# Craniosynostosis

## Diagnosis and Treatment

Weill Cornell Medicine | NewYork-Presbyterian | Columbia

## ABOUT THE CRANIOSYNOSTOSIS PROGRAM

The Craniosynostosis Program of the combined pediatric neurosurgery and plastic surgery services at NewYork-Presbyterian (Komansky Children's Hospital and Morgan Stanley Children's Hospital,) takes a multidisciplinary approach to treating craniosynostosis and related disorders. Co-directed by Dr. Caitlin Hoffman of Weill Cornell Medicine and Dr. Thomas Imahiyerobo of Columbia, the program is dedicated to ensuring a successful outcome for every child and family. This includes a comprehensive clinical evaluation, selection of the best treatment option, and utilization of the most advanced technology to optimize outcomes for each patient. The team understands that the trust developed before surgery is equally important after surgery in order to support a child through a positive recovery.

Our Craniosynostosis Program brings together a unique team of experts that offer the very best treatment for children with craniofacial abnormalities. Because disorders of the face and skull impact both functional and morphologic aspects of a patient's quality of life, systemic evaluation, genetic analysis, and family planning are all implemented when appropriate.

Weill Cornell Medicine Pediatric Neurosurgery
525 E. 68th Street, Box 99, Room 2207
New York, NY 10065



**For more information about the Craniosynostois Program**

**craniosynostosis@med.cornell.edu**

**212-746-2363**

Weill Cornell Medicine | NewYork-Presbyterian | Columbia

## DEAR COLLEAGUES:

The art and science of craniofacial surgery has advanced at a remarkable pace over the past three to four decades, with new techniques and treatments producing outstanding results for most children born with craniofacial anomalies. Today we can identify children at risk for certain birth defects before they are even born and can repair and reconstruct an extremely wide variety of anomalies with excellent outcomes.

We also now understand the need for multi-disciplinary teams to address these conditions from a multi-systems standpoint, as well as the importance of early diagnosis. Newer endoscopic surgical options allow for faster recovery times, lessen the need for transfusion, and reduce surgical risk—this option, however, is only available to infants under four months of age. We are leaders in advancing the endoscopic approach, and work closely with pediatricians to identify candidates for this option as early as possible. At the same time we continue to offer the full slate of all surgical options, including traditional open craniofacial surgery as well as craniofacial distraction ontogenesis.

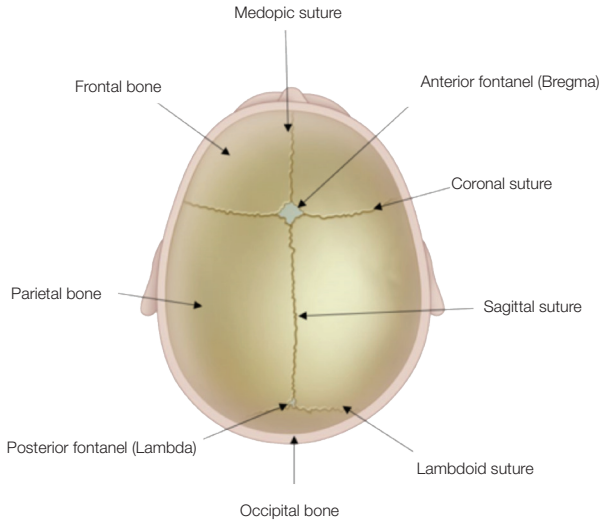
For patients with complex needs, we work with a team of professionals from a range of specialties, including otolaryngology, plastic and reconstructive surgery, dentistry, ophthalmology, genetics, and neurological surgery, to name just a few. Our young patients and their parents may also need support from psychologists, social workers, and child life specialists to help them through the process. We are proud to offer one of the most skilled and experienced comprehensive teams of craniofacial specialists in the country to serve the needs of our patients and families. Through this interface, we use advanced virtual surgical planning to determine the most appropriate treatment and to achieve optimal outcomes. We also partner with basic science labs to further our understanding of the cellular and molecular mechanisms of synostosis and hopefully define potentially non-surgical treatment options in the future.

This booklet is meant as an introduction to the clinical diagnostic methods for simple non-syndromic craniosynostosis. Please share this complimentary copy with other professionals or use it as an instructional resource guide with your families of children with craniosynostosis. The Craniofacial Program team at Weill Cornell Medicine and Columbia looks forward to working with you to provide your patients with the most contemporary and advanced care available.

**Caitlin Hoffman MD,** and **Thomas Imahiyerobo, MD**
Co-Directors, Craniofacial Program

Weill Cornell Medicine | NewYork-Presbyterian | COLUMBIA

## ANATOMY OF THE INFANT SKULL




Vertex view of normal skull

## EXAMINATION OF THE INFANT SKULL

Requisite observations/views most important to utilize in making diagnoses of single suture synostosis:

- Vertex
- AP (Anterior/Posterior)
- Lateral

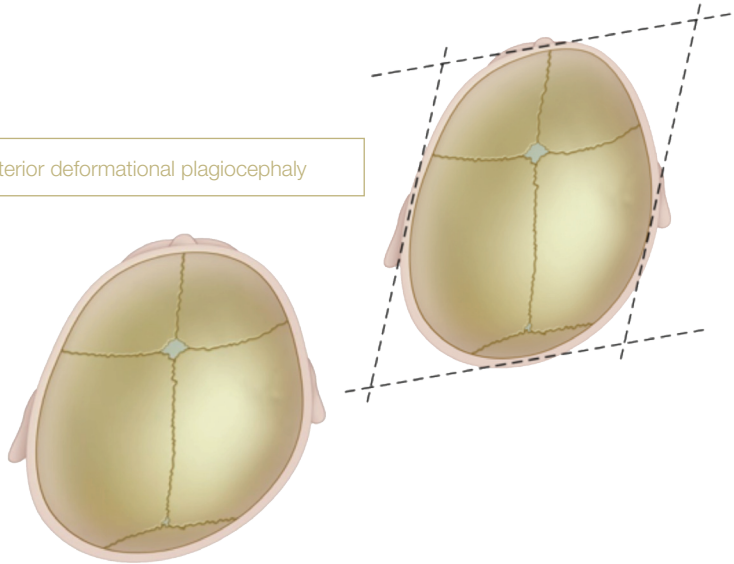
Asymmetrical Findings:

- Ocular malalignment (dystopia)
- Auricle displacement
- Flattening/Prominence of forehead/occiput

Symmetrical Findings:

- Narrow biparietal dimension with a Cephalic Index measuring within normal limits (74-83)
- Elongated AP dimension (frontal ± occipital prominence)
- Forehead retrusion (oxycephaly)

## DEFORMATIONAL (POSTERIOR) PLAGIOCEPHALY



Right posterior deformational plagiocephaly

Also known as positional molding, deformational plagiocephaly is a common cranial deformity in children and the most common cause of misshapen skull in infants. It is a term used to describe flattening on one side of the head, the major cause of posterior plagiocephaly. Flattening on both sides of the skull is known as deformational brachycephaly.

Clinical Signs of Deformational Plagiocephaly:

- Unilateral occipital flattening
- Anterior displacement of the ipsilateral forehead (frontal bossing)
- Anterior displacement of the auricle
- Parallelogram shaped skull

Clinical Signs of Deformational Brachycephaly:

- Bilateral occipital flattening (disproportionately wide head when evaluated from the front)
- NO ipsilateral frontal bossing or auricular anterior displacement

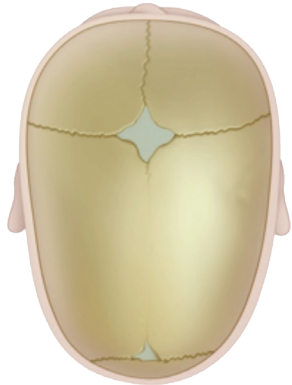
## SAGITTAL SYNOSTOSIS

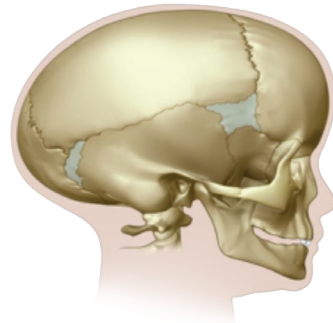
Sagittal synostosis, also known as scaphocephaly, is the most common form of craniosynostosis.

Clinical Signs of Sagittal Synostosis:

- Biparietal narrowing
- Frontal bossing (compensation)
- Occipital bulging (compensation)
- Palpable ridging overlying the sagittal suture
- Cephalic index measuring < 74

Sagittal synostosis





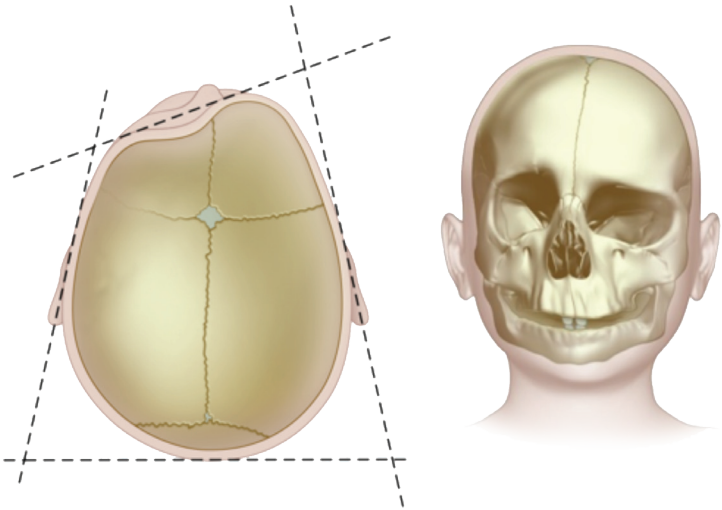
## UNILATERAL CORONAL SYNOSTOSIS

Unilateral coronal synostosis is also known as anterior plagiocephaly.

Clinical Signs of Unilateral Coronal Synostosis:

- Flattening of the forehead
- Retrusion of the orbital rim with enophthalmos
- Nasal root and midface angulation
- Anterior displacement of the ipsilateral auricle
- Ridging overlying the ipsilateral coronal suture
- Vertical dystopia of eyes (unilateral elevation)
- Retrusion/flattening of the ipsilateral forehead
- Trapezoid shaped skull

Left-sided unilateral coronal synostosis

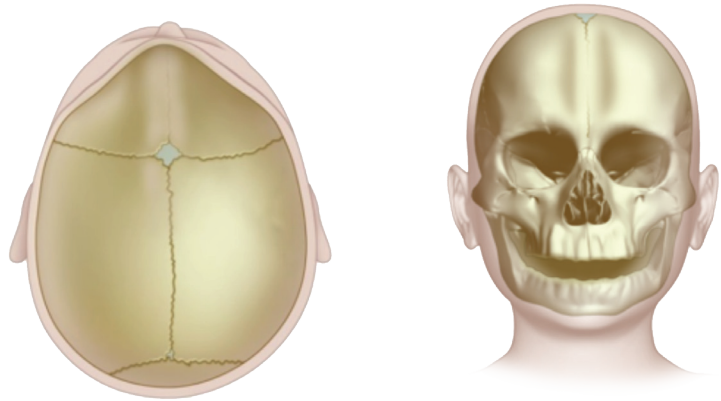


## METOPIC SYNOSTOSIS

Metopic synostosis, also known as trigonocephaly, is a less common form of craniosynostosis; however, metopic ridging is very common. The metopic suture can begin to fuse as early as 2 months of age and it is not uncommon for the ridging to be visible along the midline of the forehead. It is paramount to correctly distinguish metopic synostosis from metopic ridging.

