Since the first report by White et al. in 2009,1 posterior calvarial distraction (PCD) has been adopted and popularized in many craniofacial units worldwide, specifically in children with syndromic and multisuture craniosynostosis.2–8 Various groups have proposed PCD to be the primary treatment for children with syndromic and multisuture craniosynostosis because it adequately treats the raised intracranial pressure (ICP) and negates or delays a second anterior cranial procedure.9 As a surgical technique, PCD offers a large volume expansion per dis-
traction distance, confers a lower theoretical risk of nonviable osteotomized bone and extradural blood loss, allows a tension-free scalp closure, and prevents collapse of the expansion when the child is in the supine position.\textsuperscript{5,10,11} It has been proposed that posterior distraction imparts secondary expansion to the frontoorbital region and posterior skull base, due to modification at open cranial base sutures or the growth direction of the cranial vault itself.\textsuperscript{5,12,13}

Cerebellar tonsillar descent, or Chiari malformation type I (CM-I), is often associated with craniosynostosis, especially in syndromic cases, and with multisuture and lambdoid synostosis.\textsuperscript{14} The association of CM-I and syndromic craniosynostosis was first reported by Saldino et al. in 1972.\textsuperscript{15} CM-I occurs in 70% of patients with Crouzon’s syndrome, 75% with oxycephaly, 50% with Pfeiffer’s syndrome, and 100% with the Kleeblattschädel deformity.\textsuperscript{16–18} Cinalli et al. and Goodrich attributed the high prevalence of CM-I in Crouzon’s syndrome to the early closure of the lambdoid and skull base sutures and synchondroses.\textsuperscript{19–21} Thompson et al. demonstrated a correlation between the extent of tonsillar descent and raised ICP.\textsuperscript{22} Rijken et al. showed that children with craniosynostotic syndrome have a smaller foramen magnum.\textsuperscript{23} Craniocephalic mismatch is believed to be a key pathophysiological mechanism for CM-I.

The conventional surgical management of symptomatic CM-I or CM-I with syringomyelia in noncraniosynostotic patients is foramen magnum decompression (or suboccipital craniectomy) with or without C1 laminectomy and duraplasty.\textsuperscript{24,25} The optimal surgical treatment of CM-I with or without syringomyelia in children with syndromic craniosynostosis or with multisuture and lambdoid synostosis (referred to as complex synostosis hereafter) is unclear, and currently there is no consensus on the treatment type and timing.\textsuperscript{26} Cinalli and colleagues advocated posterior calvarial vault expansion/remodeling as the first procedure, but suggested that in selected cases this could be combined with suboccipital decompression in the same sitting.\textsuperscript{16,17} Some authors routinely perform foramen magnum decompression at the same time as posterior calvarial surgery.\textsuperscript{2} Posterior calvarial distraction or expansion has been suggested to be effective in case reports or based on a small number of cases in a larger series whose primary objective was not to assess the CM-I.\textsuperscript{2,3,6,9,27–29}

Following the earlier promising clinical results, since 2006 we have routinely offered PCD as first-line treatment for children with complex craniosynostosis requiring surgical supratentorial volume expansion and a concurrent CM-I (see management algorithm, Fig. 1). For the symptomatic patients, the primary surgical aim was reversal of the signs and symptoms of raised ICP and disease progression, and the secondary aim was to reduce the CM. For the asymptomatic patients, the primary aim was disease stabilization and to prevent the development of symptoms, and the secondary aim was CM reduction. The decision for surgery in asymptomatic patients was based on the current knowledge of the condition’s natural history. It was believed that without surgery these patients would eventually deteriorate. In all patients, where possible, an osteotomy extending inferior to the torcula was performed to increase the posterior fossa (PF) volume. In some cases this carried significant intraoperative risk, and the procedure was converted to osteotomy above the torcula. On follow-up, if patients developed hydrocephalus they underwent ventriculoperitoneal shunt insertion. If they had persistent symptomatic CM, conventional foramen magnum decompression would be considered.

Thus far, there has been no systematic, qualitative, and
quantitative study on the clinical and radiological outcome of PCD. The aim of this study was to examine the safety and efficacy of PCD in the treatment of raised ICP and CM-I, to identify surgical steps or parameters that improved effectiveness, and to evaluate whether PCD avoided further surgery of the anterior cranium and foramen magnum.

Methods

Patient Population

A retrospective review of all children with complex craniosynostosis and concurrent CM-I who were treated with PCD between 2006 and 2015 was performed. They were identified from the prospectively maintained departmental electronic database. The diagnoses of craniosynostosis, CM-I, and raised ICP were based on clinical examination, ophthalmologic assessment, and radiological investigations. CM was defined as a ≥ 5-mm cerebellar tonsillar herniation beyond the foramen magnum or < 5-mm herniation but one that caused significant cranio-cervical junction compression and/or CSF disturbance. Some patients were asymptomatic in terms of raised ICP. However, they were considered for surgery if the progression of disease indicated that they would eventually require treatment. Such a surgical decision was based on the current knowledge of the condition’s natural history. All cases were discussed in the craniofacial multidisciplinary meeting. The surgical team comprised 2 neurosurgeons, 2 maxillofacial surgeons, and 2 plastic surgeons. All children who were offered PCD proceeded with surgery.

Preoperative CT venography was performed in cases in which dilated extracranial venous drainage was anticipated during surgery. Syndromic cases were confirmed by genetic testing. The technical aspects were previously described by our group and are again described here.1,3

Surgical Technique

With the patient under general anesthesia, reinforced endotracheal intubation is performed. The patient is first placed in the supine position and undergoes a complete head shave. Lines are drawn on the parietotemporal areas to mark the planned distractor rod position and orientation (Fig. 2A). A curved wave bicoronal incision line is marked. The patient is then turned prone. The posterior cranial vault is fully exposed in the subgaleal plane initially. At 1 cm superior to the superior nuchal line, the dissection continues deep to the suboccipital muscles directly on bone (Fig. 2B).

Multiple 14-mm burr holes are placed along the planned osteotomy site, including the parasagittal regions at the vertex, both sides of the distractors, both sides of the lateral transverse sinuses, and the paramedian areas across the PF keel approximately 2–3 cm inferior to the transverse sinus. The osteotomy is completed using a craniotome (Fig. 2C and G). If an osteotomy inferior to the torcula and transverse sinus is not possible, due to large extracranial draining veins or complex and grossly abnormal PF anatomy, the osteotomy is modified to end just superior to the torcula (Fig. 2I and J). The resultant bone flap remains adhered to the dura mater.

Following the osteotomy, the distractors are secured to the calvaria via footplates. The orientation of the distractors is in the direction of the desired calvarial expansion. The turning ends of the devices are passed out of the scalp anterior to the incision (Fig. 2D–F). All patients had bilateral distractors (Fig. 2H). None of the patients had a foramen magnum decompression at the same time as PCD.

Following a period of latency of less than 1 week, the distraction was applied by turning the external segment (Fig. 2I). The patients’ parents or caregivers were first trained to do this in the hospital. Following discharge, they continued to do this at home. The target distraction was 1 mm per day, provided there was no complication. The distraction distance was calculated by the difference in inter-footplate distance measured before and after distraction on lateral skull radiographs. The distance was calibrated against the length of the footplate, to eliminate the magnification error on a lateral radiograph (Fig. 2I).

Follow-Up

Following the removal of the distractors, the patients were reviewed in the multidisciplinary clinic with ophthalmology assessment at 6 weeks, 3 months, 6 months, 12 months, and annually thereafter. The patients also had a 12-month postoperative MRI scan and further imaging when indicated.

