Chapter

40 Craniosynostosis

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A 12-month-old, 9.2 kg healthy child presents for an open cranial vault repair for sagittal synostosis. All preoperative laboratory studies (CBC, electrolyte panel, coagulation studies) are normal, with the exception of an elevated PTT (46.0, normal 22.0–36.0).

Physical examination, family history, and review of systems are unremarkable. There was no history of prior general anesthesia, and his surgical history was notable for an uneventful circumcision at birth. A repeat PTT was drawn the morning of surgery and remained elevated (41.0).

DIAGNOSIS

What Is Craniosynostosis?

Craniosynostosis is a premature closure of one or more of the cranial sutures.

What Is the Incidence of Craniosynostosis in Children?

The incidence is 1:2000 births, males more commonly affected than females.

What Is the Natural Progression of Cranial Suture Closure?

The anterolateral fontanelle closes by approximately three months of age, the posterior fontanelle by three to six months, the anterior fontanelle by 9–18 months, posterolateral fontanelle by two years.

What Are the Four Cranial Sutures Involved in Craniosynostosis?

The four cranial sutures are sagittal, coronal, lamboid, and metopic. The premature closure of the sagittal suture is the most common simple craniosynostosis (50%) followed by coronal (20%) and metopic (10%) (Figure 40.1).

The growth of the skull is in the direction perpendicular to the fused suture, resulting in characteristic skull appearances.

Simple versus Complex Craniosynostosis

Simple Synostosis. Nonsyndromic or one suture prematurely closed, 80% of diagnoses.

Complex Synostosis. Syndromic or involving the premature closure of two or more sutures, 20% of diagnoses.

Which Syndromes Are Most Commonly Associated with Craniosynostosis?

Craniosynostosis can be a component of many named syndromes; however, the most common syndromes encountered are: Crouzon, Apert, Pfeiffer, Saethre-Chotzen, and Carpenter.

What Are the Common Clinical Characteristics of Apert Syndrome?

Apert syndrome is a spontaneously occurring autosomal dominant syndrome occurring in approximately 1:100,000–1:160,000 patients. A multitude of systems can be affected including:

- Cranio-facial with coronal synostosis, proptosis, hypertelorism, midfacial hypoplasia, cleft palate, choanal or tracheal stenosis (Figure 40.2).
- Respiratory with a 40–50% incidence of obstructive sleep apnea.
- Cardiac with a 10% incidence of congenital heart disease, especially ventricular septal defect (VSD) and pulmonary stenosis.
- Genitourinary with cryptorchidism and hydronephrosis.
- Musculoskeletal with syndactyly.
- Neurologic with developmental delay, and elevated intracranial pressures.

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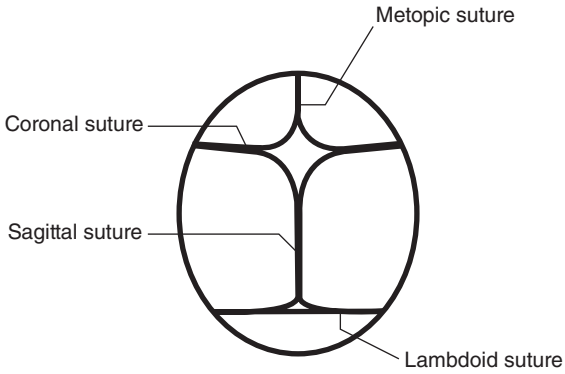

Figure 40.1 Pictorial representation of cranial sutures. Image by Estrellita and Rob Swenker, reproduced here under CC BY 3.0 license https://creativecommons.org/licenses/by/3.0/

