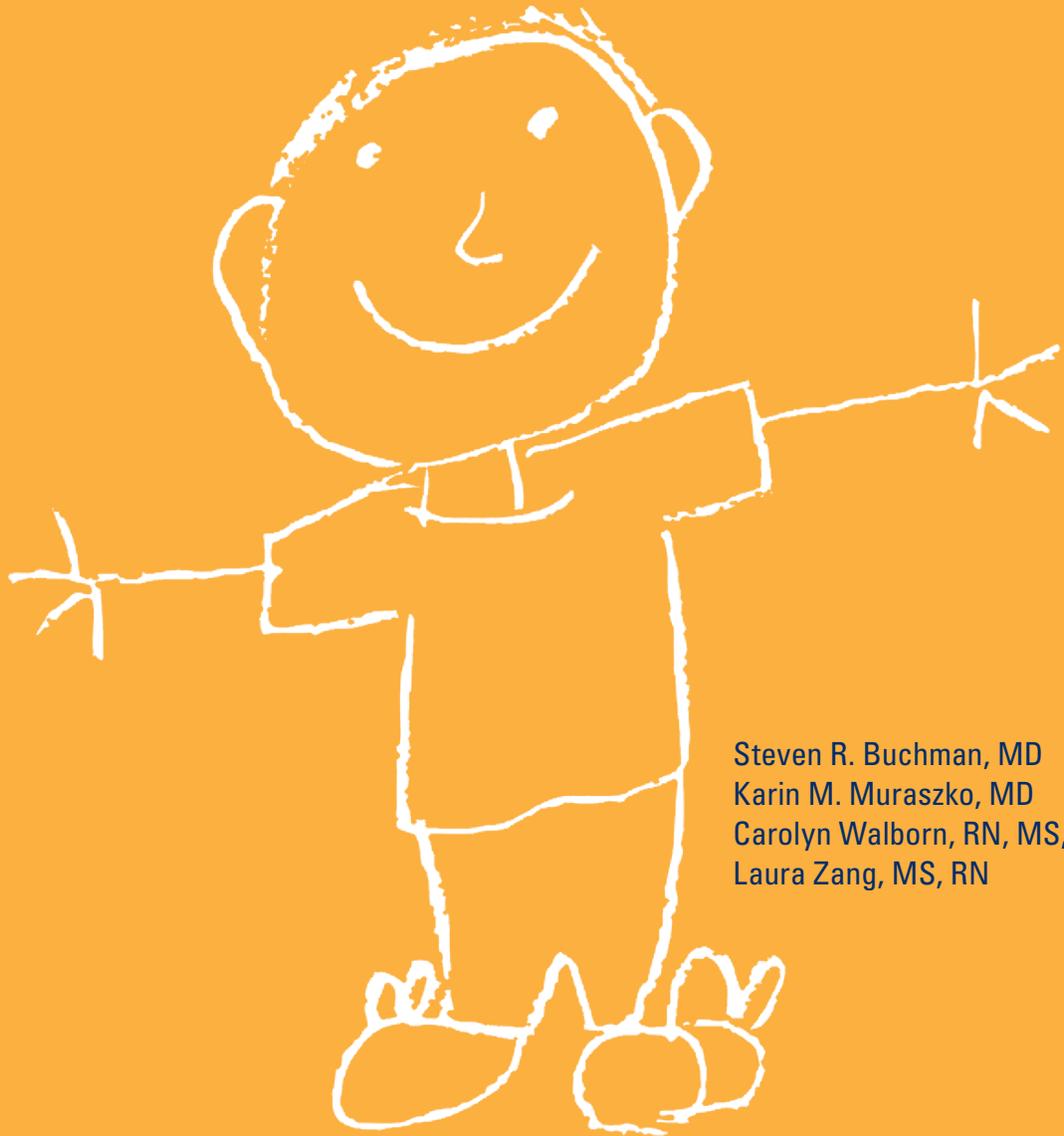


Craniosynostosis & Craniofacial Surgery

A Parent's Guide

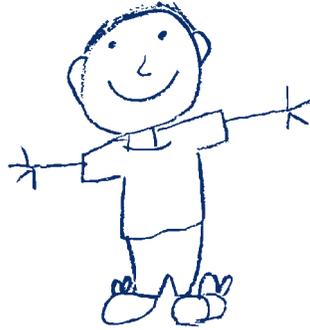


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C.S. Mott Children's Hospital
Craniofacial Anomalies Program



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What is Craniofacial Surgery?

Craniofacial surgery is a special type of surgery developed over 30 years ago. It involves surgery of the face and skull for reasons such as tumors, trauma and deformities such as birth defects. This type of surgery is performed by highly trained surgeons who are supported by a highly skilled team. The surgical team includes a pediatric neurosurgeon and a pediatric craniofacial surgeon. The neurosurgeon works on and around the brain while the craniofacial surgeon works on the skull and face. This combined approach offers the highest level of safety with the best possible results.

