Original Article

Surgical Management of Craniosynostosis

Hosam A.M. Habib*, Hazem M. Negm, Medhat Momtaz Elsawy and Ihab M. Helmy
Departments of Neurosurgery, Menoufia University, Neurosurgery, Menia University, Maxillofacial Surgery Alexandria University

ARTICLE INFO

ABSTRACT

Background: Craniosynostosis is defined as the premature closure of one or more of the sutures normally separating the infant's skull bony plates, causing abnormal growth of the cranial vault and skull base, which may affect brain growth and development. The management of craniosynostosis varies widely with no consensus on the optimal timing or type of surgical intervention. Treatment varies depending upon patient’s age at presentation, type of synostosis, and the severity of its deformity, and ultimately the surgical team’s preferences. Objective: Aim of this study is to assess the peri-operative complications and operative outcome of the surgical treatment of craniosynostosis. Patients and Methods: Retrospective analysis of the medical records of twenty three patients operated for craniosynostosis. Results: The average age: 9.3±4 months; range: 4-19 month. The follow-up ranged from 1 year to four years (median: 32 months). Cranial vault remodeling was performed in all patients; while orbital advancement was performed additionally in four patients (17.4%). The mean operative time was 169±40 minutes. Postoperative hospitalization averaged of 5.8 days. The mean intraoperative blood loss was 37.5% of the estimated blood volume for age. Operative and postoperative complications were observed in six patients (26.1%); three cases (13%) suffered hypovolemic shock during surgery. A dural tear, extradural hematoma and wound infection each in a patient. All patients had improvement of their preoperative cosmetic status. Conclusion: Performing aggressive cranial or cranio-orbital remodeling provides the best cosmetic results, it should better be performed after sixth month of age to reduce any anticipated morbidity and not be delayed beyond the first year of life if a good cosmetic outcome is desired.

© 2015 Egyptian Journal of Neurosurgery. Published by MEDC. All rights reserved

INTRODUCTION

Craniosynostosis is defined as the premature closure of one or more of the sutures normally separating the infant's skull bony plates, causing abnormal growth of the cranial vault and skull base, which may affect brain growth and development. The skull growth is restricted perpendicular to the fused sutures while promoted parallel to it (Virchows law), in association with compensatory growth in the skull's unfused bony plates. craniosynostosis can happen as an isolated defect (non syndromic) or as part of a syndrome. It is referred to as simple craniosynostosis, when only one suture is involved and as compound craniosynostosis when two or more sutures are involved. There is a male preponderance, which can be explained by the role of androgens in sutural osteogenesis.

The most commonly affected suture is the sagittal suture, which is involved in 40% to 60% of cases, followed by the coronal suture (in 20% to 30% of cases), then the metopic suture (in less than 10% of cases); while true lambdoid synostosis is rare. Syndromic craniosynostosis is less common than the non syndromic types (20%), although more than 150 syndromes with craniosynostosis have been identified. The etiology of non syndromic craniosynostosis is still unknown, and the condition is sporadic in most instances.

The management of craniosynostosis varies widely with no consensus on the optimal timing or type of surgical intervention. Treatment varies depending upon patient’s age at presentation, type of synostosis, and the severity of its deformity, and ultimately the surgical team’s preference.

There are two main indications for the surgical intervention in craniosynostosis; the first is to correct the skull shape for cosmetic and psychosocial considerations, and the second is to provide an adequate space for normal brain growth. From a cosmetic perspective, the deformities associated with craniosynostosis are generally progressive for the first
year of life, and their social and psychological impact upon the affected child is in itself sufficiently concerning to justify intervention. It is important to know that after birth, the cranial vault grows most rapidly during the first year of life, with brain volume doubling during the first 6 months and again by the age of two years. Then, the calvaria continue to grow in a linear fashion until the age of 6 to 7 years, when growth via sutures is essentially complete.

Sagittal synostoses are the most common type of craniosynostosis. Nearly 80% of them are non-syndromic and 6% run in families with dominant transmission. The skull shape is elongated in an anteroposterior direction giving the skull a boat-shaped appearance (scaphocephaly).

Metopic suture closure starts in the third trimester at the level of the glabella moving upwards towards the anterior fontanelle, and is usually fused by the 9th month. It may take up to 2 years to fuse. Premature fusion of the metopic suture can occur either prenatally or before 9 months, giving rise to a triangular-shaped head or trigonocephaly. Of the cases postnatally identified, 75% are isolated and 25% are syndromic.

Bilateral coronal synostosis is commonly associated with syndromes. There is reduction of the anteroposterior dimensions, while the transverse dimensions are increased. The most consistent feature of the coronal synostosis is the “harlequin” deformity of the orbit, as a consequence of the superior elevation of the lesser wing of the sphenoid. Coronal synostosis if associated with skull base involvement, resulting in proptosis and exophthalmos and mid-face hypoplasia.