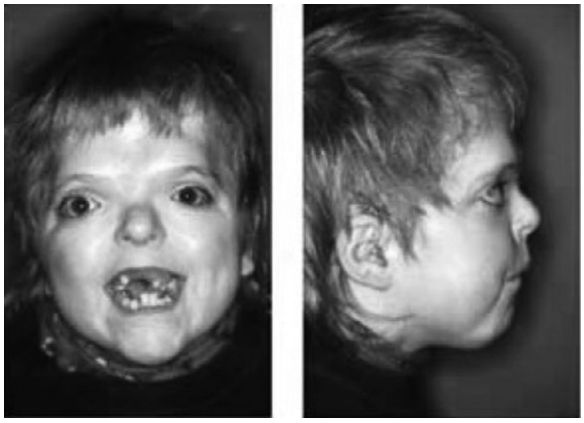

Figure 40.2 Child with Apert syndrome, hypertelorism, and midfacial hypoplasia. Reproduced from: Hohoff A., et al . The spectrum of Apert syndrome: phenotype, particularities in orthodontic treatment, and characteristics of orthognathic surgery. *Head & Face Medicine* (2007) 3:10 reproduced here under CC BY 2.0 license https://creativecommons.org/licenses/by/2.0/

What Are the Common Clinical Characteristics of Crouzon Syndrome?

Crouzon syndrome can be sporadically occurring or inherited. A multitude of systems can be affected including:

- Craniofacial including cranial synostosis (most commonly coronal and lamboid), fontal bossing, midface hypoplasia, and airway obstruction leading to obstructive sleep apnea (Figure 40.3).
- Optic atrophy occurs in roughly 20% of patients.
- Neurologic: these patients can have mild developmental delay and increased intracranial pressure.

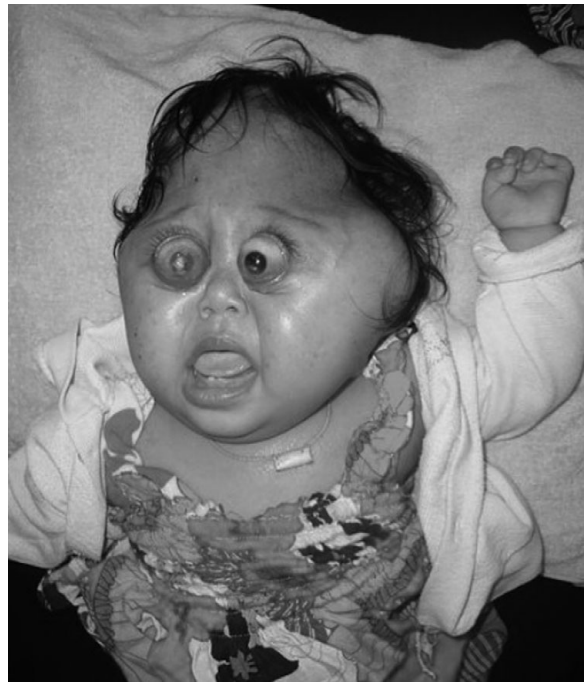

Figure 40.3 Child with a severe case of Crouzon syndrome. Reproduced under CC0 license

These patients can be difficult to mask ventilate due to their facial features.

What Are the Common Clinical Characteristics of Saethre–Chotzen Syndrome?

Saethre–Chotzen syndrome is a rare syndrome causing craniosynostosis (most commonly coronal), brachycephaly, maxillary hypoplasia, renal anomalies, cryptorchidism, and mild developmental delays. These patients can be difficult to intubate especially due to the maxillary hypoplasia.

Which Structure Is Most Commonly Associated with Syndromic Craniosynostosis?

The coronal suture is most commonly associated with syndromic craniosynstosis.

What Are Some Indications for Cranial Vault Reconstruction?

Indications for cranial vault reconstruction include poor or slowed brain growth, increased intracranial

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pressure (ICP), severe exophthalmos, obstructive sleep apnea, craniofacial deformity, and psychosocial and cosmetic reasons.

Why Do Surgeons Recommend Surgical Repair in Children by One Year of Age?

Patients without correction of craniosynostosis are at an increased risk for visual loss and developmental delay. Children repaired after the age of one year were found to have a lower IQ than those repaired prior to one year of age. In addition, better cosmetic results can be achieved if corrected during the first year of life when rapid brain and skull growth occurs.