The Craniofacial Anomalies Program at the University of Michigan is one of the largest programs of its type in the country. Our multidisciplinary program is a group of experienced professionals with diverse expertise who collaborate to provide the highest quality, family-centered care for each child. The outlook for these children in terms of appearance, function and psycho-social well-being is greatly improved by this innovative approach.

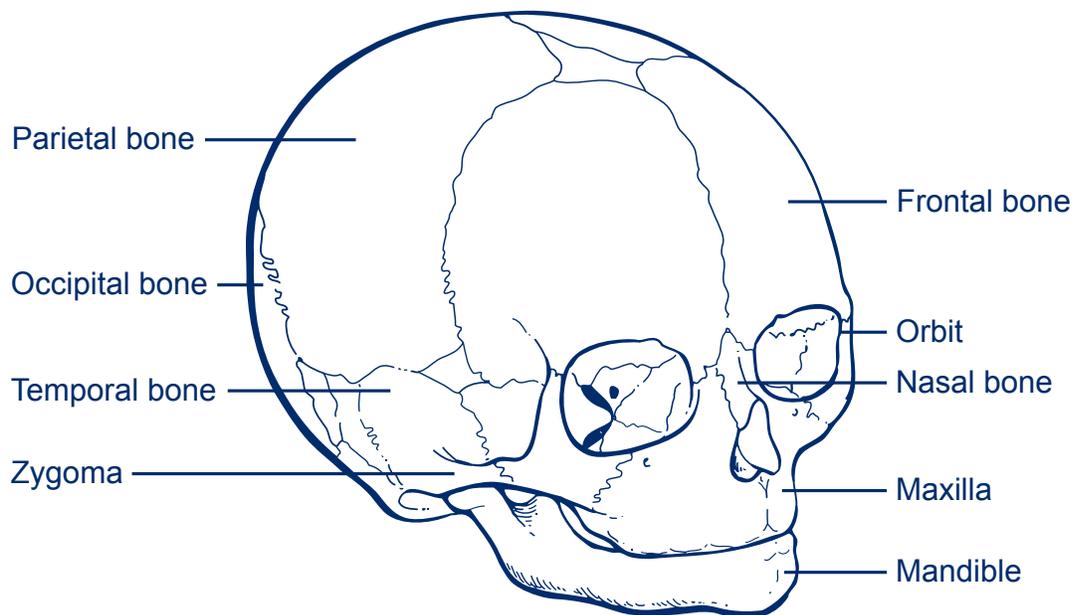


Figure 1. Normal bones of the face and cranial vault.



What is Craniosynostosis?

The skull is important because it holds and protects the brain. Several pieces of bone make up the skull (Figure 1). These pieces of bone are held together by loose connections called **sutures** (Figure 2). The sutures between the bones allow the skull to grow as the child's brain grows. Sometimes a suture will close or **fuse** too early. When one or more of the sutures of the skull fuse too early it restricts skull growth. This is called **craniosynostosis**. Craniosynostosis can cause unusual head shape, unusual facial features and in rare cases, damage to the brain due to increased pressure inside the skull. There are many different types of craniosynostosis. Different names are given to different types depending on which sutures are involved.

The cause of craniosynostosis is unknown. It occurs in about 1 in every 2,000 to 3,000 births. Craniosynostosis most often occurs by chance but sometimes can be passed on from parent to child (inherited). When craniosynostosis is inherited it often occurs as part of a genetic syndrome such as Aperts Syndrome or Crouzon's Syndrome. This type of craniosynostosis is much less common. Your surgeons may suggest that your child see a genetics doctor, (**Geneticist**) to help determine the cause of craniosynostosis and/or any other possible associated diagnosis. This information will also be helpful as the child grows older and thinks about having their own family. Sometimes craniosynostosis is diagnosed at birth, other times it appears later on in infancy.

