Information
For families

Children with microtia and atresia

We use the term 'deaf' to refer to all types of hearing loss from mild to profound. This includes deafness in one ear or temporary hearing loss such as glue ear.
We use the term 'parent' to refer to all parents and carers of children.

Introduction

This factsheet is for families who have a child diagnosed with microtia or atresia. It covers:

- what microtia and atresia are
- different types and causes of deafness that may be related to microtia or atresia
- how microtia, atresia and any associated deafness can be managed
- other medical syndromes that have microtia or atresia as part of them
- organisations which can support children with microtia or atresia and their families.

What are microtia and atresia?

Microtia is a term used to describe underdevelopment of the outer ear (pinna). It can vary from quite minor changes (such as the ear being smaller than expected) to 'classic microtia' where the pinna is missing.

Classic microtia is often associated with absence of the ear canal – known as atresia. This is because the baby's outer ear and ear canal develop together during pregnancy. In some cases, the ear canal can look present from outside but ends at a 'blind alley' inside.

Types of microtia

Lobular type microtia: the outer ear is present but small and underdeveloped (peanut shaped). This is the most common type of microtia.

Conchal type microtia: the child has an ear canal although it may be very narrow (canal stenosis) or a blind ending with no eardrum. The outer ear is present and has a conchal bowl (middle part of the ear cartilage), but it is much smaller than normal. The upper part is often underdeveloped.

Small conchal type: the ear is smaller than usual but the key features of the outer ear are present, although they may have small differences in shape or form, such as a small conchal bowl. The ear canal is either missing (atresia) or has a narrow and blind ending.

Microtia happens more often in boys than in girls.[1, 2] It usually affects one side (unilateral microtia), often the right ear.[2] About one in 10 children are affected on both sides. This is called bilateral microtia.[2]

Microtia is known to affect about one baby in every 6,000 births.[2] Around one in 10 children who have microtia or atresia will have another family member with the same condition.[3]

Most children with microtia have no other medical problems. However, microtia can also be one sign of a syndrome. 'Syndrome' means a collection of signs or symptoms that commonly occur together and that doctors recognise as being related. Microtia is known to occur as part of several different syndromes. You can find more information on these syndromes at the end of this factsheet.

What causes microtia and atresia?

Microtia or atresia happens when the outer part of the ear does not develop fully during the early stages of pregnancy. The exact reasons for isolated (or non-syndromic) microtia or atresia remain unclear although it's usually a random, one-off event. It's not caused by anything the mother did wrong before or during the pregnancy.

Medical research has suggested that sometimes certain prescribed medications taken during pregnancy, genetic and/or environmental factors may be involved. Associated factors will be explored at the first meeting with your specialist multidisciplinary team (healthcare professionals with specialised skills and expertise).

My child has microtia – what happens now?

Audiology service

Your child will be referred to your local audiology service for hearing tests. This will normally be done routinely shortly after birth. If not, you can ask your GP, health visitor or paediatrician to refer you. Your local audiology team will regularly assess your child's hearing and is responsible for fitting and maintaining hearing aids that may be used in the management of any deafness.

Specialist microtia/atresia clinic

You should also be referred to a specialist microtia/atresia clinic. There are several clinics around the country and your local audiology team will liaise closely with them. In the specialist clinic you will meet members of the multidisciplinary team. The team supporting your child may seem quite large and daunting at first but they will explain their different roles in your child's care when you meet them.

Team members include:

- a plastic surgeon
- an ear, nose and throat (ENT) surgeon
- an audiovestibular (hearing and balance) physician or paediatrician
- audiologists
- a clinical psychologist
- specialist nurses.

The team will also have access to a prosthetist, geneticist, speech and language therapist, craniofacial (bones of the skull and face) team and other professionals who may be involved in the overall care of your child.

The specialist team will usually meet with you soon after birth. At this stage, the priority is to encourage your baby's listening and communication development with support for any deafness. The specialist team will liaise with your local audiology department to make sure your child's hearing is assessed and hearing aids are fitted if appropriate. During this first early consultation with the multidisciplinary team, they will also discuss and alleviate any concerns you may have and advise about the options available in the future.