PREOPERATIVE TESTING

What Preoperative Laboratory Tests Are Required for a Craniosynostosis Repair?

Most centers require a baseline hematocrit and blood type and cross-match as the minimum laboratory studies. Some centers perform other testing including a CBC, electrolyte panel, and coagulation studies.

What Would You Expect the Hemoglobin of a Term Neonate to Be? Two-Month-Old? Six-Month-Old? What Causes This Difference?

Neonates are born with a hemoglobin of approximately 17 g/dL composed solely of fetal hemoglobin. The increased density of hemoglobin in the newborn is a residual from the child's need to extract oxygen from its mother's blood across the placenta while in utero.

Between the two- to three-month-old period, the fetal hemoglobin present at birth has reached the end of its lifespan (approximately 90 days) and the adult hemoglobin has not reached its full production capability. Therefore, it is normal for children aged two to three months to have a hemoglobin of 10 g/dL. Adult levels of hemoglobin are reached by four to six months of age.

INTRAOPERATIVE MANAGEMENT

What Procedures Are Performed to Repair Craniosynostosis? What Are the Benefits/Risks Associated with Each?

- Endoscopic strip craniectomy: performed on children within the first four months of life, least invasive repair, requires a protective helmet to be worn after surgery, many discharged postoperative day one, may avoid intensive care unit admission (institution dependent).
- Spring-assisted cranioplasty: for sagittal suture craniosynostosis only, reduced intraoperative blood loss, short hospital stay.
- Total cranial vault reconstruction: endoscopic or open, associated with 50–100% blood volume loss, 90–100% of infants require transfusion, PICU care postoperatively, no protective helmet required (Figure 40.4).

A Z-shaped opening allows for tension-free approximation and later coverage by hair growth which improves cosmetic outcomes (Figure 40.4).

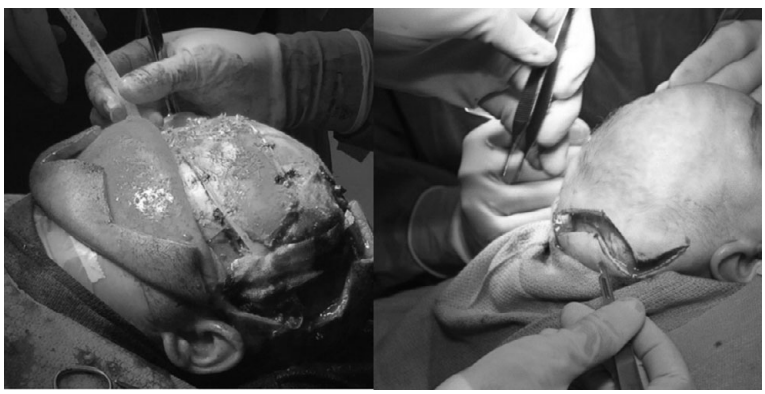

Figure 40.4 (Left) Anterior cranial vault remodeling showing the retraction of the skin and muscle and removal of bone and replacement of the remodeled skull bones with absorbable plastic spacers to allow for bony growth. (Right) Z-shaped opening allows for tension free approximation and later coverage by hair growth. Courtesy of Adam C. Adler, MD

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What Are the Anesthetic Concerns Related to Craniosynostosis Repair?

- Potential for difficult airway, especially if the child is syndromic, diagnosed with obstructive sleep apnea, has abnormal neck mobility, or midface hypoplasia.
- Blood loss: open cranial vault reconstruction is associated with a 50–100% blood volume loss. Less blood loss is associated with other repairs; however, laceration of the sagittal sinus during any type of repair can be catastrophic.
- Cardiac arrest: increased incidence, particularly in total cranial vault reconstructions, secondary to sudden massive blood loss or venous air embolism.
- Hypothermia: secondary to exposure of the head during surgery, resuscitation with large quantities of fluids/blood.
- Other organ involvement in syndromic patients (example: congenital heart disease occurs in 10% of patients with Apert syndrome)
- Increased ICP: more common in syndromic craniosynostosis, associated with greater hemodynamic instability than patients without elevated ICP, may require elevated mean arterial pressure (MAP) to maintain adequate cerebral perfusion.

What Are the Risk Factors and Treatment for Venous Air Embolism (VAE)?

Craniofacial procedures are high risk for development of VAE. VAE occurs when air is entrained into the venous system which can collect in the right ventricular outflow and impede cardiac output. Patients are at increased risk with head-up position, exposed dural venous sinuses, and relative hypovolemia.

The most sensitive method for detection of VAE is transesophageal echocardiograph. Precordial Doppler can be applied to the chest for continuous auscultation and the mill wheel murmur sound can be heard when air is being entrained.

Treatment includes stopping surgery by flooding the surgical field with saline, returning the table to a flat or head down (Trendelenburg) position. Treatment is supportive with volume resuscitation, 100% oxygen, addition of inotropic support as needed, institution of positive end-expiratory pressure (PEEP) to raise intrathoracic pressure, and advanced cardiac life support/pediatric advanced life support as needed for cardiac arrest.

How Does Positioning Influence Anesthetic Management?

- Reverse Trendelenburg: reduces blood loss, increased risk for venous air embolism.
- Trendelenburg: increased blood loss, reduced risk of venous air embolism.
- Prone: skin break down, pressure on the eyes/face/nose.
- Superman: extubation with extension of the head/neck.

What Are Some Risk Factors for Increased Intraoperative Bleeding?

Risk factors are weight less than 5 kg, age <18 months, earlier surgery date, syndromic craniosynostosis, surgical time >5 h.

What Is the Incidence of Blood Transfusion in Craniosynostosis Repairs?

Incidence is variable depending upon the surgical procedure, simple vs. complex craniosynostosis, and the surgeon's expertise. Performing a cranial vault reconstruction without resuscitation with blood products is rare; 90–100% of infants in a literature review required transfusion for this procedure. A minimum of 2 units of cross-matched packed red blood cells (PRBCs) is recommended prior to initiation of the case.

BLOOD TRANSFUSIONS

What Are the Current Recommended Ratios of PRBC:FFP:Platelets for Transfusion of a Child?

FFP and platelets are recommended if the blood loss is greater than 1 blood volume. A variety of resuscitation practices have been described for craniosynostosis repair including the use of whole blood,

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reconstituted PRBC and fresh frozen plasma (FFP), PRBC, crystalloids and colloids. Individual institutions maintain their own protocols for these procedures.

What Are the Risks of Resuscitating with Only PRBC or Crystalloid?

Dilution coagulopathy is a risk if the blood loss is greater than one blood volume.

What Surgical and Pharmacological Adjuncts Help Reduce the Rate of Transfusion?

- Preoperative management strategies: Preoperative recombinant human erythropoietin in combination with the use of cell saver results in decreased allogeneic blood transfusions. Directed blood donation from older children or from a family member may also help reduce the number of blood unit exposures.
- Intraoperative management strategies: Techniques such as acute normovolemic hemodilution, antifibrinolytics, induced hypotension, and cell salvage are used to help reduce intraoperative blood loss.
- Surgeon specific: Placement of blocking stitches or surgical clips on the skin flaps helps minimize blood loss during open cranial vault reconstruction. Additional techniques such as infiltrating the subcutaneous tissue with epinephrine, needle tip cautery, and bone wax have also aided in reducing blood loss.

POSTOPERATIVE MANAGEMENT

What Are Common Postoperative Complications Related to Craniosynostosis Repair?

Ongoing blood loss possibly requiring transfusion, cerebral edema, visual changes, cerebrospinal leak, infection, and transfusion reactions have all been reported in the postoperative period.

What Electrolyte Abnormalities May Be Present Postoperatively?

1. Hyponatremia secondary to cerebral salt wasting syndrome.
2. Hyperchloremic metabolic acidosis in large volumes of normal saline were used for resuscitation.

Suggested Reading

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