

A GUIDE TO UNDERSTANDING MICROTIA

What is Microtia? Microtia is an incompletely formed ear. The term "micro" means small and "otia" means ear. Hence, when translated literally, "microtia" means small ear. At times a bump of tissue is present in the location where an ear would normally be found. In other cases, the **lobule**, the lower part of the ear, and the **concha**, the hollow part of the ear, may be partially formed and the entire upper part of the ear is missing.

Are both ears usually involved? Although microtia can involve one ear or both ears, 80% of the time only one is affected.

Can microtia be associated with other conditions or syndromes? The most common condition in which microtia is seen is **Hemifacial Microsomia**. This is a condition which involves one ear. This is called **unilateral microtia** or one-sided. In this case, one half of the face does not grow in proportion to the other. Hemifacial Microsomia is discussed in further detail in another booklet entitled, *A Guide to Understanding Hemifacial Microsomia*.

What if both ears are involved? If both ears are involved the condition is considered to be **bilateral** or two-sided. A child with this condition could have **Bifacial Microsomia**, which is similar to Hemifacial Microsomia. However, in this case, both sides of the face are affected.

Are there other craniofacial conditions involving the ears? Another condition to consider if both ears are microtic is **Treacher Collins syndrome**. In Treacher Collins syndrome the eyes are also affected and they appear to have a downward slant or "pulled down appearance." This syndrome is explored in greater detail in the booklet entitled, *A Guide to Understanding Treacher Collins Syndrome*.

Why does microtia happen? At the present, no one knows why microtia occurs. Further, there is no evidence that the mother's actions or activities during her pregnancy contribute to this condition. It is most probable that the child received a normal set of genes from both parents. Yet something occurred early in the development stage to prevent the normal growth of the ear. Some doctors believe that microtia may be a very mild form of Hemifacial Microsomia. Further research and discoveries in the

understanding of human genes and fetal growth are necessary before many questions such as this may be answered.

What are the chances of producing a child with microtia? An estimate of the occurrence of microtia is one in 6000 births. Adults with microtia are unlikely to pass the condition to their offspring. In fact, the probability of this happening is less than 6 percent.

How will my child's hearing be affected? It would seem that your child will be unable to hear from the microtic ear. However, in most cases, there is only an approximate 40% reduction in hearing. The effect might be similar to your sticking fingers in both of your ears. This is true because one can hear via "**bony conduction**." This means that sound travels through the skin and the bones of the skull and into the inner ear.

Is the inner ear also abnormal? Most children with microtia have a normally formed inner ear in the absence of an external ear opening. Therefore, there is still an ability to hear from the affected side. Children with unilateral microtia will have problems locating the direction from which a sound comes. Children with unilateral microtia usually do not require hearing aids.

Are ear infections a problem? They can be. It is very important to aggressively treat ear infections in the unaffected ear to preserve hearing in the *normal* ear. It is also important to establish a close working relationship with your pediatrician or family practice doctor so that your child's ear can be checked frequently during the "cold season."

Do children with bilateral microtia have the same problems? Children with bilateral microtia will have more of a hearing problem. For these children, hearing is typically reduced by 40% on both sides. Due to this high degree of hearing loss, much of what is said is simply not heard. Children with bilateral microtia should be fitted with a special type of hearing aid before six months of age. The sounds that babies hear in the first year of life are very important for speech and language development. Future success in school also depends on one's ability to hear adequately.

What treatment is available? Although the outside of the ear contributes little to hearing, it does serve several important functions such as supporting glasses. Surgery to rebuild the outer ear is considered reconstructive surgery. Before surgery is begun, this writer believes that it is wise for the child and the family to visit with a **prosthetist**, who is a specialist who makes artificial ears. This allows the child to be involved in the decision-making process and in determining what is best for him. While artificial ears look better than reconstructed ears, they are always cooler than normal skin, have an artificial feel, and do not change color as the surrounding skin does at different times of the year.

