What condition does craniofacial surgery address?

Craniofacial surgery is a pediatric surgical subspecialty that involves the reconstructive treatment of disorders of the face and skull. Craniosynostosis is a rare condition in which a baby develops or is born with an abnormally shaped skull. It is caused by premature closing of one or multiple cranial sutures (skull bones) on a baby’s head. Babies are born with floating skull bones in order to allow them to come through the birth canal and to allow for rapid head growth during the first year of life. Craniosynostosis can be diagnosed as early as birth, however most infants are diagnosed in the first few months. Craniosynostosis leads to a restriction in the growth of the skull, which can cause unusual head shape and facial features. In unique cases, it can also cause damage to the brain due to increased pressure inside the skull. The cause of craniosynostosis is unknown. However, in extremely rare cases, craniosynostosis can be inherited and part of a genetic syndrome. Neurosurgeons, with or without plastic surgeons, correct this abnormality depending on the type and severity of the synostosis.

What does craniosynostosis surgery accomplish? When should it be performed?

The operative management of craniosynostosis is tailored to the type and severity of the condition as well as the age of the patient. The principle goal in the treatment of craniosynostosis is to remove and remodel fused sutures in the skull. At ANA, surgery is typically not done until the infant is about three months old and/or as close to 12 pounds as possible. The earlier the better, as the child’s skull is softer, and more malleable. (The older the child, the harder the skull.)

In regards to craniosynostosis, will my child require multiple surgeries?

When only one suture is fused, it is likely that only one surgery will be required to correct the abnormal skull shape. When multiple sutures are involved, as in craniofacial syndromes, children often require multiple skull surgeries during the course of childhood and adolescence. Staged surgeries for certain craniosynostosis syndromes include Pfeiffer, Apert or Crouzon syndromes.

Children with syndromic craniosynostosis often suffer from progressive synostosis of multiple sutures of the skull and skull base.

These patients require addition surgical consideration, which include:
- Treatment for hydrocephalus if present (more common in children with multiple synostoses)
- Management of airway obstruction
- Ophthalmological consideration for potential eye problems
- Feeding issues related to abnormal jaw or oral abnormalities

Some children are candidates for endoscopic craniofacial surgery, also performed by our expert surgeons at ANA. The incision is smaller (only about one inch), the operative time is shorter and there is less blood loss. After the surgery, the child wears a cranial molding helmet for a period of time. The key to having this surgery is an early diagnosis (under six months of age). ANA has formed craniofacial teams at many hospitals throughout New Jersey in order to provide comprehensive care and provide patients with all necessary specialists in the same location.

Who comprises a craniofacial surgical team?

In addition to an expert pediatric neurosurgeon, children with craniofacial problems require surgical care from a variety of specialties, including plastic surgery, ENT, pediatric ophthalmology, dentistry and oral surgery. There is also associated care in clinical genetics, nursing, pediatrics, speech pathology, oral-motor therapists, occupational therapy, orthotics, physical therapy and social services. ANA has formed craniofacial teams at many hospitals throughout New Jersey in order to provide comprehensive care and provide patients with all necessary specialists in the same location.
What is the expected surgical outcome for craniosynostosis?

Using modern surgical techniques, studies have shown that craniosynostosis can be corrected with positive outcomes and relatively low morbidity and mortality. This is particularly true for otherwise healthy, non-syndromic infants. (Most craniosynostosis cases are non-syndromic. In other words, not genetically caused.) However, the right surgical team is key. This team largely relies on the pediatric neurosurgeon and plastic surgeon specifically trained in craniofacial procedures. ANA has renowned expertise in this area and has combined forces to produce the Craniofacial Specialists of New Jersey.

What is positional plagiocephaly and how do you treat it?

Positional plagiocephaly refers to a misshapen (asymmetrical) shape of the head from repeated pressure to the same part of the head. (In this case, there is no fusion of the skull sutures.) Since infants are often placed on their backs to sleep to reduce the incidence of sudden infant death syndrome, this repeated pressure can flatten the skull. Positional plagiocephaly is commonly found in twins/multiples due to positioning in utero. It can also be associated with torticollis (shortening of the neck muscles causing movement restriction).

Treatment of deformational plagiocephaly generally includes adjusting positioning to avoid pressure on the back of the head, physical therapy, and in severe cases cranial reshaping helmets. The first step is to try to reposition the infant - provide tummy time when awake, partake in neck stretching (in the case of torticollis) as directed by pediatric physical therapy, and limit the amount of time spent on the back of the head. If by seven to eight months the head shape is still moderately to severely flattened, a cranial molding helmet may be recommended. The helmet is worn 23 hours a day for seven days a week for an average of two-and-a-half-month duration. Occasionally, a second helmet is needed, but this is rare.