

# What is Craniosynostosis?

## A little background information:

- Newborns and young babies have skulls made up of 5 separate bones. The areas in the skull where these bones meet are referred to as sutures. These sutures are what allow the skull to compress as the baby passes through the birth canal and for the head and brain to grow properly as the child grows.
- Craniosynostosis is an early growing together, or fusion, of two or more bones along these sutures. The location, number of fusions, and when the fusion occurs will all affect the shape of the baby's head. This fusion occurs in approximately 1 in 2000 births and affects boys more often than girls.
- Once a suture is fused, the brain continues to grow in the direction of least resistance. This limits the skull's ability to grow normally. When the skull is unable to grow in all directions, it will eventually take on a very abnormal shape and this abnormality will worsen with time.

## Types of craniosynostosis:

There are many different types of craniosynostosis.

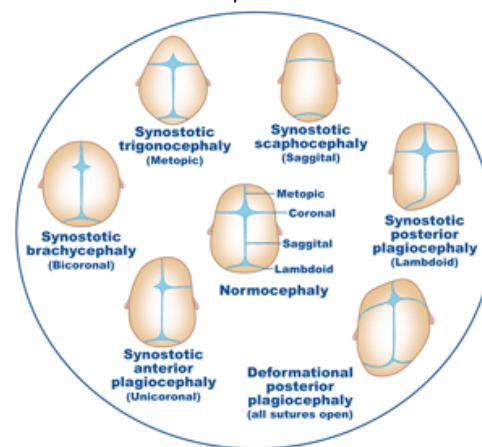
Some of the most common types are:

- **Sagittal synostosis (scaphocephaly)**  
Premature fusion of the suture at the top of the head (sagittal suture) forces the head to grow long and narrow, rather than wide. Scaphocephaly is the most common type of craniosynostosis, and it is more common in boys.
- **Coronal synostosis (anterior plagiocephaly):**  
Premature fusion of one of the sutures that run from each ear to the sagittal suture on top of the head may force your baby's forehead to flatten on the affected side. It also may raise the eye socket and cause a deviated nose and slanted skull. This second most common type of craniosynostosis is more common in girls. Untreated, it may lead to vision loss on the affected side.
- **Bicoronal synostosis (brachycephaly):**  
When both of the coronal sutures fuse prematurely, your baby may have a flat, elevated forehead and brow.
- **Metopic synostosis (trigonocephaly)**  
This form of synostosis is relatively uncommon (less than 10% of cases) and is characterized by a bony ridge in the midline of the forehead, a triangularly shaped head, a narrow forehead and eyes that are positioned close together.

## Lambdoid synostosis

In lambdoid synostosis, there is a flattening at the back of the skull and the ear is towards the back of the head.

A diagram of different craniosynostotic skulls and their characteristic head shapes:



## What causes craniosynostosis?

While the cause of many cases of craniosynostosis is unknown, in cases related to a hereditary genetic syndromes the cause is due to an abnormal chromosome or gene. Suspected physical causes may include intrauterine restraint or early engagement of the head.

## How is craniosynostosis diagnosed?

If a baby's skull is abnormally shaped, a ridge is present along one of the cranial sutures, a soft spot (fontanel) is closed, or if they have an abnormality in their facial appearance, their examining physician may suspect craniosynostosis. The doctor will order an X-Ray or CT scan to rule out or confirm craniosynostosis as the cause.

## What problems can craniosynostosis cause?

The biggest concern is a buildup of pressure within the brain cavity inside the skull, this is called increased intracranial pressure. If an excess of pressure builds up inside the skull, it may limit the brain's development, lead to blindness, cause seizures or very rarely death. The appearance of the skull is also a concern when a child has craniosynostosis. If left untreated, the head will continue to grow improperly and leave the child with a permanently deformed skull. Depending on the severity of the deformity, vision and/or breathing problems can occur.

## My child has craniosynostosis, how is it treated?

Due to the nature of craniosynostosis, and the strength of the fused suture, surgery is currently the only effective treatment available. There are different types of surgery you may discuss with your physician. In the procedure, a surgeon will remove parts of the skull so that it may grow normally and reduce the chance of increased intracranial pressure. This is typically accomplished in one procedure with most children going on to live completely normal lives. Most doctors recommend the surgery be performed between 3 and 8 months of age. The traditional procedure is extremely reliable and has a low risk of complications.

## What do I do now?

- Enjoy your baby!
- Ask your pediatrician about locations for treatment and for some names of surgeons who may be able to perform the surgery.
- Talk to someone who's been there. Find someone who has gone through this or a support group to talk to. Ask your physician if they know of one in your area.
  - <http://www.cappskids.org> and <http://www.craniokids.org> are two such support groups online.