Having a new baby with a visible difference can be a shock for some parents. At this stage, you may have questions about changing the cosmetic appearance of the ears. The team will be able to answer your questions, share photos of other children and adults they have worked with, and may use computer-aided software to show expected outcomes of ear reconstruction surgery. However, surgery is not usually done until a child is at least 10 years old. By this time, many children and parents decide that they do not want reconstruction of the ear and are happy with the child's appearance.

Medical tests

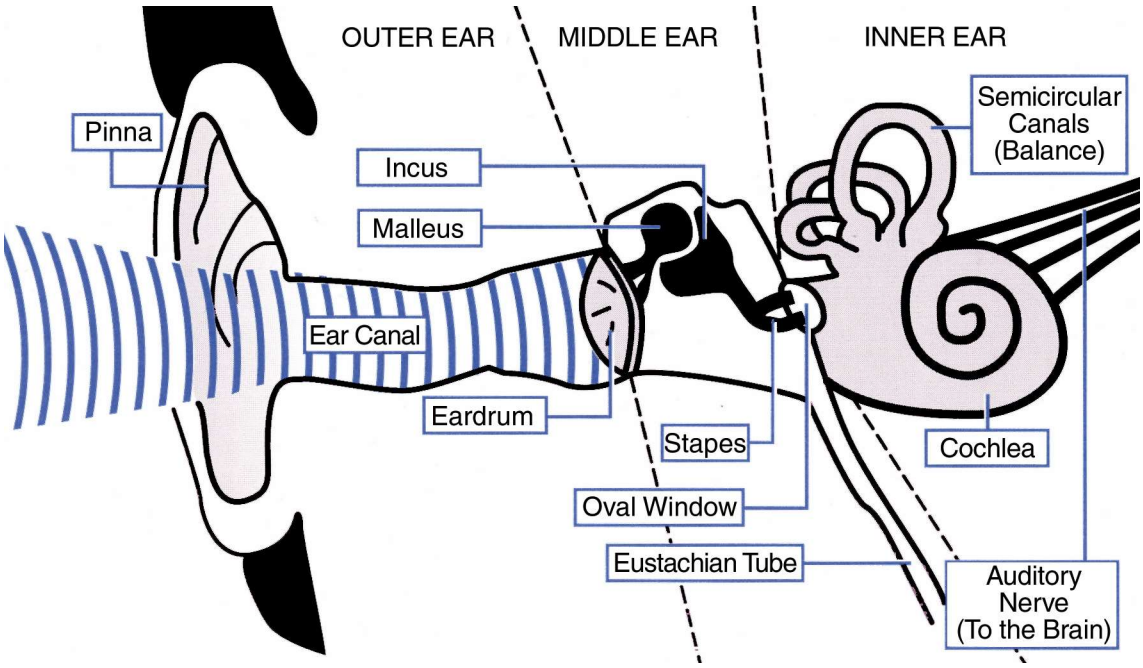
Because microtia can occur as part of several different syndromes that may involve other organs, you may be offered a range of medical tests for your child, either by your local audiology department or the specialist team. These will include blood and urine tests, renal (kidney) ultrasound and a CT scan of the inner ear. The tests are used to help your doctor assess both the structure and function of these organs and the inner ear. They may help rule out other medical conditions and help to show the best way to manage your child's microtia and any deafness. Your doctor will explain when they feel the best timing for these tests are.

You can find out more about these medical tests in our resource, **'Understanding your child's hearing tests.'** Download or order a copy of the resource at **ndcs.org.uk/understand**.

The specialist team will keep your child under regular review throughout their childhood. They will work closely with local audiology teams. Your team will again discuss options for reconstructive procedures as your child grows up and procedures develop. As they get older, your child will become more involved in discussions and decisions about their own care.

Types of deafness that may be associated with microtia and atresia

There are different types of deafness – conductive, sensorineural or mixed – that can be associated with microtia and atresia, depending on which part of the ear is affected.



Conductive deafness is when sound cannot pass efficiently through the outer and middle ear to the inner ear (cochlea and auditory nerve), such as when there is atresia of the ear canal.

Microtia can cause conductive deafness. Microtia and atresia may be associated with variations in the structure of the middle ear that will affect the level of deafness experienced. Some of the variations encountered include:

- underdevelopment of the middle ear bones (malleus, incus and stapes – collectively known as the ossicles)
- other middle ear differences
- incomplete development of the bone surrounding the ear (mastoid bone) leading to poor air circulation in the middle ear.

Sensorineural deafness is caused by a fault in the inner ear or the auditory nerve (the nerve that carries the electrical signals from the cochlea to the brain). Sensorineural deafness is unusual in children with microtia and may indicate a co-existing condition or associated syndrome.

Mixed deafness is when children have both conductive deafness and sensorineural deafness.

Unilateral deafness describes deafness that affects one ear. This is often associated with unilateral (single-sided) microtia and atresia. Unilateral deafness is often referred to as 'one-sided hearing loss' or 'single-sided deafness'. One ear has a normal level of hearing and most children with unilateral deafness manage very well in most situations.

Many children with unilateral deafness develop clear speech. However, there are situations where children will find it more difficult to hear and may need support to:

- hear sounds or speech on the side with the deafness
- identify the source of a sound or the direction a sound is coming from
- understand speech when there is background noise.

For more information, visit **ndcs.org.uk/unilateral**.

Glue ear is a common cause of temporary conductive deafness in childhood. Glue ear is a build-up of sticky fluid in the middle ear. Because it is so common, children with unilateral microtia may be affected by glue ear in their fully developed ear at some point and this may need to be managed by the specialist team.

For more information, see our resource **'Glue ear: A guide for families'** or visit our webpage, **ndcs.org.uk/glue-ear**.

Hearing tests

Because children with unilateral microtia are relying on the hearing in their fully developed ear, they should have regular hearing tests to monitor their hearing while they're young. Depending on the level of deafness and whether hearing aids are worn, this might be every three to six months for young children and every year for older children. If you're worried about your child's hearing or feel that their hearing has changed, ask for an earlier appointment.

It's possible to test the hearing of all children from birth. There are two different types of test that can be carried out to find out about your child's hearing – objective tests and behavioural tests. Responses to hearing tests depend on how the ear and its nerve connections are working, as well as the stage of general development a child has reached. Usually, several different tests need to be done over a period of time to be able to build up an accurate picture of your child's hearing.

For more information on deafness and hearing tests see our resource, **'Understanding your child's hearing tests.'** Download or order a copy of the resource at **ndcs.org.uk/understand**.

Managing deafness caused by microtia and atresia

If your child has a hearing loss, your local audiology and education services will give you and your child support. Your audiologist will refer you to a Teacher of the Deaf (ToD), who'll be able to give you advice on encouraging good communication, using hearing aids, schooling and who is responsible for making sure your child has any support they need in school. You may also be offered an appointment with a speech and language therapist. There may be specialist equipment you can use to support your child's hearing and listening. The options available depend on your child's type and level of deafness.

Hearing aids

Hearing aids can be useful for children with any level of deafness. The type of hearing aid that will be suitable for your child will depend on the type and level of deafness they have and the type of microtia present. Hearing aids work by amplifying (making louder) sounds going into the ear. Hearing aids come in a range of styles. Good quality digital hearing aids are available free of charge for all children on the NHS. Most children use behind-the-ear hearing aids.

The hearing aid sits on the top of the pinna (the outside part of the ear) and is connected to an earmould that is specially made to fit in the child's ear. However, children with microtia or atresia may find it hard to wear behind-the-ear hearing aids. For example:

- a very small pinna can make it difficult to keep a behind-the-ear hearing aid in place
- smaller ear canals can sometimes make it difficult to get well-fitting earmoulds.
- narrow ear canals can cause sound from the hearing aid to 'bounce' back off the wall of the ear canal, causing whistling or 'feedback' from the hearing aids.

If your child has any of these problems, talk to their audiologist about possible solutions. Fitting hearing aids in children with microtia can be challenging and they may need to see a specialist audiologist.

Every hearing aid is programmed for the ear it will be worn in. When the hearing aids are fitted, you'll be shown how to use them effectively, as well as how to clean and maintain them and how to change the batteries. You should be given written information to take home. Your audiologist and ToD will discuss when the best times are to use the hearing aids.

For more information about hearing aids, see our resource **'Hearing aids: A guide for families'** or visit our webpage **ndcs.org.uk/hearingaids**.

Bone-conduction hearing aids and implantable hearing devices

Bone-conduction hearing aids

Children with absent or very underdeveloped outer ears (pinna), or absent ear canals, will not be able to use a conventional behind-the-ear hearing aid and may benefit from a different type of hearing device that allows the child to hear sounds using bone-conduction. This is called a bone-conduction hearing aid, sometimes known as a BAHA.

Bone-conduction hearing aids can be worn on metal or soft fabric headbands. They use a vibrating pad that allows sound to be conducted through the bone rather than through the middle ear. The vibrator is worn behind the ear, resting on the mastoid bone (part of the skull behind the ear).

Bone-conduction hearing implants

Children who find a bone-conduction hearing aid effective may be suitable for a bone-conduction hearing implant (BCHI). A bone-conduction hearing implant is designed for people who have a functioning cochlea, but the middle or outer part of the ear prevents the information reaching the cochlea in the usual way. It consists of a sound processor that is held on the head behind the ear. The processor might be attached in one of two ways.

- The processor can be clipped to a fixture called an abutment, which is a small titanium screw that has been implanted in the skull just behind the ear. This is known as a bone-anchored hearing device or system.
- The processor can be attached with a magnet.

BCHIs conduct sound through the bone rather than through the ear canal and middle ear. This allows sound waves to be transmitted directly to the cochlea in the inner ear. This procedure is usually offered for children over the age of four.

In young children, the sound processor of a bone-anchored hearing system may be worn on a soft headband, similar to a bone-conduction hearing aid. The soft headband is taken on and off and can be used permanently or temporarily, during the assessment stage.

For more information on bone-conduction and bone-anchored hearing aids, visit **ndcs.org.uk/boneconduction**.

Middle ear implants

A small number of children and young people with microtia may be offered a middle ear implant. Middle ear implants may be suitable for some children who cannot use conventional hearing aids and have mild to severe sensorineural hearing loss, or conductive or mixed hearing loss. The implant in this case works by converting sound into mechanical vibrations. This mechanical energy directly stimulates the middle ear bones.

Your audiology team will discuss the various options with you to help find the one most suitable for your child. If your child is likely to have cosmetic surgery or prosthetic ears fitted later, it's important that their surgeon is aware of this when deciding the exact position of the device.

For more information about middle ear implants, visit **ndcs.org.uk/middle-ear-implants.**

Surgery

Your child may be offered surgery to improve either the hearing and/or the appearance of their ear. Surgery for microtia is not essential, and lots of children and their families choose not to undergo any surgical reconstruction.

Whether or not surgery can be done to improve a child's hearing depends on whether the middle and inner ear are present and developed. This type of surgery is not normally carried out until children are about nine years old. This is because young children are likely to develop bony regrowth, but this risk reduces as the child gets older. These future options will be discussed with you at your first consultation with the multidisciplinary team.

There are always risks with surgery. You should discuss any operation with your doctor and make sure you understand the expected outcome of the surgery before you agree to go ahead. As far as possible, children should be fully involved in any discussions about surgical options and be able to give their opinion before consent is given to any procedure. Some families will decide not to have any kind of surgery or may decide to let their child decide when they're older.

Many older children and adults with microtia are comfortable with their 'little ear' and don't feel a need to have surgery to change this. However, if the child and family decide that surgery is the best option for them, there are three options to surgically improve the appearance of the ear. These are:

- reconstruction with autogenous cartilage
- reconstruction with an artificial framework
- prosthesis (artificial ear).

Reconstruction with autogenous cartilage

Autogenous reconstructive surgery involves having an ear built from the child's own rib cartilage and body tissues. Because the ear is sculpted from the child's own tissues, it is alive and grows with the child. It is believed that ears constructed in this way are likely to last a lifetime.

This type of surgery is not normally done until a child is nine or ten years old, to allow time for sufficient rib cartilage to have developed. Leaving surgery until children are a little older also means that they can be involved in discussion and consent to the procedure, and that surgical aftercare is easier.

