a guide to understanding microtia

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this parent's guide to microtia is designed to answer questions that are frequently asked by parents of a child with microtia. It is intended to provide a clearer understanding of the condition for patients, parents and others.

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This booklet is intended for information purposes only. It is not a recommendation for treatment. Decisions for treatment should be based on mutual agreement with the craniofacial team. Possible complications should be discussed with the physician prior to and throughout treatment.

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what is microtia?

This condition can range in severity from a small ear that has all of its parts, including an ear canal and a normal-appearing shape, to one that has no ear canal and only a small amount of skin and cartilage. It can occur either on one or both sides. Microtia often occurs as an isolated problem and is not associated with other abnormalities. There are, however, syndromes such as Treacher Collins and Hemifacial Microsomia in which microtia also occurs. These syndromes are further discussed in other booklets.

why was my child born with microtia?

o one knows the causes of microtia. It is not passed down in the family. The pregnancy itself is often uneventful. The chances of having a baby with microtia are about 1 in 6000. The cause may be related to poor blood supply to the tissue during ear development; however, this is only speculation. Since the causes are unknown, prevention is therefore difficult.

can my child hear?

child with microtia often has a normal inner ear; therefore, in those cases children with microtia generally have some hearing in the affected ear. The presence of the ear canal and middle ear are variable. So, the amount of hearing also varies depending on the presence of these structures. If both of these structures are present, hearing in the affected ear can approach normal. It is important to note that if the opposite ear is unaffected, the child will have normal hearing in that ear, regardless of the condition of the affected ear. Since the inner ear is normal in the majority of children with microtia, those children with microtia on both sides still have the ability to hear. This type of hearing is often further enhanced with hearing aids to ensure normal speech and language development.

does microtia affect normal development?

enerally, having either one or two sided microtia does not affect development. This is true if early steps, within the first few months from birth, are taken to ensure that hearing is normal. These steps include seeing an ear, nose, and throat (ENT) surgeon (often a member of the craniofacial team) who can assess ear function and direct hearing aid placement if necessary. If the microtia is part of a syndrome, development may be affected depending on the syndrome and its severity.

what are my child's treatment options for A) hearing and B) appearance?

multidisciplinary approach is required. This is often provided by a craniofacial team usually located within a tertiary care or university center. The advantage of this type of approach is that the child's care is coordinated among all of the members of a craniofacial team, optimizing results. All members work together at each step to ensure that all of the child's treatment and life needs (as they relate to microtia) are met.

A) Hearing – Children with one-sided microtia who have one normally hearing ear essentially have normal hearing in one ear and do not require specific treatment. There are centers that offer the creation. of an external opening and ear drum when relatively normal middle ear structures are present. An advantage of this approach is obtaining normal hearing in the affected ear. Disadvantages, however, include a variable success rate meaning that normal hearing may not be achieved after lengthy surgery. The new ear canal may also close, resulting in a scar in the area. A detailed examination, investigations and discussion with an ENT surgeon are necessary prior to proceeding down this treatment path. Commonly, in children with both sides affected, hearing aids are recommended to ensure normal hearing and speech and language development. Broadly, two types of

hearing aids exist: those that are worn on a headband and others that are anchored to the skull bone. Communication between the surgeon doing the ear reconstruction and the one placing the boneanchored hearing aids is very important to ensure that the hearing aids do not interfere with appropriate positioning of the ear. This type of communication occurs readily within a craniofacial team environment.

- B) Appearance The child will live with having microtia and the consequences of its treatment his entire life. Therefore, it cannot be overstated that the principles of treatment for facial appearance should be guided by the child. Surgery or prosthesis fitting generally occurs between 6-10 years of age. This timing varies, depending upon how the child is coping socially and psychologically and also on the growth of the face and body. Treatment options include no intervention versus reconstruction of the external ear. The reconstructive options include: having a prosthesis made, reconstruction that uses both artificial and the patient's own tissue, and total surgical reconstruction using only the patient's own tissues.
- i) No Intervention Ear reconstruction is not a necessity. This is true especially if the child does not perceive having microtia as a problem both psychologically and emotionally. The options for the creation of a new ear change very little as the child

with microtia grows and so the subject of ear creation can be discussed and performed at any time including adulthood.

