**Anesthesia for a Toddler with Osteogenesis Imperfecta undergoing Craniosynostosis Repair**

James C. Layton MD, Jennifer K. Hansen MD
Department of Anesthesiology, University of Kansas Medical Center

**Introduction:** One in 20,000 live births is affected with the autosomal dominant disorder Osteogenesis imperfect (OI) and the disease process has several implications during the peri-operative period. Through this case report of a 10 month old female with Type I OI undergoing repair of craniosynostosis, we highlight the various important peri-operative considerations when caring for these patients.

**Case:** VR presented at 10 months of age for open surgical repair of unilateral coronal craniosynostosis. Past medical history included Type I OI complicated by bilateral femoral fractures with minimal trauma. Family denied issues with dentition, heart defects, or hearing. Her only medication was IV pamidronate every 8 weeks (last dose was the day before surgery). Preoperative vital signs were normal and appropriate. GETA was induced by mask, and with careful in-line stabilization during direct laryngoscopy the trachea was intubated after intravenous access was obtained. Extreme care was used during all body maneuvers and positioning. Anesthesia was maintained with sevoflurane, rocuronium, and intermittent fentanyl injections. Serial arterial blood gas analysis was used to guide ventilatory and fluid management. This seven hour procedure proceeded uneventfully and the patient was extubated and taken to the pediatric intensive care unit in stable condition. She received 500 ml crystalloid fluids, 200 ml 5% albumin, and 140 ml packed red blood cells. No apparent coagulopathy developed perioperatively.

Her three month post operative surgical follow up visit suggested the patient was doing well with no apparent complications.

**Discussion:** Osteogenesis imperfecta is a rare autosomal dominant disease with variable clinical manifestations secondary to a defect in the production of type 1 collagen. There are 4 types of OA, type I is the most common and usually milder with types III and IV being much more severe and type II being lethal in the perinatal period. The collagen defect results in blue sclera and extremely brittle bones. Fractures are very common with the femurs being particularly susceptible. In the perioperative setting it becomes very important to handle transfers and positioning with extreme care. Even non-invasive blood pressure cuffs and succinylcholine-induced fasciculations can result in fractures. Airway management is also very important given the risks of cervical spine instability, loose dentition, and risk of fracture to the mandible. Some patients may have kyphoscoliosis creating a restrictive ventilatory pattern. Associated cardiac abnormalities include patent ductus arteriosus, aortic regurgitation, and septal defects. Preoperative coagulation status should be considered as some patients may have impaired platelet function that is sometimes amenable to desmopressin.

**Anesthetic considerations:**
- Brittle bones increasing fracture risk
- Abnormal platelet function
- Kyphoscoliosis decreasing chest wall compliance
- Cervical spine involvement necessitating special care during airway management
- Temperature elevation

**Serial preoperative CT images of the skull in this patient showing the skull asymmetry.**

References: