Advantages of Calvarial Vault Distraction for the Late Treatment of Cephalocranial Disproportion

Jonathan S. Black, MD,* Jordan Deschamps-Braly, MD,** and Arlen D. Denny, MD, FACS§

Purpose: Cephalocranial disproportion is a symptomatic condition related to a volume discrepancy between the calvarial vault and the brain. Traditional expansion techniques are unfavorable in older children due to inadequate dural ossification, lack of bone pliability, and limited future growth potential. The authors review their experience using distraction to close bone defects in this setting.

Methods: A retrospective analysis was performed of all patients treated with distraction in this setting by a single surgeon. Demographic and outcomes data were collected. The efficacy of ossification of bone defects after expansion by distraction was measured using volume analysis of three-dimensional computed tomography (CT) scans. This required a CT scan at the completion of device activation and a follow-up CT scan 6 months or more beyond activation.

Results: Sixteen patients (17 distractions) met the imaging-based inclusion criteria. The average age at surgery was 3.97 (2.14–6.89) years. The mean initial bone defect volume after asymmetric transverse distraction was 7.26 (5.45–13.73) mL. The mean final defect volume was 2.18 (0.00–5.90) mL with a mean change of 5.08 (1.21–12.79) mL and mean interval time of 27.85 (7.13–56.39) months. This represents a mean percent defect closure of 72.30 (20.38–100.00).

Conclusion: Distraction osteogenesis is a very effective tool for treating the older child with cephalocranial disproportion. The ability to ossify the bone defects without a donor site provides a considerable advantage in these patients.

Key Words: Cephalocranial disproportion, cranial defect, craniosynostosis, distraction osteogenesis

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Cephalocranial disproportion refers to a volume discrepancy between the brain and the intracranial space. The calvarial vault is the major determinant of this space. If the brain is unable to be properly accommodated, increased intracranial pressure results. Elevated pressure may lead to visual loss, cognitive impairment, and behavioral issues in children.1,2 Due to the invasive nature of pressure monitoring, indirect signs of elevated pressure are used to monitor patients at risk. These include chronic headache, behavioral changes, irritability, a “copper-beaten” skull pattern on imaging, and papilledema. This condition has been reported to occur after treatment for hydrocephalus and craniosynostosis.3,4 Craniosynostosis is typically recognized early in life based on changes in head shape resulting from the fused suture(s). Traditionally, calvarial vault remodeling is performed in infancy to expand the volume and improve head shape.1,3 This procedure often leaves large full thickness bone defects and capitalizes on the infant’s ability to spontaneously ossify to achieve vault closure. Symptomatic cephalocranial disproportion, by comparison, has a nonspecific presentation later in childhood. This condition is treated similarly using cranial vault expansion, but results in a higher rate of defects due to the older age.5 Cranioplasty to close defects is also more challenging in children compared with adults due to diminished diploe formation and less donor availability.

Distraction osteogenesis is ideally suited for treatment of this condition. Ossification of the regenerate provides an increased volume of bone without a donor site. The older child provides thicker bone to better mechanically support the distraction process. Considerable volume increases have been documented with distraction directly addressing the space disproportion.6 We present a volumetric analysis of bone defect closure of the calvarial vault using the distraction process without donor material.

METHODS

This study proposal was reviewed and received institutional IRB approval. A retrospective chart review was performed of all patients treated using distraction osteogenesis of the calvarial vault by the senior surgeon. Demographic information including age, sex, diagnosis, age at surgery, age at the most recent follow-up, and length of follow-up time was recorded. Additional data collected included previous treatments, presentation type, signs of elevated intracranial pressure, and craniotomy pattern used. Outcome data collected included operative complications, improvement in signs of elevated pressure, and the need for additional procedures.

Patients were included in the study population if they were diagnosed with cephalocranial disproportion and older than 2 years at the time of surgery with adequate imaging. Adequate imaging was defined as having computed tomography (CT) scans preoperatively, postoperatively following completion of device activation, and at least 6 months following completion of device activation. For the purpose of imaging analysis, CT scans needed to be in a recorded Digital Imaging and Communications in Medicine format, which began in 2006 at our institution.

Three-dimensional CT Digital Imaging and Communications in Medicine data was analyzed using Amira software (Visage Imaging, San Diego, CA) for all included patients. Finite element mesh models were created of the segmented image (cranial defect) to...
achieve accurate volumetric measurement given its irregular dimensions. The cranial defects were segmented from the postoperative CT scans to measure interval change. This included the postoperative CT scan at the completion of device activation and the patient’s most recent CT scan. If the patient underwent calvarial bone grafting after distraction, the CT scan prior to grafting was used. The time interval between imaging sets was recorded. If the interval was less than 6 months, the patient was excluded.

The defect thickness was referenced to the thickness of the adjacent cranial bone. This created a segmented three-dimensional image and its volume was measured (mL) (Amira software). This represented the total defect volume after device activation. This time point corresponds to the junction of the activation and consolidation periods after the expansion process has been completed. Any defects present prior to the distraction process were measured in the same fashion on the preoperative CT scan and subtracted. This accounted for defects from other procedures as some patients were treated in a secondary fashion. Cranial defects were similarly segmented and measured on the patient’s most recent CT scan. The change in volume (mL) was measured and represents the amount of ossification attributed to the process of distraction. The final volume was recorded and related to the initial volume as the percent of defect closure.

RESULTS
A total of 31 patients were treated by calvarial distraction for cephalocranial disproportion. One patient was treated twice in a planned, staged manner. Three were excluded due to inadequate imaging yielding 16 patients (17 distractions) as the study population. The average age at surgery was 3.97 (2.14–6.89) years. The average follow-up time was 3.78 (1.11–7.28) years with an average age at follow-up of 7.35 (4.42–13.39) years.

Thirteen patients underwent distraction using the asymmetric transverse posterior expansion pattern. Nine were diagnosed with sagittal synostosis. One patient with bicoronal synostosis and 1 patient with a trans-sphenoidal encephalocele each underwent fronto-orbital advancement by distraction. Two patients (brothers) had pansynostosis. One underwent occipital distraction and the other underwent asymmetric transverse posterior distraction twice in a planned, staged manner. This was performed using a left followed by right parasagittal pattern.

Seven patients were treated using distraction as their initial (primary) intracranial expansion procedure and the remaining 9 had undergone a previous intracranial expansion procedure (secondary). There were no repeat distractions or subsequent intracranial remodeling procedures performed. There were no complications. Two patients (12%) underwent subsequent calvarial bone grafting to close defects.

The mean initial bone defect volume after asymmetric transverse expansion was 7.26 (5.45–13.73) mL (Table 1). The mean final defect volume was 2.18 (0.00–5.90) mL with a mean change of 5.08 (1.21–12.79) mL and mean interval time of 27.85 (7.13–56.39) months. This represents a mean percent defect closure of 72.30 (20.38–100.00). Representations of the imaging analysis are included in Figures 1–4.

The mean initial defect volume after fronto-orbital advancement was 7.14 (1.53–15.49) mL. The mean final defect volume was 3.45 (2.55–4.34) mL with a mean change of 6.14 (2.90–9.39) mL and mean interval time of 40.54 (16.33–64.74) months. This represents a mean percent defect closure of 60.78 (53.18–68.37) in 2 patients. The initial defect volume after occipital expansion in 1 patient was 4.46 mL. The final defect volume was 0.32 mL with a change of 4.14 mL and interval time of 19.68 months. This represents a percent defect closure of 92.77.

DISCUSSION
Children with cephalocranial disproportion represent a unique group of patients as their symptoms develop after infancy when many features of the infant skull have been lost. It is widely

<table>
<thead>
<tr>
<th>TABLE 1. Bone Defect Volumes (All Patients)</th>
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<td>Asymmetric transverse posterior expansion</td>
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*Shunt-induced synostosis.
FIGURE 1. Patient #12 from Table 1. Lateral (upper left) and top-down (lower left) Amira reconstructions of intracranial volume (blue) and bone defect volume (red) at the completion of device activation. The corresponding three-dimensional computed tomography images are included for reference (upper and lower right). This patient had sagittal synostosis treated secondarily using distraction at the age of 3.04 years.

FIGURE 2. Patient #12 from Table 1. Lateral (upper left) and top-down (lower left) Amira reconstructions of intracranial volume (blue) and bone defect volume (red) 2 years after surgery. The corresponding three-dimensional computed tomography images are included for reference (upper and lower right). This patient had 79.35% volumetric closure of the bony defects resulting from the distraction process.

FIGURE 3. Patient #6 from Table 1. Lateral (upper left) and top-down (lower left) Amira reconstructions of intracranial volume (blue) and bone defect volume (red) at the completion of device activation. The corresponding three-dimensional computed tomography images are included for reference (upper and lower right). Distraction was performed at 6.89 years of age. This patient had sagittal and left lambdoid synostosis treated primarily at the age of 6.89 years.

FIGURE 4. Patient #6 from Table 1. Lateral (upper left) and top-down (lower left) Amira reconstructions of intracranial volume (blue) and bone defect volume (red) 5 years after surgery. The corresponding three-dimensional computed tomography images are included for reference (upper and lower right). This patient had 98.4% volumetric closure of the bony defects resulting from the distraction process.
accepted that the ability to spontaneously ossify cranial bone defects diminishes with age.\textsuperscript{1,6,8}\textsuperscript{1,6,8} Incomplete cranial ossification occurs in as many as 50% of children and early as 9 months age.\textsuperscript{9,10} Secondary reconstruction is employed in up to 18% of patients with helmets often used in the interim.