Morphometric Measurements

The PF anterior-posterior (AP) distance was measured on the midsagittal section on the pre- and postoperative scans, from the tuberculum sellae to the torcula. The PF width was the widest distance on the axial section, often between the bilateral transverse–sigmoid junction of the venous sinuses (Fig. 2K and L).

Statistical Analysis

Statistical analyses were performed using Microsoft Office Excel. Continuous variables are expressed as means. The independent t-test was used to compare continuous variables.

Results

This study included 16 patients (8 boys, 8 girls). Fourteen had pansynostosis and 2 had lambdoid synostosis. All had cerebellar tonsillar herniation. Eight children had a genetically confirmed syndrome: 4 with Crouzon, 2 with Apert, 1 with Pfeiffer, and 1 with Saethre-Chotzen (Table 1). The mean age at surgery was 5.1 years (range 8 months–18 years) (Table 2). The primary indications for surgery were papilledema (n = 7), raised ICP without papilledema (n = 3), and synostosis with CM-I and/or syrinx (n = 6). Clinically, 9 children improved, 7 remained stable, and none deteriorated.

Two children had previous craniofacial surgeries: frontoorbital advancement and remodeling (FOAR) in a different unit, and midface distraction. Two had further craniofacial surgeries, both FOAR, one of which was for volume expansion and the other for cosmetic reasons. Two children required CSF shunt surgeries after PCD. One of

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A: The patient was in the prone position, with the head resting on a padded Mayfield neurosurgical head support. The 2 parallel lines were drawn parallel to the Frankfort plane. They marked a band on which the footplates and the distractor would be positioned. Note the marking anterior to the curved bicoronal incision line, indicating the exit point of the distractor (arrowhead).

B: Posterior view showing the posterior dissection of the bicoronal flap (black asterisk). Initially, the dissection was along the subgaleal plane, leaving the periosteum (white asterisk) intact. Approximately 1 cm superior to the superior nuchal line (white arrowhead), the dissection continued deep to the suboccipital muscle down to bone. In this photograph, an osteotomy below the torcula and a 1-cm strip craniectomy was performed (black arrowhead). This was to avoid impingement of the bone flap against the inferior bone edge during distraction.

C: Lateral view of the completed osteotomy. The periosteum has been divided to allow the osteotomy to be performed.

FIG. 2. (continued)
FIG. 2. D: Following the osteotomy, a distractor was secured via footplates on the right side. The distractors are placed parallel to each other and parallel to the preplanned line, on the flat parts of the calvaria. The distractors were aligned in the direction of the desired calvarial expansion. During placement of the distractor footplates using a drill and screws, the underlying dura is protected with malleable brain retractors. The actual footplate width (FP<sub>a</sub>) is shown here (double-sided arrow). This was for the calculation of distraction distance (see panel I). E: Passing the turning end of the distractor anterior to the bicoronal flap on the left side. F: Closure of the bicoronal flap. Note the protruding end of the distractor (arrowhead). G: Illustration showing the positions of the curved wave bicoronal incision (dashed line), the 14-mm burr holes, osteotomy cuts, and the distractors. Burr holes designated as follows: 1 = parasagittal at the vertex; 2 and 3 = on both sides of distractors; 4 and 5 = on both sides of transverse sinus. Note that the cut between 4 and 5 can often be irregular in order to negotiate across the bony ledge and protect the sinus. Two further paramedian burr holes over the PF are not shown here. H: Distractors (Stryker Modular Internal Distraction System [Stryker Leibinger]). The distractor at the top was turned over to show its underside. Note the screw threads within the shaft. The screw can be turned using the attachable part (arrowhead), which lengthened the distance between the footplates. Note: 3 different distractors from 2 manufacturers were used as our technique evolved and the availability of the distractors changed. The other 2 were Synthes mandibular distractor device and the modular Synthes Cranioaxillofacial Distraction System device (both from DePuy Synthes). I: Lateral plain radiograph of an osteotomy below the torcula, taken at the end of the distraction period. Arrowhead designates the external segment, which was turned to distract. Actual distraction distance (in mm) = [actual footplate width (FP<sub>a</sub>)/measured footplate width on radiograph postdistraction (FP<sub>m-post</sub>)] − [actual footplate width (FP<sub>a</sub>)/measured footplate width on radiograph predistraction (FP<sub>m-pre</sub>)] − measured distraction distance on radiograph postdistraction (DD<sub>m-post</sub>) − measured distraction distance on radiograph predistraction (DD<sub>m-pre</sub>)

