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Evaluation of Endoscopic Strip Craniectomy and Orthotic Therapy for Bilateral Coronal Craniosynostosis

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Abstract

Background: Bilateral coronal craniosynostosis is the premature fusion of both coronal sutures. Traditionally this condition is treated by frontal-orbital advancement. Endoscopic strip craniectomy with cranial orthotic therapy, which has gained popularity in treating single suture craniosynostosis, has recently been adapted for treatment of bicoronal synostosis. There have been few studies documenting the outcomes of this treatment. The objective of this study is to compare the morphological outcomes of endoscopic strip craniectomy and FOA in patients with bicoronal synostosis.

Methods: A retrospective case series was done on twenty-four patients with bilateral coronal synostosis treated with endoscopic strip craniectomy or FOA at two institutions. Patients with pre-operative and one-year post-operative computed tomography scans were included. Multiple measures of cranial shape and size were assessed: cephalic index, turricephaly index, basofrontal angle, circumference Z-score, and cranial vault volume Z-score.

Results: The two groups were similar statistically in terms of morphology at pre-operative scan. There were no statistical differences between the FOA group and endoscopic group at post-operative scan in any of the morphologic outcomes measured. Cranial vault volume z-scores increased post-operatively in both the Endoscopic ($p < 0.001$) and FOA ($p = 0.034$) groups.

Conclusions: One year after repair there were no significant morphological difference between patients with bicoronal synostosis treated with the endoscopic approach and those treated by FOA.

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Keywords

Bilateral Coronal Craniosynostosis; Endoscopic Strip Craniectomy; Orthotic Helmeting

Introduction:

Bilateral coronal craniosynostosis is the premature fusion of both coronal sutures. It is the most common pattern of multi-suture fusion, with two-thirds of patients having an associated syndrome. It causes a characteristic growth pattern notable for growth restriction in the anterior-posterior (AP) dimension (brachycephaly) and increased growth in the vertical plane (turriccephaly), leading to turribrachycephaly (Figure 1a). In addition to the cranial deformity, the rates of increased intracranial pressure (ICP), and of cognitive impairment is higher than in patients with single suture synostosis. ¹

There are a few surgical options for correction of the cranial deformity in patients with bilateral coronal craniosynostosis. Traditional treatment involves a bilateral fronto-orbital advancement (FOA), in which the frontal bones and superior orbital rims are removed, followed by their reshaping and repositioning². FOA is usually performed after 6 months of age due to increased safety and stability of construct³. Early posterior vault distraction is increasing in popularity⁴. This allows for rapid cranial growth in the AP dimension which can help correct turriccephaly, increase intracranial volume and decrease ICP. This procedure is then followed with a delayed FOA, to facilitate the correct relationship between the superior orbital rims and the cornea.

Endoscopic strip craniectomy followed by orthotic helmeting has been shown to be a successful treatment option for single suture craniosynostosis⁵. This procedure is associated with significantly lower blood loss, fewer transfusions, shorter operative time, decreased length of stay, and fewer ICU admissions⁶. However, patients are required to wear the orthotic helmet for 23 hours per day, until approximately one year of age. This requires frequent follow-up with a trained cranial orthotist. Jimenez et al and Rottgers et al have reported on using this technique to treat bilateral coronal craniosynostosis ^{7,8}. However, to date there has been no direct comparison between the FOA procedure and the endoscopic strip craniectomy followed by orthotic therapy for treatment of bilateral coronal craniosynostosis. The purpose of this study is to use craniometric analyses to compare morphologic outcomes of these two procedures.

Material and Methods:

Following IRB approval, a retrospective review of patients with bilateral coronal craniosynostosis was performed at two institutions. Patients with pre and post-operative computed tomography (CT) scan data, with post-operative scan date roughly one-year after repair, were included. Patients with scans of unsatisfactory quality were excluded. All patients either underwent FOA or endoscopic strip craniectomy with subsequent orthotic therapy at either Boston Children's Hospital (BCH) or St. Louis Children's Hospital (SLCH).

