Children's Hospital of Philadelphia®
Division of Plastic & Reconstructive Surgery

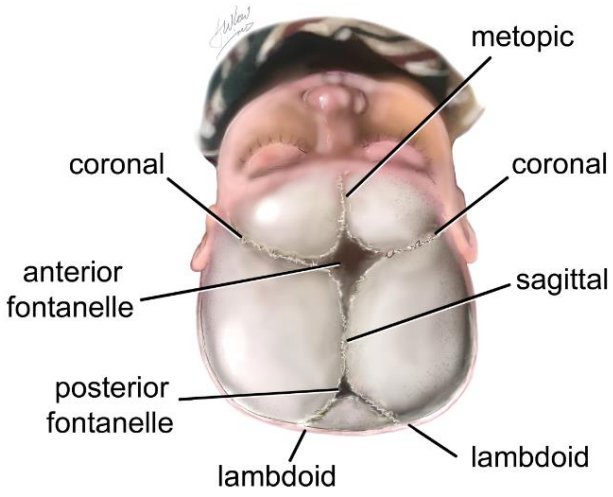
Caregiver Handbook: CRANIOSYNOSTOSIS

Craniosynostosis is a condition in which the sutures (growth seams) in an infant's skull close too early, causing problems with normal brain and skull growth. The most common type is single suture non-syndromic craniosynostosis. This means that only one of the sutures has closed early and the child does not have any related anomalies, like disorders of the limbs, ears or heart.

In a normal infant skull, all six major sutures are open at birth. The metopic suture closes sometime during early childhood (before 1 year of age), the rest of the cranial sutures stay open until late adolescence.

Cranial sutures:

- **metopic** suture in the forehead area,
- **two coronal** sutures, one on each side of the head from the soft spot (called the anterior fontanelle) to the ear
- **sagittal** suture that runs from the soft spot in the front of the head to the soft spot at the back of the head
- **two lambdoid** sutures that run from the middle of the back of the skull down to the base of the skull



MAKING THE DIAGNOSIS

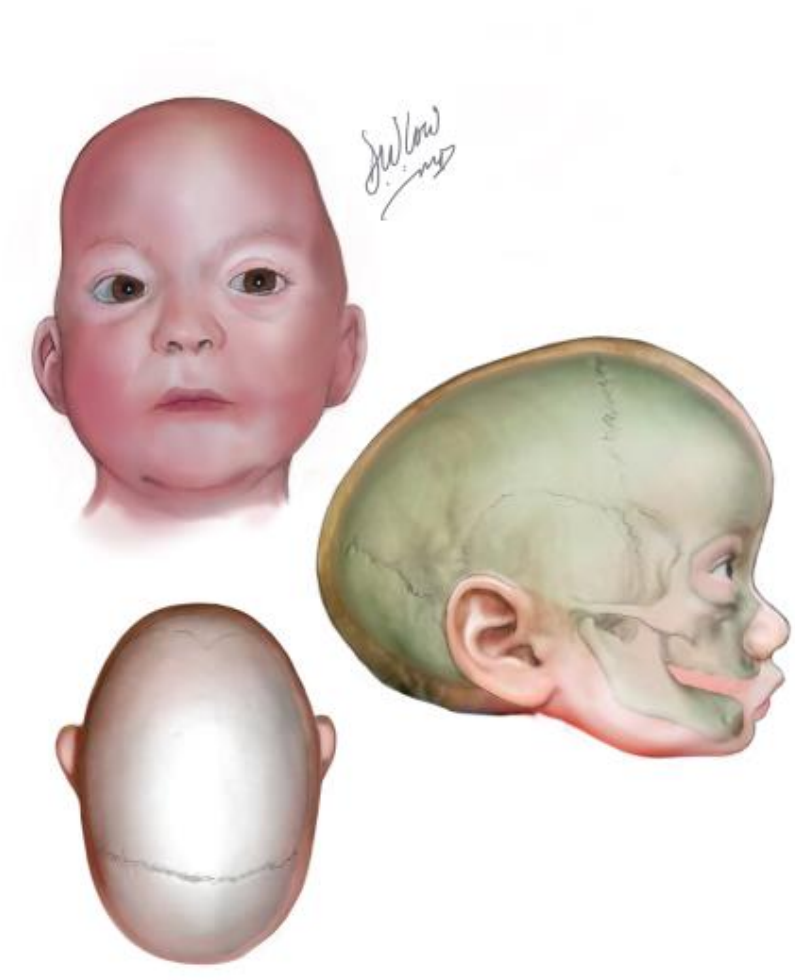
The diagnosis can often be made by a physical exam performed by an experienced craniofacial surgeon. Your craniofacial surgeon may also get a CT scan to a) confirm the diagnosis, and b) assist in planning surgery.

