Title:
Anesthetic management of a neonate with hypoplastic left heart syndrome (HLHS) and intact atrial septum (IAS) for emergent immediate postnatal transcatheter atrial septostomy and stent placement

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Introduction:
Neonates with hypoplastic left heart syndrome (HLHS) and intact atrial septum (IAS) present with severe respiratory cyanosis, pulmonary edema, and hemodynamic instability leading to profound cardiovascular shock shortly after birth. Immediate relief of the obstruction at the atrial level is essential to allow survival. These patients are extremely difficult to manage and stabilize in the early postnatal period and present a challenge for the pediatric cardiac anesthesiologist.

Case report:
A 33 4/7 weeks gestational age male with a prenatal diagnosis of HLHS and IAS was born via emergent cesarean section in a cardiac operating room at our institution after spontaneous rupture of the membranes and signs of severe hydrops fetalis. Immediately after delivery the 2.8 kg neonate was received by the anesthesia staff and taken to the adjacent hybrid suite. His APGAR score at 1 minute was 2 (body and extremities blue, no respiratory effort, pulse of 51/min, minimal muscle tone, no response to suctioning).

Our initial attempt to bag/mask ventilate with an oral airway was difficult and required high airway pressures, leading to immediate intubation. Through the endotracheal tube surfactant 6 mg was given and ventilation improved.

An umbilical venous and arterial line were placed and initial arterial blood gas analysis on an FiO2 of 1.0 showed a oxygen saturation of 56% with a pH of 6.88, paCO2 of 140 mmHg, PaO2 29 mmHg, BE of -10.8, glucose 52 mg%, and Hb of 15.6 gm%. The patient was immediately paralyzed with pancuronium 0.5 mg, fentanyl 15 mcg were given, dopamine drip was started at 5 mcg/kg/min as well as alprostadil at 0.01 mcg/kg/min. Intravenous fluid resuscitation with dextrose 10% in ringers lactate was started and the acidosis was treated with THAM solution iv boluses.

Cardiac catheterization was performed via a left femoral vein 5Fr sheath. Under TEE guidance the atrial septum was punctured by a transseptal needle followed by balloon septostomy and 12 mm stent placement. A rapid change in the clinical condition following stent placement was noted with improvement in oxygenation, lung compliance and decreased left atrial size on TEE as well as a drop of the mean left atrial pressure from 22 mmHg to 12 mmHg. The patient was transported to the cardiac intensive care unit for recovery. Upon transport the oxygen demand had decreased to such a degree that we were able to reduce the FiO2 to 0.5 followed by weaning to 0.21 within 24h.

Discussion:
Despite increasing fetal diagnosis and immediate postnatal intervention, the survival in infants with HLHS and restrictive atrial septum remains low (1). As surgical and transcatheter techniques for safe and effective early opening of the restrictive septum are explored and developed, the anesthetic management of these patients will require research and innovation for a successful outcome.

References: