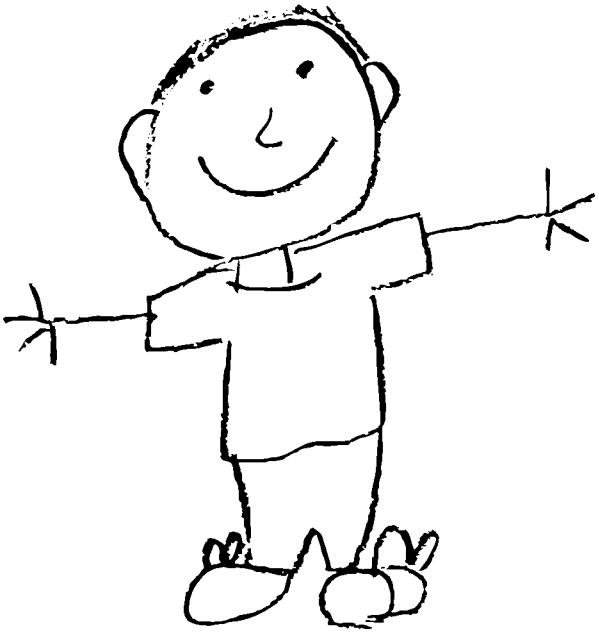
# Craniosynostosis & Craniofacial Surgery

## A Parent's Guide

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# Introduction

Parents of children with *craniofacial anomalies* (disorders of the skull and face) have many questions about the cause, nature, and treatment of their child's disorder. This booklet provides parents of children with craniofacial disorders a summary of various anomalies and the possible surgical means of treating these problems.

The skull is important because it houses and protects the brain. Several pieces of bone make up the skull. Loose connections called *"sutures-* connect the bones. The hard bones of the skull protect the brain. The sutures between the bones allow the baby's brain to grow. (See Figure 1).

Much of the growth of the brain happens by three years of age. Then, the baby's brain and skull grow slowly in size until about six years of age, when most of the sutures heal shut permanently, or *'fuse."*

Sometimes the sutures fuse too soon, while the baby's brain and skull are still growing. Specialists refer to this condition as *"craniosynostosis."* This can cause an unusual head shape, unusual facial features, and in some cases can cause damage to the brain because of increasing pressure inside the skull.

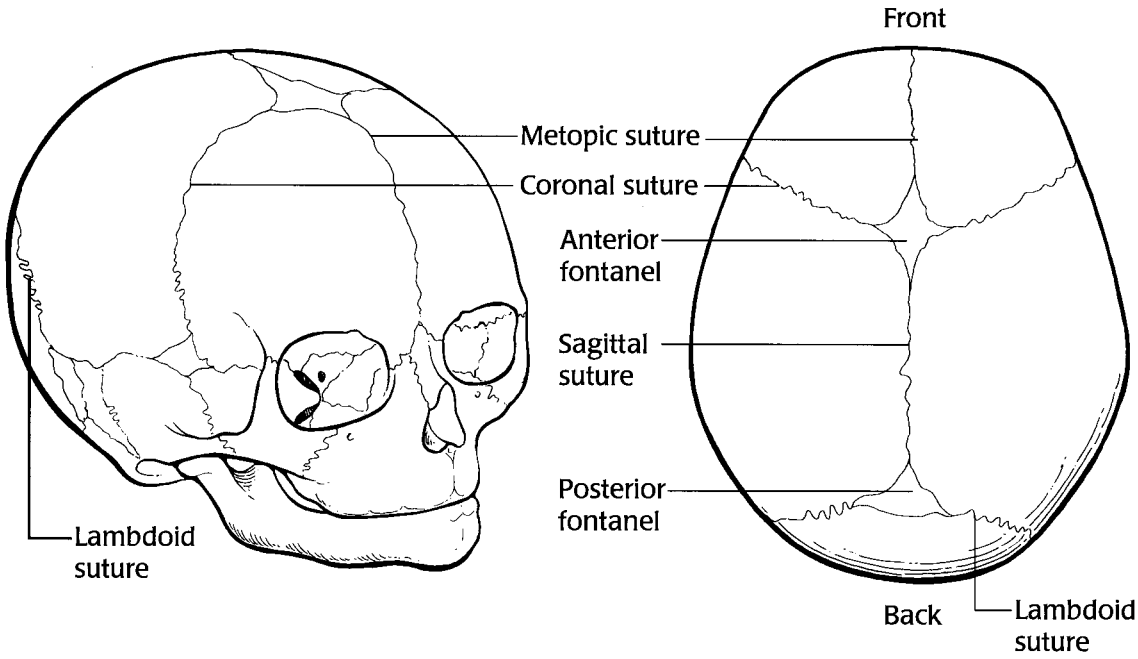


**Figure 1.** *Major cranial sutures.*

When a suture closes too soon in life, the skull can grow in an abnormal manner. This changes the shape of the skull, face, orbits (eye sockets), and jaw. (See Figure 2). Craniosynostosis may cause damage to the growing brain because there is not enough room for it to grow inside the skull. This can lead to developmental delay, brain damage, and damage to vision. The incidence of craniosynostosis is about 1 in 2,000 births.

Some craniofacial anomalies run in families. in most cases, however, specialists cannot find a cause. Often, a craniofacial anomaly is apparent at birth. Sometimes, the problem appears later in infancy and childhood. Genetic counseling may be necessary to diagnose your child's condition and to determine if it is an inherited condition (passed on from parent to child). The geneticist can also help a family in making decisions about having more children.

It may be difficult to diagnose your child's problem. You may need to bring your child to a major center where there are specialists in Neurosurgery (surgery of the brain) and Craniofacial surgery (surgery of the bones of the skull and face), among others. This team of specialists will plan your child's care, and provide follow up on a regular basis.

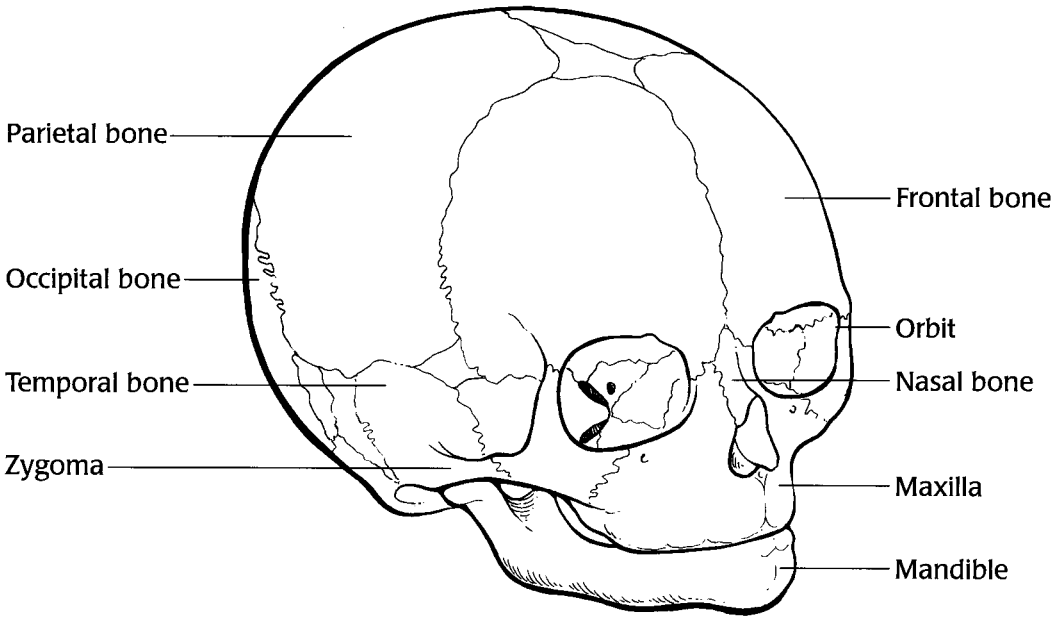


***Figure 2.*** *Bones of face and cranial vault.*

# Test and Procedures

Many tests may be necessary in order to diagnose your child's condition and make a treatment plan for a specific condition.

*Skull X- rays* may be the first step in determining if a child has a craniofacial anomaly. The physician may request this test because of a noticeable abnormality in the shape or size of the baby's head.

