

Craniosynostosis: Clinical Presentation and Outcome of Surgical Treatment

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ABSTRACT

Objective: Craniosynostosis presents sclerosis cranial sutures, ossification and fusion involving the vault of the base, sutures involved can be one or more sutures. This study aimed to determine the surgical outcome of craniosynostosis.

Material and Method: The prospective study was conducted at the neuro spinal and cancer care institute, Karachi. Patients presented with sagittal, metopic, unicoronal, lambdoid, bicoronal craniosynostosis were included in the study, while patients with coagulopathy, previously operated cases were excluded from the study.

Results: We had 26 children in our study, age range about 1–3 years. Patients were cleaved into groups depending on their age. Most of the children 15 (57.6%) were in 1–2 years age group and 11 (42.3%) were in 2–3 years of age. Boys were 18 (69.2%) and girls were 8(30.7%). Coronal 11 (42.3%) was the most common suture involved, followed by sagittal 9 (34.6%). Lambdoid suture 3 (11.5%), metopic 2 (7.6%) and 3 (11.5%) case presented with raised intracranial pressure with multiple sutures closed involved. Strip craniectomy was done in all cases. We did bicoronal flap and Scalp flap turned into a supraorbital region while in metopic suture Fronto-orbital advancement and remodeling approach were used. No major complication was observed.

Conclusion: Cases which are managed early age have given good acceptable results in follow up, proper surgical expertise, perioperative management of temperature, blood loss, relieving the restriction of sutures and normalizing raised intracranial pressure can decrease the morbidity and mortality.

Keywords: Craniosynostosis, strip craniectomy.

INTRODUCTION

Craniosynostosis presents sclerosis cranial sutures, ossification and fusion involving the vault of the base, sutures involved can be one or more sutures. The identified occurrence varies from 0.3/1000 to 3.1–6.4 in 10,000 live births and still at rise.^{1,2,18} Because of early fusion of sutures in Craniosynostosis leading to the altered shape of the cranial vault and this results in associated anomalies that cause Cosmetic deformity and problems such as limited brain development, elevated intracranial pressure, and seizures.³ Etiologically craniosynostosis is recognized as the substantial heterogeneity in underlying causes, the complex interplay of potentially causative factors lies on the pathological difference between fusion of

different cranial sutures involving, polygenic background, intrauterine environment, development and growth of the brain and chromosomal disorders.⁴ It is estimated that around 85% of the disorders are nonsyndromic and isolated, which involves only a single suture. Syndromic craniosynostosis such as Pfeiffer, Crouzon, Apert, syndromes affects multiple sutures additionally linked with clinical signs of cardiac abnormalities, hand and foot malformations, delays in growth, structural problems.⁶ The diagnostic approach to craniosynostosis based primarily on physical examination and skull deformity, craniometrics measuring, and the radiological assessment of x-ray skull and CT scan brain.⁵ Lane's first surgical treatment of craniosynostosis occurred in

1892, with various procedures going in and out of favor. But the advent of craniofacial surgery by Paul Tessier, brought with it a more reliable and standard of care, become popular. Over the past decade, surgeons have witnessed a resurgence of strip craniotomies for the treatment of single and multi-suture craniosynostosis. The current protocol is an open skull vault expansion modified Tessier's descriptions. But, previous research into the safety of these procedures has either focused mainly on blood loss, Protocols are more creative and have improved the outcomes.⁷ Current intervention methods are based on open calvarial reconstruction, minimally invasive strip cranial surgery, endoscopic expanded strip cranial surgery and endoscopic cranial distraction with help of postoperative helmet for molding purpose, cranial distraction, mini invasive strip craniectomy having spring implantation can be effective. Many surgical options are available, including the open approach. Early intervention gives favorable outcomes.^{8,9} Rationale of the study was to increase awareness regarding this disease and prevent mismanagement that are common in developing countries due to lack of information regarding treatment.

