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Perioperative management of a child with hypoplastic left heart syndrome following the hybrid stage I procedure presenting for laparoscopic gastrostomy tube placement

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Keypoints

1. Hypoplastic left heart syndrome (HLHS) is a complex congenital heart condition which includes abnormal development of left-sided cardiac structures leading to inadequate systemic perfusion following post-natal closure of the patent ductus arteriosus (PDA). The hybrid approach for surgical palliation includes bilateral pulmonary artery (PA) banding and placement of a stent into PDA.
2. The goals of preoperative management include stabilization of the patient's clinical status, a complete definition of the cardiac anatomy, and identification of non-cardiac diagnoses that may impact perioperative care.
3. Preoperative echocardiography is used to evaluate the atrial level communication, myocardial function, degree of tricuspid regurgitation, adequacy of the PA bands, and degree of retrograde aortic arch obstruction.
4. Problems that may be encountered perioperatively include excessive pulmonary blood flow, increased pulmonary vascular resistance (PVR), and coronary ischemia. Coronary ischemia may result from a combination of retrograde arch obstruction, low diastolic blood pressure, and abnormal coronary vasculature.

Abstract

Hypoplastic left heart syndrome (HLHS) is a complex congenital heart condition which includes abnormal development of left-sided cardiac structures leading to inadequate systemic perfusion following post-natal closure of the patent ductus arteriosus (PDA). Surgical palliation may be accomplished through a 3 staged process known as the hybrid approach. Shortly after birth, the first procedure includes bilateral pulmonary artery (PA) banding to restrict pulmonary blood flow and pla cement of a stent into PDA to allow for systemic blood flow without the ongoing need for prostaglandin therapy to maintain ductal patency. We present a 2-month-old patient, 2.5 kg infant who presented for laparoscopic placement of a gastrostomy tube following stage I of the hybrid procedure. The intraoperative implications of the hybrid anatomy are discussed, options for anesthetic care presented, and previous reports of anesthetic care for such patients reviewed.

Keywords: hypoplastic left heart syndrome, laparoscopic surgery, congenital heart disese, hybrid stage 1.

Introduction

Hypoplastic left heart syndrome (HLHS) is a complex congenital heart condition which includes abnormal development of left-sided cardiac structures leading to inadequate systemic perfusion following post-natal closure of the patent ductus arteriosus (PDA).[1,2] Various surgical strategies have been employed for the palliation of HLHS in the immediate newborn period including the Norwood procedure and more recently, the hybrid procedure. The hybrid procedure includes bilateral pulmonary artery (PA) banding through a median sternotomy and placement of a stent into PDA.[3,4] The stent is placed through a sheath inserted directly into the main pulmonary artery with a multidisciplinary approach involving the surgeon and an interventional cardiologist in the hybrid operating room suite. The technique avoids the need for CPB during the neonatal period. One to two weeks after the initial procedure, a balloon atrial septostomy (BAS) is performed to ensure unimpeded mixing of blood at the atrial level. At 4-6 months of age, the comprehensive stage II is performed. The comprehensive stage II includes removal of the bilateral PA bands, removal of the PDA stent with PDA ligation, reconstruction of the aortic arch, and the creation of a cavopulmonary anastomosis (Glenn procedure). At two years of age, the Fontan procedure is completed in the same manner as the Norwood pathway for HLHS. We report a 2-month-old patient, 2.5 kg infant who presented for laparoscopic placement of a gastrostomy tube following the stage I hybrid procedure. The intraoperative implications of the hybrid anatomy are discussed, options for anesthetic care presented, and previous reports of anesthetic care for such patients reviewed.

