Craniosynostosis
Diagnosis and Treatment
ABOUT

The Weill Cornell Craniofacial Program takes a multidisciplinary approach to treating Craniofacial Disorders. Co-directed by Dr. Mark Souweidane of Pediatric Neurological Surgery and Dr. Vikash Modi of Pediatric Otolaryngology, the program is dedicated to ensuring a successful outcome for every child and family. This includes a thorough evaluation of the case, selecting the best option, and utilizing the most advanced technology. The team understands that the trust developed before surgery is equally important after surgery in order to support the child through a positive recovery.

Our Craniofacial Program brings together a team of experts that offer the very best of non-operative and surgical treatment for children with congenital (inborn) or acquired skull abnormalities. Because disorders of the face and skull can involve more than just the child’s appearance, systemic evaluation, genetic analysis, and familial planning are all available when appropriate.

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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 those at risk for birth defects before they are even conceived, and we 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 problems—patients with craniofacial issues do best when treated by a team of professionals from a range of specialties, including otolaryngology, plastic and reconstructive surgery, dentistry, ophthalmology, 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. At Weill Cornell, we are proud to offer one of the best comprehensive teams of craniofacial specialists in the country to serve the needs of our patients and families.

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 looks forward to working with you to provide your patients with the most contemporary and advanced care available.

Sincerely,

Mark Souweidane, M.D.
Director, Pediatric Neurosurgery
Weill Cornell Brain and Spine Center
ANATOMY OF THE INFANT 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)
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
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
**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
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
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/craniofacial

or call Program Coordinator Charlotte Beam, MS, CGC, at 212-746-1274