ndcs
every deaf child

A child with microtia and atresia

A guide for families

Our vision is of a world without barriers for every deaf child.

Content

Introduction

This booklet is for families who have a child diagnosed with microtia or atresia. It explains what microtia and atresia are, about different types and causes of deafness that may be related to microtia or atresia and how microtia, atresia and any associated deafness can be managed. Other medical conditions that have microtia or atresia as components of the condition are reviewed. Throughout the booklet there are references to other publications that give more detail about some topics. If you would like more information about any of the topics covered in this booklet or would like to order a copy of these booklets, phone our Freephone Helpline on 0808 800 8880 (voice and text), send an email to helpline@ndcs.org.uk, or visit our website at www.ndcs.org.uk. For other sources of information go to p23, 'Where can I get further help?'

NDCS uses the word 'deaf' to refer to all levels of hearing loss.

Acknowledgements

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We would also like to thank Changing Faces for kindly allowing us to reprint some photos in this publication.

What are microtia and atresia?

Microtia is a term used to describe malformation of the pinna (external part of the ear). Mictrotia varies in severity from quite minor changes (such as the ear being smaller than expected) to 'classic microtia' where the pinna is missing and is associated with absence of the external auditory meatus (ear canal). Absence of the ear canal is known as atresia. Microtia is frequently accompanied by atresia because the baby's outer ear and the ear canal develop together during pregnancy. In some cases the ear canal can look normal from outside but ends at a 'blind alley' inside.

Microtia happens more often in boys than in girls. It usually affects one side: this is known as unilateral microtia. Unilateral microtia more often affects the right ear than the left.

Approximately one in ten children with microtia are affected both sides. This is known as bilateral microtia.

Microtia is known to affect about one baby in every 8,000–10,000 births.

In families where there is already one person with microtia or atresia there is some evidence that suggests the chance of future children having the same condition is about 1 in 20.

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. Microtia is known to occur as part of several different syndromes. There is more information on these syndromes in the section 'Syndromes and other medical conditions that are associated with microtia' on p20.

What is the cause of microtia and atresia?

Microtia or atresia occurs when the outer part of the ear fails to develop normally during the early stages of pregnancy. The exact reasons for isolated (or non-syndromic) microtia or atresia remain unclear although it is known that it is usually a random, one-off event. It is important for parents to understand it was not caused by anything the mother did wrong before or during the pregnancy. Medical research has suggested that occasionally certain prescribed medications taken during pregnancy, or genetic and/or environmental factors may be involved. Associated factors will be explored at the initial meeting with the multidisciplinary team.

My child has microtia – what happens now?

Your child will need to 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 assess your child's hearing at regular intervals and is responsible for fitting and maintaining any hearing aids that may be used in the management of any deafness.

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. At the specialist clinic you will see a multidisciplinary team that may include a specialist ENT surgeon, plastic surgeon, audiological physician, audiologists and speech and language therapists. The team will also have access to a geneticist and other professionals who may need to be involved in the overall management of your child.

The specialist team will usually meet with you soon after birth. They will discuss the options available for your child and develop with you a long-term management plan. At this stage parents often have lots of questions about improving the cosmetic appearence 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 demonstrate expected outcomes of ear reconstruction surgery. In the meantime they will liaise with your local audiology department to ensure your child's hearing is assessed and hearing aids are fitted if appropriate. The purpose of the first early consultation with the multidisciplinary team is to ensure that any deafness is managed appropriately, alleviate concerns you may have and advise about the options available in the future.

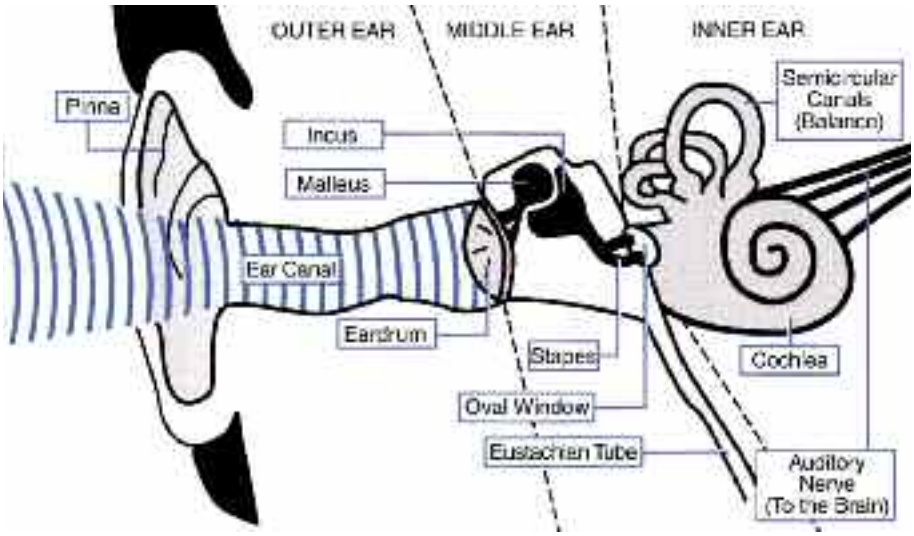
Because microtia is known to occur as part of several different syndromes that may involve other organs you may be offered a range of medical tests for your child, either by your local audiology department or the specialist team. These tests will include blood and urine tests, renal (kidney) ultrasound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