PATIENTS AND METHODS

The medical records of twenty three patients treated for craniosynostosis (2008 – 2013) were retrospectively reviewed. The collected data included age at operation, gender, the involved sutures, other medical diagnoses and operation records with details of the surgical procedures.

Data concerning the estimated intraoperative blood loss (calculated as percentage of estimated blood volume for age (EBV)), operative duration, postoperative hospital stay and perioperative complications were gathered.

Preoperative Roentgenograms or three-dimensional CT scans (3D CT) in addition to brain MRI had been performed for all patients, to detect any concomitant pathology, and radiological signs of increased intracranial tension (Fig. 1).

Visual analogue scale for cosmesis with a ten point scale was used to assess the parent’s satisfaction with the cosmetic result, where the parent’s perception of the postoperative cosmetic result in comparison to the preoperative deformity was recorded in a retrograde manner.

Clinical follow up was performed to assess bony healing, the presence or absence of a palpable defect and head contour. Due to the cumulative data concerning the long term radiation exposure hazards in children, postoperative radiographic examinations were only performed in three patients due the occurrence of postoperative complications.

The cranial vault bone flaps were raised and contoured to correct the deformities. Barrel-stave osteotomies and green stick fracturing of selected areas of the calvarial bone were performed in an attempt to enlarge the cranial volume (Fig. 2). Fronto-orbital advancement was performed in four cases. After reaching the desired modeling, the bone fragments were held together with stainless steel wires and/or microplates. In children above one year of age, with large bone defects, the defects were filled by hydroxyapatite to promote osteo-conduction thus minimizing possible disfigurement.
RESULTS

Twenty three patients (fifteen males and eight females; average age: 9.3±4 months; range: 4-19 month) were surgically treated for craniostenosis. Seven patients (30%) presenting below six month of age, eight patient (35%) presented beyond one year of age, while remaining eight patient (35%) were operated between sixth and twelvth months of age.

Non syndromic simple synostosis was present in nineteen patients (82.6%), of which fourteen (60.8%) were sagittal (Scaphocephaly) (Fig. 2) and four cases (17.4%) were metopic synostosis (Trigonocephaly) and a single case (4.3%) of bilateral coronal synostosis (Fig. 3). Four cases (17.4%) were syndromic; of which two were Crouzon syndromes (Fig. 4) with oxycephaly and two were Apert syndrome (Fig. 5) with oxycephaly and syndactyia. The follow-up ranged from 1 year to four years (median: 32 months).

Twelve cases had signs of raised intracranial tension on presentation (six cases of scaphocephaly, two cases of Apert syndrome, two cases of Crouzon syndrome, and finally two case of trigonocephaly).

Fig. 2 a-d: a & b: Intraoperative calvarial remodeling in a thirteen month old child. c: Preoperatative and d: Thirty nine months postoperative follow up showing correction of the scaphocephaly, with mild frontal irregularities.

Fig. 3 a-c: a: Preoperative nine month male child with bilateral coronal synostosis. b: Early postoperative subgialal CSF collection due to a dural tear. c: One year follow-up.
Cranial vault remodeling was performed in all twenty-three patients; orbital advancement was additionally performed in four patients (17.4%). The operative time ranged from 120 to 240 minutes (mean 169 ± 40 minutes). Postoperative hospitalization time ranged from 4 to 13 days, with an average of 5.8 days. The mean estimated intraoperative blood loss was 37.5% ± 10 of EBV (range 20 – 55 %). The amount of blood loss during the orbital advancement step (when it is performed) averages 16% of EBV, with a concomitant increase in the operative duration by an average of 45 minutes. A mean of 9% of EBV was lost, in addition, through the suction drain over the next three days.

Operative and postoperative complications were observed in six patients (26.1%) (Table 1); three cases (13%) suffered hypovolemic shock during surgery, one of these patients had cardiac arrest that was revived. Hypovolemia was corrected, but the surgical procedures were terminated before performing the initially planned fronto-orbital advancements; the postoperative period was uneventful for these cases (the advancement
procedures in these three patients were not performed later due to parents refusal). A dural tear occurred in one of the patients resulting in a subgial cerebrospinal fluid collection in the postoperative period that was controlled single time aspiration under complete aseptic techniques followed by tight bandaging. One patient developed an extradural hematoma necessitating its evacuation and one patient had wound infection that was treated by antibiotic and daily wound dressing.

The mean visual analogue scale (VAS) for cosmetic outcome of the different groups, as perceived by the parents against the VAS of the child’s preoperative cosmetic status, is summarized in table 2.

