Craniofacial Ireland

RECONSTRUCTING FUTURES
PIONEERING CRANIOFACIAL SURGERY AND CARE

UniCoronal (Anterior Plagiocephaly)

Unicoronal Craniosynostosis

Unicoronal craniosynostosis is the second most common type of single suture (non-syndromic) craniosynostosis and occurs when one of the 2 coronal sutures fuses prematurely before birth. The coronal sutures (1 on the right and 1 on the left) separate the forehead bone from the bones at the sides of your child's head and meet in the middle of the forehead at the anterior (front) fontanelle (soft spot).

Unicoronal craniosynostosis can be associated with other conditions; the Craniofacial Surgeon will examine your child closely. The head shape which occurs because of unicoronal craniosynostosis is known as plagiocephaly which means oblique (slanted) skull in Greek.

Sometimes both coronal sutures are prematurely fused before birth. When more than one suture is affected, it is called multi suture craniosynostosis. This may happen as part of a syndrome (collection of symptoms) and is known as syndromic craniosynostosis.

How common is unicoronal craniosynostosis?

It is the second most common type of premature suture fusion affecting approximately 1 in every 10,000 births with a 2:1 female: male ratio which means that it is 2 times more common in girls.

What causes unicoronal craniosynostosis?

The cause of unicoronal craniosynostosis is not yet known. Unicoronal craniosynostosis seems to affect more females than males but we are not yet sure why this is the case.

There may be a genetic basis to the condition as it seems to be passed on from parent to child in a small number of families and in approximately 20-30% of patients, we may be able to identify the gene responsible. When some children are born with unicoronal craniosynostosis a known syndrome (collection of symptoms) can be identified. Unicoronal craniosynostosis can occur as a part of craniofacial syndromes such Muenke and Craniofrontonasal Dysplasia. More research is needed to identify all the causes of unicoronal craniosynostosis.

What are the symptoms of unicoronal craniosynostosis?

Some children with unicoronal craniosynostosis may also have a speech and language delay where they tend to start to speak later than other children, this usually improves with help from a speech and language therapist.

Raised intracranial pressure (ICP) seems to develop in approximately 14% of children 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.

What does unicoronal craniosynostosis look like?

The main sign of unicoronal craniosynostosis is the flatter appearance of the forehead and eye socket on the affected side and the prominence (protrusion) of the forehead on the opposite side. This protrusion on the unaffected side can also cause the eyelid to look as if it is hanging downwards (droopy). The eyes often look different, with one being larger or 'more open' than the other.

When looked at from above your child's head shape will look flat on the affected side as their forehead slants backwards and it will look bigger (protruding) on the opposite side of the forehead. The affected coronal suture itself may be easily seen as a prominent bony ridge extending from the 'soft spot' to the side of their forehead on the affected side.

Also, your child's eye will look more prominent on the affected side because the bones (orbital) surrounding the eye are recessed (sunken) and your child may look amused or surprised. As the eye on the affected side is not in a normal position in the eye socket, it may cause a squint (strabismus) to develop. A squint will cause the eyes to look in different directions such as turning inwards, outwards, upwards or downwards while the other eye looks forwards. Children with unicoronal craniosynostosis must be seen by an Ophthalmologist (Eye Specialist) who will examine your child closely for this. If a squint is not treated it can affect the development a vision. The root of the nose (where the nose is attached to the forehead) may also seem to be tilted (skewed) towards the unaffected side

A child with Right sided Unicoronal Craniosynostosis.

Does unicoronal craniosynostosis affect the brain?

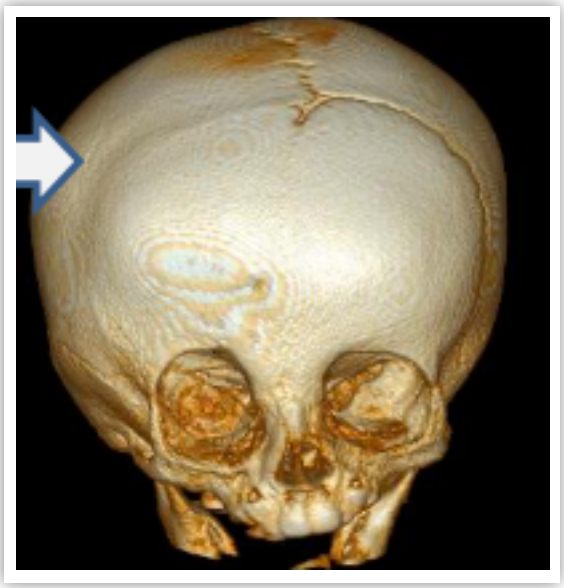
Unicoronal craniosynostosis is associated with brain growth and development problems in approximately 5% of children leading to cognitive (intellectual) and behavioural (developmental and communication) difficulties.

