Endoscopic-Assisted Craniosynostosis Surgery

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Abstract

Over the last decade, endoscopy has been increasingly utilized in craniosynostosis surgery. In 2006, the author added endoscopy followed by helmet therapy to the treatment of young craniosynostosis patients. Since then, 73 children have been successfully treated utilizing endoscopic techniques with a transfusion rate of 23%. Most children are discharged on the first postoperative day; helmet therapy begins one week later. A helmet is worn for 4 to 6 months with one helmet replacement. Complications were limited to three reoperations to address suboptimal results, and one reoperation for a persisting skull defect. One sagittal sinus injury was addressed successfully, with resolution of a small intrasinus thrombus and no adverse brain sequelae. Although not applicable to every craniosynostosis patient, properly applied endoscopic-assisted craniosynostosis surgery is safe and effective, adding another option to the treatment armamentarium for craniosynostosis.

Keywords

- ▶ craniosynostosis
- ▶ endoscope
- ▶ pediatric

Surgery to correct craniosynostosis has evolved over the decades, from simple strip craniectomy to extensive cranial vault reconstructions to achieve superior results. All surgeries evolve over time to accomplish the same surgical goals with smaller incisions and less trauma. Vicari in 1994¹ and Drs. Barone and Jimenez in 1998² introduced endoscopic-assisted craniosynostosis surgery followed by application of a molding helmet. Over the years, the results have proven durable. Recently, more centers have undertaken this technique to enlarge their surgical options addressing skull and facial deformities.^{3–14} The author presents his series of endoscopic-assisted craniosynostosis surgery at Cook Children's hospital in Fort Worth, Texas.

How to Start an Endoscopic Program

In the fall of 2006, the author attended Drs. Jimenez and Barone's workshop on endoscopic-assisted craniosynostosis surgery (accompanied by a neurosurgical registered nurse first assistant [RNFA] and a local orthotist). After careful consideration, the orthotist and I concluded this was a worthy endeavor. Attention was then turned to making sure we had the appropriate surgical instruments. Most of the equipment was already available in our hospital. There are several

instruments crucial to the surgery. A serviceable endoscope is needed along with large pituitary bone rongeurs, including a downward curved and straight rongeur. A lighted retractor is useful to improve operative illumination. The disposable suction coagulator used in many ear, nose, and throat (ENT) procedures is used to coagulate the cut bony edges to decrease bleeding. Coated malleable brain retractors and Frasier suckers protect dura/brain from the suction coagulator. Finally, an experienced assistant is needed for the surgery. There is a learning curve for the team as they manipulate multiple instruments in a coordinated fashion. For my surgeries, the same RNFA assists. After careful planning, the first endoscopic surgery was performed in November 2006. Since that time, 73 surgeries have been completed (▶ Table 1).

How to Choose between Open versus Endoscopic Approach

All families are offered the choice of endoscopic versus traditional open surgery, and the pros and cons are each discussed thoroughly. However, aging and concomitant increasing thickness of the skull render endoscopic craniosynostosis surgery more difficult. At this time, the author, having performed endoscopic craniosynostosis surgery on several

Table 1 Series data

Type	Number	Mean age (days)
All	73	115
Sagittal	43	105.6
Metopic	14	114.2
Coronal	12	151.8
Lambdoid	0	0
Multisuture	5	109.2

children older than 6 months and achieving suboptimal improvement of head shape, no longer offers endoscopic surgery to these children. The decreased blood loss and shorter operative times associated with surgery on the relatively thin calvarium of younger patients renders endoscopic repair safer in very young children. Our earliest endoscopic intervention was carried at 32 days. Another advantage with a younger child, their rapid head growth assures prompt head-shape correction following surgery and molding helmet.

If the child is close to 6 months old and harbors a severe calvarial deformity related to craniosynostosis, I recommend open surgery. However, in younger children, I will still proceed with endoscopic repair with severe deformity. It remains a judgment call with no hard rules.

Multisuture craniosynostosis can be managed with an endoscopic approach. However, cranial orthosis treatment can be difficult. I have only performed one syndromic synostosis (Pfeiffer syndrome) with many postoperative complications and reoperations. I would be willing to do it again, but would proceed very cautiously.