*A Computerized Tomography Scan (CT or CAT Scan)* may confirm the presence of prematurely fused sutures, and other brain abnormalities, and may determine the presence of increased pressure in the brain. *A CT* scan of the brain and skull is an X-ray of 'slices' of the brain and skull. When stacked up, these images can create a two-dimensional view of the bones of your child's skull. This study helps the surgeon diagnose problems and to plan surgery. In order to hold still for the CT scan, your child may need sedation or even general anesthesia during the test.

*A Magnetic Resonance Imaging Scan (MRI Scan) is* a high-powered magnetic wave machine. This scanner, together with a computer, produces images of the soft tissues (such as the brain) of the body. *As* with the CT scan, children may need sedation to undergo this test.

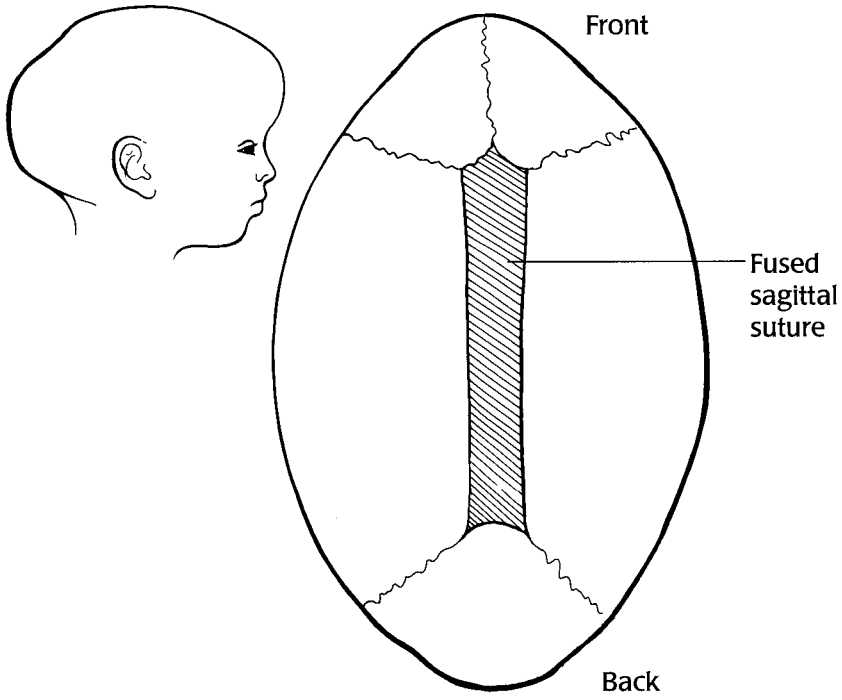


***Figure 3.*** *Scaphocephalic head viewed from above.*

# Sagittal Synostosis

Sagittal Synostosis is the most common type of suture synostosis. In this type, the suture that runs from the front to the back fuses prematurely. This causes a condition which specialists refer to as "scaphocephaly" or "boat-shaped skull (See Figure 3). The skull becomes long from front to back, and appears narrow when viewed from the front. There may be a prominent ridge along the top of the head extending from the "soft spot' to the back of the head. The forehead may also be too prominent.

If specialists diagnose this condition early in life, the surgeon performs an operation called a "Sagittal Synostectomy." In this operation, the neurosurgeon removes the sagittal suture and places cuts in the side bones of the skull, allowing for proper growth of the brain and skull, and a normal cosmetic appearance. If doctors do not diagnose this condition in infancy, the child may need a more complex operation. This operation is called a total cranial vault reshaping (See Figure 4).

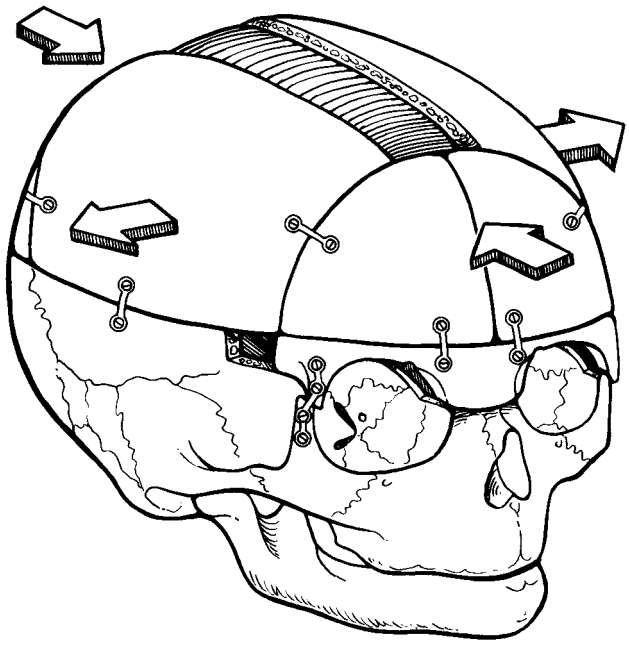

***Figure 4.*** *The surgeon reshapes the cranial vault, allowing for proper growth of the brain and skull, and a normal cosmetic appearance.*

