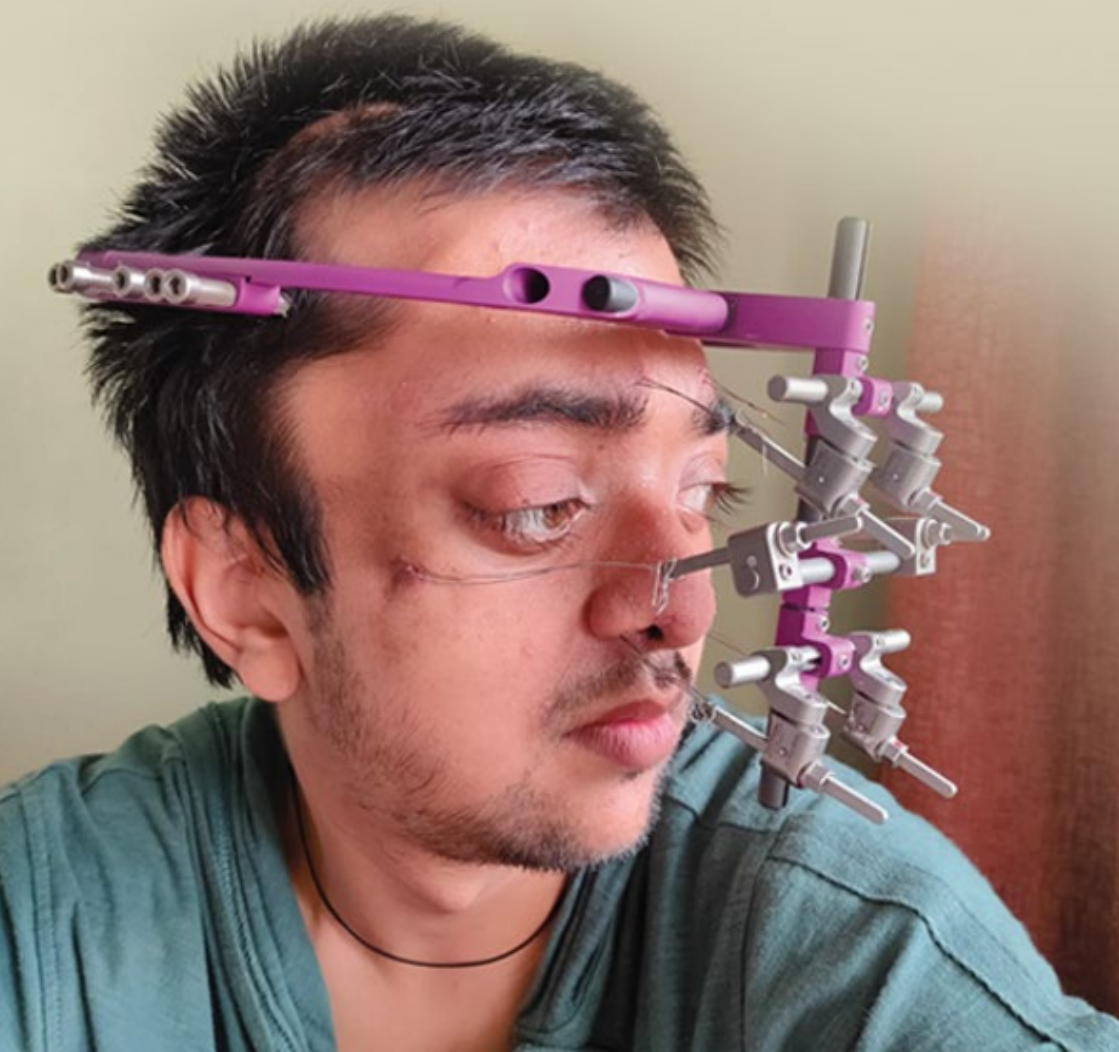
SRCC CHILDREN'S HOSPITAL

Managed by Narayana Health

Haji Ali Park
Mumbai

Bizarre Brain Structure

could not stop Yusuf from pursuing his love for History & Literature



Crouzon Syndrome is a rare genetically inherited disorder with an incidence rate of 1 in 60,000 newborns worldwide.*

Cranial sutures or anatomical lines are the fibrous tissue bands that join the bony plates of the skull together. In Crouzon Syndrome, the sutures, in an infant, merge prematurely affecting the normal growth of the skull and head leading to an alteration in the shape and therefore the development of the skull. The seriousness of craniosynostosis does vary from a child to another. The symptoms primarily include abnormalities of the face and head. Intelligence is usually not affected.

