ORIGINAL ARTICLE

# Fronto-Orbital Advancement and Total Calvarial Remodelling for Craniosynostosis

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## ABSTRACT

**Objective:** To describe the results of fronto-orbital advancement and remodelling for craniosynostosis in children.
**Study Design:** Case series.
**Place and Duration of Study:** Department of Plastic Surgery, Combined Military Hospital, Rawalpindi, from June 2009 to June 2012.
**Methodology:** All the patients with cranial suture synostosis operated were included in the study. Those patients who were lost to follow-up were excluded. Variables considered were age, gender, type of synostosis, intracranial pressure, and history of previous surgeries for the same problem. Outcome measures were studied in terms of improvement of skull measurements (anteroposterior and bicoronal), duration of surgery, hospital stay, blood transfusions, complications and parents satisfaction.
**Results:** A total of 36 patients were included in the study. Male to female ratio was 3:1. The age ranged from 5 to 54 months. Thirty two patients presented with non-syndromic and four with syndromic craniosynostosis. Fronto orbital advancement and total calvarial remodelling was done in 26 and 10 patients respectively. There was improvement in the skull measurements and the parents were satisfied in all cases with the skull shape. Complications occurred in 11.1% including chest and wound infection and one death.
**Conclusion:** Fronto-orbital advancement and remodelling is an effective procedure for the correction of craniosynostosis, however, individual cases may require other procedures like total calvarial remodelling.

**Key Words:** *Craniosynostosis. Fronto-orbital advancement and remodelling. Total calvarial remodelling.*

## INTRODUCTION

The era of modern craniofacial surgery dates back to 1970's when Tessier, the father of modern craniofacial surgery, introduced his dynamic and versatile surgical approaches.[1] Craniofacial surgery is thought to be one of the youngest and most challenging fields of plastic surgery.[2] Ever since its rebirth, it is undergoing a rapid evolution. It encompasses wide range of anomalies from the common cleft lip and palate to rare craniofacial syndromes and different varieties of craniosynostosis, rare and complicated craniofacial clefts and craniopagus twins.[3] Correction of these deformities often requires correction of overlying soft tissue as well as sculpturing the underlying skeleton which can provide an infrastructure for future normal growth and development of the craniofacial features.[4] This requires a dedicated and specifically trained multidisciplinary team and highly sophisticated instruments.[5]

Soft tissue corrections and minimal bony work including strip craniectomies are being done by plastic surgeons and neurosurgeons since a long-time in the country.[6] Local data in this regard is lacking, so the aim of this study was to describe the different aspects of fronto-orbital advancement and total calvarial remodelling for craniosynostosis in the setup.

## METHODOLOGY

A retrospective review of patients presented to the Department of Plastic and Reconstructive Surgery, Combined Military Hospital, Rawalpindi, from June 2009 to June 2012, with craniofacial anomalies was done. All the patients who underwent bony correction and remodelling of their craniofacial deformities were included in the study. Exclusion criterion were patients who received only soft tissue correction or were lost to follow-up. A total of 36 such cases were operated upon. The clinical data including the patient's detailed history, clinical findings, investigations and follow-up notes and pictures were retrieved. Each patient underwent detailed history and clinical examination on their first visit. Clinical examination included noting any visible deformity of head, palpation of sutures and fontanelle, scars of

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previous surgery if any, interorbital distance and nasal deformities. Detailed systemic examination was done in all patients to rule out other associated anomalies. Three standard head measurements, which included circumference, antero-posterior and bi-parietal distance, were taken in all cases. Ophthalmological and neurosurgical consultations were also done. Radiological investigations performed were antero-posterior and lateral X-rays of head, and CT scan of craniofacial region with 3-D reconstruction. “Copper beaten” appearance of skull on plain radiographs was assessed as an indication of raised intra-cranial pressure. Echo cardiography and cardiac consultations were obtained where indicated. Blood complete picture, clotting profile and pre-anaesthesia evaluation were done about a week before surgery.

Patients were admitted at least two days prior to surgery. Detailed counselling of parents was done by the senior author and high-risk consent taken. Each patient received at least 4 doses of prophylactic antibiotics, one at the time of induction and 3 in the postoperative period. In the first 4 cases, only two peripheral intravenous lines were used but afterwards arterial lines were also passed either through the radial or posterior tibial artery in all cases. To keep the children warm and prevent hypothermia in these lengthy surgeries, all patients were wrapped intra-operatively in a thick layer of cotton wool exposing only the head and neck area. In addition to that, warm air blowers were also used in winter season. Temporary tarsorrhaphy sutures were done in all cases to protect the cornea in these long surgeries. These were removed at the end of surgery. In those cases in which surgery had to be performed in prone position, special measures were undertaken to avoid pressure on eye balls and facial skin. They included the use of horse shoe head rest, extra padding on pressure prone areas and regular head lifts.