**Table 1: Clinical picture and complication of patients**

<table>
<thead>
<tr>
<th>Type of synostosis and Number</th>
<th>Patient age (month)</th>
<th>Number</th>
<th>Age (month)</th>
<th>Increased ICT</th>
<th>Orbital advancement</th>
<th>Operative time (minutes)</th>
<th>Blood loss (%EBV)</th>
<th>Complications</th>
<th>Visible defects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non Syndromic Sagittal N = 12</td>
<td>&lt; 6</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>120</td>
<td>49±1.4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>6-12</td>
<td>5</td>
<td>1</td>
<td></td>
<td>133±11</td>
<td>33.8±4.3</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>&gt;12</td>
<td>7</td>
<td>4</td>
<td></td>
<td>142±12</td>
<td>29.4±6.3</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Non Syndromic Metopic N = 6</td>
<td>&lt; 6</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>150</td>
<td>46</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>6-12</td>
<td>2</td>
<td>1</td>
<td>2</td>
<td>205±7</td>
<td>52±4.2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>&gt;12</td>
<td>1</td>
<td>1</td>
<td></td>
<td>180</td>
<td>36</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Non Syndromic Bilateral coronal N = 1</td>
<td>6-12</td>
<td>1</td>
<td>240</td>
<td>52</td>
<td>Subgial CSF leak</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Apert Syndrome N = 2</td>
<td>&lt; 6</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>210±42</td>
<td>40±21.2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Crouzon Syndrome N = 2</td>
<td>&lt; 6</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>205±15</td>
<td>36±8.5</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Table 2: Visual analogue scale of the preoperative and postoperative cosmetic status**

<table>
<thead>
<tr>
<th>Type</th>
<th>Non Syndromic Sagittal N = 12</th>
<th>Non Syndromic Metopic N = 6</th>
<th>Non Syndromic Bilateral coronal N = 1</th>
<th>Apert Syndrome N = 2</th>
<th>Crouzon Syndrome N = 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Remodeling</td>
<td>Preop. VAS</td>
<td>Postop. VAS</td>
<td>Preop. VAS</td>
<td>Postop. VAS</td>
<td>Preop. VAS</td>
</tr>
<tr>
<td>Orbito-Cranial</td>
<td>-</td>
<td>-</td>
<td>4</td>
<td>9</td>
<td>-</td>
</tr>
<tr>
<td>Cranial</td>
<td>4.5±1</td>
<td>7.9±0.8</td>
<td>4±0.8</td>
<td>7.5±0.6</td>
<td>3</td>
</tr>
</tbody>
</table>

**DISCUSSION**

Surgery remains the main therapeutic modality for craniosynostosis, aiming to provide an adequate intracranial volume, to accommodate brain growth, to minimize possible cognitive impairment and to create an aesthetically normal skull shape.9, 15, 31

The optimal timing for the surgical intervention in craniosynostosis is still controversial. As the majority of brain growth takes place during the first year of life, the deformation is expected to progress with age. Early surgery permits normal brain growth that will help in molding the skull shape. A general agreement among surgeons is to operate at the earliest opportunity, if there is any evidence of increased intracranial pressure (e.g. bulging fontanelles, progressive optic atrophy, seizures, or multiple-suture synostosis).4,6, 9, 21 In the present study surgery was scheduled upon the patients presentation unless there were general contraindications, where surgery was postponed beyond six month of age upon the anesthesiologist recommendation, unless there were signs of raised intracranial tension necessitating early
intervention, which was the case in seven patients (30%) presenting below six month of age.

Performing surgery in early infancy reverts the abnormalities in the cranial base which results in abnormal facial growth and asymmetry of the maxilla and the mandible that are expected if surgery is delayed. Positron emission tomography scans have demonstrated that, the raised intracranial pressure (ICT) and the reduced cortical blood supply underlying the prematurely fused suture were normalized following surgery.  

Early operative techniques included linear suture cranietomy and cranial vault fragmentation that were performed mainly to correct the functional abnormalities. Unfortunately, these procedures were accompanied by a high rate of reossification and gave only modest results. Surgical results were improved with the introduction of calvarial remodeling with or without orbital advancement techniques. Criticism of these more complicated procedures is based on the presumption of increased operative risk, prolonged hospitalization and the risks associated with significant blood transfusion requirement (placing patients at risk for pathogen transmission, development of transfusion reactions, and other short- and long-term sequelae).  

Currently, surgical correction of craniosynostosis can be divided into 3 main surgical procedures: (1) Surgical procedure involving suture release, cranial vault decompression and upper orbital reshaping and advancement in infancy (6-12 months); (2) Surgical operations to correct midface deformities in childhood (6-12 years); and (3) Orthognathic surgery in adolescence (14-18 years). The utilization of one or more of the aforementioned surgical procedures is determined by the functional and the psychological needs of the patient. The current study population had undergone aggressive cranial and cranio-orbital remodeling, with over correction of the deformity to compensate for postoperative correction reduction. Midface deformities and orthognathic procedures if needed were managed by our maxillofacial college in the years to follow. 

