Lambdoid craniosynostosis

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

Normal Skull of the Newborn

What is lambdoid craniosynostosis?

Lambdoid craniosynostosis is a very rare type of non-syndromic craniosynostosis and occurs when one of the lambdoid sutures at the back of the head fuses before birth. It may be associated with other forms of syndromic craniosynostosis where more than one suture is fused.

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 lambdoid suture leads to an asymmetric skull shape flattened at the back.

What causes lambdoid craniosynostosis?

The cause of lambdoid 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. Another theory for the cause of lambdoid craniosynostosis is that the position of the baby while in the womb may affect skull development. More research is needed to identify the cause of lambdoid craniosynostosis.

What are the signs and symptoms of lambdoid craniosynostosis?

The main symptom of lambdoid craniosynostosis is that the back of the head is flattened, usually on one side where the lambdoid suture has fused too early. The ear on that side is also affected, being positioned lower down and further back than the other.

In some children, the abnormal skull shape causes raised intracranial pressure so regular monitoring will be suggested – this is done using regular eye examinations.
How is lambdoid craniosynostosis diagnosed?

Lambdoid craniosynostosis may be confused with positional or deformational plagiocephaly – a disorder that affects the skull, making the back or side of a baby’s head appear flattened. Unlike craniosynostosis, the skull plates are not fused, but moulded into a different shape – a condition that does not require surgical treatment.

Imaging scans, such as x-ray, CT or MRI may be suggested to identify whether the sutures have fused or not to confirm the diagnosis. Imaging scans will also be suggested to monitor bone growth before, during and after treatment. As the gene mutation causing lambdoid craniosynostosis has not yet been identified, genetic testing may not be helpful in most cases but will help us to investigate all forms of craniosynostosis.

How is lambdoid craniosynostosis treated?

Although lambdoid 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 continue 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 lambdoid 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.

Compiled by the Craniofacial team in collaboration with the Child and Family Information Group
Great Ormond Street Hospital for Children NHS Foundation Trust, Great Ormond Street, London WC1N 3JH
www.gosh.nhs.uk