Duke Cleft &
Craniofacial Program

Care for Children with Craniosynostosis

What Is Craniosynostosis?

Infants are born with skulls composed of six bone plates, separated by areas of soft fibrous tissue known as sutures. These sutures expand throughout early childhood to accommodate the growth of the brain. Brain growth is rapid through the first two years of life, then tapers over time.

Under normal circumstances, the sutures fuse during predictable age ranges. When sutures fuse at the appropriate times, the result is the symmetric round head shape we're accustomed to seeing. If, however, one or more of the sutures closes prematurely, the brain will grow in the direction of sutures that are still open.

This abnormal pattern of brain growth results in the first sign of craniosynostosis: an abnormally shaped skull. It may also be possible to detect the fused suture—which may feel like a ridge of bone—by touch. The condition can also cause increased pressure on the growing brain. Although uncommon, craniosynostosis can be associated with developmental delay, mental retardation, and/or seizures.

What Causes Craniosynostosis?

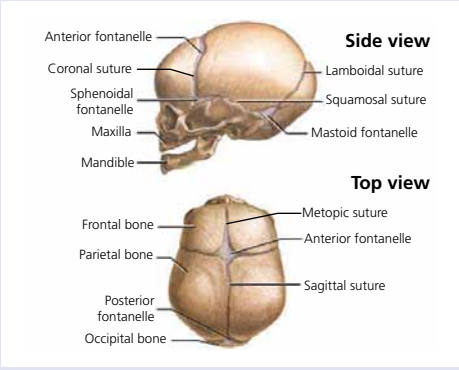
The cause of craniosynostosis is still relatively unknown. Certain congenital conditions, such as Apert, Crouzon, Pfeiffer, and Saethre-Chotzen syndromes, include craniosynostosis. The majority of affected children, however, have "non-syndromic" craniosynostosis, which may be caused by a combination of several factors. Parents should never feel that they have done anything to cause craniosynostosis in their child.

The Cranial Sutures

The six cranial sutures most commonly affected in craniosynostosis are:

- Two coronal sutures and two lambdoid sutures, running across the skull on either side
- One sagittal suture, running down the length of the skull
- One metopic suture, running longitudinally down the forehead

The areas where sutures intersect are the fontanelles, often referred to as the "soft spots."



Craniosynostosis and Head Shape

Even when a suture fuses prematurely, the brain continues to grow at a relatively normal rate. As a result, the shape of the head will become abnormal not only near the affected suture but in other parts of the skull as well, as they expand more than usual to accommodate the growing brain.

Craniosynostosis can cause various patterns of abnormal skull growth. For **sagittal and metopic synostosis** (see illustration), the abnormal shape is symmetric (the same on both sides) because these sutures run along the midline.

In **unilateral coronal synostosis**, lack of growth on the affected side leads to compensatory overgrowth on the normal side to accommodate the growing brain. This asymmetrical growth is often easier to see, even though the condition is not necessarily any more severe than other forms of craniosynostosis.

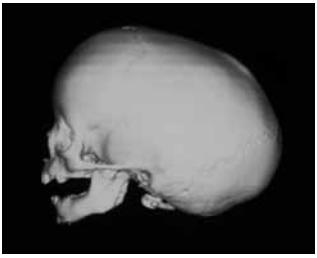
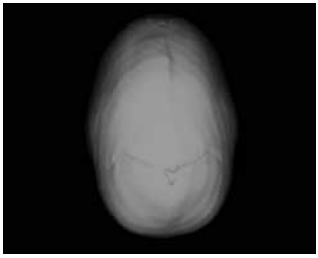
Treatment of Craniosynostosis

The Duke craniofacial team evaluates each child individually, weighing many factors before making a consensus treatment recommendation. Among these factors include the severity of the aesthetic deformity, the presence of a known syndrome, and the potential for increased pressure within the skull.

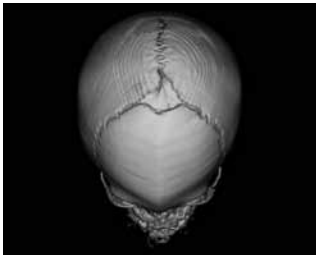
Treatment varies according to the suture(s) involved. It generally consists of surgery to relieve pressure on the brain and the cranial nerves. However, not every child diagnosed with craniosynostosis will be recommended for surgery. Based on a detailed exam and the latest clinical experiences, our team will recommend reconstructive procedures that are specific to each child's particular condition.

All craniosynostosis treatments share the goal of releasing fused sutures and increasing the volume within the skull. These techniques are also designed to restore the normal shape and dimensions of the head while allowing for future growth. We prefer to proceed with reconstruction between the ages of six and nine months; however, reconstruction can be performed safely in older children as well.

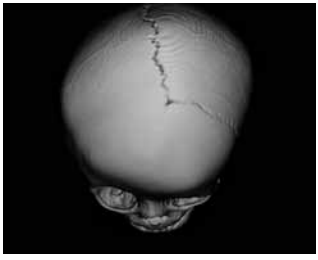
Note: Children with sagittal synostosis are candidates for specific reconstructive procedures if they can be performed prior to six months of age.