TYPES OF CRANIOSYNOSTOSIS

The head shape changes are dependent on which suture has closed prematurely. In some cases more than one suture is fused, this is called "multi-suture craniosynostosis".

Sagittal synostosis

- Suture involved: sagittal suture, the seam that runs from the front to the back of the skull
- Head shape is long and narrow, also called scaphocephaly (Greek for "boat-shaped head")
- May have a forehead that sticks out, called frontal bossing

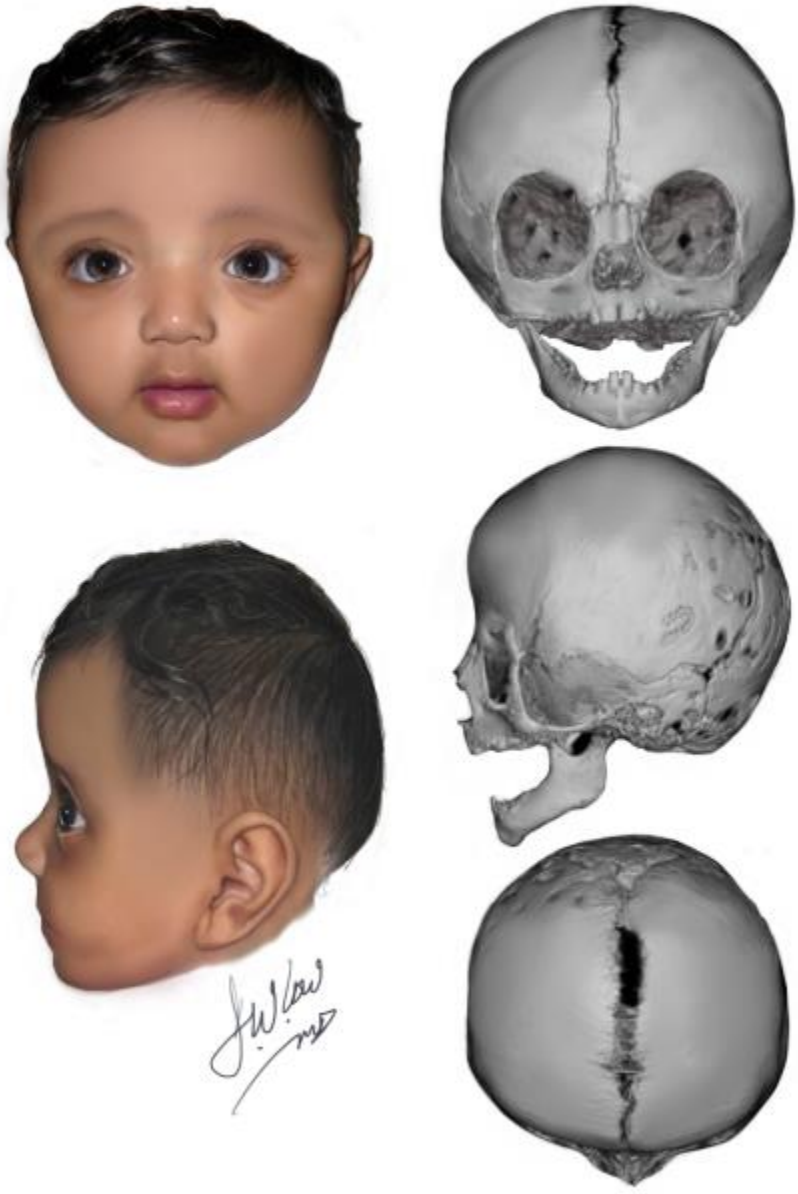


Unicoronal synostosis

- Suture involved: one of the coronal sutures, the seam that runs from the ear to the soft spot on top of the baby's head
- One side of the forehead and brow is much flatter and pulled backwards relative to the other side
- The eye on the flat side of the forehead appears taller and larger, the bridge of the nose is bent to the flat side

