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Peri-Operative Considerations and Surgical Outcomes in Craniosynostosis: A Tertiary Care Centre Experience

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ABSTRACT: Aim : To evaluate the surgical outcomeof total and subtotal calvarial remodelling with or without fronto-orbital advancement, strip craniectomy and distraction osteogenesis in the management of syndromic as well as non-syndromic craniosynostosis.

Methods:A retrospective study was conducted at our institution between 2010 and 2020 for both non-syndromic craniosynostosis (3/10) and syndromic craniosynostosis (7/10).There were a total of ten cases (six males and four females) of craniosynostosis which were operated for skull deformity, developmental delay and neurological disability. Amongst these ten cases, age at the time of surgery ranged from 23.2 months (range 3.5 months to 5 years). Patients were monitored for intra-operative, post-operative complications, short and long-term outcomes in terms of neurological sequelae and progression of cranial morphology. Follow-up data was collected for all of these patients at 4 weeks, 3 months, 6 months and one year.

Results: The results of cranial vault remodeling withFronto-orbital advancement and cases of total cranial vault remodeling without fronto-orbital advancement were stable in majority of the cases (n=7/10) howeverthere were patients (n=2/10) who developed post-operative neurological sequelae (vision change, Headache) and one case (n=1/10) which was fatal.On evaluating our results as per Whitaker grading system 60% cases (n=6) required no surgical refinements , 20% cases (n=2) were planned for midface distraction later and 20 % cases (n=10) were taken up for revision surgeries.

Conclusion:Open surgical techniques like total and subtotal calvarial remodelling with or without fronto-orbital advancement, strip craniectomy and distraction osteogenesis in syndromic as well as non-syndromic craniosynostosis are considered reliable. The long term stability may confer greater long-term skull shape correction and decreased neurological sequelae.

Keywords: Craniosynostosis, Calvarial reconstruction , Syndromic Craniosynostosis, Non syndromic-craniosynostosis, Neurological sequelae

I. INTRODUCTION:

Craniosynostosis is a developmental anomaly which results in impairment of brain development and abnormally shaped skull[1]. The main cause of craniosynostosis is premature closure of one or more cranial sutures. It can occur as an isolated deformity or in combination with other malformations as part of complex craniofacial syndromes. When left untreated, craniosynostosis can result in serious complications, such as developmental delay, facial abnormality, sensory, respiratory and neurological dysfunction, anomalies affecting the eye, and psychological disturbances.[2]

It affects approximately 1 in every 2000–2500 newborns and is the second most common craniofacial anomaly after oro-facial clefts[2].During the embryonic development, the cranial vault develops from the mesenchymal tissue. It is first arranged as a capsular membrane around developing brain. Gradually, the outer mesenchymal layer is formed through the process of intramembranous ossification. This intramembranous bone growth depends mainly on the direction of the forces that are defined by the growth of the brain. In the developmental period, the brain is surrounded by dural fibers, which are closely related and strongly attached to the sutural system[3-4].

Calvarial sutures are formed during the embryonic development at the sites of approximation of the membranous bones and later represent the major sites of bone expansion. This

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process is a combination of deposition of osteoid at the sutural margins, ii) surface apposition and resorption (remodeling) of the bone, and iii) centrifugal displacement by the expanding brain .The fusion of the sutures is mainly regulated by the dura mater, which provides many important regulators of growth, such as intercellular signals (for example, signaling mediated by fibroblast growth factor [FGF] and transforming growth factor beta [TGF-β], mechanical signals, and cells which undergo transformation and migrate to the sutures. This complex signaling cascade can be disrupted by a large number of genetic mutations, leading to an abnormal development and premature fusion of one or more sutures, which is called craniosynostosis .[5-6]In this study we intend to present our institutional experience in the management of both syndromic and non-syndromic craniosynostosis using an array of surgical modalities.

Patients and methods:

Hospital records of all multidisciplinary craniofacial cases were evaluated between Jan 2010 and Jan 2020 at Department of oral and maxillofacial surgery at our tertiary care center.A multidisciplinary team comprising of members from plastic surgery, neurosurgery, pediatric, neuro-anesthesia, ophthalmology, genetics, and intensive care departments is dedicated for comprehensive care of syndromic and non-syndromic patients.

