

a guide to understanding  
**craniosynostosis**

a publication of children's craniofacial association

## a guide to understanding craniosynostosis

**T**his parent's guide to craniosynostosis is designed to answer questions that are frequently asked by parents of a child with craniosynostosis. It is intended to provide a clearer understanding of the condition for patients, parents and others.

## how can children's craniofacial association (cca) benefit my family?

**C**CA understands that when one family member has a craniofacial condition, each person in the family is affected. We provide programs and services designed to address these needs. A detailed list of CCA's programs and services may be found on our Web site at [www.ccakids.com](http://www.ccakids.com) or call us at 800.535.3643.

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This booklet is intended for information purposes only. It is not a recommendation for treatment. Decisions for treatment should be based on mutual agreement with the craniofacial team. Possible complications should be discussed with the physician prior to and throughout treatment.

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## what Is craniosynostosis?

**C**raniosynostosis is a medical term that literally means fused bones of the skull. It is a condition that some children are born with or later develop. The skull is abnormally shaped because of the fusion of skull bones.

To better understand craniosynostosis, it is helpful to know that our skulls are not made up of one single “bowl” of bone. Instead, different bones that fit together like a jigsaw puzzle make up the skull. The areas where the bones meet one another are called sutures. As a baby grows, the brain rapidly increases in size. According to current theories of growth, the growing brain pushing on the bones of the skull causes the skull bones to expand or grow. Much of this growth occurs in the areas of the sutures where the bones meet. When one of the sutures fuses, it is called craniosynostosis. There will be no growth in this area. This inability to grow in one area may lead to overgrowth in another area. This results in an abnormally shaped skull.

## how do I recognize this condition in my own child?

**T**he diagnosis of craniosynostosis can only accurately be made by X-rays such as a CT scan. However, any child with an unusually shaped head is certainly suspect. Some children will experience a marked improvement in head shape when they begin to sit up and support their heads alone. However, sometimes this condition will worsen with growth. Sometimes it will remain the same. Another sign some parents note is a small ridge of bone that may run along the skull in different locations. Early closure or sealing of the fontanelle (“soft spot”) may be another sign of craniosynostosis.

## what kinds of craniosynostosis are there?

There are numerous types of craniosynostosis. Different names are given to the various types of craniosynostosis. The names depend on which suture or sutures are involved. This booklet will discuss plagiocephaly, trigonocephaly, scaphocephaly, and Crouzon syndrome. There are other syndromes involving craniosynostosis. Most of these other syndromes, however, are similar in some way to the conditions discussed here.

Plagiocephaly occurs most often. It happens in approximately one out of 2,500 births. It involves fusion of either the right or left side of the coronal suture. Normally, the coronal suture extends from ear to ear over the top of the head. The fusion, or early closure, of the coronal sutures on one side causes the normal forehead and brow to stop growing forward. This causes a child with plagiocephaly to look as if the forehead and brow are pushed backward. The eye on the affected side also has a different shape.

Trigonocephaly is a fusion of the metopic suture. This suture runs from the top of the head, down the middle of the forehead, toward the nose. Early closure of this suture may result in a prominent ridge running down the forehead. Sometimes the forehead looks quite pointed. It resembles the bow of a boat. Frequently, the eyes are closer together.

Scaphocephaly is an early closure or fusion of the sagittal suture. This suture runs from front to back, down the middle of the top of the head. This fusion causes a long, narrow skull. The skull is long from front to back and narrow from ear to ear.

Crouzon's involves fusion of both sides of the coronal suture. This suture runs from ear to ear, over the top of the head. This fusion prevents the entire forehead from growing in a forward direction. This results in the brain pushing the top of the skull higher. It then leads to a flattened, tall forehead. The bones protecting the eye are also kept from growing forward. This makes the eyes look very large.