Sculpting the rib cartilage is an art and technically challenging to do within the operation time. Making sure it looks the same as the opposite ear is another challenge. This type of reconstruction may take two or more operations to perform and is done by a specialist surgeon within the multidisciplinary microtia team.

Reconstruction with an artificial framework

The ear is built from the child's own tissue and skin around a plastic framework. This type of reconstruction can be carried out on children from the age of three. However, the ear will not grow with the child, so careful consideration needs to be given to the expected size of the opposite ear in the future. There are also risks associated with possible extrusion (where the framework is pushed out through the skin) or infection following any future trauma or surgery to the ear.

Prosthesis (artificial ear)

As the results of autogenous ear reconstruction improve, it has become less usual to choose prosthesis unless there is no other option. However, some families will choose prosthetic ears over reconstructive surgery. Sometimes reconstructive surgery may not be possible, for example if the tissues or blood supply at the site of the missing ear have been very badly damaged, either by trauma, disease or by previous surgery. Occasionally a prosthetic ear may be considered following reconstructive surgery that was not as successful as hoped. Prostheses are made in soft, durable silicone. They're cast from impressions taken of the other ear and hand-coloured to closely match skin tone.

Prosthetic or false ears are attached to titanium fixtures that have been implanted in the bone on the side of the head during two separate operations. New bone forms around the titanium implant, known as osseointegration, in the same way as with bone-anchored hearing aid surgery. The prosthetic ear is attached using either clips or magnets.

Surgery to implant the titanium fixtures cannot be done until the skull bone has developed sufficient thickness (usually after the age of four). The prosthetic ear will need to be replaced fairly regularly. It can be removed at night for sleeping. It's important to maintain good hygiene of the skin around the fixtures with careful cleaning to avoid infection.

Deciding on the right option

If you and your child are considering surgery for their microtia or atresia, you'll need to discuss all the options with your surgeon. Here are some questions you might like to consider.

- What are the pros and cons of each type of procedure?
- How many operations of this type have you performed before?
- Can we see photos of outcomes for other children?
- How often is further or revision surgery necessary?
- What is the timing of other procedures that are likely in the future, for example atresia (ear canal) surgery?
- What risks are associated with the surgery?
- Will there be any restrictions on the child's activities following the procedure?
- How long will the hospital stay be?
- How long is the recovery period?
- What aftercare is necessary?

If your child is going to have bone conduction or middle ear implant surgery and is likely to have cosmetic surgery in the future, it's very important that the exact placement of the device abutment is discussed with your surgeon so that it does not prevent reconstructive surgery or proper placement of prosthetic ears.

Syndromes and other medical conditions that are associated with microtia

Most children with microtia have 'isolated' or non-syndromic microtia, which means that it isn't associated with any other medical conditions. However, microtia can also happen as one sign of a syndrome. 'Syndrome' means a collection of signs or symptoms that commonly occur together and that doctors recognise as being related. Listed below are some of the syndromes known to include microtia. They are listed with the most common first.

Treacher Collins syndrome (also known as mandibulofacial dysostosis) is a genetic condition characterised by underdeveloped facial bones. Most children with Treacher Collins syndrome have underdeveloped or absent external and middle ears. Conductive deafness is caused by anomalies of the outer or middle ear.

Treacher Collins syndrome is caused by a genetic mutation (a change to a specific gene). People with Treacher Collins syndrome have a 50% chance of passing it on to their children.

Visit the Treacher Collins Family Support Group at **treachercollins.org.uk** or find them on Facebook at **facebook.com/TreacherCollinsFamilySupportGroupUK.**

Goldenhar syndrome (also known as hemifacial macrosomia) is a congenital (born with) condition which usually affects one side of the face. It can occasionally affect both sides of the face – this is known as **craniofacial microsomia**. Children with hemifacial microsomia are born with underdevelopment of parts of the face. This might include the orbit (bony socket around the eye), the maxilla and mandible (jaw), the ear, the facial nerve and the soft tissues.

Goldenhar syndrome can affect other body parts, including the spine. Conductive deafness is caused by anomalies of the outer or middle ear. Some children may have sensorineural deafness. The cause of Goldenhar syndrome is currently unknown.

Visit the Goldenhar Family Support Group at **goldenhar.org.uk** or find them on Facebook at **facebook.com/Goldenhar**.

