INTRODUCTION
An 11 month old 10 kg Amish male patient with Simpson-Golabi-Behmel (SGB) syndrome presented for emergent exploratory laparotomy for toxic megacolon secondary to Clostridium difficile infection. The patient had a complex past medical history related to SGB syndrome, including type B interrupted aortic arch with severe left ventricular outflow tract obstruction. This was palliated with a Norwood procedure and Blalock-Taussig Shunt at age 6 days, and later transitioned to a bidirectional Glenn (BDG) procedure at age 4 months. The patient had airway obstruction requiring tracheostomy, difficult intravenous access requiring surgical line placement, inability to feed requiring gastrostomy tube, craniosynostosis with surgical repair, and Wilm’s tumor that was resected and treated with chemotherapy.

CASE REPORT
The patient initially presented with a 2 week history of diarrhea, vomiting, and abdominal pain. His condition progressively worsened over the next 3 days. In spite of aggressive fluid therapy and antibiotics, he developed coagulopathy, hematemesis, and oliguria requiring pediatric intensive care unit (PICU) transfer for sepsis management. His parents initially refused surgical intervention. With continuing medical management, the patient required escalating doses of vasopressors. Stress dose steroids were given, antibiotic coverage was broadened, and arterial and venous access were obtained. The patient worsened, developed acute renal insufficiency, and required 100% oxygen. His parents agreed to surgery despite the poor prognosis.

When the anesthesia team arrived in the PICU, the patient had systolic blood pressures (SBPs) in the 70-80’s while on vasopressors and SaO2 levels of 60-70%, down from 70-80% usual with BDG physiology. Arterial blood gases (ABGs) showed mixed respiratory and metabolic acidosis with a serum lactate level of 5.2mmol/L. Upon arrival in the operating room, SBPs were between 50-60mmHg with SaO2 of 50%. He was placed on pressure control of 28cm H2O. Positive end expiratory pressure (PEEP) was decreased from 6 to 4cmH2O with the goal of decreasing intrathoracic pressure and control of 28cm H2O. Positive end expiratory pressure (PEEP) was decreased from 6 to 4cmH2O with the goal of decreasing intrathoracic pressure and increasing pulmonary edema, which further worsened ventilation and oxygenation. This resulted in decreased oxygen delivery, increasing lactate acidosis, and death of the patient.

Due to potential catecholamine depletion and myocardial depression, ketamine infusion was discontinued and fentanyl was given alternatively. Packed red blood cells (PRBCs) were given to replace blood loss and to maximize oxygenation. Fresh frozen plasma (FFP) was given to correct coagulopathy with an INR of 2.3. The patient slowly improved his SaO2 and SBP, likely from the combination of fluid therapy, milrinone infusion and steroid administration. Serial ABGs showed improvement in acidosis, and lactate had decreased to 3.8mmol/L by the completion of the operation.

Overnight in the PICU, the patient continued to require a large volume of fluid and PRBCs to maintain hemodynamics, and he progressed to pulmonary edema with severe oxygen desaturation. Despite an increase in vasopressor medications, his lactate level rose to 10mmol/L. In consultation with the PICU and surgical teams, the parents made the decision to withdraw support and the patient died.

DISCUSSION
Although this patient had a rare syndrome and complex medical history, the treatment of sepsis in a patient with palliated congenital heart disease may be an expected occurrence for the pediatric anesthesiologist. BDG is the second palliative surgery to correct single ventricle diseases prior to Fontan procedure. The superior vena cava (SVC) is anastomosed to the pulmonary artery, which helps decrease the volume load on the ventricle. The blood from the SVC that passes through the lungs is oxygenated and mixes in the ventricle with deoxygenated blood returning from the IVC. The blood pumped to the body is therefore not fully oxygenated. Elevated PVR decreases the amount of oxygenated blood returning to the ventricle.

Due to the body’s higher oxygen demand during sepsis, patients with BDG physiology rely on an increased cardiac output (CO) to provide increased oxygen delivery. In our case, CO was supported through volume and inotropic support. We attempted to decrease PVR through supplemental oxygen, decreased PEEP and correction of acidosis. Although we were able to stabilize this critically ill patient with a poor prognosis intraoperatively, the continued need for volume postoperatively to maintain CO ultimately resulted in increasing pulmonary edema, which further worsened ventilation and oxygenation. This resulted in decreased oxygen delivery, increasing lactic acidosis, and death of the patient.