Perioperative Management of Pediatric Patients with Craniosynostosis

Jeffrey L. Koh, MD, MBA a,b,*, Heike Gries, MD, PhD a

aDepartment of Anesthesiology and Perioperative Medicine, Oregon Health and Sciences University, 3181 SW Sam Jackson Park Road, Portland, OR 97201, USA
bDivision of Pediatric Anesthesia and Pain Management, Doernbecher Children’s Hospital, Portland, OR, USA

The growth of an infant’s skull is intimately intertwined with the growth of the infant’s brain. At birth, a term newborn will have nearly 40% of his adult brain size and the nine-year-old brain and skull will be approximately 90% of its adult size [1]. A normal newborn skull accommodates this rapid growth via the presence of unfused sutures and open fontanelles. The skull has two sets of paired sutures, the coronal sutures and the lamboid sutures. There are also two single sutures, the metopic suture and the sagittal suture (Fig. 1). Premature closure of these sutures results in craniosynostosis and can result in a dysmorphic appearance if left untreated. Impaired brain growth and cognitive development may be another untoward effect of uncorrected craniosynostosis.

Although reported frequency varies, the incidence of craniosynostosis is believed to be approximately 0.6 per 1000 births [2] and the exact sutures involved differ somewhat between patients. There are a variety of ways that these different presentations can be categorized. One of the most common is nonsyndromic versus syndromic. Nonsyndromic craniosynostosis typically affects only one suture and is not associated with any other syndrome. The most common form of craniosynostosis (approximately 50% of cases) affects only the sagittal suture [1,3]. This form is also called scaphocephaly. The next most common is fusion of the coronal suture or plagiocephaly. This form accounts for approximately 20% of craniosynostosis. Finally, fusion of the metopic suture accounts for 10% of cases and is also called trigonocephaly (Fig. 2). Fusion of the lamboid suture has been
reported but is extremely rare. A familial pattern of nonsyndromic craniosynostosis has been identified and has been shown to be an autosomal dominant disorder. Two to six percent of infants with sagittal synostosis and 8% to 14% of infants with coronal synostosis are familial in nature [1].

Syndromic craniosynostosis accounts for approximately 20% of cases and usually affects two or more sutures. Over 150 syndromes have been identified that can include craniosynostosis as part of their presentation, but the most common are Apert’s syndrome and Crouzon’s syndrome. Both coronal sutures are usually affected (brachycephaly) in patients with these syndromes. In addition, patients with Apert’s or Crouzon’s syndrome may have midface hypoplasia and obstructive apnea because of their abnormal airway anatomy, which requires consideration when making plans for airway management.

Although not truly craniosynostosis, deformational plagiocephaly is another form of cranial deformity that is often discussed along with craniosynostosis. In this case, there is no suture fusion that has occurred. Rather, frequent positioning of an infant on their back results in the distinctively misshaped skull. The distinguishing feature of deformational plagiocephaly is the lack of suture fusion and a different pattern of cranial deformation as compared with true lamboid synostosis. Although initially felt to require surgical intervention, surgery is no longer felt to be indicated for this problem.

Other causes of craniosynostosis are much less common and are often termed “secondary” forms. These include metabolic disorders (hypothyroidism), fetal teratogen exposure (phenytoin, valproic acid) and certain forms of mucopolysaccharidosis (Hurler’s syndrome). Finally, congenital malformations involving the brain or skull can have associated craniosynostosis.

Researchers examining children with both syndromic and nonsyndromic craniosynostosis have recently identified a possible underlying genetic
alteration in some patients. Mutations have been found in genes involved in
the osteogenic process of the skull [4]. Although a full discussion is beyond
the scope of this article, it is clear that the development of craniosynostosis
is part of a complex process that is likely multifactorial in nature.

