a guide to understanding

# craniosynostosis

a publication of children's craniofacial association

## a guide to understanding craniosynostosis

this 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?

CCA 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 website at www.ccakids.org or call us at 800.535.3643.

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## what is craniosynostosis?

Craniosynostosis is a medical term that means premature fusion of the bones in the skull. It is a condition that some children are usually born with or more infrequently later develop. Since normal head shape and growth rely on the joints between these bones to remain open, early fusion seen in craniosynostosis results in an abnormal shape.

To better understand craniosynostosis, it is helpful to know that our skulls are not made up of one single "bowl" of bone. Instead, the skull is made of many bones that fit together like a jigsaw puzzle. The areas where the bones meet one another are called sutures. As a baby grows, the brain rapidly increases in size pushing on the bones of the skull to expand. Since the sutures are the growth centers of the skull, suture fusion results in no growth in that area. This inability to grow in one area may lead to overgrowth in another area. This results in an abnormally shaped skull.

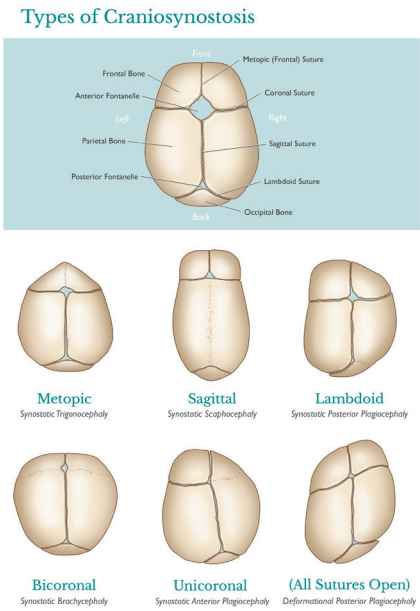
## how do I recognize this condition in my own child?

The first clue that a child has craniosynostosis is an abnormally shaped head. However, not all abnormally shaped heads are from craniosynostosis. Preferential head positioning while laying in their crib and weak or stiff neck muscles can result in persistent pressure on the back of the head and flattening. This condition is called positional plagiocephaly and usually improves once the child begins to sit up and support their own head. While this condition does not require surgery, occasionally physical

therapy or molding helmets are beneficial, but must be determined on a case-by-case basis. Most pediatricians are qualified to determine if abnormal head shape is from positional plagiocephaly or craniosynostosis. Other signs some parents note are small ridges of bone running along the skull or early closure of the fontanelle or "soft spot" before one year of age. However, when in question, referral to a craniofacial surgeon may be required. Definitive diagnosis of craniosynostosis is then often confirmed with an X-ray such as a CT scan.

## what kinds of craniosynostosis are there?

There are numerous types of craniosynostosis, each with a different name. In general, the types of craniosynostosis are subdivided into syndromic and nonsyndromic categories. Nonsyndromic craniosynostosis is much more common and usually involves premature fusion of only a single suture and without any associated medical problems. Nonsyndromic craniosynostosis is currently thought to be a spontaneous defect without underlying



genetic abnormality. Syndromic craniosynostosis is much more rare and usually involves premature fusion of two or more sutures, associated with other medical problems, and often linked to genetic mutations.

Different names are used to describe the various types of craniosynostosis and depend on which suture or sutures are involved. This booklet will discuss scaphocephaly, anterior deformational plagiocephaly, brachycephaly including Crouzon syndrome, trigonocephaly, and posterior deformational plagiocephaly. There are many other syndromes involving craniosynostosis, including Apert, Pfeiffer, and Crouzon syndromes, which are covered in more detail in separate booklets.

**Scaphocephaly** is the most common type of craniosynostosis and results from 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. This is also referred to as sagittal craniosynostosis.

**Anterior deformational plagiocephaly** occurs next most commonly, involving fusion of either the right or left coronal suture. The coronal suture extends from ear to ear over the top of the head. The fusion of the coronal sutures on one side causes the forehead and brow to stop growing forward. This results in a child with anterior plagiocephaly to look as if the forehead and brow are pushed backward on the affected side. The orbit on the affected side also has a different shape and the tip of the nose is often pointed slightly away from the affected side as well. This is also referred to as unicoronal craniosynostosis.

**Brachycephaly** involves fusion of both the left and right coronal sutures. 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, wide, and tall forehead. The bones protecting the eyes are also kept from growing forward making the eyes look very large. This is also referred to as bicoronal craniosynostosis and seen in many types of syndromic craniosynostosis such as Crouzon syndrome.

**Trigonocephaly** is the third most common type of craniosynostosis and results from fusion of the metopic suture. This suture runs from the top of the head, down the middle of the forehead, towards the nose. Early closure of this suture may result in a prominent ridge running down the forehead. Sometimes the forehead looks quite pointed, resembling the bow of a boat. Frequently, the eyes are closer together. This is also referred to as metopic craniosynostosis.