Essentially, the risks of each approach are the same, except for the need for transfusion. Transfusion rates for endoscopic surgery are 23% overall (► **Table 2**) compared with a nearly 100% transfusion rate with open surgery. However, I am much more conservative with transfusions in the endoscopic series, allowing the hematocrit to drift into the upper teens as long as the children remain clinically stable. I would not do this with the open surgery due to the continued fall of the hematocrit postoperatively, a phenomenon not nearly so common with endoscopic surgery. Coagulating the skull edges at the end of the case, assures little change in the hematocrit postoperatively (► **Table 3**).

Table 2 Transfusion rates

Type	Transfusion rates
All	17/74 = 23%
Sagittal	12/43 = 28%
Metopic	2/14 = 14.3%
Coronal	0/12 = 0%
Multisuture	3/5 = 60%

Surgery

The goal of endoscopic surgery is to remove the abnormal suture, then allow brain growth and the postoperative molding helmet to reshape the skull. Because head-shape correction occurs slowly after surgery, helmet therapy is just as important as the surgery. All families have preoperative orthotics consult to explain helmet therapy.

For sagittal synostosis, the strip craniectomy is made 4 to 5 cm wide to accommodate continued calvarial expansion as the brain grows and skull reossifies. If the craniectomy is too small, the bone will regrow before the head shape has corrected. For metopic, lambdoid, and coronal synostosis, the strip craniectomy is usually 1 cm in width along the abnormal suture.

The endoscope is only used to help visualization while working. I've found it is only necessary while coagulating the cut bony edges. This allows precise and safe coagulation to prevent dura or brain injury. The endoscope also allows the assistant to see, which allows more instruments to be used. A brief description of each procedure follows. Please see Jimenez and Barone's publications for any further clarification.^{2,15–22}

The children are transferred from the recovery room to a hospital room. Most children are sent home the next day (mean stay 1.1 days), after we check their hematocrit level. They are fitted for their helmet one week after surgery.

Sagittal Craniosynostosis

The patient is prepared for surgery in the usual fashion. I recommend an arterial line, but central venous access is not required. The patient is placed prone, the head elevated in a swan-dive position. This can be accomplished with a beanbag, although there are foam-padding devices that can be purchased for assistance. Hip and shoulder rolls are placed to prevent compression of abdomen and great care is used for padding at all pressure sites. Skin prep is the surgeon's preference; we infiltrate the skin incisions with a half percent lidocaine with epinephrine. Two incisions 4 to 5 cm long are made perpendicular and across the sagittal suture: The first lies 2 cm behind the anterior fontanelle and the other 2 cm in front of the lambda (intersection of sagittal and lambdoid sutures) (► **Fig. 1**).

Extensive subgaleal dissection is made. A single bur hole is fashioned at each incision, and then using a Kerrison rongeur, a trough of bone is removed parallel to each incision, crossing the fused sagittal suture. Liberal use of thrombin-soaked Gelfoam assures hemostasis. Using a periosteal freer, the dura is dissected from the skull from each trough to the anterior fontanel and to the lambda. With bone scissors under direct vision, a small triangular craniectomy is made from the anterior trough to the anterior fontanelle and from the posterior trough to the lambda. By removing this bone, the endoscope may be used to dissect the dura from trough to trough under the sagittal suture. When possible, bridging veins can be coagulated and cut before they are torn, to minimize bleeding. However, I find this tedious and time consuming; so the endoscope or blunt dissector (a #1 Penfield is helpful) is passed from trough to trough to gently free the dura from the bone.

Table 3 Change in hematocrit

Type	Mean preop hematocrit	Mean immediate postop hematocrit	Mean postop day (POD) 1 hematocrit	Mean change in hematocrit from postop to POD 1
All	32.1	26.8	25.1	-1.7
Sagittal	32.1	26.1	24.6	-1.5
Metopic	32	26.6	24.7	-2.0
Coronal	33	27.1	25.9	-1.2
Multisuture	30	32.6	29.1	-3.6

Small bridging veins may be torn, but stop with application of epidural FloSeal (Baxter, Deerfield, IL) or direct coagulation. Large scissors are then used to cut a 4 to 5 cm strip craniectomy extending from trough to trough. Occasionally, the bone has to be divided while still under the scalp to allow removal via smaller incisions. This cutting is done blindly. One must remember to keep the scissors tips up so the tips will not dig into the dura and produce tears. We have produced a large dural laceration with this maneuver on one occasion, which necessitated an additional scalp incision to allow dura closure. Once the bone has been removed, the endoscope is used to inspect for any dura lacerations or vigorous bleeding. Two triangular-shaped barrel-staving cuts in each parietal bone laterally will facilitate skull expansion. At this point, the endoscope is used with a suction-coagulator to coagulate all bony edges to stop bone oozing. An assistant places a coated malleable between the skull and dura to prevent arcing from the monopolar coagulator to the dura. The scalp is also lifted with a lighted retractor to protect the scalp. The incisions are then closed in the usual fashion. Surgery time is usually 45 minutes.

