



# Craniosynostosis - A Case Series and a Brief Review of Literature

Ganapathy S<sup>1\*</sup> and Gopal S<sup>2</sup>

<sup>1</sup>Department of Neurosurgery & Spine Surgery, Manipal Institute of Neurological Disorders, India

<sup>2</sup>Department of Neurosurgery, Institute of Neurosciences, Sakra World Hospital, India

## Abstract

Craniosynostosis is a uncommon disorder affecting the growing skull. Timely surgery and physical therapy can give excellent results restoring neurological function and cosmesis. However, miss the appropriate juncture, and severe consequences will follow. We present our institutional experience of varied syndromes along with a short review of literature.

**Keywords:** Craniosynostosis; Strip Craniotomy; Suture excision

## Introduction

Craniosynostosis is defined as a premature closure of one or more of the cranial sutures leading to raised intracranial pressure and developmental disorders of the brain [1]. The incidence and occurrence are sporadic with certain recognized risk factors. Occasionally, syndromic Craniosynostosis also exists, where a combination of cranial and extra cranial abnormalities give rise to a complex set of problems often difficult to handle [2-4].

## Case Presentation

We present a simple summary of the disease data from our institution, collected over 5 years with patients followed up for 5 years post-surgery. The results, and inferences gleaned from these patients is presented below.

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#### \*Correspondence:

Sibhi Ganapathy, Department of Neurosurgery & Spine Surgery, Manipal Institute of Neurological Disorders (MIND), 13 'Waterwoods', Varthur Road, Ramagondanahalli, Bangalore-560066, India, Tel: 09686617831;

E-mail: sibhig09@gmail.com

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**Primary Craniosynostosis:** A primary defect of ossification.

Suture lines across the skull are described below

**Metopic suture:** This exists between the 2 growing frontal bones. It closes by 3 to 9 months of age. It exists to allow the growth of the massive frontal lobes responsible for personality, emotion, cognition and speech [5].

**Coronal suture:** This is the next most commonly affected suture by craniosynostosis. It fuses normally by 24 years of age completely. Coronal suture allows the growth of the parietal and temporal lobes of the brain [6].

**Sagittal suture:** The longest and most important suture present fuses by the 3<sup>rd</sup> decade of life. It allows the complete development of the hemisphere. The anterior fontanelle, the point of convergence of the coronal, metopic and sagittal sutures closes by 18 months of age. A bulging Anterior Fontanelle (AF) is generally a sign of raised ICP, and requires investigation [7].

**Lamboidal suture:** This occurs most commonly as a consequence of not moving the child from the bed, leading to a positional pressure related deformity. Usually if unilateral they result in occipital plagiocephaly. If bilateral, (rarely) it results in a tower skull deformity (especially if associated with coronal synostoses as well) known as Turricephaly or Oxycephaly (Figure 1) [8].

**Craniosynostosis is classified as either:**

**Primary:** (When 1 or more sutures fuse prematurely, skull growth can be restricted perpendicular to the suture. If multiple sutures fuse while the brain is still increasing in size, intracranial pressure can increase. The cause has been identified as a primary defect in the mesenchymal layer ossification in the cranial bones. A gene locus for single suture craniosynostosis has not been identified [9,10].

**Secondary:** Craniosynostosis is described as a failure of brain growth, which precipitates a small and often misshapen skull. This is less common. The disease can also be classified as:

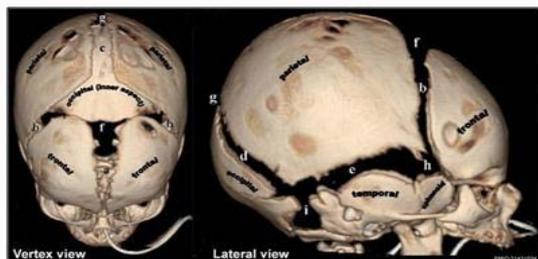


Figure 1: Showing sutures of the skull.

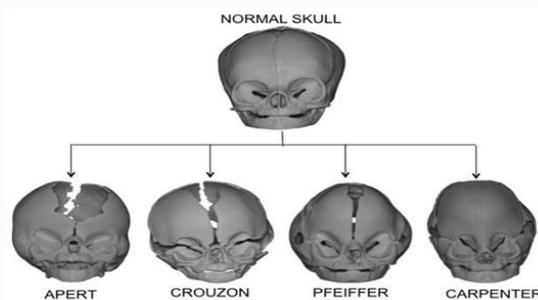


Figure 2: Skull deformity in syndromic Craniosynostosis.