Procedures:

The FOA procedures were performed between 1996 and 2012 at SLCH. Endoscopic-assisted strip craniectomy cases were done between 2007 and 2014 at SLCH as described previously. Endoscopic repairs at BCH were performed between 2005 and 2014. They involved an 8–10mm width craniectomy of the affected fused sutures (Figure 2a), followed by the use of an orthotic helmet. A five day post-operative CT scan of a patient who underwent an endoscopic strip craniectomy is shown in figure 1b.

Craniometric Analysis:

Each scan was analyzed using Analyze 12.0 (Mayo Clinic, Rochester, MN) by a single operator (CW). After reconstruction and orientation to the Frankfort horizontal, cranial index (Figure 2b) and head circumference were calculated using the volume render and ROI tools respectively. The scans were then bisected in the mid-sagittal plane. The vertex, glabella, nasion, and sella turcica were landmarked, allowing measurement of the cranial height (distance between sella turcica and vertex), turricephaly index (AP diameter divided by cranial height) (Figure 2c), and basofrontal angle (Figure 2d).

After clipping the rendered volume to isolate the cranium, the volume was converted and exported as a mesh object in STL file format. This mesh was then imported into Blender (Version 2.78, www.blender.org). Cranial vault volume was determined using the shrinkwrap tool in Blender.

Nine scans were randomly selected for duplicate analysis to assess reliability. A minimum of one week elapsed between each paired evaluation.

Due to the age-and sex-dependent nature of head circumference and cranial vault volume, Z scores were calculated as follows: An online calculator was used for circumference. Unpublished data acquired for a previously published study of unaffected controls was used for cranial vault volume. After applying a log transformation on subject age, linear regression analysis of the male (n = 67) and female (n = 56) subsets was performed on this unpublished data. Cranial Vault Volume z-scores of the subjects accounting for both sex and age at scan were then calculated based on the regression model.

Demographic data of the two groups (FOA vs. endoscopic) were compared. Mean values of the five craniometrics indices (cephalic index, turricephaly index, basofrontal angle, circumference, and cranial vault volume) were compared between the two groups at both pre-operative and post-operative scan time points. Similarly, pre-to post changes within groups were assessed. 3D reconstructions of the one-year post-operative CT scans of the endoscopically treated patients were reviewed for evidence of re-synostosis. Peri-operative parameters: operative time and estimated blood loss data were available for the endoscopic repair group and obtained via electronic chart review. Perioperative data were not available for patients in the FOA group.

Statistical Analysis:

Intra-class correlation coefficients were calculated to determine intra-rater reliability. Categorical variables were assessed with Fisher's Exact test. Pre to post-operative

differences within groups and between group z-score differences were determined using two-tailed Student's t-tests. Other differences between groups were assessed using regression analysis. Regression models on the pre-operative data account for age at scan. Age at surgery was used as a covariate of pre to post-operative change. Models comparing the two groups' post-operative data use pre-operative severity as a covariate. Significance was set *a priori* at $p < 0.05$. All calculations were performed using SPSS ver. 22 (IBM SPSS, Chicago IL).

Results:

Demographics: 24 Patients were included in the analysis: 15 endoscopic patients and 9 FOA patients. 10 endoscopic procedures were completed at BCH, 5 were completed at SLCH, and all 9 in the FOA group were performed at SLCH. The FOA group included 3 males and 6 females. The endoscopic group included 8 males and 7 females. The FOA group and endoscopic group significantly differed in age at pre-operative scan (4.2 ± 1.8 vs. 1.5 ± 1.1 months, $p < 0.001$) and age at time of operation (5.3 ± 1.8 mos. vs. 2.7 ± 0.9 mos., $p < 0.001$). Age at post-operative scan in the FOA group was 18.6 ± 5.1 mos. and in the endoscopic group was 16.7 ± 5.1 mos., $p = 0.403$. 44% of the patients in the FOA group had an associated syndrome (Apert, Crouzon, Saethre-Chotzen, Di George, Muenke), whereas 73% of the endoscopic group were syndromic (Apert, Saethre-Chotzen, Muenke, Craniofrontonasal dysplasia) ($p = 0.146$). Intra-rater reliability: Intra-class correlation coefficients for morphologic parameters ranged between 0.856 and 0.999.