# Coronal Synostosis

*Coronal Synostosis is* the second most common type of suture synostosis. *Bilateral* (occurring on both sides of the head) coronal synostosis is common in many inherited craniofacial syndromes. In coronal synostosis, the coronal suture (the suture that is from ear to ear across the top of the head) heals prematurely on both sides of the head (See Figure 5). This leads to a condition which specialists call *"brachycephaly"* or a wide shaped head. in bilateral coronal synostosis, the forehead is flat, broad, and too tall. An operation is necessary to allow the brain room to grow, and to reshape the skull.

Babies with coronal synostosis must see a neurosurgeon and craniofacial surgeon to plan for surgery. The goal of surgery is to open the prematurely fused sutures, reshape the forehead and allow for normal brain and skull growth. In this operation, called a *Fronto-orbital advancement,* the surgeon accesses the bones of the skull through an incision that goes from ear to ear across top of the head. The surgeon removes the bones of the forehead, reshapes the bones, and puts them back in place with small plates and screws that are permanent (See Figure 6).

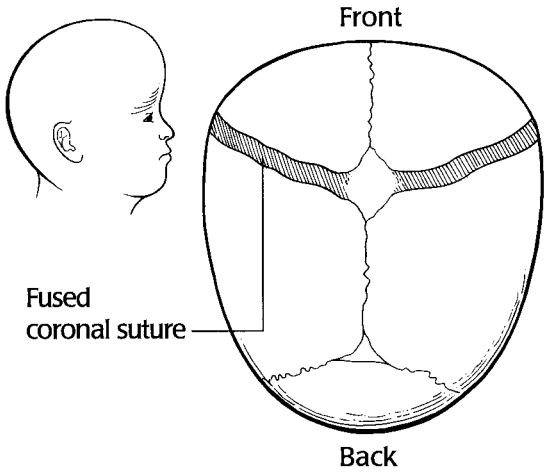


***Figure 5.*** *Brachycephalic head viewed from above. A brachycephalic head is flatter and taller than normal.*

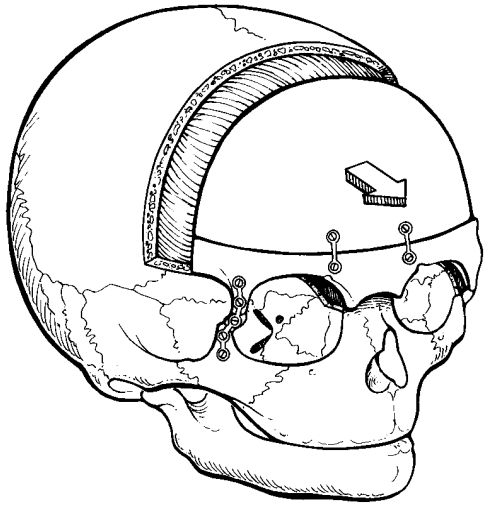

***Figure 6.*** *The surgeon removes the bone of the forehead and advances the forehead to allow for growth of the brain, and to allow the skull to resume a normal shape.*

In *unilateral coronal synostosis,* the coronal suture on one side of the head fuses prematurely (See Figure 7). This leads to a condition which specialists refer to as *"plagiocephaly"* or skull asymmetry. In this disorder, the forehead on the involved side appears flat, and the opposite side may bulge out. The bony rim above the eye on the involved side is flatter and may look higher than that on the uninvolved side. The nose points away from the fused suture. An operation is necessary to allow the brain room for growth, and to restore normal shape to the forehead. If left untreated, this condition causes a skull deformity that will get worse over time. It also causes facial disfigurement, and could in rare cases cause brain damage. Babies with unilateral coronal synostosis should see a neurosurgeon and craniofacial surgeon to plan for surgery. The goal of surgery is to open the prematurely fused suture, allow for normal forehead shape, and allow for normal brain growth. In this operation, the surgeon accesses the bones of the skull through an incision that goes from ear to ear across the top of the head. The surgeon removes the bones of the forehead, reshapes and advances the bones with small plates and screws that are permanent (See Figure 8).