Clinical Signs of Metopic Synostosis

- Bifrontal narrowing
- Biparietal widening (compensation)
- Hypotelorism
- Midline pointedness of the forehead
- Ridging overlying the metopic suture



Metopic synostosis

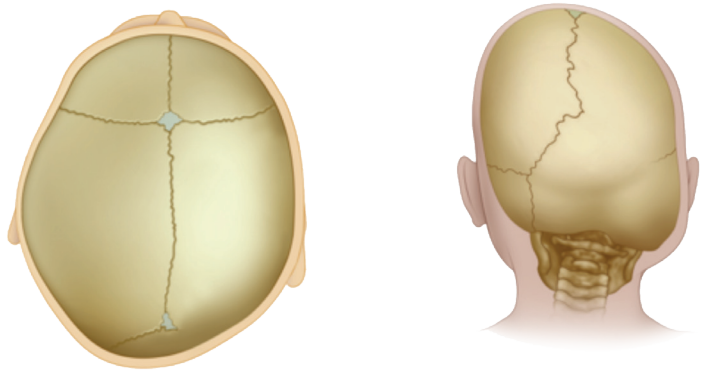
## LAMBDOID SYNOSTOSIS

Lambdoid synostosis is a much less common form of synostosis and the less common cause of posterior plagiocephaly.

Clinical Signs of Lambdoid Synostosis:

- Posterior and inferior displacement of the auricle of the ear
- Vertex slanting and shortened cranial height on the affected side
- Prominent mastoid bulging on the affected side
- Ridging overlying the ipsilateral lambdoid suture

Right lambdoid synostosis



## IMAGING

It is not necessary to order imaging to confirm or rule out a diagnosis of single suture craniosynostosis. Computed tomography is costly, often requires sedation, and involves low-dose ionizing radiation. It is impractical to have every child with cranial flattening undergo imaging because the majority of infants with cranial asymmetry will have deformational plagiocephaly and not synostosis.

If a diagnosis of synostosis is made or suspected based on physical examination, the next step is to refer the child to a pediatric neurosurgeon. A trained specialist can usually distinguish deformational plagiocephaly from synostosis easily based on history and physical examination. Only in very rare cases is radiologic imaging necessary.

For more information, visit weillcornellbrainandspine.org/craniosynostosis

or call Program Coordinator **Michelle Buontempo** at 212-746-**2363**

## TREATMENT OPTIONS

Surgery is the only effective treatment for craniosynostosis, because fused sutures must be opened to allow the brain to expand. This is not something that will happen naturally, and a child cannot "outgrow" craniosynostosis. A skilled surgeon must create new openings in the skull to allow for the rapid brain growth that takes place in the first year of life. The surgery is safe and produces excellent results.

There are several surgical options for treating craniosynostosis, depending on which type it is. It's usually best to perform surgery at just a few weeks to a few months of age, since the skull bones are the softest and most malleable then. The craniofacial team that evaluates a child will recommend the best surgery based on which suture closed prematurely and the degree of deformity.

### TRADITIONAL OPEN SURGERY

Traditional open surgical procedures are called cranial vault remodeling and vertex craniectomy, which are safe and produce excellent results. In this surgery, a neurosurgeon and plastic surgeon remove the affected or closed suture and then "remodel" the skull. The surgery usually takes between two and six hours and requires three to five days in the hospital, depending on the age of the child and which suture is involved. Some children need blood transfusions during the surgery; a compatible parent may donate his or her blood to be used in the event that a transfusion is needed. No helmet therapy is needed after traditional surgery.

### ENDOSCOPIC SUTURECTOMY

Endoscopically assisted strip craniectomy (also called endoscopic assisted suturectomy) is a newer, minimally invasive approach to craniosynostosis surgery. As in the traditional approach, a neurosurgeon and plastic surgeon remove the closed suture – but unlike the traditional approach, the endoscopic procedure does not include cranial remodeling during surgery. This minimally invasive procedure is typically performed on infants younger than four months of age, since it depends on extremely rapid brain growth to help reposition the cranial bones. Endoscopic assisted suturectomy usually takes less time in the operating room and requires a shorter hospital stay. After endoscopic surgery for craniosynostosis, the child will wear a cranial remodeling helmet to help reshape the skull. For more information, visit **suturectomy.org**

**Weill Cornell Medicine**
**Pediatric Neurosurgery**
525 East 68th Street, Box 99 Starr
Pavilion, Suite 651
New York, NY 10065 212.746.2363

Like us on Facebook for news and updates
facebook.com/WCMNeurosurgery
facebook.com/WCMPediatricNeurosurgery

Visit us online for more information:
neurosurgery.weillcornell.org

Weill Cornell Medicine | NewYork-Presbyterian | Columbia

June 2024