



Unicoronal craniosynostosis

This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of unicoronal craniosynostosis and where to get help.

What is unicoronal craniosynostosis?

Unicoronal craniosynostosis is a type of non-syndromic craniosynostosis and occurs when one of the two coronal sutures fuses before birth. The coronal sutures run from the front fontanelle down to the side of the forehead.

The skull is made up of several 'plates' of bone which, when we are born, are not tightly joined together. The seams where the plates join are called 'sutures'.

As we grow older, the sutures gradually fuse (stick) together, usually after all head growth has finished. When a child has craniosynostosis, the sutures fuse before birth. It can affect one suture or several.

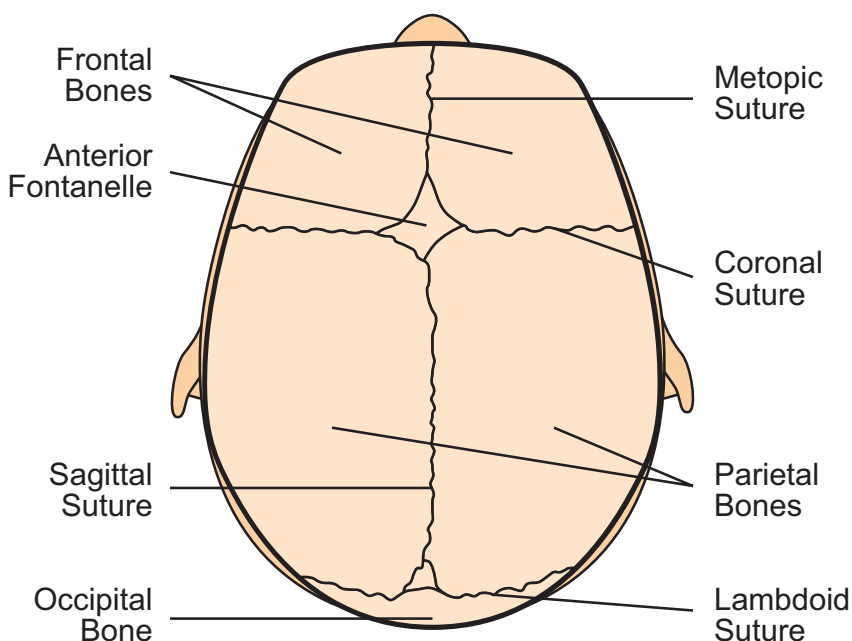
The premature fusing of the coronal suture makes the forehead and eye socket on one side flatter while the opposite side of the forehead moves forward to compensate.

What causes unicoronal craniosynostosis?

Unicoronal craniosynostosis may occur as part of Muenke syndrome but in most cases, the cause is unknown.

Unicoronal craniosynostosis can be associated with other clinical conditions, such as cranio-fronto-nasal dysplasia so doctors will examine your child closely to check if this is the case.

Normal Skull of the Newborn





What are the signs and symptoms of unicoronal craniosynostosis?

The main sign of unicoronal craniosynostosis is the flatter appearance of the forehead and eye socket on one side and the prominence (bossing) of the opposite side of the forehead. The root of the nose may also seem to be skewed towards the unaffected side. As the eye on the affected side is abnormally placed within the eye socket, it may cause a squint to develop. If untreated, this can affect visual development in childhood so a referral is usually made to an ophthalmologist (eye specialist).

How is unicoronal craniosynostosis diagnosed?

As children with unicoronal craniosynostosis have a characteristic appearance, no specific diagnostic tests are needed. A DNA/genetics test may be done to see if it is part of Muenke syndrome. Imaging scans, such as x-ray, CT or MRI may be suggested to monitor bone growth before, during and after treatment.

How is unicoronal craniosynostosis treated?

Although unicoronal craniosynostosis mainly affects the skull, treatment is best delivered at a specialist centre where a multidisciplinary team approach can be taken. The multidisciplinary team will usually comprise craniofacial (skull and face) surgeons, neuro (brain) surgeons, ophthalmologists (eye specialists), geneticists and speech and language therapists with other specialists brought in as needed.

Unicoronal craniosynostosis is not usually associated with raised pressure in the head so treatment is indicated primarily for cosmetic reasons. It consists of skull re-shaping surgery which takes place within the first year or two of life. There are different types of surgery depending on the age of your child at diagnosis – the team will discuss the options with you.

There are several methods of treating a squint – what is needed will depend on the severity of the squint. The aim of all methods of treatment is to align the eyes so that they look normal and work properly. Non-surgical methods like glasses or patches may be tried first. Only if these methods do not correct the squint is an operation considered.

As the bone continue to grow during childhood and adolescence, further surgery is occasionally needed to make, usually minor, corrections to the skull shape and forehead area.



What happens next?

The outlook for children with unicoronal craniosynostosis is good with the vast majority growing up to lead a normal life, working and raising a family.

Further information and support

Headlines – the Craniofacial Support Group – is the main support organisation in the UK for families of children and young people affected by a craniofacial disorder. Visit their website at www.headlines.org.uk.

Changing Faces is another organisation that will be able to offer help and support to anyone living with a condition that affects their appearance. Visit their website at www.changingfaces.org.uk or telephone their helpline on 0845 4500 275.