Your child should be assessed by a team of specialists such as a Craniofacial Surgeon, Neurosurgeon and Ophthalmologist (Eye Specialist) to monitor for signs of raised intracranial pressure as this can happen in 14% of children with unicoronal craniosynostosis, especially when they are between 3 and 5 years of age.

Does my child need an x-ray?

We do not recommend x-rays to diagnose unicoronal 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 (3D CT scan) may be used to:

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



The arrow is pointing at the fused Right coronal 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. The team will confirm if your child has unicoronal craniosynostosis and discuss the options available to you. Your child will be referred to an Ophthalmologist (Eye Specialist) and a 3D CT Scan 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 is 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 unicoronal craniosynostosis. The Craniofacial Coordinator will also give you the information about your child's date of surgery.

Will my child need surgery?

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 as per our protocol is indicated.

Early surgery is usually recommended for children with unicoronal craniosynostosis because of the effects on the bones and other features of the face and jaws. If left untreated, unicoronal craniosynostosis may result in a severe abnormality (defect) of the growth of the forehead, orbits and nose which may become more noticeable as your child grows. Whilst the severity of the condition may not worsen, it does become more obvious as the child loses there 'babyface' and ultimately grows into a teenager, where the asymmetry can often be very obvious. If it is very severe it may also affect your child's occlusion (the way the jaw and teeth fit together) and possibly how they chew their food, along with their speech and growth of their lower face.

https://www.craniofacial.ie/wp-content/uploads/2020/02/NPCC-Care-Pathway-Unicoronal-2020.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.

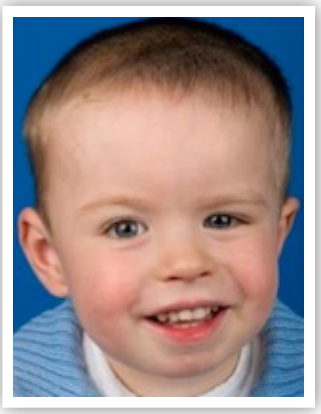
What will my child's surgery involve?

There is a close relationship between the growth of the skull and facial growth. Children with abnormal skull growth will have abnormal facial growth. 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. 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

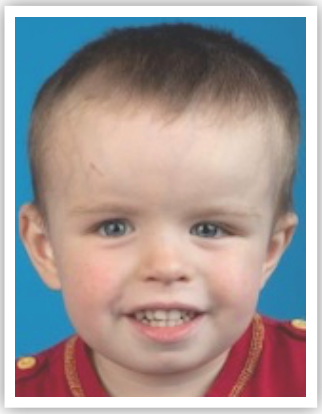
Surgical correction of unicoronal craniosynostosis involves a procedure called a Fronto-Orbital Advancement / Anterior Cranial Vault Remodelling (FOA/ACVR) generally; this surgery is performed when your child is between 15 and 18 months of age and is healthy and strong. It is during this time frame that the bones of the skull are thick enough and yet still malleable (pliable) to reconstruct. This surgery involves the removal of the frontal bones (forehead) and of the supra orbital rims (eyebrow bones) and the fused unicoronal suture. Once the bones are reconstructed the forehead and eyebrow bones are brought forward to allow the brain to grow.

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

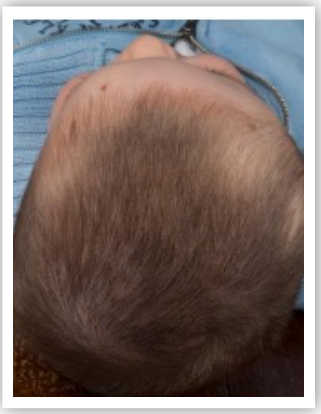
soft spots or defects left after the reconstruction will be filled in naturally by new bone being laid down. The surgery takes about 5 hours from start to finish, this includes putting your child to sleep and waking them up. Your child will stay in hospital for approximately 4 to 5 days following surgery.

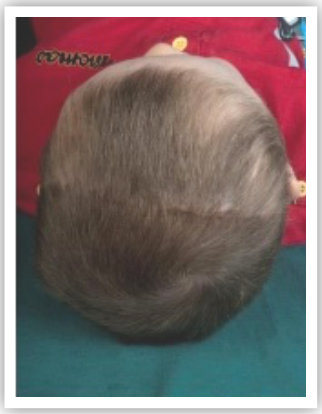


Before Surgery



After Surgery





A child with right sided Unicoronal Craniosynostosis

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