Craniofacial Ireland

RECONSTRUCTING FUTURES
PIONEERING CRANIOFACIAL SURGERY AND CARE

Sagittal (Scaphocephaly)

Sagittal Craniosynostosis

Sagittal craniosynostosis is the most common type of single suture (non-syndromic) craniosynostosis and occurs when the sagittal suture fuses before birth. The sagittal suture separates the bones on either side of the head from the midpoint of the top of the forehead at the anterior (front) fontanelle (soft spot) towards the back of the head where the posterior (back) fontanelle is located.

Isolated sagittal craniosynostosis is rarely associated with problems affecting other parts of the skull, face or body. The head shape which occurs because of sagittal craniosynostosis is known as scaphocephaly which means boat-shaped in Greek.

How common is sagittal craniosynostosis?

It is the most common type of premature suture fusion affecting approximately 1 in every 2000 births with a 3:1 male: female ratio which means that it is 3 times more common in boys.

What causes sagittal craniosynostosis?

The precise cause of sagittal craniosynostosis is not yet known. For some reason the immature bone cells in the suture, mature into adult bone cells.

There may be a genetic basis to this as it seems to be passed on from parent to child in a very small number of families, but the gene causing this has not yet been identified. If ultimately we do find a genetic basis to this condition, it is likely, that your child is a new case. The chances of passing this on however are very small. More research is needed to identify the cause of sagittal craniosynostosis.

What are the symptoms of sagittal craniosynostosis?

The main sign of sagittal craniosynostosis is an abnormal head shape. There is often a bony ridge over the prematurely fused sagittal suture. Depending on whether the entire sagittal suture has fused or only part of it, children may have a prominent (protruding) forehead and the back of the head (occipital region) may be quite pointed or ‘bullet’ shaped. The head shape can be quite long and narrow.

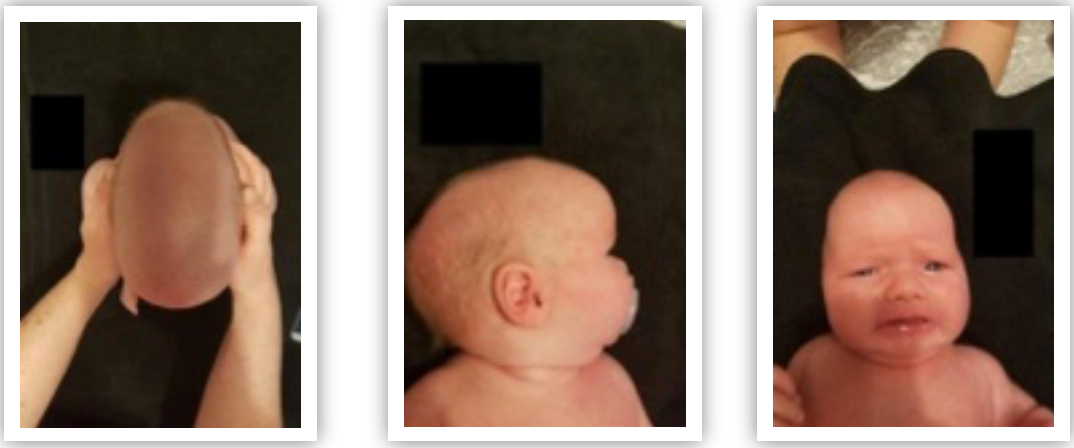
Most children with sagittal craniosynostosis may have no symptoms apart from the appearance of their skull. The incidence of learning or behavioural issues in children with non-syndromic craniosynostosis is only slightly higher than the average population. While others may also have a speech and language delay where they tend to start to speak later than other children, this usually improves with from a speech and language therapist .

Raised intracranial pressure (ICP) seems to develop in less than 5% of children and often when they are between 3 and 5 years of age. Raised intracranial pressure means the pressure inside the skull around the brain increases, which can cause pressure on the brain itself. If your child reports having headaches and are otherwise well with no other signs of an infection such as temperature, sore throat or ears or has a cough it is important to discuss this with a member of the craniofacial team as it may be a sign of raised intracranial pressure.

We are not sure why raised intracranial pressure happens, but it can occur in children who have had surgery to correct their head shape as well as in those who have not had surgery.

What does sagittal craniosynostosis look like?

The main sign of sagittal craniosynostosis is an abnormal head shape. Your child's head shape will be typically narrow from side to side and lengthened from front to back. Children may have a prominent (protruding) forehead and the back of the head (occipital region) may be quite pointed or 'bullet' shaped. The sagittal suture itself may be easily seen when your child is very young as a prominent bony ridge extending from the 'soft spot' to the back of the head.



A child with sagittal craniosynostosis

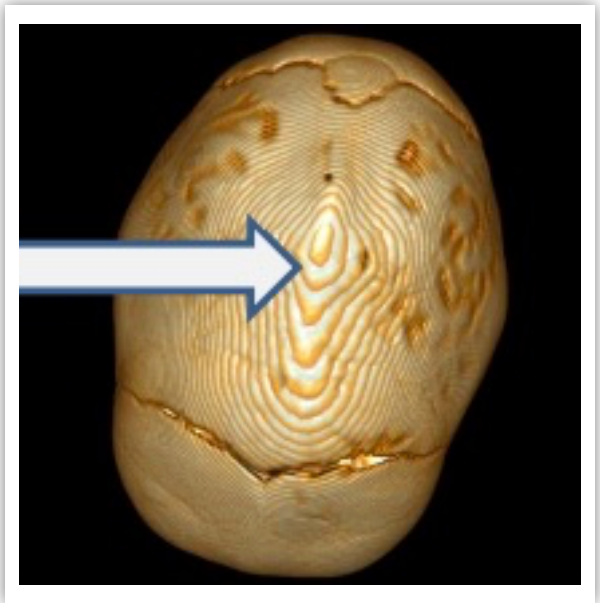
Does sagittal craniosynostosis affect the brain?

Sagittal craniosynostosis is rarely associated with brain growth and development problems. However, your child should be assessed by a team of specialists such as a Craniofacial Surgeon and Neurosurgeon to monitor for signs of raised intracranial pressure.

Does my child need an x-ray?

We do not recommend x-rays to diagnose sagittal craniosynostosis. The Craniofacial Surgeon and Neurosurgeon will make a diagnosis with careful assessment of your child's skull. However, a CT-Scan with 3D reconstruction (3DCT scan) may be used to:

- Verify the diagnosis of sagittal craniosynostosis
- Assess brain growth
- Evaluate if there is evidence of raised intracranial pressure
- Assist in surgical planning



3D CT scan of Sagittal Craniosynostosis
The arrow is pointing at the fused sagittal suture

What happens in the craniofacial clinic?

You and your child will meet the Craniofacial Surgeon, Neurosurgeon, Craniofacial Nurse Specialist and Craniofacial Coordinator. The team will ask you about the pregnancy and your child's medical /surgical history to date. It is always helpful to have all this information available for your appointment, so take note of any other doctors or specialist your child has seen or due to see. The Craniofacial team will confirm if your child has sagittal craniosynostosis and discuss the options available to you. Your child may be referred for a 3D CT scan and other investigations may also be arranged. Once these assessments have taken place you will once again meet with the team at the craniofacial clinic where they will discuss any further questions you may have and the options for surgery to correct your child's head shape. If your child has other medical conditions, your Craniofacial Consultant may decide to delay surgery until some of these problems are treated, this to ensure your child is fit and well for surgery. Your expectations and conservative management (no surgery) will also be discussed at this visit.

Once a decision for surgery has been made the Craniofacial Nurse Specialist will meet you in clinic to discuss the pre-operative (before surgery) planning including any further assessments or blood tests which must be completed. The Craniofacial Nurse Specialist will also discuss your child's admission to hospital before surgery including preparation for surgery, expected length of stay in hospital and your child's expected post-operative (after surgery) recovery course including managing their pain and care of their wound (scar) and the follow up appointments you will need to bring your child to after surgery.

