Anesthetic Management of Craniosynostosis Repair

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Case Description:

A 6 month-old, 5 kg infant with Down syndrome and sagittal craniosynostosis presents for cranial remodeling. The baby was born at term, after which the diagnosis of ventricular septal defect (VSD) was made. The patient had a URI beginning 6 weeks prior to scheduled surgery, with runny nose and a cough for 10 days. Physical exam was remarkable for stigmata of Down syndrome and sagittal synostosis as well as a harsh, III/VI holosystolic heart murmur. Lab tests revealed a hematocrit of 27%. EKG revealed bilateral ventricular hypertrophy. Cardiac echo showed a small patent foramen ovale (PFO) and a moderate VSD with left-to-right shunting.

Questions:

1. Should any additional preoperative evaluation/testing be done? If so, what tests and why?
2. The baby has moderate wheezing after induction with sevoflurane and tracheal intubation. What should be done?
3. What monitoring and vascular access is indicated?
4. Should trans-esophageal echocardiography (TEE) be performed under anesthesia prior to surgery? If so, what findings might be contributory to the anesthetic management?
5. Blood loss during the surgery is approximately 450 ml and there is moderate “oozing” from surgical sites. What blood products should be administered? Are specific tests indicated to guide this at this point?
6. During the case the BP and ETCO2 drop suddenly. What is the differential diagnosis?
7. To what extent is this baby at risk for intraoperative stroke? From what source would this be more likely, the PFO or VSD?

Discussion:

Craniosynostosis is a relatively common disorder with an incidence of 1 in 2000 to 1 in 2500 live births. It may be associated with congenital syndromes, the commonest being Apert, Pfeiffer and Cruzon’s syndromes. Associated mid-face deformity can pose significant airway challenges, including difficult face mask and/or intubation.

Surgical remodeling of craniosynostosis is often performed before 1 year of age to facilitate normal brain growth. Surgery before 1 yr of age is associated with an IQ greater than 70 in more than 50% of children, whereas surgery beyond 1 yr of age is associated with an IQ greater than 70 in only 7%. Other indications for surgery include hydrocephalus, raised intracranial pressure and psychosocial issues (e.g. what?).

The surgical procedure may vary from strip craniectomy to total calvarial reconstruction with the duration of procedure anywhere between 1 hr to 5 hours. Strip craniectomies generally are shorter procedures with minimal blood loss, especially when done with endoscopic assistance. Total calvarial reconstruction, on
the other hand, may involve considerable blood loss, massive transfusion, DIC, venous air embolism, positional injury and hypothermia.

Attention to securing the tracheal tube during a prolonged surgery with potential head movement is essential. The available options include nasotracheal intubation (may need changing the tube to orotracheal in the middle of the procedure!), using enhanced adhesive tapes, suturing the tube to the adjacent structures and circum-mandibular fixation of the tube.

Adequate vascular access includes two relatively large IV catheters and an arterial catheter. Central venous access should be considered if peripheral IV access is limited. For all but the smallest cases blood should be in the room before surgery begins. Point-of-care testing for hematocrit, electrolytes and acid-base balance is highly desirable.

Calvarial reconstruction and sagittal craniosynostosis repair may be done with the child in the prone position. Generous padding of the pressure points is indicated to avoid pressure injuries. Fronto-orbital and midface advancements are done with the child in supine position. For all such procedures at LPCH the table is turned 180 degrees with minimal access to the child under the drapes. Careful planning is needed to ensure the monitoring cables, IV and arterial lines, bladder catheter and anesthetic circuits are free from entanglement.

Common sources of blood loss are the subgaleal tissues and the fresh ends of the cut bones. It may be impossible to accurately measure blood loss, as much of the loss is absorbed by and/or under the drapes, on sponges and in suction tubing and reservoirs mixed with saline irrigation. For predictably bloody procedures it is best advised to start transfusion at the start of skin incision and to keep up with the ongoing losses. Communication with the surgeons prior to and during surgery regarding blood loss is essential. Blood conservation techniques may include pre-operative autologous and directed blood donation, isovolemic hemodilution, preoperative erythropoietin therapy, intraoperative red cell salvage, use of antifibrinolytics and induced hypotension. Splitting blood units may help conserve this precious resource.

Because the large head of the child is exposed for a long time, considerable body heat is lost from evaporation and blood loss from the wound during surgery. Measures to prevent hypothermia include adequate covering of extremities, elevated room temperature and use of forced air warming blankets and IV fluid warmers. Core body temperature should be continuously monitored.

Extensive exposure of osteal veins and head elevation to reduce venous congestion increases the risk for venous air embolism (VAE) in these procedures. It has been noted that the incidence of VAE could be as high as 85% (30-85%); the incidence of clinically significant air embolism is, fortunately, only1-2%. Methods of monitoring for VAE include close attention to vital signs (sudden changes in heart rate, rhythm and blood pressure), capnography, endtidal nitrogen analysis, transthoracic Doppler and TEE. If VAE is suspected the head should be lowered and the wound flooded with saline and covered immediately. Efforts to aspirate air from the heart, even when a central venous catheter is in place, are unlikely to be useful.
References:

1. Perioperative management of pediatric patients with craniosynostosis.  
Koh JL, Gries H.  

2. Comparison of perioperative blood salvage and postoperative reinfusion of drained blood during surgical correction of craniosynostosis in infants.  
Paediatr Anaesth. 2003 Nov;13(9):797-804.


4. Early management of craniosynostosis using endoscopic-assisted strip craniectomies and cranial orthotic molding therapy.  
Jimenez DF, Barone CM, Cartwright CC, Baker L.  

Venous air embolism in craniosynostosis surgery: what do we want to detect?  

6: Faberowski LW, Black S, Mickle JP.  
Incidence of venous air embolism during craniectomy for craniosynostosis repair.  