MATERIAL AND METHODS

Study Design and Setting

A Prospective descriptive study was conducted from 23. 3. 2012 to 20.04.2019 at the Neuro-Spinal and Cancer Care Institute, Karachi.

Sample Size & Sampling Technique

Sample size was calculated on 0.3/1000 live birth,¹⁸ that was calculated as 10 patients with 95% confidence interval through WHO Open Epi software.

Inclusion Criteria

Inclusion criteria of patient included those with metopic, sagittal, unicoronal, lambdoid, bicoronal, > 4 months to withstand surgical procedure, relevant skull deformity.

Exclusion Criteria

Cases with coagulopathy, previously operated cases and age less than 4 months were excluded.

Data Collection

Data was collected as Non-probability consecutive

sampling, a patient presenting with craniosynostosis were admitted via OPD at neurospinal and cancer care institute, Karachi after diagnosis based on history taking, an examination of cranium and involved suture, radiological imaging skull x-ray with front and lateral view and 3D CT reconstruction, CT scan brain. Suture involvement, unilateral coronal, sagittal bilateral coronal, bilateral lambdoid, unilateral lambdoid, metopic or multiple suture were included in study. Nine month was the average age of surgery. The follow up was done in OPD for six months. It's a rare disorder which presents to OPD, not many published articles available in Pakistan based on single center study and usually craniosynostosis study based on limited number of patients (n = 24-45), for a single center this is a decent number, the patients were admitted a day before to procedure, and parents counselling was done by the senior consultant neurosurgeon. Outcome of surgery was assessed based on irregularities and residual deformity, associated complications, or need of additional surgery. Data was analyzed in SPSS 23.00.

RESULTS

Age Range

We had 26 children in our study, age range about 1 – 3years. Patients were cleaved into groups depending on their age. Most of the children 15 (57.6%) were in 1 – 2 years age group and 11 (42.3%) were in 2-3 year of age shown in (Table 1).

Gender Distribution

Boys were 18 (69.2%) and girls were 8 (30.7%).

Clinical Presentation

Coronal 11 (42.3%) was the most common suture involved, followed by sagittal 9 (34.6%). Lambdoid suture 2 (7.6%), metopic 3 (11.5%) and Multiple sutures in 1 (3.8%) patient while, 1 (3.8%) case presented with raised intracranial pressure with multiple sutures closed involved.

Surgery

Strip craniectomy was done in each and every case. We did bicoronal flap and Scalp flap turned into a supraorbital region while in metopic suture Fronto-

orbital advancement and remodeling approach were used.

The minor dural tear was in 2 cases, 100cc blood was arranged for 70% cases and the operation theatre temperature was maintained. Prophylactic antibiotics were given for 5 doses. 3 (15%) patient had fits for which anti-epileptics were given.

The involved intracranial strain was found most frequently in involvement (metopic or sagittal), a suture of the midline rather than where a single coronal suture was fused. We inferred that intracranial hypertension exists with single-suture craniosynostosis in a large proportion of the babies.

Complications

No major complication was observed (**Table 2**). Two patients had pre procedure fits and one patient developed post procedure fits.

Table 1: Demographical Presentation of the Patient.

Parameter	n = 26
Age	
1 to 2 year	15 (57.6%)
2 years to 3 years	11 (42.3%)
Gender	
Male	18 (69.2%)
Female	08 (42.5%)

Table 2: Location of Involved Suture.

Involved Suture	Presentation	n = 26
Coronal suture	Plagiocephaly	11 (42.3%)
Sagittal suture	Scaphocephaly	09 (34.6%)
Metopic suture	Trigonocephaly	03 (11.5%)
Lambdoid suture	Posterior Plagiocephaly	02 (7.6%)
Multiple sutures	Craniosynostosis	01 (3.8%)

DISCUSSION

Craniosynostosis is a congenital disorder need to be addressed a multidisciplinary approach and correcting certain factors can impact the outcome of surgery and least is written is on this disorder in our region. In rural areas of Pakistan mishandled and presentation is

late because of lack of knowledge and awareness about this disorder in the community.