Eleven of 16 children underwent osteotomy extending inferior to (below) the torcula, while in 5 it was entirely superior (above) (Figs. 1, 2I, and 2J). The mean latency period for the distraction was 3 days (range 1–6 days), the mean distraction period was 23 days (range 3–27 days), and the mean consolidation period was 112 days (range 1–390 days). The mean distraction distance was 23 mm (range 16–30 mm). One girl developed a CSF leak in the 3rd week of distraction and so the process was abandoned (case 11 in Table 3). Her distraction could not be calculated because she did not have a further lateral skull radiograph. Both patients were included in subsequent analyses. Excluding these 2 patients, the mean distraction period was 25 days. The mean last clinical assessment was 50 months postoperatively (range 9–116 months). The last imaging session was performed at a mean of 33 months postoperatively (range 10–115 months).

Comparing the preoperative and last postoperative imaging, the PF AP distance increased from 73 to 84 mm (p = 0.000002) (Table 4). The PF width increased from 101 to 106 mm (p = 0.009) due to growth. The PF AP distance/width ratio increased from 0.73 to 0.80 (p = 0.0004). The PF AP distance/width ratio increase was higher in those with osteotomy below the torcula compared to those with osteotomy above (12.6 vs 5.4 mm, p = 0.24) (Fig. 3). There

TABLE 1. Characteristics of 16 patients with complex craniosynostosis and cerebellar tonsillar herniation

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Age at Op (yrs)</th>
<th>Genotype</th>
<th>Affected Suture(s)</th>
<th>Raised ICP</th>
<th>Papilledema</th>
<th>ICP Monitoring</th>
<th>Syringomyelia</th>
<th>Osteotomy Below/ Above Torcula</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>0.7</td>
<td>No</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Below</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>4.3</td>
<td>Thalassemia major</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Above</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>2.6</td>
<td>Crouzon</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Below</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>6.9</td>
<td>Crouzon</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Above</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>5.5</td>
<td>No</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Below</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>0.8</td>
<td>Crouzon</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Below</td>
<td></td>
<td></td>
<td>Above</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>2.3</td>
<td>No</td>
<td>Lambdoid</td>
<td></td>
<td>Below</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>3.9</td>
<td>No</td>
<td>Lambdoid</td>
<td></td>
<td>Below</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>18.9</td>
<td>Apert</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Below</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>4.4</td>
<td>Saethre-Chotzen</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Above</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>1.2</td>
<td>Apert</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Below</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>M</td>
<td>3.1</td>
<td>Pfeiffer</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Above</td>
<td></td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>F</td>
<td>6.4</td>
<td>Crouzon</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Below</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>1.4</td>
<td>No</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Below</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>F</td>
<td>1.4</td>
<td>No</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Below</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>M</td>
<td>17.3</td>
<td>No</td>
<td>Pansynostosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Below</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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was no correlation between the distraction distance and the change of PF AP distance/width ratio (Pearson coefficient = 0.39).

In 13 of 16 children, the CM improved. The mean herniation reduced from 9.3 to 6.0 mm (p = 0.011) (Fig. 4). There was no correlation between the PF AP distance/width ratio and change in CM size (Pearson coefficient = 0.16). The reduction of herniation was 3.0 and 3.7 mm for osteotomy below and above the torcula, respectively.

