Laparoscopic Nissen Fundoplication in a 3-month-old s/p Norwood Procedure

Peggy P. McNaull, MD
University of North Carolina

A 4.5 kg, 13-week-old infant with hypoplastic left heart syndrome presents for a laparoscopic Nissen fundoplication and gastrostomy tube (G-tube). On day of life 4, the infant underwent the Norwood procedure (stage 1 single-ventricle palliation) with a right ventricle-to-pulmonary artery conduit (Sano modification). The baby now has severe gastroesophageal reflux despite maximal medical therapy. He has had 2 episodes of presumed aspiration pneumonia and has just completed a 10 day course of intravenous antibiotics. His hemoglobin is 14 g/dL and oxygen saturation is 78% on room air. Vital signs also include a respiratory rate of 40 breaths per min, a pulse of 120 beats per min, and a of BP 80/50 mmHg. The CXR shows cardiomegaly and chronic patchy infiltrates with evidence of pulmonary vascular congestion.

Objectives:

1. Review the indications for a Nissen fundoplication and G-tube in an infant with hypoplastic left heart syndrome s/p the Norwood Procedure.
2. Discuss the outcomes observed in children with congenital heart disease, specifically hypoplastic left heart syndrome, after noncardiac surgery.
3. Review the physiologic effects of laparoscopic surgery in an infant with complex congenital heart disease.
4. Develop an anesthetic management plan for an infant with hypoplastic left heart syndrome s/p the Norwood Procedure for a laparoscopic Nissen fundoplication and G-tube.

Introduction:

Congenital heart defects are the most common birth defects in the United States. Approximately 7.5% of those infants born with congenital heart disease (CHD) have hypoplastic left heart syndrome (HLHS). HLHS describes a group of cardiac malformations characterized by inadequate systemic flow secondary to an underdeveloped left heart and aorta. Palliative surgical repair for children with HLHS involves a 3-stage process whereby the single ventricle is ultimately dedicated to the systemic circulation and venous return is directed to the pulmonary circulation. Performed in the first few weeks of life, stage 1 single-ventricle palliation, commonly referred to as the Norwood procedure, includes the creation of a new aorta, from the hypoplastic ascending aorta and main pulmonary artery. This directs blood from the right ventricle to the systemic circulation. The Norwood procedure also provides the patient a new source of pulmonary blood flow through either a modified Blalock-Taussig (BT) shunt or a right
ventricle-to-pulmonary artery conduit, commonly referred to as the Sano shunt or modification (Figure 1).

Over the last 30 years, major advances in the surgical repair and medical management of children with HLHS have occurred. As a result, an increasing number of infants with palliated HLHS are surviving and subsequently requiring operative interventions for their extracardiac problems such as gastroesophageal reflux disease (GERD), oral aversion, and pulmonary disease. Nissen fundoplication is one of the most commonly performed noncardiac procedures in children with HLHS².

**Indications for surgery:**

Research has indicated that infants with HLHS have a high incidence of GERD resulting in a greater risk of failure to thrive, aspiration, and acute life-threatening events³. GERD in the patient with CHD is diagnosed by clinical observations of persistent emesis, aspiration, or respiratory distress after attempted feedings. Surgical intervention may be planned when an infant with HLHS exhibits: (1) intolerance to feedings and/or poor weight gain despite maximal medical therapy for GERD (2) a need for feeding access in conjunction with recurrent aspiration 3) aspiration pneumonia or sustained apnea after feedings⁴.

The nutritional status of infants with HLHS is often poor secondary to inadequate oral intake in the setting of ineffective oromotor coordination, high metabolic demands, and frequent nil per os (npo) status for various indications. No consensus exists regarding the optimal route for providing nutritional support to this patient population. Consequently, many infants with HLHS are referred for surgical placement of a G-tube.

There is little evidence that Nissen fundoplication and/or gastrostomy tube placement improves outcomes in patients with HLHS. One retrospective review of 112 infants with CHD, of which 33% had single ventricle physiology, demonstrated adequate weight gain with an increase in median weight percentile from 1.5% preoperatively to 4% at 3 months and 20% at 5 years. There was not, however, a comparative nonsurgical control group⁵.