Craniosynostosis and cognitive function

Early intervention has traditionally been recommended in an effort to
maximize the opportunity for normal brain growth and cognitive develop-
ment. Traditionally, it has been believed that infants with nonsyndromic
craniosynostosis had otherwise normal brains and cognitive function that
would be negatively affected by the restriction caused with craniosynostosis.
There has been a growing amount of research over the past few years that
has sought to clarify the influence of craniosynostosis, brain development,
and cognitive development.
The largest body of research has attempted to explore the neurodevelopmental status of children with nonsyndromic craniosynostosis, with somewhat varied results. Kapp-Simon [5] reported a longitudinal study, evaluating mental development and learning disorders of infants and young children with single suture craniosynostosis (nonsyndromic). This study found that most of the children had mental development within the normal range for infancy; however, a high rate of learning disorders was identified. Becker and colleagues [6] found that nonsyndromic craniosynostosis was associated with cognitive, speech, and behavioral abnormalities, regardless of which sutures were fused. This study included both patients who had surgical repair and those who did not. Magge and colleagues [7] found that 50% of children with nonsyndromic sagittal craniosynostosis had learning disabilities that affected their reading or spelling. However, similar to the findings of Kapp-Simon, the children in the study group were found to fall in the normal range of intelligence. Panchal and colleagues [8] evaluated the neurodevelopment of infants with single suture craniosynostosis and children with deformational plagiocephaly. Neither group had undergone surgical intervention at the time of the study. Results indicated that both groups demonstrated delay in cognitive and psychomotor development. The investigators suggest that postintervention assessment is required to determine if these delays can be reversed with treatment.

In contrast to the studies described above, Warchausky and colleagues [9] found no difference in cognitive and motor development between children with untreated metopic craniosynostosis and normative data. The severity of synostosis did not seem to influence development negatively. In another study, Da Costa and colleagues [10] used global intellectual evaluations to test intellectual outcomes of children and adolescents with both syndromic and nonsyndromic craniosynostosis. This study found that the children with nonsyndromic craniosynostosis had no intellectual impairment, as compared with normative data. This study also observed that the majority of children with syndromic craniosynostosis were of normal intelligence.

There has been much less work evaluating the potential benefit of surgical intervention on cognitive development in patients with craniosynostosis. Virtanen and colleagues [11] examined the cognitive outcome of children who had undergone surgical repair of scaphocephaly. This study found that the neurocognitive performance of these children did not reach the same level as matched controls on follow up at school age. The investigators also found that a small subgroup of children who were repaired before 1 month of age tended to have better neurocognitive outcome, suggesting early repair may be important to long-term cognitive performance. Cohen and colleagues [12] performed a prospective study seeking to evaluate the effect of surgical intervention on cognitive development for children with nonsyndromic craniosynostosis. This study showed that the study group had mild baseline (preoperative) deficits in both motor and mental development. Improvement in motor function was found on follow up one year after
surgery, however mental function remained delayed. There was no correlation between patient age at the time of surgical intervention and outcome. Finally, Gewalli and colleagues [13] evaluated the effect of dynamic cranioplasty repair of sagittal synostosis on cognitive development. They found no difference in pre- and postoutcome measurements.

In summary, it appears that children with nonsyndromic craniosynostosis may suffer from underlying neurodevelopmental deficits that can lead to learning disabilities once school age is reached. Overall intelligence seems to be in the normal range. It is unclear whether the underlying etiology of this cognitive dysfunction is a result of abnormal brain development unrelated to the presence of craniosynostosis, a comorbid condition caused by some of the same factors that cause craniosynostosis, or the impairment of growth of an otherwise normal brain due to the premature fusion of sutures. The impact of surgery is unclear at this point; however, performing a true randomized controlled trial to evaluate the effect of surgery on outcome is difficult to do because of ethical considerations. Therefore, most clinicians recommend repair of craniosynostosis as early as feasible after the diagnosis is made.

**Anesthetic management**

*Preoperative assessment*

As with any surgery, preoperative assessment is an extremely important part of the anesthetic care for cranial vault surgery. The basics are the same as any preoperative assessment: evaluating preexisting medical conditions, medication history, allergies, problems with previous anesthetics, family history of problems with anesthetics, and a physical exam. These patients should have had a complete multidisciplinary evaluation by a craniofacial team before surgery and the anesthesiologist should take advantage of this process by reviewing the records and recommendations from this evaluation. In addition, special attention should be directed to signs of increased intracranial pressure, such as visual difficulties, nausea and vomiting, somnolence, or headaches. These findings are unusual in infants presenting with craniosynostosis, but may be seen in older children or in cases with fusion of multiple sutures [3].