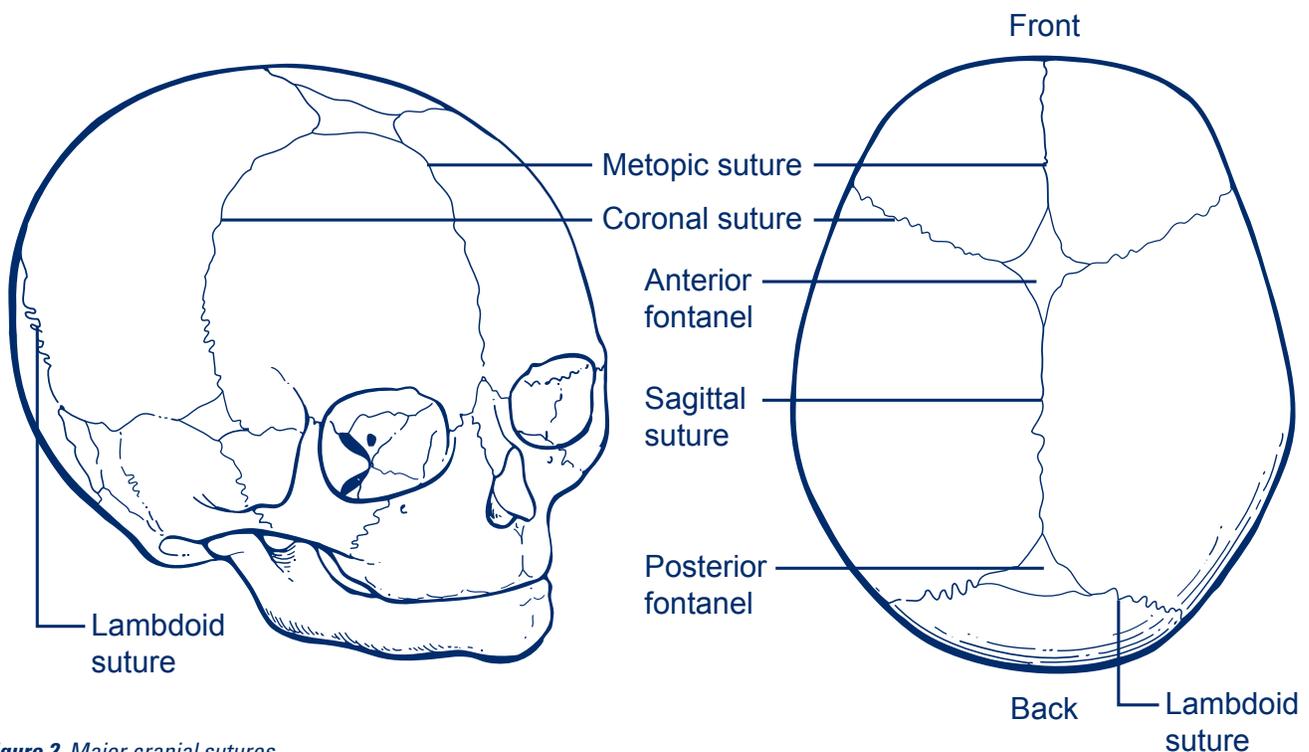


Figure 2. Major cranial sutures.



Tests and Procedures

Many tests may be necessary in order to diagnose your child's condition and make a treatment plan. Every child is different and specific treatment plans and operations will vary with the type of problem your child has, the age of your child, as well as several other factors. Your doctor will feel your child's head for bumps or **suture ridges** while also looking for facial deformities. If a problem is suspected, several tests may be ordered.

CT scan (Computerized Tomography scan):

A CT scan of the brain and skull creates a two or three dimensional view of the bones of your child's skull. A CT scan may confirm the presence of fused sutures, brain abnormalities and increased pressure in the brain. Infants and children need to stay very still during this procedure and most often need general anesthesia for a CT scan.

MRI scan (Magnetic Resonance Imaging): A MRI is a noninvasive procedure that uses a powerful magnetic field and a computer to produce detailed pictures of organs, soft tissues, bone and internal body structures. Infants and children need to stay very still during this procedure and most often need general anesthesia for a MRI scan.

Genetic testing: Sometimes skull deformities can be caused by an underlying hereditary syndrome or a specific genetic defect. These genetic syndromes can have different patterns of inheritance and different chances for reoccurrence. Genetic tests can help identify syndromes and causes. Tests sometimes require a blood sample which is sent to a special lab for analysis.

Neuropsychological testing: These are tests developed by a psychologist in order to evaluate a child's development in many areas. Although many children with craniofacial deformities do not have any developmental difficulties, a number of craniofacial conditions, such as craniosynostosis do include risks for certain learning and thinking skills as well as developmental delays.

Ophthalmology evaluation and testing: Your doctors may suggest that your child have a complete eye exam (ophthalmologic exam) by an eye doctor (ophthalmologist). Sometimes infants and children with skull abnormalities may develop increased pressure inside their brain. Their skulls don't expand enough to make room for their growing brain. An eye doctor will check for **papilledema** to help determine if there is any increased pressure inside your child's brain.



Sagittal Synostosis

Sagittal synostosis is the most common type of craniosynostosis. In this type, the suture that runs from front to back, down the middle of the head, closes too early. This causes your child's skull to become long and narrow. This is sometimes called "**scaphocephaly**" (Figure 3). Your child's head may appear narrow when viewed from the front and long when viewed from above. They may also have a large forehead that may or may not stick out.

Your neurosurgeon and plastic surgeon will perform an operation called a **cranial vault reshaping** to repair your child's skull (Figure 4). A wavy incision is made over the top of your child's head from ear to ear (Figure 5). The neurosurgeon removes the skull and dissects the bone from the brain. The plastic surgeon makes cuts in the bones of the skull to reshape the head. The bones are sometimes held in place with absorbable plates or sutures.