Case report

Institutional Review Board approval is not required for publication of isolated case reports at Nationwide Children's Hospital (Columbus, Ohio). A 2-month-old, 2.5 kg infant with trisomy 18 who was status post stage I of the hybrid procedure for HLHS presented for laparoscopic G-tube placement. The patient's medical history included an unbalanced atrioventricular canal defect (AVC) with right-sided dominance and aortic coarctation following hybrid stage I palliation. There was ongoing hyperbilirubinemia due to cholestasis related to the prolonged need for parenteral nutrition. There was a past history of acute kidney injury (AKI) due to hypotension and intravascular volume depletion related to aggressive diuretic therapy following stage I of the hybrid procedure. From birth, there was acute and chronic respiratory failure requiring tracheostomy and ventilator dependence. Past surgical history included hybrid stage I procedure on day of life 11. The current feeding regimen included 14 mL of human milk delivered via a nasogastric tube every 3 hours. Medications included vancomycin (40 mg) every 12 hours and ampicillin-sulbactam (125 mg) every 6 hours for a recent nosocomial tracheal infection. Cardiac medications included enteral chlorothiazide twice a day and digoxin once a day. Phenobarbital was administered once a day to treat cholestasis. The preoperative hematocrit and hemoglobin were 40.4% and 12.6 gm/dL, respectively. Coagulation studies, serum electrolytes, blood urea nitrogen, creatinine, and blood glucose were within normal limits. Preoperative transthoracic echocardiogram showed HLHA (small left ventricle, hypoplastic and abnormal mitral valve, a large sub-aortic ventricular septal defect), no stenosis across the PDA stent, an unrestrictive atrial septal defect, mildly abnormal pulmonary valve, trivial tricuspid regurgitation, moderately dilated and hypertrophied right ventricle, mild to moderately depressed ventricular function, and adequate gradients across the PA bands.

The patient was held *nil per os* for 6 hours during which time 10% dextrose in 0.45% NS was infused at a maintenance rate of 10 mL/hour. After transportation from the Pediatric Cardiothoracic Intensive Care Unit (CTICU) to the operating room, routine American Society of Anesthesiologists' monitors were placed. Ane-

sthesia was induced via the incremental inhalation of sevoflurane in 30-40% oxygen which was administered via the tracheostomy. This was supplemented with fentanyl (total intraoperative dose of 10 µg/kg). Neuromuscular blockade was provided by rocuronium. Intraoperatively, the FiO_2 was maintained at 22-23% and maintenance anesthesia provided by isoflurane (expired concentration of 0.4-1.4%). During the procedure, mechanical ventilation was provided by pressure-limited ventilation with a peak inspiratory pressure (PIP) of 21-24 cmH2O and a positive end-expiratory pressure (PEEP) of 4 cmH2O. Exhaled tidal volumes varied from 8-10 mL/kg. Oxygen saturation (SpO2) ranged from 90-97% and the end-tidal carbon dioxide (EtCO2) varied from 34-37 mmHg during the procedure. Forced air warming was used to maintain normothermia of 36.4-37^{o} Centigrade during the procedure. Following the induction of anesthesia, the patient's abdomen was prepped and draped in standard surgical fashion. A 5 mm infraumbilical incision was made and carried down to the fascia which was directly incised. A laparoscopy trocar was placed in this location. A second 5 mm trocar was placed in the anticipated site for the G-tube. Insufflation was provided using an intra-abdominal pressure of less than 6-8 mmHg. The stomach was clearly visualized. It was grasped at the G-tube site and brought into sight. An open incision into the stomach was performed, a purse string suture placed, and the G-tube inserted. The two trocars were removed and the incisions closed. A local anesthetic agent (0.25% bupivacaine) was infiltrated into the surgical incision sites. Total intraoperative time was 95 minutes. During the procedure, heart rate ranged from 111 to 148 beats/minute, systolic blood pressure ranged from 85 to 110 mmHg, and diastolic blood pressure ranged from 25 to 55 mmHg. The patient tolerated the procedure well and was transferred back to the CTICU with standard ASA monitoring. The patient returned to his preoperative baseline status as far as hemodynamic and respiratory status without any perioperative complications.

Discussion and conclusion

HLHS encompasses a spectrum of intracardiac and extracardiac anomalies, including abnormalities of the ascending aorta, aortic valve, left ventricle, and mitral valve. The left ventricle and mitral valve may be poorly developed or totally absent. With this combination of defects, perfusion of the systemic circulation depends partly or completely on flow from the PA across the ductus arteriosus. Although these infants frequently appear normal at birth, signs of cardiorespiratory failure occur when the ductus arteriosus closes, usually during the first 2 weeks of life. Initial stabilization requires the infusion of prostaglandin to maintain or reopen the ductus arteriosus and provide systemic flow. The goals of surgical palliation of HLHS include provision of systemic blood flow while limiting pulmonary blood flow. At our institution, surgical palliation is provided by the hybrid approach. Shortly after birth, the first procedure includes bilateral pulmonary artery (PA) banding to restrict pulmonary blood flow and placement of a stent into PDA to allow for systemic blood flow without the ongoing need for prostaglandin therapy. A balloon atrial septostomy is then performed to ensure unrestricted flow of pulmonary venous blood returning from the lungs across the atrial septum into the right atrium. This blood is then ejected from the single ventricle into the pulmonary artery. The blood that is ejected into the pulmonary artery can then either flow to the lungs or to the body. The distribution of this blood flow (systemic versus pulmonary) is determined by the differential resistance to flow of the two circulations. Flow to the systemic circulation is determined by the systemic vascular resistance (SVR) or any mechanical obstruction in the PDA stent/aorta versus the restriction imposed by the degree of pulmonary banding or the pulmonary vascular resistance (PVR).

