When Two Wrongs Don't Make a Right; Anesthetic Management for Congenital Diaphragmatic Hernia (CDH) in a Neonate with Hypoplastic Left Heart Syndrome (HLHS) - A Case Report

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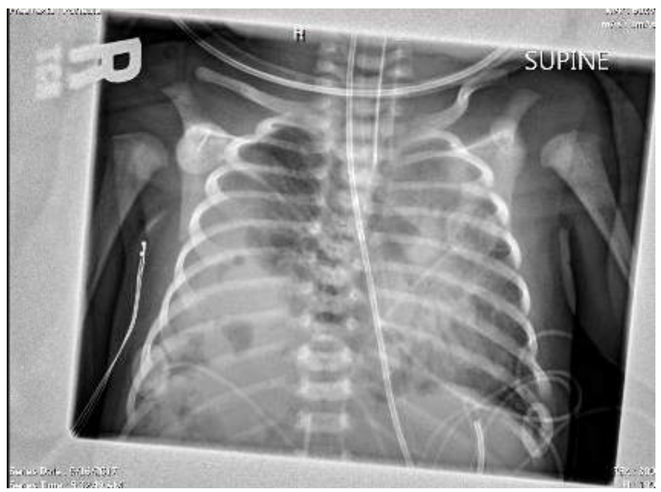
Introduction

Hypoplastic left heart syndrome (HLHS) is a congenital cardiac anomaly associated with high morbidity and mortality. The focus of perioperative management is on maintaining adequate pulmonary-to-systemic circulation ratio while keeping a low inspiratory fraction of oxygen (FiO2) (1). Congenital diaphragmatic hernia (CDH) results in various manifestations of pulmonary hypertension, with the major problem often encountered is persistent fetal circulation (PFC) due to high pulmonary arterial pressures. Therefore, neonates with CDH are often treated with a high FiO2 (1). Given competing cardiorespiratory goals, the management of combined HLHS and CDH can presents challenges. We present a case of CDH repair in a neonate with multiple comorbidities including HLHS.



Case

A 13 day old female neonate born at 37 weeks was transferred to our institution from an outside hospital. At the previous institution, she was deemed inoperable because of her co existing, competing medical conditions. Medical abnormalities included CDH, coarctation of aorta, double outlet right ventricle complicated by HLHS, syndactyly of fingers and choanal atresia. Upon transfer she was immediately admitted to the cardiac intensive care unit (CCU) in preparation for her surgery. Ultrasound guided arterial and central access was obtained. Echocardiogram showed preserved right ventricular systolic function and severe pulmonary hypertension. The patient was intubated and mechanically ventilated from outside hospital. Patient remained on prostaglandin infusion to maintaining potency of the ductus arteriosis. Preoperative discussion with neonatology, pediatric surgery and anesthesia, agreed upon an open repair versus a laparoscopic approach. Induction of anesthesia was conducted with rocuronium and fentanyl. Anesthetic maintenance was achieved with infusions of midazolam, fentanyl and dexmedetomidine with intermittent bolus doses of neuromuscular paralytic agents. To minimize hemodynamic perturbations, a total intravenous anesthetic was utilized with avoidance of volatile anesthetic agents. Frequent arterial blood gasses were drawn to monitor patient's pH status, hemoglobin and electrolytes. The CDH repair was successful without any catastrophic intra operative events, despite the diaphragmatic defect encompassing almost the entire right heme-diaphragm. Patient received intermittent PRBC transfusion to maintain hematocrit between 40 and 45%. No vasopressors or NO was utilized during the case, but were available. At the end of the procedure, the patient was placed on a paralytic infusion in continuation with her sedatives and transferred smoothy back to the CCU.

Conclusion/Discussion

The isolated presence of congenital cardiac anomalies presents challenges for the pediatric anesthesiologist in the perioperative setting. The addition of co-morbidities often times further complicates the perioperative management. The presence of CDH with congenital heart disease presents a unique challenge with an inherent high risk for mortality. Although HLHS occurs in 7.5 percent of infants with congenital heart anomaly, the incidence of associated extra- cardiac abnormalities is low in patients with HLHS (1). In an analysis of 122 cases of HLHS, no patient was found to have associated gastrointestinal disease(2). Furthermore, though CDH is sometimes associated with heart anomalies, it's association with HLHS is rare (2,3). Minimal literature or reference support exists for managing this unique constellation of co-morbidities. Balancing analgesia, amnesia, and muscle relaxation while ensuring adequate pulmonary-to-systemic circulation was the primary anesthetic goal. Avoidance of volatile anesthetic helped eliminate sudden changes in systematic vascular resistance. The surgical outcome was successful and the patient remains critically ill due to her co-morbidities, but stable and awaiting cardiac palliation.

References

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