Headlines
Craniofacial Support

SAGITTAL 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.

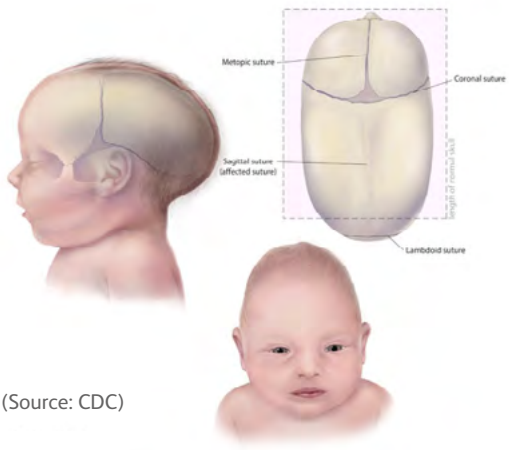
What is sagittal craniosynostosis?

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.

Sagittal craniosynostosis is the most common type of non-syndromic craniosynostosis, affecting around 1 in 5,000 births. It occurs when the sagittal suture fuses before birth, leading to the skull being long from front to back but narrow from side to side. It's also known as 'scaphocephaly' – from the Greek for 'boat-shaped'.



(Source: CDC)

The resulting head shape can vary from being only mildly different from normal to evidently abnormal. It's generally more common among boys than girls, but only rarely associated with problems affecting other parts of the skull, face or body.

What causes sagittal craniosynostosis?

At present, we do not fully understand what causes the sagittal suture to close too early. 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. However, more research is needed to identify the specific causes.

What are the signs of sagittal craniosynostosis?

The main sign is a bony ridge over the fused sagittal suture, and a change in the head shape. Depending on whether the entire sagittal suture has fused or only part of it, children have a prominent forehead and a low pointed back of occipital region (back of head).

What are the effects of sagittal craniosynostosis?

Because skull growth is so rapid in the early years of life, the fused suture can cause the skull and forehead to distort as they grow.

Problems that can be associated with sagittal craniosynostosis are speech and language delay and raised intracranial pressure (ICP).

Some children with sagittal craniosynostosis may have early speech and language difficulties, but with support from parents and local Speech and Language therapy (SLT), children can be supported to develop their speech and language skills.

Raised intracranial pressure seems to develop in some children . We are not sure why this happens, but it seems to occur whether or not a child has had skull re-shaping surgery.

Most children with sagittal 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 be mostly hidden by their hair. The scar can sometimes be quite tender in the initial period following surgery and should be protected against the sun if the hair is very short.

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 prevent raised pressure in the brain (raised intracranial pressure) from developing when the child is older.

However, 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 sagittal craniosynostosis are referred to one of 4 highly specialised 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 Units can also assist if a GP or parent is unsure about the diagnosis.

Details of how to contact the Units are given on our website: www.headlines.org.uk/specialist_hospitals.asp.

In many cases, initial skull re-shaping surgery takes place within the first 18 months of life. However, it is still possible to perform some types of surgery on older children.

In the UK, the vast majority of cases are performed as a single procedure.

What are the different types of skull reshaping surgery?

There are various different approaches to surgery, all aimed at improving the head shape and reducing the risks of intracranial pressure.

Generally, individual surgeons use the techniques that they have most experience with and feel most comfortable performing. Timing of surgery also varies between the Units, being influenced by when your baby gets referred, and the surgical technique used.

As with any operation there are risks. Your child's surgeon will explain the surgery to you, discuss the potential complications and answer any questions. The specialist craniofacial nurses are key contacts who are there to support your family throughout the craniofacial journey.

Remember that there are no 'rights' or 'wrongs' - your team should discuss options with you, so that you can make a fully informed choice.

What happens after surgery?

After surgery your child will have regular follow up appointments. The timing of these will vary according to where your child is treated. Long term post -operative surveillance usually stops around 12 years when the child's head growth is nearly complete . Children who have not had surgery are generally followed up at the same time intervals.

At these appointments the surgeon will review the head shape and also ask questions to check if your child is showing any signs of raised intracranial pressure. Symptoms of raised pressure include waking on a regular basis with a headache, behavioural changes, increased aggression and a decline in skills. Although this sounds alarming, the changes happen very slowly and any problems can be identified and treated to prevent long-term harm.

At some of these appointments your child will have an assessment with a speech and language therapist and/or a clinical psychologist at key points in their development. The reason for these assessments is that each child with sagittal craniosynostosis is different and will develop on their own trajectory. The professionals who conduct these assessments will monitor your child's individual progress as they develop. In most children with sagittal craniosynostosis, the speech and language therapists and clinical psychologists do not expect to identify any developmental delays, but if any difficulties are identified, children can be referred to local services to support their development. In the majority of cases, with early intervention, most speech and language delay can be resolved before the child starts school.

What is the outlook for children affected by sagittal craniosynostosis?

Generally children with sagittal craniosynostosis grow up to lead a normal life, working and raising a family.

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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