Crouzon syndrome (also known as craniofacial dysotosis) is a genetic disorder causing premature fusing of the skull bones, preventing normal bone growth. Different patterns of growth of the skull occur leading to characteristic head and facial features. Children with Crouzon syndrome sometimes have irregularities of the outer and/or middle ear. Conductive deafness may be caused by glue ear, perforation of the eardrum or anomalies of the middle ear. Some children may have sensorineural deafness.

Crouzon syndrome is caused by a genetic mutation (a change to a specific gene). People with Crouzon syndrome have a 50% chance of passing it on to their children.

DiGeorge syndrome (also known as 22q11.2 deletion syndrome, Velocardiofacial syndrome or VCFS) is a genetic condition. Many children with this syndrome have differently formed external ears. The syndrome can also affect lots of other parts of the body, including the heart, kidneys, bones, muscles, joints, mouth, hormones and immune system. Some children have a degree of learning disability and some are at higher risk of developing mental health conditions. Children with DiGeorge syndrome are more prone to long-standing glue ear, leading to conductive deafness. Some children may have sensorineural or mixed deafness.

DiGeorge syndrome happens when a small piece of genetic material is missing from a person's DNA. This usually happens at random. About 1 in 10 cases of DiGeorge syndrome are inherited.

The charity Max Appeal provides support to families affected by DiGeorge syndrome, VCFS and 22q11.2 deletion. Visit **maxappeal.org.uk.**

Pfeiffer syndrome is a genetic disorder that causes premature fusing of the skull bones, preventing normal bone growth. Different patterns of growth of the skull occur leading to characteristic head and facial features, which can affect the hearing, vision and breathing and cause learning disabilities and difficulty feeding. Pfeiffer syndrome often causes differences in the outer or middle ear, leading to conductive deafness. Some children have mixed deafness.

Pfeiffer syndrome is caused by a genetic mutation (a change to a specific gene). People with Pfeiffer syndrome have a 50% chance of passing it on to their children.

Nager syndrome is a very rare congenital (present at birth) condition. Many children with Nager syndrome have underdeveloped outer or middle ears. Some children might also have underdeveloped facial features or arms. Conductive deafness is caused by anomalies of the external or middle ears.

Doctors suspect that Nager syndrome could be caused by a genetic mutation (a change to a specific gene). However, the specific genetic mutation which causes Nager syndrome hasn't yet been identified.

Where can I get further help?

Microtia UK is a charity campaigning for improved awareness of microtia and atresia, and access to technologies. They support families by providing evidence-based information and organise annual events where families and medical professionals can come together to share information and discuss issues. Visit **microtiauk.org**

Microtia Mingle UK is a Facebook support group for families and adults affected by microtia.

Headlines is a charity for children affected by craniosynostosis (when one or more of the plates in the skull fuses). They produce a newsletter and children's newsletter three times a year and have an extensive library of leaflets, medical articles, videos and other resources.
Visit **headlines.org.uk**.

Changing Faces is a UK charity that supports and represents people who have disfigurements of the face or body from any cause. Visit **changingfaces.org.uk.**

The London Centre for Ear Reconstruction is run by a UK-based consultant plastic surgeon specialising in ear reconstruction. The website has useful information on the surgery and step-by-step photos of surgical outcomes. Visit **earreconstruction.co.uk**

Thanks

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References

1. *UK Care Standards for the Management of Patients with Microtia and Atresia*
2. *Microtia – A new parents guide,* Microtia UK.
3. *Microtia-Atresia.* 2011. *Possible Cause & Incidence (Etiology).* [ONLINE] Available at: www.microtia.us.com/microtia-atresia-possible-cause-and-incidence.html. [Accessed 13 July 15].

Useful resources from the National Deaf Children's Society

'Understanding your child's hearing tests': **ndcs.org.uk/hearingtests**
'Glue ear: A guide for families': **ndcs.org.uk/glueear**
'Hearing aids: A guide for families': **ndcs.org.uk/hearingaids**
Unilateral deafness: **ndcs.org.uk/unilateral**
Bone conduction hearing implants and bone-anchored hearing aids: **ndcs.org.uk/boneanchored**