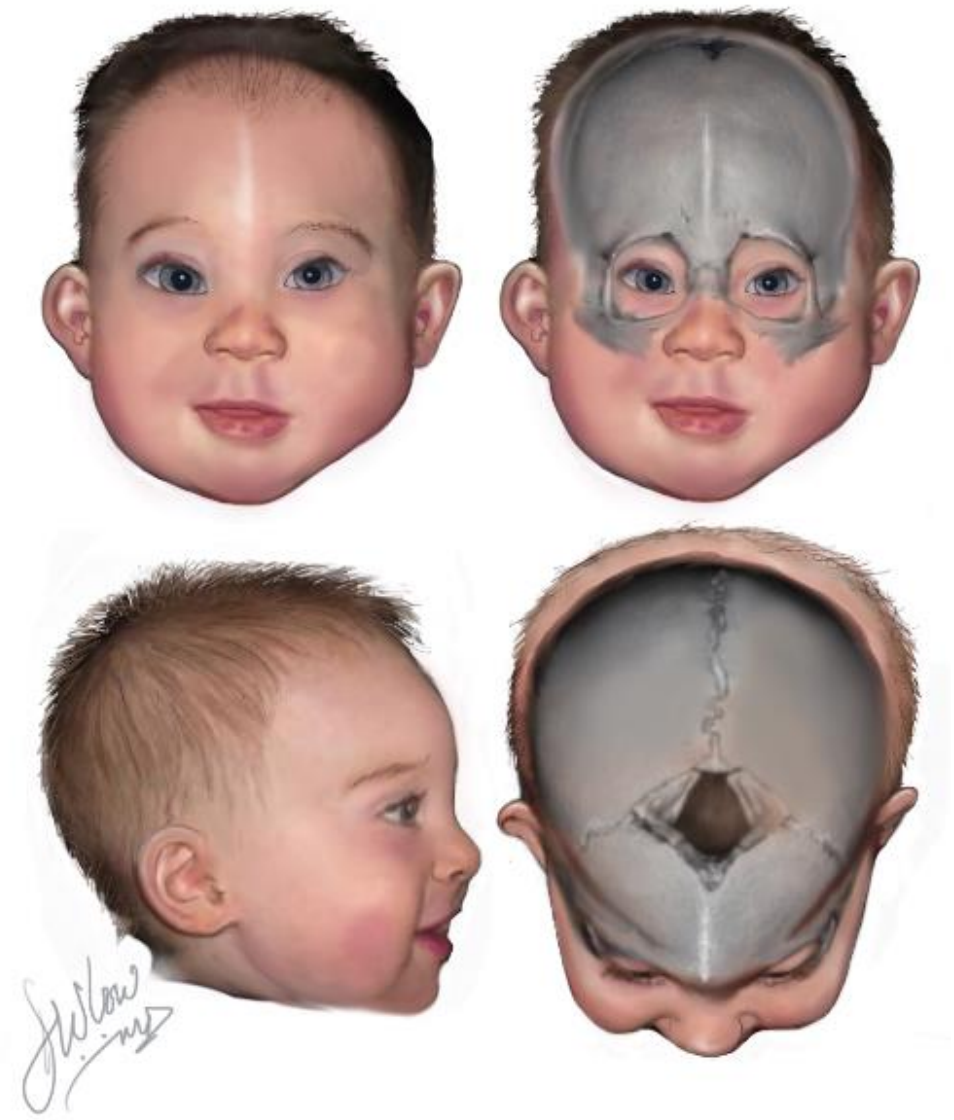
Bicoronal synostosis

- Suture(s) involved: both of the coronal sutures, the seams that run from ear to ear over top of the head and meet at the soft spot in the middle
- The head is forced to grow side to side and not front to back causing a wide and flat head with a tall forehead
- Bicoronal may exist in isolation, but it is the type of craniosynostosis most frequently associated with a syndrome, when other anomalies co-exist with craniosynostosis



Metopic synostosis

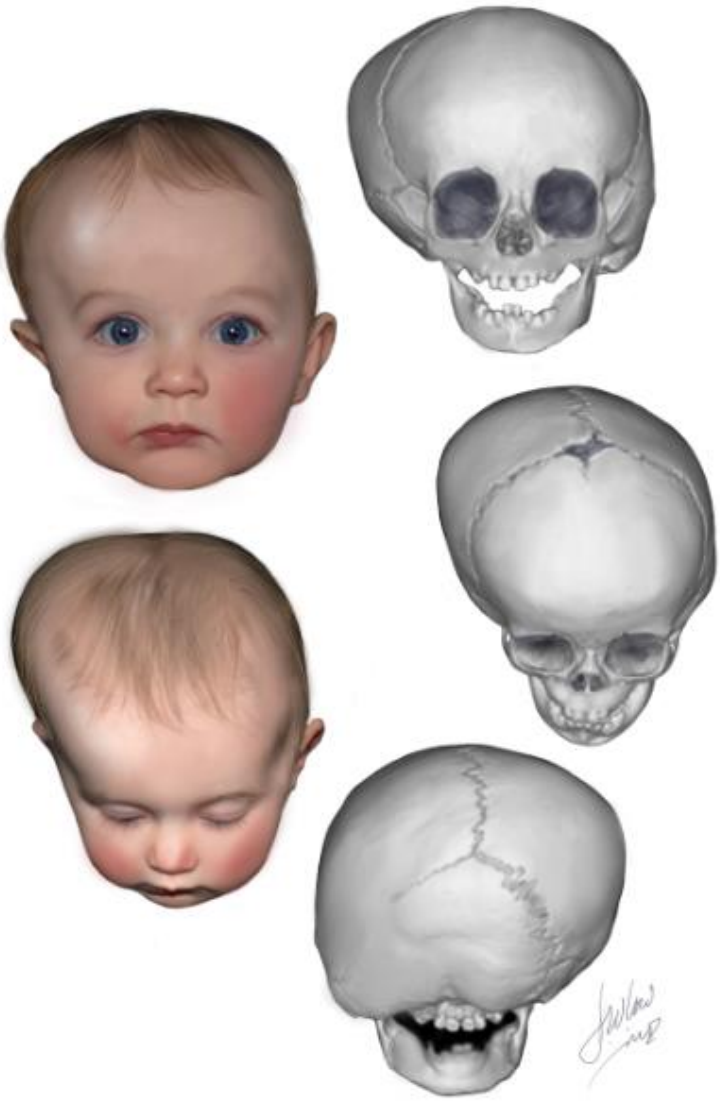
- Suture involved: metopic suture, the seam that runs from the top of the head down the middle of the forehead to the nose
- Head shape when viewed from above has a triangular shaped forehead, also called trigonocephaly
- Eyes may appear close together



