

a guide to understanding
**hemifacial
microsomia**

a publication of children's craniofacial association

a guide to understanding hemifacial microsomia

this parent's guide to hemifacial microsomia is designed to answer questions that are frequently asked by parents of a child with hemifacial microsomia. 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 hemifacial microsomia?

hemifacial microsomia is a condition that simply comes “out of the blue.” It does not run in families and is not the result of a disease process. Simply stated, it is a birth defect. Some cases of hemifacial microsomia are severe; therefore, the degree of deformity is considerable. Other cases are mild with a wide spectrum of manifestation. A doctor or medical team makes the clinical diagnosis. No DNA has been identified in the condition.

how do I recognize this condition in my child?

hemifacial microsomia involves the entire face. The skull may be underdeveloped on the affected side and the orbit, or eye socket, can be small. There may be a small non-seeing eye or the eye may be entirely absent. The eye may show a range of problems—from normal to absent. There may also be eye abnormalities including dermoids, or notches in the eyelid. The cheek is flat because the bone beneath has not grown properly. The face is vertically short in this area. The mandible, or lower jaw, is involved in a similar way. The mandible can be underdeveloped or a portion of the mandible can be missing, usually vertically. The ear may be normal, but it may be underdeveloped or even absent. Hearing is defective. There may be skin tags in front of the ear or in a line between the ear and the corner of the mouth.

are there different degrees of this condition?

Craniofacial microsomia is a variant of hemifacial microsomia. In addition to the things normally seen in hemifacial microsomia, the forehead is flattened and the eye may be pulled downward. Another condition, Goldenhar syndrome, is very similar; however, children with this syndrome also have benign growths on the eye. The growths are called epibulbar dermoids. Patients with Goldenhar syndrome may also have neck problems. A fusion, or bony bridges between the bones of the neck, causes the most common problem. Goldenhar syndrome is also called oculoauricular dysplasia or OAV.

what treatment is available for hemifacial microsomia?

This condition is a very complex, three-dimensional deformity. Due to this complexity, the correction is difficult and can rarely be achieved in one stage. If the skull and the skull base are underdeveloped and asymmetrical, then intracranial surgery may be necessary to enlarge the skull by putting the bones in the correct position. A neurosurgeon helps with this procedure. Usually at this time, it is possible to enlarge the orbit and to reconstruct the zygomatic arch, or cheek bones. The operation can be performed with either bone from the skull or from elsewhere. This reconstruction can be done at an early age or when the patient presents for treatment.

Before the child goes to school, cutting the upper jaw transversely, or crosswise, can lengthen the face. This is called an osteotomy and allows the deformed side to be lengthened. A bone graft is put into the gap. At the same time, the lower jaw can be altered to lengthen it. This brings the teeth down into the

correct plane and allows them to meet the teeth of the upper jaw correctly. If there is no joint and the vertical part of the lower jaw is missing, then it can be replaced with a rib graft, which has cartilage on its upper surface. This allows the doctor to form a joint. If the tissue of the face is underdeveloped, it can be replaced by skin and fat. Sometimes muscle is used, which is taken from another part of the body. This procedure is called a free flap. This tissue has its own blood vessels, and these vessels can be joined to other blood vessels in the neck so that the tissue is kept alive.

After this work is done, there will be other small operations to revise scars and to shift the soft tissue around to obtain a better contour or shape. It may be necessary to bone graft various areas. The bone grafts are taken from the skull or the hip. If there is no eye, it may be necessary to make an eye socket for an artificial eye.

what other problems and treatments might we expect?

Children with this condition require long-term follow-up. Several operations may be needed over the period of facial growth, and often there is a tendency for poor growth on the affected side. When there is no eye, the ocularist will make an artificial one. When the teeth do not meet properly, an orthodontist will plan and carry out appropriate orthodontic treatment. For some aspects of jaw surgery, an oral and maxillofacial surgeon will be consulted. If there are problems with the ear, such as hearing defects, the neuro-otologist will examine the child and advise as to procedures for restoring hearing. The goal is for normal hearing, which is possible in most cases.

Another problem with this condition is the absence of the external ear. There are two methods for dealing with this. One

method, called osseointegration, involves placing small metal studs into the bone in the ear region. A framework is then attached to the studs and a silicone ear, a replica of the normal ear, can be clipped onto this framework. These are usually very secure and provide a very realistic reconstruction. This ear obviously has no feeling and it is possible for this ear to detach and become lost. An example might be while swimming. The ear color may fade with time, but this can be corrected. The ear may have to be altered as the child grows.

An alternative method is to make an ear from rib cartilage. This is a staged procedure, in which a cartilage framework is carved according to the pattern designed from the other ear. Then it is placed under the skin on the side of the head. After a length of time determined by the surgeon, the ear is elevated and another skin graft is placed behind it. Frequently, it takes several operations to create an ear that is cosmetically acceptable and that matches the normal ear. It is important to understand that this ear may not be an accurate match to the other ear. It may vary in color and will have no normal sensation. However, it is more convenient than a prosthetic ear.

There are often abnormalities of the internal ear, and the ear canal can be completely absent. There may be varying degrees of deafness. Neuro-otologists, specialists in this area who assess all children with hearing problems, have made great advances in the treatment of deafness.

Although children with hemifacial microsomia often look different, it is important to mention that most of these children are of normal intelligence and function completely normally. They may experience learning difficulties. A common difficulty is a language problem due to deafness. It is important for parents to realize the rehabilitation will take a long time. The complexity of the deformity is the cause of the lengthiness. Another reminder is that as the child grows, changes occur, and these changes may make further surgical procedures necessary.

where is the best place to have my child treated?

hemifacial microsomia is a complex condition. It requires the expert skill of several different specialties working together. Craniofacial teams experienced in the management of these patients best treat these conditions. Centers with 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. As there are only a few experienced centers in the country, it is quite common for families to travel quite a distance to get the best care. Children who are treated locally by inexperienced teams or by individual physicians not working together as a team, are more likely to have unsatisfactory results. It sometimes requires two or three additional operations to correct what has been done. Another advantage of traveling to larger centers is the opportunity to meet other families and children affected with similar problems who can offer advice. These families often share their experiences, which provides moral support.

how can children's craniofacial association (cca) benefit my family?

CCA understands that when one family member has a craniofacial condition, each person in the family is affected. We provide programs and services designed to address these needs. A detailed list of CCA's programs and services may be found on our Web site at www.ccakids.com or call us at 800.535.3643.



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