Craniosynostosis
What is craniosynostosis?

Craniosynostosis is a condition that causes the skull to form an unusual shape.

The skull is formed by bones that fit together like a jigsaw puzzle. The skull gives shape to the head and protects the brain. The areas where the skull bones connect to each other are called sutures. The sutures allow the skull to expand as the brain and other parts of the head grow. In the first few years, the brain grows a lot. Nature allows for this by keeping the sutures "open" for a few years.

Craniosynostosis is the result of one or more of these sutures closing (fusing) too early. The spot where the sutures have fused can not expand as it normally would. This results in the skull forming an unusual shape.

Each suture has a different medical name. The different forms of craniosynostosis take their names from the suture(s) that have fused early.
Why have the bones fused?

About one in 2,000 babies are born with craniosynostosis. Why this happens is, in most cases, not known. Medical research is looking into some possible causes:

- Genetics: perhaps a gene (inherited pattern of growth)
- Pressure on the head while in the womb: perhaps from the baby’s position or some other reason
- Biochemistry: perhaps there are unusual amounts of some of the hormones, proteins, and minerals etc. that occur naturally in our body.
- A part of another medical condition or a premature birth: about 5% of children with craniosynostosis have one of the craniofacial syndromes explained in this pamphlet.

95% of children with this condition have no other medical problems and no known cause for craniosynostosis.

Does it cause brain damage?

In most (not all) cases the brain can grow and develop normally despite the fused suture. The other open sutures will expand enough to prevent any pressure on the brain. In some cases all of the sutures fuse, which means that the baby’s brain will not have enough room to grow.

How is craniosynostosis diagnosed?

Most of the time, the doctor can diagnose craniosynostosis simply by looking at the shape of the head. Sometimes, as the baby grows, others may be the first to notice that the baby’s head is not a normal shape. They may also notice:

- a small ridge of bone running along parts of the skull,
- the child’s ‘soft spot’ (anterior fontanel), disappears before it is supposed to (usually closed between 18-24 months of age).

The family doctor will then refer the child to a pediatric neurosurgeon (brain and nerve specialist) and, perhaps, a craniofacial surgeon (specialist surgeon for the head and face). Their experience allows them to diagnose by examining the skull and looking at the head.

If the surgeon needs more information, s/he may ask for X-rays or CT scans of the skull. These are not usually needed but, the surgeon may want to see some images of the skull before deciding on the best way to correct the problem.

Common forms of craniosynostosis:

The shape of the baby’s head is a clue to which suture(s) have fused. The common forms have only one fused suture and do not involve the face. Most craniosynostosis can be corrected with surgery. 90% of baby’s with only one fused suture can have their craniosynostosis repaired with one operation.

If the craniosynostosis affects more than one suture and also the shape of the face, more than one surgery may be needed to get the best results. A plastic surgeon and a neurosurgeon work together as an expert team to reshape the head and face.
1. Sagittal Synostosis (Scaphocephaly):

This is the most common form of craniosynostosis treated at BC Children’s Hospital. The sagittal suture runs front to back, along the middle of the head. Fusion of this suture causes a long narrow skull. Sometimes the front and the back of the head will bulge.

Surgical correction of sagittal synostosis:
The neurosurgeon removes the bone around the sagittal suture and widens the skull by opening the coronal and lambdoid sutures on both sides of the head. The surgeon may place pieces of the bone that was removed back into the new spaces, which will slowly grow in and reshape the head. Babies are usually 4-12 months old when they have their surgery.

2. Coronal Synostosis (Plagiocephaly or Brachycephaly):

This is another common form of craniosynostosis. There are left and right coronal sutures that run from the ear to the fontanel at the top of the head. One or both of these sutures may fuse.
- If the suture is fused on only one side, **plagiocephaly** the forehead and brow seem to arch in, rather than out, in the normal way. Plagiocephaly may be caused by fusion of both the coronal or lambdoidal suture. **Positional plagiocephaly** may not require surgical intervention and alternative options should be discussed with your doctor.

- When both sides are fused **(brachycephaly)**, the head is flatter and wider than normal. The eyes bulge further out of the skull. The skull, around the ears, bulges. This is the fusion found most often in Crouzon’s and Apert’s Syndromes.

Diagram of plagiocephaly, in which one coronal suture (right side) has closed prematurely and needs surgical correction.