The goals of pre-operative management include clinical stabilization, complete definition of the cardiac anatomy, recognition of non-cardiac diagnoses, and family education. Transthoracic echocardiography is used to determine the presence of an adequate atrial level communication, myocardial function, degree of tricuspid regurgitation, adequacy of the PA bands, and degree of retrograde aortic arch obstruction. If needed, the anatomy can be further defined or interventions provided during cardiac catheterization as indicated. Problems that may be encountered perioperatively include excessive pulmonary blood flow, increased pulmonary vascular resistance (PVR), and coronary ischemia.

Coronary ischemia may result from a combination of retrograde arch obstruction, low diastolic blood pressure (BP), and abnormal coronary vasculature. These factors may lead to an increased frequency of ventricular fibrillation and the possibility of perioperative myocardial ischemic events.[5,6] Although the dependence on retrograde aortic perfusion of the coronaries may partly explain these events, it has also been postulated that there may be primary abnormalities in the coronary circulation with HLHS related to the persistently high intraventricular pressure with retrograde flow into the coronary vasculature, leading to intimal injury.[7,8] Given these concerns, control of factors regulating myocardial oxygen consumption and delivery should be considered in these patients. In particular, control of heart rate and maintaining normal diastolic blood pressure is suggested.

The primary goal in the management of patients with single ventricle physiology is optimizing systemic oxygen delivery and perfusion pressure. Maintenance of systemic oxygen delivery is dependent on optimizing cardiac output and arterial oxygen content. Optimal cardiac output requires attention to volume status (preload), vascular resistance (afterload), heart rate, rhythm, and myocardial contractility, whereas arterial oxygen content is predominately dependent on hemoglobin content.

Another major issue concerning the hemodynamic and respiratory status of infants with HLHS is the balance between pulmonary and systemic blood flow. Provided that they have an appropriate gradient, PA bands generally prevent excessive pulmonary blood flow related to decreases in PVR due to hypocarbia or high inspired oxygen concentrations. However, significant increases in PVR may result in a decrease in pulmonary blood flow and decreases in oxygen saturation. An oxygen saturation of 75% to 80% is generally optimal in infants with univentricular anatomy as a saturation in excess of 80% implies a pulmonary-to-systemic flow (Qp:Qs) ratio of greater than 1.5 to 1. These ratios not only decrease peripheral oxygen delivery but also result in overperfusion of the pulmonary bed, leading to pulmonary edema and chronic volume overload of the ventricle. A second, less common cause of pulmonary edema is pulmonary venous congestion from obstruction to pulmonary venous return from a restrictive atrial septum. These problems should be identified on preoperative echocardiography and if severe enough, may require intervention during cardiac catheterization. Simple intraoperative maneuvers to maintain the Qp:Qs ratio include adjusting the inspired oxygen concentration to maintain an oxygen saturation of 75-80%, maintaining normocarbia, a normal pH, and normothermia. Direct measurements of cardiac output are not possible by conventional means. Intraoperative data have demonstrated a progressive increase in the base deficit when the PaO_2 exceeds 50 mmHg.[9] Conversely, an increased SVR can generate lactic acidosis and a low cardiac output state due to the high afterload.