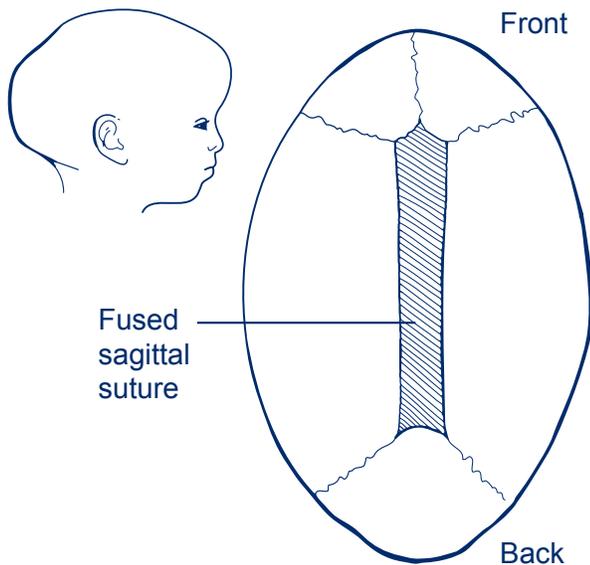


Figure 3. Scaphocephalic head viewed from above.

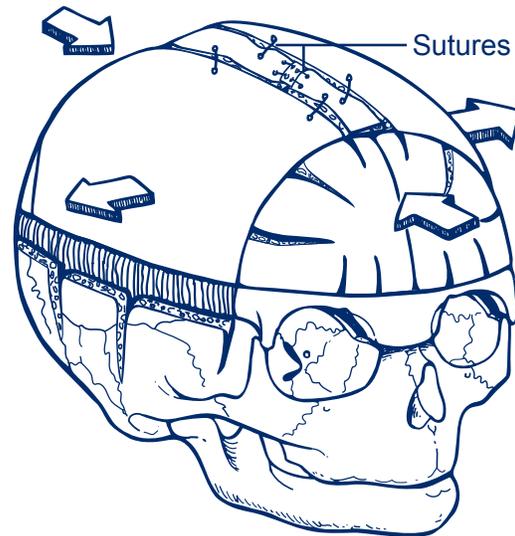


Figure 4. Cranial vault reshaping: the surgeon reshapes the cranial vault allowing for proper growth of the brain and skull and a more normal appearance.

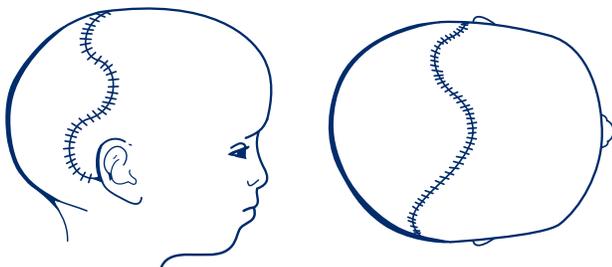


Figure 5. Wavy incision across top of head from ear to ear for craniosynostosis surgery. Hair will eventually hide much of this scar.



Coronal Synostosis

Coronal synostosis is the second most common type of synostosis. This occurs when both sutures that run across the top of the head from ear to ear are closed (**bilateral**). This gives an infant a wide head with a forehead that is flat and too tall. Your doctor or surgeon will call this shaped head "**brachycephaly**" (Figure 6). Sometimes coronal synostosis can occur only on one side of the head (**unilateral**). When this happens, your child's forehead on the affected side appears flat and the other side may appear to bulge out. This can also cause the eyes to look different and cause the nose to point away from the flat side. This head shape is often referred to as "**plagiocephaly**" (Figure 7).

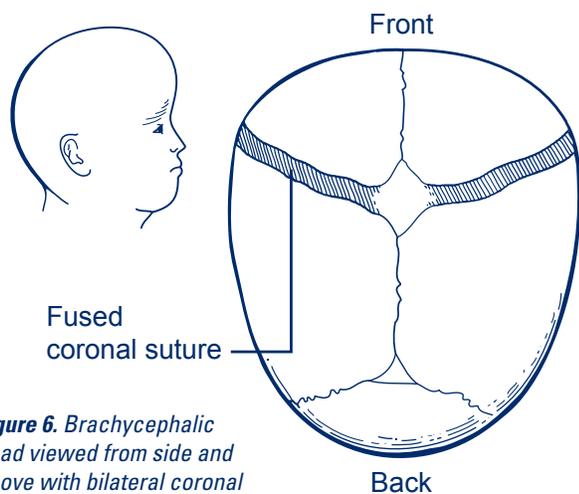


Figure 6. Brachycephalic head viewed from side and above with bilateral coronal synostosis.

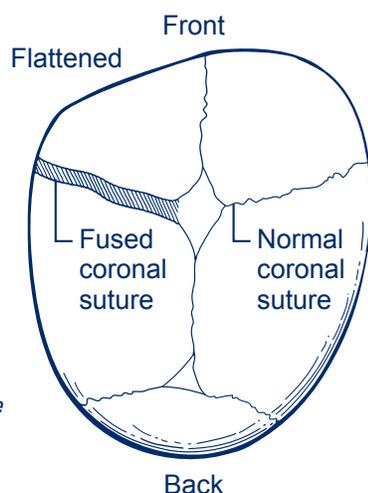


Figure 7. Plagiocephalic head viewed from above with unilateral coronal synostosis.