For intracranial volume, regression models on unaffected control generated the following:

Male: Cranial Vault Volume = $590.0 + 172.2 \times \ln(\text{age in months})$. R-squared = 0.861.

Female: Cranial Vault Volume = $482.7 + 179.8 \times \ln(\text{age in months})$. R-squared = 0.785.

Morphologic outcomes:

The two groups were similar statistically in terms of morphology at pre-operative scan (accounting for age), $p = 0.385$. Table 1. Median circumference Z score in the FOA group rose from 0.0 to 1.6 post-operatively. In the endoscopic group, median circumference Z score rose from 0.2 to 1.8 (Figure 1a). No significant differences were found between the cranial vault volume of FOA and Endoscopic groups pre-or post-operatively. Cranial vault volume z-scores increased post-operatively in both the Endo ($p < 0.001$) and FOA ($p = 0.034$) groups. (The two subjects less than 1 week of age at pre-operative scan were excluded as outliers from the cranial vault volume z-score analysis.) (Figure 1b). Photographic images and CT scans of single endoscopically treated patient at pre-operative and post-operative time points are shown (Figure 2) to demonstrate morphologic change. Two patients in the endoscopic group required subsequent FOAs after their post-operative CT scans, one patient at 14 months and the other at 31 months. Both of these patients had syndromic craniosynostosis. Secondary fronto-orbital advancement was done due to inadequate globe protection. One of these patients was found to have incomplete craniectomy of the coronal sutures. Three patients in the endoscopic group (20%)

demonstrated at least some re-synostosis (2 partial, 1 complete) at the 1 year post-operative scan. None of these patients underwent a subsequent FOA.

The endoscopic procedure had a reported mean EBL of 35 cc (range 0–50) and a mean operative time of 74 min (range 38–116).

Discussion:

Although Jimenez et al.¹ and Rottgers et al have reported on outcomes with the endoscopic repair and several groups have reported on outcomes for single suture synostosis², a direct comparison between endoscopic suturectomy/orthotics and FOA including morphometric analysis has not been previously published for bilateral coronal craniosynostosis. This study found no differences in morphologic outcomes for fronto-orbital advancement and endoscopic strip craniectomy followed by orthotic helmeting in the treatment of bilateral coronal craniosynostosis. The two groups of patients had similar morphology and age-adjusted size at the time of pre-operative scan and again at post-operative scan. The equivalence held for all five of the metrics studied (cranial index, turriccephaly index, basofrontal angle, circumference, and cranial vault volume). It is notable that the endoscopic procedure seemed to show a significant difference in all morphologic outcomes between the pre-and post-operative scans, while the FOA did not. This is likely due to the fact that the FOA group contained fewer patients, thus requiring a greater change to detect a significant difference.

The equivalence of the means of the measurements comes with a caveat. While endoscopic strip craniectomy with appropriate helmeting generally led to favorable morphologic outcomes, two of the fifteen patients required subsequent FOA and three of the patients demonstrated at least partial re-synostosis at the one-year post-operative scan. We have identified two shortcomings that may be responsible for these inferior results. Surgically, inadequate resection of bone near the squamosal suture might limit the ability of the orthotic helmet to guide frontal growth leading to re-synostosis and/or the need for secondary FOA. To avoid this, at SLCH we now use two 1 cm incisions on each side to insure proper completion of the craniectomy. Second, suboptimal post-operative helmeting may have played a role. Careful attention to the helmeting protocol may reduce the need for secondary repair. This includes ensuring helmet therapy begins as early as possible, often utilizing preoperative scanning, the use of orthotists trained in postoperative helmeting, and monthly communication between the orthotist and surgical team with surface scan data to assess morphological progress. In a previous study with longer follow-up after fronto-orbital advancement, Rottgers et al. showed that a high proportion of patients with syndromes require secondary FOA to address clinical concerns and “symptoms suggestive of intracranial hypertension.” They identified re-synostosis of the coronal sutures in many of those who required secondary FOA.