When is the best time to begin reconstruction? Reconstruction of the outer ear usually takes in approximately four separate operations which are spaced at least three to six months apart. In the first operation, rib cartilage is taken from the chest, carved to resemble the shape of an ear, placed in a pocket under the skin, and then allowed to heal. At the present, there is no adequate, artificial material which can substituted for the child's rib without increasing the risk of complications. Nor is it possible for a donated rib from a parent to be used. The donated rib would eventually be rejected.

What is involved in the other operations? The second stage varies with each doctor. However, the usual procedure involves making an incision around the ear to lift it slightly. A skin graft is then placed on the backside of the cartilage. This procedure helps to create the space normally found behind the ear, but it does not usually help the ear to stick out any further from the head.

In the third and fourth stages of surgery, small amounts of tissue are moved around in order to improve the appearance of the ear. These stages are usually scheduled at least three months apart. Small parts of the opposite ear are used to rebuild the microtic ear. The normal ear is sometimes "set back a bit" in order to match the reconstructed ear.

How long will my child be hospitalized? In our surgical center, children usually spend two nights in the hospital for the first stage of surgery. The remaining stages are done as day surgery and the child leaves the hospital on the same day as surgery.

Are the surgeries the same for patients with bilateral microtia? Reconstruction for bilateral microtia can take more operations to rebuild both ears depending on procedures of the particular surgical center. At the Dallas center, both ears are being reconstructed at the same time. This reduces the number of operations and decreases the time spent in the hospital. Rebuilding the ears simultaneously trims the medical costs, a very important notion in a time of rising health-care costs. Nevertheless, it is not wrong to perform constructive procedures in separate operations if the doctor wishes.

Is there surgery to improve the hearing? There is a surgical procedure to improve hearing. It involves drilling out an ear opening and rebuilding an eardrum. This is done by an otolaryngologist (ENT). An **otolaryngologist** is specialized in working with ears, noses, and throats. The results of this operation depend on how much of the middle ear is present, as well as the experience and expertise of the surgeon. Most children with bilateral microtia are good candidates for this procedure. It is best to perform this surgery after the external ear has been rebuilt. If the surgery for impaired hearing is done first, it is sometimes impossible to later build the outer ear. For children with unilateral microtia, it is more difficult to decide if surgery is necessary. The patient and his family should make this decision only after consulting with the otolaryngologist and after weighing the possible benefits against the possible risks.

Where should I go to seek treatment for my child? Your child should be treated by a qualified craniofacial team. Microtia is a complex problem. It requires the expert skill of many different specialties working together. These problems are best treated by craniofacial teams experienced in the management of these patients.

What is a craniofacial team? A craniofacial team is a group of specialists specifically trained in the surgical management of problems involving the face and head. Your child might not see all of these specialists. However, the basic team members and their roles in the treatment of craniofacial children are detailed in the information that follows.

Anthropologist - This specialist takes multiple measurements of the face and skull. These measurements are compared to tables of normal values. An example of these measurements is the distance between the eyes. Craniofacial surgeons use the information gathered by the anthropologists to see how a child is growing and developing. This also assists in the planning for corrective surgery.

Craniofacial Surgeon - This individual is usually the craniofacial team leader and frequently coordinates the appointments with other doctors. The craniofacial surgeon will have usually completed general surgery training, plastic surgery training, and a fellowship in craniofacial surgery at a children's hospital.

Geneticist - This physician specializes in categorizing different syndromes. This is very important to the overall care of the child. After the geneticist makes the diagnosis, the team members can then look for problems, which may be associated with that condition or syndrome and perhaps prevent them. The geneticist also counsels families as to the possibility and probability of future generations having Microtia.

Medical Photographer/Medical Artist - The photographer takes photographs that are enlarged to life-size. From these life-sized photographs, a medical artist can suggest changes in appearance using life-sized drawings.