Surgery is required to open the fused sutures, reshape the head and allow for normal brain and skull growth (Figure 8). A wavy incision is made over the top of your child's head from ear to ear. The neurosurgeon removes the skull and dissects the bone from the brain. The plastic surgeon removes the bones of the forehead, reshapes them and puts them back in place with absorbable plates.

After surgery to repair coronal synostosis it may seem that the forehead protrudes or sticks out more than it should. This is normal and called **overcorrection**. Overcorrection is necessary to make the forehead look more natural over time. The forehead bones naturally recede or become pulled back toward their original position from the tension of the overlying skin and scalp tissue. The surgeons intentionally overcorrect so that natural movement back returns the forehead to a more natural shape. This often takes 6 to 18 months. Try not to be alarmed when you see overcorrection in your child's forehead.

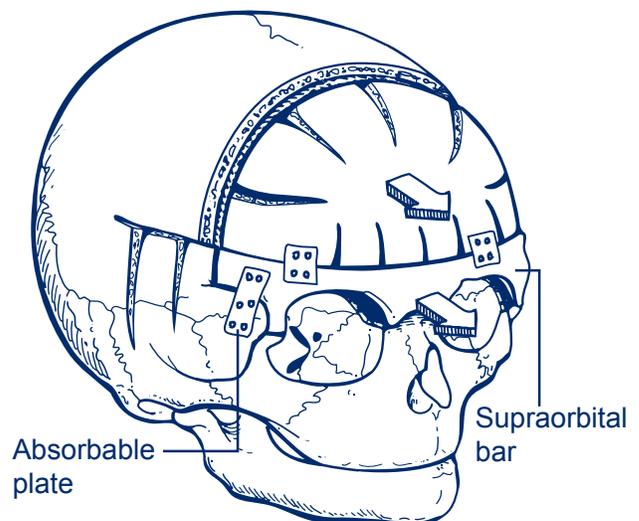


Figure 8. Frontal orbital advancement: the surgeon removes the bones of the forehead as well as supraorbital bar and advances the forehead to allow for growth of the brain and to allow the skull to resume a more normal shape.



Metopic Synostosis

Metopic synostosis occurs when the suture in the middle of the forehead closes too early. This gives an infant a forehead that often looks pointed or triangular from above. The sides of the forehead appear pinched causing the eyes to look like they are too close together (*hypotelorism*). Sometimes you may be able to feel a ridge in the middle of the forehead. Your doctor or surgeon may call this type of head shape “*trigonocephaly*” (Figure 9).

Surgery is required. The goal of the surgery is to open the suture that is closed and to restore the natural shape of the forehead (Figure 10). A wavy incision is made over the top of your child’s head from ear to ear. The neurosurgeon removes the skull and dissects the bone from the brain. The plastic surgeon removes the bones of the forehead, reshapes them and puts them back in place often with absorbable plates.

After surgery to repair metopic synostosis it may seem that your child’s forehead protrudes or sticks out too much, especially at the outer edges. Just as in coronal synostosis mentioned previously, this is done on purpose by the surgeon and called *overcorrection*. Overcorrection is necessary to make the forehead bone look more natural over time. After surgery, forehead bones will recede or move back toward their original position from the tension of the overlying skin and scalp. The surgeon intentionally overcorrects so that the movement back returns the forehead to a natural shape. This often takes 6 to 18 months. Try not to be alarmed when you see overcorrection in your child’s forehead.

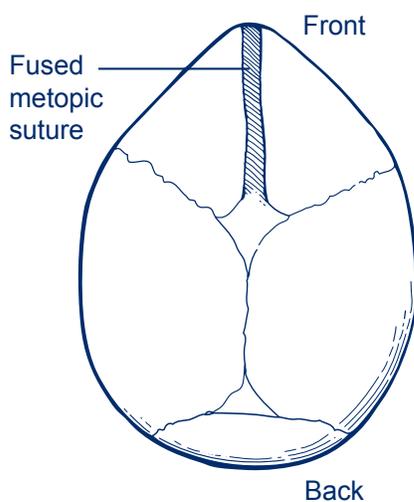


Figure 9. Trigonocephalic head viewed from above.

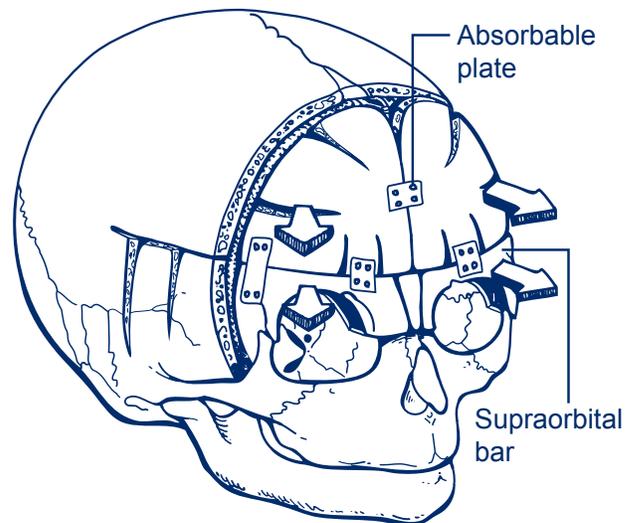


Figure 10. Frontal orbital advancement: the surgeon removes the bone of the forehead as well as supraorbital bar and advances the forehead to allow for growth of the brain, and to allow the skull to resume a more normal shape.