Patients with nonsyndromic craniosynostosis are usually otherwise healthy infants. However, patients with syndromic craniosynostosis can have associated anomalies that will need to be considered when planning a safe anesthetic. For instance, patients with Crouzon’s syndrome or Apert’s syndrome can have very abnormal airway anatomy and may require fiberoptic intubation. In addition, identifying a history of obstructive apnea will be important in planning the postoperative care. A history of congenital heart disease should also be sought, as many syndromes, including Apert’s syndrome, can have associated congenital cardiac defects.
Laboratory evaluation usually includes a preoperative hematocrit (Hct), platelet count, and coagulation studies. Appropriate additional laboratory evaluations should be obtained based on the patient’s medical history. Probably the most important laboratory evaluation is a type and cross for packed red blood cells. The availability of an appropriate volume of packed red blood cells must be confirmed before surgery is started.

As with any preoperative evaluation, adequate time should be dedicated to addressing parental concerns and questions. The surgical repair of craniosynostosis is a major procedure and the parents deserve whatever reassurances the anesthesia team can provide. The parents should also be fully informed about the likelihood of transfusion, as well as the possibility of postoperative mechanical ventilation. Assistance from a child life specialist is often very helpful in helping parents to understand the process and ensure appropriate communication.

Intraoperative management

Many patients undergoing craniosynostosis repair will be young infants and will not require any premedication. For patients old enough to display separation or situational anxiety, a discussion with the parents about the best way to ease the transition from parents to the operating room may be helpful to both the anesthesiologist and the parents. These children may benefit from premedication, such as oral midazolam, before separation from parents, or a parent present during induction of anesthesia may be preferable. The use of nonpharmacologic interventions, such as video games, has also proven useful. A history of obstructive apnea may limit the option for pharmacologic premedication; however, nonpharmacologic means can still be used. Patients with a potentially difficult airway may also need an intravenous tube (IV) placed before induction. In this case, topical anesthetic creams can be used to decrease pain associated with IV placement.

Monitoring should include standard monitoring (American Society of Anesthesiologists guidelines), as well as blood pressure measurement with an arterial line. Central venous pressure monitoring is not routine, but can be considered should especially large blood loss be anticipated. Venous air embolism has been a reported complication of craniosynostosis repair. Fabrowski and colleagues [14] reported an 83% incidence of venous air embolism in a prospective study of 23 patients undergoing craniosynostosis repair, although most were without hemodynamic symptoms. The investigators recommended routine use of a precordial Doppler to increase the chance of early diagnosis. Careful patient positioning should also be considered to minimize the occurrence of venous air entrainment, as surgical indications allow [15].

Finally, adequate venous access is critical, with two large bore IVs being most common. For an infant less than 6 months old, 22- to 20-gauge
Catheters are adequate. Central access is not routine, but is used if adequate peripheral access cannot be obtained. Central venous access can also be considered if concern of venous air embolus is high.

There are few studies in the literature evaluating different anesthetic techniques for craniosynostosis repair. One study has reported the safe use of remifentanil during repair [16]. Another compared the use of sevoflurane and remifentanil to isoflurane and remifentanil [17]. Outcome variables in both study groups, including intraoperative physiologic parameters and time to wake up, did not differ significantly. A balanced neurosurgical technique using opioid and inhalation agents is often chosen as the anesthetic of choice. In most circumstances, the real focus of anesthetic clinical care is on management of intraoperative blood loss.

**Intraoperative blood loss management**

The most challenging part of the anesthetic management for craniosynostosis repair is managing the inevitable blood loss that occurs with these procedures. Management can be especially tricky, given that most of these surgeries are performed on small infants. Even though the majority of patients will be infants who weigh more than 5 kg, the blood volume in these infants is still very limited: only 80 mL/kg. In a retrospective study, Meyer and colleagues [18] found a red cell volume loss of 91 in plus or minus 66% of patient’s estimated red blood cell volume during the perioperative period in surgical repair of craniosynostosis. Kearney and colleagues [19] reported a mean blood loss of 24% of the estimated blood volume for sagittal suture repair, 21% for unicoronal, 65% for bicoronal suture repair and 42% for metopic suture repair. Another study reported that 96.3% of their patients received a blood transfusion and concluded that transfusion for craniosynostosis repair is almost unavoidable [20]. Finally, in a retrospective review, Eaton and colleagues [21] found that transfusion rates differed among types of suture repair, but also varied between neurosurgeons and anesthesiologists caring for the patients. Mean intraoperative transfusion in Eaton and colleagues’ investigation was 72.1 in plus or minus 55.6% estimated red cell mass.