Although we were unable to obtain data on our FOA group, perioperative parameters around blood loss and time under anesthesia in the endoscopic group were lower than published values for patients treated by fronto-orbital advancement (550 cc, 240 min). These findings

are in line with reported reductions in blood loss and operating time in patients with single suture craniosynostosis treated by endoscopic strip craniectomy.”

While the population was bolstered by data from two institutions, this study remains limited by the small sample size. Furthermore, although not statistically significant, there was a higher prevalence of syndrome-associated craniosynostosis in the endoscopic group (73% vs. 44%). It remains to be determined if one or the other procedure obtains better morphological outcomes when restricting the patient pool to either the syndromic or non-syndromic population. Future studies with greater numbers of patients with this rare condition would be required to determine if there is any difference between these two procedures for syndromic or non-syndromic subgroups. In addition, these groups will require longer term follow up to see if the one year data remains consistent over time.

Future studies are also required to elucidate the differences, if any, between the endoscopic procedure and the FOA for neuropsychiatric outcomes.

We believe that endoscopic strip craniectomy and fronto-orbital advancement are both viable treatment modalities for bilateral coronal craniosynostosis. Each procedure has specific risks and benefits that must be tailored to individual patients. The fronto-orbital advancement has the risk of increased blood loss and longer operative time. The endoscopic procedure has an upper age limit and a learning curve, regarding complete removal of the fused suture and optimal postoperative orthotic therapy, to avoid secondary procedures.

Conclusion:

At one-year postoperative, we did not find significant differences in morphological outcomes in patients with bilateral coronal synostosis treated by fronto-orbital advancement and endoscopic craniectomy with orthotic helmeting.

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Figure 1a:

A 3D rendering of a computed tomography scan of a patient with turribrachycephaly, with profile (*left*) and vertex (*right*) views.

Figure 1b: Five-day post-operative CT scan (lateral view, left, and vertex view, right) of a patient who underwent endoscopic strip craniectomy.

Figure 2a:

Pre-operative picture demonstrating craniectomy made in endoscopic suture release.

Figure 2b: Cephalic index is the width of the skull at its widest point divided by its length.

Figure 2c: Turricephaly index is the length of the skull in the anterior-posterior dimension divided by the height, from the sella turcica to the vertex.

Figure 2d: The basofrontal angle is the angle between the sella turcica, the nasion, and the glabella.

Figure 3a:

Change in circumference Z scores from pre-to post-operatively.

Figure 3b: Change in cranial vault volume Z scores from pre-to post-operatively. Z-scores based on normal control data from Kamdar et al.

Figure 4: Photographic images and computed tomography scans of a patient treated endoscopically are shown at preoperative (*left*) and postoperative (*right*) time points to demonstrate morphologic change.

Table 1:

Regression coefficients for models comparing the two procedures. FOA: Fronto-orbital advancement.
Endoscopic: Endoscopic strip craniectomy.

		Coefficients (β)	<i>p</i>	r^2
Cranial Index	FOA vs endoscopic	-0.031	0.451	0.065
	ln(age)	0.012	0.243	
Turricephaly Index	FOA vs endoscopic	0.059	0.385	0.133
	ln(age)	0.009	0.613	
Basofrontal Angle	FOA vs endoscopic	0.049	0.992	0.125
	ln(age)	-1.448	0.222	