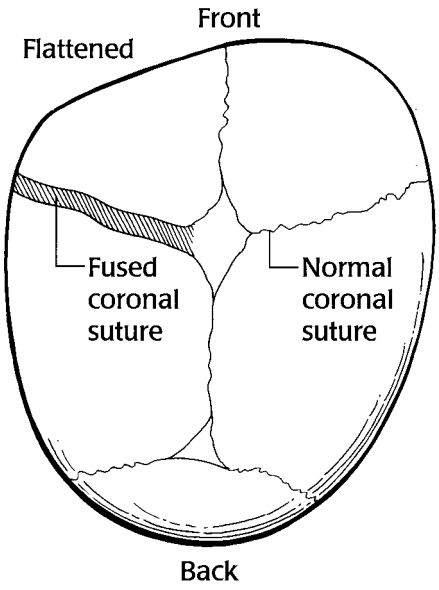


***Figure 7*** *Plagiocephalic head viewed from above.*

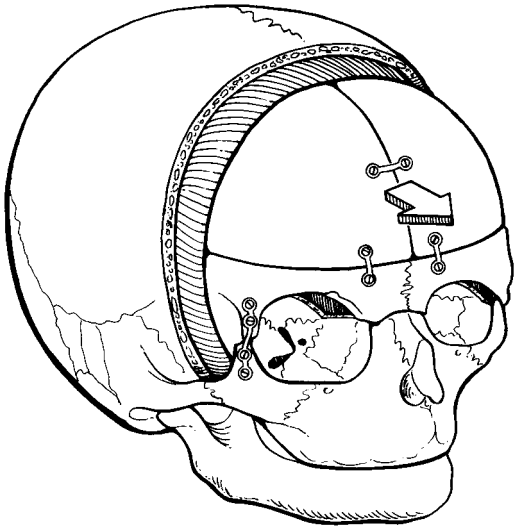

***Figure 8.*** *The surgeon removes the bone of the forehead and advances the forehead to allow for growth of the brain, and to allow the skull to resume a normal shape.*

# Metopic Synostosis

*Metopic synostosis is* the premature fusion of the suture in the middle of the forehead. This causes a condition which specialists call *"trigonocephaly,* "or triangle shaped head (See Figure 9). This condition may be an inherited trait. In this case, the forehead is narrow, the temples appear pinched, and the eyes are too close together (hypotelorism). There may be a noticeable ridge in the middle of the forehead. A child with this condition may require surgery to correct the shape of the head, or to prevent it from getting worse. A child would also need surgery if there were too much pressure on the brain.

Children with metopic synostosis should see a neurosurgeon and craniofacial surgeon to plan for surgery if needed. The goal of surgery is to open the prematurely fused sutures, to allow for normal brain growth, and to restore the normal shape of the forehead. In this operation, the surgeon accesses the bones of the skull through an incision that goes from ear to ear across the top of the head. The surgeon removes the bones of the skull and forehead, reshapes them, and puts them back in place with small plates and screws that are permanent (See Figure 10).

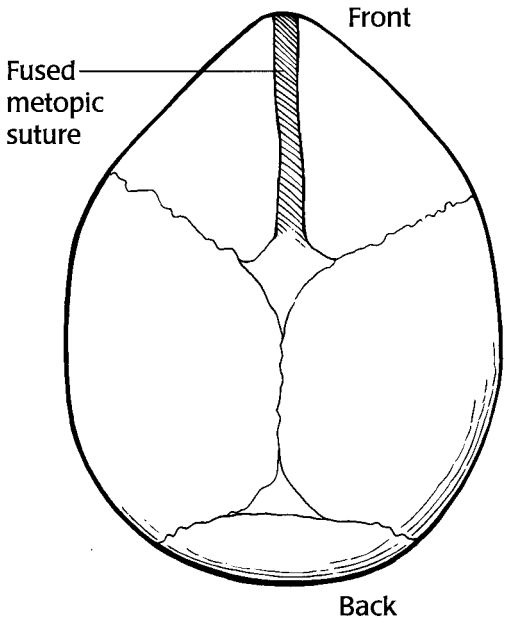


**Figure 9.** Trigonocephalic head viewed from above.

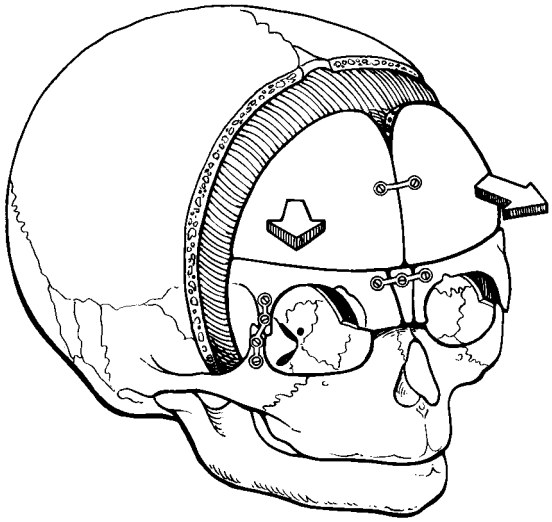

***Figure 10.*** *The surgeon removes the bone of the forehead and advances the forehead to allow for growth of the brain, and to allow the skull to resume a normal shape.*

# Surgical Repair of the Skull and Face

Often times it is necessary to surgically treat the problems of the bones of the skull and face to improve a child's appearance and to prevent possible pressure on the brain, which can cause headaches, irritability, decreased mental capacity, and visual changes. The goals of surgery are to relieve pressure and to allow for normal growth of the skull and face.

Before surgery, a team of specialists, including plastic surgeons and neurosurgeons, will evaluate your child. Your child will undergo tests, such as a CT Scan, X-rays, and a physical exam. Your child may need blood work such as a hemoglobin, hematocrit, and a test of the blood type. A photographer will take photos in order to document the deformity and measure progress. You will also take a tour of the recovery room and pediatric intensive care unit.

You can help to prepare your child for the recovery period by bringing a favorite toy, pillow or book to the hospital. In addition, you can bring tapes of familiar music, or tapes of favorite books or stories to make the hospital stay more comfortable.

Because of the possibility of blood loss during an extensive procedure, your child may need a blood transfusion. If there is a chance that your child may need a transfusion, the surgeon or family may request "Directed Donation" (the donation of blood by a friend or family member). You can arrange this procedure through your local Red Cross. You may also choose to have the surgeon use blood from the hospital's blood bank.

# Postoperative Care

Your child may spend several days in the pediatric intensive care unit after surgery. Here, specialists will monitor your child's vital signs, neurological, and respiratory status. Once your child is stable, he will continue to recover on the general care unit. Your child's progress will determine the length of the hospital stay. Usually, a child spends five to seven days in the hospital after the procedure. Once the child is able to take feedings and has no sign of infection and some of the swelling has decreased around the eyes, you will be able to take your child home from the hospital.

**Appearance**

After surgery, your child will have an incision across the top of the head. Hair will eventually hide this scar. Initially, your child will have a head dressing. Before discharge home, doctors will remove the dressing. Your child will have swelling of the face and eyes. The child's eyes will be swollen shut for several days. Your child may have one or more IV tubes for fluid and medication after surgery. It may be necessary to keep the breathing tube for a day or so after surgery, until your child is stable and breathing on his own. While in the pediatric intensive care unit, heart rate and breathing monitors will also be used.

**Medications**

You may need to put an antibiotic ointment on your child's incision. Postoperatively, your child may have some pain. Your child will receive a prescription for pain medication after the operation and before you go home. Tylenol® or lbuprofen® are generally adequate for pain relief after a few days at home.

**Diet**

Whether it be drinking from a cup or using a bottle, once your child's respiratory status is stable, doctors will encourage fluids. Your child will start taking fluids by mouth the day after surgery, if vital signs are stable. Your child will not require a special diet after surgery. Generally, your child will be able to eat in the same manner as preoperatively.