Lambdoid synostosis

- Suture involved: one or both of the lambdoid sutures, the two seams located on the back of the skull
- Head shape is flat on one side with a large bulge in the mastoid region (back of the skull where the head connects to the neck)
- The ear of the flat side is moved to the back of the head or set lower than the other ear

  THIS TYPE OF CRANIOSYNOSTOSIS IS VERY RARE. MANY INFANTS HAVE FLATNESS ON ONE SIDE OF THE BACK OF THE HEAD AS A RESULT OF POSITIONING; THIS CAUSE OF FLATNESS DOES NOT NEED SURGERY. YOUR CRANIOFACIAL SURGEON WILL BE ABLE TO DIFFERENTIATE LAMBDOID CRANIOSYNOSTOSIS FROM OTHER CONDITIONS.



SURGICAL TREATMENT

Why is surgery recommended?

For most children diagnosed with craniosynostosis, some form of cranial surgery is recommended. The goals of surgery are:

*to release the constriction of the fused suture and normalize skull volume

*to normalize skull and facial shape in order to produce a normal appearance

An average newborn brain weighs about ¾ lb but an adult brain weighs around 3 lbs! The growing brain pushing out on a skull that cannot expand can cause increased intracranial pressure. Long periods of increased intracranial pressure can cause irreversible brain damage.

Unfortunately, the only way to measure intracranial pressure directly is surgery performed by our neurosurgery colleagues. A pressure monitor is placed through the skull and outer covering of the brain. Since intracranial pressure can fluctuate throughout the day, the child would then need to remain in the Intensive Care Unit with the brain pressure monitor in place for several days.

We do have other less invasive tests to look for pressure, such as a dilated eye exam or appearance of the skull bone on CT scan, however, these tests do not identify a significant number of children who actually do have increased pressure. A focus of our current research at CHOP is how we can better identify the children who are at highest risk of increased intracranial pressure.

The risk of increased intracranial pressure depends on the type and number of sutures closed and the age of the child. Talk to your craniofacial surgeon about your child's risk of increased intracranial pressure***. Again, this is a chronic problem and not something that happens overnight. In most cases the risk of it happening at all within the first year of life are very low.***

What type of surgery is available?