## how do these syndromes occur?

**a**t present, no one is sure why these birth defects occur. Studies do not show that there is anything in particular the mother did or did not do which results in these defects. Most likely, some accident occurred very early in development to one of the baby's genes. In the normal population, plagiocephaly occurs in one of 2,500 births. This is the most common form of craniosynostosis. Some of the more rare craniosynostosis happen one in 50,000 births. If one child has craniosynostosis, there is a slim chance that a second child will have this problem. The chances are between 0 and 4%. When your child with craniosynostosis grows up, the chance of having a child with craniosynostosis is just as small. Of the types of craniosynostosis discussed here, Crouzon syndrome is the exception to the rule. When Crouzon syndrome develops, children with this condition have a 50% chance of passing it on to their children. For example, if a person with Crouzon's has four children, it is expected that two children would also have the syndrome.

## what are the treatments available for craniosynostosis?

**m**any children with craniosynostosis do not need any treatment. Each of the different types of craniosynostosis can occur in various degrees of severity. In the mildest form of craniosynostosis, only a small ridge can be felt. There is no abnormal skull shape. In some cases, the problem will worsen with growth. For some, it will stay the same. For others, it will improve with time.

Children with obvious deformities should be treated. Those children with deformities that are predicted to worsen should also be treated. One of the greatest concerns is intracranial pressure. As the brain is growing, it needs to be able to push the skull bones apart, giving it more room to grow. If there is a fusion of a suture, as in craniosynostosis, growth is restricted, and the brain is squeezed. As the brain grows larger, the skull cannot expand. This may lead to a buildup of pressure inside the skull. This increased pressure may cause a delay in development or a permanently damaged brain.

## if my child needs surgery, when is the best time to operate?

**T**he timing of surgery varies with the type of craniosynostosis and with the severity of the deformity. Generally, it is best to wait until the child is at least 2-3 months old, as there may be a lower risk at this age. When surgery is performed at less than one year of age, the results are usually better than when performed later. With the exception of certain syndromes, one operation will correct the craniosynostosis. About 10% to 20% of patients need a second operation later to correct small remaining deformities.

## where is the best place to have my child treated?

**C**raniosynostosis is a complex problem that requires the expert skill of many different specialties working together. These problems are best treated by large craniofacial teams experienced in the management of these patients. Centers with large craniofacial teams working together have the advantage of a greater experience. This definitely leads to better results and fewer complications. In addition, ongoing research at these centers offers patients the latest breakthroughs in treatment. As there are only a few experienced centers in the country, it is quite common for families to travel quite some distance to get the best care. Children who are treated locally by inexperienced teams or by individual physicians not working together as a team, are more than likely to have unsatisfactory results. It sometimes requires two or three additional operations to correct what has been done. Another advantage of traveling to busy centers is the opportunity to meet other families and children affected with similar problems who can offer advice. These families often share their experiences, which provide moral support.

## what is the surgical procedure for repairing this condition?

**t**he surgical technique for correcting the problem varies with the type of craniosynostosis, but all have certain things in common. Surgery is only considered for these children after a pediatrician, trained in this field, certifies the child can tolerate the anesthesia and the operation. One of the greatest risks to the child comes from the general anesthetic. It is necessary for an anesthesiologist, well experienced in this type of surgery in young children, to be present during the entire procedure. The surgery is usually performed by two specialists working together. One is a craniofacial surgeon and the other is a pediatric neurosurgeon. The craniofacial surgeon is a plastic surgeon who has received additional training in pediatric craniofacial surgery. It is common for an incision to be made in the hair from one ear to the other ear, across the top of the head. This is usually the only scar from surgery. The hair usually hides the scar. After this incision is made, the neurosurgeon removes the affected areas of the skull and forehead. The craniofacial surgeon reshapes these bones and returns them to a normal position. Once the procedure is finished, the incision is closed—usually with dissolving sutures. The child is then taken to the pediatric intensive care unit.

The routine is different among the various centers. Children typically spend the first night or two in the intensive care unit. They then go to the regular pediatric floor. Children are normally sent home on the third to fifth day following surgery. Generally, children experience only minor discomfort from this operation. There is little pain from the cutting of skull bone. By the second day after surgery, most children need nothing more than Tylenol®. It is also common for both eyes to swell shut for about three days after surgery. Not being able to open one's eyes annoys the child the most. After the child is discharged from the hospital, the family may be asked to stay in the area for another few days before returning home. This allows the treating doctors to make sure there is a good chance that there will be no major complications. Between six weeks to three months after surgery, the child returns for follow-up visits. The surgeon usually sees the child once a year thereafter.

