

Cochlear implants

Information for families



NHS

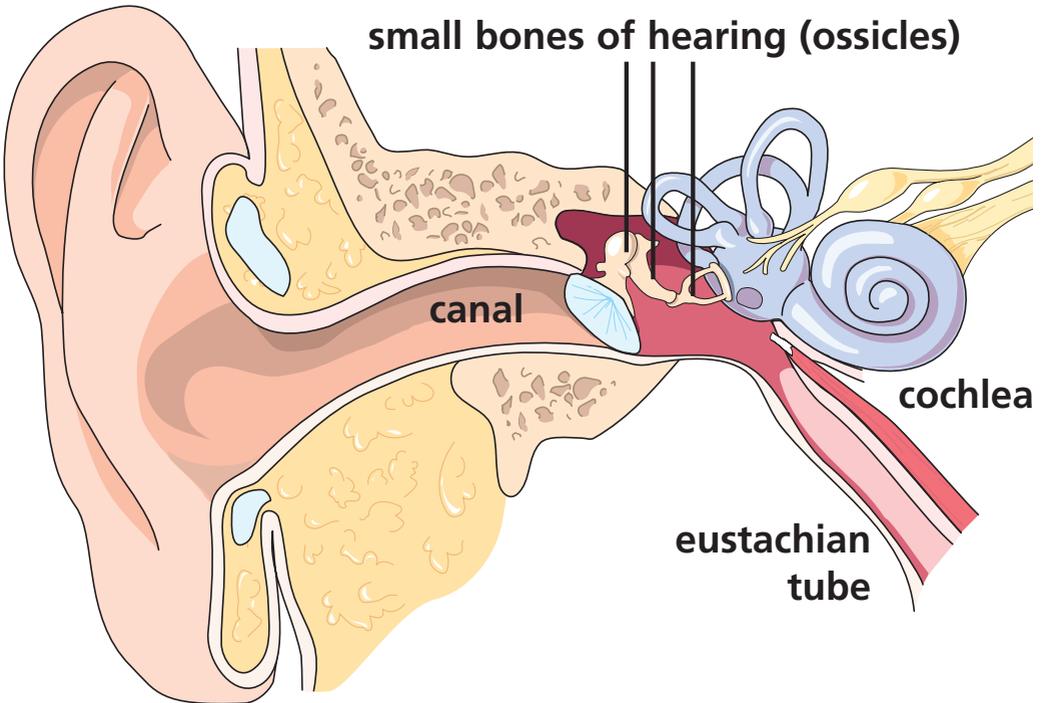
Great Ormond Street
Hospital for Children
NHS Foundation Trust



Welcome to the Cochlear Implant Programme at Great Ormond Street Hospital (GOSH)

This booklet explains cochlear implants and what to expect when your child comes to GOSH to be assessed to see whether one is suitable. It also explains the operation to fit a cochlear implant and what happens afterwards.

The Cochlear Implant Programme at GOSH is one of around 20 in the UK and to date has carried out over 1100 cochlear implants. The implant team is multidisciplinary, involving people from many different specialist areas. These people include the Consultant Audiovestibular Physician, Ear, Nose and Throat (ENT) Surgeons, Audiologists, Speech and Language Therapists, Teachers of the Deaf and Clinical Psychologists.



How do we hear?

The ear consists of three parts: the outer ear, the middle ear and the inner ear. Sound waves enter the ear canal and make the eardrum vibrate. The sound then passes through the middle ear via the three small bones of hearing, onto the inner ear that is filled with fluid.

The movement of fluid in the cochlea stimulates hair cells inside it to trigger a nerve impulse, which is carried to the brain by the auditory nerve. The brain then interprets these nerve impulses as sound.

The most common reason to have hearing loss is related to problems with the hair cells in the inner ear. This causes 'sensorineural' deafness. Although sound travels through the ear in the usual way, the hair cells are not stimulated and so no nerve impulse is sent to the brain.

