Craniosynostosis

What is craniosynostosis?

The skull is made up of 8 bones and joints where the bones of the skull meet (sutures). These sutures allow the skull to grow as an infant grows and develops. Over time, the sutures close and the bones fuse together. This forms the skull into a solid piece of bone.

Craniosynostosis is a condition where one or more of the sutures close too early. This may cause:

- Problems with normal brain and skull growth
- More pressure than normal inside the head
- Skull or facial bones to become irregular in shape

The problem occurs in 1 in every 2,000 live births. It affects boys slightly more often than girls.

What causes craniosynostosis?

Craniosynostosis most often happens by chance. But it may occur more often in some families. It may be passed down (inherited) in 1 of 2 ways:

- **Autosomal recessive.** This means that both parents carry the gene that causes the condition. Carrier parents have a 1 in 4 chance to have a child with craniosynostosis with each pregnancy.
- **Autosomal dominant.** This means that only one parent must carry the gene. The parents have a 1 in 2 chance that each child will have the condition.

The condition may also be part of many other genetic syndromes.

There are many types of craniosynostosis. Different names are given to the various types. The types are named by which suture or sutures are involved. They are also named by how the shape of the skull is affected. Common types of craniosynostosis are listed below.

Plagiocephaly

This refers to a twisted skull shape. The forehead and brow on the affected side are flat. The forehead on the opposite side may stick out. The back of the head may also be flat. The eye on the affected side may also have a different shape. This type can be caused by an early fusion of the coronal or lambdoid sutures.

But in the most common form of plagiocephaly, none of the sutures are fused. This form is called positional plagiocephaly. It is not linked with any early suture closure. Rather, it happens when a baby always lies on the same area of the back of their head. Some babies with this form have a motor disorder that prevents them from changing position when they are lying down.

Trigonocephaly

This means a "triangle-like" shape skull in which a ridge may stick out from the forehead. The eyes may be close together, and the forehead may look pointed and narrow. It is caused by the fusion of the forehead (metopic) suture. This suture runs from the top of the head down the middle of the forehead, toward the nose.

Scaphocephaly

This means a skull that is long and narrow from front to back and narrow from ear to ear. It is caused by early fusion of the sagittal suture. This suture runs front to back, down the middle of the top of the head. This is the most common type of craniosynostosis.

What are the symptoms of craniosynostosis?

The first and only symptoms may be changes in the shape of your baby's head and face. A child may also have:

- One side of the face that is not the same when compared with the other side
- Small or absent soft spot on the top of the head (anterior fontanelle)
- Higher pressure than normal in the skull. But this isn't common. It is especially true when more than one suture fuses.

Symptoms of increased pressure in the skull include:

- Full or bulging soft spot on the top of the head (anterior)
- Sleepiness or baby is less alert than usual
- Very noticeable scalp veins
- Increased irritability or fussiness
- High-pitched cry
- Poor feeding
- Projectile vomiting
- Increasing head size
- Seizures
- Bulging eyes or trouble looking
- Poor development
- Trouble breathing (in certain cases when multiple sutures are involved)

The symptoms of craniosynostosis may look like other health conditions. Make sure your baby sees a healthcare provider for a diagnosis.

How is craniosynostosis diagnosed?

Craniosynostosis may be present at birth (congenital). Or it may be found later, during a physical exam. The diagnosis is based on a physical exam. It includes measuring your child's skull. During the exam, your child's healthcare provider will ask a lot of questions about your pregnancy and your child's birth. You may be asked if you have a family history of head or face defects. You may also be asked about your child's development, because the condition can cause delays. Developmental delays may need more medical follow-ups.

Diagnostic tests are no longer routinely done for this condition. But your baby may need these tests to confirm the diagnosis of craniosynostosis:

- X-rays of the head
- CT scan of the head, which shows more detail than X-rays

An ultrasound during pregnancy can sometimes diagnose craniosynostosis before the baby is born.

How is craniosynostosis treated?

The key to treating craniosynostosis is early detection and treatment. Treatment will depend on your child's symptoms, age, and general health. It will also depend on how severe the condition is.

When the diagnosis is made before a child is age 1, surgery is usually the recommended treatment. The goal of surgery is to fix the deformities of the face and skull bones. It can also ease pressure in the skull, if it is present.

It is usually best for your baby to have the surgery before they reach age 1. This is because the bones are still very soft. It is easier to fix the deformities at that time. Your baby may need surgery at a much earlier age if the condition is more serious. Most surgeries are done between ages 3 and 8 months old. Before surgery, your child's healthcare provider will explain the procedure. After surgery, your baby may need to wear a helmet for several months to help recovery.

When the diagnosis is made after 1 year of age, nonsurgical treatment is often recommended if the skull's shape appears close to normal.

Positional plagiocephaly is usually not treated surgically. Parents are instructed to increase the amount of tummy time when the baby is awake. The baby should continue to sleep on their back. A helmet can be a choice.

What are possible complications of craniosynostosis?

Some forms of craniosynostosis can affect the development of the brain.

How can I help my child live with craniosynostosis?

Most children who have surgery early live healthy lives. But long-term complications may occur. A child with craniosynostosis needs regular medical checkups to make sure that the skull, facial bones, and brain are developing normally. Your child's healthcare provider, surgeon, and other specialists will work with you and your child. The medical team will work with your family to provide education and guidance.

Your child's provider may recommend genetic counseling. This is to check for genetic conditions that may be present in your family.

Key points about craniosynostosis

- Craniosynostosis is a condition where one or more of the sutures of the skull close too early. It causes problems with normal brain and skull growth.
- Craniosynostosis usually occurs by chance.
- The first and only symptoms are usually changes in the shape of the baby's head and face.
- Surgery is usually the recommended treatment.
- Most children who have surgery early live healthy lives.

Next steps

Tips to help you get the most from a visit to your child's healthcare provider:

- Know the reason for the visit and what you want to happen.
- Before your visit, write down questions you want answered.
- At the visit, write down the name of a new diagnosis and any new medicines, treatments, or tests. Also write down any new instructions your provider gives you for your child.
- Know why a new medicine or treatment is prescribed and how it will help your child. Also know what the side effects are.
- Ask if your child's condition can be treated in other ways.
- Know why a test or procedure is recommended and what the results could mean.

- Know what to expect if your child does not take the medicine or have the test or procedure.
- If your child has a follow-up appointment, write down the date, time, and purpose for that visit.
- Know how you can contact your child’s healthcare provider after office hours, and on weekends and holidays. This is important if your child becomes ill and you have questions or need advice.