For more information on these medical tests read the NDCS booklet *Understanding your Child's Hearing Tests*.

When your child is about six years of age the specialist team will arrange to see you again. At this time they will discuss with you again any possible reconstruction procedures available. At the meeting your child can also be involved in the discussion and decisions. Often the child or parents may decide that they do not want any further management or reconstruction of the ear and indeed are happy with the child's appearance.

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 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).

Microtia can cause conductive deafness. Microtia and atresia may be associated with deformities of the middle and inner ear that will affect exactly 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.

Grades of microtia

Grade I – a slightly small ear with identifiable structures and a small but present ear canal. Grade I microtia is not usually associated with deafness.

Grade II – a partial pinna with a closed off ear canal. Grade II microtia causes conductive deafness.

Grade III – the pinna, ear canal and eardrum are missing. There may be a small, peanut-shaped lobe near where the pinna would be. Grade III microtia causes conductive deafness.

Grade IV – may be known as anotia and means 'no ear'.

Depending on which parts of the ear are affected, typically a child with microtia and atresia will have a moderate deafness (40–70 dB) in the affected 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. This type of deafness may indicate another abnormality or an 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. Most children with unilateral deafness develop clear speech. There are a few situations where they may find it more difficult to hear well:

- hearing sounds or speech on the side with the deafness
- identifying the source of a sound or the direction a sound is coming from
- understanding speech when there is background noise.

For further information read the NDCS booklet *Unilateral Deafness*.

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 better ear at some point and this may need to be managed by the specialist team. For more information on glue ear read *Glue Ear: A guide for parents*.

Because children with unilateral microtia are relying on the hearing in their better ear, it is wise to have regular hearing tests to monitor their hearing while they are young. Depending on the degree 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 between appointments, 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 read the NDCS booklet *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 appropriate 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 grade 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.

This photo is to show a behind the ear hearing aid. The child in the picture does not have microtia.

The hearing aid sits on the top of the pinna (the outside part of the ear) and is connected to an ear mould that is specially made to fit in the child's ear. Children with microtia often have a very small pinna that can make keeping a behind the ear hearing aid in place difficult. Smaller ear canals can sometimes make it difficult to get well-fitting ear moulds.

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 read the NDCS booklet *Hearing Aids: Information for families*.

Bone conduction and bone anchored hearing aids

Children with grade II, III or IV microtia will not be able to use a conventional behind the ear hearing aid and may benefit from a different type of hearing aid. Bone conduction hearing aids use a vibrating pad that allows sound to be conducted through the bone rather than through the middle ear. They are worn on a headband with the vibrator behind the ear, resting on the mastoid bone (part of the skull behind the ear).

This photo is to show a bone conduction hearing aid. The child in the picture does not have microtia.

In very young children a bone anchored hearing aid on a soft headband can be used. The bone anchored hearing aid soft headband is taken on and off like other bone conduction hearing aids and can be used permanently or temporarily during the assessment stage.

Children who have trialled and found a bone conduction hearing aid effective may also be suitable for a bone anchored hearing aid. These hearing aids have a sound processor that clips onto a fixture (known as an 'abutment') attached to a small titanium screw that has been implanted in the skull just behind the ear. Through the abutment, the sound processor is directly connected to the skull bone which the cochlea (inner ear) forms part of. This allows sound to be transmitted more directly to the inner ear. This procedure is usually offered after the age of four years.

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 abutment.

This photo is to show a bone anchored hearing aid. The young person in the picture does not have microtia.

For more information on bone conduction and bone anchored hearing aids read the NDCS booklet *Bone Anchored Hearing Aids: Information for parents and families.*

Surgery

Depending on the nature and degree of the deformity, it may be possible to have surgery that improves the hearing. It may not be possible to wear hearing aids and surgery may be required. This will depend on whether the middle and inner ear are present and developed normally. 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.