Myocardial function may be compromised in infants with HLHS related to chronic volume overload, a decreased ability of the morphologic right ventricle to sustain systemic pressures, and myocardial ischemia (see above). Myocardial function should be assessed preoperatively using echocardiography and inotropic agents such as milrinone and epinephrine administered as nee-

ded to improve myocardial performance. Chronic diuretic therapy may lead to electrolyte imbalance, and preoperative monitoring of potassium and magnesium are suggested. When ventricular function is normal, we have found that non-invasive monitoring of hemodynamic status is generally sufficient for surgical procedures of a limited duration. Furthermore, non-invasive monitoring of tissue oxygenation using near infrared spectroscopy may provide an early warning of decreased oxygen delivery.[10] Invasive monitoring may be warranted in patients with depressed ventricular function or for more invasive surgical procedures. This provides not only access for invasive blood pressure monitoring, but also access for arterial blood gas monitoring. This may be useful to follow acid-base status, pH, and $PaCO_2$ in a setting where end-tidal CO_2 monitoring may be unreliable. Alternatively, these patients may have percutaneously inserted central (PIC) lines which allow for venous blood gas monitoring.

The induction and maintenance of anesthesia is achieved with a combination of opioids, volatile, and intravenous agents. During both the induction and maintenance of anesthesia, the anesthetic agents chosen should be modified to limit their deleterious effects on hemodynamic parameters (see above). In our patient, anesthesia was induced and maintained with a combination of a volatile agent (sevoflurane or isoflurane) and fentanyl. Although synthetic opioids such as fentanyl offer the advantage of hemodynamic stability, the use of high doses will often requires postoperative mechanical ventilation.[11,12] Given that our patient already required chronic mechanical ventilation, we chose a high dose opioid technique with fentanyl to effectively blunt the surgical stress response and maintain hemodynamic stability. Alternatively, if the goal had been early tracheal extubation, the dose of fentanyl can be decreased to 3-5 µg/kg and higher doses of isoflurane used for maintenance anesthesia.In addition to our patient's comorbid CHD, the surgical procedure may also impact intraoperative hemodynamic and respiratory function. Concerns exist as to whether or not laparoscopic surgery is safe in patients with CHD, especially those with HLHS, given the increases in intra-abdominal pressure (IAP) with insufflation and its effects on cardiac function. In the absence of associated CHD, an IAP of 12 mmHg, but not 6 mm Hg, has been shown to decrease cardiac index assessed by TEE.[13] IAP above 6-8 mmHg may decrease preload, increase afterload, and increase PVR which may be poorly tolerated in patients with HLHS.[13,14] Echo-Doppler monitoring has shown decreases in aortic blood flow, stroke volume, and cardiac index in children at an IAP as low as 10 mm Hg.[13] Additionally, increases in PaCO2 induced by laparoscopy should be expected and minute ventilation increased, as an increase in PaCO2 levels may have deleterious effects on myocardial function and pulmonary vascular resistance. A significant increase in the arterial pressure of CO2 and end-tidal CO2 gradient after abdominal insufflation has shown end-tidal CO2 to be an insensitive monitor in healthy infants.[13]

Adequate postoperative pain control in neonates has historically been difficult to achieve and continues to present a challenge to the anesthesiologist and intensive care unit physician. The challenge of adequately treating postoperative pain is particularly relevant after surgery for complex congenital heart disease such as HLHS as pain treatment must be carefully weighed against the risks associated with opioids. Our experience has suggested that nurse-controlled anesthesia (NCA) with fentanyl achieves effective analgesia with limited adverse effects as demonstrated by low pain scores, successful early tracheal extubation, infrequent need to change to an alternate opioid, and a low incidence of adverse effects.[15] Fentanyl is commonly preferred in this setting due to its rapid onset, easy titratability, lack of active metabolites, and limited effect on myocardial and hemodynamic performance. Dexmedetomidine may be added to the regimen to provide sedation and potentiate

opioid-induced analgesia thereby decreasing opioid and potentially opioid-related adverse effects.[16]

The perioperative course of infants with HLHS may present specific challenges during all phases of care related not only to the underlying CHD, but also by the challenges imposed by the surgical procedure (laparoscopy). Pre-operative goals include clinical stabilization of myocardial performance, definition of the cardiac anatomy, and recognition of non-cardiac diagnoses. Transthoracic echocardiography is used to evaluation the atrial level communication, myocardial function, degree of tricuspid regurgitation, adequacy of the PA bands, and degree of retrograde aortic arch obstruction. Problems that may be encountered perioperatively include excessive pulmonary blood flow, increased pulmonary vascular resistance (PVR), and coronary ischemia. When caring for such patients, a key to successful outcome is the cooperation of a multi-disciplinary team including Pediatric ICU physicians, pediatric cardiac anesthesiologists, and pediatric cardiologists.

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