As previously mentioned, intraoperative blood loss remains the most significant concern during open reconstruction for craniosynostosis. With a low circulating blood volume in infants, even a relatively small amount of blood loss can represent a large proportion of the total blood volume, which might lead to a potentially life-threatening hypotension and cardiac arrest. Intraoperative correction of the circulatory volume is the corner stone of successful procedure, which could be attained by proper preoperative planning that, would reduce both operative time and intraoperative blood loss.  

There is a great variability of the reported blood loss following open craniofacial repair, which had been cited between 25% and 500% EBV. In this series total cranial remodeling or orbital-cranial remodeling was scheduled upon presentation, with the operative goal to overcorrect the deformity to an extent that minimizes the risk of long-term relapse. 

In the present study the mean of blood loss of the different procedures was 37% EBV. Orbital advancement was associated with an increase of both operative time and blood loss, and although it was planned for seven cases, this step was canceled in three patients due to intraoperative hypovolemic shock. These three patients were younger than six months of age. Orbital advancement does provide a better cosmetic outcome, but to minimize the operative risks associated with a protracted operative time and blood loss, it should be performed beyond the sixth month of age (unless there are signs of raised ICT); while to minimize the progress of the deformity it should not be delayed beyond the ninth month of age. 

Although several authors consider that the supraorbital bar should be advanced and either widened to correct hypertelorism as in metopic stenosis; or narrowed to correct hypertelorism as in Apert and Crouzon syndromes; Fearon et al. demonstrated that intercanthal and interorbital distances increase significantly even if the orbits themselves are not surgically corrected, which was observed in two cases of trigonocephaly without orbital advancement in the present study. 

Calvarial remodeling techniques provide excellent immediate postoperative cosmetic results, but the long term outcome is unpredictable; as not infrequently, the patient’s forehead demonstrates an uneven surface. These irregularities and persistence of the skull base deformities are commonly encountered if surgery was performed closer to or beyond 1 year of age, as recorded by Jimenez and Barone. 

Similarly, Paige et al published that bone regeneration after subtotal calvarectomy is an age dependent process, where those under the age of 9 months are likely to close the iatrogenically created bony defect. This is supported in the current study, as cranial defects and visible irregularities had a higher incidence in children operated beyond the first year than those operated at a younger age, and this is explained by the reduced bone-regeneration capacity to fill large defects beyond the first year. This observation confirms earlier reports in the literature. In later cases of this study, hydroxyl apatite was used to fill the residual bone defect after the desired remodeling, to achieve a harmonious contour in those operated upon above one year of age. 

In this study, stainless steel wires and titanium plates were used as anchoring devices, for their availability and affordability. Migration of these metallic wires into the bone, or even deeper through the dura into the brain, has been documented. They may...
become subcutaneously visible, palpable and occasionally painful and tender. Fortunately, none of these complications were encountered; however, it is advisable to use absorbable plates and screws whenever possible. 12,13,14

There was an overall improvement of the parent’s perception of the cosmetic end results whether in syndromic or non syndromic craniosynostosis. The results seemed to be better in syndromic cases where cranio-orbital remodeling was performed. Although advancement has been canceled in three patients, the cosmetic results in these patients were found acceptable by the parents who refused further surgeries.

Rottgers et al. advocate thorough evaluations and surveillance of patients with sagittal synostosis, above 1 year of age, due to the increased propensity of them having raised ICT; this was substantiated by the current study, where of twelve patients with sagittal synostosis, six patients had signs of raised ICT; of these patients four were older than twelve month. Anderson et al. proposed that surgical treatment of sagittal synostosis should be directed towards cranial vault remodeling, rather than suture excision, as they found that sagittal synostosis had an increased intracranial volume and tension, when compared to the normal population. 5,13

On the rise are multiple endoscopic procedures, which are claimed to be safe, with markedly decreased operating time, transfusion risk, and hospital stay. But these techniques need to be performed at the age of 3–5 months to achieve a successful outcome. These procedures are hampered by the costs and patients compliance with the mandatory postoperative molding helmet therapy. 5,11,13 These procedures were not performed in this study due to our lack of the needed experience and equipment for performing them.

The small sample size of the present study and the variability of the synostotic types, made it difficult to interpret the surgical variables and outcome from a statistical point of view.

CONCLUSION
Performing aggressive cranial or cranio-orbital remodeling provides the best cosmetic results, but as it is associated with an increased operative time and blood loss, it should be performed after sixth month of age to reduce any anticipated morbidity; yet these procedures should not be delayed beyond the first year of life if a good cosmetic outcome is desired. The role of the anesthesiologist, and his ability to compensate for circulatory volume, is the main determinant of the extent of completeness of the surgical procedure.

REFERENCES