

# Craniosynostosis

BIRTH DEFECT RESEARCH FOR CHILDREN



## What is Craniosynostosis?

Craniosynostosis occurs when the bones of the baby's skull fuse together before the brain has stopped growing. The top of a baby's skull consists of five bones that are held together by fibrous material called sutures. These sutures should stay open while the baby's brain is still growing. If even one of the sutures closes prematurely, it can result in a misshapen skull or face. Craniosynostosis can happen before the baby is born or during the first few months of life.



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How many children have Craniosynostosis?

Three to five babies in every 1,000 live births have closure of the sagittal suture, the most common form of Craniosynostosis. This condition is more common in males.

What are the causes?

Craniosynostosis may be part of a chromosomal or genetic syndrome. It may also occur spontaneously.

Some cases may be associated with other disorders such as microcephaly (an abnormally small head) or hydrocephalus (the excessive accumulation of cerebrospinal fluid in the brain).

Two studies have reported that prenatal cigarette smoking may increase the risk of certain types of Craniosynostosis.

When is Craniosynostosis usually detected?

Symptoms may occur from birth through the first year of life.

Prenatal ultrasound imaging can show closed sutures and can detect an oval-shaped head.

How is Craniosynostosis treated?

Treatment usually consists of surgery performed early in a baby's life for the following reasons:

- to relieve pressure inside the skull;
- to make room for brain growth;
- to improve the appearance of the baby's head.

During traditional surgery, the top of the skull is removed. After the surgeon reshapes the bone, it is replaced over the brain. The surgery usually lasts several hours; requires blood transfusions and includes at least a five-day hospital stay. Swelling is common after this form of surgery.

Children's Hospital at the University of Missouri Health Sciences Center has developed a new treatment for Craniosynostosis called an endoscopic strip craniectomy (ESC) procedure. This procedure only requires small incisions to remove closed sutures so it results in little blood loss and a much shorter hospital stay. After three years of use, follow-ups reveal that the procedure's effectiveness is quite good. Cost of ESC is one-third that of traditional treatment. ESC, however, must be done by at least six months of age and the baby must wear a specially fitted molding helmet for a few months after surgery so the head will grow normally.

What is the prognosis for babies with Craniosynostosis?

The outlook for babies with Craniosynostosis is good with appropriate treatment; 75% of children have satisfactory results. Babies who have single suture closures have a better prognosis than those with closure of multiple sutures or if other associated abnormalities occur.

The most common form of Craniosynostosis is sagittal synostosis. Sagittal synostosis is not associated with increased cranial pressure or mental retardation. Other forms of Craniosynostosis, however, may be associated with mental retardation, seizures, or blindness.

Can Craniosynostosis be prevented?

Genetic counseling is recommended to detect chromosomal and genetic forms of Craniosynostosis. Nothing is known to prevent isolated cases of the disorder.

Pre-natal exposure to cigarette smoking should be avoided.



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The National Institute of Neurological Disorders and Stroke (NINDS) conducts and supports on-going research into neurological development that may offer hope for new ways to treat and prevent congenital anomalies, including Craniosynostosis.

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