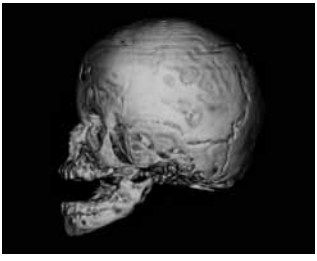
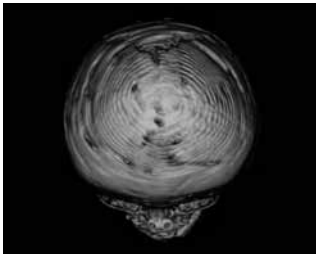
Sagittal synostosis: Growth is impaired from side to side and therefore compensates from front to back, resulting in an elongated and narrow head, referred to as scaphocephaly. The forehead and/or the back of the head may seem particularly prominent.

Metopic synostosis: Impaired growth along the forehead results in a triangular shape when viewed from above and recession of the brows, referred to as trigononcephaly.

Unilateral coronal synostosis: Results in an asymmetric shape, referred to as plagiocephaly. Flattening of the forehead and brow occur on the affected side and a bit of bulging is visible on the unaffected side. The eye is drawn upward, making it appear more open than the opposite (unaffected) side. This may make the lid of the unaffected side appear "droopy" by comparison.

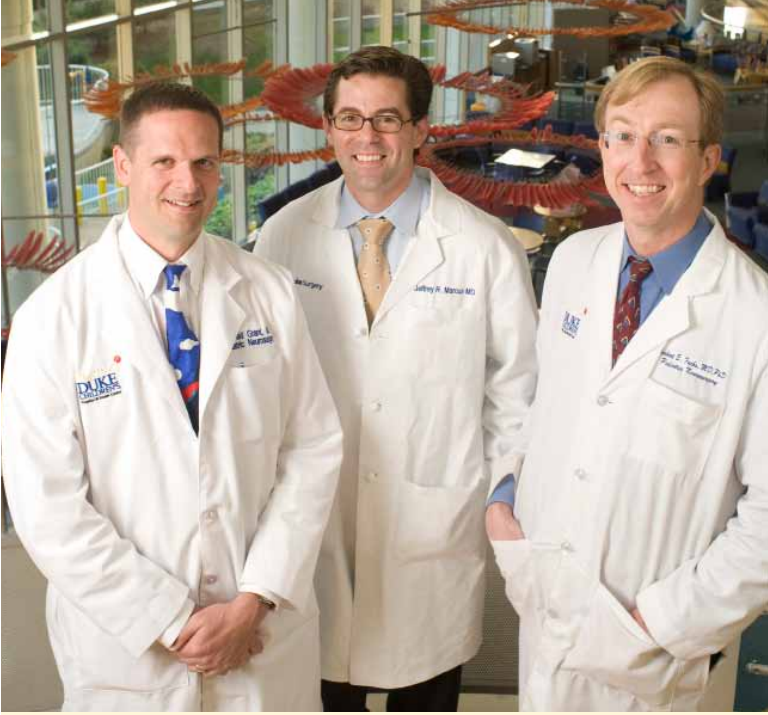
Bilateral coronal synostosis: When both coronal sutures are involved, there is relatively equal recession of the forehead and brow on both sides. The forehead appears tall and steeply angled. This is referred to as brachycephaly.

Prognosis and Follow-Up

The prognosis for craniosynostosis varies depending on whether single or multiple cranial sutures are involved, and whether other abnormalities are present. For the vast majority of children with craniosynostosis, growth and development proceed normally. Most of the children who undergo reconstructive surgery will require no further surgical procedures.

After surgery, our young patients will have follow-up appointments, alternating between the pediatric neurosurgeon and craniofacial surgeon, at one week, one month, three months, nine months, and one year, and followed by annual appointments with both surgeons until the age of five.

Children with congenital syndromes, multiple problems, or extended needs may be referred for comprehensive care by our multidisciplinary Craniofacial Program team. Our team approach allows families to meet with multiple specialists in the same place at the same time, particularly convenient for families who must travel some distance to come to Duke. After each visit, a summary and report are sent to the family and to the child's pediatrician. This approach helps to ensure continuity of care and positive outcomes as the child grows and develops.



(from l to r), Gerald A. Grant, MD; Jeffrey R. Marcus, MD; Herbert E. Fuchs, MD, PhD

The Duke Craniofacial Team

The program's craniofacial surgeon, Dr. Jeffrey R. Marcus, is chief of pediatric plastic surgery and is fellowship-trained in both pediatric plastic surgery and craniofacial surgery.

The Duke Craniofacial Program includes two specialized pediatric neurosurgeons with years of experience: Dr. Herbert E. Fuchs and Dr. Gerald A. Grant.

Other members of the multidisciplinary Duke Craniofacial Program team include specialists from the following disciplines:

- Pediatric Neurosurgery
- Craniofacial Surgery/Pediatric Plastic Surgery
- Pediatric Dentistry
- Pediatric Otolaryngology
- Pediatric Ophthalmology
- Pediatric Hand Surgery
- Orthodontia and Oral Surgery
- Genetics
- Speech Pathology & Audiology
- Social Work
- Child Life
- Neuroradiology

Duke Cleft & Craniofacial Program

Results to last a lifetime

For More Information

We welcome family inquiries and physician referrals to our service, and are happy to provide consultations to community-based physicians about youngsters in their care.

To find out more about the Craniofacial Program at Duke:

- Call **888-ASK-DUKE** (275-3853) for patients/families or **800-MED-DUKE** (633-3853) for physicians
- Visit **dukehealth.org**
- Mail c/o Ann Mabie
  DUMC 3974
  Durham, NC 27710
- Visit **dukesurgery.org** to support research in this area

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