Experience with craniosynostosis treatment using posterior cranial vault distraction osteogenesis

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ABSTRACT

Background: Craniosynostosis compromises the cranial vault volume, severely impede growth, and may lead to increased intracranial pressure (ICP). Posterior cranial vault (PCV) distraction osteogenesis (DO) offers an excellent treatment opportunity for this condition. This article intends to describe the outcomes of PCV DO. **Materials and Methods:** Nine males and seven female children indicated for PCV DO were included in the study. The single vector distraction devices with quick-disconnect distraction rods, a type of miniaturized hardware, was used in all cases. **Result:** Seven of the 16 patients had a history of one or more prior cranioplasty. All reoperations in this series were performed for the indication of raised ICP including five of the scaphocephaly patients and the syndromic patients. Clinical signs of raised ICP were present in all patients with either measured raised intracranial pressure or those with clinical signs of raised ICP preoperatively. There was substantial decrease in the ICP postoperatively. **Discussion:** The outcomes of this study were encouraging. Placing the distractor stems as flat as possible against the outer layer of the cranial bone seems to be a very important maneuver. This keeps the distractor stem less proud and less likely to sustain future trauma. Removal of the distractor stems keeps the devices further away from the risk of later traumatic dislodgement. Moreover, miniaturized distractors allow precise control of the rate and the amount of distraction.

Keywords: Craniosynostosis, posterior cranial vault distraction osteogenesis, distraction, intracranial pressure

INTRODUCTION

Craniosynostosis, especially in syndromal cases, will result in inadequate cranial vault volume. Craniosynostosis results from the premature fusion of one or more sutures of the skull resulting in a cascade of disturbances in the normal growth of the brain and skull including increased intracranial pressure (ICP).[1] Therefore, the classic presentations of elevated ICP can include headaches, nausea, and vomiting. Softer more subtle signs may manifest as a change in behavior, excessive somnolence, or retarded cognitive development noticed by either astute parents or caregivers.

Methods to treat craniosynostosis including cranioplasty with cranial vault remodeling aim to increase the intracranial volume.[2] Stability of calvarial fragments has been greatly improved by the use of resorbable fixation.[3-5] Increasing the intracranial volume by posterior cranioplasty is inherently unstable. The skin is often tight over the repositioned fragment. When the child sleeps in the supine position, the forces on the repositioned fragment of the cranium tend also to drive the fragment back to its original position. In addition, fusion of the sutures following cranioplasty may require multiple secondary revisional cranioplasties.

If the potentially fusing ends of the calvarial bones are kept apart while progressively expanding the skull, the skull volume may be gradually increased so that subsequent fusion might not be of clinical significance. Springs have been used to provide continuous traction on cranioplasty fragments.[6,7] However, the amount of tension on the fragments is not under the continuous direct control of the clinician so that the method is indirect or hidden by the scalp flaps.

Distraction osteogenesis (DO) is a biological process that promotes bone formation between cut surfaces of bone segments while continuous traction is applied to separate the bony fragments.[8-12] Modern clinical DO of the craniofacial skeleton began once McCarthy applied the concept to mandibular lengthening.[13] This led to an explosion of clinical and research activity in craniomaxillofacial DO over the past decade.[14,15] As in other sites, craniofacial DO involves five distinct periods: osteotomy, latency, distraction, consolidation, and remodeling.[14] The gradual distraction of the osteotomized fragments allows the surgeon unparalleled control in gradually increasing the gap between the cranioplasty fragments, and the consolidation period helps to prevent relapse of the fragments back to their original positions.

Posterior cranial expansion should be considered as an initial procedure particularly for syndromic cases with bilateral coronal synostosis. Operative procedures have been lacking for those cases where the shape of the head is satisfactory but where there is insufficient intracranial volume. Posterior expansion allows for an increase in intracranial volume and with minimal cosmetic detriment as the all too visible frontal areas are left undisturbed.

Recently, White *et al* described their experience with posterior calvarial vault expansion using DO in six patients.[16] While increases in intracranial volume were attained, five of the six patients in their series sustained hardware loosening complications. The current authors reported on volume increases in 10 children choosing to use miniaturized hardware with removable distractor stems. This allowed consolidation with the minimum of hardware that might be traumatized during the postoperative period.[17] This study reports updated experience of the current author with DO of the posterior cranial vault (PCV) in 16 children requiring increased intracranial volume.

MATERIALS AND METHODS

This study was conducted in accordance with the ethical principles of the Helsinki Declaration. A total of 16 patients, 9 males and 7 females with ages between 2.5 months and 7 years, were included in this series. Of the 16 patients, there were 6 with scaphocephaly 1 with Saethre–Chotzen syndrome, 4 with brachycephaly (Muenkes syndrome), and 4 with Apert syndrome. In every patient, 3D CTs, stereolythic skulls, and clinical photographs were obtained preoperatively. CT scans and plain films were taken as required postoperatively at follow-up. The follow-up time was a mean of 1 year, ranging from 6 months to 2 years. The main indication for surgery in this group of patients was raised intracranial pressure (ICP) rather than cosmetic indications. The results were tabulated to allow basic descriptive reporting of the outcomes. No other formal statistical methods were used.