Four patients had syringomyelia on preoperative imaging. All were significant. Three had osteotomy above and 1 had osteotomy below the torcula. On average, the last scan in this subgroup was done 37 months postoperatively.

The AP diameter of the syrinx improved from 7.9 to 3.1 mm, transverse diameter from 10.7 to 4.1 mm, and superior-inferior (SI) distance from 203 to 136 mm. Because the SI distance change could be confounded by growth, it was also measured in vertebral levels, which reduced from 17 to 14 levels. Because of the small number in the cohort, even though the improvement was marked, statistical testing would not be meaningful.

### Complications

The complications are listed in Table 3. There were no complications related to the venous sinuses. Eight of the 9 complications occurred in the first 11 patients and 1 occurred in the last 5 patients.

### Discussion

Since our group described PCD in 2009,1 the technique has become routinely used in many craniofacial units worldwide, specifically in children with syndromic and multisuture craniosynostosis.2-8 PCD is increasingly performed as the primary surgery for children with complex craniosynostosis. In addition to alleviating the raised ICP, it negates or delays a second anterior cranial procedure.9 Compared to anterior surgeries, PCD offers a larger volume expansion per distraction distance.5,11 Compared to other posterior surgeries, it carries a lower risk of non-viable osteotomized bone and extradural blood loss, reduces scalp tension at the time of closure, and prevents collapse of the expansion when the child is in the supine position, which places direct pressure against the occiput.12 By modifying the cranial base sutures and synchondroses, as well as the growth direction of the cranial vault, it may have a secondary effect of frontoorbital expansion and further enlargement of the PF.3,12,13

Cerebellar tonsillar descent, or CM-I, is often associated with craniosynostosis, especially in syndromic cases, or with multisuture and lambdoid synostosis.14 The association was first reported by Saldino et al. in 1972.15 The incidence of CM-I in nonlambdoid single-suture synostosis is less than 10%; in unilateral lambdoid it is 56%; in multisuture with lambdoid involvement it is 57%; and in multisuture without lambdoid it is 11%.30 The incidence of CM-I is 70% in Crouzon’s syndrome and 100% with the Kleebblattschädel deformity.16-18 Cinalli et al. attributed the high prevalence of CM-I in Crouzon’s syndrome to the early closure of the lambdoid and skull base sutures.19 Thompson et al. demonstrated a correlation between the extent of tonsillar descent and raised ICP.22 Rijken et al. showed that children with craniosynostotic syndrome have a smaller foramen magnum.23 This would further compound the normal CSF dynamics at the cranio cervical junction. In children with complex craniosynostosis, the relationships between abnormal calvarial growth, raised

###-table

**TABLE 2. Summary of demographic and clinical characteristics in 16 patients with complex craniosynostosis and cerebellar tonsillar herniation**

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of patients</td>
<td>16</td>
</tr>
<tr>
<td>Mean age at surgery (range)</td>
<td>5.1 yrs (8 mos–18 yrs)</td>
</tr>
<tr>
<td>Sex</td>
<td>8 male, 8 female</td>
</tr>
<tr>
<td>Synostosis by suture, n</td>
<td></td>
</tr>
<tr>
<td>Pansynostosis</td>
<td>14</td>
</tr>
<tr>
<td>Lambdoid</td>
<td>2</td>
</tr>
<tr>
<td>Genotype, n</td>
<td></td>
</tr>
<tr>
<td>Nonsyndromic</td>
<td>8</td>
</tr>
<tr>
<td>Syndromic</td>
<td>4 Crouzon; 2 Apert; 1 Pfeiffer; 1 Saethre-Chotzen</td>
</tr>
<tr>
<td>Primary, n</td>
<td></td>
</tr>
<tr>
<td>Papilledema</td>
<td>7</td>
</tr>
<tr>
<td>Raised ICP w/o papilledema</td>
<td>3</td>
</tr>
<tr>
<td>Synostosis w/ CM-I w/o syrinx</td>
<td>6</td>
</tr>
</tbody>
</table>