The syndrome is characterized by Craniosynostosis, and many other features.

This can cause an abnormal shape of the head and face with babies having

- SHORT AND WIDE OR LONG AND NARROW HEAD
- ENLARGED FOREHEAD
- SHALLOW ORBITS (EYE SOCKETS) LEADING TO BULGING EYEBALLS
- FLATTENED CHEEKS
- POORLY DEVELOPED UPPERJAW
- PROTRUDING LOWER JAW
- CURVED, BEAK-LIKE NOSE

The other features associated with this syndrome are

- INFLAMMATION OF CORNEA (EXPOSURE KERATITIS) DUE TO BULGING EYEBALLS (PROPTOSIS)
- SQUINT
- VISION LOSS
- HEARING LOSS
- DENTITION PROBLEMS
- CLEFT IN THE LIP/PALATE
- NARROW AIRWAYS LEADING TO RESPIRATORY PROBLEMS
- SLEEP APNOEA ETC.

There can be increased pressure in the brain and accumulation of excessive fluid in the brain (hydrocephalus).

Most of these children can have a normal life expectancy, if diagnosed early and treated timely by an experienced team. The social stigma around the abnormal facial appearance and lack of awareness about the condition leading to many children not receiving appropriate and timely diagnosis and treatment.

The treatment of this syndrome requires a multidisciplinary team approach consisting of specialists like Neurosurgeons, Plastic Surgeons, ENT Surgeons, Ophthalmologists, Dental Surgeons, Paediatric Anaesthetists, Speech and Language Therapists etc. Such children may require more than one surgical procedure during their lifetime.

*https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6128172/#:~:text=Crouzon%20syndrome%20occurs%20in%20approximately,births%20(1%3A%2060%2C000).&text=It%20is%20considered%20the%20most,all%20craniosynostosis%20cases%20at%20birth.

Yusuf Ali, a child with Crouzon's Syndrome, was recently treated by the Craniofacial Team at Narayana Health's SRCC Children's Hospital.

This 17 years old child was having serious issues with breathing, including sleep apnoea. Besides, his bulging eyeballs were at risk of exposure to keratitis. He had undergone two surgeries as a baby, namely a **Ventriculoperitoneal shunt** for excessive fluid in the brain (hydrocephalus) and a **Fronto Orbital advancement** to bring his flattened forehead forwards.

This admission was planned for a surgery to bring his forehead and mid-face forwards so as to make space for his eyeballs and increase the diameter of his airways through the nose and mouth.

The surgery is called **Monobloc Advancement** using a **RED (Rigid External Distractor) frame.**

This frame was attached to the sides of his skull, forehead, cheek and jaw bones with wires. The frame was then used to advance the forehead and face by 1 mm / day starting one week after surgery. The frame stayed on the patient's head for 2-3 months after which it was removed, confirming adequate advancement on imaging.

This process of slow advancement leads to new bone formation in the gap generated by pulling the forehead and face forwards.

A team of Craniofacial Surgeons comprising Plastic Surgeons Dr. Nitin Mokal & Dr. Amita Hiremath, and Neurosurgeons Dr. Uday Andar & Dr. Saurav Samantray performed the procedure.

Dr. Nitin Mokal
Paediatric Plastic Surgeon

Expertise:

- Craniofacial distraction osteogenesis
- Correction of cleft lip nose deformity
- Aesthetic plastic surgery

Dr. Uday Andar
Paediatric Neurourgeon

Expertise:

- Congenital scoliosis
- Spinal dysraphism
- Craniofacial anomalies

Dr. Saurav Samantray
Paediatric Neurourgeon

Expertise:

- Epilepsy surgery
- Brain and spine tumours
- Spina Bifida (Dysraphisms)
- Hydrocephalus
- Craniosynostosis
- Spasticity
- Vascular surgery

Yusuf, a very intelligent and brave boy, co-operated very well with the whole procedure. His mother noticed a discernible change in his smile and eyes immediately after the surgery, and the change turned her feel emotional and optimistic about the surgery. Yusuf could eat, drink, sleep, read, walk and do every other routine activity within a week of surgery.

On Yusuf's birth, the Akhtar family were told that Yusuf would never make it past his childhood and that if he did he would grow up with severe mental retardation and disability.

His story proves the need for awareness and sensitivity towards a condition like Crouzon's Syndrome. With correct guidance and motivation, such children, if referred to and treated by a team, experienced in the management of Craniofacial disorders, can have a normal life and, future!

Don't Let Abnormal Brain Structure Hamper Brain Potential

Appointments
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