Indications for and timing of a Nissen fundoplication and/or gastrostomy tube placement in the HLHS patient with GERD and/or poor nutritional status is a matter of debate. Opponents of early intervention fear the risks of surgery and anesthesia, while proponents argue that ongoing pulmonary insult and inadequate weight gain can be devastating in the child with single ventricle physiology.
Outcomes of surgery:

Researchers have sought to define, primarily through retrospective reviews, the risks of morbidity and mortality from noncardiac surgery in children with CHD. Baum and others evaluated data from a large multi-institutional database for the period of January 1, 1993 through December 31, 1996. They reviewed the clinical information of 191,261 patients under 18 years of age having one or more noncardiac surgeries. Three percent of the patient population evaluated had CHD. Mortality was 3.8% in patients without CHD and 6.0% in patients with CHD. Mortality was increased in patients with CHD less than 1 year of age and in patients with major cardiac anomalies. Torres and colleagues reviewed the discharge data of 2,457 children under the age of 2 with HLHS hospitalized between 1988 and 1997. Nineteen percent of children with HLHS undergoing noncardiac surgical procedures died. In 2010, data from the Pediatric Perioperative Cardiac Arrest Registry (POCA) was published addressing anesthesia-related cardiac arrest in children with heart disease (HD). From 1994 to 2005, 34% of the perioperative cardiac arrests (CA) reported to the POCA registry occurred in patients with HD. Fifty-four percent of those children with HD who suffered a perioperative CA did so during noncardiac surgical procedures. Forty-seven percent of the patients with HD who had a CA were under 6 months of age and 70% under 2 years of age. The most common cardiac defect reported was a single ventricle and the most frequent surgical procedures were gastrointestinal. The overall mortality rate after a CA for patients with HD was 33% versus 23% for patients without HD. When mortality rates for ASA physical status III-V were compared, there was no difference in mortality between patients with and without HD.

Over the last decade, several groups have evaluated outcomes specifically in the single ventricle patient undergoing a Nissen fundoplication. Garey described their institutional experience in 39 patients with HLHS s/p the Norwood procedure who had an open Nissen fundoplication between January 1990 and July 2007. Three intraoperative complications were described: 2 with hemodynamic instability and 1 with pulmonary hypertension that required ECMO and termination of the procedure. Sixteen patients (41%) suffered postoperative complications including prolonged mechanical ventilation, NEC, sepsis, recurrent GERD, ECMO, DVT, intestinal ischemia, pneumonia, cerebral infarction, and candidemia. There were 2 deaths within 30 days of the procedure. From these results, Garey concluded that gastric fundoplication should be performed only under prospective protocols in this patient population. However, the data presented by Garey does not address the potential significant risk reduction with the laparoscopic Nissen fundoplication. Furthermore, they do not account for the great advances in both surgical and medical management in children with HLHS that have occurred since 1990.

Since 2005, there have been at least 3 outcome studies evaluating laparoscopic Nissen fundoplications in patients with complex CHD that present more favorable data. Mariano
described 5 patients with HLHS s/p the Norwood procedure who presented for laparoscopic fundoplication. No patients experienced intraoperative or postoperative complications\(^8\). Slater reported on 12 patients with HLHS (9 s/p the Norwood procedure and 3 s/p the bidirectional Glenn shunt) that underwent a laparoscopic procedure. There was no intraoperative hemodynamic instability and no intraoperative or postoperative mortality. Postoperative complications occurred in 6 patients including G-tube cellulitis, UTI, and sepsis\(^9\). Cribbs described 112 patients with complex CHD (37 with single ventricle pathology) that had a 104 laparoscopic and 8 open fundoplications. Postoperative mortality at 30 days was 4.5% (5 deaths). One death occurred in the operating room in a single ventricle infant who aspirated with extubation. The other deaths occurred at 22, 26, 27 and 28 days postoperatively. Twenty-eight percent of patients had non-lethal complications, most commonly recurrent GERD (7%) and postoperative mechanical ventilation greater than 48 hours (6%)\(^5\).

Not surprisingly, clinical research reveals that noncardiac surgery is of greater risk in patients with CHD\(^2,4-9\). While the data that has come from these clinical investigations is informative and highlights increased perioperative risk, it must be interpreted cautiously. The field of congenital cardiac surgery is rapidly advancing; frequently the studies evaluating long-term complications are out of date by publication. Additionally, the complex nature of congenital heart disease and the variety of noncardiac surgeries described in the literature make it difficult to extrapolate outcome data to the individual patient. Recent literature seems to indicate procedures such as a laparoscopic Nissen fundoplication can safely be performed on the single ventricle patient with careful intraoperative and postoperative monitoring by experienced personnel.