Problems with the hair cells make it more difficult to hear quiet or subtle sounds, although in some cases not even very loud sounds can be heard. Some people can be fitted with hearing aids, which amplify (make louder) sounds until they can be picked up by the hair cells. However, there is a limit to the amount that this can help people hear sounds clearly. When a hearing aid does not help somebody hear well enough, then a cochlear implant may be suggested.

What is a cochlear implant?

A cochlear implant is an electronic system that stimulates the hearing nerve directly. It can give a sensation of sound to profoundly deaf children who have been unable to hear using the most powerful hearing aids available. After cochlear implants have been fitted, children can learn about the sounds around them and develop their listening and spoken language skills over time, in line with their communication potential.

There are different models and makes of cochlear implants, which share similar features. The cochlear implant system is made up of an internal part (implanted under the skin surgically) and an external part (worn on the skin surface of the head).

The external part (which typically looks like a hearing aid) is often referred to as a 'speech processor'. The microphone(s) on the speech processor picks up sound and converts it into an electrical signal. This electrical signal is passed to the antenna and sent to the receiver on the internal part of the implant, under the child's scalp. The antenna is held near to the internal receiver using magnets.

The internal part converts the signal into a tiny electrical current that is passed down an electrode that has been put in the cochlea. This stimulates the auditory nerve. In most people with hearing loss, the hearing nerve works normally, and stimulating it electrically can stimulate the brain and reproduce hearing.

This works remarkably well - indeed, older children and adults who receive cochlear implants report being able to hear some sounds as soon as they start wearing the device. Most children with a permanent severe to profound deafness can understand and develop speech and language using their cochlear implants. The quality of sound provided by cochlear implants is not as good as 'normal' hearing, and while it is usually the best available alternative for children with severe to profound hearing loss, **it takes a lot of time, patience and practice to use the cochlear implants as well as possible.**

Why does my child have a hearing loss?

There are lots of reasons that your child may have hearing loss. Children can be born with hearing loss, or hearing may get worse over time (usually gradually, but this can happen suddenly). For the majority of children, we never find the exact cause of their hearing loss.

We may suspect that the hearing loss has a genetic cause – but this does not mean we are currently able to identify the specific gene affected, or yet have an appreciation of what that gene does, other than it is associated with hearing loss. Our understanding increases each year, and hopefully in the future, we will be able to answer these questions.

The most common causes that we are able to identify are specific genetic causes – such as connexin 26 / GJB2, or children who have hearing loss as part of a pattern of other medical problems called a syndrome. The most common syndromes that we look for are Usher (which also causes visual impairment), Pendred (which can cause thyroid problems in older children), Waardenburg (which can cause changes to skin, eye and hair colour), Alport (which causes kidney problems) and Jervell-Lange-Nielson (which causes heart problems).

While the cause of hearing loss is usually due to the functioning of the inner ear, it can also be due to a problem with the hearing nerve itself, or a problem with the structure of the inner ear. Nerve problems are referred to as auditory neuropathy spectrum disorder (ANSO) and may be a structural problem (small or absent nerves

termed 'cochlear nerve deficiency'), or a functional problem (most commonly seen in children born very prematurely who develop jaundice).

Assessing the amount of hearing loss in ANSD can be difficult, as the hearing nerve can give confusing signals. There is a balance between wanting to restore hearing with an implant, and to see the potential of the ear to hear without an implant. Also, as a cochlear implant still relies on the auditory nerve to transmit hearing to the brain, implants are often not as effective in people with auditory neuropathy, and it may take longer to see the full benefit of a cochlear implant. Making these assessments depends on the specific cause of auditory neuropathy and so additional tests can be recommended in this situation.

Structural problems of the cochlea (called 'cochlear dysplasia') have lots of different types. Depending on the shape, it may be that a cochlear implant is less effective than in normally shaped cochleas. Cochlear implants may still be recommended in people with these problems, but they may not work as well, and the risk of surgery may be higher. These problems are more commonly seen in some syndromes, such as CHARGE syndrome (which causes issues in various parts of the body such as the heart and nose).