ii) Prosthetic Reconstruction – A prosthesis is completely artificial and usually made of silicone from a mold that often uses the opposite ear or a parent's ear as a template. The prosthesis can either be held onto the head with adhesive or using magnets or clips. The magnetic or clip-on prosthesis requires a minor surgical procedure to place bone screws permanently into the skull. Advantages include an excellent color match and natural appearing ear. There is either no or only minor surgery involved, so pain and scarring is minimal. Disadvantages include the need for prosthetic replacement every few years as the old one wears out, which may incur costs to the family depending on the insurance plan. The color of the prosthesis does not change in different weather conditions and so it can look unnatural particularly when the child is flushed or blushing. In addition there is always the possibility of the prosthesis coming loose or falling off at an inopportune time which can be socially difficult for the child (this is more likely with a prosthesis that is held on with an adhesive). Finally, cleaning and skin care around the site of retention is required daily to maintain the life span of the prosthesis.

iii) Combined Prosthetic and Own-Tissue

Reconstruction – This form of treatment uses a premade ear framework that is then covered by the child's own local tissues. The framework is made of a plastic material that has holes within it allowing blood vessels and tissue to grow into it over time. Therefore, the framework eventually becomes a part of the child. Advantages include no need for harvesting cartilage from the ribs to create an ear framework. This means less surgery and pain. The reconstruction often only needs one main operation with one or two minor ones to complete the ear reconstruction. The new ear becomes a part of the child's body image. Disadvantages include infection which is a concern anytime artificial material is used within the body. A severe infection may require removal of the framework and loss of the reconstruction. Also, the long-term outcomes of this type of reconstruction (how it lasts for the person's lifetime) are still unknown. Finally, if the person has an injury that results in exposure of the framework at any point in time, the entire reconstruction can be at risk of being lost.

iv) Total Reconstruction Using Only Own Tissue

– This type of reconstruction uses the child's rib cartilage (usually 3-4 ribs) to create an ear framework that is carved out of the harvested cartilage. The framework is then placed under the skin in the appropriate position. A second operation, 3-9 months after the first, is performed to lift the ear

into a position similar to the opposite ear. Sometimes a third and even fourth operation is necessary to fine-tune the appearance of the reconstruction and move the ear lobe. Advantages include the use of the patient's own tissue that generally does not undergo rejection and has a lower risk of infection. The reconstruction becomes a part of the child's body image. Generally, exposure of the cartilage after injury is salvageable without loss of the entire framework. Disadvantages include two or three operative sites and multiple operations. A scar is left on the rib cage. Sometimes the removal of cartilage can leave a chest wall deformity. After the second operation, skin grafting of the back of the ear is necessary. There will be additional scarring at the site of graft harvest. The cartilage in the ear can also change with time requiring additional touch-ups.

The choice of the best treatment method for your child must involve open discussions between the members of the craniofacial team, the family/caregivers and, most importantly, the child. There is not one best option for treatment in all children. The best treatment option remains the one that best suits the child's needs and desires..... social, emotional and psychological.

what other problems or treatments might we expect?

nce treatment for ear reconstruction is requested, regardless of the type of reconstruction used, care and additional surgeries may be necessary to keep the ear appearance from changing with time. If your child has microtia as part of a syndrome, additional treatments may be necessary to address syndrome-related problems. If your child wears a hearing aid, life-long maintenance will be required. In situations where both ears are involved, additional surgeries are often necessary to complete both reconstructions.

how will microtia affect my child psychologically?

his is a complex question. Having microtia may ■not affect your child at all or it may affect selfesteem, body image perception and confidence. This depends on multiple factors including family interactions, family and child education about microtia, the school environment and whether your child has developed or been taught the tools to cope with his/her differences. The social worker and child psychologist that are a part of the craniofacial team are a good resource for starting the education process. They can help with opening the avenues of communication between family members and even coordinating meetings between families who have already gone through the treatment process. These families can often be important resources for information, offering real-life experiences to families just beginning the treatment process.



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