Blood loss not only varies depending on type of craniosynostosis, but also differs with surgical technique. In general, techniques associated with more bony dissection (and therefore more blood loss) are associated with better cosmetic outcome [22]. The spring-mediated cranioplasty, a newer technique for sagittal craniosynostosis repair, might be an exception. Several investigators have found that spring-mediated cranioplasty required less blood product replacement than cranial vault remodeling and might provide the same cosmetic outcome as cranial vault remodeling [23–25]. However, other studies have shown that complex calvarial vault remodeling provides the better outcome, in comparison to less invasive procedures like synostectomy or strip craniectomy, for sagittal craniosynostosis repair [22,26].
Moreover, accurate assessment and replacement of blood loss in cranial vault repair is difficult. Most blood loss occurs during elevation of the vascular periosteum and a significant percentage is lost on the surgical gowns and drapes. Once the osteotomy is performed, blood loss is usually slow and continuous. Yet another source of bleeding can be from dural sinuses. Bleeding from the sinuses can be dramatic and require an immediate response.

**Transfusion guidelines**

There are no randomized trials of transfusion algorithms available for pediatric surgical patients, but it has been shown that institutional protocols might decrease the number of transfusions [27]. The American Society of Anesthesiologists Task Force on Blood Component Therapy excluded infants and children from the published practice guidelines in 1996 [28]. Other authorities consider adults and children older than 4 months of age equivalent, despite missing reliable data on perioperative transfusion in this population [29–31]. The British Committee for Standards in Haematology Transfusion [31] stated that for neonates and older children:

- All components other than granulocytes should be leukocyte depleted.
- Blood transfused in the first year of life should be cytomegalovirus seronegative.
- A screen filter (170μ–200μ) or alternative filtration system should be used during transfusion, for all components.
- Red blood cells stored less than 2 or 3 weeks should be used in young children.

**Red blood cell transfusion**

Red blood cell (RBC) transfusion is rarely indicated when the hemoglobin (Hb) concentration is greater than 10 g/dL and is almost always indicated when it is less than 6 g/dL. For patients more than 4 months of age, transfusion is indicated if the intraoperative blood loss is greater or equal to 15% of the total blood volume, and if the hematocrit is less than 24% in the perioperative period, with signs and symptoms of anemia [29]. Using the formula of desired Hb (g/dL) – actual Hb × weight (kg) × 3, will result in an appropriate volume (usually 10–20 mL RBC/kg).

In the intraoperative setting, it is useful to estimate the maximal allowable blood loss (MABL) by using the formula MABL = EBV (Ho – Hl): Ho, where EBV is estimated total blood volume, Ho is initial Hct and Hl is the lowest acceptable Hct (in percentage). Sometimes red blood cells need to be given before the maximal allowable blood loss is reached: for example, if hemodynamic instability occurs despite adequate volume
replacement, or if rapid blood loss occurs [30]. In many institutions, red cell transfusion is begun on incision (before the patient meeting guidelines for transfusion) for cases with a high likelihood for significant blood loss, especially in young infants. This is done by experienced care teams to prevent hemodynamic instability at times when the rate of blood loss is greater than the rate at which blood can be returned to the patient (because of the relative small size of the intravenous catheters). In addition, one might decide to transfuse the whole unit of RBC once the patient is exposed to the unit, thereby minimizing the need to transfuse a second unit postoperatively.

**Platelet transfusion**

Surgical patients with microvascular bleeding usually require platelet transfusion if the platelet count is less than $50 \times 10^9/L$, and rarely require therapy if it is greater than $100 \times 10^9/L$. The usual therapeutic dose is one platelet concentrate per 10 kg body weight [29,30]. In children under 10 kg bodyweight, 5 mL/kg to 10 mL/kg of a random donor or apheresis unit should result in a rise of platelets of 50 to $100 \times 10^9/L$.

It is useful to use ABO matched platelets, especially in small children, where the plasma volume is relatively large compared with the patient’s total blood volume [30].

