Fontan Physiology, Posterior Spinal Fusion
& ε-Aminocaproic Acid

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**Introduction:** Patients with Fontan (single ventricle) physiology have unique perioperative concerns, especially in regards to preload, mechanical ventilation and underlying procoagulant and anticoagulant abnormalities. (1) These high blood loss cases like posterior spinal fusion (PSF) have led to various strategies to minimize transfusion exposure. (2) While antifibrinolytic drugs are well documented in pediatric cardiac and scoliosis surgery for minimizing blood loss, there are no such reports in the combined setting. This is a report of the use of ε-aminocaproic acid (EACA) in a patient with Fontan physiology undergoing long segment PSF.

**Case report:** A 12 – 8/12 year old 50 kg, 160.4 cm female with Fontan physiology secondary to cardiac dextroversion and hypoplastic right heart syndrome presented for T2-L4 PSF with instrumentation. Thoracic and lumbar curves measured 82 and 71 degrees respectively. She was on no medications, cardiac work-up was significant for minimal baffle leak and mean Fontan pressure of 11 mmHg with good exercise tolerance despite moderate restrictive pulmonary disease. Baseline saturation was 91% in room air with slightly increased PT 14.7, INR 1.22, GGT 55, ALT 44, and AST 41.

Transcranial evoked potential monitoring dictated the use of propofol and remifentanil infusions. EACA was initiated prior to incision with a bolus of 100 mg/kg followed by an infusion at 10 mg/kg/h. Baseline central venous pressure (CVP) supine versus prone was 11 and 16 mmHg respectively. The patient had an unremarkable anesthetic until nearly 4 hours into surgery when rapid hemorrhaging ensued. Aggressive resuscitation with blood products and dopamine maintained hemodynamic stability and improved hemostasis. In total, the patient received 1500 ml crystalloid, 2500 ml 5% albumin, 3 units packed red blood cells, 5 units fresh frozen plasma and 1120 ml cell saver blood with estimated blood loss (EBL) of 3000 ml.

Given the large amount of fluid resuscitation, the patient was taken to the cardiac intensive care unit (CICU) intubated. After demonstrating hemodynamic and respiratory stability, the patient was extubated 5 hours after surgery, discharged from the CICU on post-operative day (POD) 1 with nasal cannula oxygen and antihypertensive medication, and discharged home on POD 6 with no sequelae. No further blood products were required postoperatively.

**Discussion:** Prior reports have documented increased blood loss in patients with Fontan physiology undergoing PSF. (3, 4) Our EBL, EBL/kg and EBL/vertebral segments of 3000 ml, 60 ml/kg, and 200 ml/segment are consistent with these reports. Given that duration of surgery and increased number of vertebral segments are independent risk factors for intraoperative blood loss, EACA was likely benefiting this patient the first 4 hours of surgery as well as into the postoperative period (no further transfusions).
Antifibrinolytics have been utilized extensively in single ventricle patients undergoing cardiac surgery without reported thromboembolic complications. EACA, a lysine analog, has been demonstrated to decrease perioperative blood loss in idiopathic scoliosis. The theoretical risk of antifibrinolytics in patients with Fontan physiology is precipitating a thromboembolic event in hypercoaguable patients. However, the advantages of decreased perioperative blood loss and transfusion requirements (and those inherent risks) with a need for a higher CVP, prone positioning and mechanical ventilation that increase blood loss in these patients likely will encourage their use. This is the first documented case of a pediatric patient with Fontan physiology undergoing PSF with EACA.

**Refs:**