**Simple** craniosynostosis, where only 1 suture fuses prematurely or,

**Complex or compound:** Craniosynostosis, where premature fusion of multiple sutures occurs. The primary factor that keeps sutures open is ongoing brain growth. Normal skull growth occurs perpendicular to each suture. The suture lines aren't fused at birth to enable the skull to be flexible enough to get through vagina. This phenomenon is called moldings. The skull as a whole however needs to be expansible enough to accommodate rapid brain growth.

**Syndromic Craniosynostosis:** Although only 10% to 20% of cases, these conditions form a complex series of disorders which make successful resolution difficult. They are usually autosomal dominant in inheritance linked to Chromosome 10q. Syndromic craniosynostosis are usually multi-sutural & complex cases requiring a multidisciplinary approach along with long term care in order to achieve good results [11,12].

Diagnosis is made clinically through analysis the shape of skull, detecting features of raised ICP, documenting delayed developmental milestones and assessing syndromic features in other organ systems (in complex synostoses) confirmation is usually made through imaging modalities [13,14]. A summary of the common syndromes resulting in craniosynostosis are listed in Table1 (Figure 2).

Common clinical presentations include

1. Trigenocephaly (Figure 3A)
2. Brachycephaly (Figure 3B)
3. Anterior Plagiocephaly (Figure 3C)
4. Dolichocephaly (Figure 3D)
5. Occipital Plagiocephaly (Figure 3E)

X-Ray of the skull is the best screening modality, where one looks for a copper beaten appearance along with premature fused sutures. Once suspected, further investigations are done with CT scans of the head.



Figure 3A: Trigenocephaly.



Figure 3B: Brachycephaly.



Figure 3C: Anterior plagiocephaly.

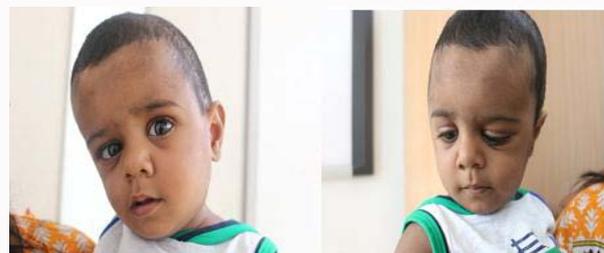


Figure 3D: Dolichocephaly

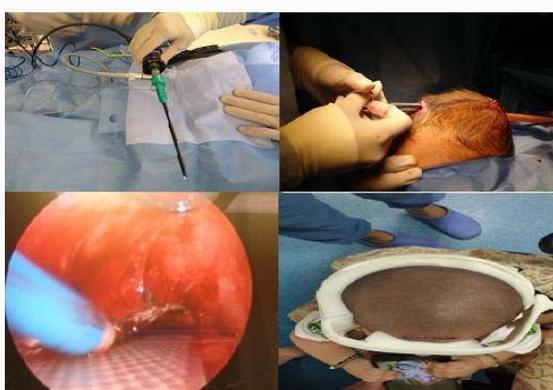


Figure 3E: Occipital plagiocephaly.

These are the best for pre-op planning (3D recon imaging) especially in craniofacial abnormalities with syndromic craniosynostosis. MRI brain is useful only in patients with developmental delay, seizures, suspected hydrocephalus or features of ICP. MRI screening of the brain is mandatory in syndromic craniosynostosis [15]. The Decision



**Figure 4:** Radiological features of craniostenosis consisting of X-ray images (A), Showing deformed shape with copper beaten appearance (B), CT skull showing the 3Dreconof the skull (c), MRI of the brain showing hydrocephalus and a missshapen ventricle (D).



**Figure 5:** Endoscopic suture excision done using a 30 degrees endoscope (A), Being used to cut the bone with a scissors (B), The suture bone being removed (C), The child given a specially designed helmet to preserve the contour of the skull (D).



**Figure 6:** Bicoronal flap with strips marked on the skull (A), the bone removed and cut into strips (B), and repositioned back to the skull (C) The Post Op CT scan showing the strip craniotomy reshaping the skull.

to operate depends upon the presence of features of raised ICP along with any neurological deficit. Particular importance is given to vision, speech and gait above others due to their vital importance in further brain development coupled with the poor prognosis in recovery once damaged. Learning regression (especially in association with other deficits) is also an important indication when clearly documented. Cosmetic considerations usually most important as it affects peer acceptance, parent-child bonding, self-image and coping (Figure 4) [16-18].