Lambdoid Synostosis

Lambdoid synostosis is one of the rarest forms of craniosynostosis. It occurs when the suture that runs across the back of the head is closed. This causes flattening of the head on the affected side or “**posterior plagiocephaly**” (Figure 11). Often times, infants can have an area of flatness on one side of their head from lying on their back in one favorite position. Surgery is only required if the lambdoid suture is closed or fused. When an infant has a flat head without closing of the sutures it is called “**positional head deformity.**”

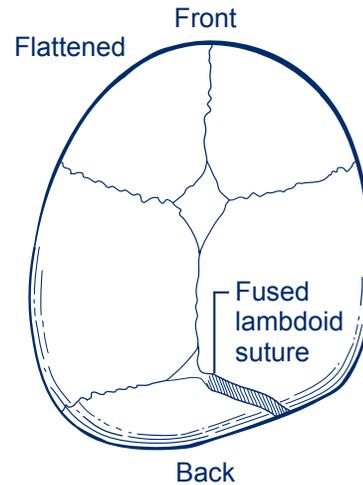


Figure 11. Posterior plagiocephalic head as viewed from above.



Positional Head Deformity

Positional Head Deformity occurs when the soft skull of an infant changes shape because of outside forces. For example, the position in the uterus may affect a baby’s head shape, and the baby’s position in the crib or while sleeping can also cause flattening of the skull if the pressure isn’t frequently relieved. **Torticollis** (bending of neck) can also cause positional head deformity. Torticollis is when your child holds his/her neck, and with it the head, towards one side. This tilted position can cause changes to your child’s face and head shape over time. Positional head deformity can be treated with helmet therapy to help correct head shape and physical therapy to help correct abnormal neck position. The child with positional head deformity often has a head shape resembling a parallelogram. The difference in overall head shape between positional head deformity and lambdoid synostosis is illustrated at right (Figure 12).

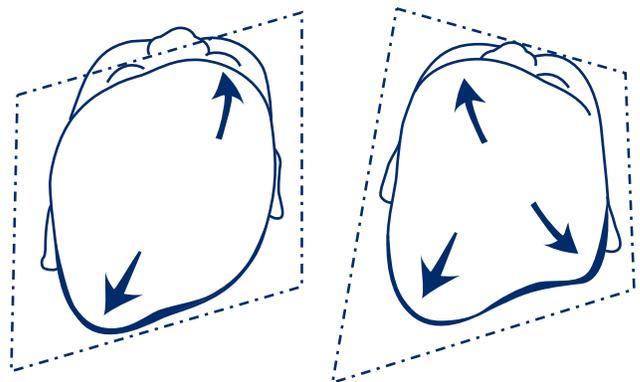


Figure 12. Positional head deformity (left) compared to lambdoid synostosis (right).



Preparing for Surgery

There are many reasons why it might be necessary to surgically treat the problems of the skull and face. One such reason is to improve a child's appearance. Another reason is to prevent possible pressure on the brain. Pressure on the brain, though rare, can cause headaches, irritability, decrease mental capacity or visual changes.

Before surgery, there may be several tests or evaluations that your child may need. He or she may need another CT scan or MRI (see page 6). Also, each child will need a complete history and physical exam completed within 1 month of the actual surgery. At this appointment, you and your child may see a Resident Doctor or Nurse Practitioner. Your child will also have blood drawn at this pre-surgery visit. You and your child will have an opportunity to speak with a nurse from anesthesiology to have questions answered about how your child will be anesthetized. Alternately, you can speak with a representative from anesthesiology on your operative day just before your child goes to surgery. Parents interested in a tour of the recovery rooms can also have that arranged. Tours take place usually once per week and may need a separate visit.

Surgery to repair craniosynostosis is an extensive procedure in which some children need a blood transfusion. A family can request "Directed Donation" blood if they choose. Directed donation occurs when a patient who may need a blood transfusion selects his or her blood donors. This is arranged through your local Red Cross. Parents are

not advised to be blood donors for their children; however aunts, uncles and siblings can be. All efforts are made to prevent transfusion during surgery including the use of medicine and other strategies during surgery to help control bleeding.

It is important to keep your child healthy prior to surgery. If your child has any of the following symptoms within six weeks of their surgery date it is important to take your child to their primary care provider and then call us:

- Croup, bronchiolitis, pneumonia
- Fever
- Cold or flu symptoms
- Wheezing
- Otitis media (ear infection).

Any sign of infection (strep throat, ear infection, sinusitis) should also be reported. If your child is taking an antibiotic just before or at the time of surgery, then surgery may need to be postponed. It is **extremely important** that your child is healthy going into craniosynostosis surgery.