**Fresh frozen plasma**

Fresh frozen plasma (FFP) is indicated for correction of microvascular bleeding when prothrombin and partial thromboplastin times are greater than 1.5 times normal, or for correction of microvascular bleeding in the setting of massive transfusion when coagulation tests cannot be obtained in a timely manner. It should be given in doses calculated to achieve a minimum of 30% of plasma factor concentration, which is usually achieved with administration of 10 mL/kg to 15 mL/kg FFP. Reference values for coagulation tests are essentially identical for children older than 6 months and adults. Nevertheless, the activated partial thromboplastin time is often prolonged in the first 6 months of life because of the lower concentrations of coagulation factors IX, X and XI in newborns and young infants.

**Cryoprecipitate**

Cryoprecipitate should be considered when microvascular bleeding is present and fibrinogen levels are below 80 mg/dL to 100 mg/dL. One unit of cryoprecipitate per 10 kg body weight raises plasma fibrinogen concentration by approximately 50 mg/dL, in the absence of massive bleeding or consumption.

Regardless of the kind of craniosynostosis repair, the transfusion of red blood cells is very common, whereas the need for platelets, fresh frozen
plasma and cryoprecipitate is rare. The risks of transfusion are well known. Besides transfusion-related morbidity from posttransfusion hepatitis, acquired immunodeficiency syndrome, hemolytic transfusion reactions, and allergic reactions, transfusion-related acute lung injury and leukocyte-platelet allogenic immunization are complications associated with massive and rapid transfusions. Rapid blood transfusion in an infant can result in hyperkalemia and cardiac arrest, primarily because of the high concentration of potassium in stored blood [31]. Furthermore, coagulopathy is associated with blood loss approaching 1.5 times of estimated blood volume [32].

Strategies for decreasing homologous transfusion

Preoperative autologous donation

Preoperative autologous donation is usually not feasible in children younger than 3 years and is, therefore, very uncommon. Beside concerns related to smaller blood volumes, younger children usually do not tolerate repeated vascular access and donation procedures without deep or general sedation.

Directed donations and limited-exposure blood donor programs

When parents wish to be a directed donor for their children, it is presumably because they believe that blood from themselves is less likely to be a source of transfusion-transmitted diseases than blood from an anonymous volunteer blood donor pool. Whether this is true or not has been a source of controversy in the literature. Several studies have shown no statistically significant differences in the results of infectious disease markers (human immunodeficiency virus, hepatitis C, hepatitis B, syphilis) in directed donors versus anonymous volunteer donors. Other studies have shown an increased frequency of positive infectious disease markers among directed donors, which could be attributed to the relatively larger number of first time donors [30,33,34].

Potential risks of parental blood transfusion are chimerism and graft-versus-host diseases (GVHD). Therefore, all cellular blood components obtained from biologic relatives must be gamma-irradiated before transfusion. In addition, it is also possible that the added complexity of providing directed donations could lead to an increased frequency of human error [35].

Acute normovolemic hemodilution

Acute normovolemic hemodilution (ANH) is a difficult strategy to use in small children because they have a lower hemoglobin level than adults, which translates to a smaller red cell volume available to dilute on a per kilogram basis. In addition, infants less than 4 to 6 months may not be good candidates because the compensatory increased cardiac output, seen in
adults during hemodilution, can be absent or decreased in infants of this age. Moreover, only small volumes of whole blood can be harvested before a postdilutional target Hct (usually 25% Hct) is reached. Hans and colleagues [36] investigated the potential benefit of ANH in the surgical repair of craniosynostosis. The investigators randomly assigned 34 infants undergoing craniosynostosis repair over a 4-year period to receive ANH or standard fluid management, and found that ANH reduces neither the incidence of homologous transfusion nor the amount of homologous blood transfused.

Perioperative blood salvage

There are several studies that have evaluated perioperative blood salvage for children undergoing craniosynostosis surgery. For instance, Deva and colleagues [37] did not find significant benefit in using autologous blood recovery and transfusion during primary cranial vault remodeling. For this study, the investigators used the Cobe-Bret 2 autologous blood recovery system for 11 subjects (mean age 8.8 months) and an equal number of consecutive comparable subjects who did not receive intraoperative autotransfusion. In addition to the autologous blood, the autotransfusion group received 34.1 mL/kg of homologous blood, and the control group 32.1 mL/kg. Four subjects did not receive recovered autologous blood because of inadequate recovery [37].

However, outcomes from other studies have shown some benefit for using perioperative blood salvage techniques. For instance, a retrospective, nonrandomized study by Dahmani and coworkers [38] found that the use of continuous autotransfusion system (CATS) was associated with a reduction in homologous transfusion during the surgical correction of craniosynostosis in infants. The use of CATS did not prevent the need for transfusion completely and all 20 infants studied received intraoperative transfusions. Using a prospective nonrandomized study design, Fearon [39] evaluated 60 consecutive children undergoing major cranial vault remodeling. The average age was 4 years old; the mean estimated blood loss was 28% of the estimated total blood volume. Only 30% of their study group required homologous blood transfusion. Likewise, in a nonrandomized prospective trial, Jimenez and Barone [40] demonstrated a cell-salvage reduced need for blood transfusion in children undergoing craniosynostosis repair.

Intraoperative blood salvage presents inherent risks, including coagulopathy, hemolysis, bacterial contamination, and damage to platelets. Studies demonstrated that cultures of blood harvested for recycling yielded positive results for up to 30% of patients. The most common microorganism isolated was coagulase-negative staphylococci. However, none of the patients who received the culture-positive autotransfusion blood showed clinical signs or laboratory findings of bacteremia [41]. Innenhofer and colleagues [42]
found, in their pilot study with adolescent and adult patients, that polymorphonuclear leukocytes, which may induce endothelial damage and increase vascular permeability or coagulopathy, are neither impaired nor activated to the priming threshold, and concluded that the use of intraoperative blood salvage is safe. There is no similar data currently available for younger children.

Autotransfusion, by using a postoperative blood salvaging system like the CBCII ConstaVac system, has the potential to be useful because of the common occurrence of postoperative ooze. Although the feasibility of this technique is not clearly established, Orliaguet and colleagues compared CATS intraoperative blood salvaging with the postoperative use of the CBCII Consta Vac system in a retrospective nonrandomized study [43]. They found the postoperative blood salvaging system was equally effective as the perioperative use of the CATS system for reducing homologous blood transfusion during craniosynostosis repair in infants weighing less than 10 kg.

**Other blood saving techniques**

*Induced hypotension*

Induced hypotension has not gained acceptance for craniosynostosis repair. Reasons include increased risk of venous air embolism and potential added hemodynamic instability associated with blood loss [3].

*Preoperative administration of recombinant human erythropoietin*

A few investigators have studied recombinant human erythropoietin (RHEPO) in craniosynostosis surgery in an attempt to decrease the need for homologous transfusion. Helfaer and colleagues [44] administered 300 U/kg erythropoietin every other day for 3 weeks to 31 children scheduled for craniosynostosis repair. The children also received additional elemental iron (6 mg/kg per day divided into three portions a day) during this period. They found that children who received RHEPO had significantly higher Hct values (43%) before surgery, compared with the control group (Hct 35%). The estimated blood loss in the erythropoietin group was about one third less than in the control group, and there was a 36% reduction in homologous transfusion for the study group. The investigators speculated that the reduction in blood loss might be caused by the higher hematocrit level and therefore associated with higher viscosity. In another prospective randomized, single blinded controlled trial by Fearon and Weinthal [45], the study group received 3 weeks of preoperative erythropoietin at a dose of 600 U/kg per week. Both the study group and the control group received supplemental iron (4 mg/kg per day). Fifty-seven percent of the patients given erythropoietin and 93% of the control group needed blood
transfusion. These studies suggest that erythropoietin may reduce the transfusion requirements, but cannot eliminate the need for transfusion.