Coronal Craniosynostosis

This is the easiest of the surgeries. I do not require an arterial line for this procedure. The patient is positioned supine with head turned slightly, with the affected side up. The 2- to 3-cm scalp incision is outlined roughly one-third of the way down

the coronal suture from the fontanelle, perpendicular to the suture (→**Fig. 2**). Galeal dissection is performed with cautery. A single bur hole made on the suture with an automatic perforator is slightly enlarged with a Kerrison rongeur. The dura is then blindly dissected with a periosteal freer. A 1-cm strip craniectomy is then performed with scissors and large pituitary bone rongeurs from the anterior fontanelle to the squamosal suture. The endoscope may be used to inspect the inferior bony removal to make sure this has reached the squamosal suture. The skull edges are coagulated with endoscopic assistance. This is more challenging than the sagittal craniosynostosis operation because there is only one small incision. This takes some practice to be able to coordinate all of the instruments (endoscope, suction coagulator, coated malleable) with two people and one incision. Sometimes a coated Frasier sucker is used rather than the bulkier malleable retractor to protect the dura. This operation usually takes 20 to 30 minutes to perform.

Metopic Craniosynostosis

This may be the most technically difficult surgery due to curvature of the forehead and the need to place the incision behind the hairline. The patient is kept supine with his or her head in a soft donut headholder. I do not hesitate to place an arterial line in these patients because of potential blood loss. A 2- to 3-cm incision is placed just behind the hairline, perpendicular to the metopic suture. Subgaleal dissection is

Fig. 1 Set-up for sagittal craniosynostosis repair. Solid lines indicate incisions. Dotted lines indicate planned skull removal.

Fig. 2 Set-up for coronal craniosynostosis repair. Solid arrow indicates the planned incision site. Dotted arrow indicates planned strip craniectomy from squamosal suture to anterior fontanelle (diamond).

performed with the endoscope and with a malleable extension on the cautery so the galeal may be dissected all the way to the nasion. A lighted retractor helps to protect the scalp from cautery injury. A single bur hole is placed over the metopic suture and widened slightly with a Kerrison rongeur. A 1-cm trough of bone is cut to the fontanelle. If the skull is thin enough, scissors are used to cut as far as possible down either side of the suture to the nasion. However, the bone tends to be too thick to get very far. Tactics are then changed to piecemeal removal of the suture with large bone pituitary rongeurs. The endoscope is very useful to make sure the dura is not getting caught in the rongeurs. Proceeding down the suture, visualization is more difficult. It is also useful to switch to downward curved rongeurs. The strip craniectomy needs to be dutifully performed to the nasion to ensure correction from the molding helmet. Once completed, coagulation of bone edges is performed to minimize postoperative bleeding. Thickness of the bone itself and the tedious piecemeal removal of bone promotes greater blood loss.

Complications

Blood Loss

Blood loss in an endoscopic procedure is much less than in an open operation. Our data is summarized and presented in ►Tables 2 and 3. It should be noted that three of the 17 were transfused due to other circumstances. Two were transfused due to congenital heart defects: The anesthesiologists wanted to keep the hematocrit higher (one transfused before the operation). The third infant was transfused before the operation started due to dilution from intravenous (IV) fluids and blood loss while starting IV and arterial lines. Six of the 17 patients were transfused after the operation (five in post-anesthesia care unit and one on postoperative day 1).

Dural Lacerations

Dural lacerations occurring during open operations are easily repaired. Dural laceration has occurred 2 (2.7%) times with the endoscopic approach. One was large, inflicted by the bone scissors used to cut the sagittal strip of bone, and required a new scalp incision to access the area requiring repair after endoscopic repair was unsuccessfully attempted. An associated diminutive cortical laceration appeared to bear no neurologic sequelae. The second case involved a small dura tear next to sagittal sinus resulting in brisk bleeding. This was incompletely closed due to bleeding. Postoperative magnetic resonance imaging (MRI) revealed a normal sagittal sinus and normal brain.

