Headlines
Craniofacial Support

NON-SYNDROMIC CRANIOSYNOSTOSIS

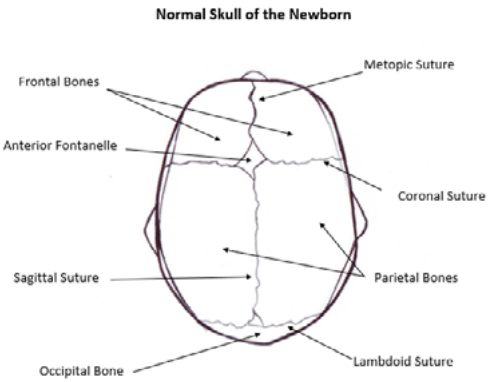
A guide for parents and carers

If your child has just received a diagnosis of craniosynostosis you may be feeling a bit overwhelmed. There can be a lot of information to take in, particularly at a time when you may only just be getting used to having a new baby. This leaflet, produced with input from a group of UK medical experts, is intended to help you understand your child's condition and the treatment that you may be offered.

What is craniosynostosis?

The skull is made up of several plates of bone which meet at gaps (sutures), called the sagittal, coronal, metopic and lambdoid. The sutures allow the bones of the skull to overlap slightly so that the baby's head can pass through the birth canal. The sutures also enable the skull to expand to accommodate the brain which grows rapidly during the first two years of life.

Normally, sutures join (fuse) during adulthood, when brain growth has finished. However, in a small number of babies one or more of the sutures fuses before birth or early in childhood resulting in a condition called 'craniosynostosis'.



When the suture is fused, bone growth ceases at the fused suture , and is re-directed to the unfused sutures, leading to the skull assuming an unusual shape. The type of shape will depend on which suture is affected.

We generally talk about two main types of craniosynostosis:

- **Non-syndromic** (sometimes referred to as ' 'single suture' craniosynostosis) - usually, but not always, involving one suture, and usually without any problems affecting other parts of the skull, face or body. There may or may not be an underlying genetic cause.
- **Syndromic** (sometimes referred to as 'complex' or 'multi suture' craniosynostosis) - where there are physical features or problems affecting other parts of the body, which follow recognisable patterns. There is often an underlying genetic cause.

It is estimated that craniosynostosis affects between 1 in 1,800 and 1 in 2,000 babies in the UK. Around 75% of these will be non syndromic, and the other 25% syndromic.

Types of non-syndromic craniosynostosis

Most non-syndromic craniosynostosis can be subdivided into four different types, according to which suture is affected. The underlying suture involved will also generally determine the type of treatment and surgery required.

Sagittal synostosis

This is the most common form of non-syndromic craniosynostosis, affecting around 1 in 5,000 births. It occurs when the sagittal suture – the growth line running along the top of the skull from front to back - fuses too early. The result is a characteristic head shape where the length of the skull is increased and the width reduced (said to be like the shape of a boat). It is traditionally referred to as 'scaphocephaly'.

Metopic synostosis

Metopic craniosynostosis is also known as 'trigonocephaly', from the Greek for 'triangleshaped head'. It occurs when the metopic suture - which runs from the front fontanelle (soft spot) through the forehead to the top of the nose - fuses too early. The head viewed from above is sometimes pointed in a triangular shape to the front and top of the skull. In some cases, the eyes may appear slightly misaligned.

Coronal synostosis

Coronal – and especially bicoronal – craniosynostosis is much more frequently associated with syndromic than non-syndromic craniosynostosis.

Unicoronal synostosis is where one coronal suture is involved and there isn't enough growth on the side of the fused suture, making the forehead and eye socket on one side flatter, whilst the opposite side of the forehead grows forward to compensate. In some cases, the eyes may be slightly misaligned. It's also known as known as 'plagiocephaly' or 'oblique skull'.

Bicoronal synostosis is when both coronal sutures are involved. , The skull becomes excessively wide and short from front to back. There may be excessive growth at the upper part of the forehead and both eyebrows are pulled up and are flattened. This short front to back appearance of the skull has given rise to the name 'brachycephaly' or 'short skull'.

Lambdoid synostosis

This is a very rare type of craniosynostosis , affecting less than around 1 in 100,000 births. It occurs when one of the lambdoid sutures at the back of the head fuses prematurely, leading to an asymmetric skull shape flattened at the back.

What effects are there?

Because skull growth is so rapid in the early years of life, the fused suture(s) can cause the skull and face to distort as they grow. In a small percentage of cases, the restrictions can also cause the pressure within the skull to rise – this condition is called raised intracranial pressure (ICP), and, if left untreated, may lead to problems with a baby's long term development. However, many of these problems can be successfully corrected by surgery.

Most children with non- syndromic craniosynostosis don't have any lasting health problems. They may have a scar across the top of their head if they have had surgery, but this will generally be hidden by their hair.

Treatment and surgery

Unfortunately, the distortion of the skull that occurs in craniosynostosis will not get better on its own, a child cannot 'outgrow' craniosynostosis, and treatment usually involves surgery. As well as improving appearance, one of the reasons for operating is to reduce the risk of raised pressure in the brain (raised intracranial pressure) from developing when the child is older.

If babies do not have a particularly different head shape, parents may choose to take a 'watch and wait' approach where the medical team monitor how the head shape is developing as the child grows older. If the child does not have surgery, the medical team will also need to check that they are not showing any signs of raised intracranial pressure (which may develop gradually over time).

Most children with craniosynostosis are referred to one of 4 highly specialised National Craniofacial Units 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), clinical psychologists, geneticists and speech and language therapists, with other specialists brought in as needed.

Specialist nurses at these centres can also assist if a GP or parent is unsure about the diagnosis.

Details of how to contact the Centres are given on our website:

www.headlines.org.uk/specialist_hospitals.asp.

Headlines produces a number of other leaflets covering different aspects of craniosynostosis and treatment. Please email helpline@headlines.org.uk for further information.

Headlines is the only national charity supporting those affected by craniosynostosis and rare craniofacial conditions. As well as providing information for parents, carers and families, we also offer a confidential helpline, a member magazine and regular newsletters, conference & information days, opportunities for members to meet, and an Annual Family Weekend.

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