Assessing your child

The reason that your child has been referred to our cochlear implant department is because the audiologists and doctors who have seen you think that your child has a hearing loss that is not going to be adequately treated with hearing aids.

That is not to say that your child should not use hearing aids – indeed it is very important that they use their hearing aids as much as possible. Hearing aids usually give a little bit of hearing, and if you're used to wearing a hearing aid it's easier to get used to wearing a cochlear implant. However, it means that more hearing than a hearing aid can provide may be required.

All children are different, and so it is important that you and your child go through a thorough assessment so the team can give you all the information you need to make a decision. You will have the chance to talk to the team about any concerns or questions and you may have the opportunity to meet other families with children with a cochlear implant.

Members of our team will visit your child's school or nursery, if appropriate. If your child has a local speech and language therapist, we will also contact them for information. It is important to have the support of such people locally, as they will also be involved in the period after the operation when your child is learning to use the cochlear implant. Your child will be helped to get the maximum benefit from their cochlear implant if you, they, the medical team and other people involved with them, for example at school, all work together.

Your child will need various tests and checks, which take place over several appointments, usually during one assessment 'week'. These include:

- **General health check** – your child's general health will be checked by the Consultant Audiological Physician (doctor who specialises in hearing).
- **Ear check** – they or the Audiologist will also check your child's middle ear regularly. If your child has glue ear this may affect their hearing and an operation to insert grommets to correct this may be suggested.
- **Hearing tests** – your child will need tests to check how much they can hear with and without hearing aids. The team may also want to try different models of hearing aids or new ear moulds. This will include behavioural tests where your child responds to sound. Your child may also need a test called an Auditory Brainstem Response (ABR). This is a test that measures the response of the hearing nerve to sounds, and it may mean your child needs sedation or a general anaesthetic. Some of these tests may have been done before you come to see us, and may or may not need to be repeated.
- **Scans of the ears** – these scans look particularly at the shape of the cochlea, and also the size of the **hearing nerve. These scans may mean your child needs sedation or a general anaesthetic.**
- **Vestibular balance and vision tests** – your child will also receive these tests as your hearing and balance systems are linked.

- **Communication skills** – the team will assess your child's communication. The more we know about your child's skills, personality and interests, the better we are able to help with the learning process afterwards.
- **Psychological assessment** – all children are routinely assessed by a Clinical Psychologist, who will find out about your child's general development and learning style. They will also discuss any concerns about your child's behaviour and emotional well-being.
- **Further tests** – additional tests may be required to look at the cause of hearing loss

Information about many of these tests is available on our website at www.gosh.nhs.uk/conditions-and-treatments/procedures-and-treatments

In addition to these tests, the team will use a questionnaire to find out about your child's ability to hear sounds at home and, if appropriate, at school. It is important that your child is a good hearing aid user when they come for the assessment, even if you are unsure about what benefit they are getting from the hearing aids.

If you agree, some of these tests will be recorded on video, so the team can refer back to them later on in the process. We will ask you to sign a consent form before the assessments start.

When we have the results of all these tests, the team will meet to decide whether to offer your child cochlear implants. You can meet with the Consultant Audiovestibular Physician to discuss the outcome of the assessment, and also an ENT surgeon.

The operation

A cochlear implant is a routine operation that is undertaken thousands of times every year in the UK. At GOSH, we insert about 80 implants every year.

What happens before the operation?

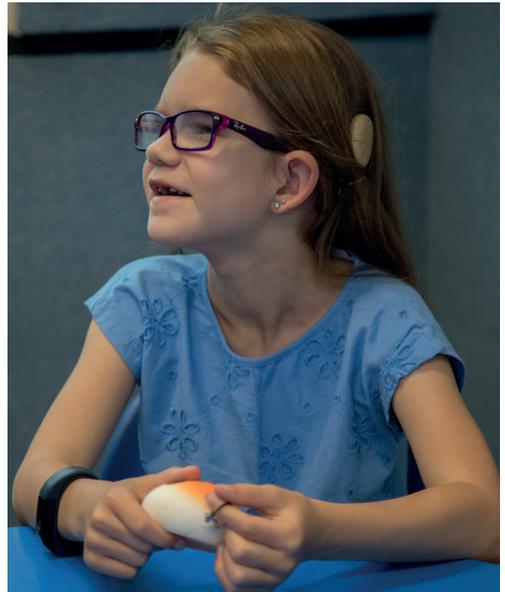
You will have met an ENT surgeon and discussed the operation. You will be seen in the pre-assessment clinic and the nurses there will ask questions about your child's general health. You may have to return to meet an anaesthetist. They will give you more information about having a general anaesthetic and pain relief after surgery.