The approach used was 'Zig Zag Bicoronal' incision except in one patient who underwent secondary surgery in our hospital for resynostosis. In this patient, previous straight bicoronal incision had been utilized for exposure at some other hospital. Barrel stave osteotomies were done using high speed drill in bone panels to allow room for brain expansion. The bony fixation was done using dental wire of 0.35 mm and titanium mini-fixation plates. Two non-suction drains were placed beneath the scalp for drainage of blood or cerebrospinal fluid in all the cases. They were removed once the collection was less than 30 ml. All cases were planned and performed in collaboration with our neurosurgical colleagues.

Data was collected in terms of patient's age, gender, diagnosis, symptoms, and duration of surgery, blood transfusion, and stay in intensive care, hospital stay and complications. Complications were divided into early and late depending on whether they occurred within 30 days of surgery or later. Follow-up visits were done at 1, 3, 6 and 12 months and then 2 yearly. On follow-up visits, parents were asked about improvement in symptoms and measurements were done to assess the growth of skull. A 3-D CT scan was repeated for all the patients at the end of one year of surgery to assess the correction and any evidence of re-synostosis.

Analysis was made from the data by using Statistical Package for Social Sciences (SPSS) version 16.0. Continuous variables were expressed as mean ± standard deviation, whereas frequencies were shown for nominal variables.

## RESULTS

A total of 36 (n = 36) patients were included in the study. Male to female ratio was 2:1. There was no family history of craniofacial deformities in these children. The age range was 3 – 54 months (Table I). The median age of patients at the time of surgery with craniosynostosis was 5 months (mean = 6.67 months). Thirty two patients presented with non-syndromic craniosynostosis, 4 patients had syndromic craniosynostosis. Out of 32 patients with non-syndromic craniosynostosis, 7 patients had unicoronal, 8 had metopic, 10 had bicoronal and 6 had sagittal and one patient had unilateral lambdoid craniosynostosis (Table I). Four patients with syndromic craniosynostosis had Apert's (n = 2), one had Crouzon's (n = 1) and one had unspecified syndrome. The patients with Apert syndrome had bilateral coronal synostosis. The patient with Crouzon's syndrome was suffering from multiple suture synostosis of his cranial vault. He had already undergone surgery at some neurosurgical unit at the age of 1 year. Details of previous surgery were not available.

Fronto-orbital advancement and remodelling was done in 26 cases of craniosynostosis. The advancement ranged from 9.7 to 14.5 mm. Total calvarial remodelling (TCR) was performed in 10 patients, 4 with multiple suture involvement and 6 with sagital synostosis. Calvarial bone grafts were used in 7 cases of unicoronal synostosis to augment the recessed frontal bone.

Most of the patients were brought by their parents with complaints of deformity of face or head. Patients with Apert's syndrome presented with craniofacial and hand anomalies. The patient with Crouzon's syndrome presented with difficulty in learning at school in addition to craniofacial deformity. Parents reported that their children were becoming more active and alert, their eye contact improved and children with walking difficulty showed better locomotor control after surgery. All patients were operated for the first time for their craniofacial problem except the one with the Crouzon's syndrome.

The radiological findings of 'copper beaten' appearance of skull, were found in 21 patients on pre-operative assessment, however, fundoscopy was normal in all these cases. The total hospital stay of the patients was

**Table I:** Detail of individual cases.

| S.No | Age (months) | Gender | Suture involved | Type of procedure |
|---|---|---|---|---|
| 1 | 12 | M | Metopic | FOAR |
| 2 | 7 | F | Right UCS | FOAR |
| 3 | 10 | M | BCS+ULS | FOAR |
| 4 | 54 | M | Multiple | TCR |
| 5 | 7 | M | BCS | FOAR |
| 6 | 12 | M | BCS | FOAR |
| 7 | 10 | M | Metopic | FOAR |
| 8 | 10 | M | Right UCS | FOAR |
| 9 | 6 | M | BCS | FOAR |
| 10 | 6 | F | BCS | FOAR |
| 11 | 5 | M | BCS | FOAR |
| 12 | 6 | F | Right UCS | FOAR |
| 13 | 4 | M | SS | TCR |
| 14 | 3 | M | SS | TCR |
| 15 | 3 | F | Left UCS | FOAR |
| 16 | 4 | F | Left UCS | FOAR |
| 17 | 5 | F | BCS | FOAR |
| 18 | 5 | F | BCS | FOAR |
| 19 | 4 | F | BCS | FOAR |
| 20 | 4 | M | Metopic | FOAR |
| 21 | 3 | M | Metopic | FOAR |
| 22 | 3 | M | SS | TCR |
| 23 | 3 | M | SS | TCR |
| 24 | 3 | M | SS | TCR |
| 25 | 3 | F | Metopic | FOAR |
| 26 | 4 | M | BCS | FOAR |
| 27 | 4 | M | Metopic | FOAR |
| 28 | 4 | F | Metopic | FOAR |
| 29 | 4 | M | Metopic | FOAR |
| 30 | 5 | M | Multiple | TCR |
| 31 | 5 | F | Multiple | TCR |
| 32 | 5 | M | Bilateral lambdoid | TCR |
| 33 | 4 | F | BCS | FOAR |
| 34 | 5 | M | SS | TCR |
| 35 | 5 | M | Right UCS | FOAR |
| 36 | 3 | M | Right UCS | FOAR |

