Role of intraoral distractors in management of cranial synostosis: An initial experience

Nandakishor Sahoo, Indranil Deb Roy, Vishal Gupta1, Ajay Premanand Desai2
Department of Dental Surgery, AFMC, Pune, Maharashtra, Departments of Oral and Maxillofacial Surgery,1AFDC, 2ADC (R and R), New Delhi, India

Address for correspondence:
Dr. Vishal Gupta, Department of Oral and Maxillofacial Surgery, AFDC, New Delhi, India.
E‑mail: drvgupta16@gmail.com

Objective: The objective of the following study is to evaluate the results of strip craniectomy with distraction osteogenesis, using the intraoral distractor devices, as a modality of treatment for craniosynostosis. Materials and Methods: Two cases of cranial synostosis were selected for this study. The cases were operated for strip craniectomy with distraction osteogenesis using a pair of miniaturized intraoral distractor devices. Distraction was carried out after a latency period of 4 days at a rate of 0.5 mm twice a day. Total separation of osteotomized segments achieved was in the range of 25‑28 mm. Results: Both patients were evaluated clinico‑radiologically at 3, 6 and 12 months postoperatively. There was an increase in the occipital frontal circumference with improvement in the contour of the skull. Both the cases showed marked improvement of bowel habits, bladder control and cognitive behavior. Radiologically copper‑beaten appearance reduced considerably suggesting improved intracranial pressure. Conclusion: Combination of distraction osteogenesis with strip craniectomy for the management of craniosynostosis is an effective treatment modality with promising results.

Keywords: Craniosynostosis, distraction osteogenesis, strip craniectomy

INTRODUCTION

Craniosynostosis is a congenital anomaly that occurs either as an isolated entity or in association with other malformations. The incidence of craniosynostosis is about as 1 in 2500 live births. The cranial vault normally enlarges rapidly during fetal development and continues to grow until the age of 8 years. By this age, most calvarial sutures are fused. However in craniosynostosis early fusion of sutures do not allow the neurocranial growth. The nomenclature of the skull deformity is derived from the specific sutureal involvement.

The etiological factors of craniosynostosis are genetic, mechanical, metastatic disturbances, bone, dural pathologies and teratogens. Abnormalities of the cranial base that result in abnormal tensions on dural attachments are also cited as one of the cause for premature fusion. Unfortunately, these conditions often remain undiagnosed until such time the parents notice growth retardation and other clinical features. The clinical manifestations are functional disorders such as optic atrophy, blindness and cerebral atrophy with reduced cerebral function, which are the manifestations of either increased intracranial pressure or abnormal cerebral development.

Only modality of treatment for these conditions is early surgical opening of the involved sutures to permit the brain growth. The various surgical interventions are strip craniectomy, surgical reconstruction of the skull and fiberoptic assisted strip craniectomy with molding head gear. Of late cranial distraction has also been employed in the treatment of craniosynostosis.

In our institution, we have treated two cases of cranial synostosis by strip craniectomy with distraction osteogenesis using the
intraoral miniaturized distractor devices after obtaining prior approval of Institutional Ethical Committee. The results achieved were promising.

**MATERIALS AND METHODS**

Two cases of craniosynostosis were referred to our hospital for management. The details of the cases and their management are as under.

**Case 1**

This was a first case report of a 17-month-old male child, case of syndromic craniosynostosis (Carpenter’s syndrome) with trigonocephaly was referred to us. He was evaluated clinico-radiologically on the clinical examination the child had trigonocephaly, squint, depressed nasal bridge, low set ears, polydactyl [Figure 1], flat feet, hypopigmented hair, torsional testis and delayed milestones. The occipital frontal circumference was 42 cm against the normal value of 47 cm. Biochemical analysis showed raised ornithine and ornithine/citrulline ratio indicating hyperornithinemia-hyperammonemia, homocitrullinuria syndrome, which is an amino acid disorder. Computed tomography (CT) scan was carried out which showed the premature fusion of metopic suture. X-ray skull posterior-anterior (PA) view showed typical beaten copper appearance [Figure 2].

After pre-anesthetic assessment patient was undertaken for surgery under general anesthesia (GA). The sagittal, coronal and metopic sutures were exposed through bicoronal approach [Figure 3].

A 10 mm wide strip was marked over the fused suture area with micro saw under copious normal saline irrigation. A cleavage point was created and a dura protector was inserted to avoid any dural tear during the ostectomy. The strip cranietomy was carried out over the sagittal and coronal suture. The triangular portion of the metopic ridge was also excised [Figure 3]. The ostectomy was completed bilaterally over supraorbital rim joining the strip cranietomy of the coronal suture posteriorly. Though the frontal bone was osteotomized into two segments, but was attached to dura. A pair of modular intraoral distractors were opened around 10 mm and fixed bilaterally across the ostectomy site fixing the parietal and frontal bone [Figure 9]. The vector and distraction was confirmed by activation of the distractor device intraoperatively. The activation port was brought out anteriorly through the bicoronal flap [Figure 10]. Wound was closed in multiple layers. Postoperative care and distraction protocol followed was similar to case 1. A total of 15 mm distraction was achieved. On completion of 4 weeks of consolidation period, the distractor devices were removed under GA [Figure 11].

**RESULTS**

Both patients were evaluated clinically and radiologically at 3, 6 and 12 months intervals.

**Case 1**

**Clinical examination**

There was an increase in the cranial circumference to 51 cm with satisfactory contour. There was marked improvement in the general condition of the patient. Patient had started recognizing the parents and had started standing with support. He showed improvement in cognitive ability. On further 6 and 12 month follow-up patient showed further improvement in general as well cognitive ability.

**Radiological examination**

At 3 months postoperative lateral skull radiograph showed well-defined radiolucent areas between the osteotomized segments. Copper beaten appearance was considerably reduced. Skull contour maintained [Figure 12].

At 6 and 12 months follow-up copper beaten appearance was hardly visible with incomplete calcification at ostectomy and distracted site.

**Case 2**

**Clinical examination**

Cranial circumference was increased to 43 cm. Patient was able to stand up with support with marked improvement in cognitive behavior. There was a significant improvement of nutritional status, bowel habit and bladder control.

**Radiological examination**

Gap was maintained between the osteotomized segments at 3 month follow-up period. Copper beaten appearance was also reduced and skull contour was maintained.
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Figure 1: Preoperative frontal view with polydactyly in syndromic craniosynostosis

Figure 2: Preoperative computed tomography scan 3D view and X-ray skull posterior-anterior view

Figure 3: Intraoperative view showing fused cranial sutures with strip craniectomy over the sagittal and coronal sutures

Figure 4: Intraoperative distractors in situ after activation

Figure 5: Postoperative frontal view after removal of the distractors

Figure 6: Case 2 preoperative view

At 6 and 12 months follow-up copper beaten appearance was not evident. Moreover, it also showed signs of bone formation at ostectomy sites [Figure 13].

Figure 14 is the pre and postoperative photograph of a 3 year old girl operated for craniosynostosis. Her preoperative and postoperative radiographs are shown in Figures 15-17. Figure 18 depicts the 3D reconstruction of the consolidated distracted bone at four years follow up.

**DISCUSSION**

Craniosynostosis is defined as the premature fusion or absence of one or more of the cranial vault sutures and is associated with clinically significant neurologic and morphologic consequences. Growth is arrested perpendicular to the fused suture and a
compensatory overgrowth occurs across the suture that remains open. The result is a characteristic craniofacial dysmorphology and lack of the cranial vault growth that restricts the growing brain.[4]

The timing of surgical intervention is a matter of controversy. Some authors believed that it should be as early as 6 months of age because it takes advantage of rapid brain growth that occurs within 1st year.[5,6] Others believe in delaying until 9th or 12th month because the bones are better developed then and there is less reliance on growth of the operated bones and brain to maintain the initial surgical results. Moreover, it gives time for various associated syndromes to manifest and diagnose.[7] Both the cases referred to us for the management of craniosynostosis were
of 17 and 30 months of age. By the time parents could realize that the child is having a delayed milestones, the child was more than a 1½ years of age.

Various treatment protocols are being followed in various centers. Earlier surgical techniques used were the removal of involved sutures via a “strip craniectomy.” Strip craniectomy is essentially reopening the prematurely fused suture line with excision of some amount of surrounding bone. It would allow unrestricted brain growth and the expanding brain would adequately recontour the bones without the need for formal craniofacial reconstruction.

Although this approach does allow for cerebral decompression, but the dysmorphic skull will not reshape itself, even in the presence of an expanding brain, leading to a residual bony deformity.\textsuperscript{[8,9]}

Cranial distraction has recently been tried in the management of craniosynostosis. Matsumoto \textit{et al.}, Greensmith \textit{et al.}, Imai
et al. have successfully treated sagittal synostosis with distraction technique.\cite{10,12} Gradual distraction not only widens the gap between the osteotomized segments to permit the brain growth but also helps in reshaping the skull without leaving any dead space. However in synostosis cases the pattern of bone healing is unpredictable. The distracted callus may undergo premature calcification and obliteration of gap hence defeating its purpose. Therefore, we decided to combine the strip craniectomy with distraction osteogenesis. This is for the 1st time that we attempted to combine strip craniectomy with distraction by using miniaturized intraoral distractor devices for the management of craniosynostosis. This technique did not strictly adhere to the principles of DO, in which a surgical fracture is created rather than removal of a bony segment. The effort was to create and maintain the adequate gap between the osteotomized segments for a longer duration. Thus, allowing more time for the brain to expand as either by strip craniectomy or by distraction osteogenesis alone it is technically difficult to create and maintain a gap of 25 mm. The other advantages of the combined procedure are:

i. Minimally invasive surgical procedure which is less time consuming
ii. Less blood loss thus avoiding the risk of transfusion
iii. Possibility of dural tear is minimal
iv. Osteotomized segment maintains its vascularity since it is attached to dura
v. No extradural dead space thus minimizing the chances of postoperative infection
vi. With the soft-tissue histiogenesis the osteotomized segment always remains covered by soft tissue flap
vii. There is no requirement of any implant to stabilize the osteotomized segment since it is held in position by the distractors
viii. Due to the callus distraction there is no breach or step deformity in the continuity of the osteotomized segment.

However, the intraoral distractors have a limitation of activation up to 25-30 mm. After strip craniectomy of 10 mm, we preactivated the distractor by 10 mm hence only 15 mm of distraction could be achieved in the second case.

Our first case had CSF leak during distraction, which was controlled by tablet diamox and cessation of the distraction for 72 h. The leak may be due to tear of dura which was not apparent during the craniectomy and osteotomy. While fixing the distractor care was taken not to puncture the dura with a drill bit. Hence we used drill bits with sleeves at 5 mm. 2 mm diameter mini screws of 5 mm length were used to fix the distractors. Due to gradual distraction there was no obvious step deformity over the osteotomized supra orbital ridge area.

The distractors were removed after 4 weeks of consolidation. The callus tissue was soft and was incompletely mineralized. Radiological evaluation at the end of 6 and 12 months showed minimal copper beaten appearance which is suggestive of a reduced intracranial pressure.

**CONCLUSION**

The initial experience of combining strip craniectomy and DO, for the management of craniosynostosis is promising. A long-term follow-up in more cases will be required to validate our results.

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