posterior vault distraction (PVD) techniques as a way of increasing intracranial volume. To date, the effect of PVD on craniofacial dysmorphology in these patients has not been well studied. This study was designed to examine the effect of PVD on craniofacial morphology in patients with syndromic multi-suture synostosis using radiologic morphometric analysis.

METHODS: With IRB approval, 22 patients (11M, 11F: age 4 to 149 months) with syndromic craniosynostosis (3 Apert, 4 Pfeiffer, 4 Muenke, 3 Crouzon, 2 chromosomal anomaly, 1 craniofrontonasal dysplasia, 5 unknown) underwent PVD for correction of elevated intracranial pressure (n=12) and/or correction of turribrachycephaly (n=16). Morphometric analysis of sequential CT scout radiographs and lateral skull radiographs assessed turricephaly index (TI=cranial length/height), vertical growth index (VGI=cranial base length/height), absolute vertical height (AVH=sella to vertex distance), and occipital inclination angle (OIA).

RESULTS: Preoperatively, patients exhibited a mean TI of 1.34 (range=1.02–1.53, SD=0.16), VGI of 0.41 (range=0.26–0.58, SD=0.08), AVH of 107.37mm (range=87.4–149.4, SD=15.38), and OIA of 44.54 degrees (range=12.5–67.2, SD=12.17). PVD (n=18) demonstrated association with increased TI (mean=1.53,p<0.001) and VGI (mean=0.44,p=0.001), while flattening the OIA (mean=37.5,p=0.004). Additional frontal advancement procedures after PVD (n=15) yielded further increase in TI (mean=1.59,p=0.004) and VGI (mean=0.47,p=0.012). AVH did not change significantly once distraction was initiated.

CONCLUSIONS: PVD results in significant improvements in craniofacial dysmorphology by increasing calvarial length relative to AVH and flattening occipital inclination, effectively normalizing turricephalic proportions. These results are improved by further frontal advancement surgery. AVH does not appear to increase or decrease after PVD initiation, suggesting aesthetic benefits are due to calvarial length attaining proportion to pre-treatment cranial height.

The Use of Distraction Osteogenesis in the Treatment of Rickets-Associated Craniosynostosis

Mitchell A. Stotland, MD, MS; David F. Bauer, MD

INTRODUCTION: There is a known increased incidence of craniosynostosis in patients affected by the metabolic condition of rickets.1–3 Due to abnormal bone development, and a tendency toward presentation at an older age than most craniosynostosis patients, cranial remodeling surgery in rickets patients may be complicated by inadequate post-operative cranial growth and a greater risk of sutural re-fusion. We present two cases of 3-year old boys with rickets-associated sagittal craniosynostosis, and demonstrate the effectiveness of distraction osteogenesis in the surgical management of this condition.

METHODS: Two 3-year-old boys with rickets presented with sagittal synostosis and marked scaphocephalic deformity as an indication for surgical intervention. A single midline sagittal osteotomy extending from the bregma to the lambda was made. A sagittal suturectomy was not performed in order to ensure bone edge proximity across the sagittal osteotomy site at the commencement of the distraction phase. Transverse osteotomies were made bilaterally behind the coronal suture to the pterion, and just anterior to the lambdoid sutures to the asterion to allow for biparietal distraction of the bilateral parietal region. Transverse wedge ostectomies were performed anterior to the lambdoid sutures, and the occipital bone was barrel-staved to encourage anterior occipital movement resulting from scalp tension during the ensuing transverse distraction. The devices were activated for 30 days. Consolidation phase lasted 12 weeks. Breadth of distraction and quality of regenerate were confirmed clinically at the time of device removal. Head shape was documented photographically.

RESULTS: In both cases:

- Distraction breadth was verified intraoperatively as 30mm.
- Clinical exam confirmed that the regenerate was solid bone, without palpable areas of incomplete osteogenesis.
- A significant improvement in cranial proportion was achieved in both patients, as assessed clinically.
- Estimated blood loss was high in both cases, presumably related to the hyperemic nature of rickets bone, rather than the surgical distraction approach employed.