Certain medications can have an effect on bleeding. Children should NOT take ibuprofen or other nonsteroidal medications such as Advil, Aleve, Motrin or Pediaprofen for a full 2 weeks before surgery. Alternately, children may take Tylenol (acetaminophen). Also, childhood immunizations or vaccines should not be given to your child for 2 weeks before surgery and for at least 2 weeks after surgery.



After Surgery

Postoperative Care

Parents or caregivers may see their child in the Recovery room after surgery. Parents may accompany their child as they are moved from the Recovery room to the Pediatric Intensive Care Unit, (PICU). In PICU, your child's vital signs, respiratory and neurological status will be very closely monitored. Your child may receive a blood transfusion if necessary. A doctor called a **Pediatric Intensivist** may be involved in taking care of your child. This is a doctor that specializes in the care of ill children in intensive care units. Your child's progress will determine the length of hospital stay. A child may stay an average of 5 to 7 days after craniosynostosis surgery. Once your child is able to take their feedings, has no sign of infection and some of the swelling around the eyes has gone down enough for your child to open at least one eye, your child will be able to go home.

Appearance

When you first see your child in the recovery room, they will have a large head dressing on. Underneath is a wavy incision across the top of their head from ear to ear (Figure 13). Hair will eventually hide much of this scar. This large head bandage is usually removed on the second or third day after surgery. Your child will also have swelling of the face and eyes. Most often the child's eyes will be swollen shut for several days.

They will also have one or more IV tubes for fluid and medication after surgery. Many children will have a Foley catheter in place. This is a tube that allows urine to drain from the bladder. Sometimes it is necessary to keep the breathing tube for a day or two after surgery until your child is stable and breathing on his/her own. In the PICU, you may stay with your child at almost all times, however you will be unable to sleep by your child's bedside. A family waiting room is located down the hall for you to rest in.

Medications

After surgery your child will receive pain medications through their IV tubes. Once your child can take fluids, pain medication can be given by mouth. You will receive a prescription for pain medication and an antibiotic when discharged. Often, Tylenol and Motrin are adequate for managing pain relief a few days after surgery. You may be instructed to put antibiotic ointment on your child's incision for a few days after surgery. Once your child is able to at least open one eye and is eating and drinking a little bit, he or she can be discharged home. After that, generally Vaseline is all that is needed on the incision site. While in the hospital and after discharge, the incision site will often drain. It is important to keep the incision free from scabbing and crusting by using Vaseline and gentle washing with a non detergent soap.

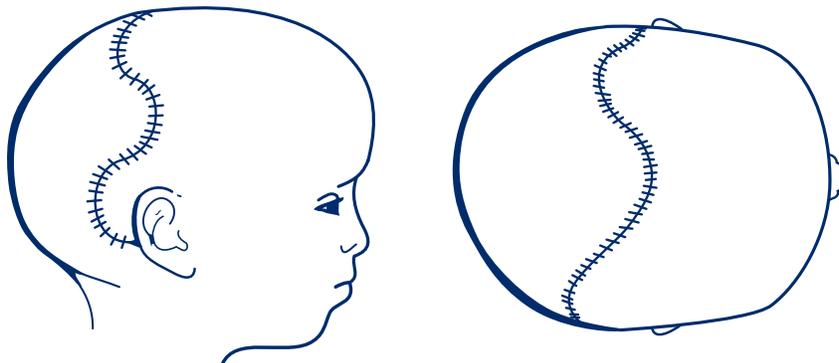


Figure 13. Scalp incision for craniosynostosis surgery.

Diet

Once your child's breathing tube is out and their respiratory status is stable, fluids are encouraged. Your child may resume nursing or drinking from a bottle or a cup. Generally fluids are started slowly with a little bit of clear liquids. If tolerated well your child may quickly resume their normal diet. Generally though, it is often several days before their appetite returns.

Activity

When you are discharged home, close supervision at all times is necessary. Your child may not resume any rough activity. If your child walks, it is very important to pad surfaces that he or she may be in danger of bumping their head on. If your child does not yet walk, they need to be kept in an environment where another toddler will not bump or accidentally hurt them.

While at home, your child will need to sleep with the head of their bed slightly elevated to help continue to decrease the swelling in their head. Sometimes sleeping in a car seat or bounce seat is easier. It is not necessary to wear a helmet after surgery. When your surgeon puts the bones of the face and skull together, they are held in place by dissolvable plates. It takes approximately 12 weeks for the bones in your child's head to heal and regain full strength. You will have follow up appointments periodically after discharge, most often 1-2 weeks after discharge, again in 1-2 months, 4-6 months after discharge, and then yearly for several years.

Changes in Behavior or Personality

In the first few days after surgery, you may see changes in your child's emotions and/or behaviors; these are normal responses. Your child may be unusually "fussy" or "clingy." There may be changes in their sleep patterns and eating patterns for several weeks or more. Occasionally you may see some "regression." For example, a child that was holding their own bottle may not want to do that for a short time. Gradually, children adjust and progress just as they did before surgery.