There are three options to surgically improve the appearance of the ear:

- autogenous reconstruction
- reconstruction using an artificial framework
- prosthetic ears.

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 six years to allow time for sufficient rib cartilage to have developed. Additionally, leaving surgery until children are a little older can mean 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 and ensuring it looks the same as the opposite ear. 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 as their preferred option. 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.

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 anchored hearing aid surgery and is likely to have cosmetic surgery in the future, it is very important that the exact placement of the bone anchored hearing aid abutment is discussed with your surgeon so that it does not prevent reconstructive surgery or proper placement of prosthetic ears in the future.

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

Treacher Collins syndrome is a genetic condition characterised by underdeveloped facial bones. Most children with Treacher Collins syndrome have malformations of 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 microsomia

Hemifacial microsomia affects the development of the lower half of the face, most commonly the ears, the mouth and the lower jaw. Most children with hemifacial microsomia have malformations of the external or middle ear. Conductive deafness is caused by anomalies of the outer or middle ear. Occasionally children may have a sensori-neural deafness.

For information on hemifacial microsomia see the Goldenhar Family Support Group **www.goldenhar.org.uk**

Goldenhar
Goldenhar syndrome is characterised by incomplete development of the ear, nose, soft palate, lip and mandible (jaw). Other parts of the body affected may include the heart, kidneys and lungs. Either the organ is not present on one side or will be underdeveloped. Usually just one side of the body is affected but occasionally both sides are affected. Conductive deafness is caused by anomalies of the outer or middle ear.

Goldenhar Family Support Group **www.goldenhar.org.uk**

Crouzon
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)
22q11.2 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 longstanding glue ear, leading to conductive deafness. Occasionally children may have a sensori-neural or mixed deafness.

Max Appeal! **www.maxappeal.org.uk**

Pfeiffer syndrome
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
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?

NDCS
15 Dufferin Street, London EC1Y 8UR
Phone: 0808 800 8880 (voice and text)
Email: helpline@ndcs.org.uk
www.ndcs.org.uk

Headlines The Craniofacial Support Group is a UK charity set up by a group of parents whose children were affected by craniosynostosis. Headlines produces a newsletter and children's newsletter three times a year and has an extensive library of leaflets, medical articles, videos and other resources.
www.headlines.org.uk

Contact a Family is a UK charity providing support, advice and information for families with disabled children, no matter what their condition or disability.
www.cafamily.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/microtia.php

Children's Craniofacial Association (CCA) is a USA nonprofit organisation dedicated to improving the quality of life for people with facial differences and their families. Nationally and internationally, CCA addresses the medical, financial, psychosocial, emotional and educational concerns relating to craniofacial conditions. CCA's mission is to empower and give hope to individuals and families affected by facial differences.
www.ccakids.com

www.microtia.us.com is a comprehensive website written for families and patients with microtia by an experienced USA-based plastic surgeon

www.microtia.org.uk is a UK-based online discussion forum for anyone affected by microtia.

NDCS provides the following services through our membership scheme. Registration is simple, fast and free to parents and carers of deaf children and professionals working with them. Contact the Freephone Helpline (see below) or register through www.ndcs.org.uk

- A Freephone Helpline 0808 800 8880 (voice and text) offering clear, balanced information on many issues relating to childhood deafness, including schooling and communication options.
- A range of publications for parents and professionals on areas such as audiology, parenting and financial support.
- A website at www.ndcs.org.uk with regularly updated information on all aspects of childhood deafness and access to all NDCS publications.
- A team of family officers who provide information and local support for families of deaf children across the UK.
- Specialist information, advice and support (including representation at hearings if needed) from one of our appeals advisers in relation to the following types of tribunal appeals: education (including disability discrimination, special educational needs (SEN) and, in Scotland, Additional Support for Learning (ASL)); and benefits.
- An audiologist and technology team to provide information about deafness and equipment that may help deaf children.
- Technology Test Drive – an equipment loan service that enables deaf children to try out equipment at home or school
- Family weekends and special events for families of deaf children.
- Sports, arts and outdoor activities for deaf children and young people.
- A quarterly magazine and regular email updates.
- An online forum for parents and carers to share their experiences, at www.ndcs.org.uk/parentplace.
- A website for deaf children and young people to get information, share their experiences and have fun www.buzz.org.uk.

NDCS is the leading charity dedicated to creating a world without barriers for deaf children and young people.

NDCS Freephone Helpline:
0808 800 8880 (voice and text)

Email: **helpline@ndcs.org.uk**

www.ndcs.org.uk

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