[N-74] Anesthetic Challenges for Craniosynostosis Repair in a Patient with Beare-Stevenson Syndrome

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Introduction: Beare-Stevenson Syndrome (BSS) is an extremely rare craniofacial syndrome caused by a mutation in fibroblast growth factor receptor (FGFR) 2, and consists of craniosynostosis resulting in a cloverleaf skull and hydrocephalus, ocular proptosis, fragile vasculature, palate abnormalities, hypertelorism, and acanthosis nigricans. Anesthetic challenges include difficulties in airway and ventilator management, increased intracranial pressure (ICP), challenges with intravenous access and positioning due to redundant, corrugated skin (cutis gyrate), and risk of corneal abrasions. These patients present for craniosynostosis repair earlier due to multiple suture involvement and have the potential for significant blood loss due to the FGFR mutation causing abnormal angiogenesis, making careful anesthetic planning essential (1).

Case: A 12 week old, ex-28 week, 3.8kg male with BSS presented for anterior and posterior cranial vault expansions for fused bilateral lamboid and saggital sutures. Pertinent history included severe chronic lung disease, laryngomalacia with tracheostomy at birth, and significant ocular proptosis (Fig.1). He required early surgical repair due to elevated ICP, but had poor respiratory mechanics to tolerate an extensive procedure. With his limited respiratory reserve and early age for surgical repair, planning was critical. Anesthetic preparation included placement of large bore venous access and an arterial line with use of ultrasound to assist with the challenge of his redundant tissue structure and fragile vessels. The tracheostomy was replaced with a microcuffed endotracheal tube (ETT) sutured in place; the distal position of the cuff on the microcuffed ETT decreased the risks of mainstem intubation and of dislodgement. Blood products were immediately available and an aminocaproic acid infusion was used in anticipation of coagulopathy. Meticulous attention was paid to proper prone positioning due to ocular proptosis and limited room for movement of the ETT in a patient with limited respiratory reserve. Shortly after incision, due to the abnormal vasculature and extreme vascularity of the tissues, bleeding resulted in tenuous hemodynamics requiring transfusion of greater than one blood volume and initiation of vasopressors. Discussion between the anesthesia and surgery teams led to the decision to stage the surgery due to the patient’s limited cardiovascular and respiratory reserve.

Conclusion: Craniosynostosis repair is a major surgery in otherwise healthy infants and is especially high risk in syndromic patients. Patient with BSS present unique challenges above and beyond those expected for this procedure, requiring careful planning and continuous interdisciplinary communication to ensure optimal patient safety.

1 Upmeyer S, et al. Pediatr Anesth 2005;15;1131-6.