Support and Resources

Patients and families with craniofacial anomalies can often use some extra support, both financial and emotional. Our team provides several ways to assist you. Our social worker can speak with you about insurance called ***Children's Special Health Care Services***. This is supplemental insurance which provides comprehensive medical care to children under the age of 21 who have congenital or acquired physically handicapping conditions. Specific medical and financial criteria have to be met by the applicant before financial assistance is approved.

We also have networks of parents with special needs children. In addition, several of our parents have given consent to have other parents contact them regarding their own child's diagnosis. Speaking to other parents who have gone through a similar experience can often be very helpful. Please feel free to speak to our Nurse Practitioner or Social Worker for more details.



Syndromic Craniosynostosis

Sometimes craniosynostosis is inherited and part of a genetic syndrome. This type of craniosynostosis occurs much less often. Below are some of the more common types of syndromes in which children often have craniosynostosis. A complete evaluation by a geneticist will help identify syndromic craniosynostosis.

Apert Syndrome: This is a congenital or birth condition in which children have abnormal growth in several bones of the body. The skull sutures are fused causing craniosynostosis. They also have abnormal growth in the mid face or the part of their face between their eyes and mouth. Their face often looks as though it is retruded or sunken. Children with Apert syndrome also have bones or tissues in their hands and feet which are fused. Treatment for Apert syndrome starts early with skull surgery and continues for many years. Your child may be treated by a craniofacial team. This is a group of doctors (neurosurgeon, plastic surgeon, and orthodontist, just to name a few) and other health care professionals such as a speech pathologist and audiologist.

Crouzon Syndrome: Crouzon syndrome is a congenital condition in which the bones of the skull and face fuse abnormally. This causes abnormal skull shape, jaw problems and changes in the facial bones especially around the eyes and cheeks. Cheeks may appear flat and eyes may appear too prominent or bulging. Children with Crouzon syndrome will need care and treatment by a number of specialists in a medical center with a craniofacial team.

Pfeiffer Syndrome: Pfeiffer syndrome is a congenital condition in which children have craniosynostosis along with wide thumbs and great toes as well as partially webbed fingers and toes. Often there are several skull sutures which are fused causing abnormal growth of the skull and face. Many children with Pfeiffer syndrome have hearing loss. The severity of deformities associated with Pfeiffer syndrome varies quite widely. The care of children with Pfeiffer syndrome begins at birth and can include multiple operations, including surgery for craniosynostosis. These children will need care and treatment by a number of specialists in a medical center with a craniofacial team.



Definitions

- **Bilateral:** Pertaining to both sides of the body.
- **Brachycephaly:** A wide shaped head that is often flat, short from front to back and associated with a tall forehead.
- **Coronal Suture:** The loose connection that separates the frontal and parietal bones of the skull.
- **Craniofacial:** Referring to the face and skull.
- **Craniosynostosis:** The premature fusion of the bones of the skull.
- **CT Scan:** Computerized Tomography Scan: an x-ray that creates a two or three dimensional view of a portion of the body.
- **Frontal Orbital Advancement and Cranial Vault Reshaping:** An operation in which the bones of the skull and orbit are removed and reshaped to allow for growth of the brain.
- **Fusion:** Healing shut; establishing a bony union.
- **Geneticist:** A doctor specializing in the diagnosis of inherited disorders.
- **Hypotelorism:** An abnormally small distance between the eyes.
- **Intracranial:** Referring to the space within the skull.
- **Lambdoid Suture:** The loose connection that separates the occipital and two parietal bones.
- **Maxilla:** The bone that makes up the upper jaw.
- **Metopic Suture:** The loose connection that runs up and down the middle of the forehead between the two frontal bones.
- **MRI Scan:** Magnetic Resonance Imaging: a detailed picture of a part of the body created by a magnetic field and a computer.
- **Occiput:** The back part of the head.
- **Orbit:** The bony cavity that contains the eyeball.
- **Overcorrection:** The intentional placement of skull bones in a position that appears overly expanded or protruding. Over time, the gentle tension of the scalp and skin returns the bones to a natural position.
- **Papilledema:** Swelling in the optic disc of the eye caused by increased intracranial pressure.
- **Pediatric Intensivist:** A medical doctor that specializes in the care and treatment of critically ill children.
- **Plagiocephaly:** Flattening of the head on one side, also known as slanting type of skull asymmetry.
- **Positional Head Deformity:** Flattening of the skull because of outside forces.
- **Posterior Plagiophaly:** Flattening of the back of the head caused from a fused suture or lying in one favorite position.
- **Sagittal Suture:** The loose connection that runs down the middle of the head from front to back between the two parietal bones.
- **Scaphocephaly:** A head shape that is long and narrow from front to back, most often caused from premature fusion of the sagittal suture.
- **Sutures:** The open, fibrous areas that join the bones of the skull.
- **Synostosis:** Premature union between bones.
- **Torticollis:** The holding of a child's neck and with it head, towards one side which may cause changes to a child's face and head.
- **Trigonocephaly:** A head shape in which the forehead appears pointed and head appears triangular from above which most often occurs when the metopic sutures fuse prematurely.
- **Unilateral:** Pertaining to one side of the body.
- **Zygoma:** Cheekbone.



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