I have damaged the sagittal sinus, tearing the lateral wall in two cases (one previously discussed). One had a postoperative clot in the sagittal sinus without sinus occlusion; the patient was started on aspirin. After 3 months, MRI showed a normal sinus with resolution of the clot; aspirin therapy was discontinued. She remained asymptomatic.

Brain Injury

Besides the aforementioned minor brain laceration, there are no known brain injuries. Several postoperative scans have

been performed for various reasons and no injuries have been noted. No new clinical neurologic deficits have been noted in follow-up.

Skull Defects

Usually, calvarial regrowth is slow while the head shape changes during application of postoperative molding helmets. Once the head shape reaches the desired configuration, bone regrowth is rapid. Older children exhibit slower progression of head shape and bone regrowth. Even large defects from sagittal craniosynostosis reossify well. Full repair of the skull is usually accomplished by the end of helmet therapy. I have not seen any skull defects associated with coronal, lambdoid, or metopic correction. It is not uncommon to see minor skull defects (less than a centimeter in diameter) present one year postoperatively.

Three sagittal patients had larger defects more than one year postoperatively. One has been repaired with autologous cranioplasty and repair of harvest site with synthetic bone product (Mimix, Biomet, Jacksonville, FL). This produced excellent functional and cosmetic results. Two patients have recently returned to clinic with defects that will require repair. One, now 4 years old, harbors a large defect. His preoperative scan shows significant compensatory changes to both parietal bones (►Fig. 3). Postoperative MRI and computed tomography (CT) show no encephalocele or dura damage as the source of the defect (►Fig. 4). My working hypothesis is that his poor calvarial regrowth is related to an underlying metabolic/dura abnormality. Workup for this is pending. The other child's defect is smaller, but prominent dura/brain pulsations at the defect are preventing skull regrowth. An MRI is pending to see if there is an underlying structural defect that is the source.

Fig. 3 Preoperative computed tomography scan showing significant skull changes due to sagittal craniosynostosis.

Fig. 4 Computed tomography scan obtained 3 years postoperatively from craniosynostosis surgery showing large residual skull defect over the vertex.

Regrowth of the skull after sagittal repair sometimes is not smooth. Usually, these are minor variations, but occasionally ridging and divots do occur. These are always in the hairline and well hidden. When explained to family that these are not problematic, they are well tolerated. No family has asked me to repair these. This seems to be no different than the “lumpy-bumpy” results seen with open repair.

Reoperations

We have had to reoperate three times for poor cosmetic result (two unilateral coronal reconstructions and one sagittal reconstruction). The sagittal reconstruction went well, but it appears the helmet therapy induced poor growth of the inferior frontal bone and orbital rims resulting in high frontal bossing. Postoperative CT showed open coronal sutures. We recommended close observation over time, anticipating that with head growth these abnormalities would slowly improve. However, the mother was displeased with the results and demanded open reconstruction. Repair of frontal bossing and advancement of the orbital rims was undertaken successfully.

Both of the coronal reoperations were due to residual recessed orbital rim and inferior frontal bone. The superior forehead was normal. Both of these were repaired with open reconstruction. I have had the same results with an open operation requiring reoperation.

Open reoperation is not rendered more difficult by previous endoscopic surgery. The previous incisions are easily incorporated into the new incision. There is no increased scarring of the dura to the skull. The galea can adhere to the skull (as with any reoperation), but causes minimal problems. Because the coronal endoscopic repair has limited exposure, the open craniotomy is straightforward.

Helmet Complications

Helmet therapy is well tolerated. Minor skin irritation is easily treated by not wearing the helmet for a few days. There have been no open wounds. Helmet therapy lasts approximately 6 months. Patients usually only require two helmets over the treatment span; rarely will they need more. Occasionally, approval for insurance coverage can be tricky, but rarely have they refused. A letter sent to the insurance carrier at the time of approval of surgery to explain the use of postoperative helmet therapy has limited the hassle factor.

The use of helmet therapy is meticulously explained to families before surgery, but we still have families that do not use the helmets as prescribed. My impression is that their cosmetic results are inferior to our other patients. There are numerous visits to the orthotics office when starting helmet therapy. For those families traveling long-distance (not uncommon due to our large referral area), we make sure families understand the commitment necessary for helmet follow-up. Several families have had to decline the surgery due to travel hardships; open surgery was performed instead.

Conclusion

Although not a cure-all, endoscopic-assisted craniosynostosis surgery is safe and effective. It adds another option in our treatment armamentarium for craniosynostosis.

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