**TABLE 3. Summary of surgical complications in 9 patients with complex craniosynostosis and cerebellar tonsillar herniation**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Complication</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Traumatic dislodgement of distractor</td>
<td>Early removal of distractor</td>
</tr>
<tr>
<td>2</td>
<td>Spontaneous loosening of footplate</td>
<td>Extra distractor insertion</td>
</tr>
<tr>
<td>3</td>
<td>CSF leak from 6-mm screw site</td>
<td>Reposition of distractors w/ 4-mm screws</td>
</tr>
<tr>
<td>5</td>
<td>Wound infection</td>
<td>Shorter distraction (14 days) &amp; early removal</td>
</tr>
<tr>
<td>6</td>
<td>Infected pin site w/ collection</td>
<td>Removal of distractor</td>
</tr>
<tr>
<td>9</td>
<td>Lt distractor malfunction</td>
<td>Manual distraction of 9 mm under general anesthesia</td>
</tr>
<tr>
<td>10</td>
<td>CSF leak in 3rd wk</td>
<td>Abandoned distraction, removal of distractors, dural repair</td>
</tr>
<tr>
<td>11</td>
<td>CSF leak 4 days into distraction</td>
<td>Abandoned distraction (3 days)</td>
</tr>
<tr>
<td>14</td>
<td>Rt distractor site wound infection</td>
<td>Conservative</td>
</tr>
</tbody>
</table>
ICP, hydrocephalus, venous hypertension, obstructive and central sleep apnea, and CM are complex. Nonetheless, it is widely believed that craniocephalic mismatch is a key pathophysiological mechanism for CM-I.

The conventional surgical management of symptomatic CM-I or CM-I with syringomyelia in patients without craniosynostosis is foramen magnum decompression (suboccipital craniectomy) with or without C1 laminectomy and duraplasty. The optimal surgical treatment of CM-I with or without syringomyelia in children with complex craniosynostosis is unclear, and according to the Dutch national working group guidelines, there is no clear optimal surgical technique and timing. Cinali et al. advocated posterior calvarial vault expansion/remodeling as the first procedure, but suggested that in selected cases this could be combined with suboccipital decompression in the same sitting. Komuro et al. routinely perform foramen magnum decompression at the same time as posterior calvarial surgery. Posterior calvarial distraction or expansion has been suggested to be effective in case reports, or based on a small number of cases in a larger series whose primary objective was not to assess the CM-I. Thus far, there has been no systematic, qualitative, and quantitative study on the clinical and radiological outcome of PCD for patients with CM-I.

The aim of this study was to examine the safety and efficacy of PCD in the treatment of raised ICP and CM-I, identify surgical steps that improved effectiveness, and evaluate whether PCD avoided further surgery of the anterior cranium and foramen magnum.

In the 2000s, our unit developed the PCD technique primarily to treat raised ICP secondary to supratentorial craniocephalic mismatch in children with complex craniosynostosis. The development of CM-I in syndromic synostosis, or in multisuture and lambdoid synostosis, was attributed to the similar pathophysiological mechanism in the PF (infratentorial) to its supratentorial counterpart. The pathogenic sutures can be supratentorial, lambdoid, or basal sutures and synchondroses. The main difference in the PF compared to the supratentorial compartment was that the brain could herniate through the foramen magnum as an outlet. Therefore, we hypothesized that PCD in which osteotomy also expanded the PF (i.e., extended below the torcula) would improve the CM-I. Our treatment paradigm was as follows (Fig. 1). In patients with multisuture and lambdoid synostosis with raised ICP (evident on clinical grounds, ophthalmological assessment, or radiological imaging), or with significant CM-I or syrinx, PCD was performed as first-line treatment. The primary objective was to normalize the ICP. An osteotomy extending inferior to the torcula was attempted where possible, because it was believed that this would directly increase the PF volume and thus be most effective in reducing the tonsillar herniation, which was the secondary objective. However, this was not achieved in 5 of 16 cases in our cohort. The main reason was a combination of abnormal dilated extracranial veins caused by raised ICP and a difficult surgical approach due to grossly abnormal PF bony anatomy. Following PCD, if the patients continued to have clinical or radiological deterioration related to the CM-I and syrinx, we would consider the conventional foramen magnum decompression.

**The Clinical Objectives Were Met by PCD**

Clinically, 9 of 16 patients improved, 7 remained stable, and none deteriorated. The 7 patients who had a stable outcome were mainly those whose indications were...
not primarily symptomatic, and therefore they could not improve, so to speak. In these patients, surgery was performed based on clinical and radiological progression of diseases and the natural history of the conditions. If they have not had an operation, they were likely to have developed symptoms. In other words, the indication for surgery was not to improve symptoms per se, but to stabilize the disease and prevent the development of symptoms. The study also showed that no patients deteriorated clinically. Therefore, these results showed that the primary surgical aim was achieved and the treatment was effective.

Choice of Morphometric Measurements

The PF AP distance was used to assess the PF size and its change. Because the last scan was performed on average 33 months postoperatively, to eliminate or “correct” the confounding factor of growth, the PF AP distance was divided by the PF width, and the PF AP distance/width ratio was used to assess the effect of the surgery. Any change to the width was due to growth and not distraction because the distraction vector was perpendicular to the width. It follows that any change to the PF AP distance/width ratio was due to the surgery and not growth. Because of the irregular shapes of the PFs studied, a standardized measurement of their heights was not possible; therefore, height was not used for analyses.

A number of nomograms plotting intracranial volumes against age for healthy boys and girls have been constructed. In studying the effect of craniofacial surgery on intracranial volume, some authors have correlated the pre- and postoperative intracranial volumes to these normative data. Some authors have approximated intracranial cavities to ellipsoids and estimated the volume change using formulas based on geometrical shapes. Recently, efforts have been made to produce nomograms for children with syndromic craniosynostosis, not dissimilar to the pioneer work by Kreiborg in the 1980s, and identify predictors or surrogate markers of intracranial volumes in the healthy and craniosynostotic populations. However, to date, there is no ideal and universally agreed-upon methodology to assess the postoperative volumetric change in craniosynostosis. The current study population was heterogeneous in terms of genetic syndrome. Also, for each syndrome, there is significant phenotypical variation. Specific to the PF, many studies of CM-I use the PF/intracranial volume ratio to standardize PF volume measurement. This methodology was not applicable in this study because the supratentorial volume was also changed by the operation. Therefore, in this study the change in PF dimensions was assessed by comparing the intrapatient postoperative to preoperative measurements as discussed above.

Posterior Calvarial Distraction Results in PF Expansion and Reduction in Cerebellar Tonsillar Herniation

The significant increase in PF AP distance/width ratio indicated that PCD led to PF (infratentorial) as well as supratentorial expansion. There was an associated improvement in CM-I; i.e., the secondary surgical objective was fulfilled. However, there was no positive correlation between PF expansion and distraction distance. According to the current study, a distraction distance between 16 and 30 mm was effective, and no conclusion could be drawn regarding distraction outside this range.

Osteotomies Inferior and Superior to the Torcula Are Both Effective

It was initially hypothesized that only those who underwent osteotomy extending below the torcula would have PF expansion. However, a similar effect was also observed in those with osteotomy above the torcula. Such effect was due to the sequential change of the supratentorial compartment and the tentorium cerebelli. Because the posterior calvaria moved posteriorly, consequently the tentorium and the torcula shifted superiorly and posteriorly. There was a trend of more PF expansion in patients with osteotomy below the torcula (Fig. 3), but this was not significant. Interestingly, comparable improvement in CM-I was observed in both subgroups. These observations have two important implications. First, preoperatively, a radiological finding of dilated extracranial collateral draining veins in the occipital bone, including the mastoid emissary vein region, is not a contraindication to PCD as a treatment of CM-I. Second, intraoperatively, if an osteotomy below the torcula is difficult or carries too much risk, an immediate conversion to an osteotomy above the torcula will still be reasonable and effective. A larger sample size is required to elucidate whether the osteotomy below the torcula, which is technically more challenging and carries more risk, confers significantly more PF expansion and CM-I reduction.

