Anesthetic Management of a Recurrent Sagittal Craniosynostosis in a Child with Von Willebrand’s Disease

A. Kydes MD, B. Chon DO, A. Nicolai MD, S. Barst MD
Department of Pediatric Anesthesiology at New York Medical College at Westchester Medical Center, Valhalla NY

SUMMARY:
• Six-year old boy with von Willebrand’s disease (vWD)
• For repair of a recurrent sagittal craniosynostosis by cranial vault reconstruction
• Discussion highlights the anesthetic considerations and management of this potentially challenging patient

CASE REPORT:
• 18 kg boy with vWD who had previously undergone a sagittal craniosynostosis repaired at 7 months of age
• Presented with intermittent headaches over the past 2 months and found to have elevated intracranial pressure (ICP)
• Received Antihemophilic factor/von Willebrand factor complex (Humate-P) and sedation with midazolam preoperatively
• Standard ASA monitors and two large bore IVs were placed along with a 22G arterial line to monitor blood pressure and facilitate ABG and hematocrit monitoring
• Following induction and intubation, the patient received tranexemic acid bolus (10mg/kg) and was placed on continuous infusion (10mg/kg/hr) throughout surgery
• Sevoflurane with controlled ventilation and fentanyl for analgesia
• One unit of PRBCs and FFP given at the time of surgical incision
• Case proceeded uneventfully with approximately 400 mL of blood loss
• Patient remained intubated and transported directly to PICU
• Patient required a second transfusion of blood during his hospitalization
• Discharged home on post-op day 4

DISCUSSION:
• Craniosynostosis is premature closure of > 1 suture in infant skull
  • Occurs in approximately one in 2000 live births
  • Sagittal synostosis is the most common type
  • Most commonly occurs sporadically as an isolated condition
  • Otherwise can be part of known craniofacial syndromes (including Alpert’s and Crouzon’s)
  • Cause of non-syndromic craniosynostosis remains unknown
• On presentation findings include:
  • Symptoms of ICP - vomiting, headache, papilledema, altered mental status, or bulging fontanel
  • Obstructive sleep apnea (OSA)
  • Neurobehavioral impairment.
  • Difficult airway is a concern (especially when associated mid-facial deformities present)
• Cranial vault reconstructive surgery associated with significant blood loss
• Kearney and colleagues reported ‘sagittal craniectomies were associated with a mean blood loss of 24% of estimated blood volume (or 20ml/kg)’ and that ‘approximately 95% of patients undergoing cranial vault reconstruction required blood during surgery’
  • Rapid transfusion associated with transient electrolyte abnormalities (including hypocalcemia and hyperkalemia) - could lead to cardiac arrhythmias and arrest
• Recurrence of surgically corrected craniosynostosis increased in syndromic patients and if surgically corrected early (<6 month age)
• Anticipation, preparation and multidisciplinary approach required in recurrent craniosynostosis surgery, ESPECIALLY when there are confounding factors (such as history of coagulation disorder)

REFERENCES: