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ORIGINAL PAPER

Posterior calvarial vault expansion using distraction osteogenesis

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Abstract
Objectives Management of raised intracranial pressure in syndromic multi-suture craniosynostosis by cranial vault expansion can be achieved by a number of techniques. We present our initial experience in treating this group of patients with posterior calvarial distraction.
Materials and methods Six patients underwent distraction osteogenesis of their posterior calvarial vault.
Results The mean period of distraction was 28 days. The mean consolidation period was 49 days. The mean distance of advancement was 24 mm. Five out of six patients completed their period of distraction and three of these cases also completed their period of consolidation. Significant calvarial expansion and improvement of head shape was achieved in all cases.
Conclusions Posterior calvarial distraction is a safe and more efficient method of calvarial expansion than conventional techniques. These are early promising results, and future modification of the distraction devices will be needed if the effective consolidation time is to be increased.

Keywords Craniofacial surgery · Syndromic craniosynostosis · Posterior fossa surgery · Distraction osteogenesis

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Introduction

Cranial vault expansion is one element in the management of raised intracranial pressure due to craniosynostosis in both syndromic and non-syndromic patients [1]. Calvarial remodelling with fronto-orbital or posterior calvarial advancement are established surgical techniques for expanding the skull vault [2]. It is thought that posterior calvarial movement offers a far greater volumetric increase than traditional fronto-orbital advancement [3]. However, surgery in the posterior fossa can be technically difficult because of the anatomical constraints of the area and fixation techniques. Scalp closure in these cases can be tight and lead to wound healing problems [4]. When young children are placed "back to sleep" (i.e. supine), the postural forces causing posterior relapse are significant and will challenge any fixation technique after open remodelling. The use of distraction osteogenesis for calvarial vault expansion is becoming an accepted alternative to open calvarial vault surgery [5] with potentially less morbidity [6]. This paper describes a preliminary series of six patients with syndromic multi-suture craniosynostosis and raised intracranial pressure managed by posterior calvarial distraction osteogenesis.

Materials and methods

Patient population and outcomes

Data was prospectively collected for six patients who underwent distraction osteogenesis of their posterior calvarial vault for raised intracranial pressure between October 2006 and July 2007. The diagnosis of syndromic multi-suture craniosynostosis and raised intracranial pressure was

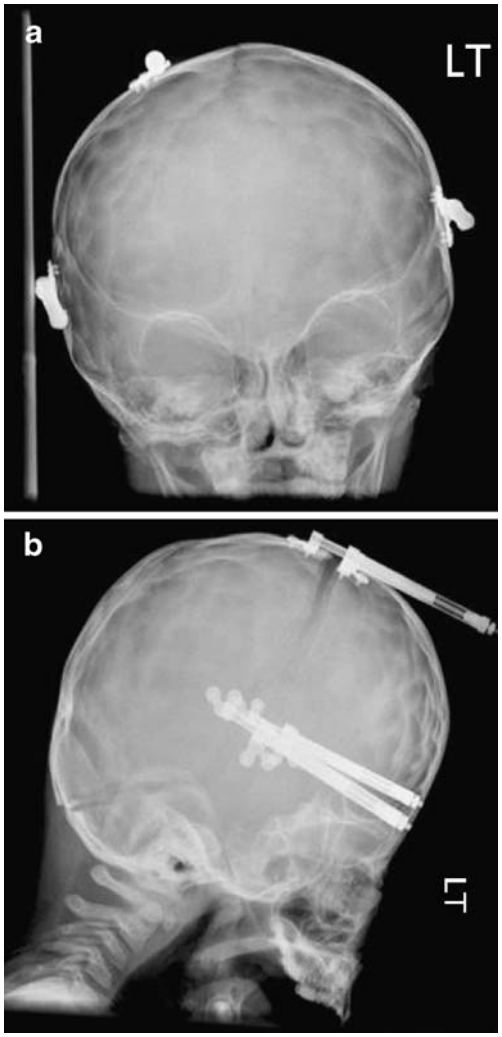

Fig. 1 An anterior/posterior (**a**) and lateral (**b**) skull radiograph illustrating the placement of the osteotomies and distractors

based on clinical examination, radiological investigation and genetic testing. Data was collected for the length of the distraction period, the length of the consolidation period and the distance of the advancement achieved. The advancement distance was calculated from a lateral skull radiograph measuring the distance from the posterior aspect of the anterior footplate to the anterior aspect of the posterior footplate of a distractor on the skull apex; the immediate post-operative distance was subtracted from the distance at the time of distractor removal (using the dimensions of the distractor arm to correlate a true distance). Outcome was assessed by clinical examination and comparison of pre- and post-operative imaging.

Technical aspects

In the prone position, surface scalp markings were drawn before incision to facilitate the vector orientation of the distractors. Three internal 30 mm titanium single vector distractors (Synthes, Pennsylvannia, USA) were used for each case. The zigzag bi-coronal incision was placed anterior to the calvarial osteotomy, which extended from the vertex to below the estimated position of the torcula, within 2 to 3 cm of the foramen magnum. The calvarial segment was not elevated from the dura. The placement of the lower horizontal posterior osteotomy below the torcula and the avoidance of trans-osseous communicating veins decreased the chance of intra-operative bleeding. The distractors were positioned in a horizontal vector parallel to the Frankfort plane. There were two lateral distractors positioned in each temporal region and a superior one placed off centre to avoid the sagittal sinus (Fig. 1). Given the young age of this cohort, the latency period was 3 days and the distraction rate was 1 mm/day with a frequency of 0.5 mm every 12 h. The consolidation period was individualised to each patient. At the end of the consolida-

Table 1 Patient details and outcome

Patient no.	Diagnosis	Age at surgery	Distraction period (days)	Consolidation period (days)	Advancement achieved (mm)	Complication
1	Aperts	1 year 4 months	29 (total), 14 (distraction), 10 (interval), 5 (distraction)	0	18	CSF leak, loosening of footplate
2	Aperts	1 year 2 months	27	19	24	Wound dehiscence, external trauma to distractor
3	Crouzons	1 year 7 months	31	95	30	None
4	Aperts	1 year 2 months	31	115	28	Loosening of footplate, CSF leak
5	Aperts	1 year 5 months	24	7	22	External trauma to distractor
6	Crouzons	9 months	28	7	24	Loosening of footplate

Fig. 2 Improvement in head shape with both posterior and anterior calvarial expansion. **a** Pre-operative, **b** immediately post-operative, **c** after completion of distraction, **d** following removal of distractors

tion period, the distraction devices were removed under general anaesthetic.

Results

Six patients underwent posterior calvarial vault expansion using distraction osteogenesis, each case is discussed individually below and summarised in Table 1. In this series, there were four patients with Aperts syndrome and two with Crouzons syndrome. The mean age at the time of surgery was 1 year 3 months (range 9 months to 1 year 7 months). The indication for calvarial expansion in all cases was raised intracranial pressure. This was confirmed by a combination of ocular disc changes on fundoscopy (four cases), computed tomography scan appearance showing bony erosion in the occiput (three cases) and magnetic resonance imaging (MRI) demonstration of Chiari malformation/tonsillar descent (three cases) with abnormal surface cerebrospinal fluid (CSF) distribution on T2-weighted images (four cases).

Three of the cases required 1 U of blood to be transfused in the perioperative period. Average operating time was 3 h 10 min (range 2 h 30 min to 3 h 45 min). Inpatient hospital stay varied from 4 days to 13 days with a mean of 7 days.

An improved head shape and expanded cranial vault was achieved in all cases with expansion taking place in both the posterior and the anterior calvarium (Fig. 2). Five out of six patients completed their period of distraction and three of these cases also completed their period of consolidation. In all cases, there was radiographic confirmation of ossification (Fig. 3) and, in one case, biopsy taken at the

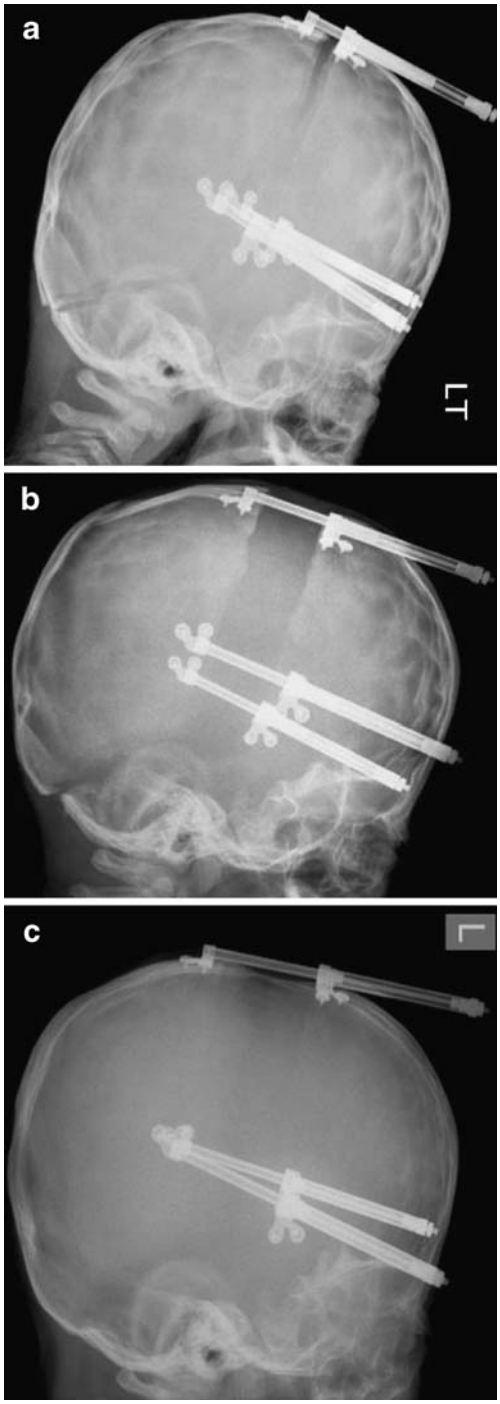

Fig. 3 Example of radiological confirmation of ossification. **a** Post-operative, **b** post-distraction, **c** post-consolidation