In the present study patients were reviewed with regard to demographic data, surgical technique, intraoperative factors and post-operative complications. Demographics included age, gender, timing of intervention and type of synostosis. Different surgical techniques comprising of total and subtotal calvarial remodelling with or without fronto-orbital advancement, strip craniectomy and distraction osteogenesis were opted for these patients based on a thorough pre-operative clinical and radiographic assessment. The goals of surgical intervention included the release of the affected suture allowing the unrestricted development of the visceral components and three dimensional reconstruction of the skeletal components establishing a more normal anatomic position and contour .Intraoperative variables included total surgical time, blood loss, blood transfusion and dural tear. Post op evaluation comprised of anaesthesia associated complications, post-operative ICU stay, ophthalmic complications, CSF leak, poor cosmetic result and total length of hospital stay.We assessed our surgical results using Whitaker classification.

Investigations:

As per instituitional protocol, all cases were evaluated by a pediatrician, ophthalmologist and a neurosurgeon on first visit and after a systematic clearance they were subsequently examined by plastic surgeon and maxillofacial surgeon for feasibility & planning of surgical treatment. A pre-operative CT scan and MRI in majority of the syndromic cases was done in the same institution as per standardised pediatric CT norms ($CTDI_{vol}$ of 31.26 mGy or lower).All selected cases were also subjected to a detailed pre-anestheticcheck up.

Surgical Techniques:

Strip Craniectomy

A total of five cases of brachiocephaly and one case of plagiocephaly were treated using this technique. Cases were taken up under GA with armoured ETT(Oral intubation).1 in 100,000 adrenaline saline solution was used for hemostasis. Coronal incision was placed and subgaleal flap was elevated. Dissection of the scalp was done anteriorly and posteriorly as per the sutures involved. 3-cm to 4-cm strip was marked along the marked sutures and osteotomy using piezoelectric saw was carried out, encompassing the fused suture. The bone flap was elevated followed by restructuring of the osteotomised bone strips and fixation with bioresorbable plates and screws.

Distraction Osteogenesis :

Two cases of trignocephaly underwent DO. After the calvarial exposure the synostosed sagittal suture and coronal suture was osteotomized to release it from the parietal and frontal bones. Distractors were placed in parallel with the vector of distraction in a coronal plane and transverse plane.

Total Cranial Vault Remodelling

This technique involved complete exposure of the calvarium followed by multiple craniectomies and replacement of the bone flaps in altered positions to remodel the overall shape of the skull in a single stage. The technique was used in four cases of non-syndromic craniosynostosis amongst which one was previously operated incomplete correction of Pi procedure from a different institution. Fixation was carried out using miniplates and screws.

Fronto-orbital Advancement

Exposure was the same as used in other techniques .The periosteum was incised about 2 cm above the supraorbital ridge and then rest of the

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dissection was done in the subperiosteal plane to expose the roof of the orbit and the lateral orbital margins. The supraorbital neurovascular bundle was gently teased off the and in some cases an osteotomy was required to reflect the flap downwards. Bifrontal craniotomy was done by the neurosurgeon followed by osteotomy of the supraorbital bandeau segment. After reshaping of the segment and preliminary fixation of this segment it is replaced in the advanced position to the frontal lobe and fixed laterally. The frontal component is placed on top of the advanced bandeau and fixed.

II. RESULTS

A total of 10 patients were included in this series. The mean age at the time of surgery was 23.2 months (range 3.5 months to 5 years). Sixty percent of the patients were male. The most common indication for surgery was skull deformity (50%) followed by hydrocephalus ,raised ICP, associated ocular changes and to risk of further developmental delay. Cases were grouped into syndromic and non-syndromic groups amongst them most common was plagiocephaly (40%), followed by metopic synostosis (30%), sagittal synostosis (20%), and Brachycephaly(10%)

Majority of plagiocephaly cases underwent total calvarial remodeling with fronto-orbital advancement (30%) and distraction osteogenesis (10%) followed by total calvarial remodeling with fronto-orbital advancement (10%) in brachycephaly cases while metopic craniosynostosis (30%) and sagittal craniosynostosis cases underwent strip craniectomy (20%).