Operative procedure

The operations were carried out through a wavy line or zig-zag coronal incision anterior to the vertex of the skull. The posterior cranium was exposed allowing the planned craniotomies as required. In two of the patients with scaphocephaly, the cranial fragments were elevated as three segments and then were reconstituted in an expanded arrangement to further increase cranial volume using distractors. These segments were fixated into one solid fragment using resorbable plates of 1.5 mm PLGA (Inion CPS® baby, Tampere, Finland). In the remaining eight cases, the PCV was mobilized as one piece. In 15 cases, the cranial bone fragments were totally detached from the underlying dura leaving one case where the cranial bone was not totally detached from the underlying dura.

The single vector distraction devices with quick-disconnect distraction rods (Biomet Microfixation 1.5 mm CMF Quick-Disconnect Distractor®, Biomet Microfixation, Jacksonville, FL) were fixed on either side of the skull osteotomies to the parietal, temporal, or occipital bones to provide a posterior vector of distraction [Figure 1]. The upper part of the devices was fixed to the slightly recontoured calvarial segments, each device being secured with four 1.5-mm titanium self-drilling screws. The rods of the distractors were bent downward toward the bone to keep their profile close to the plane of the cranial bone, to minimize skin distension.

Once the distractor rods were attached, the devices were opened to ensure that the vectors were all complimentary and not interfering with each other. The devices were then backed down with 3 mm of a distraction gap remaining [Figure 1]. The devices were then buried beneath the scalp, and the quick-disconnect rods were attached to the distractor devices so they would emerge through the skin through small stab incisions, distant to the wavy-line skin incisions. All incisions were closed. No drains were used. The distraction protocol was started in all patients within 5 days of distractor placement [Figure 2].

RESULTS

Further expansion of the cranium of 1 mm/day was performed during 2–4 weeks either posteriorly or upward, thus gaining cranial expansion in two to three directions. The cranium was distracted from 20 to 30 mm [Figure 2]. The distractor rods were removed under general anesthesia once the desired distraction distance had been attained. No additional incisions were necessary for this part of the procedure. The distractors themselves were removed at a third anesthetic following the consolidation period [Figure 3]. None of the 16

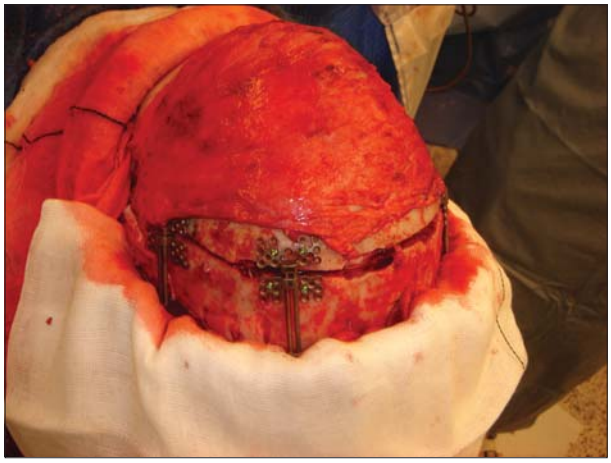

Figure 1: Clinical intraoperative photograph showing distractors in position on cranium with gap of 3 mm at the time of distractor placement surgery

patients showed signs of infection through any of the phases of distraction care.

In all cases, the parents had been taught to apply the distraction forces postoperatively. This allowed early discharge of all patients home from the hospital. None of the 16 parents reported any difficulties with the application of the distraction or the schedule of distraction. They all felt comfortable to be an integral part of their child's treatment.

Seven of the 16 patients had a history with one or more prior cranioplasty. All reoperations in this series were performed for the indication of raised ICP including five of the scaphocephaly patients and the syndromic patients. Clinical signs of raised ICP were present in all patients with either measured raised intracranial pressure or those with clinical signs of raised ICP preoperatively [Figures 4].

DISCUSSION

The distraction goals were met in all the patients. There were no cases of hardware loosening in this series. All patients tolerated their devices right to the planned end of their consolidation phases without any interruptions. There were no infections involving the peri-distractor tissues or the zig-zag scalp incisions.

Immediately following the initiation of distraction, the appearance of the skulls began to show marked improvement in their shapes [Figure 5]. The length of stay of the distraction patients was no longer than for cranioplasty patients at our center, and the parents were able to perform their child's distraction at home. There were no complications such as premature disconnection of the quick-disconnect distraction stems or dropping out of the distraction stems occurred. Distractor stem removal was simple with the quick-disconnect couplings. This occurred at a second short outpatient general anesthetic once the desired amount of distraction had been attained. The remainder of the devices then remained buried in their subcutaneous positions to provide retention during the consolidation phase.