time of distractor removal demonstrated neo-ossification at the distraction site. The mean period of distraction was 28 days (range 24 to 31 days). The mean consolidation period for the five patients who completed their distraction was 49 days (range 7 to 115 days). The distraction distance ranged from 18 to 30 mm with a median of 24 mm.

Successful reduction of raised intracranial pressure was achieved in all cases. This was confirmed by fundoscopy and radiological investigation (Figs. 4 and 5).

There were eight complications in total. There were two temporary CSF leaks, two episodes of external trauma to the distractors, the footplates of the distractors loosened in three cases and there was one case of partial dehiscence of the bi-coronal incision. These complications are discussed individually below.

Case 1

A male child with Aperts syndrome who had previously undergone posterior calvarial augmentation at the age of 8 months underwent posterior distraction at 1 year 4 months. A CSF leak developed 14 days after distraction was started; distraction was halted at this point and restarted after another 10 days for a further 5-day period. At this point, there was loosening of a distractor footplate that necessitated removal of the distractors and rigid internal fixation of the distracted skull; therefore, there was no period of consolidation. The patient later underwent fronto-orbital advancement at 2 years 7 months of age.

Case 2

A female child with Aperts syndrome underwent posterior distraction at 1 year 2 months of age. During the distraction period, there was an area of wound dehiscence that was left to heal by secondary intention and a distraction period of 27 days was successfully completed. Nineteen days into the consolidation period, there was external trauma to a distractor, which was loosened at this point, and the distraction devices were removed.

Case 3

A female child with Crouzons syndrome underwent posterior distraction at 1 year 7 months. There were no complications; a distraction period of 31 days and a consolidation period of 95 days were achieved.

Case 4

A male child with Aperts syndrome underwent posterior distraction at 1 year 2 months of age. During the distraction period of 31 days, there was loosening of a footplate. The distractor was repositioned in theatre under general anaesthesia. There was an extended period of consolidation of 115 days as removal of the distraction devices was delayed to coincide with surgery for a cleft palate repair. One week following removal of the distractors, there was a CSF leak which was treated successfully by re-suturing of the bi-coronal incision.

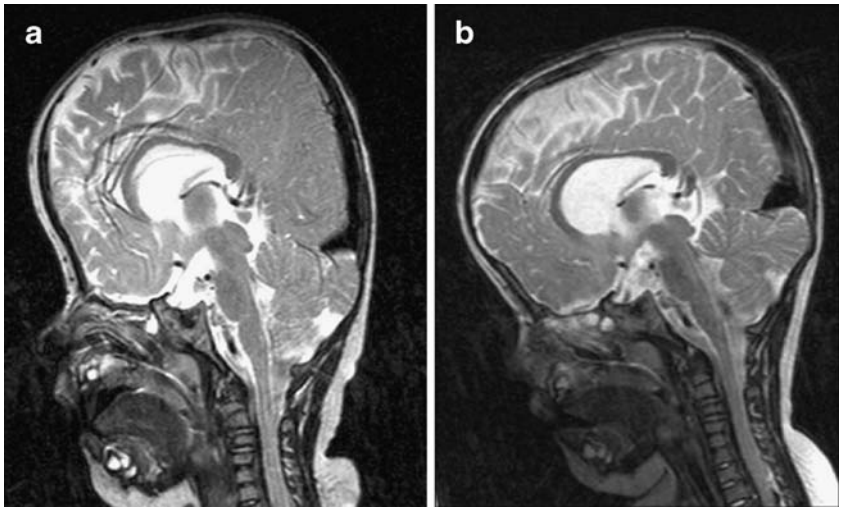

Fig. 4 MRI with a reduction of tonsillar herniation and a more normal CSF distribution. **a** Pre-operative, **b** 3 months post-operative