In a study by Synèse¹⁰ there was male predominance with average age 13.11 months. Craniosynostosis subjugated 40.38%, by brachycephaly which is followed by another scaphocephaly (21.15%) plus had Apert Syndrome in two patients. The surgical techniques were suturectomy in the majority of cases, with blood transfusion at the time of the incision compared to our study we also had male dominance in cases, the coronal suture was the common suture involved and we did strip craniectomy with blood transfusion in all cases, compared to our study we did not receive any patient with the syndrome in our study.

A regional survey published with orbital development of Frontal and calvarial total remodeling of the calvary. They had increased skull dimensions in their analysis and all instances the parents were pleased with the skull form with problems in 11.1% including wound infection and 1 death¹¹ while we did advancement in metopic sutures without having major complication.

The study by Lionel¹² stated that: Twenty-two patients, 9 (40.9%) or were females, with standard age of -21.4 months. Another very ordinary suture implicated was coronal in eighteen patients (81.8%), followed by sagittal thirteen patients (59.1%), metopic in twelve patients (54.6%) and lambdoid in eleven (50%) respectively. 7 (31.8%) had involvement of multiple sutures, while 13(59.1%) had syndromic syndromes (Apert's, Crouzon's and Down's syndrome) compared to our study we had no syndrome patient but similarly, we had coronal, sagittal, followed by metopics. A study by Sharma¹³ they also had a patient up to 7 years and older while compared with our study age was up to 3 years.

Shim¹⁴ in the study showed that Craniostenosis not merely considered a clinical condition, families of the patient make more complex demands, especially for aesthetic appearance but also demand good outcome in terms of cognitive consequences. The presented view of the clinician will be oriented towards holistic supervision, respecting both presentation and function.¹⁴

One study showed that the complication rate was 3.1%, reoperation rate of 2.45%, and readmission rate of 2.8%. Overall, 67% of the patients received a blood transfusion. Duration of surgery and anesthesia are significantly associated with blood transfusion¹⁵ while we did not observe any major complication

dural tear was in two cases; blood was transfused in 70% cases.

The study by Fearon¹⁶ Children with lambdoid suture have associated chiari malformation in the majority of cases and require routine by considering magnetic resonance imaging. Treatment with cranial remodeling procedures for lambdoid craniosynostosis may result in suboccipital decompression. In their studies, 96% was managed in the same sitting procedure. While, we had two patients of lambdoid suture MRI cervical spine was done pre-operative to clear any associated chiari malformation.

In another study, it is said that Endoscopic method effective, safe & it is durable in correcting craniosynostosis. It helps normal head growth with decrease need of blood transfusion while Early identification of craniosynostosis and referral by pediatricians helps in better result.¹⁷ Compared to our study we had experience team to while blood transfusion was in 70% cases and the temperature was on priority per operatively while in a different part of Pakistan people still have a dilemma about treatment, they consider such child special and give no consideration to treatment due to which they present late although education and awareness at physician level are provided to get the best treatment in early months of the disorder.

CONCLUSION

Cases which are managed early age have given good, acceptable results in follow up, proper surgical expertise, perioperative management of temperature, blood loss, relieving the restriction of sutures and normalizing raised intracranial pressure can decrease the morbidity and mortality.

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Additional Information

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Human Subjects: Consent was obtained by all patients/participants in this study.

Conflicts of Interest:

In compliance with the ICMJE uniform disclosure form, all authors declare the following:

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AUTHORS CONTRIBUTIONS

Sr.#	Author's Full Name	Intellectual Contribution to Paper in Terms of:
1.	Aurangzeb Kalhoro	Study design and methodology. Proposed topics and Basic Study Design, & data collection. Data collection and calculations.
2.	Abdul Sattar M. Hashim	Referencing, data calculations, and manuscript writing. Analysis of data and interpretation of results etc.

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