**Activity**

When you go home, doctors will restrict your child from engaging in any rough activity. When the surgeon puts the bones of the face and skull in the correct position, they are held together with permanent metal plates and screws. However, it takes six to eight weeks for these bones to heal together properly. These plates and screws are not visible to the naked eye, but are visible on X-ray. The plates and screws are permanently left in place, unless there is a problem such as infection, pain, or dislodgment.

**Monitoring**

You will need to monitor your child for signs and symptoms of infection, such as redness of the incisions, pus or other drainage, swelling or fever. Your child will need yearly follow up, until eighteen years of age, when facial and dental growth are complete.

**Psychological Adjustment**

In the first few days after surgery, you may see emotional changes in your child that are normal responses to any hospitalization. Your child may be unusually "fussy' or "clingy.' You may see changes in sleep patterns, such as difficulty in getting to sleep or waking in the night. You may see some -regression.' For instance, if your child was potty trained prior to surgery, he may need diapers for a short time after surgery. Generally after a few days at home, children begin to adjust and progress as they had before surgery.

**Resources**

Emotional and financial resources are available for the treatment of a child with a craniofacial anomaly. Private health insurance will usually cover at least a portion of the cost of the child's care. Your child may be eligible for Children's Special Health Care Services, which in some instances, provides financial assistance for the medical care of children under 21 years of age with congenital or acquired physical conditions. The team social worker will be able to help you determine what types of assistance are available.

**Conclusion**

The craniofacial team will follow your child on a yearly basis. The Craniofacial Anomalies Program at the University of Michigan Medical Center began more than 70 years ago. Since then, we have become one of the largest programs in the country. For information regarding our program, please write: Craniofacial Anomalies Program University of Michigan Medical Center 1500 East Medical Center Drive 7859 Mott Hospital Ann Arbor, Michigan, 48109-0219 or call 1 **(800) U. OF M. CAP (863-6227).** Also, you may call Pediatric Plastic Surgery at (734) 763- 8063, or Pediatric Neurosurgery at (734) 936-5016. Our team has a great deal of experience in taking care of children with craniofacial anomalies. The outlook for these children in terms of appearance, function, and psycho-social wellbeing is excellent.

# Definitions

• ***Anomaly:*** A disorder that a person is born with.

• ***Bilateral****:* Pertaining to both sides of the body.

• ***Brachycephaly****:* A wide shaped head caused by premature fusion of the coronal sutures.

• ***Coronal Suture:*** The I line of junction between the frontal and two parietal bones.

• ***Craniofacial:*** Craniofacial refers to the bones of the skull and face.

• ***Craniosynostosis:*** The premature fusion of the bones of the skull.

• ***CTScan:*** (Computerized Axial Tomography Scan) An X-ray picture that shows "slices' of a portion of the body.

• ***Fronto-Orbital Advancement:*** An operation in which the bone of the forehead is removed and the forehead is advanced to allow for growth of the brain, and for the skull to resume a normal shape.

• ***Fusion:*** Healing shut; establishing a bony union.

• ***Geneticist:*** A physician specializing in the diagnosis of inherited disorders.

• ***Hypotelorism****:* An abnormally small distance between the eyes.

• ***Hydrocephalus****:* An accumulation of cerebral-spinal fluid in the brain, causing pressure in the brain.

• ***Intra Cranial:*** This refers to the space within the skull.

• ***Lambdoid Suture:*** The line of junction between the occipital and two parietal bones.

• ***Malocclusion:*** A condition in which the upper and lower teeth are not properly positioned in relationship to one another. *Maxilla.* The bone that makes up the upper jaw.

• ***Metopic Suture:*** The suture that runs up and down in the middle of the forehead. *MRI Scan:* Magnetic resonance imaging scan. An X-ray of a portion of the body taken using a highpowered magnet and a computer. *Occiput:* The back part of the head *Orbit:* The bony cavity that contains the eyeball. *Plagiocephaly:* Skull asymmetry.

• ***Sagittal Suture:*** The suture that runs down the middle of the head from the back to the front.

• ***Scaphocephaly:*** Condition where the skull is long and narrow from front to back.

• ***Sutures:*** The open, fibrous areas that join the bones of the skull.

• ***Synostosis:*** Union between the bones.

• ***Trigonocephaly:*** Triangle shaped forehead.

• ***Unilateral****:* Pertaining to one side of the body.

• ***Zygoma:*** Cheekbone.

# Acknowledgments

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