If you have any particular concerns about coming into hospital, or operations in general, please ask the ENT surgeon, anaesthetist or pre-assessment nurse. If anything specific needs to be done to make it safe for your child to have an operation, the pre-assessment nurses will arrange it. You will receive information about how to prepare your child for the operation in your admission letter.

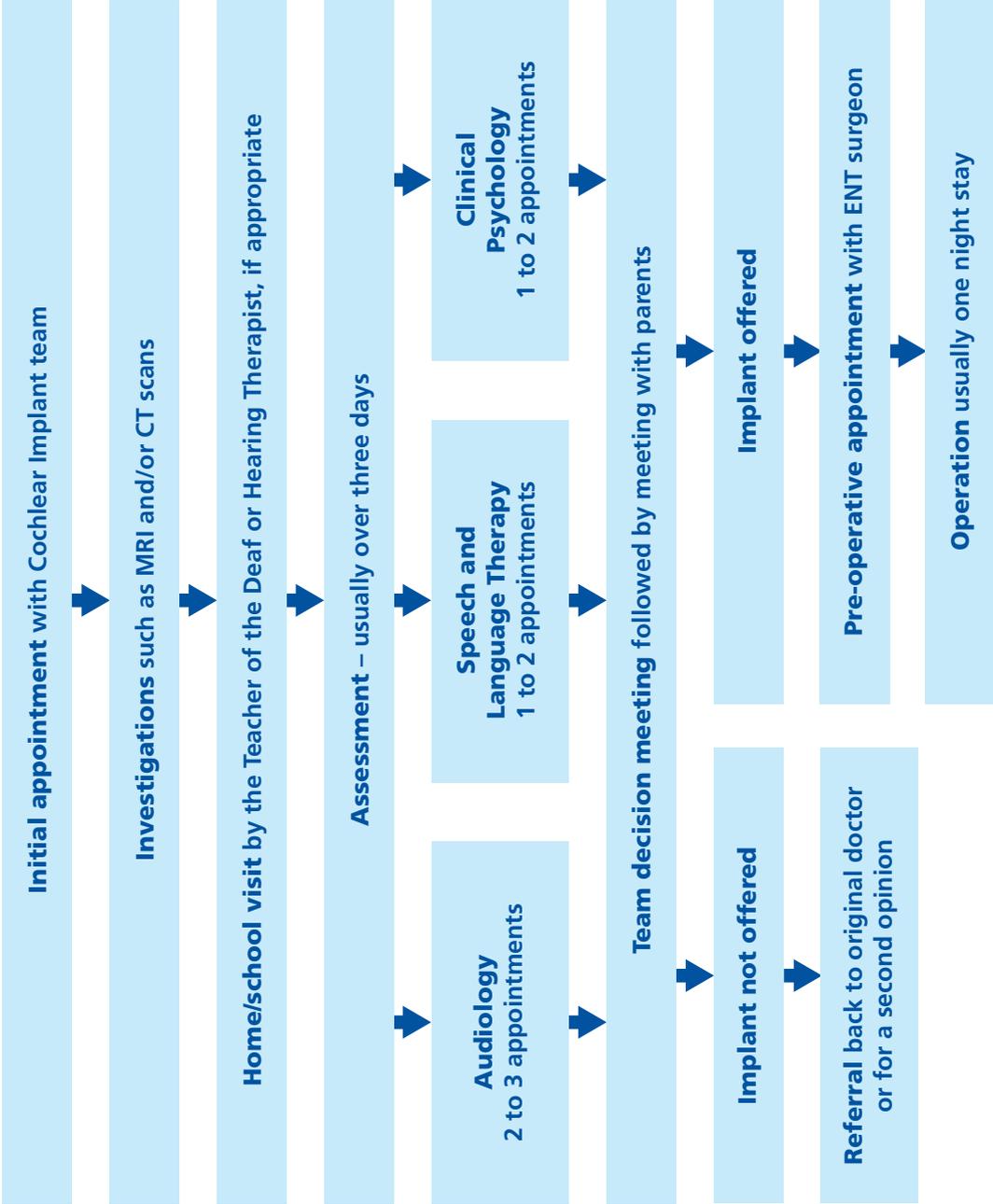
Most children come to hospital on the day of your operation. Your child will not be allowed to eat or drink anything for a few hours before surgery. Usually, we will call you the day before the operation to tell you what time to come for your operation and the time after which your child should not eat or drink. Please note, that the time of your operation is variable – we carry out a number of operations each day, and we cannot always predict how long an operation will take – every operation will take as long as is required to be completed safely.