### Surgical Correction

Surgical correction once decided upon can assume many forms ranging from distracter placement to the time and tested strip craniotomy concept. Surgery for craniostenosis has undergone a massive improvement over the last decade, with a spectrum of procedures available to suit different situations thereby reducing the morbidity of surgery and improving post-surgery. Types of surgery range from below.

#### Suture excision

Here there is cutting of the fused suture allowing normal brain expansion to occur. It can be done endoscopically with small incisions, thereby reducing blood loss and effect early discharge (2 to 3 days alone) a 30-degree endoscope is used through a remote incision and bone cuts are made by strong scissors followed by removal of fused bone and placement of the 3D printed helmet (Figure 5) [19].



**Figure 7:** Burrholes made to start the craniotomy (A), With removal of the frontal bones and orbital ridge as seen in (B), With the before surgery (C), and postop images (D).

#### Strip craniotomy

The commonest and most successful procedure done is the strip craniotomy. It is ideally performed in young infants less than 2 yrs of age, and only if brain growth is stifled. Sometimes skin closure and healing can be challenging recovery and cosmesis. After surgery a special helmet is placed over the head to ensure molding of the now

**Table 1:** Distinguishing features of syndromic Craniosynostosis.

|           | <b>Muenke</b> | <b>Crouzon</b> | <b>Jackson-Weiss</b> | <b>Apert</b>     | <b>Pfeiffer</b>        |
|-----------|---------------|----------------|----------------------|------------------|------------------------|
| Thumbs    | Normal        | Normal         | -                    | Fused to fingers | Broad & Deviated       |
| Hands     | Carpal fusion | Normal         | Variable             | Bone Syndactyly  | Variable Brachydactyly |
| Great Toe | Broad         | Normal         | Broad & deviated     | Fused to toes    | Broad Deviated         |
| Feet      | Tarsal fusion | Normal         | Abnormal tarsals     | Bone syndactyly  | Variable brachydactyly |

mobile skull strips (Figure 6) [20-21].

### Orbital advancement

Orbital advancement the commonest procedure for facial dimorphisms especially in syndromic craniosynostosis. It combined with strip craniotomies offers excellent results [22-25]. It is usually done usually after 2 years of age. Here bone pieces cut, reshaped and put back along with a remodeled skull. A CT scan after surgery confirms the results (Figure 7).

### Discussion

Complications of Surgery include blood loss, hypothermia, post-operative seizures, CSF leak, wound infection, meningitis, non-healing of sutures, implant failure (if using distracters). There may be a need for multiple procedures as well in the future. Impairment of ocular mobility, dissociated movements, amblyopic and refractive errors. Pre- and post-op impairments seen most frequently with unilateral coronal and metopic synostoses. Thus, long term follow-up remains vital to ensuring the gains of surgery do not gradually end up lost. Follow up must be done regularly upto 12 years (until bone maturity develops) with special emphasis given to vision, speech, feeding & swallowing. Lastly genetic counseling is vital for compliance of the family to potentially distressing therapy and surgery in very young children.

Certain special conditions may mimic craniosynostosis but may not need surgery. These conditions include:

**VP shunting** cause scaphocephaly, and chronic hydrocephalus thickening the skull. In such conditions surgical Indications are definitive. Here an OFC (Occipital Frontal Circumference) >50 cm (4 to 5+ STDs), along with when VP shunt performed on very low birth weight babies.

**Prematurity**, leading to deformational scaphocephaly associated with impaired mobility due to prolonged positioning. If not corrected, it may persist until adulthood. Prevention of craniosynostosis can be effected by using donut-shaped head supports, and waterbed mattresses. Most conditions do not warrant intervention.

**Microcephaly:** Here surgical correction not indicated despite an abnormal OFC as in primary craniosynostosis, OFC remains normal yet oddly shaped. These are rare cases of multicultural craniosynostosis restricting head growth, but which manifest with increased ICP.

**Positional deformation:** This is the most common cause of an abnormal skull shape. Usually only forehead asymmetry occurs sometimes associated with torticollis. Coronal or lamboidal sutures maybe involved as well. Around 40% of new-borns.

### Conclusion

Craniosynostosis is treatable with minimally invasive surgery tailored to suit the needs of each individual patient. Proper follow up and counseling give rise to excellent long-term results with

prevention of brain damage.

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