Endoscopic-Assisted Craniosynostosis Procedure

Part 2 of 3

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In Part 1, the relevant anatomy, pathology, epidemiology, and types of craniosynostosis was discussed. Part 2 will discuss the specifics regarding the endoscopic-assisted craniosynostosis procedure including surgical patient position, steps of the procedure, expected clinical outcome including neurocognitive outcome, and postoperative helmet therapy.

ADVANCEMENTS IN CRANIOSYNOSTOSIS SURGERY

Rudolf Virchow, MD, German physician known as the "Father of Modern Pathology," was the first to describe the modern-day pathophysiology of craniosynostosis in 1851.[2] In 1881, the French surgeon Odilon Lannelongue, MD, first described strip craniectomy to treat craniosynostosis in which he made a straight incision along the fused suture releasing the constriction to allow cranial expansion, thus preventing further cranial deformity and decreasing the intracranial pressure.[2] However, in 1894, Abraham Jacobi, MD, known as the "Father of American Pediatrics", described the high morbidity and mortality rates associated with surgical treatment of craniosynostosis which led to the procedure not being performed for almost three decades.[2,3] The issues that were linked to the morbidity and mortality rates included inability to adequately control intraoperative bleeding, inappropriate patient selection, for example, operating on a patient with microcephaly instead of craniosynostosis, incomplete knowledge of the cranial anatomy, and the lack of advanced surgical and anesthetic techniques.[2]

LEARNING OBJECTIVES

- ▲ Explain the advancements of craniosynostosis surgery that led to the development of the endoscopic technique
- ▲ Describe the specific surgical patient position that is used
- ▲ Discuss the perioperative factors of the surgery including imaging studies, anesthetic monitoring, antibiotic prophylaxis, hemostatic control, and postoperative care
- ▲ List the steps of the surgical procedure
- ▲ Recall the instrumentation and equipment that are required
- ▲ Detail the expected patient surgical outcomes
- ▲ Communicate the reasons for the use of postoperative helmet therapy

KEYWORDS
craniosynostosis, helmet therapy, neurocognitive, sphinx surgical position, 3D stereophotogrammetry

DEFINITIONS

Calvarial: Also called the neurocranium or skullcap. Refers to the calvaria, the upper, dome-shaped section of the skull that protects the brain. It is formed by the frontal bone, two parietal bones, and occipital bone.

Emissary veins: Valveless veins that drain into the diploic veins that are located between the internal and external layers of the skulls compact bone (Figure 1). The presence and distribution of emissary veins vary in people. However, in infants and children, they occur more frequently and have larger foramen.[1] The lack of valves allows bidirectional blood flow to provide equalization of intracranial pressure and contributes to cooling of the brain.[1] Knowledge of the presence and position of the veins during surgery to repair craniosynostosis can prevent complications such as unintentionally damaging the veins causing air embolism, hemorrhaging, and venous thrombosis.

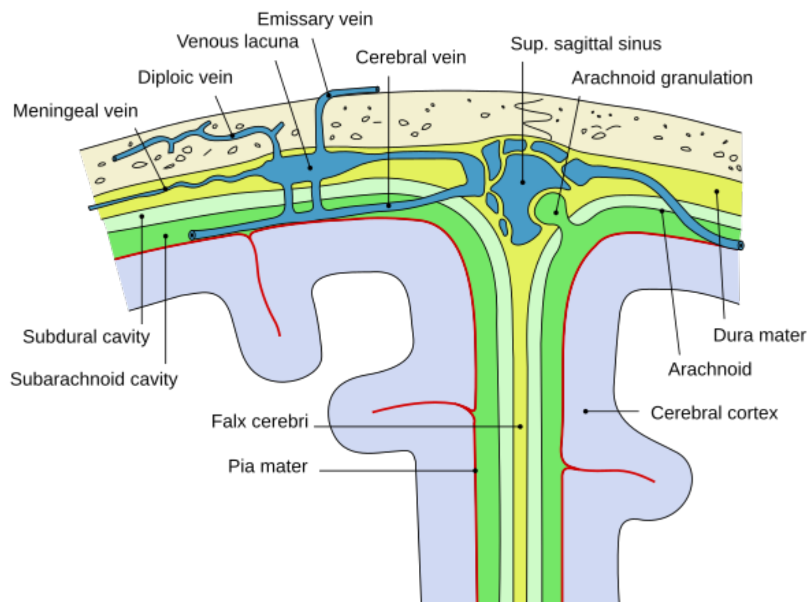

Figure 1 *Diploic and emissary veins*

> "…the development of minimally invasive techniques was crucial to better address the perioperative complications associated with craniosynostosis surgery."

