

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.

What are microtia and atresia?

Microtia is a term used to describe under-development 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 under-developed (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, and is known as unilateral microtia. It more often affects the right ear than the left.² About one in 10 children are affected on both sides – this is known as 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. There is more information on these syndromes at the end of this factsheet.

What is the cause of 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 is usually a random, one-off event. It is 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, or 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 – see next section).

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, ear, nose and throat (ENT) surgeon, audiovestibular (hearing and balance) physician or paediatrician, audiologist, clinical psychologist and specialist nurse. 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 and at this stage you may have questions about improving the cosmetic appearance of the ears. The team will be able to answer your questions, share with you 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, any decision to go ahead with surgery is usually not until about the age of 10 years. At this time many children and parents decide that they do not want reconstruction of the ear and are happy with the child's appearance.

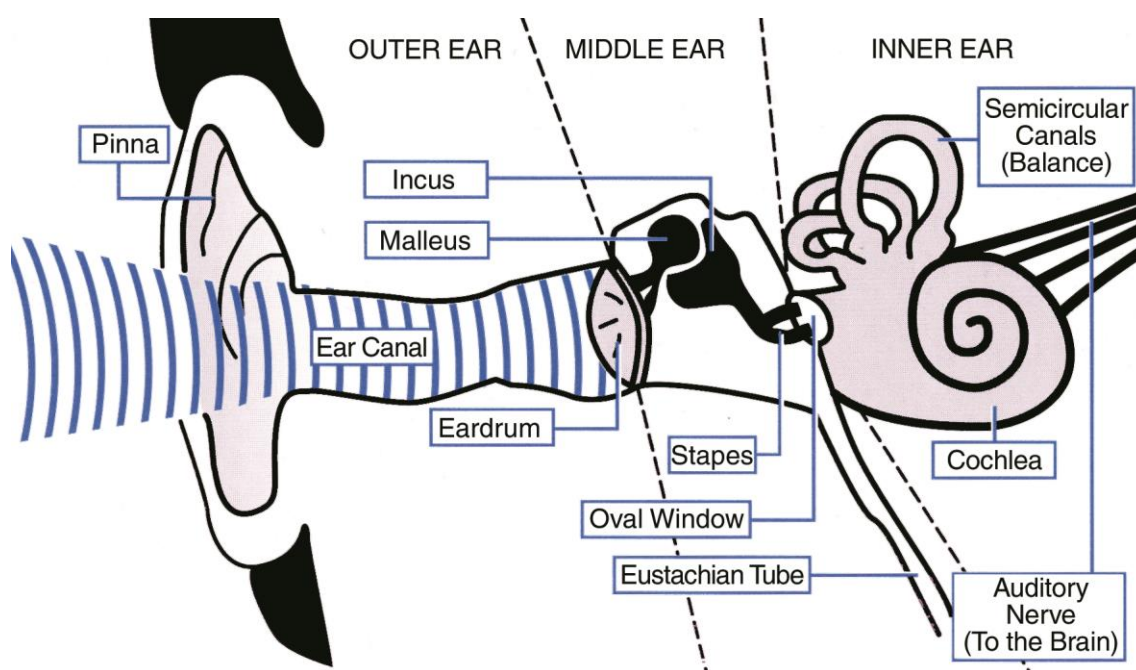
Medical tests

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. They 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 may 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. For more information on these medical tests see our resource *Understanding your Child's Hearing Tests*.

The specialist team will keep your child under regular review through childhood. They will work in close liaison with local audiology teams. Your team will again discuss options for reconstructive procedures as your child grows up and procedures develop. Your child will increasingly be included 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, sensori-neural or mixed – that can be associated with microtia and atresia, depending on which part of the ear is not formed or working as it should.



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 deformities of the middle and inner ear that will affect the level of deafness experienced. Some of the deformities encountered include:

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

Sensori-neural 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). Sensori-neural deafness is unusual in children with microtia and may indicate a co-existing condition or associated syndrome.

Mixed deafness is when children who have conductive deafness also have sensori-neural 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. But 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 further information visit www.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 on glue ear see our resource *Glue Ear*.

Hearing tests

Because children with unilateral microtia are relying on the hearing in their fully developed ear, it is wise to have regular hearing tests to monitor their hearing while they are 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 are worried about your child’s hearing or feel that their hearing has changed, ask for an earlier appointment.

It is 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 both 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 types of deafness and hearing tests see our resource *Understanding your Child's Hearing Tests*.

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 who will 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 help improve your child's hearing. Depending on the type and level of the deafness, there are several options available.

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 in each ear.

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. Children with microtia often have a very small pinna that 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.

The narrow ear canals can cause some children difficulty in wearing behind-the-ear hearing aids as sound from the hearing aid 'bounces' 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 of hearing aids in children with microtia may be challenging and difficult. It may need special expertise.

Every hearing aid is programmed for the ear it will be worn behind. When the hearing aids are fitted you will 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 Teacher of the Deaf will discuss when the best times are to use the hearing aids. For more information about hearing aids see our resource *Hearing Aids*.

Bone-conduction hearing aids and implantable hearing devices

Bone-conduction hearing aids

Children with absent or very under-developed 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.

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 have trialled and found 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:

1. *Clipped to a fixture, known as 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 aid or system.*
2. *With a magnet.*

Sound is conducted 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 after the age of four years.

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

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 who have mild to severe sensori-neural hearing loss, as well as for 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 it is likely that your child will have cosmetic surgery or prosthetic ears fitted later it is important that you discuss with your surgeon the exact position of the device.

For more information on bone-conduction and bone-anchored hearing aids visit www.ndcs.org.uk/boneanchored.

Surgery

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

Whether or not the surgery improves the hearing depends on whether the middle and inner ear are present and developed typically. This type of surgery is not normally carried out until about the age of nine years. This is because bony regrowth is likely in very young children and 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 any surgery before you agree to go ahead with it. As much 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 leave the decision to the child when they are 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:

- 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 about the age of nine to ten years to allow time for sufficient rib cartilage to have developed. Additionally, leaving surgery until children are a little older 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 – ensuring 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 from the age of three years. 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 and are 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. This is usually over the age of four years. The prosthetic ear will need to be replaced on a fairly regular basis. It can be removed at night for sleeping and it is important to maintain good hygiene of the skin around the fixtures with careful cleaning to avoid infection.

Deciding on the right option

Whichever option you might be considering, ask your surgeon how many operations of this type he or she has performed and for photos of outcomes for other children. Ask questions about the pros and cons of each type of procedure. Some questions you might like to consider:

- 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 is 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

The majority of children with microtia have no other medical problems. However, microtia also happens 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 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 Family Support Group www.treachercollins.net

Hemifacial macrosomia (Goldenhar's syndrome) 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 under development 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. Hemifacial microsomia may be associated with other birth anomalies including anomalies of the spinal vertebrae.

Conductive deafness is caused by anomalies of the outer or middle ear. Occasionally children may have a sensori-neural deafness.

Goldenhar Family Support Group www.goldenhar.org.uk

Crouzon syndrome 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 malformations of the outer ear and/or the middle ear. Conductive deafness may be caused by glue ear, perforation of the eardrum or anomalies of the middle ear. Occasionally children may have a sensori-neural deafness.

Crouzon Support Network www.crouzon.org

22q11.2 deletion syndrome (sometimes known as DiGeorge syndrome and Velocardiofacial syndrome) is a genetic condition. Many children with this syndrome have malformations of the external ear. Other parts of the body affected may include the heart, kidney and/or immune system. Some children have a degree of learning disability. Children are more prone to long-standing glue ear, leading to conductive deafness. Occasionally children may have a sensori-neural or mixed deafness.

Max Appeal! Supporting families affected by DiGeorge Syndrome, VCFS and 22q11.2 deletion www.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. Many children with Pfeiffer syndrome have malformations of the external or middle ear. Conductive deafness is caused by anomalies of the outer or middle ear. Occasionally children have a mixed deafness.

Nager syndrome is very rare. Many children with Nager syndrome have underdeveloped external or middle ears. Sometimes underdevelopment of other facial features and/or the arms is also present. Conductive deafness is caused by anomalies of the external or middle ears.

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.

www.microtiauk.org

Microtia Mingle UK is an online support group (on Facebook) 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.

www.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.

www.changingfaces.org.uk

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.

www.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 National Deaf Children's Society resources

Understanding your Child's Hearing Tests www.ndcs.org.uk/hearingtests

Glue Ear www.ndcs.org.uk/glueear

Hearing Aids www.ndcs.org.uk/hearingaids

Unilateral deafness www.ndcs.org.uk/unilateral

Bone conduction hearing implants and bone anchored hearing aids

www.ndcs.org.uk/boneanchored

This information can be requested in large print or as a text file.

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For resource references or to give us your feedback email informationteam@ndcs.org.uk.

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