**Posterior deformational plagiocephaly** is the least common type of craniosynostosis resulting from fusion of the right or left lambdoid sutures at the back of the skull. This results in flattening of the back of the head and downward displacement of the ear on the fused side. There is also commonly bulging on the side of the skull of the non-fused side. This is also referred to as lambdoid craniosynostosis.

## how do these syndromes occur?

At present, no one is sure why these birth differences occur in nonsyndromic craniosynostosis. Studies do not show that there is anything in particular the mother did or did not

do which results in these differences. Overall, in the normal population, craniosynostosis occurs in one of 2,500 births. 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 a child with craniosynostosis grows up, the chance of having a child with craniosynostosis is just as small. Of the types of craniosynostosis discussed here, syndromic craniosynostosis is the exception to the rule. When Apert, Crouzon, or Pfeiffer syndromes develop, children with this condition have a 50% chance of passing it on to their children. For example, if a person with Crouzon has four children, it is expected that two children would also have the syndrome.

## who should receive surgery for craniosynostosis?

As previously discussed, not all abnormal head shapes are created by craniosynostosis. Positional plagiocephaly is the most common reason for abnormal head shape, but does not require surgical correction since there is no restriction of head growth. Craniosynostosis, on the other hand, does restrict head growth, which can lead to increased pressure around the developing brain because the skull cannot expand. Evidence suggests that prolonged increased pressure around the brain can cause developmental delay. For this reason, surgery is recommended for all children with craniosynostosis to allow proper head growth and avoid developmental delay.

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

The timing of surgery varies with the type of craniosynostosis and with the severity of the difference. Generally, it is best to wait until the child is at least 3 months old, which minimizes surgical risk. The type of surgery recommended also affects timing, with some procedures better performed closer to one year of age. However, when surgery is performed within the first year of life, the results are usually better than when performed later. With the exception of certain syndromes, usually only one surgery is required to correct craniosynostosis. About 10% to 20% of patients need a second smaller operation later to correct small remaining differences.

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

Craniosynostosis 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 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 relatively few experienced centers around the country, it is quite common for families to travel quite some distance to get the best care.

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

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.

The type of craniosynostosis affecting a child and the age at which it is discovered help determine which type of surgery should be performed. Very basically, the timing of surgery varies with the technique that a surgeon prefers, but will likely occur between 12 weeks and as late as 15 months of age.

In general, children with sagittal and metopic craniosynostosis that are identified early in life (< 3 months of age) can be treated in a minimally invasive fashion in a procedure call **endoscopic assisted suturectomy**. In this procedure, one or two small incisions are made in the scalp and a piece of skull is removed to include the fused suture. This allows the skull to then continue to expand in a normal fashion by the rapidly growing brain at this young age. The piece of skull removed eventually heals after head growth slows. However, a molding helmet is required to

recreate a normal head shape from the distortion caused by the craniosynostosis. This is usually worn for 6 – 8 months following surgery, and needs to be worn at all times unless bathing. The helmet is adjusted several times as the child grows, which requires multiple visits to the surgeon and helmet specialist, called an orthotist. Molding helmet therapy is as important as the surgery itself to create a good result, so a family's ability to follow this treatment plan should be carefully considered when choosing this surgical option.

Sagittal and metopic craniosynostosis children who are identified after 3 months of age or are not unable to undergo molding helmet therapy, in addition to children with coronal and lambdoid craniosynostosis, are better treated later in life with an **open procedure**. It is common for a zigzag incision to be made in the hair from one ear to the other across the top of the head. This is usually the only scar from surgery and the hair usually hides the scar. After this incision is made, the neurosurgeon removes the affected areas of the skull or forehead. The craniofacial surgeon then reshapes these bones and returns them to a normal position. Once the procedure is finished, the incision is closed, usually with dissolving sutures. Since this surgery requires more extensive dissection, the amount of time required and blood lost during the procedure are both increased compared to a minimally invasive surgery. It is routine for children to require a blood transfusion during this surgery, which is often avoided in minimally invasive procedures. Thus surgery is usually performed between 6 – 12 months of age depending on the type of craniosynostosis.

The routine after surgery is different among the various centers. Children may spend the first night or two in the intensive care unit before going to the regular pediatric floor, or go to the pediatric floor directly. Children who undergo a minimally invasive surgery are often able to go home the following day. Children who undergo an open surgery are normally sent home on the second to fifth day depending on the surgery performed. 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 a significant amount of head swelling, which may cause 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 two weeks to six months after surgery, the child returns for regular follow-up visits. The surgeon usually sees the child once a year thereafter.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