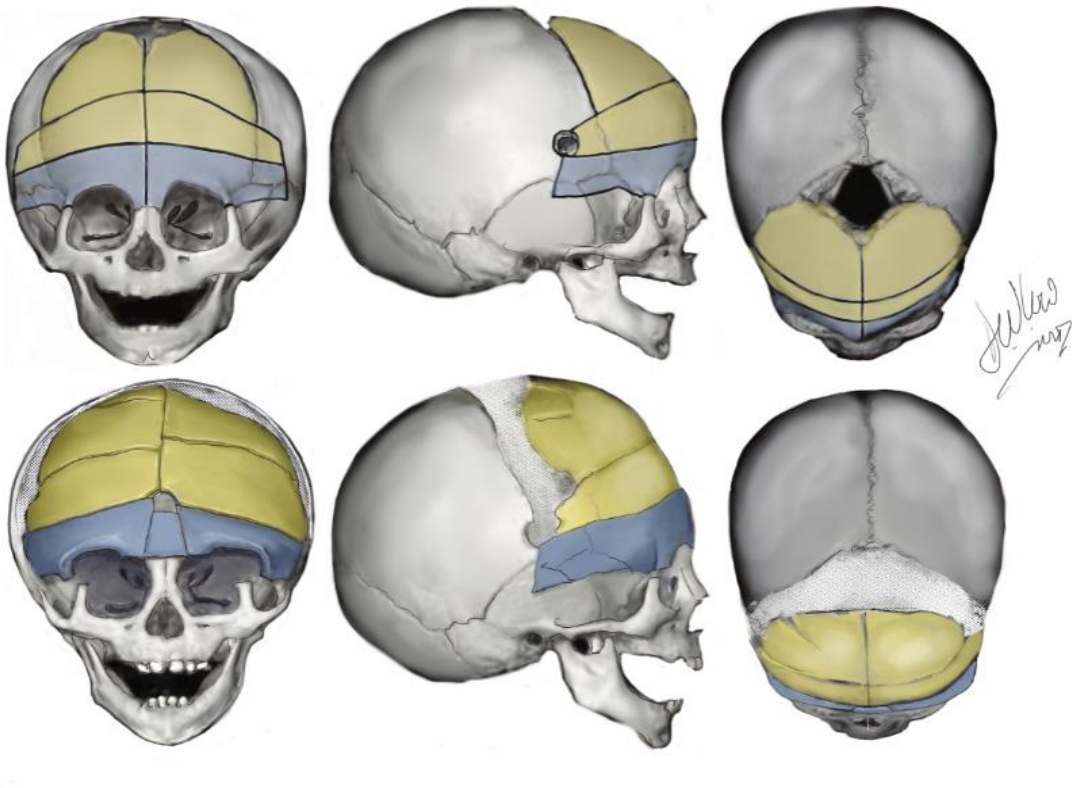
The type of surgery will depend on the sutures that are closed, the severity of the head shape changes and the age of your child. The surgeries all have one common goal; to remodel the skull so provide more room for the growing brain and to restore a more normal head shape.

We have provided a list of all potential surgeries here. It is important that you are aware of all options; however, you and your surgeon will work together to choose the best option for your child's specific needs.

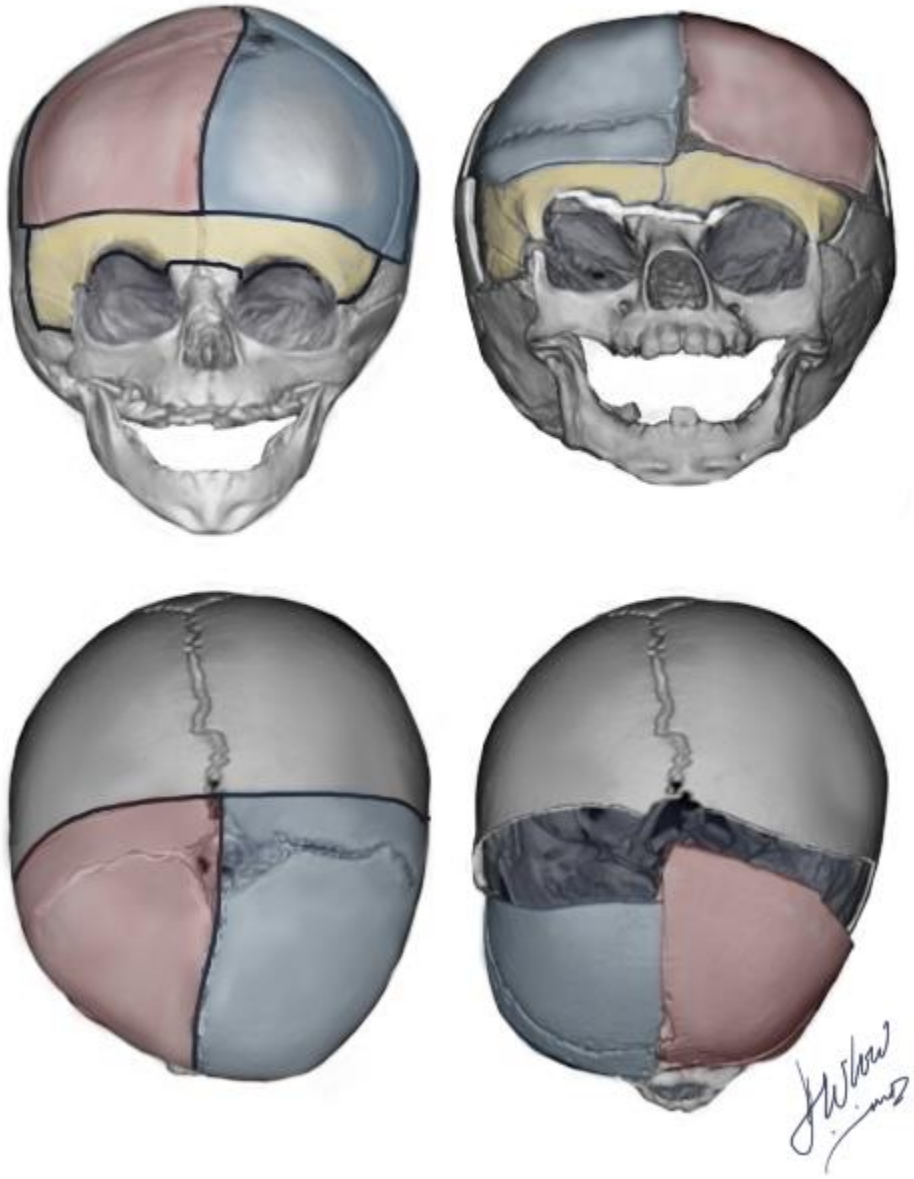
Anterior Cranial Vault Expansion and Remodeling or Bifronto-orbital advancement

- In this surgery, an incision is made across the skull from ear to ear. The incision is curved in such a way that hair regrowth should easily hide it.
- A neurosurgeon removes the front part of the skull with the closed suture. The craniofacial surgeon then reshapes the bones of the skull using absorbable plates and screws to hold the bones in the new position. Surgery takes about 4-5 hours.
- This surgery is usually done with the child is between 10-13 months old
- Your child will be in the hospital for about 4-5 days after surgery
- Your child will need a blood transfusion during this surgery
- Recovery will take about 2 weeks, though if done on older children we will limit strenuous physical activity for about a month.
- At home you will need to clean the incision with soap and water and apply a topical antibiotic ointment
- Very swollen eyes are expected and can last for about a week before starting to improve
- Most children do not require prescription pain medication once they leave the hospital
- For most children this will be the only cranial surgery they will ever need

The anterior skull expansion and remodeling below shows the positions of the skull bones in a patient with metopic craniosynostosis, pre and post surgery.

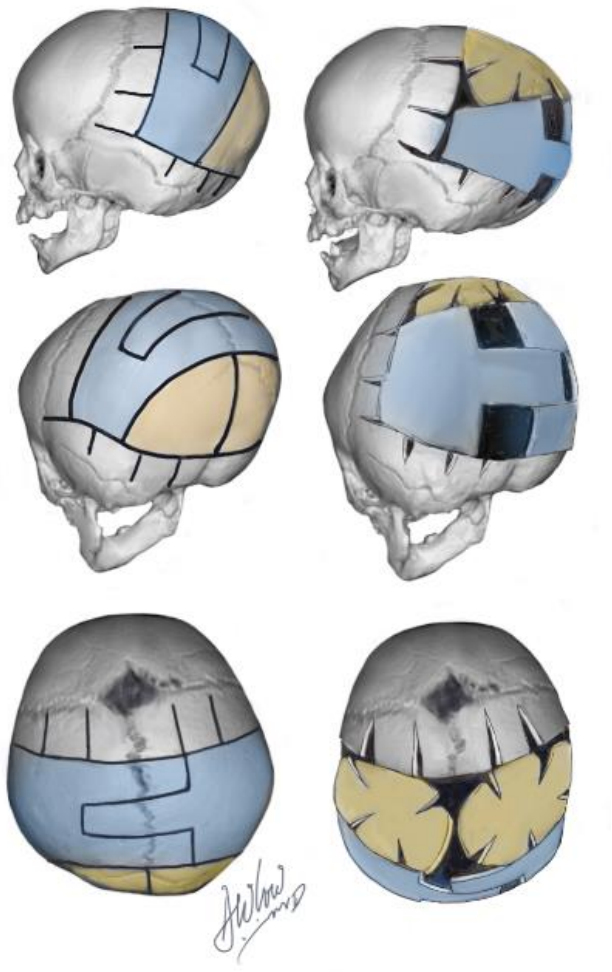


The anterior skull expansion and remodeling below shows the positions of the skull bones in a patient with right unicoronal craniosynostosis, pre and post surgery.



Posterior Cranial Vault Expansion and Remodeling

- In this surgery, an incision is made across the skull from ear to ear. The incision is curved in such a way that hair regrowth should easily hide it.
- A neurosurgeon removes the back part of the skull with the closed suture. The craniofacial surgeon then reshapes the bones of the skull using absorbable plates and screws to hold the bones in the new position. Surgery takes about 4-5 hours.
- This surgery is usually done when the child is between 10-13 months old
- Your child will be in the hospital for about 4-5 days after surgery
- Your child will need a blood transfusion during this surgery
- Recovery will take about 2 weeks, though if done on older children we will limit strenuous physical activity for about a month.
- At home you will need to clean the incision with soap and water and apply a topical antibiotic ointment
- Most children do not require prescription pain medication once they leave the hospital
- For most children this will be the only cranial surgery they will ever need

