

# A GUIDE TO UNDERSTANDING HEMIFACIAL MICROSOMIA

**What is Hemifacial Microsomia?** Simply stated, this is a condition in which the lower half of one side of the face does not grow normally. The most obvious sign of this condition is a partially formed ear or a total absence of an ear. Parents may notice that the child's chin is off-center or closer to the affected ear. One corner of the mouth may be higher than the other. The term, "hemifacial", refers to one side of the face and "microsomia" refers to having a small body. When translated, "hemifacial microsomia" means "one half of the face has a small body."

**Are there other names for this condition?** Hemifacial microsomia was previously called first and second branchial arch syndrome. It may also be referred to as oral-mandibular-auricular syndrome.

**Are there other conditions similar to Hemifacial Microsomia?** If the forehead or eye is affected, **Craniofacial Microsomia** is the common diagnosis. In this condition, the forehead is flattened and the eye may be pulled downward.

**Goldenhar syndrome** is another very similar condition. Children with Goldenhar syndrome have the same characteristics as those with Hemifacial Microsomia. However, they also have benign growths of the eye. These growths are called **epibulbar dermoids**. Patients with Goldenhar syndrome also have neck problems, which are mostly commonly caused by a fusion or bony bridges between the bones of the neck. Goldenhar syndrome is also called **Oculoauricular Dysplasia** or OAV.

**Why does Hemifacial Microsomia happen?** At this time, it is uncertain why this happens. Studies do not link this condition with the mother's actions or activities during her pregnancy. It is most probable that the child with Hemifacial Microsomia received a normal set of genes from both parents. However, something occurs in the early stages of development to cause this condition. Studies using mice who are still in the womb show that if a small blood vessel near the ear breaks open and bleeds, the mice present with a condition that looks like Hemifacial Microsomia. There is no evidence that trauma or excessive motion of the mother causes this problem. Further, researchers are uncertain

that this evidence in mice is related to the development of Hemifacial Microsomia in human beings.

**What are the chances of producing a child with this condition?** The estimate of occurrence of Hemifacial Microsomia is one in 3500 births. For parents with one child with Hemifacial Microsomia, the chances of producing a second child with the condition is between zero and 1%. Adults born with Hemifacial Microsomia have approximately a

3% chance of passing the condition to their offspring.

**Do these numbers hold true for Goldenhar syndrome?** For adults with Goldenhar syndrome there is also a predicted 3 % chance of passing the condition to their children. There *are* families, however, who have a 50% chance of passing Goldenhar syndrome to their children.

**When is the best time to correct these problems?** The timing for surgical procedures varies with the severity of the condition. Surgical correction usually begins at approximately five years of age. The first operation involves lowering the upper jaw on the affected side to match the opposite side. In addition, a small piece of rib is taken from the child and is used to lengthen the lower jaw. This allows the chin to be moved back to the center of the face. This can be done without wiring the jaws together. This allows the child to begin eating soft foods shortly after the operation. The rib is the substance of choice because medical experience has shown that it will continue to grow as the child grows. For many children, only one operation is needed on the jaws. A few of the larger surgical centers are now experimenting with jaw lengthening devices which may reduce the number of surgeries needed.

**When can work begin on the ears?** The next surgical procedure takes place when the child is about six years old. This surgery is the first of three or four operations to rebuild a child's outer ear. External ears can be built from the child's own rib cartilage. This is discussed in greater detail in another booklet entitled, *A Guide to Understanding Microtia*.

**Besides surgical procedures on the jaws and ears, are there other surgeries?** In addition to these operations, additional procedures are sometimes recommended. Occasionally, it may be necessary to add bone to build up the cheek bones. In our center, we believe that it is always best to a patient's own bone in lieu of artificial material. A second operation involving the jaw may be necessary to correct either an undergrowth or overgrowth of the rib graft in the jaw. Finally, patients with Craniofacial Microsomia may need surgery to correct the flattened forehead and to re-position the eye.

**Where should I go to seek treatment for my child?** Your child should be treated by a qualified craniofacial team. Hemifacial Microsomia 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. The basic team members and their roles in the treatment of your child 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 Hemifacial Microsomia or Goldenhar syndrome.

**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 Hemifacial Microsomia or Goldenhar syndrome 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.

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