Craniofacial Ireland
RECONSTRUCTING FUTURES
PIONEERING CRANIOFACIAL SURGERY AND CARE

Lambdoid (Posterior Plagiocephaly)

Lambdoid craniosynostosis

Lambdoid craniosynostosis is a very rare type of single suture (non-syndromic) craniosynostosis and occurs when one of the 2 lambdoid sutures fuses prematurely before birth. The lambdoid sutures (1 on the right and 1 on the left) separate the bones at each side of the head from the bone at the back of the head.

Lambdoid craniosynostosis can be associated with other conditions; the Craniofacial Surgeon will examine your child closely. The head shape which occurs because of lambdoid craniosynostosis is known as plagiocephaly which means oblique (slanted) skull in Greek.

Sometimes both lambdoid sutures are prematurely fused before birth. When more than one suture is affected, it is called multi suture craniosynostosis. This may happen as part of a syndrome (collection of symptoms), and is known as syndromic craniosynostosis.

How common is lambdoid craniosynostosis?

Isolated lambdoid craniosynostosis is the least common type of premature suture fusion affecting approximately 1 in 150,000 births. It is more commonly associated with the complex, syndromic craniosynostosis, where there are other sutures are also affected.

Lambdoid craniosynostosis on its own is only seen in 1% of all children with single suture craniosynostosis which means that out of 100 children with craniosynostosis only 1 of them will have lambdoid craniosynostosis. Because the numbers are so small, we do not know if it affects boys or girls the most.

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, but the gene causing this has not yet been identified. More research is needed to identify the cause of lambdoid craniosynostosis.

What are the symptoms of lambdoid craniosynostosis?

Most children with lambdoid craniosynostosis may have no symptoms apart from the appearance of their skull. While others may also have a speech and language delay where they tend to start to speak later than other children, this usually improves with help from a speech and language therapist.

Raised intracranial pressure (ICP) seems to develop in very few of children, with isolated lambdoid craniosynostosis, and usually when they are between 3 and 5 years of age. Raised intracranial pressure means the pressure inside the skull around the brain increases, which can cause pressure on the brain itself. If your child reports having headaches and are otherwise well with no other signs of an infection such as temperature, sore throat or ears or has a cough it is important to discuss this with a member of the Craniofacial team as it may be a sign of raised intracranial pressure. We are not sure why raised intracranial pressure happens, but it can occur in both children who have had surgery to correct their head shape as well as in those who have not had surgery.

What does lambdoid craniosynostosis look like?

Your child's head shape will look flat at the back of the head on the affected side and there may be a bulging on the top opposite the flat Side. The ear on the same side will look as if it is pulled towards the back of the head and it may also look slightly lower than your child's other ear. You will see this most easily when you look at the top of your child's head. You may also see a prominent bony ridge extending across the bones between the side and back of the head on the affected side.

A child with lambdoid craniosynostosis

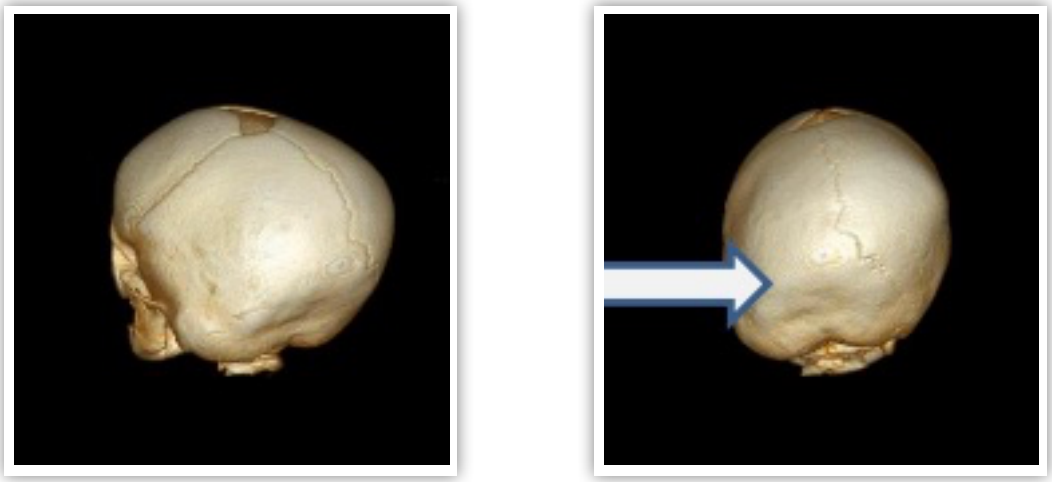
Does lambdoid craniosynostosis affect the brain?

Lambdoid craniosynostosis is rarely associated with brain growth and development problems. However, your child should be assessed by a team of specialists such as a Craniofacial Surgeon and a Neurosurgeon to monitor for signs of raised intracranial pressure, especially when they are between 3 and 5 years of age.

Does my child need an x-ray?