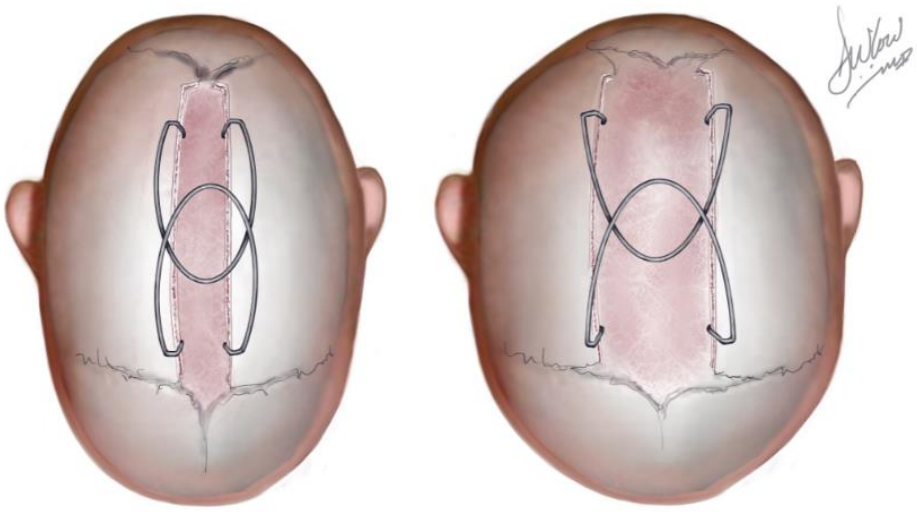


Endoscopic Strip and Prolonged Helmeting

- Appropriate for infants who are under 4 months of age
- The fused sagittal suture is removed through two small incisions in the top of the head, they are each about 3-4cm long. To keep the area from immediately closing again the child must wear a custom molding helmet at least 23 hours per day for up to one year post-op. This will require regular trips to an orthotist, a professional who will fit and reshape the helmet throughout the process.
- One surgery, that takes about 1-2 hours. It is done with a neurosurgeon and requires a 1-2 night hospital stay
- Most patients do not need a blood transfusion
- 1-2 week recovery with no specific activity limitations or wound care at home

Endoscopic Strip with Spring Mediated Cranial Vault Expansion

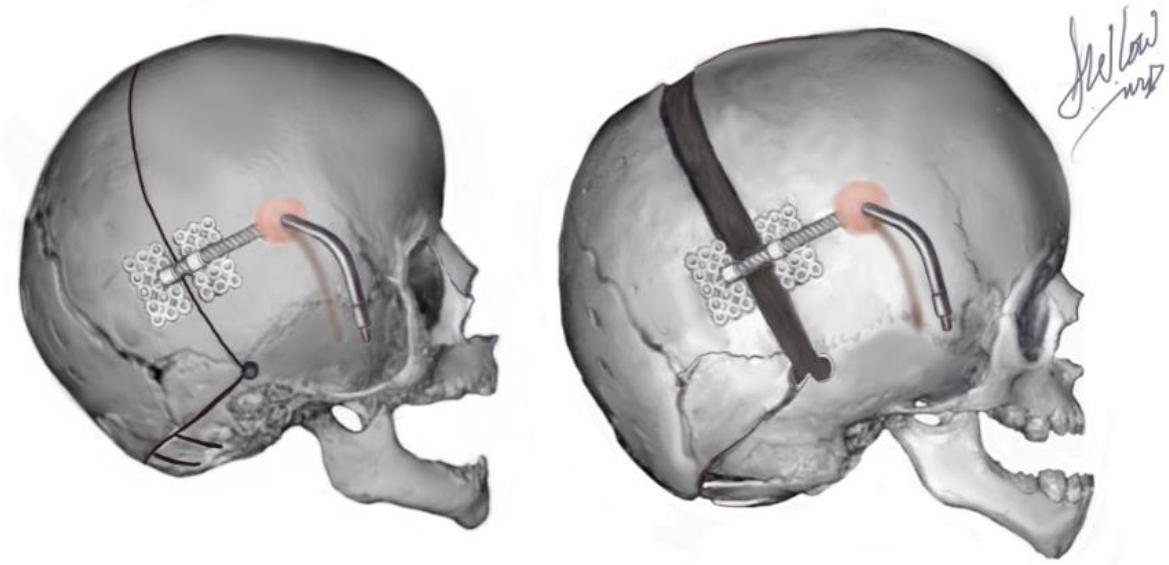
- Appropriate for infants who are under 4 months of age
- The fused sagittal suture is removed through two small incisions in the top of the head, they are each about 3-4cm long. To keep the area from immeadiately closing again custom springs are placed to slowly move the bone pieces further apart.
- This requires two surgeries approximately 3 months apart. The first surgery is done with the craniofacial surgeon and a neurosurgeon; it takes about 1-2 hours and requires a 1-2 night hospital stay. Just the cranial facial surgeon performs the spring removal and no overnight stay is needed.
- Most patients do not need a blood transfusion
- 1-2 week recovery with no specific activity limitations or wound care at home following the first stage. Generally infants are recovered by the next day following spring removal
- The child may need a short period of wearing a cranial molding helmet before and after the surgery



The first picture below is the skull immeadiately following endoscopic strip with placement of springs, the second is the shape of the skull after three months of spring mediated cranial vault expansion.