*FOAR = Fronto orbital advancement and remodeling; TCR = Total calvarial remodeling; M = Male; F = Female; SS = Sagital synostosis; UCS = Unicoronal synostosis; BCS = Bicoronal synostosis; ULS = Unilateral lambdoid synostosis.*

from 5 to 7 days (mean = 6.11 ± 0.747 days). All the patients were kept in intensive care for the first few days, on average it was 2.1 days (minimum = 2 days, maximum = 3 days). The average operating time was 7.31 ± 1.102 hours (minimum = 5 hours, maximum = 10 hours, SD ±1.102). The amount of blood transfused during one operation was on average 290 ± 71.43 ml (minimum = 200 ml, maximum = 500 ml).

Dural tears were commonly encountered during these surgeries. Most of these were small (1 - 2 mm) which were sealed with fibrin glue (Beriplast). Two patients had larger tears (5 - 10 mm) which were repaired with absorbable sutures and then sealed with fibrin glue. One patient had CSF leak in postoperative period due to unidentified dural tear.

Early complications were recorded in 4 patients which were chest and local wound infection and cerebrospinal

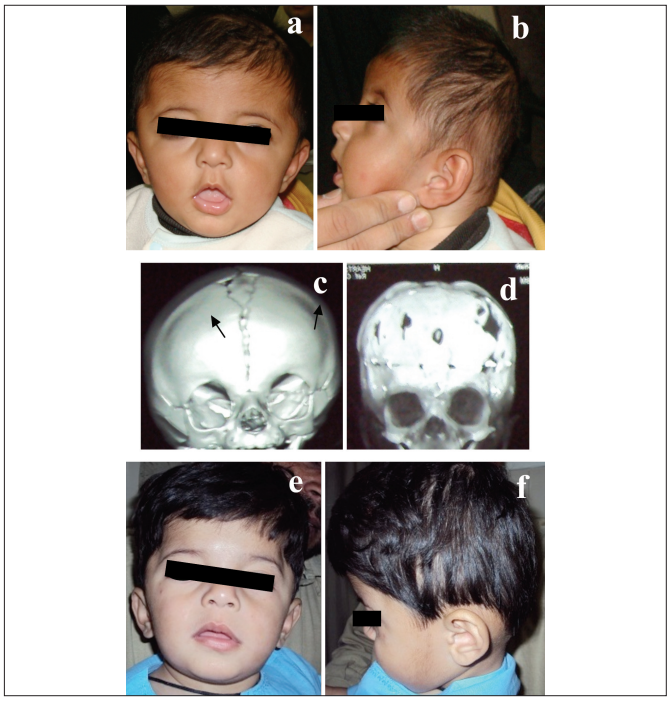


**Figure 1 (a,b,c,d,e,f):** Six months old child with Brachycephaly **(a, b)** Pre-operative picture, anterior and lateral views, **(c)** pre-operative 3-D CT scan of skull, black arrows showing fused coronal sutures, **(d)** one year follow-up 3-D CT scan **(e, f)** anterior and lateral views after one year of surgery.

fluid leak. They responded well to the intravenous antibiotics, chest physiotherapy and local wound care. Low to high grade fever was recorded in all patients, which settled after 24 hours. There was no case of intracerebral hematoma or brain injury requiring re-exploration. One patient had postoperative fits on the first postoperative day which were managed successfully by inj. Valproate. The late complication occurred in only one patient who had gross wound infection which required wound wash and debridements. This child ultimately recovered but had a large area of bone resorption in the area of frontal region. Unfortunately, one of our patients died per-operatively, most probably due to hypothermia related complications. So overall morbidity in these case series was 11.1% and mortality was 2.7%.

Complete follow-up of all 35 patients is available at one year after surgery, showing good craniofacial shape and improvement in cranial dimensions. 3-D CT scan shows satisfactory bony union with no evidence of re-synostosis.

## DISCUSSION

This study describes an early experience in a relatively new field in Pakistan. Over a period of three years, 41 patients were referred who required either FOAR or total calvarial remodelling. Out of these 41 patients, 36 patients were operated and included in this study. The exact incidence of craniofacial anomalies is not known, however, for craniosynostosis, it is estimated to be 1 in

2500 live births.[7,8] The most common suture involved in craniosynostosis is the sagittal suture.[9] In our series 10 patients had brachycephaly, 7 had plagiocephaly, 8 had trigonocephaly, 6 had scaphocephaly, 1 had bilateral lamdoid synostosis and 4 had multiple suture involvement. As it is a small case series so these numbers may not be the true representatives of the population in Pakistan. A majority of these children either remain undiagnosed or the parents do not seek treatment due to financial reasons, lack of knowledge about the disease or are being managed by some basic procedures by our neurosurgical colleagues.

Fronto-orbital advancement and remodelling was the most common procedure performed. It was done in 26 patients with involvement of a single suture while total calvarial reconstruction (TCR) was carried out in 10 cases of sagital and multiple suture synostosis. Usually the common procedures performed for these patients are strip craniectomy and wide strip craniectomy. These procedures rely on the brain growth for skull expansion rather than the primary correction of the deformed skeleton. These procedures have a higher incidence of re-synostosis and have a little or no effect on future normal development of forehead and face.[10]

Now it is widely accepted that fronto-orbital advancement and remodelling should be performed before the age of 6 months.[11] Operating at this early age has got certain advantages e.g. there is no need of bone grafts as bone gaps have the ability to reossify at this age, the associated facial deformity is limited and the bones are malleable making the operative procedure easier.[12] The median age of these patients at the time of surgery was 5 months.

The failure of cranial cavity to expand in the presence of a synostosed suture can lead to various complications. The most dreaded one is the raised intracranial pressure which can cause blindness, mental deterioration and even death. It is more common in cases of multiple suture synostosis.[13] According to Renier *et al*. 47% children with multiple suture fusions have raised intracranial pressure as compared to 14% children with single suture involvement.[14] The signs of raised intracranial pressure (ICP) were seen in 21 (58%) patients. The radiological changes on X-ray and fundoscopic examination were used for the detection of raised intracranial pressure. Various authors have studied the reliability of different methods being used to detect raised ICP in children with craniosynostosis.[15-17] In general intracranial devices are found to be more reliable although technically more demanding. Presence of papilledema has got upto 98% specificity but its sensitivity is age dependent.[18] Presence of diffuse copper beaten appearance on skull radiograph is related significantly to the raised ICP levels.[16,18]

Blood loss during craniofacial surgery can be devastating for such small children.[19] Every effort is made to minimize the blood loss. On an average, 290 ml of RCC (red cell concentrate) were given to patients in per and postoperative period. This is comparable to other studies.[20] The authors used the stored blood from blood bank to replace the blood loss, although some centres are using autologous transfusions, recombinant erythropoietin in the pre-operative period and transfusion of the drained blood postoperatively.[21,22]

In the majority of cases, 0.35 mm dental wires were used for the approximation of bone panels. Worldover, there is an increasing trend towards the use of biodegradable plates in paediatric craniofacial surgery but are much more costly.[23] The use of dental wires has significantly reduced the total cost of surgery and so far the authors have not come across any complication related to the use of these wires.

Few postoperative complications were encountered besides one death. The death of the child occured near the completion of the surgery when he suddenly went into cardiac arrest and despite of all measures could not be revived. Two patients had chest infection, one had minor wound infection and one had major CSF leak. Patients with chest infection were dealt with physiotherapy, steam inhalation and intravenous antibiotics. Local wound infection responded well to the topical wound care. The patient who had major CSF leak was initially treated with antibiotics and general measures such as head elevation. The CSF leak stopped on the 5th postoperative day and his drains were removed. He presented after 3 weeks of his discharge from the hospital with pus discharge from the wound. Formal debridements were performed. Although we were able to control his infection, he had bone resorption in the fronto-parietal region, for which we are planning to put a bone graft in the defect once the child is 5 years or older.

Chromosomal analysis and gene mapping is one of the new advancement in the field of craniofacial surgery. Detection of mutation in fibroblast growth factor receptors 1, 2 and 3 and transcription factor gene TWIST helps in diagnosing and differentiating non-syndromic and multiple syndromic craniosynostosis.[24] This facility is not available at the study centre, so far.

## CONCLUSION

Fronto-orbital advancement and remodelling is an effective and safe procedure for the correction of craniosynostosis and should be the first choice. However, individual cases may require other procedures like total calvarial remodelling.

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