The Craniofacial Coordinator will liaise with all services both inside and outside of Children's Health Ireland (CHI) at Temple Street to ensure that your child's entire pre-operative plan is completed before surgery. The Craniofacial Coordinator will also be able to help you connect with other families who have recently gone through similar circumstances and surgery after their child was diagnosed with sagittal craniosynostosis. The Craniofacial Coordinator will also give you the information about your child's date of surgery.

Will my child need surgery?

95% of children will not suffer any functional problems with a craniosynostosis, however, the abnormal shape of the skull can cause very significant psychological problems at a later stage and particularly in teenage years. The decision for surgery is based on severity of the craniosynostosis, the findings of the craniofacial team (with input from other teams where necessary) and parental involvement. All families are offered the option of surgical management and treatment for their child's head shape. For the more severe cases surgery to correct your child's head shape is almost always necessary. For mild to moderate cases surgical management and treatment is offered. For the cases that are considered mild often conservative (no treatment) management with follow up care as per our protocol is still indicated. https://www.craniofacial.ie/wp-content/uploads/2018/08/NPCC-Care-Pathway-Sagittal-Craniosynostosis.pdf

The team is always available should families wish to discuss surgery in the future if their child's head shape becomes a problem for their child particularly from a psychological and or social perspective. It is important that families understand the only treatment to change your child's head shape to a normal head shape is with surgery.

What will my child's surgery involve?

There is a close relationship between the growth of the skull and facial growth. Children with abnormal skull growth may have abnormal facial growth but this is rare. Since the moulding and shape of the skull is directly dependent on the growth of the brain the best appearance changing results are obtained with early surgery. For sagittal craniosynostosis there are 2 broad categories of surgery depending on how early your child's diagnosis is made. If surgery can be done before six months then there are a number of options. If however the child is not suitable for surgery for six months, then we usually Delay it until they are approximately 15 months.

Surgery for craniosynostosis starts with the Craniofacial Surgeon making a cut in the skin across the top of your child's head from ear to ear. This cut is shaped like a zigzag which makes it easier for your child's hair to grow over it after surgery. The Craniofacial Surgeon will decide which parts of the skull bone need to be removed and reshaped (reconstructed) to give your child a normal head shape. Once all the bones are in place the Craniofacial Surgeon will stitch your child's skin together.

Surgery with diagnosis before 6 months of age:

Surgical correction of sagittal craniosynostosis can be performed when your child is between 4 and 6 months of age with a procedure called an Extended Strip Cranial Vault Remodelling (ESCVR). This involves the removal of the fused sagittal suture and the placement of cuts along the bones of the skull to allow the brain to push out the bones of the skull into a more normal head shape. The surgery takes about 5 hours from start to finish, this includes putting your child to sleep and waking your child up. Your child will stay in hospital for approximately 4 to 5 days following surgery. There are other potential surgical options, which your Craniofacial surgeon with you.

Surgery with diagnosis after 6 months of age:

When your child receives their diagnosis after 6 months of age the surgical correction of sagittal craniosynostosis involves a procedure called a Total Cranial Vault Remodelling (TCVR). Generally, this surgery is performed when your child is between 15 and 18 months of age and is healthy and strong. It is during this time frame that the bones of the skull are thick enough and yet still malleable (pliable) to reconstruct. This is a more complex and longer surgery and involves the removal and reconstruction of the bones in the front, back and sides of the skull.

The skull bones are held in place with plates and screws (usually bio-resorbable ones which gradually dissolve between 12 and 18 months after surgery). Usually, there is not enough bone to reconstruct the entire skull and several “soft spots” or defects will remain. These will heal spontaneously over time, up to 24 months after surgery the brain is still growing rapidly, and any soft spots or defects left after the reconstruction will be filled in naturally by new bone being laid down. The surgery takes between 5 to 7 hours from start to finish; this includes putting your child to sleep and waking your child up. Your child will stay in hospital for approximately 5 to 7 days after surgery.

Following surgery, infants and children generally stay in St Gabriel’s ward. Before going home, the Craniofacial Nurse Specialist will discuss your child’s discharge and home care with you.

https://www.craniofacial.ie/wp-content/uploads/2018/08/3.Following-Craniofacial-Surgery.pdf