The perioperative complications is as shown in Table 4. The average operative time was 131 minutes (54–313 minutes). The average estimated blood loss was in the range of 10-20 mL/kg with an average transfusion rate of 34 mL/kg. Blood conservation procedures like induced hypotension, antifibrinolytic agents, preoperative erythropoietin (EPO) were undertaken on a case to case basis to minimise blood loss.The average hospital stay was 5 days. 40% of patients had postoperative complications. There was one fatal case and one case with vision loss. The second complication was a small area of scalp necrosis requiring a skin graft.

Table 1: Indications for undergoing treatment

Indication for treatment	No of cases
Skull deformity	5
Hydrocephalus	4
Raised ICP	3
Ocular symptoms	3
Risk of developmental delay	1

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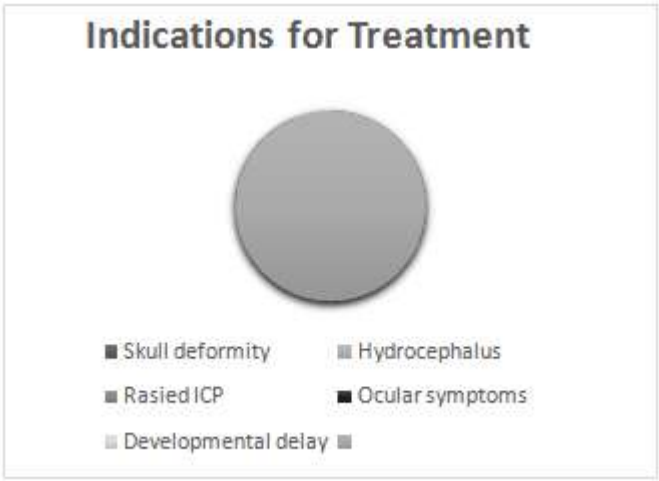

Table 2:Age and Gender distribution in Syndromic craniosynostosis subjects

Diagnostic group (Syndromic craniosynostosis)	Average Age at Primary surgery (months)	Gender	No of cases
Sagittal synostosis	13.4	M	02
Brachycephaly	12.3	M	01
Plagiocephaly	60	M & F	02
Metopic synostosis	12	F	02
Total			07

Table 3: Age and Gender distribution in Non-Syndromic craniosynostosis subjects

Diagnostic group (Non-Syndromic craniosynostosis)	Average Age at Primary surgery (months)	Gender	No of cases
Plagiocephaly	18	M & F	02
Metopic synostosis	24	M	01
Total			03

Chart 2: Surgical techniques undertaken in the subjects

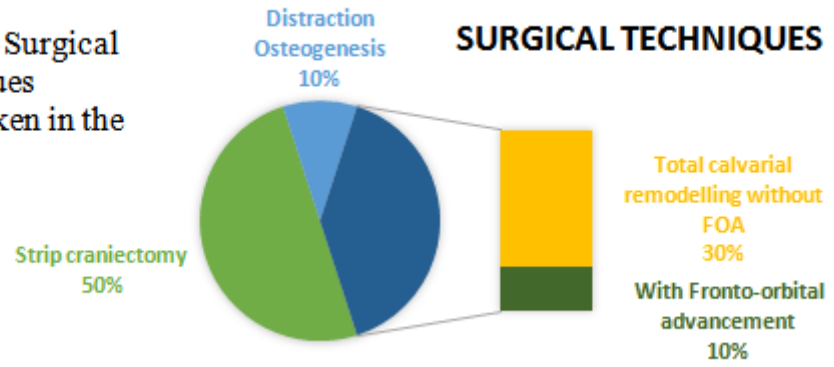

Table 4: Intra and postoperative complications in the subjects

Complications	No of Cases
Haemodynamic changes	**01**
Infection	**0**
Vision changes	**01**
Headache	**01**
Poor cosmetic result	**02**
CSF leak	**01**
Death	**01**
Anaesthesia associated complications	**02**
Scalp necrosis	**01**

Whitaker Classification System

Category A	No refinements or surgical revisions considered advisable or necessary
Category B	Soft tissue or lesser bone contouring revisions advisable; could be done on a outpatient basis or a maximum of 2 days hospitalization
Category C	Major alternative osteotomies or bone-grafting procedures advisable; these procedures were not as extensive as original procedures
Category D	Major craniofacial procedure advisable, duplicating or even exceeding original procedure.

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Categories (whitaker classification)	No of cases
Category A	06
Category B	00
Category C	02
Category D	02

III. DISCUSSION

In our case study majority of the cases were male (60%)(Table 2,3).Male preponderance can be observed in other studies such as that of Sloan et al.[4] where there were 157 men (62.8%) and 93 women (37.2%) in the study, with most of the male preponderance accounted for by the large sagittal synostosis group. In another study by Oliveira et al.[5]there was a male preponderance (71.3% men versus 28.7% women).

The incidence of syndromic craniosynostosis was 9.2% in Sloan et al[4].study, 20.5% in Oliveira et al.[5] study, 6% in Zakhary et al.[8] study, and 70 % in our study.In our study mean age was 23.2 months which was slightly on the higher side because of late reporting at the hospital.(Table 2,3)

The primary goal of syndromic craniosynostosis surgery is to reduce the intracranial pressure, ocular and neurologic complications and to improve the normal growth of the brain. However the primary goal in the management of non-syndromic isolated craniosynostosis is the improvement in cosmetic function. In both the scenarios early operative intervention allows for rapid correction of cranial deformity and facilitates normal brain growth of the child[3]. The skull deformity followed by the risk of mental retardation and developmental delay are the most important factors which governs the surgical treatment in craniosynostosis[4].Bannink et al.[11] in his study observed that papilledema was present in 51% of the syndromic patients with craniosynostosis. In another study by Tim de Jong [16]the incidence of intracranial hypertension was 53% in Crouzon/Pfeiffer syndrome, 33% in Apert syndrome, and 21% in SaethreChotzen syndrome.In the present study subjects skull deformity (50%) was the primary indication for treatment in of cases followed by hydrocephalus (40%), ocular symptoms(30%), raised ICP(30%) and developmental delay(10%).(Table 1)

An array of surgical techniques have been described in literature to manage and treat different types of craniosynostosis. Nonetheless the literature does not identify or state one technique to be superior to other[5-7]. We have performed open surgical techniques like total and sub-total calvarial remodelling with or without fronto-orbital advancement, strip craniectomy and distraction osteogenesis(Chart 2) in our cases with stable results both functionally and cosmetically on follow up (Table 4)[2,5,8,9].

Early studies on surgical treatment of craniosynostosis focused on low rates of mortality and major morbidity[10-13]. Over time complications and certainly mortality have decreased; therefore, more recent studies focus on individual risk factors for complications. Identified risk factors include complex craniosynostosis, increased intraoperative blood loss (over 60 mL/kg), larger volume transfusions (> 45 mL/ kg), longer operative times, and comorbidities[14]. Our post-operative complications (Table 4) included a small area of scalp necrosis, one fatal case, one case with partial vision loss and two patients with sub-optimum cosmetic outcome of which one patient had almost immediate bony ingrowth and required a larger sagittal strip craniectomy at the repeat operation. The other child had significant behavior problems after initial corrective surgery also signs of elevated intracranial pressure so revision surgery was performed after one year.

On evaluating our results as per Whitaker grading system 60% cases (n=6) required no surgical refinements , 20% cases (n=2) were planned for midface distraction later and 20 % cases (n=10) were taken up for revision surgeries for symptomatic postoperative calvarial growth restriction.Sloan et al.[4] in his study observed that there were two deaths (0.8%), both with Kleeblattschadel patients and morbidity/mortality were significantly associated with secondary versus primary operations and syndromic versus non-syndromic patients. Looking at factors related to reoperation we found a trend toward a higher age at the time of initial operation; however, this did not reach statistical significance.

