

CRANIOSYNOSTOSIS

WHAT IS CRANIOSYNOSTOSIS?

Craniosynostosis is a condition where one or more of the sutures in a child's skull closes too early. Sutures are fibrous joints located between the bones in a baby's skull. Open sutures allow the skull to grow at the same rate as the brain. In the first few years of life this growth is very rapid.

When a suture closes too early, the skull cannot grow normally. This can cause pressure on the growing brain. As the brain continues to grow, it pushes against or expands the areas of the skull that are not fused. This leads to a change in the shape of the head. Depending on which of the four sutures is fused, a typical head shape is produced.

The 4 sutures in the skull are metopic, sagittal, lambdoid and coronal. Only one suture - the metopic - normally closes during the first few years of life, and the others remain open into adulthood.

Most commonly, only one suture is affected. This is called single suture craniosynostosis. With single suture craniosynostosis, babies are usually healthy and have no other problems or differences.

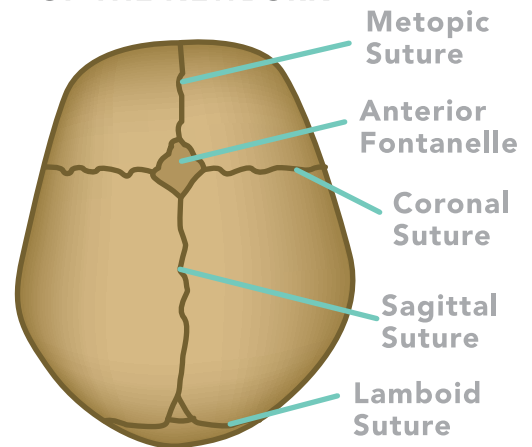
Less commonly, two or more sutures can fuse early. In this case, children are more likely to have a *craniofacial syndrome*, especially when both coronal sutures are affected. The more sutures involved, the higher the level of concern for pressure on the brain.

HOW COMMON IS CRANIOSYNOSTOSIS?

Craniosynostosis usually occurs while the baby is still in the womb. It is thought to occur in 1 out of every 2000 live births. Babies with this condition typically have an unusual head shape when they are born and it becomes more noticeable as weeks and months pass.

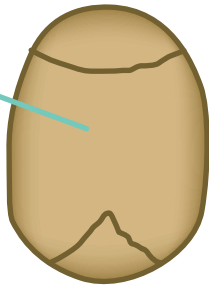
The most common reason for a baby to have an unusual head shape is not craniosynostosis, but a condition called deformational plagiocephaly which does not involve a fused suture. That is why it is important to see a craniofacial specialist for an evaluation.

NORMAL SKULL OF THE NEWBORN



THERE ARE FOUR TYPES OF SINGLE SUTURE CRANIOSYNOSTOSIS.

Fused
Sagittal
Suture



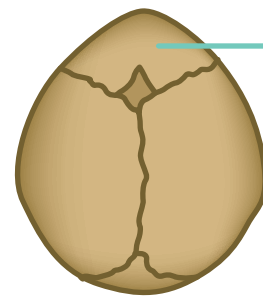
◀ Sagittal Craniosynostosis (Scaphocephaly)

Sagittal craniosynostosis is the most common form of craniosynostosis, occurring in about 1 in 2000 live births. The sagittal suture runs from the baby's soft spot and goes straight back. When it fuses too early, a ridge can often be felt or even seen over this area. The soft spot may be absent or small.

As the brain grows, the skull can no longer get wider, so it gets longer and bulges at the front and the back. This is called scaphocephaly. A baby with sagittal craniosynostosis may seem to have a large or prominent forehead.

Metopic Craniosynostosis (Trigonocephaly) ▶

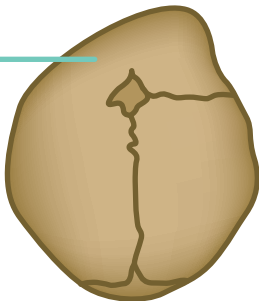
The metopic suture runs from the baby's soft spot to the forehead. When it fuses too early, a ridge can often be felt or even seen over this area. The soft spot may be absent or small. As the brain grows, the forehead can no longer get wider, and appears pinched.



Fused
Metopic
Suture

When viewed from above, the head shape looks like a triangle. This is called trigonocephaly. A baby with metopic craniosynostosis may seem to have his or her eyes close together.

Fused
Coronal
Suture
(1 side)

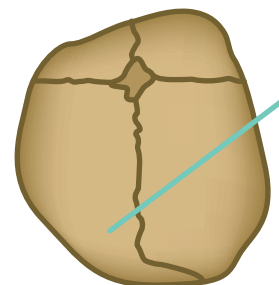


◀ Unicoronal Craniosynostosis (Anterior Plagiocephaly)

The coronal suture runs from a baby's soft spot towards the ear. When it fuses too early, the forehead looks flat on the affected side. The eyebrow can appear higher. As the brain grows, the forehead can appear prominent on the unaffected side. This is called anterior plagiocephaly.

Lambdoid Craniosynostosis (Posterior Plagiocephaly) ▶

The lambdoid suture is in the back of the head. When this suture fuses too early, the back of the head is flattened and the ear may be lower on the affected side. This is called posterior plagiocephaly and is the rarest type of craniosynostosis.



Fused
Lambdoid
Suture
(1 side)

WHAT CAUSES SINGLE SUTURE CRANIOSYNOSTOSIS?

The exact cause of craniosynostosis is not yet clear. In about 20 % of children with single suture craniosynostosis, a mutation or change can be identified in one of the genes. If a couple has a child with single suture craniosynostosis, the chance their next baby will also have

craniosynostosis is believed to be less than 2%. Sometimes craniosynostosis is part of a syndrome, although this is more common with multiple sutures. Craniofacial syndromes are thought to result from a genetic mutation. Our center is currently investigating both the molecular mechanisms and the underlying genetic causes of craniosynostosis.

HOW DO YOU DIAGNOSE CRANIOSYNOSTOSIS?

If a family or pediatrician notices an unusual head shape, the infant should be referred for evaluation by a craniofacial specialist. Sometimes children are referred based on an unexpected change in head size (circumference) or early closure of the soft spot (fontanel).

An experienced pediatric craniofacial surgeon or pediatric neurosurgeon can make the diagnosis by physical exam. In some cases, a CT scan may be obtained to assist with the evaluation.

HOW IS CRANIOSYNOSTOSIS TREATED? WHY?

Surgical treatment is recommended when craniosynostosis affects the shape of the head in a significant way. The more the shape of the head is affected, the greater the worry about the effect on the child's brain. Surgery is believed to give a child the best possible chance to develop normally. It also normalizes the child's appearance. An unusual head shape can have profound effects on a child's personality, self esteem, and social interactions.

WHAT TYPE OF SURGERY IS RECOMMENDED?

The goal of surgery is to expand the volume of the skull, thereby relieving pressure on the brain. This will create a normal head shape. The operation is called "cranial vault remodeling." Current evidence suggests that the skull does not grow normally in the affected region. A slight overcorrection of the head shape is performed at the initial surgery to allow for future growth.

Surgery is performed by the craniofacial surgeon and the pediatric neurosurgeon working together. Each specialist performs the part of the operation where he or she is most expert. Working efficiently together as a team, the surgeons are able to reduce the amount of time a child requires under anesthesia. At our center, only specially trained pediatric anesthesiologists experienced in craniosynostosis work with our children.

AT WHAT AGE IS SURGERY RECOMMENDED?

Surgical treatment is generally performed before the child reaches a year of age. Exact timing is based on which suture or sutures are fused, and can vary from 4 to 10 months of age. If all the sutures are fused, earlier treatment may be recommended. When children are diagnosed after 1 year of age, operative treatment may be recommended at the time of diagnosis.

ONCE THE SURGERY IS SCHEDULED, WHAT SHOULD FAMILIES EXPECT?

Families are given as much time as needed to feel comfortable with the treatment plan and have all their questions answered. In addition, families are provided with written information about the surgery and hospitalization. Being prepared for surgery can help reduce your family's stress and there are many ways to help you and your child prepare for surgery. Concentrate on preparing yourself. If parents feel at ease, their child is usually able to sense this and reacts in the same way. Guidance is available on how to discuss treatment with older children and siblings on our website.

There is a chance that a child may require a blood transfusion. If a family chooses to donate blood, our coordinators can help set up the donation at the best time. A drug called erythropoietin may help reduce the need for a blood transfusion. The use of this drug is discussed with families, and written information is provided.

WHAT SHOULD FAMILIES EXPECT ON THE DAY OF SURGERY?

Children go to sleep by breathing in an anesthetic gas. Nothing that may cause pain to a child is done while awake. It takes about two hours to put in place all the IVs and monitoring equipment needed to keep children safe. The actual cranial vault remodeling procedure at our center takes 3 to 4 hours.

During the surgery, no hair is shaved. The incision is made in a wavy pattern. The wavy line allows the hair to cover the scar naturally even when the hair is wet. Every effort is made to keep the incision away from the ear so that it will be less likely to be seen with a short hair cut. We do not use bandages or plastic drains in young children.

Families are given a pager to carry and given updates every hour. After the operation is completed, children are brought to the pediatric intensive care unit (PICU) for careful monitoring. Most children spend a total of two nights in the hospital. At home, families are asked to wash their child's hair every other day. Regular baby shampoo is used. There is no other special care needed. Follow up appointments are scheduled 2-3 weeks after the surgery.

JENNIFER L. RHODES, MD

RUTH TRIVELPIECE, MD

GARY W. TYE, MD

JOANN TILLET, RN

FOR MORE INFORMATION, CONTACT THE VCU CENTER FOR
CRANIOFACIAL CARE AT 828-3042 OR VISIT OUR WEBSITE