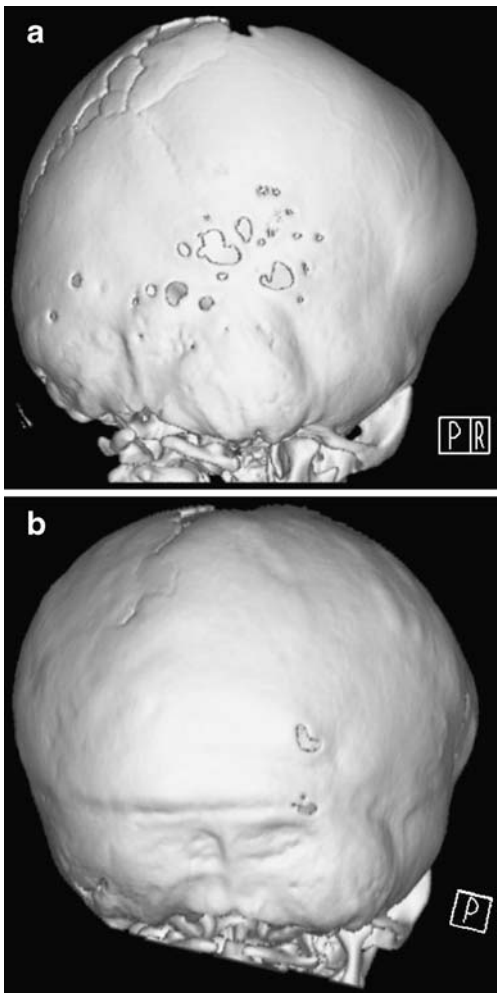

Fig. 5 Reduction of occipital bone erosions following posterior calvarial distraction. **a** Pre-operative, **b** 5 months post-operative

Case 5

A male child with Aperts syndrome underwent posterior distraction at 1 year 5 months of age. Twenty-four days of distraction were successfully completed. Consolidation was stopped after 7 days due to external trauma to a distractor, and the distractors were removed at this point and replaced with rigid internal fixation.

Case 6

A male child with Crouzons syndrome underwent posterior distraction at 9 months of age. Twenty-eight days of distraction were successfully completed. Seven days into the consolidation period, there was loosening of a distractor footplate. The distractors were removed and replaced with rigid internal fixation.

Discussion

Distraction of the calvarium has been demonstrated as a viable technique both experimentally and clinically [7, 8]. Various distraction devices and techniques have been reported that include single vector, multiple vector and spring-assisted vault expansion [9–11]. Distraction osteogenesis for anterior and combined anterior/midface (monobloc) expansion has been widely reported for the management of various craniosynostoses [12, 13]. However, whilst its use for posterior remodelling is at present not established, it does theoretically offer a number of advantages.

As in other areas of the craniofacial skeleton, distraction allows bone transport over distances that would be difficult with conventional techniques. We have found that the aesthetic results are equal to any conventional technique.

Posterior distraction directly targets the region of the brain that needs expansion in the presence of Chiari malformation. The large area of the bone encompassed by the calvarial osteotomies allows a large increase in cranial volume per centimetre of advancement, and the distractors resist relapse secondary to supine positioning of the child.

Interestingly, it has been observed in this series that posterior distraction also has an anterior affect (Fig. 2). Significant anterior fossa expansion was observed during distraction. This is probably a reflection of the calvarial plasticity in babies below 2 years of age as well as the horizontal direction of the distraction vectors. The paediatric skull expands in accordance with Newton's Third Law (every reaction has an equal and opposite reaction).

The technique described does not involve lifting the calvarium off the dura, which results in shorter operating times, reduced blood loss and less morbidity. The expansion is gradual and, therefore, wound closure is not under tension and there is less chance of problems with healing. The use of posterior calvarial expansion by distraction leaves the anterior fossa surgically untouched so that future procedures (such as frontal orbital or monobloc advancement) will not be impeded. There is a significant chance of further surgery to the anterior skull base and midface in this group of patients as the incidence of recurrent raised intracranial pressure in syndromic children with Aperts or Crouzons syndrome is high and mid facial deformity is a feature [14].

There was a significant rate of distractor complications in this series. Direct trauma due to the patients' activity is inevitable, but other published studies of anterior calvarial distraction have had lower distractor loosening rates [15]. The footplate loosening rate of the distractors used in this series is high. This resulted in a shorter than planned period of consolidation in some cases which necessitated replacement of the distractors with internal fixation plates. The use of internal spring distraction of calvarial osteotomies or for open calvarial sutures, rather than internal distraction rods with footplates, may be an alternative method of posterior calvarial vault expansion. Modifications of the described technique that we are currently investigating to decrease footplate slippage include the use of four rather than three distractors positioned as two pairs on either side of the head to provide a more stable construct. The commencement of distraction immediately, without a latency period, also decreases the chances of footplate loosening as once the skull is released there is immediate expansion of the brain which may lift the footplates off the distraction device. We believe that the design for the optimal distractor does require further development.

Posterior calvarial distraction is a safe and more effective method of calvarial expansion than more conventional procedures. These are early results which show promise. Future modification of the distraction devices will enable further progress with this technique.

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