Endoscopic-assisted repair of craniosynostosis

GREGORY J. A. MURAD, M.D., MARK CLAYMAN, M.D., M. BRENT SEAGLE, M.D.,
SNO WHITE, M.D., LEIGH ANN PERKINS, A.R.N.P., AND DAVID W. PINCUS, M.D., PH.D.

Department of Neurosurgery, Divisions of Plastic Surgery and Pediatric Anesthesiology, and the University of Florida Craniofacial Center, University of Florida College of Medicine, Gainesville, Florida

Object. The goal of the craniofacial surgeon has always been the correction of form and function with prevention of associated morbidity and death. Through the pioneering work of Jimenez and Barone, minimally invasive approaches to the surgical correction of craniosynostosis are now gaining wider acceptance. Here the authors review the technique for endoscopic-assisted repair of craniosynostosis from the perspective of a new minimally invasive approach. They also assess the safety, efficacy, and results of the early treatment of infants with craniosynostosis in a small series of children who underwent surgery at this institution.

Methods. Data regarding synostosis type, operative time, patient age, blood loss, transfusion rates, duration of hospitalization, and complications were collected. Nineteen patients (12 girls and seven boys) between the ages of 1.2 and 5 months of age were treated with the endoscope-assisted technique. The mean operative time was 97 minutes. Five (26%) of 19 children received a blood transfusion. Most patients were discharged home the morning after surgery. The clinical courses of two patients who required additional major craniofacial reconstructions are discussed. There were no deaths, dural sinus tears, cerebrospinal fluid leaks, neurological injuries, or infections, and there were no complications related to the use of helmet therapy. Seventeen of the 19 patients achieved excellent cosmetic results with a single surgery.

Conclusions. This small series supports larger experiences and indicates that early treatment of craniosynostosis with minimally invasive, endoscope-assisted techniques is safe; limits blood transfusion, hospital stay, and operative time; and represents a valuable alternative to the traditional calvarial reconstruction methods.

KEY WORDS • endoscopic surgery • minimally invasive surgery • craniosynostosis • pediatric neurosurgery • strip craniectomy

Craniosynostosis, or the premature closure of calvarial sutures, may result in progressive skull deformity in children. It is a common cause of deformational cranial changes and has an estimated frequency of 0.4 of 1000 persons.¹⁰ Approximately 80 to 90% cases involve isolated defects, while the remaining cases are part of a recognized syndrome such as Crouzon or Apert. Although the cause is frequently unclear, recent work suggests that mutations of the fibroblast growth factor signaling pathway play a role.^{1,23} In the isolated cases, the sagittal suture is affected most often (55%), followed by the coronal (20%), lambdoid (5%), and metopic (5%) sutures.⁴ The fused suture restricts growth of the calvaria, thus leading to a characteristic deformation, each associated with a different type of craniosynostosis. Premature closure of skull sutures is associated with compensatory cranial and facial deformational changes that by 6 months of age often present with changes requiring major reconstructive procedures.³

The first surgical treatment of craniosynostosis was un-

dertaken by Lannelongue¹⁷ in 1892, and involved the correction of a sagittal synostosis. Since then, multiple procedures have been used for the treatment of this condition, ranging from simple suturectomies to extensive calvarial vault remodeling.^{2,5,14,18,19} Experience has shown that more extensive reshaping yields excellent results, particularly in older children with moderate to severe deformity.²⁵ On the other hand, children undergoing more extensive reconstructions have a high requirement for blood transfusion, may experience hypothermia caused by prolonged surgery, and often have lengthy stays in the ICU. Surgery for craniosynostosis has evolved rapidly over the past two decades, with increased emphasis on early operations and on less invasive procedures.²¹ Because of changes in surgical timing and techniques, earlier series may not accurately reflect more recent experience. Currently, surgeons at many craniofacial centers favor surgical correction before the age of 6 months to avoid the morbidity caused by extensive calvarial vault remodeling in older children.

Jimenez and Barone¹² first described endoscopic synostosis repair in 1998. This technique allows for a less invasive method of craniosynostosis repair. When detected early, minimally invasive repair combined with a postop-

Abbreviations used in this paper: CT = computerized tomography; ICU = intensive care unit; 3D = three-dimensional.

Fig. 1. Photograph of patient positioned prone for sagittal synostosis repair with face supported in the horseshoe beanbag. The neck is extended, providing excellent access to the entire calvarium.

erative molding device can result in excellent longstanding reconstruction of the cranial skeleton. These methods may decrease some of the morbidity, such as that caused by blood loss, involved with traditional reconstructions. The length of the incisions is shorter, surgery is less prolonged, and durations of ICU and hospital stays are reduced. Although these procedures are unlikely to ever completely replace standard ones using bicoronal incisions, multiple craniotomies and osteotomies, and plate and screw reconstruction (particularly in children > 6 months of age), they should be part of the armamentarium of the modern craniofacial surgeon for the treatment of craniosynostosis in the neonatal period.

We began using minimally invasive techniques with endoscopic assistance at the University of Florida Craniofacial Center 2 years ago. Our goal was to minimize the need for blood transfusion, shorten scalp incision length, and reduce operative time and postoperative recovery in patients younger than 6 months of age undergoing craniosynostosis repairs. As in the series published by Jimenez and Barone,¹² all children were fitted for calvarial orthoses postoperatively to optimize long-term outcomes. The experience of and techniques used at the University of Florida are described.

CLINICAL MATERIAL AND METHODS

Patient Selection, Preoperative Imaging, and Anesthesia

We prefer the endoscopically assisted, minimally invasive repairs for children younger than 4 months of age. No preoperative blood tests are done unless the family wishes to have donor-directed blood available. For children with sagittal synostosis, we do not obtain any preoperative images. Computerized tomography scans are obtained in children with all other types of craniosynostosis. The patient is brought to the major operating suite, and general anesthesia is induced. Our pediatric anesthesiologists prefer to place two intravenous catheters and an arterial line similar to that used in major craniofacial procedures; however, if difficulty is encountered in arterial line placement, this step can be deferred. Bladder catheters are not used. A precordial Doppler ultrasonography procedure is advantageous for the identification of intraoperative air emboli. A single preoperative dose of cefazolin (30 mg/kg) is administered. Warmers are placed under and on top of the child.

Positioning and Skin Preparation

For metopic, unicoronal, or bicoronal repairs the patient is positioned supine with the head supported in a pediatric horseshoe headrest (Mayfield; Integra Lifesciences Co., Plainsborough, NJ). For sagittal repairs, the child is positioned prone in a beanbag horseshoe (Soule Co., Lutz, FL) with the neck extended (Fig. 1). This provides good access to most of the calvaria. No hair is shaved. The scalp is then prepped for 5 minutes with povidone iodine scrub. Povidone iodine paint is then applied. Incisions are marked for the particular repair being performed and 0.25% bupivacaine with 1:100,000 epinephrine is injected. The head is then draped in the standard fashion.

Incision and Subgaleal Dissection

Incisions are made by scoring the skin with a No. 15 blade and dividing the scalp using monopolar cautery and a Colorado needle (Stryker Instruments, Portage, MI) at 15 W. This technique results in little, if any, blood loss. Monopolar cautery at 25 to 35 W with a suction coagulator (Valleylab, Boulder, CO) is then used to separate the galea from

Fig. 2. *Left:* Schematic drawing showing an angled, lighted retractor in position for subgaleal dissection. *Center:* The rigid endoscope is used for dissection of the dura from the calvarium. *Right:* Right endoscopic view of sagittal sinus dural dissection.

Endoscopic-assisted repair of craniosynostosis

the underlying pericranium. Taking advantage of this dissection plane rather than the subpericranial plane is important in minimizing blood loss. An angled, lighted rhinoplasty retractor (Aufricht; Techman International Co., Charlton City, MA) allows for visualization during this step (Fig. 2). To avoid damage to the skin when cauterizing, hemostasis is achieved with the monopolar cautery and insulated bipolar cautery.

Initial Craniectomy and Dural Dissection

A high-speed drill is used to fashion a burr hole over the stenotic suture. Dural stripping is begun using Penfield dissectors and curettes. Rigid endoscopes (0 and 30°) are then used to aid in visualization during the stripping of the dura mater from the involved suture (Fig. 2). This maneuver is usually a straightforward one that is performed with suction

Bone Removal and Hemostasis

Once the dura has been adequately separated from the inner table of the skull, curved Mayo scissors are used to cut a strip craniectomy of the involved suture. Additional bone removal is performed using rongeurs. We find the Jansen Middleton nasal rongeur (Weck Surgical Instruments, Research Triangle Park, Raleigh, NC) particularly helpful for the most distal aspect of certain bone cuts. Bone bleeding is controlled with bone wax and the use of the suction coagulator at a setting of 50 W (Fig. 3). Care is taken to retract the dura away from the coagulator with a flexible insulated brain retractor. Similarly, the galea is protected with the lighted angled retractor. Additional hemostasis is obtained with a combination of spray thrombin, thrombin-soaked Gelfoam, and a slurry of collagen-absorbable hemostat (Instat; Ethicon, Somerville, NJ) mixed with normal saline. This is applied with a syringe and large angiocath catheter. All topical hemostatic agents and debris are then irrigated from the epidural and subgaleal space with warm saline. The incisions are closed with interrupted inverted subcuticular 4-0 absorbable suture (Polysorb or Vicryl) and a simple running 4-0 absorbable suture (Vicryl Rapide) on the skin. The head is then thoroughly washed and the incisions are dressed with antibiotic ointment only.

Postoperative Care

Patients are extubated in the operating room. We require the children to spend 1 night in the pediatric ICU, but the regular pediatric surgical floor is completely acceptable. A single postoperative hematocrit is checked. We do not use a specific cutoff for blood transfusion and have allowed children to go home with hematocrit values as low as 16%. Packed cells are administered to patients with symptomatic anemia only. All patients are allowed to leave the morning following surgery. Additional hospital time has primarily been for alleviation of parental anxiety.

Posthospital Care and Cranial Orthoses

Parents are instructed to wash their child's hair 4 days after surgery and with their normal bathing routine thereafter. Helmets are fitted by an orthotist 1 to 2 weeks after surgery. Although several types of cranial molding devices are available commercially, we prefer a custom-made helmet

Fig. 3. Endoscopic view of suction monopolar cauterization of craniectomy edge.

with a closed top (Fig. 4). We have noticed some protrusion of skull and osseous knob formation in children treated with open devices. Frequent visits are made to the orthotist for the 1st month to ensure proper fit and lack of pressure points. Follow-up visits are then made every 3 months for a recommended minimum of 12 months. Most children will require two helmets due to head growth. Patients with initially good cosmetic results who come out of the helmet prematurely because of poor compliance or parents' inability to pay for subsequent helmets may have some reversion of their head shape (Fig. 4).

Suture-Specific Considerations

Sagittal Synostosis. The patient is positioned prone. The malar eminences are supported and the beanbag positioner is connected to suction to maintain extension (Fig. 1). The anterior fontanelle and lambda are palpated and marked. A 5-cm midline strip craniectomy is marked on the scalp. Additional craniectomies are marked bilaterally just posterior to the coronal sutures and just anterior to the lambdoid sutures. Two small transverse incisions approximately 2 cm in length are then marked just posterior to the anterior fontanelle and anterior to the lambda (Fig. 5). After scalp preparation, draping, and skin incision, the galea is dissected from the pericranium, exposing the anterior and posterior fontanelles and the coronal and lambdoid sutures as far laterally as can be reached. Burr holes over the sagittal suture are fashioned at each incision site. The dura is dissected around each burr hole and craniectomies 5-cm wide are performed using rongeurs. The dura is stripped and the craniectomy is extended into the anterior fontanelle. The sagittal sinus is then separated from the skull using the endoscope, suction dissection, and Penfield dissectors. Opposing bone cuts are then made with Mayo scissors from the anterior and posterior incisions, resulting in a 5-cm craniectomy. This bone plate must then be cut in half to deliver the pieces through the scalp incision. Once complete hemostasis has been achieved, the dura is stripped immediately posterior to the coronal sutures and immediately anterior to the lambdoid sutures ex-

Fig. 4. Lateral (*left*) and anterior (*right*) views of a custom-made cranial helmet with a closed top.

Fig. 5. Photograph of a patient positioned prone in horseshoe beanbag for sagittal synostosis repair. Skin incisions are marked in *black* and the fontanelles in *green*. The midline craniectomy and bilateral parietal bone cuts are outlined in *blue*.

Fig. 6. Schematic drawing showing bone cuts for sagittal synostosis repair. The midline strip is approximately 5 cm wide and the parietal cuts are 1 cm wide.

Fig. 7. Schematic drawing of a 1-cm-wide synostectomy for right unicoronal synostosis.

Fig. 8. Schematic drawing illustrating bone removal (1 cm) for metopic synostosis.

tending to the squamous sutures bilaterally. Craniectomies of approximately 1 cm width are then made as far as possible using Mayo scissors and the Jansen Middleton rongeur (Fig. 6). Hemostasis and closure are then performed as described previously.

Coronal Synostosis. The incision is placed over the midportion of the involved coronal suture. A burr hole is made and the dura is dissected with endoscopic assistance. Dural separation is extended medially into the anterior fontanelle. A removal of the suture approximately 1 cm wide is performed with scissors and rongeurs (Fig. 7).

Metopic Synostosis. An incision is made over the metopic suture just behind the hairline. A burr hole is fashioned and the dura is separated from the metopic suture posteriorly into the anterior fontanelle. The dura is dissected anteriorly to the nasofrontal suture with endoscopic assistance. A bridging vein is frequently found near the nasion that must be cauterized with the bipolar. The metopic suture is typically thicker and more vascular than the other sutures. The suture is removed with rongeurs as part of a 1-cm-wide craniectomy down to the nasion (Fig. 8). Due to the thick bone, the high-speed drill with a long, curved guarded bit (Midas Rex T12 Dissector; Medtronic, Inc) is used for the most anterior portion of the craniectomy.

RESULTS

Nineteen patients between 1.2 and 5 months of age (12 girls and seven boys) were treated with the endoscopic technique over a 2-year period. Eleven patients had isolated sagittal suture involvement, five patients had metopic

synostosis, two patients had coronal synostosis with one bicoronal involvement, and one patient had both sagittal and metopic synostosis. The mean operative time was 97 minutes. Longer operative times were required in the patients with multiple suture synostosis. The mean estimated blood loss was 39 ml; two patients underwent intraoperative blood transfusion, and three patients (26%) had postoperative blood transfusion. Most patients were discharged the morning after surgery.

Complications were minimal. No deaths, dural sinus tears, cerebrospinal fluid leaks, neurological injuries, or infections occurred, and there were no significant complications related to the use of helmet therapy. Seventeen of 19 patients had good cosmetic results with a single surgery (Figs. 9–11).

Two patients had outcomes of note. One, a girl, underwent uneventful sagittal synostosis repair at 7 weeks of age with an initial good result. A 3D CT scan performed elsewhere prior to surgery indicated isolated sagittal synostosis (Fig. 12). Postoperatively, she was fitted with an open-topped helmet. She returned after 5 months with irritability and a bulging fontanelle. Computerized tomography scanning did not reveal hydrocephalus or intracranial mass but on 3D reconstructions pansutural synostosis was evident (Fig. 12). When she underwent surgery a second time, an open technique was used. Hyperostotic bone was encountered in the craniectomy site, and the metopic, coronal, and lambdoid sutures were stenotic. A complete calvarial remodeling was performed and her irritability resolved. Five months later she had recurrent signs and symptoms after moving to another state, and a repeat calvarial reconstruction was performed. Results of a fibroblast growth factor

Fig. 9. Preoperative (*left*) and 3-month postoperative (*right*) photographs of a boy with sagittal synostosis.

receptor analysis were nondiagnostic. The other child had multiple medical problems and underwent simultaneous sagittal and metopic repair. He had a prolonged hospital stay because of airway problems associated with his unknown genetic disorder. He subsequently underwent orbitofrontal advancement and tarsorrhaphies for corneal exposure caused by severe proptosis.

DISCUSSION

Although clinical descriptions of craniosynostosis date

back to Hippocrates and Galen,⁷ the first modern scientific investigator to describe the anatomical structure of calvarial sutures and the results of their premature closure was Sömmerring²² in 1800. In 1894, Jacobi¹¹ reported a series of complications and deaths associated with craniosynostosis surgery that led to its discontinuation for the next 30 years. Surgical intervention was subsequently reintroduced and has evolved over the years to include a variety of reconstructive procedures. Several different techniques for the repair of sagittal synostosis have been described, including complete calvarial remodeling, the pi procedure, and ex-

Fig. 10. Preoperative (*left*), 2-week postoperative (*center*), and 5-month postoperative (*right*) photographs of a boy with metopic synostosis.

tended vertex craniectomy. The most common procedure performed for metopic, unicoronal, or bicoronal synostosis is an orbitofrontal advancement. The reported blood loss, operative time, need for transfusion, hospital stay, and complications associated with these procedures can be significant and are well described in the craniofacial literature.^{8,9,13,15,16,20,24} In addition, traditional craniosynostosis repairs typically require a bicoronal scalp incision, which has the potential for suboptimal cosmesis due to alopecia and hair parting. The long-term cosmetic outcomes with these procedures are excellent, however, and cranial orthoses—with their associated expense and inconvenience—are avoided.

Minimally invasive techniques for craniosynostosis repair have several potential advantages over traditional open

surgery for selected patients with synostosis. Transfusion rates, operating room times, and ICU and hospital stays are reduced. Postoperative swelling, discomfort, and the associated parental anxiety are lessened. Finally, scars are substantially smaller and are often cosmetically superior to bicoronal incisions. Drawbacks include less exposure and control of dural sinuses and the importance of a postoperative helmet. We have found the latter to be the greatest obstacle to success for our patients. Although compliance is nearly universal and the devices are well tolerated, they are expensive and are not paid for by most insurance carriers. Cost is not an issue in the value analysis of these procedures compared with open surgery because the savings in ICU costs and hospital days dwarf the helmet expense,

Fig. 11. Preoperative (*left*), 6-month (*center*), and 15-month postoperative (*right*) photographs of a girl with left unicoronal synostosis. Note progressive improvement of nasal deviation. Some mild flattening of the left forehead can still be appreciated at 15 months postoperatively.

which varies from approximately \$800 to \$2000. Nonetheless, several families of limited means have chosen traditional surgery instead of endoscopic-assisted surgery because they could not pay for a helmet.

CONCLUSIONS

New, less invasive techniques for the early treatment of infants with craniosynostosis have been presented. Our small series supports the excellent results of previous reports of these methods, including low morbidity and no deaths, as well as decreased need for blood transfusion and

shorter ICU and hospital stays. Early diagnosis of craniosynostosis and prompt referral for surgical evaluation, as well as postoperative helmet therapy are paramount in obtaining the best possible results. Although minimally invasive methods will likely never replace traditional calvarial reconstructions, these techniques should be considered as important additions to modern craniofacial surgery.

Acknowledgments

We thank David Jimenez, M.D., for sharing his techniques and expert advice. We also wish to acknowledge David Peace, M.S.,

Fig. 12. *Left*: Preoperative 3D CT reconstruction obtained in a 2-month-old girl with isolated sagittal synostosis. *Right*: A repeated 3D CT reconstruction obtained at 7 months revealing pansutural synostosis (reconstruction).

C.M.I., and Robin Barry, M.A., C.M.I., for assistance with figure preparation and Bridget Richter for data gathering and manuscript preparation.

References

1. Aleck K: Craniosynostosis syndromes in the genomic era. **Semin Pediatr Neurol** 11:256–261, 2004
2. Alvarez-Garijo JA, Cavadas PC, Vila MM, et al: Sagittal synostosis: results of surgical treatment in 210 patients. **Childs Nerv Syst** 17:64–68, 2004
3. Barone CM, Jimenez DF: Endoscopic craniectomy for early correction of craniosynostosis. **Plast Reconstr Surg** 104:1965–1973, 1999
4. Behrman RE, Kuelman R, Jenson H: Craniosynostosis, in Kliegman R (ed): **Nelson Textbook of Pediatrics**, ed 16. Philadelphia: WB Saunders, 2000, pp 1831–1832
5. Boulos PT, Lin KY, Jane JA Jr, et al: Correction of sagittal synostosis using a modified Pi method. **Clin Plast Surg** 31:489–498, 2004
6. Breugem CC, van R Zeeman BJ: Retrospective study of non-syndromic craniosynostosis treated over a 10-year period. **J Craniofac Surg** 10:140–143, 1999
7. Cohen MM: **Craniosynostosis: Diagnosis, Evaluation and Management**. New York: Raven Press, 1986, pp 1–20
8. Faberowski LW, Black S, Mickle JP: Blood loss and transfusion practice in the perioperative management of craniosynostosis repair. **J Neurosurg Anesthesiol** 11:167–172, 1999
9. Faberowski LW, Black S, Mickle JP: Incidence of venous air embolism during craniectomy for craniosynostosis repair. **Anesthesiology** 92:20–23, 2000
10. Hunter AGW, Rudd NL: Craniosynostosis I. Sagittal synostosis: its genetics and associated clinical findings in 214 patients who lacked involvement of the coronal suture(s). **Teratology** 14:185–193, 1976
11. Jacobi A: Non nocere. **Med Rec** 45:609, 1894
12. Jimenez DF, Barone CM: Endoscopic craniectomy for early surgical correction of sagittal craniosynostosis. **J Neurosurg** 88:77–81, 1998
13. Jimenez DF, Barone CM: Intraoperative autologous blood transfusion in the surgical correction of craniosynostosis. **Neurosurgery** 37:1075–1079, 1995
14. Johnston SA: Calvarial vault remodeling for sagittal synostosis. **AORN J** 74:632–647, quiz 655–662, 2001
15. Kanev PM, Lo AK: Surgical correction of sagittal craniosynostosis: complications of the pi procedure. **J Craniofac Surg** 6:98–102, 1995
16. Kearney RA, Rosales JK, Howes WJ: Craniosynostosis: an assessment of blood loss and transfusion practices. **Can J Anaesth** 36:473–477, 1989
17. Lannelongue M: De la craniectomie dans la microcéphalie. **Compt Rend Seances Acad Sci** 50:1382–1385, 1890
18. McCarthy JG, Glasberg SB, Cutting CB, et al: Twenty-year experience with early surgery for craniosynostosis: I. Isolated cra-

- niofacial synostosis—results and unsolved problems. **Plast Reconstr Surg** **96**:284–295; 1995
19. Panchal J, Marsh JL, Park TS, et al: Sagittal craniosynostosis outcome assessment for two methods and timings of intervention. **Plast Reconstr Surg** **103**:1574–1584, 1999
 20. Ririe DG, David LR, Glazier SS, et al: Surgical advancement influences perioperative care: a comparison of two surgical techniques for sagittal craniosynostosis repair. **Anesth Analg** **97**:699–703, 2003
 21. Shillito J, Matson DM: Craniosynostosis: a review of 519 surgical patients. **Pediatrics** **41**:829–853, 1968
 22. Sömmering ST: **Vom Baue des Menschlichen Körpers**. Leipzig: Voss, 1800
 23. Thomas GP, Wilkie AO, Richards PG, et al: FGFR3 P250R mutation increases the risk of reoperation in apparent ‘non-syndromic’ coronal craniosynostosis. **J Craniofac Surg** **16**:347–354, 2005
 24. Tuncbilek G, Vargel I, Erdem A, et al: Blood loss and transfusion rates during repair of craniofacial deformities. **J Craniofac Surg** **16**:59–62, 2005
 25. Vollmer DG, Jane JA, Park TS, et al: Variants of sagittal synostosis: strategies for surgical correction. **J Neurosurg** **61**:557–562, 1984

Manuscript received October 15, 2005.

Accepted in final form November 21, 2005.

Address reprint requests to: David W. Pincus, M.D., Ph.D., P.O. Box 100265, Gainesville, Florida 32610. email: pincus@neurosurgery.ufl.edu.