



Metopic craniosynostosis

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

What is metopic craniosynostosis?

Metopic craniosynostosis is a type of non-syndromic craniosynostosis that occurs when the metopic suture fuses before birth.

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.

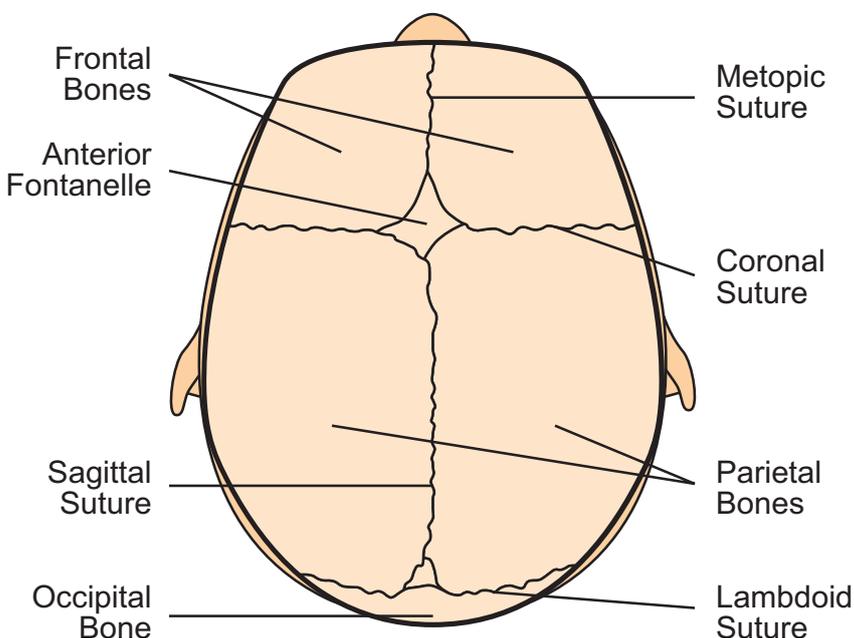
The metopic suture runs from the front fontanelle (soft spot) through the forehead to the top of the nose. Metopic craniosynostosis is also known as trigonocephaly – from the Greek for triangle-shaped.

What causes metopic craniosynostosis?

The cause of metopic craniosynostosis is not yet known. 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. There also seems to be a link between the mother taking an anti-epilepsy medicine called sodium valproate during pregnancy and her baby being born with metopic craniosynostosis. Another theory for the cause of metopic craniosynostosis is that the position of the baby while in the womb may affect skull development.

Metopic craniosynostosis seems to affect more males than females but we are not yet sure why this should be the case. The number of babies born with metopic craniosynostosis also seems to be increasing but again, more research is needed to discover the reason for this increase. It can be associated with other conditions, so the 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 metopic craniosynostosis?

The main symptom of metopic craniosynostosis is the abnormal shape of the forehead which is pointed and triangular. There may also be a bone ridge over the prematurely-fused suture running down the forehead from the front fontanelle to the top of the nose. This early fusing of the metopic suture often makes the eyes closer set than usual

Children with metopic craniosynostosis can have developmental delay – that is, they reach their 'milestones' later than other children of a similar age. Over time, many children catch up so do not need much additional support in school.

There is no evidence currently that this developmental delay is a direct result of the craniosynostosis – it appears to be an association only. Corrective surgery does not change the progress of development.

How is metopic craniosynostosis diagnosed?

As children with metopic craniosynostosis have a characteristic appearance, no specific diagnostic tests are needed. Imaging scans, such as x-ray, CT or MRI may be suggested to monitor bone growth before, during and after treatment. As the gene mutation causing metopic craniosynostosis has not yet been identified, genetic testing will not be helpful in most cases.

How is metopic craniosynostosis treated?

Although metopic 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.

In many cases, initial skull re-shaping surgery takes place within the first few years 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. As the bone continues to grow during childhood and adolescence, further surgery may be needed to make minor corrections to the skull shape and forehead area.

What happens next?

The outlook for children with metopic craniosynostosis is good with the vast majority growing up to lead a normal life, working and raising a family, although it will vary depending on any other medical conditions present. Children can sometimes develop behavioural problems at school age, but with input and support from specialists, these are overcome in most cases.

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.