The person bringing your child for the operation should have 'Parental Responsibility' for them. Parental Responsibility refers to the individual who has legal rights, responsibilities, duties, power and authority to make decisions for a child. If the person bringing your child does not have Parental Responsibility, we may have to cancel the operation.

The doctors will discuss any worries you may have and ask you to give permission for the surgery by signing a consent form. Another doctor will also visit you to explain about the anaesthetic. If your child has any medical problems, particularly allergies, please tell the doctors about these. Please also bring in any medicines your child is currently taking.



Child care pathway within the Cochlear Implant Programme



Switch on appointment (x2) about 2-4 weeks after operation

Mapping appointments

Map 1 at 1 week later
Map 2: 2 weeks later
Map 3: 4 weeks later

Speech and Language Therapy appointments

Up to monthly therapy sessions (at GOSH or with prearranged telephone review) in the period following Map 2

Review appointments

At 3, 6, 9, 15, 18 and 21 months after switch on

Assessment reviews

Speech and language therapy reviews annually until age of 5 years. Audiology reviews annually until age of 16 to 18 years. Transfer to adult Cochlear Implant Programme. Psychology, Teacher of the Deaf and SLT follow-up as appropriate

School/home visits

As appropriate by Speech and Language Therapists and Teachers of the Deaf



What does the operation involve?

The operation usually takes about two hours per cochlear implant and is carried out when your child is under general anaesthetic. A small amount of hair behind the ear will be shaved while your child is under anaesthetic, and there will be a cut approximately 5cm long behind the ear. This will leave a small scar, but this is hidden behind the ear, and after time it is unusual for this to be noticeable.

In the first half of the operation, the surgeon drills through the bone behind the ear. This involves drilling between the nerve that moves the face and a taste nerve, into the middle ear and the cochlea. Small needles are placed in the face to warn the surgeon if they come close to the nerve that moves the face.

In the second half of the operation, the implant is put into a 'bed' behind the ear, and the electrode array is inserted into the cochlea as gently as possible. The surgeon will then use dissolvable stitches to close the incision. Audiologists may perform some initial testing while your child is still in the operating theatre. This testing confirms that the electrodes are working and can help them make an initial program for your child's implant.

Are there any risks?

Cochlear implantation is a routine surgical operation and complications are unusual, however, they can occur, and it is important you are aware of them. If you have any questions about them, please ask your surgeon.

Drilling close to the nerve that moves the face means there is a very small risk of injury to the nerve. This would cause weakness to movement of the face which has a number of implications. It is very rare and when it does occur, it is usually temporary, although recovery can take many months. Injury to the taste nerve can cause an unusual taste in the mouth that usually settles after six to eight weeks.