CONCLUSIONS: Distraction osteogenesis promotes bone growth and cranial remodeling in patients with craniosynostosis due to rickets. The technique allows for continuous, incremental expansion of both bone and scalp tissue to overcome the limitations of abnormal bone healing due to rickets, as well as inadequate scalp compliance due to older
We recommend consideration of distraction osteogenesis specifically for the treatment of craniosynostosis in older children with severe deformity, including those with rickets.

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Outcomes of Patients with Unicoronal Craniosynostosis Treated by Endoscopic Strip Craniectomy

Suresh N. Magge, MD; Aparna Sajja, BS; Robert Mccarter, ScD; Robert F. Keating, MD; Gary F. Rogers, MD

BACKGROUND: Unicoronal craniosynostosis (UCS) results in craniofacial deformities including recessed forehead, orbital dystopia, and angulation of the nose and face. This condition traditionally has been treated with bifrontoorbital advancement, but facial and nasal angulation can remain as a significant issue. Endoscopic strip craniectomy (ESC) and helmet therapy is being used more frequently to treat craniosynostosis. This study evaluated the change in fronto-facial asymmetry in infants with UCS who were treated with ESC.

METHODS: This IRB-approved, retrospective study included 16 patients who underwent ESC and postoperative helmet therapy. Cranial anthropometric data was collected preoperatively and during follow-up visits after surgery. Preoperative and follow-up photographs of patients were analyzed using ImageJ software. Craniofacial analysis was conducted for forehead asymmetry (defined by difference of midline occipital to left frontal and midline occipital to right frontal measurements), nasal angulation, and facial angulation.

RESULTS: The mean follow-up was 30 months. In the ESC patients, mean nasal tip angulation improved from 12.3 degrees preoperatively to 4.3 degrees at follow-up (p<0.001). Mean facial midline angulation improved from 4.8 degrees preoperatively to 1.35 degrees at follow-up (p<0.001). Forehead asymmetry improved from 8.4 mm preoperatively to 3.1 mm at follow-up (p=0.006). The majority of the correction happened within the first 12 months but was maintained out to 30 month follow-up.

CONCLUSIONS: Our study provides evidence of statistically significant improvement of forehead asymmetry, nasal angulation, and facial angulation in patients who underwent ESC followed by helmet therapy.

DISCLOSURE: None of the authors have any commercial or financial interests related to this study.

Evaluation of a Patient with Metopic Synostosis Treated using Cranial Orthosis

Kamlesh B. Patel, MD; Dennis C. Nguyen, MD; Gary B. Skolnick BS; Sybill D. Naidoo PhD, RN; Matthew D. Smyth, MD

PURPOSE: Outcomes in patients with metopic synostosis are focused on improvements in head shape due to surgical intervention.1,2 Most patients with true pathological trigonocephaly are managed surgically, therefore literature is lacking on frontal morphology in untreated patients.3 We present and analyze the early result with orthotic therapy alone for a patient with metopic synostosis.

CASE PRESENTATION: A 6-month-old twin boy born at 30-weeks via a surrogate presented to the clinic with chief complaint of abnormal head shape. Patient’s history was significant for VACTERL and had undergone repair of tracheoesophageal fistula at 2 days of life. He also required prolonged respiratory support due to primary pulmonary hypertension. On physical exam the head circumference was in the 26th percentile with both occipital brachycephaly and trigonocephaly with frontal width of 62mm. Intercanthal distance was 27mm. Computed tomography scan showed 7 of 8 characteristic findings consistent with metopic synostosis as described by Birgfeld and colleagues (metopic ridge, pulled anterior fontanelle, posteriorly displaced lateral frontal bone, tangent drawn along lateral frontal bone intersects...