Diagram of brachycephaly, in which both coronal sutures had closed prematurely and needed repair.
4. **Lambdoid Synostosis (Lambdoidal stenosis):**

The lambdoid suture is located at the back of the head and runs from side to side. If both sutures fuse, the back of the head becomes flat, pushing the sides of the skull out. Lambdoid synostosis is mainly caused by pressure on the back of the skull from positioning. Some doctors believe that the best treatment for this is to use skull 'moulding' devises which reshape the skull as the child grows. The doctor may provide advice on how to position your child in a way that decreases the pressure on the back of the skull and prevent the flattening of the back of the head. Your doctor will discuss the best option for your child, if this is a concern. **Positional plagiocephaly** is another term used to describe misshapen skulls that may be treated without surgery.

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3. **Metopic Synostosis (Trigonocephaly):**

The metopic suture runs from the top of the head, down toward the nose. If this is fused, the child will have a large ridge running down the forehead. The forehead will look pointed rather than rounded. In many children the difference in shape is so slight that they do not need surgery. But, if the forehead makes the child look odd, surgery can be done to correct it.

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*Surgical correction of metopic synostosis:*

The bone around the fused suture is removed making more room. Then the bones of the forehead are shaved and rounded. It is best to do the surgery when the child is between 6 months and 1 year of age.
Craniosynostosis syndromes:

There are some forms of craniosynostosis that are part of a more complex condition. Besides the fusion of sutures, these children also have other health problems. When problems come as a "set", and are present at birth, the conditions are known as syndromes. Syndromes of this kind are caused by genes that have somehow changed their normal form. Apert and Crouzon are the most common of the craniosynostosis syndromes. Others include Pfeiffer, Saethre-Chotzen and Carpenter's syndrome.

These syndromes are very similar. They involve:
- Two or more fused skull sutures as well as fusion of other bones in the head and face.
- Misshapen face bones - for example eye sockets that are too shallow.

Other common problems are:
- Weakness in some of the muscles that move the eyes causing visual problems.
- Blocked or misshapen ear canals causing ongoing infections that lead to hearing loss.
- Cleft palate (openings in the roof of the mouth) and blocked nasal passages causing breathing difficulty and changes in speech sounds and patterns.

The fingers and toes of children with Apert's are webbed so the hands look like mittens rather than gloves.

Corrective surgery for craniosynostosis syndromes:

Corrective surgery for children with these syndromes is done in stages over several years. A plastic surgeon and neurosurgeon work as a team. The children have their first surgery by age one. This opens the sutures and gives the brain room to grow. The shape of the face and skull may be reshaped a few times as the child grows. The aim is to get the best possible results.

About craniosynostosis surgery:

The care before and after surgery is similar for children with primary craniosynostosis and those with a syndrome which includes craniosynostosis.

Before surgery: meeting with the doctor:

- Well before surgery parents meet with the surgeons to discuss the surgery. The surgeons explain what they will do and outline the risks and the expected results of the surgery. A date for surgery will be set and you will be given information about the recovery process.
- This meeting is a chance for you to ask questions and discuss any concerns that you have. We suggest that you write your questions down before this meeting so you don’t forget what you want to ask. (Note: A manual called ‘Partners’ has a helpful section on how to ask health professionals questions and teach parents how to make informed decisions. The manual is also available in video. You can find both items in the Family Resource Library).
- You may be asked to sign the consent for surgery at your first meeting. If you need some time to think about the surgery, tell your doctor. Most times parents can take as much time as they need to provide consent.
- The doctor will also give you information on how to prepare your baby for the surgery. Please follow the directions carefully.

The surgery:

- The surgery takes from 2 hours (for sagittal synostosis) to 6 hours (if there is more than one fused suture and some additional reshaping that is needed).
- Your child will be admitted to Day-care Surgery and, when the operation is over, they will go to the post anesthesia care unit (PACU) until they are awake. Once your child is awake and
After the surgery:

- A nurse will check your child’s neurological status frequently following the surgery. This is to ensure there is no bleeding or swelling in the brain. Even though this may appear disruptive when your child is trying to sleep, it is very important to ensure your child is recovering and not experiencing any complications.

- Your child will have a large bandage on their head for at least 48 hours after surgery. This dressing protects the incision while it is healing. Your nurse will check the dressing often to ensure it is clean and dry.

- The first couple of days after surgery, your child will be given pain medication at regular times so that they are as comfortable as possible. Your doctor or nurse can explain the type of medication that your child will be receiving.

- There is often some blood loss during this kind of surgery. This is a concern because a child’s total amount of blood is small, so even a small loss can affect health and healing. The surgeon may order blood tests to check if your child has enough **haemoglobin** (Hgb) in their blood. Haemoglobin is part of the blood that carries oxygen to the body. Low **haemoglobin** is treated with an iron supplement or if the **Hgb** stays low, your child may need a blood transfusion. Less than 20% of children who have surgery at B.C.’s Children’s Hospital, for all types of craniosynostosis, need a blood transfusion. Blood transfusions are only given with your consent. Ask for the booklet that explains blood transfusions, if you have any questions.