When the surgeon inserts the implant they will be as gentle as possible to preserve any small amount of remaining hearing, although this cannot be guaranteed. This can be useful to hear fire alarms or similar sounds when not wearing the implant. Residual hearing also helps get the most out of an implant as it gives context to sounds you hear. We also want to be gentle to avoid injury to the balance organ. This can make your child feel dizzy and affect their balance, although this is almost always temporary. The operation can also cause a ringing sound in the ear, although this usually stops on its own, it can take some time. This can also stop when your child starts using the implant. In some cases, the implant may not be fully inserted, and although this may not have any noticeable effects, it might mean that your child gets less hearing.

Bleeding or infection are unusual, but they may occur, and can mean that another operation is needed. We give a dose of antibiotics at the time of the operation to reduce this risk. The anaesthetist will discuss any particular risks with general anaesthetic your child may have, although general anaesthetics are very safe, and the anaesthetist you see will be very experienced. After the operation, it is not unusual to have a sore throat or some vomiting as a consequence of the anaesthetic.

If your child has two implants, the surgeon will try to get the devices as symmetrical as possible on each side of your child's head. The implants inserted are electrical devices, and as such, they have a life-expectancy. We would expect an implant to last for 20 to 30 years, although they may last much longer, or equally they may stop working after a much shorter period. Injuries to the head from falling over increase this risk by a very small amount, and so implants in children tend to have a slightly shorter life expectancy. If an implant fails, it can be replaced.

In a small proportion of people, there is a connection between the fluid in the cochlea and the fluid in the inside of the head. Sometimes this means that when a cochlear implant is inserted, this fluid can then leak out into the ear. If this happens, it has to be repaired, as this increases the risk of meningitis. If this does not happen, but there is still a connection, then there is still a very small risk of meningitis. To reduce this very small risk, we recommend vaccination which your child may already have had.

What happens after the operation?

After the operation, your child will return to the ward to wake up fully from the anaesthetic. Your child will have a large bandage around their head. Once your child feels comfortable and has had a drink they will be able to get up and play.

Children generally spend one night in hospital following surgery. During the morning after the surgery, your child will have the bandage removed followed by an x-ray to document the position of the electrode. Afterwards, they will usually be able to return home.

After the operation, it is important to keep the ear and wound dry for ten days to stop the wound from becoming infected. If you have concerns during this period, such as a swelling behind the ear, please contact the cochlear implant department (in hours), or otherwise the ward (out of hours).

Your child will need simple painkillers such as paracetamol for the first few days – particularly before going to bed to make sure your child gets as much rest as possible. After the operation, you may be able to feel the cochlear implant under the skin, this will be more obvious in smaller children. Your child may also experience some bloody discharge from the ear for several days. They need to be off school for two weeks and avoid sports and rough play.

Learning to use the cochlear implant

The first 'switch on' appointment usually takes place around two to three weeks after surgery and will then involve frequent visits to the hospital on a regular basis. This is so that the settings on the implant can be altered as it settles into the cochlea and as your child gets more used to detecting sounds. An ENT doctor will attend the first appointment to check that your child's scar is healing well.

The Audiologist will start by connecting the speech processor to a computer and activating the implant gradually to work out your child's level of hearing. They will also program the processor so that the volume of sound your child can hear is comfortable and not too loud.

This process is known as 'mapping'. The Audiologists tend to be cautious, and set the levels of stimulation low to begin with, so that your child is not overwhelmed by sound. The level is gradually increased as your child becomes accustomed to the sound from the implant.

The 'switch on' process takes place on two separate sessions in the cochlear implant department. Following switch on, regular mapping appointments are required to ensure that your child's speech processor remains programmed to the best level.

It takes time to get used to electrical hearing. Adults with implants say voices sound electronic, or like a cartoon character talking, when they first use the implant. However, over time, typically a few months, users will become used to listening until voices sound normal. This learning process involves a collaboration between your child, the team at GOSH, your family and your child's Teachers and Speech and Language Therapist.

Members of the team will regularly liaise with your child's local team to discuss progress and plan next steps. They will also offer you and your child intervention sessions at GOSH as a supplement to any therapy your child may be receiving locally. The local Speech and Language Therapist and/or Teacher of the Deaf will be invited to attend one of these sessions to plan the process.

