Anesthesia for Emergent Noncardiac Surgery in Neonates with Unpalliated Hypoplastic Left Heart Syndrome
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Introduction: Neonatal surgery for emergent correction of congenital anomalies increases the risk of morbidity and mortality in the perioperative period. Although rare, co-existing congenital heart defects make the perioperative management of these neonates a formidable challenge, especially when palliation or repair of the cardiac lesion cannot precede the noncardiac procedure. Here we describe the anesthetic management of two neonates with a prenatal diagnosis of hypoplastic left heart syndrome (HLHS) and a tracheoesophageal fistula (TEF) in one and meningomyelocele in the other.

Case Reports:
Case 1: A 2.4 kg female was delivered at 37 ½ weeks following induction of labor for IUGR. The diagnosis of TEF was made shortly after birth when a chest x-ray confirmed nasogastric tube placement in an atretic esophagus with air in the stomach. The trachea was intubated for airway control. An echocardiogram revealed a hypoplastic left ventricle with mitral and aortic atresia and a diminutive ascending aorta. Prostaglandin E₁ therapy was promptly initiated. The patient was taken to the operating room on day of life one for repair of her TEF. Prior to surgery, umbilical venous and arterial lines were placed for intraoperative monitoring. The patient was already intubated and sedated on arrival in the operating room. Anesthesia was maintained with air, oxygen and isoflurane and FiO₂ was reduced to maintain SpO₂ at about 80-85%. The patient was breathing spontaneously initially but eventually required controlled ventilation by hand. Systolic blood pressure was consistently maintained in the 50-60 mmHg range. Prostoglandin E₁ infusion was continued intra- and post-operatively. Five days later the patient successfully underwent a modified Stage I Norwood procedure.

Case 2: A 3.3 kg male was delivered at 37 weeks gestation by caesarian section. He had an intact 5x5x7cm meningomyelocele. An echocardiogram revealed a hypoplastic left ventricle, mitral atresia, double outlet right ventricle, large subpulmonary VSD, and a mildly hypoplastic aortic arch. Prior to surgery for closure of the meningomyelocele, umbilical venous and arterial lines were placed for intraoperative monitoring. The patient was induced with fentanyl 10mcg/kg and pancuronium. The trachea was intubated uneventfully and the patient was ventilated in the lowest possible FiO₂ delivered by the anesthesia machine (25%). Because the SpO₂ remained elevated, the FiO₂ was reduced to 21% by changing to a VIP Bird ventilator and circuit. After 3 hours of anesthesia, the systolic blood pressure had decreased into the 40’s and serum lactated had increased, although the SpO₂ remained in the high 80’s. Suspecting diminished systemic perfusion secondary to pulmonary runoff, the FiO₂ was decreased to 18% by adding N₂ to the fresh gas flow. The systolic pressure subsequently rose to between 50-6O mmHg and SpO₂ fell to the low 80’s indicating improved systemic perfusion. The patient had an uneventful recovery and two days later underwent successful PA band placement and atrial septectomy.

Discussion: Emergent noncardiac surgery in neonates with HLHS requires an appreciation of the complex and evolving pathophysiology of this lesion. Both congenital anomalies, TEF and meningocele, required immediate repair in order to avoid increased morbidity in an already hemodynamically compromised neonate. In both cases, we were able to manage and minimize excessive pulmonary blood flow by (1) pharmacologic therapy (prostaglandin E₁) in the first case and (2) decreasing the inspired oxygen concentration below 21% by adding nitrogen to the gas mixture in the second case. Both maneuvers provided stable hemodynamics throughout the initial surgical procedure and the subsequent period leading up to the surgical correction of the HLHS.

References:
1. Andropoulos DB et al., Paediatr Anaesth. 8:313-19, 1998