Split calvarial bone is used as a means of cranioplasty, but is difficult in children less than 5 years age. This is due to the underdeveloped diploe.\textsuperscript{11} Numerous other materials have been utilized with varying success in closure including alloplasts, hydroxyapatite cement,\textsuperscript{12} and bone dust. Alloplasts do not incorporate or grow with the patient and are therefore not typically recommended. Bone dust has been reported to have poor success in defect closure owing to high resorption.\textsuperscript{13–18} Particulate bone is the most promising current autologous material and has been shown superior to bone dust for inlay cranioplasty in the rabbit model.\textsuperscript{13} This resulted in a 99% defect closure at 16 weeks compared with a 41% closure with bone dust and a 38.6% closure without an implant. Despite this success, particulate bone graft requires a donor site with associated morbidity and additional harvest time.

Distraction techniques are uniquely suited to manage the older child with less ability to ossify defects. Distraction provides regenerate bone for closure of expansion defects. In our population, this resulted in a 72.15% average defect closure in all patients with a mean volume of 5.15 mL of bone formed. Similar to the experimental study using bone dust and particulate bone, we found greater closure with increased time. Our patient population demonstrated a greater percentile of defect closure if the time interval to their final CT scan was greater than 20 months (Table 2). This suggests that a greater time than 2 to 3 times the activation period would be beneficial. Additional study to determine the rate of defect closure and time period to the peak of defect closure would be useful to guide the timing of device removal.

All patients treated using distraction as their primary intracranial expansion procedure had a greater than 80.71% defect closure rate. The mean age of this group was 4.14 (2.30–6.89) years. All patients treated in a secondary fashion had less than 79.53% closure of their bone defects. This was despite a similar mean age of 3.86 (2.17–5.15) years. The 1 exception in the secondary group was the single study patient treated with occipital expansion. This patient ossified 92.77% of the bone defect volume. This finding suggests an advantage for patients who have not had prior expansion procedures. Finally, patients younger than 4 years at the time of surgery had greater percentile closure (81.60 versus 64.71), but the results were varied as seen by the ranges (Table 2). Clearly, the process of ossification is complex and warrants further investigation to determine which patient factors are significant.

limitations exist in this analysis beyond that of its retrospective nature. We do not perform traditional vault remodeling procedures on patients beyond infancy at our institution due to the concern for inadequate spontaneous ossification, lack of bone pliability, and inability to capitalize on the rapid brain growth phase. Without this comparison group, we are limited in performing a direct assessment of the efficacy of distraction compared with traditional techniques. The study population lacks the power to compare craniotomy patterns. This is partly due to the rigid inclusion criteria related to imaging analysis. Due to the small population, we were also unable to distinguish which patient factors (eg, age, consolidation length, prior procedures) led to a greater efficacy of distraction in ossifying defects. Due to the small incidence of those with cephalocranial disproportion, we need centers to combine their experience and generate recommendations to improve screening and overall care.

Distraction osteogenesis is a very effective tool for treating the older child with cephalocranial disproportion. The ability to ossify the bone defects incurred by the expansion process provides a considerable advantage in older patients unable to spontaneously ossify large calvarial defects. The presence of de novo bone generation obviated the need for donor site grafting in most patients. This technique should be considered in older children requiring cranial expansion.

\begin{table}[h]
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\begin{tabular}{|c|c|c|}
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\textbf{Diagnosis} & \textbf{Mean Defect Vol Change (mL)} & \textbf{\% Change} \\
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Isolated suture fusion & 5.92 (1.22–12.79) & 87.25 (76.26–100.00) \\
Fusion with additional features & 3.56 (1.99–7.67) & 45.39 (20.38–86.56) \\
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\textbf{Previous expansion procedure} & & \\
No (primary) & 6.68 (2.06–12.79) & 90.95 (80.72–100) \\
Yes (secondary) & 3.48 (1.99–6.78) & 53.64 (20.38–79.54) \\
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\textbf{Time interval} & & \\
Greater than 20 mo & 6.27 (1.22–12.79) & 85.36 (68.37–100.00) \\
Less than 20 mo & 3.55 (1.99–6.77) & 53.27 (20.38–92.77) \\
\hline
\textbf{Age} & & \\
Younger than 4 yr & 4.15 (2.06–6.97) & 81.60 (50.50–100.00) \\
Older than 4 yr & 5.34 (1.22–12.79) & 64.71 (20.38–98.41) \\
\hline
\end{tabular}
\caption{Bone Defect Ossification—Asymmetric Transverse Distraction}
\end{table}

\*Pansynostosis, chromosomal anomalies, shunt-induced.

\begin{thebibliography}{99}
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\end{thebibliography}