Neuro-ophthalmologist - This physician will closely follow your child's eyesight and closely monitor any problems. The doctor can perform surgery to balance the eye muscles if there are problems in looking straight ahead with both eyes.

Neuro-radiologist - This physician is specially trained to read x-rays and scans of the brain and the skull. This specialist provides important information to the craniofacial surgeon and neurosurgeon.

Pediatric Anesthesiologist - This doctor is a very important part of any craniofacial team. Children with craniofacial problems often have problems associated with the airways that create breathing difficulties. It is essential that this doctor be well trained in pediatric anesthesiology, but it is just as important

that he/she have substantial experience in dealing with these special children. The pediatric anesthesiologist's amount of experience with craniofacial problems perhaps has the greatest effect on the overall safety of the surgery.

Pediatric Dentist - Since children with craniofacial problems often have problems with their teeth, the pediatric dentist will care for these specialized problems. There are circumstances in which teeth are absent or a patient may be unable to open to mouth. This makes the care of their teeth quite difficult; therefore, the special skills of a pediatric dentist are needed.

Pediatric Intensivist - This is a pediatrician who specialized in the care of children in intensive care units. This specialist's expertise is called upon to follow children during the first night following surgery to insure that all goes well.

Pediatric Neurosurgeon - This doctor works with the craniofacial surgeon in the operating room and contributes in a big way to the safety of the procedure. This doctor has completed training in neurosurgery and has taken advanced training in pediatric neurosurgery.

Pediatric Nurse - Of all the team members, the pediatric nurse will probably spend the most time with your child. This nurse has specialized training not only in the treatment of children, but specifically the treatment of children with craniofacial conditions.

Pediatric Otolaryngologist - This specialist plays an important role in monitoring the child's hearing. (It has been found that even small improvements in a child's ability to hear can greatly affect his performance in school.)

Pediatric Psychologist - This individual performs two important functions. The first function is to monitor a child's development to determine a need for intervention in helping your child reach his potential. Second, this individual helps your child to cope with the stress and pressures arising from the medical condition. The psychologist can often provide parents with suggestions for dealing with interpersonal relationships. This is especially helpful with handling other children at school.

Social Worker - This person often introduces children to the hospital and helps them prepare for surgery. With the rising costs of medical care, the social worker can also help families by providing important financial information.

Speech Therapist - This specialist evaluates your child's ability to communicate. The trained ear of the speech therapist can sometimes catch early problems that can be corrected with simple speech exercises.

What are the advantages of treatment at a craniofacial center? Centers with large craniofacial teams working together have the advantage of greater experience. This definitely leads to better results and fewer complications. In addition, ongoing research at these centers offers patients the latest breakthroughs in treatment and technology. As there are only a few experienced centers in the country, it is not uncommon for families to travel long distances to get quality care. By contrast, children treated locally by individual physicians not working as a team or by inexperienced teams are at risk of unsatisfactory results. These children sometimes require two or three additional operations to correct procedures performed under these conditions.

Are there other advantages of receiving care at a craniofacial center? Another advantage of receiving care at one of the large craniofacial centers is that often more than one operation can be performed by different specialists at the same time. This decreases the total number of surgeries a child will need. Every effort is made to minimize the time a child spends in the hospital. This is important for your child's development, as well as for financial reasons. Having a child with Microtia can place an enormous financial hardship on the family. It is important to provide surgical correction with the lowest complication rate and with the shortest hospital stay. Craniofacial centers with qualified staff are equipped to accomplish this with the least amount of physical, emotional, and financial strain.

Are there other benefits? Another benefit of traveling to busy, qualified craniofacial centers is the opportunity to meet other children and families affected with similar conditions. These families often share their experiences and offer valuable advice. This provides a tremendous amount of emotional and moral support.

Written by Jeffrey Fearon, M.D.

Edited by Carolyn Johnson, M.Ed.