- Your child’s head and face may be quite swollen after surgery. The swelling will get worse on the 2nd and 3rd day after surgery and will start to get better on the 4th day. It helps if you hold ice packs and cool cloths on the swelling. Keep the head of the bed up. If the head is up, the fluid and blood can drain away from the head. Sometimes, if the incision is close to the face, the eyes will swell shut. It is important not to let your child scratch their head or face if they are swollen.

- The incision is usually large and may be in a straight line or in a zig-zag pattern. The location of the incision will depend on what suture(s) are fused. Stitches or staples are used to close the wound. Although the incision looks very big at first, it will fade over a few months and as your child grows, the scar will be hidden by hair. Unfortunately, hair will never grow within the scar itself.

Going home

- The hospital stay is 3-5 days. Your child can go home once they are eating/drinking well and the swelling is starting to come down. Your nurse will give you a pamphlet called ‘Care at home after neurosurgery’ which will give you information about how to care for your child at home. This pamphlet is available from the Family Resource Library.

- The neurosurgeon and the plastic surgeon will want to see your child at 3 months and 1 year after the surgery. Talk to your doctor or nurse about making the follow-up appointments.
Common questions and concerns about craniosynostosis

1. When is the best time to operate?

For a very few babies, the timing of the surgery is not even a question. Because the amount of pressure in their skull (intracranial pressure or ‘ICP’) is life-threatening, they must have emergency surgery within the first two weeks of life. For other children, the surgery is not considered an emergency.

To achieve the best results, most surgeries should be done between 4 months of age and 1 year. The reason for this is that in the first year:

- the bones of the skull are softer and easier to shape.
- the tissues covering the brain can make new bone.
- the growing brain can continue to reshape the skull and face after the surgery.

2. Will my child’s brain be damaged?

Most babies with fused sutures have normal brain development and intelligence. Very few children are at risk of brain damage as a result of fused sutures. Some children with craniosynostosis syndromes may have some delay as part of their syndrome, not the craniosynostosis itself. Because surgical repair involves the brain and the skull, there are additional risks. Ensure that you discuss all of these risks with your doctor prior to your child’s surgery so that you can make an informed decision about your child’s treatment.

3. It is upsetting when people stare at my child!

This is a real challenge for most parents of a child with a facial difference, such as those with a craniosynostosis syndrome. Children will learn how to deal with their condition from your responses. Remind yourself that staring is a natural response to understand anything that looks different. There are several ways to deal with someone who is curious:

- You can make contact with the person – say hello, smile or introduce your child by name. This usually reminds the other person that your child is a real person.
- An explanation is often the best approach with other children. You can start by saying: “This is Kate. I can see you are wondering about her face. Sometimes faces or bodies or feet don’t turn out quite right and need a little fixing. Kate’s face needs a little fixing. Would you like to say hello to Kate? I bet you can make her smile.”
- If you don’t want to engage others you do not have to. Simply distract the person with some everyday chat-chat, or simply say nothing at all.

Good tip from a parent:
“I made a list of things that people said or did that upset me. I asked a friend to help me come up with as many responses I could for these situations. I practised saying a few of them in a clear voice looking right into the person’s face. I was well prepared and stares and comments did not bother me after that. Bonus was that over the years my child learnt how to respond. As he got older, he became a pro at answering back to teasing. His confidence won him lots of friends”.

- The main thing is not to let your anger, sadness or fear overcome you. Take control by making eye contact. If you can look comfortable you show others how you want them to be with your child. It takes courage. Your child will learn this confidence from you.

4. I worry about my child’s self image and how I can be supportive.

Most children with craniosynostosis will grow up without any sense of being different. The surgery will correct the condition before self-image becomes an issue.
Children with facial differences that will take many surgeries to reconstruct may have some social challenges. With your support your child can learn to cope and grow in courage and strength. Ask your nurse about different supports that are available to your child, you and your family. You may also contact the Family Resource Library for more information.

**Additional resources for families:**

*Children with Facial Difference: A Parent's Guide*
Author: Hope Charkins  
Published By: Monarch Books of Canada  
Trade Paperback  
Published: January 1996  
Resource to help parents cope with medical, emotional, social, educational, legal and financial challenges presented by facial differences of their children.

*Family Resource Library*  
BC Children’s Hospital  
Room K2-126, Ambulatory Care Building  
4480 Oak Street  
Vancouver, BC V6H 3V4  
tel: 604-875-2345 local 5102  
tollfree: 1-800-331-1533 ext. 2  
fax: 604-875-3455  
email: famreslib@cw.bc.ca  
web: www.cw.bc.ca/library