It is important that your child learns to wear the speech processor all their waking hours. The sooner they learn to enjoy improved access to sound, the sooner they will start to make progress with listening.

Children can respond at different rates to the sound signal they are receiving. The sound that the implant produces through electrical stimulation is very different from the sound produced from a hearing aid. Therapy following implantation aims to help your child understand the new sounds they hear. It also aims to encourage your child's listening and spoken language development through carefully graded steps.

The team will monitor your child's listening, language and communication skills over time. For the first five years after implant, you will be seen by both the Speech and Language Therapy and Audiology teams to monitor your child's progress with language and listening. After this, your child will continue to be monitored annually by the Audiology team until they transfer to adult services.

If you or any professional working with your child have concerns about your child's progress, appointments may be arranged with an appropriate member of cochlear implant team at any stage. Intervention can be offered as needed from the Clinical Psychologists, Teachers of the Deaf and Speech and Language Therapists.

Outcomes and long term implications

Cochlear implants are immensely effective at rehabilitating the hearing of people with low levels of hearing, and people with cochlear implants can usually hear very well, and go to school, get jobs, and undertake daily activities entirely normally. In adults whose hearing loss means that even with hearing aids they are typically only able to identify less than half the words spoken at a set volume, cochlear implants will typically enable them to hear over 80 or 90 per cent of words at the same volume. In children who, without an implant, would never have developed speech, some will develop speech more quickly than children without hearing problems. They just need to wear their implants to hear like people need to wear hearing aids to hear.

However, each child is an individual, and the amount a cochlear implant can do for them does vary. The vast majority of people benefit hugely from an implant, but a small proportion (around two to four per cent) of people find that they cannot get used to electrical hearing, and decide not to wear their implant much, or at all. Also, we know that people who use cochlear implants tend not to like music as much as people with normal hearing.

There are factors that we know make it more likely that your child will do really well with a cochlear implant. We know that in children who are born with very little hearing, that the younger they are they receive their implant (particularly if they are less than one year old), the more

likely they are to benefit. Children without additional difficulties typically do well with learning to listen with cochlear implants. In those children and adults that develop progressive hearing loss, we know that the shorter the length of time they struggle with hearing, the more an implant will help them. We know that support during rehabilitation is very important.

We know that some forms of hearing loss, like some specific genetic hearing losses, respond particularly well to cochlear implants. There are surgical factors that are important too – fully inserting the cochlear implant helps, as does keeping any remaining ‘acoustic’ hearing mechanisms. Despite everything we know about who will do best with a cochlear implant, there is still much we don’t know – indeed the main factors (such as the age at implantation and amount of hearing present) only account for about a third of the variation that is seen in outcomes.

People with cochlear implants can swim. There are swimming accessories to put the processor in, or otherwise, the processor can be removed and they can swim without hearing. We do, however, advise against scuba diving at any stage having had a cochlear implant. Also, for sports which require protective headwear, such as boxing or horse riding, you may need to make specific measures.

If your child needs an MRI scan after having an implant, this can be an issue. The magnet in the cochlear implant will be pulled on by the magnet in the MRI scanner. Although a scan is almost always possible, there are some important considerations. Your child may have to have a bandage to wrap around their head to hold the magnet in place. The scan may be painful, and there have been cases where the implant has had to be replaced as the internal magnet has moved out of position. The quality of the image that the scan produces may also be affected by the implant (particularly the images of the head). Your child may have a device that is compatible with MRI scans, which makes things more straightforward. Having an implant may also cause difficulties with other medical treatments, such as radiotherapy or electrosurgery, such as gamma knife surgery.

Which type of cochlear implant will my child have, and should they have one or two?

There are a number of hearing benefits to having two cochlear implants. Two implants also help listening to speech in noisy environments – particularly when someone is talking to your child from one side, rather than straight ahead. Having two implants is also useful should an implant have reliability issues, as your child would then have a working hearing ear to rely upon while the implant is fixed. Spatial hearing and localisation of sound is also important – and being able to hear from both ears lets your child orientate themselves in the world around them much more effectively. We know that people with deafness on one side have worse visuospatial memory for example, working out from which direction a sound is originating.