Posterior Calvarial Distraction Improved Syringomyelia

Four patients had significant syrinx. Three had osteotomy above and 1 below the torcula. In all 4 cases, the syrinx reduced in the AP, transverse, and SI dimensions. Such findings confirmed that the PF volume expansion and the resultant CM-I reduction also improved the CSF dynamics at the craniocervical junction.

Posterior Calvarial Distraction Prevents Further Surgeries

Of 16 patients, 2 required further anterior cranial surgery, namely FOAR. One (6%) was primarily for volume expansion and 1 (6%) for cosmetic indication. Although the number is small, this series suggested that only 13% required further anterior surgery for all indications. It also suggested that anterior cosmetic surgery can be delayed to a later age when anesthetic and surgical risk is lower and a superior cosmetic outcome can be achieved. Two patients required CSF shunt surgery. Most importantly, none of the patients required foramen magnum decompression at a later stage. This implied that a combined posterior calvarial and foramen magnum surgery was not necessary in the first sitting. Therefore, PCD was an effective first-line, single-stage treatment for CM-I in this group of patients.

Complications

The complication rate was comparable to other published PCD series, including the 50-patient series from our unit (Table 3). Eight of the 9 complications occurred in the first 11 patients and 1 occurred in the last 5 patients.
suggesting a learning curve and that the complication rate has improved with experience. Most complications are distractor related. There was no venous sinus injury. However, the surgical technique described in this study carried an inherently higher hemorrhage risk than previously described PCD surgeries. This was because of the surgical intent to create an osteotomy extending below the torcula. This approach involved 1) dissection around the abnormal dilated extracranial veins, which were more abundant in the suboccipital and retromastoid regions; and 2) performing cuts across the transverse sinuses and the midline keel. Data from 16 patients were not sufficient to draw conclusions regarding the rate of venous sinus injury and significant hemorrhage.

Conclusions
This 10-year series is the first systematic evaluation of PCD for treating children with multisuture and lambdoid synostosis and concurrent CM-I. It shows that PCD is effective in treating supratentorial raised ICP and concurrent CM-I. It avoids a second anterior cranial surgery for volume expansion in 94% of patients and foramen magnum decompression for the CM-I in 100%. PCD negates the need for a combined posterior calvarial and foramen magnum surgery in the same setting. The CSF shunt surgery rate is low (13%) following PCD. A distraction distance between 16 and 30 mm is effective. Osteotomies both above and extending below the torcula lead to PF expansion in 94% of patients and foramen magnum synostosis and concurrent CM-I. It shows that PCD is effective. Volumetric analysis of anterior versus posterior cranial vault expansion in patients with syndromic craniosynostosis. J Craniofac Surg. 2012;23(2):455–458.

References


**Disclosures**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Author Contributions**

Conception and design: Lo, Rodrigues. Acquisition of data: Lo, Thant, Kaderbhai. Analysis and interpretation of data: Lo, Thant, Kaderbhai. Drafting the article: Lo. Critically revising the article: Lo, Kaderbhai, White, Nishikawa, Dover, Evans, Rodrigues. Reviewed submitted version of manuscript: Lo, Kaderbhai, White, Nishikawa, Dover, Evans, Rodrigues. Approved the final version of the manuscript on behalf of all authors: Lo. Statistical analysis: Lo. Study supervision: Rodrigues.

**Supplemental Information**

**Previous Presentations**

This work has been accepted for presentation in the 47th Annual Meeting of the International Society for Pediatric Neurosurgery.

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