We do not recommend x-rays to diagnose lambdoid craniosynostosis. The Craniofacial Surgeon and Neurosurgeon will make a diagnosis with careful assessment of your child's skull. However, a CT-Scan with 3D reconstruction (3DCT scan) may be used to:

- Verify the diagnosis of lambdoid craniosynostosis
- Assess brain growth
- Evaluate if there is evidence of raised intracranial pressure
- Assist in surgical planning



3D CT scan of Right sided Lambdoid Craniosynostosis
The arrow is pointing at the fused lambdoid suture

What happens in the craniofacial clinic?

You and your child will meet the Craniofacial Surgeon, Neurosurgeon, Craniofacial Nurse Specialist and Craniofacial Coordinator. The team will ask you about the pregnancy and your child's medical /surgical history to date. The team will confirm if your child has lambdoid craniosynostosis and discuss the options available to you. Your child may be referred for a 3D CT Scan and other investigations may also be arranged. Once these assessments have taken place you will once again meet with the team at the craniofacial clinic where they will discuss any further questions you may have and the options for surgery to correct your child's head shape. If your child has other medical conditions, your Craniofacial consultant may decide to delay surgery until some of these problems are treated, this is to ensure your child is fit and well for surgery. Your expectations and conservative management (no surgery) will also be discussed at this visit.

Once a decision for surgery has been made the Craniofacial Nurse Specialist will meet you in clinic to discuss the pre-operative (before surgery) planning including any further assessments or blood tests which must be completed. The Craniofacial Nurse Specialist will also discuss your child's admission to hospital before surgery including preparation for surgery, expected length of stay in hospital and your child's expected post-operative (after surgery) recovery course including managing their pain and care of their wound (scar) and the follow up appointments you will need to bring your child to after surgery.

The Craniofacial Coordinator will liaise with all services both inside and outside of Children's Health Ireland (CHI) at Temple Street to ensure that your child's entire pre-operative plan is completed before surgery. The Craniofacial Coordinator will also be able to help you connect with other families who have recently gone through similar circumstances and surgery after their child was diagnosed with lambdoid craniosynostosis. The Craniofacial Coordinator will also give you the information about your child's date of surgery.

Will my child need surgery?

The decision for surgery is based on severity of the craniosynostosis, the findings of the Craniofacial team with input from other teams where necessary and parental involvement. All families are offered the option of surgical management and treatment for their child's head shape. For the more severe cases surgery to correct your child's head shape is almost always necessary. For mild to moderate cases surgical management and treatment is offered. For the cases that are considered mild often conservative (no treatment) management with follow up as per our protocol is indicated.

https://www.craniofacial.ie/wp-content/uploads/2018/08/NPCC-Care-Pathway-Lambdoid.pdf

The team is always available should families wish to discuss surgery in the future if their child's head shape becomes a problem for their child particularly from a psychological and or social perspective. It is important that families understand the only treatment to change your child's head shape to a normal head shape is with surgery.

What will my child's surgery involve?

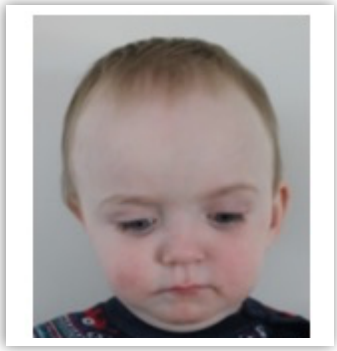
There is a close relationship between the growth of the skull and facial growth. Children with abnormal skull growth will have abnormal facial growth. Since the moulding and shape of the skull is directly dependent on the growth of the brain the best appearance changing results are obtained with early surgery. Surgery for craniosynostosis starts with the Craniofacial Surgeon making a cut in the skin across the top of your child's head from ear to ear. This cut is shaped like a zigzag which makes it easier for your child's hair to grow over it after surgery. The Craniofacial Surgeon will decide which parts of the skull bone need to be removed and reshaped (reconstructed) to give your child a normal head shape. Once all the bones are in place the Craniofacial Surgeon will stitch your child's skin together.

Surgical correction of lambdoid craniosynostosis involves a procedure called a Posterior Cranial Vault Remodelling (PCVR) generally, this surgery is performed when your child is between 15 and 18 months of age and is healthy and strong. It is during this time frame that the bones of the skull are thick enough and yet still malleable (pliable) to reconstruct. This surgery involves the removal of the bones at the back and sides of the head and the fused lambdoid suture. Once the bones are reconstructed the bones at the back of the head are brought forward to allow the brain to grow.

The skull bones are held in place with plates and screws (usually bio-resorbable ones which gradually dissolve between 12 and 18 months after surgery). Usually, there is not enough bone to reconstruct the entire skull and several "soft spots" or defects will remain. These will heal spontaneously over time, up to 24 months after surgery the brain is still growing rapidly, and any soft spots or defects left after the reconstruction will be filled in naturally by new bone being laid down. The surgery takes about 5 hours from start to finish. this includes putting your child to

sleep and waking your child up. Your child will stay in hospital for approximately 4 to 5 days following surgery.

Before Surgery



After Surgery

A child Pre and Post Lambdoid Craniosynsotosis correction

Following surgery, infants and children generally stay in St Gabriel's ward. Before going home, the Craniofacial Nurse Specialist will discuss your child's discharge and home care with you. Please click on link below for more details.

https://www.craniofacial.ie/wp-content/uploads/2018/08/3.Following-Craniofacial-Surgery.pdf