It wasn't until 1921, when the German physician Arndt Mehner, MD, performed and reported the first successful strip craniectomy.[2] This revived interest in surgically treating patients with craniosynostosis led by Harold K. Faber, MD, and Edward B. Towne, MD, who performed multiple extensive craniectomies beginning in 1927.[2] Faber and Towne are also credited with advocating that surgery should be performed early in the child's life to prevent neurological complications caused by craniosynostosis.

A significant advancement in postoperative treatment proposed by John A. Persing, et al, is the importance of postoperative external cranial vault molding devices, such as helmet therapy.[4] The authors stated that surgery alone was not sufficient to correct craniofacial deformities.

Over time, multiple surgical techniques were developed, such as cranial vault remodeling, strip craniectomy, and hybrid midline craniectomies. The development of minimally invasive techniques was crucial to better address the perioperative complications associated with craniosynostosis surgery. In 1998, David F. Jiminez, MD, neurosurgeon, and Constance M. Barone, MD, plastic and reconstructive surgeon, introduced endoscopic-assisted surgery to treat sagittal craniosynostosis followed by postoperative helmet therapy.[5] This was another significant advancement in craniosynostosis surgery that laid the foundation for further innovations in minimally invasive techniques. They are also well known for establishing that the surgery should be performed prior to six months of age.

EMERGING TECHNOLOGIES AND RESEARCH

As with other surgical specialties and procedures, progress in treating craniosynostosis will be driven by technology and research. The use of 3D printed imaging that is currently being used for surgically treating pectus excavatum (see "Of Interest in the Medical Arena") is being investigated for use in creating implants for craniosynostosis patients that would contribute to improving the precision of the procedure, improve patient outcomes including aesthetic appearance, and decrease intraoperative complications.[2] Along the same line, artificial intelligence and the subfield of machine learning are expected to eventually make impacts in improving preoperative planning of the procedure by the surgeon, improve risk prediction based on patient factors, and reduce human errors.

Genetic research is promising for advancements toward non-surgical treatments. Identifying genetic pathways that are the cause for premature suture fusion would contribute to the development of therapies that delay or prevent fusion allowing the patient to avoid surgery.[2] Advancements in non-surgical methods would obviously lead towards a decrease in the number of minimally invasive procedures resulting in improved patient outcomes and lowering healthcare costs.

ENDOSCOPIC-ASSISTED REPAIR OF SAGITTAL CRANIOSYNOSTOSIS

Overview

Despite decades of research the surgical community has not been able to agree upon an ideal treatment that provides advantages over other surgical techniques. The challenge in the field of craniosynostosis surgery is to arrive at what is the best surgical technique or combination of techniques – open, endoscopic-assisted craniosynostosis surgery (EACS), spring-assisted – that provides the best outcomes for the patient.[6] However, the goals continue to focus on establishing surgical techniques that reduce morbidity, provide satisfying cosmetic results for the patient, and lower costs. This has led to the development of the endoscopic-assisted procedure that is based on making small incisions to minimize the invasiveness of the procedure.

The two primary goals of the endoscopic procedure are the same as with open procedures:

- aesthetic correction of the cranial deformity to give the child a completely normal appearance, and
- prevention or treatment of neural disorders when increased intracranial pressure has been diagnosed.

The two goals are achieved through the endoscopic approach by:

- improving aesthetic appearance,
- increasing the volume of the cranium,
- normalizing the dynamics of the skull,[3] and
- redirecting the course of the cranial growth in a normal manner.[3]

The advantages of EACS include:

- Less scarring
- Shorter operative time: EACS tends to have a shorter operative time as compared to open procedures, but depends on factors such as complexity of the procedure, the number of cranial sutures that are involved, and the surgeon's experience in performing the procedure.
- Decreased recovery time: Patients are usually taken to the neurosurgical ward without being required to spend time in the Pediatric Intensive Care Unit (PICU) and discharged within one to two days.
- Two small skin incisions
- Decreased healthcare costs
- Decreased blood loss during surgery: Typically, EACS has minimal blood loss compared to open procedures. The estimated blood loss will vary depending on the surgical technique used by the surgeon, the patient's medical condition, and complexity of the procedure.[3] The need for blood transfusions is based on the same factors to obviously include the amount of blood loss that is occurring during the procedure. Because of the low blood volume of pediatric patients even the loss of a small amount of blood can have a significant effect.

The surgeon performs a suturectomy and short parietal osteotomies with EACS to treat scaphocephaly. The procedure itself is not used to reshape the cranial vault but meant to relieve the effect scaphocephaly has on the growth of the skull. The procedure relies on the ability of the developing brain to reshape the skull to achieve normocephaly. Additionally, remodeling of the skull is achieved with the use of a postoperative helmet that provides support by inhibiting the longitudinal growth vector of the skull to encourage the lateral growth vector.[3] One factor many surgeons agree, based upon the research by Jiminez and Barone, is that the procedure, whether open or endoscopic-assisted, is best performed on infants under six months of age, as there is rapid growth of the brain soon after and the bones are softer and mold easier.[2,3] Early surgery allows bone ridges that develop along the osteotomies to be covered by the overgrowth of bone tissue during the first year of life.[3] If surgery is performed on children after six months of age, particularly after one year of age, the bone defects can be palpated and remain visible.[3]

The effect of surgery on the long-term neurocognitive outcomes of patients has not been clearly established by studies. The probability of neurocognitive impairment appears to be low with one study reporting it to be up to 9% but presents as being slight symptoms.[7] Study results are contradictory thereby lending them to the inability to establish long-term definitive conclusions. However, the key factors previously discussed – age at the time of surgery, surgical technique, type of anesthesia, surgeon's experience, and duration of the surgery, all influence the patient outcomes with age combined with surgical technique being very important. Several studies have shown that children undergoing surgery within the first six months of age have better outcomes compared to children operated on at a later age.[8-10] Age combined with surgical technique has a significant final impact on neurocognitive outcomes.[10]

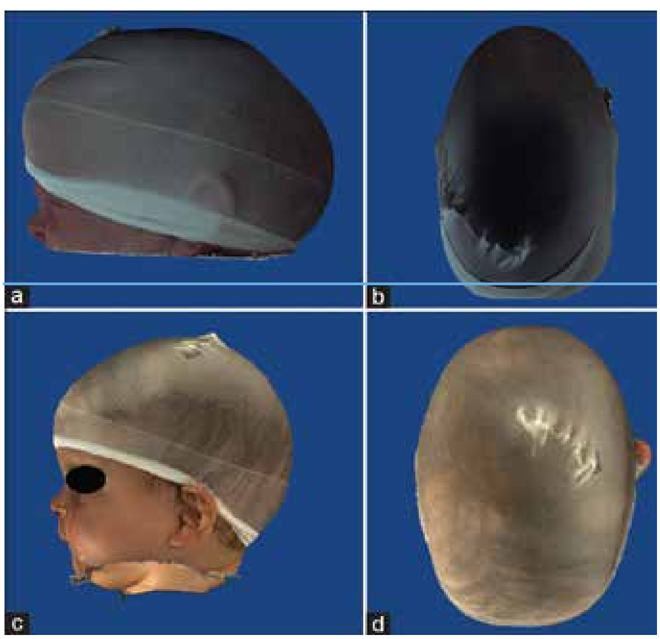

Figure 2 *(a and b) preoperative 3D stereophotogrammetry of a scaphocephalic patient. (c and d) 11 months postoperative 3D stereophotogrammetry of same patient. Frontal bossing has declined, occipital pointing is resolved, mid-parietal breadth normalized*

Imaging Studies

Imaging studies are not primarily used for diagnosing craniosynostosis, but to aid the surgeon in preparing for the procedure. Details regarding diagnosis are presented in "Craniosynostosis, Part 1" in the December 2025 issue of *The Surgical Technologist*. The gold standard for both the diagnosis and procedural preparation is a 3D CT scan with 3D stereophotogrammetry becoming the commonly used imaging modality. This system offers the advantages of minimal invasiveness, fast capture speeds, high degree of accuracy and precision, ability to save the images for ensuing analysis, expanded surface coverage, elimination of radiation exposure, and reliability of data procurement.[11] The systems are capable of accurately reproducing the surface geometry of the face to include realistic color and texture to produce a lifelike rendering **(Figure 2)**.

Upon review of the imaging studies, the surgeon will note the opacities in the cranial bone that can be present in infants to avoid breaking or perforating the bone.[6] The surgeon will also take note if the affected cranial suture is incompletely synostotic to avoid unnecessarily removing the open part of the cranial suture. Additionally, this helps the surgeon to avoid causing a dural laceration because dura is more firmly attached to the open suture as compared to the synostotic suture.[12] Thickness of the bone that can be irregular is noted by the surgeon because they use a pair of heavy-duty scissors to cut the bone. Lastly, the surgeon will plan skin incisions based on all factors.

Anesthetic Intraoperative Monitoring, Antibiotic Prophylaxis, and Postoperative Care

Standard anesthetic monitoring techniques are used including blood loss monitoring, electrocardiography, noninvasive blood pressure monitoring, pulse oximetry, and temperature monitoring.[12] Because blood loss is minimal and surgery time is shorter as compared to open procedures, arterial and central venous lines are not necessary with two peripheral intravenous (IV) lines being sufficient. Antibiotic prophylaxis, usually cefazolin, is administered prior to the skin incisions.[6] Because of the short operative time, averaging 30 – 60 minutes, and minimal blood loss, there is no need for a urinary catheter to be inserted during or after surgery.[6]

The patient typically does not need to be transferred to the PICU and as previously noted, the infant is discharged on the first or second postoperative day. Hemoglobin and hematocrit levels are closely monitored immediately beginning at the end of the procedure and for approximately six to eight hours postoperatively.[6] Additionally, a complete blood count is checked four hours postoperatively.[12] Postoperative pain is controlled with acetaminophen and low-dose morphine administered through the IV until the first postoperative day.[6]

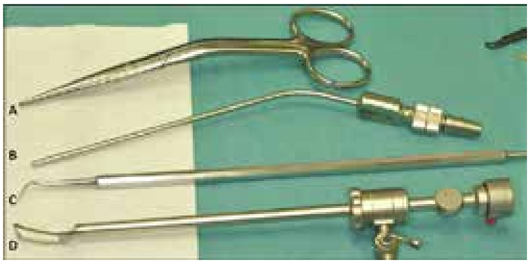

Figure 3 *Instruments commonly used in EAC surgery. A: bone cutting scissors, B: small suction device, C: bended spatula for dura dissection, D: 0-degree endoscope with footplate*

Instrumentation and Intraoperative Hemostasis

Except for the addition of endoscopy instruments and equipment, the typical neurosurgical instrumentation will be needed for the procedure **(Figure 3)**. The surgeon will use bipolar and suction cautery. A 0-degree or 30-degree endoscope without irrigation or suction, depending on surgeon's preference, with footplate attached is used. A separate small diameter suction device, such as a 10 French Frazier suction tip, is positioned parallel to the endoscope to aspirate blood. The craniectomy is performed with a pediatric craniotome and the opening enlarged with Kerrison rongeurs. For scaphocephaly procedures, the surgeon will need strong angled bone cutting scissors with blunt tips to remove the segment of bone. A scalp dural retractor will be used when the surgeon is cauterizing the bone edges towards the end of the procedure.

Depending on the surgeon's preference, there can be differences in the instrumentation for the endoscopic procedure. Some surgeons may use an optical dissector and ultrasonic bone cutter instead of the angled bone cutting scissors.[13] Additionally, when the surgeon makes the incision over the anterior fontanelle rather than at the posterior prelambdoid region, rongeurs can be used for making the burr hole rather drilling because the bone in this region is thinner and avascular.[13]

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Depending on surgeon's preference, Gelfoam™ sponges (may be soaked in thrombin, again based on surgeon's preference), bone wax, or FloSeal® Hemostatic Matrix (Baxter International Inc., Deerfield, IL, USA) will be used after the craniectomies have been performed to control bleeding at the bone edges.[6,12] Many surgeons use FloSeal® Hemostatic Matrix to control bleeding from the epidural space and bone edges which is injected towards the end of the procedure.[6,12] The CST is responsible for preparing the solution – the kit contains the needed "ingredients" including the pre-filled sodium chloride solution syringe, thrombin vial, and FloSeal® gelatin matrix. The following link is to a page provided by Baxter International Inc. with instructions on how to mix the solution: https://advanced-surgery.baxter.com/sites/g/files/evysai3391/files/2019-09/Floseal%20Fast%20Prep%20QRG_US.pdf

Sphinx Surgical Position

The sphinx patient position is often used for the two-incision procedure **(Figure 4)**. It was first described by Suzuki et al in 1984.[14] Since then it has undergone adjustments and is used in many surgeries requiring access to the top of the head and posterior aspects of the cranial vault. It is indicated for calvarial reconstruction and strip craniectomy, both open and endoscopic procedures.[15]

The patient is placed in a prone position with the head supported at the malar area, typically using a horseshoe headrest **(Figure 4)**. Reverse Trendelenburg at 30 to 45 degrees is utilized to prevent cervical hyperextension

Figure 4 *Sphinx surgical patient position*

and allow improved access to the anterior and posterior regions of the skull.[15] The arms must be well padded, and the torso is stabilized by applying a padded strap across the gluteus or thighs. When the patient has been positioned, the anesthesia provider will check breath sounds bilaterally. The surgical team should periodically assess the patient's position throughout the procedure.[15]

The advantages of the sphinx position include full access to the cranial vault and is particularly advantageous in patients with scaphocephaly with excellent anterior and posterior approaches.[15] It also facilitates venous return providing an improved view of the surgical field. The disadvantages of the position includes extra caution from the surgical team when positioning the infant to prevent inadvertent extubation, because the position involves hyperextension, it is contraindicated in patients that present with a cervical spine abnormality.[15]

It has a risk of venous air embolism due to the surgical site is superior to the right atrium,[15] of the large head of infants as compared to neonates and adults, of the potential for an increase in blood loss for craniosynostosis procedures, and the blood loss can contribute to a decrease in central venous pressure.

Surgical Procedure

The craniectomy is typically performed from the anterior to the posterior fontanelle but may be adjusted if a section of the suture line is open. The endoscopic approach is performed through two small incisions on either end of the fused suture; however, the single incision procedure has also gained popularity. The single incision offers several advantages – the cosmetic advantage of only one incision, decreased risk of wound complications, early control of emissary veins, and easier identification of the lambdoid sutures. Whether one- or two- incisions, the width of the strip of bone to be removed is according to the surgeon's preference with or without barrel stave osteotomies, also referred to as wedge osteotomies, performed posterior to the coronal sutures and anterior to the lambdoid sutures. Surgeons may remove a 2.5 to 6 cm width strip of bone.[3,6,16]

The following are links to videos of the one- and two-incision procedures.

- One-incision: https://pmc.ncbi.nlm.nih.gov/articles/PMC9542309/
- Two-incision: https://vimeo.com/513939623

The two-incision procedure will be described. Prior to the start of the procedure the surgical team should confirm with the blood bank that emergency blood is available in case transfusion is necessary. The team should also pre-warm the operating room and keep it warm throughout the procedure to aid in maintaining the core temperature of the infant.

1. Patient is transported into the OR and placed under general anesthesia in the supine position. Two peripheral IVs are placed and secured. Tranexamic acid is administered IV to aid in controlling bleeding.[16] An antibiotic, usually cefazolin, administered IV.[6,12]
2. The patient is then carefully turned and placed in the prone sphinx position with the head positioned in the headrest, aligning the sagittal suture with the horizontal plane.[15]
3. Using the marking pen, two skin incisions are indicated. Each incision is two to three cm in length. The first incision is behind the anterior fontanelle and the second incision is in front of the lambda (Figure 5).
4. Local anesthesia is injected into the area of the skin incisions, usually lidocaine 2% with or without epinephrine depending on surgeon's preference.[6]
5. Using a #15 knife blade, the surgeon makes the two skin incisions. The incision is carried through the scalp with electrosurgery.[12]
6. Next, the surgeon separates the galea from the underlying pericranium to connect the two skin incisions.[12,16]
7. Through the anterior incision, using electrosurgery on the cutting cycle, the periosteum is incised, and the dissection is carried down to the anterior fontanelle.[6,12,16]
8. The burr hole is made with the pediatric craniotome.
9. A curette and Kerrison rongeur are used to widen and

"Depending on the surgeon's preference, there can be differences in the instrumentation for the endoscopic procedure. Some surgeons may use an optical dissector and ultrasonic bone cutter instead of the angled bone cutting scissors."

complete the osteotomy. The osteotomy is 2.5 cm, the same length as the skin incision.[16] Gelfoam® sponges possibly soaked with thrombin, bone wax, or the FloSeal® Hemostatic Matrix, depending on surgeon's preference, is used to control bleeding from the bone edges.

10. Steps seven through nine are repeated through the posterior skin incision.
11. The endoscope and suction device are inserted through the anterior incision under the bone and advanced towards the posterior incision. The placement of the suction device also contributes to dura dissection.[6,16]
    a. The footplate on the endoscope contributes to providing visualization of the operative field under the bone, identification and bipolar coagulation of emissary veins to control bleeding, and protection of the dura mater during bone resection.[6]
12. The dura dissection from the overlying bone and synostotic suture is completed using curettes and Penfield dissectors.[12]
    a. The dissection is usually easily accomplished because the dura mater is barely attached to the synostotic suture.

Figure 5 *Skin incisions*

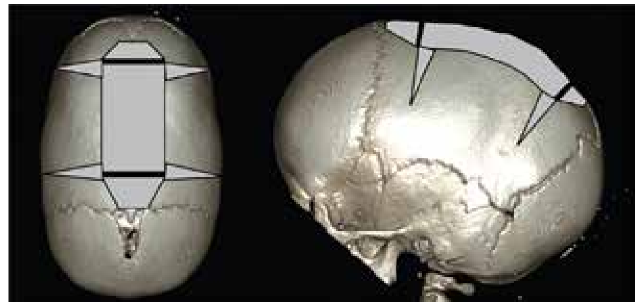

Figure 6 *3D scan showing extent of craniectomy in scaphocephaly. Thick black line indicates skin incision, grey area depicts craniectomy size*

13. Once the dura dissection is completed, the periosteum is dissected and lifted from the suture.
14. Using Jackson-Tessier bone scissors, the strip craniectomy is performed **(Figure 6)**. The strip of bone may be divided into two pieces.
15. A long clamp, such as a straight Schnidt clamp, is used to remove the strips of bone.[16]
16. The dura is stripped immediately behind the coronal sutures and directly in front of the lambdoid sutures towards the squamous sutures bilaterally.[12] Using bone scissors, the surgeon creates barrel stave osteotomies 1 cm in width behind the coronal sutures and in front of the lambdoid sutures to allow an increase in the biparietal width **(Figure 6)**.[6,12]
17. A scalp dural retractor is inserted and the bone edges are cauterized using suction cautery.
18. The syringe with Floseal® Hemostatic Matrix is inserted through the anterior incision and the solution injected to aid in controlling bleeding from the bone edges.[16]
19. No drains are necessary.
20. The scalp is closed with suture of the surgeon's preference; options include 4-0 Monocryl™ suture, 4-0 Vicryl™ or Dermabond™ applied to the skin.
21. A small compressive head bandage is applied and kept in place for 24 hours to prevent subcutaneous hematoma development.[6]
22. The patient is transported to the Post-Anesthesia Recovery Unit and then to the neurosurgical ward.
23. The patient can be orally fed three to four hours post-

"The child's head shape is corrected within three to four months postoperatively, but the skull shape can relapse if the helmet therapy is discontinued too early."

operatively and usually discharged from the hospital the next day.[6]

POSTOPERATIVE HELMET MOLDING THERAPY

The success of the scaphocephaly surgical procedure is dependent on the use of the cranial helmet. Helmet therapy controls the direction of calvarial growth.[6] The flexibility of the child's cranial vault combined with rapid brain growth provides the optimal time for skull remodeling to occur postoperatively and prevent regression of the deformity. The helmet may be one piece or two pieces depending on the preference of the orthotist and craniofacial team. Advantages of the two-piece helmet include being lighter in weight and thinner, improved aeration, easier to take on and off the child, and less movement (sliding).[6]

Three days prior to surgery the child will have a helmet evaluation completed by an orthotist. A 3D digital scanner is used to provide precise data for the orthotist to design and fit a helmet that will reshape the skull as the brain grows **(Figure 7)**. The helmet is usually delivered and fitted onto the child within seven to 11 days after surgery.[6] A typical schedule involves the child building up the amount of time the helmet is worn, for example, day one the child will wear the helmet one to two hours, remove for three hours, then repeat until bedtime. By day six the child will be wearing the helmet 21 to 23 hours a day until they are nine to 12 months old.[6] During times when the helmet is not worn, the parent is responsible for examining the child's head for redness and pressure ulcers. The child will also have regular follow-up appointments with the orthotist to check the helmet's fit and adjust if necessary.

Figure 7 *Helmets used for orthotic treatment. Left: 2-piece thermoplastic helmet used for trigonocephaly/anterior plagiocephaly. Right: one-piece resin helmet used for scaphocephaly.*

During appointments, the craniofacial team will also check the progress of the head's growth. The child's head shape is corrected within three to four months postoperatively, but the skull shape can relapse if the helmet therapy is discontinued too early.[6] The craniofacial team and orthotist will determine when the therapy can stop.[6]

FEBRUARY CE ARTICLE

To wrap up the series of articles addressing the repair of sagittal craniosynostosis, "Spring-Assisted Cranial Expansion for Sagittal Craniosynostosis, Part 3" will be published in the February edition of *The Surgical Technologist*.

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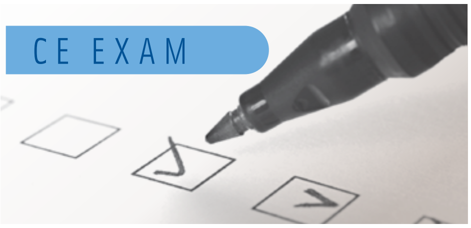

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1. How old is the child when the helmet therapy can be discontinued?
- **a.** 9 – 12 months
- **b.** 12 – 15 months
- **c.** 15 – 18 months
- **d.** 18 – 21 months

2. Which instrument is used to dissect the dura from the synostotic suture?
- **a.** Craniotome
- **b.** Bone scissors
- **c.** Penfield dissector
- **d.** Kerrison Rongeur

3. At what age should EACS be performed?
- **a.** Day after birth
- **b.** Before 6 months
- **c.** Between 9 - 12 months
- **d.** Between 15 – 18 months

4. Which of the following statements regarding the location of the two skin incisions is correct?
- **a.** Behind the anterior fontanelle; in front of the lambda
- **b.** In front of the anterior fontanelle; behind the lambda
- **c.** Lateral to anterior fontanelle; lateral to lambda
- **d.** Medial to anterior fontanelle; medial to lambda

5. Which of the following drains is inserted just prior to incisional closures?
- **a.** Jackson-Pratt
- **b.** Penrose
- **c.** Hemovac
- **d.** No drain inserted

6. A disadvantage of 3D stereophotogrammetry is exposure to radiation.
- **a.** True
- **b.** False

7. Which of the following is not used to control bleeding during the procedure?
- **a.** Bone wax
- **b.** Hemostatic matrix
- **c.** Gelfoam™
- **d.** Avitene™

8. Which of the following veins must the surgeon avoid damaging during the surgical procedure?
- **a.** Diploic
- **b.** Cerebral
- **c.** Emissary
- **d.** Dural

9. The barrel stave osteotomies are made:
- **a.** behind coronal sutures and in front of lambdoid sutures.
- **b.** in front of coronal sutures and behind lambdoid sutures.
- **c.** lateral to coronal sutures and medial to lambdoid sutures.
- **d.** medial to coronal sutures and lateral to lambdoid sutures.

10. Once the patient is placed in the sphinx position, slight Trendelenburg is used.
- **a.** True
- **b.** False

ENDOSCOPIC-ASSISTED CRANIOSYNOSTOSIS PROCEDURE *PART 2 OF 3*
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