Distractor mediated cranial vault expansion

- A neurosurgeon removes the part of the skull with the closed suture. The craniofacial surgeon then places devices along side the cuts in the skull bone. A part of this device will stick out through the child's skin.
- This surgery takes about 3-4 hours and your child will be in the hospital for 4-5 days
- Your child may need a blood transfusion during this surgery
- Recovery will take about 1-2 weeks, though if done on older children we will limit strenuous physical activity while the distractors are in
- At home you will need to turn the distractors every day to slowly expand the bone and soft tissue, most children do not find this painful
- You will come to the office every 1-2 weeks and get skull Xrays so we can check on progress
- It usually takes about 3 weeks of turning the distractors every day to move the bone to its final position
- After the turning is completed most of the distractor device that can be seen through the skin is removed in the office, this is painless
- Three months after the first surgery a second surgery will be needed to remove the remaining distractor device that is buried under the child's skin. This surgery takes about 1 hour and your child may stay over one night, though many go home that same day.
- Most children do not require prescription pain medication once they leave the hospital



PRE SURGERY CHECKLIST

- You will **meet with your craniofacial surgeon** and other members of the plastic surgery team; including the Craniofacial Nurse Navigator, Plastic Surgery Nurse Practitioners and the surgical scheduler. We will review the plan for surgery and are available to answer any questions you have. It is important that you arrive for the day of surgery fully understanding the plan and confident in the care your child will receive.

- You will also **meet the Neurosurgeon** who will be involved in your child's operation. Sometimes we are able to arrange for this to happen on the same day as your Plastic Surgery visit, otherwise you will need to schedule the neurosurgery consultation for another day.

- You will not meet the anesthesiologist until the day of surgery. The **Anesthesia Resource Center** (ARC) Nurse Practitioners do pre-operative anesthesia assessments. The ARC team will review your child's medical history and discuss the anesthesia plan with you. The ARC team will review when your child needs to stop eating and drinking prior to surgery. Instructions are provided in this booklet.

- Your child will need **blood work** prior to surgery; the ARC team will arrange this. The testing related to potential blood transfusions must be done at a CHOP lab and within a specific time period. The ARC team will help coordinate this for you. If your child has other medical conditions we may ask for additional tests or consultations. Although this may mean additional appointments, we want to fully understand your child's needs well so we can coordinate the safest surgical plan.

- We may refer your child to an **ophthalmologist** to get a baseline dilated eye exam prior to surgery. A concern with craniosynostosis is potential increased pressure in the brain, one way to screen for this is with a specific eye exam. The number for CHOP Ophthalmology is 215-590-2791. We may also ask your permission to perform a specialized eye exam while your child is in the operating room.

- We may ask you to **consider participation in a clinical trial**. Care for children with craniosynostosis has made significant advancements over the past decade. However, we are committed to searching for new, better, safer ways to care for children. This is only possible through research. You are never required to participate in our research studies; however, we ask that you be open to considering the studies for which your child is eligible.

- We may refer your child to **genetics**. This evaluation does not need to be done prior to surgery. Many children with craniosynostosis do not have a genetic cause. The number for CHOP genetics is 215-590-2920.

- To **find out your arrival time** call 267-435-4699 the afternoon before the procedure between 3:30 and 6pm. If your child's surgery is scheduled for a Monday please call the Friday before.

- Give your child a bath the night before surgery and perform the **CHG treatment**. Please see specific CHG treatment instructions provided in this booklet.

HAVE WE ANSWERED THESE QUESTIONS FOR YOU?

1. How long will I be away from my child on day of surgery? _____
2. How long will my child be in the hospital? _____
3. What special care will my child need at home? _____

   _____
4. How long will my child need to stay home from school or daycare? _____
5. How long before going back to sports/playground/gym?_____
6. How long before swimming, bathing? _____
7. How often will I see you while my child is in the hospital? _____
8. When will my child's first post-operative appointment be? _____
9. What if I have an urgent question after surgery and it is a night? Or a weekend? _____

   _____
10. What are the risks of surgery? _____

    _____
11. What are the risks of not doing surgery? _____

    _____
12. What are the major signs I should look out for post-operatively to know if something is wrong?

    _____