

Patient information sheet – Multisuture craniosynostosis

What is Multisuture craniosynostosis?

It is a condition where more than one of the soft joints of the skull fuses earlier than usual resulting in an abnormal shaped head (Brachycephaly, Turribrachycephaly, Microcephaly). It is also associated with an abnormal gene usually, causing one of the many craniofacial syndromes.

How does it affect my child?

It results in an abnormal shaped head like a flat head (Brachycephaly), tall head (Turricephaly), flat and tall head (Turribrachycephaly) and small head (Microcephaly). It can also be associated with many other anomalies like hand anomalies, ear anomalies, brain anomalies and internal organ anomalies. Very often, it can compress the brain to increase the intracranial pressure leading to blindness and damage to brain. Many children have midface hypoplasia which results in airway issues and obstructive sleep apnoea. Along with the flat face deformity, they can have exorbitism, where the eyeballs are placed ahead in a very exterior position because of a shallow eye socket. This can lead to recurrent dislocations of the eyeball and damage to the eye. A significant proportion of children also might have some degree of mental retardation.

How is it diagnosed?

Your Craniofacial surgeon will perform a thorough clinical examination. After that, a Skull Xray, a 3D CT scan and MRI scan of head is usually required. Intracranial pressure monitoring also may be required occasionally.

Which are the common syndromes associated with Multisuture craniosynostosis?

Some of the common syndromes are Crouzon syndrome, Apert syndrome, Pfeiffer syndrome, Muenke syndrome and Saethre-Chotzen syndrome. Genetic testing is usually done to detect the abnormal genes.

How is it treated?

The treatment of this condition requires surgery by a multidisciplinary approach mainly involving the Craniofacial surgeon, Neurosurgeon, Orthodontist, Respiratory physician, ENT surgeon, Ophthalmologist, Speech Pathologist, Paediatrician and Clinical Psychologist.

Many children require surgery to improve the raised intracranial pressure and improve the shape of the head. Your child may require more than one surgery in different phases of their childhood.

In the first year of life, surgery like Posterior cranial vault expansion is done to release the pressure inside the brain and improve intracranial anomalies like Chiari malformation.

Surgery to bring the forehead ahead and reshape the skull is delayed as much as possible to allow for growth, but is usually done in the later part of first year or the second year of life. It could be a combination of Fronto orbital advancement and Cranial vault remodeling.

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After the age of 3, if your child has issues with breathing and sleep apnoea, a surgery to advance the midface might be required to improve breathing (Lefort III advancement).

Multiple other procedures might be required depending upon the condition of your child and Dr Vybhav Deraje will organize regular multi-disciplinary meetings to decide the right course of action for your child.

What can I expect with the Posterior Cranial vault Expansion?

After a pre operative assessment by a paediatric neuroanaesthetist and ophthalmologist, your child will be admitted on the day of surgery. The surgery is done under general anaesthesia and usually takes 3-4 hours. Your child might have a lot of tubes connected for monitoring like central line and arterial line. Almost all the children need a blood transfusion during and after the surgery. There will be a cut made from one ear to the other to gain access to the skull. The neurosurgeon protects the brain, while the craniofacial surgeon does expansion of the skull bones on the back of the head to create more space in the skull for the brain. Bones are usually fixed with Stainless steel wires or stitches. Rarely, absorbable plates and screws also may be used. The wound will be closed with absorbable stitches. There will be a head bandage. Your child will spend the first night in the ICU and subsequently be shifted to the ward. You will notice significant swelling of the head and face and the eyes may close over for 3-4 days. The swelling gradually reduces over the week. Your child will be discharged usually on the 6th or the 7th day after removal of head bandage. You will be able to give head shower to the baby after that.

What happens after discharge?

You will have follow up visits with the craniofacial surgeon, neurosurgeon and ophthalmologist periodically. You will notice some gaps in the skull which will close over in 2-3 years time. The scar also is usually well hidden in the hair. Your child will need follow up CT scans and MRI scans to keep a close eye on the progress.

What can I expect with the open Fronto orbital advancement surgery or Cranial vault remodeling surgery?

After a pre operative assessment by a paediatric neuroanaesthetist and ophthalmologist, your child will be admitted on the day of surgery. The surgery is done under general anaesthesia and usually takes 4-5 hours. Your child might have a lot of tubes connected for monitoring like central line and arterial line. Almost all the children need a blood transfusion during and after the surgery. There will be a cut made from one ear to the other to gain access to the skull. The neurosurgeon protects the brain, while the craniofacial surgeon brings the forehead to a more forward location and reshapes the forehead and skull bones. Bones are usually fixed with Stainless steel wires or stitches. Rarely, absorbable plates and screws also may be used. The wound will be closed with absorbable stitches. There will be a head bandage. Your child will spend the first night in the ICU and subsequently be shifted to the ward. You will notice significant swelling of the head and face and the eyes may close over for 3-4 days. The swelling gradually reduces over the week. Your child will be discharged usually on the 6th or the 7th day after removal of head bandage. You will be able to give head shower to the baby after that.

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What complications can I expect?

This is a major surgery and you need to be well informed about all complications. Infection, bleeding, scar, swelling and pain are common complications that can happen. All these can be managed by our well trained team easily. Minor tears in the covering of the brain is sometimes seen which is usually repaired during the surgery. Very rarely there might be injury to the eye or the brain. Death is also a remote theoretical possibility, but we have never seen one in our practice. Rise in the intra cranial pressure can happen anytime and your child might need repeated surgeries if that happens.

What can I expect from the midface advancement surgery?

It is called as Lefort III advancement, where the middle part of the child's face including the upper jaw and cheek bones are brought to a forward location to give support to the eyeballs and improve the airway and sleep apnoea issues. Your child will have an appointment with the orthodontist to correct the alignment of the teeth. A CT scan also will be done which will be useful for virtual surgical planning. After assessment by a paediatric anaesthetist, your child will be admitted on the day of the surgery. The surgery usually takes 6-7 hours. Your child will definitely need blood transfusion during and after surgery. Cuts will be made from one year to the other and inside the mouth as well. The upper jaw along with the cheek bones are cut and brought forward. The bones are fixed in the new position using Titanium plates and screws. Absorbable stitches are used to close all the wounds. There will be a head bandage at the end of the procedure. Your child will spend the first night in the ICU and subsequently be shifted to the ward. You will notice significant swelling of the head and face and the eyes may close over for 3-4 days. The swelling gradually reduces over the week. Your child will be discharged usually on the 6th or the 7th day after removal of head bandage. You will be able to give head shower to the baby after that. Your child will be on soft diet for the first few weeks after surgery. Sleep study is usually repeated after a few months to assess the improvement after surgery.

I have a few more questions. What do I do?

Kindly write to us at contact@drvvybhavderaje.com. We will be happy to reply to any of your questions and concerns.

Disclaimer: This information sheet is for you to get a general idea about the condition and surgery. This is in no way a substitute for a formal consultation with your doctor.

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