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Fig 1a,1b,1,c

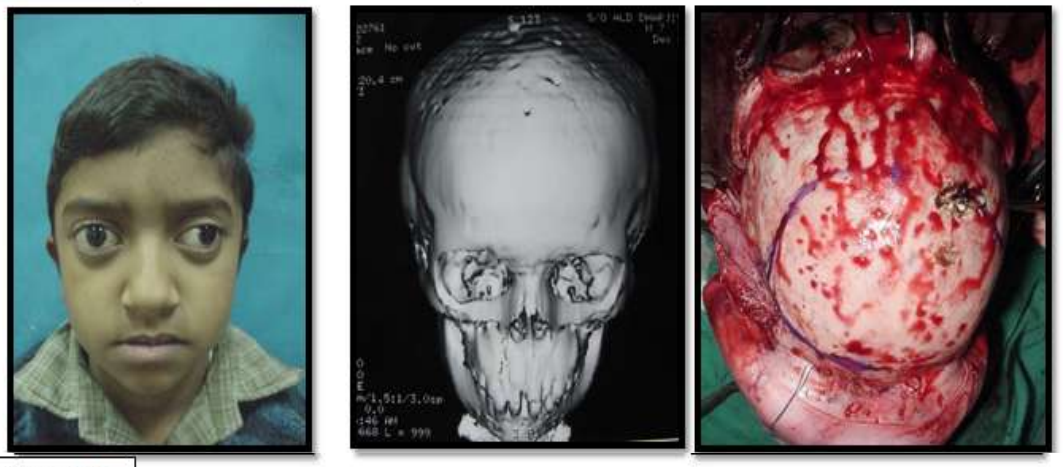

Fig 2a,2b,2c

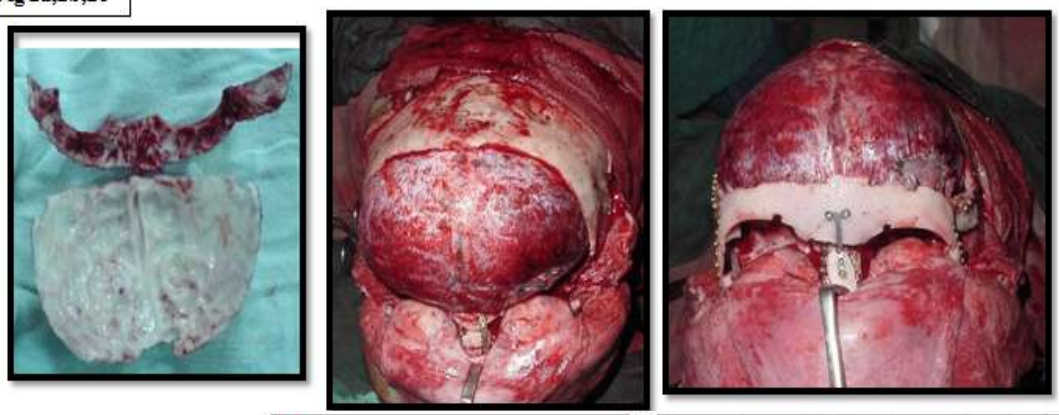

Fig 3a,3b

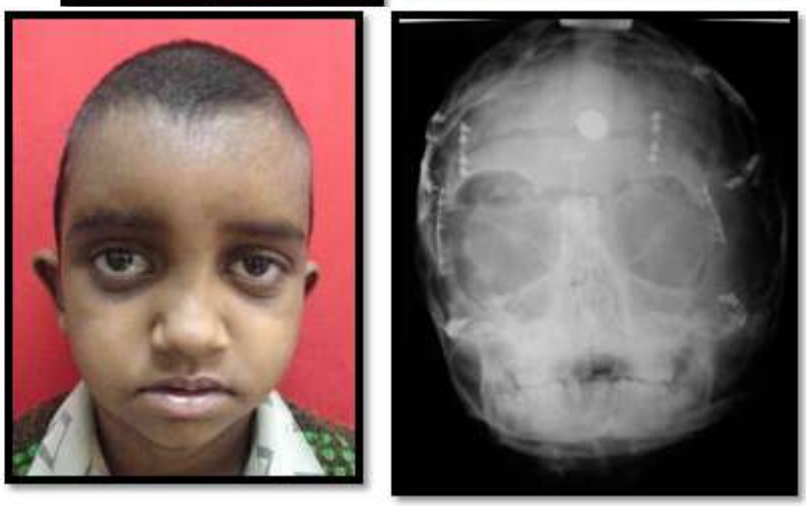

Figure- 1a-Preoperative frontal view of the patient ,1b-Three-dimensional reconstruction of CT obtained in the pa- tient with multiple-suture synostosis 2a-Calvarial exposure with bi-frontal and fronto-orbital osteotomy markings, 2d,e,f,- fronto-orbital calvarial segment remodelling and fixation using miniplates,3a,b-Postoperative frontal profile view and radiograph at I year follow up

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