**Combination of blood sparing modalities**

Meneghini and colleagues [46] performed a retrospective study to evaluate whether pretreatment with erythropoietin and iron combined with ANH could decrease homologous blood transfusion in craniosynostosis surgery. The study group consisted of 16 infants who were given erythropoietin at a dosage of 300 U/kg two times a week, and 10 mg/kg per day of iron for 3 weeks before surgery. Acute normovolemic hemodilution (Hct target 25%) was performed in all 16 subjects at the time of surgery. Five out of the 16 infants in the study group required homologous transfusion, while 7 out of 9 infants in the control group (no treatment) received homologous transfusion. In a similar retrospective study, Meara and colleagues [47] found that transfusion requirements were lower in the study group that received recombinant human erythropoietin and iron in conjunction with other blood conserving modalities (ANH, hypervolemic hemodilution, controlled hypotension) versus the control group, that was not pretreated with erythropoietin but was managed with blood conserving techniques. The investigators concluded that erythropoietin pretreatment combined with blood-conservation modalities was associated with a decreased need for blood transfusion. They also found that when ANH was used in combination with preoperative erythropoietin, the allowable blood loss was doubled, which might present a major advantage of preoperative erythropoietin. Finally, Rohling and colleagues [48] found that a multimodal strategy using erythropoietin, preoperative autologous donation, ANH, intraoperative blood salvage and deliberate hypotension for craniofacial procedures in a large cohort of children and adults (between 8 and 71 years) resulted in a significant reduction in homologous transfusions. Interestingly, they also reported that no transfusion was required in any of the subjects who received erythropoietin in combination with other blood conserving modalities. This finding is similar to the results of Helfaer and colleagues [44] noted above.

**Colloid versus crystalloid fluid replacement**

In addition to blood replacement, children undergoing craniofacial surgery require additional fluid administration to provide maintenance fluid, replace third space losses, and potentially to replace a portion of the blood loss. The decision about using colloid versus crystalloid is always present, but unfortunately there are few studies comparing colloids versus crystalloids in the pediatric population undergoing surgery. Oca and colleagues [49] did perform a randomized trial in newborns with acute hypotension. They found that administration of normal saline was as effective as 5% albumin for the treatment of neonatal hypotension.
Studies in the adult population have failed to clearly support the benefit of colloid replacement as compared with crystalloid [49–51]. In their consensus statement, the American Thoracic Society stated that the influence of albumin and synthetic colloids on microvascular integrity is still uncertain [52]. Jones and colleagues [53] investigated the influence of crystalloid and colloid replacement solutions in ANH and found changes in factor VIII, activated partial thromboplastin time, and thromboelastography in the group receiving hetastarch and dextran 70. The investigators hypothesized that these changes may attenuate hypercoagulability related to surgery.

In light of the lack of empirical evidence, it seems most prudent in the circumstance of craniofacial reconstructive surgery to replace blood loss with packed red blood cells if necessary, and to use crystalloid for other fluid replacement. Colloid can be used for acute volume resuscitation when packed red blood cells are not indicated or not available.

Postoperative course

Postoperative management varies by surgeon and institution. In most circumstances, the patient can be extubated at the end of surgery and transported to the perioperative anaesthetic care unit. If the patient has been particularly unstable, has undergone an unusually lengthy surgery, or has a history of a difficult airway, it may be prudent to leave the endotracheal tube in place for a period postoperatively to allow more time for stabilization. The patient’s hematocrit should be followed postoperatively, as continued oozing is common. Coagulation studies may be indicated should postoperative bleeding be excessive. Often, the patient will undergo a postoperative computed tomography at some point after leaving the operating room.

As with any postoperative patient, pain management is crucial. Appropriate dosing of an opioid is usually required, with follow-up to ensure adequate analgesia is achieved. A careful balance may be required for patients with a difficult airway or obstructive apnea. Patients are routinely watched in the pediatric intensive care unit for at least 24 hours, to insure close nursing care. The use of nonsteroidal anti-inflammatory drugs is usually deferred because of the fear of platelet inhibition and increased postoperative bleeding. Acetaminophen can be a useful adjunct to intravenous opioids.

Summary

Craniosynostosis is a complex condition that is most likely caused by multiple influencing factors. There is now some evidence that even patients with nonsyndromic craniosynostosis may have some underlying abnormality in brain development that can lead to learning disorders or more severe cognitive disabilities. The impact of early repair of craniosynostosis on the incidence of cognitive delay remains to be clearly determined. Nonetheless,
it is the current standard of care to consider early surgical intervention for most forms of craniosynostosis. The young age of most patients, combined with the high likelihood of significant blood loss, make these some of the most challenging cases for a pediatric anesthesiologist. With proper preoperative planning, and careful attention to intravascular volume status during the case, patient outcome is usually excellent.

References