However, having two implants commits your child to cochlear implantation on a lifelong basis, and having two implants does involve operations on both ears, and their associated risks. When your child has two implants, they should always use their left processor on their left implant, and their right processor on their right. This is not a safety issue, but will help your child get the most hearing, and avoid hearing uncomfortably loud sounds. In most cases, the speech processors will only work when connected to the correct implant.



Overall we usually recommend implants in both ears, as we feel the additional benefits are worth the additional risks. This view is also held by the National Institute of Health and Care Excellence (NICE) who fund bilateral implants in children (but not in adults). Children who have previously received one implant may have another, however the funding does not clearly allow for people to choose to have one and then subsequently a second (although this may medically be needed).

If there are specific surgical or audiological reasons to have a particular type of implant, we will let you know which implant is recommended. Sometimes, there will be a specific reason to have one device – for example if your child has a brother or sister with a cochlear implant, it is advisable to have the same manufacturer.

If there are no specific reasons, we will discuss the available devices with you. There are four cochlear implant manufacturers currently in use throughout the UK. These are:

- Advanced Bionics
- Cochlear
- Med-El
- Oticon

All of the devices have their own strengths, however the differences between any two devices are tiny compared to the differences between a cochlear implant and a hearing aid. All companies invest large amounts in developing their products. At any one time, one manufacturer may be ahead in one area (such as the size of their processor, or the MRI compatibility of their magnet, or any one of a hundred other areas), but over subsequent years, other manufacturers will catch up in those areas, and get ahead in other areas. The external processor will be upgraded by the implant department every few years. Although the internal package is not straightforward to replace, it is almost always compatible with new technology in the external processor.

Reliability and device failure are important considerations. All manufacturers publish reliability reports, which are freely available on the internet. Some new devices may be found to have higher rates of device failure as companies refine and develop their innovations – there is a limit to the amount that testing their devices in laboratories can show. When such an issue is identified after some years of a device being in use, that device would not be inserted any more, and devices already inserted can be replaced if they stop working. Examples of this in the companies we use regularly would include the Advanced Bionics Ultra V1 failures and the Cochlear N5 failures. There is therefore a balance between wanting the latest, newest device, with the most up to date technology, and wanting a device that has been used for many years without problems.

Magnet type is also a consideration. Advanced Bionics, Med-El and Cochlear all offer MRI compatible magnets, which can be very helpful. However, these magnets are less powerful than conventional magnets, and although this is not usually a problem, this may mean the antenna is more likely to become dislodged. Ways to deal with this, if it does occur include cutting hair over this area, using sticky tape, or wearing a head band. Under exceptional circumstances, the magnet can be replaced, although this carries small risks.

If you need more advice from the Cochlear Implant Department at GOSH, please telephone 020 7813 8316 from Monday to Friday, 9am to 5pm or email cochlear@gosh.nhs.uk.

You can get further advice, and contact with people in similar situations, from:

■ **British Cochlear Implant Group**

Tel: 0121 204 3830

E-mail: info@bcig.org.uk

Website: www.bcig.org.uk

■ **Cochlear Implant Children's Support Group**

Website: www.cicsgroup.org.uk

■ **Ear Foundation**

Tel: 0115 942 1985

Website: www.earfoundation.org.uk

■ **National Deaf Children's Society (NDCS)**

Information line: 0808 800 8880

Voicetext open 10am – 5pm Monday – Friday

Email: helpline@ndcs.org.uk

Website: www.ndcs.org.uk

■ **The Royal National Institute for Deaf People (RNID):**

Information line: 0808 808 0123

Textphone: 0808 808 9000

Email: Informationline@rnid.org.uk



We hope you have found this introductory booklet helpful. If you have any questions or require further information, please contact the team.



NHS

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Compiled by the Cochlear Implant department
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