A third short outpatient general anesthetic was necessary for the removal of the titanium distraction devices where a short skin

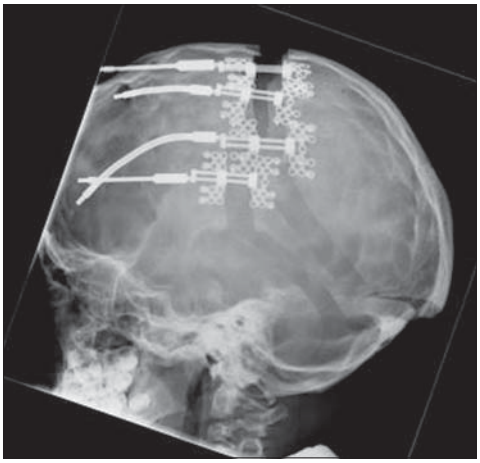

Figure 2: Plain lateral skull radiograph at end of distraction showing posterior displacement of the cranial flap

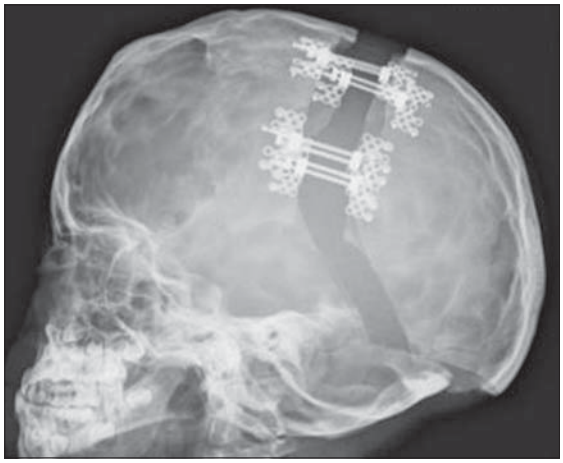

Figure 3: Plain lateral skull radiograph at time of distractor rod removal with distractor left in to provide retention during consolidation phase

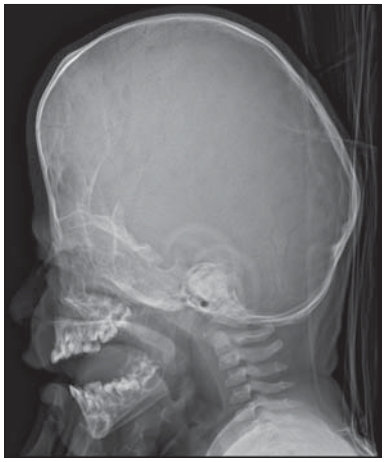

Figure 4: Preoperative radiograph of patient with raised ICP and digital impressions both in anterior cranial fossa and posterior regions

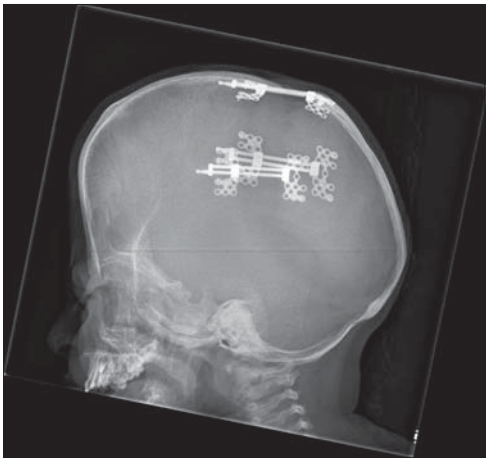

Figure 5: Radiograph following distractor stem removal during consolidation phase showing resolution of digital impressions

incision was used. The distracted gap had ossified with the same appearance as the surrounding cranial bone.

All distracted patients developed normally following the distraction protocol. There were no signs of cognitive delay or behavioral disturbances. Ocular examinations did not show signs of papillary edema nor were there other signs of raised ICP. There were a few differences between this case series and the report of White *et al*.[16] In 9 of the 10 cases, the cranial bone fragments were totally detached from the underlying dura unlike in the White *et al* series. There were no instances of distractor hardware loosening in this group. The current authors believe that placing the distractor stems as flat as possible against the outer layer of the cranial bone is a very important maneuver. This keeps the distractor stem less proud and less likely to sustain future trauma. Removal of the distractor stems keeps the devices further away from the risk of later traumatic dislodgement. None of these devices came loose and all patients were able to keep their hardware up to the planned end of their consolidation phases.

One of the major advantages of a distraction technique using miniaturized hardware versus springs is the inherent control that such hardware provides. Springs by their nature are totally buried beneath soft tissue and their control is indirect at best. Miniaturized distractors allow precise control of the rate of distraction, the amount of distraction, and when to stop distraction.

Posterior calvarial vault expansion using DO is a safe technique. There were no infections despite hardware that was only partly covered by skin. A larger scale trial will help to further establish the safety of this technique and whether two, three, or four distractors are required to provide the desired increase in intracranial volume in a predictable manner. Future studies to correlate the increase in intracranial volume with the extent of cosmetic improvement and showing cosmetic changes over time with serial photography should also be performed.

In the future as resorbable devices become more reliable, hybrid metal and resorbable distractors, or completely resorbable distractors[18] should decrease the number of postoperative anesthetics that are necessary. Such reductions of interventions will make this technique even more patient and family friendly. This preliminary series shows that cranial bone distraction is a useful method for cranial expansion with low morbidity in children with craniosynostosis.

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