WHAT IS CRANIOSYNOSTOSIS?

A Baby’s Skull

A baby’s skull is made up of several large bones. The places where these bones touch are flexible connections called sutures. The location where four of these large bones meet in the front of the head is called the anterior fontanelle, or soft spot (a smaller one is sometimes present at the back of the head as well). This flexibility allows the head to fit through the birth canal, and permits the brain to grow. The brain quadruples in size the first two years of life, and the bones of the skull must grow and not restrict its growth. Bone growth occurs at these sutures.

Craniosynostosis

When a suture is not formed or closes too soon, it is called craniosynostosis. It is estimated that this defect occurs in one out of every 2000 live births.

Craniosynostosis causes the head shape to be deformed and in certain instances, can prevent the brain from having enough room to grow. The specific abnormality of the head shape depends on the suture(s) involved. An abnormal head shape is noticed after birth.

Diagnosing craniosynostosis is done by performing a physical exam and an x-ray.

SUTURE LOCATIONS

- Metopic
- Coronal
- Sagittal
- Lambdoid

THERE ARE TWO BROAD CATEGORIES OF SURGERY FOR CRANIOSYNOSTOSIS:

MINIMALLY INVASIVE SURGERY

Children’s Hospital Oakland is the only facility in Northern California with extensive experience with endoscopic craniosynostosis surgery. In the 10 years we have been performing this surgery, we have not needed to perform any revision surgeries on endoscopic surgery patients with sagittal synostosis.

About the procedure:

- “Minimally invasive” or endoscopic surgery requires an early diagnosis because it can be done only during the first few months of life before the bone thickens and the compensatory changes are set. It is generally performed when the baby is about 3 months old.
- Two small incisions are made and a tiny endoscope or lighted retractor is inserted to remove the fused suture and strips of bone.
- Following surgery, the child wears a helmet for several months that is custom-designed by our staff orthotist to help mold the skull into a normal shape. The orthotist makes any necessary helmet adjustments at Children’s.

The advantages over invasive surgery:

- The less-invasive procedure results in less blood loss, smaller scars.
- A shorter hospital stay is required.
- Endoscopic surgery produces better cosmetic results and less chance of having to perform revision surgeries.

INVASIVE SURGERY

The more traditional, invasive surgery is typically performed when the child is 6 to 12 months old. With this approach, the surgeons rearrange the bones in the skull to achieve a normal skull shape and size. The time spent in the operating room and the duration of the hospital stay are usually longer for this surgery than for endoscopic surgery.

In a traditional repair for older children, an incision is made across the scalp, from the top of one ear to the top of the other ear, behind the patient’s hairline. Major portions of the skull are removed to mechanically reconstruct the skull. The long-term outcomes are excellent.
HEAD SHAPE AND APPEARANCE
Craniosynostosis can include the following:

**Sagittal synostosis**
Sagittal synostosis is the most common suture to close too soon, and it inhibits growth of the skull on both sides. The skull compensates by growing longer in the front and back, with a very large forehead and narrow protruding back of the skull. This is called dolicocephaly (or scaphocephaly). In addition to a deformed skull, there is a real risk that the brain may not have enough room to grow in sagittal synostosis. Elevated intracranial pressure and subsequent neurologic damage can occur.

**Trigonocephaly**
When the metopic suture closes too soon, it may or may not be a problem. It is not a problem when the only noticeable feature is a ridge on the forehead. It is a problem when the forehead becomes keel shaped (just like the front of a boat). This is called trigonocephaly. Additionally, the eye sockets may not grow laterally, giving the appearance that the eyes are too close together. Also, the temporal area on each side of the skull (near the temples) cannot grow laterally, creating a hollow appearance to the child’s temples.

**Coronal suture synostosis**
Coronal suture synostosis can involve one or both coronal sutures. However, it is much more common for only one coronal suture to be involved. This causes flatness of the forehead on the affected side and the eye socket is pulled up on that side. When both coronal sutures are fused too soon, the skull cannot grow front to back. The skull instead grows side-to-side, which is called brachycephaly.

**Lambdoid suture synostosis versus Plagiocephaly**
The last and least common suture to close too soon is the lambdoid suture. It causes flatness in the back of the head on the affected side. It can look like positional plagiocephaly, a benign flatness of the back of the head caused by babies spending too much time that side. Unlike lambdoid synostosis, positional plagiocephaly is not caused by craniosynostosis (premature closure of one or more cranial sutures) and is not treated with surgery. Instead, treatment involves repositioning techniques or helmet therapy.