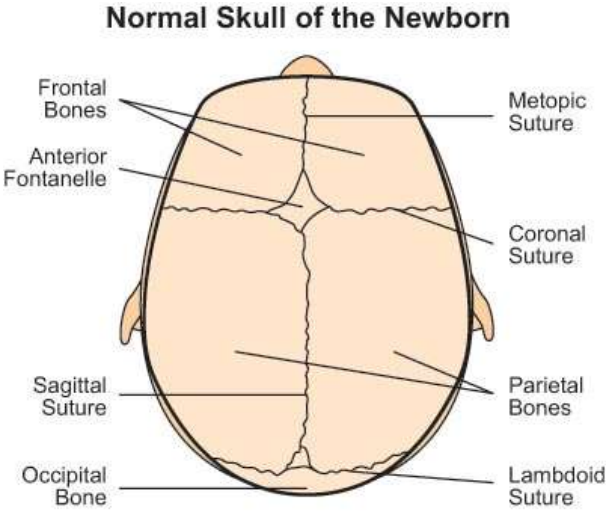
Bicoronal craniosynostosis: information for families

Bicoronal craniosynostosis is a type of craniosynostosis which may be part of a syndrome (collection of symptoms often seen together) or non-syndromic. This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of bicoronal craniosynostosis.

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 sutures makes the forehead and eye sockets flatter as the rest of the skull compensates.

What causes bicoronal craniosynostosis?

The cause of bicoronal 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.

Several genes have been identified as being associated with bicoronal craniosynostosis. Gene mutations can be passed on from parent to child but in many cases develop sporadically (out of the blue). More research is needed to identify the cause of bicoronal craniosynostosis.

What are the symptoms of bicoronal craniosynostosis?

The main symptoms of bicoronal craniosynostosis are the flatter appearance of the forehead and eye sockets and a head shape that is shorter front to back and taller than average.

How is bicoronal craniosynostosis diagnosed?

As children with bicoronal 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. Some gene

mutations causing bicoronal craniosynostosis have been identified. Genetic testing will most likely be undertaken to check whether there is a genetic reason for the bicoronal craniosynostosis.

How is bicoronal craniosynostosis treated?

Although bicoronal 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.

There is a chance that children with bicoronal craniosynostosis may develop raised pressure in the head. This is often without symptoms initially, but if left untreated, may result in visual or neurological problems. For this reason, children with bicoronal craniosynostosis are monitored with regular eye examinations, usually every few months.

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

What is the outlook for children and young people with bicoronal craniosynostosis?

The outlook for children with bicoronal 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.

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 offers 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.