**Physiologic effects of laparoscopic surgery:**

Over the last 20 years, there have been major advances in the field of pediatric laparoscopic surgery. The first report of a pediatric laparoscopic Nissen fundoplication was in the early 1990s. This technique has virtually replaced the open approach. The benefits of the laparoscopic approach include a reduction in postoperative pain, analgesic requirements, respiratory complications, wound infections, and hospital stay\(^9-14\).

Despite the benefits of laparoscopy, there are many physiologic changes that occur as a result of abdominal insufflation. Abdominal insufflation causes cephalad displacement of the diaphragm. Consequently, there is a decrease in total lung compliance and functional residual capacity that may result in an increase in mean airway pressure, atelectasis, and ventilation-perfusion mismatch\(^12\). The reverse-Trendelenberg position necessary for the laparoscopic Nissen procedure may offset some of these respiratory effects.
Insufflated CO₂ is absorbed during laparoscopic surgery and eliminated for up to 10 minutes after deflation of the abdomen. Elevations in CO₂ will increase pulmonary vascular resistance even in healthy patients. There is often a large discrepancy between PaCO₂ and ETCO₂. Wulkan demonstrated in 7 children with HLHS undergoing laparoscopic Nissen fundoplication that there was a significant increase in the PaCO₂ and ETCO₂ gradient after abdominal insufflation compared with baseline. The gradient increased from a mean of 5.7 pre-insufflation to 13.4 post-insufflation.

The increase in abdominal pressure and PaCO₂ that occurs during laparoscopic surgery also has significant effects on the cardiovascular system. Research indicates that increasing intra-abdominal pressures leads to a proportional increase in systemic and pulmonary vascular resistances and decrease in cardiac index. In a series of 12 healthy males, age 6-30 months, having laparoscopic orchiopexies at 10 mmHg of intra-abdominal pressure, continuous esophageal aortic blood flow echo-doppler demonstrated a 30% decrease in cardiac output and a 162% increase in systemic vascular resistance indexed to body surface area (BSA). A limitation of this study is that aortic blood flow measurements may overestimate the decrease in cardiac output during abdominal insufflation. Sakka evaluated the effects of abdominal insufflation using transesophageal echocardiography in 12 healthy children having hernia surgery. A 13% decrease in cardiac index was noted at 12 mmHg of intra-abdominal pressure. Cardiac index returned to baseline when the intra-abdominal pressure was decreased to 6 mmHg. Finally, the reverse-Trendelenberg position causes a significant reduction in venous return which further compounds the observed hemodynamic changes.

The physiologic consequences of laparoscopy previously described above are well tolerated by the healthy child. Recent case series indicate that laparoscopic conditions are also tolerated by the child with HLHS s/p the Norwood procedure. Nonetheless, it is imperative the pediatric anesthesiologist understand how a laparoscopic procedure may impact the physiology of this fragile patient population.

**Anesthetic management:**

As stated earlier, the patient with HLHS s/p the Norwood procedure relies upon his/her single right ventricle to pump to the systemic circulation via the new aorta, and to the pulmonary circulation via either a BT shunt or Sano shunt. The single right ventricle is volume overloaded as a result of the parallel circulation. Furthermore, there is frequently an imbalance in the ratio of pulmonary to systemic blood flow (Qₚ/Qₘ). The Qₚ/Qₘ is determined by the balance between the pulmonary and systemic vascular resistance. Initially, the BT shunts or Sano shunts are often large in comparison to the BSA of the infant to allow for growth. Therefore, pulmonary vascular resistance may be low relative to the systemic circulation in the first weeks to months after the Norwood procedure. Pulmonary over-circulation may be problematic in
this setting. However, at around 3 to 6 months, the $Q_p/Q_s$ often becomes less than 1:1 secondary to the infant outgrowing the shunt, shunt calcification or stenosis, and/or the development of pulmonary hypertension$^{16}$.

The preoperative evaluation should include an assessment of the balance between the pulmonary and systemic blood flow. Assessment of the patient’s oxygen saturation, chest x-ray, and recent echocardiogram will likely reveal the presence of either pulmonary over- or under-circulation. Careful consideration should be given to how the effects of laparoscopy will affect the patient’s $Q_p/Q_s$. Signs and symptoms of cardiac failure and/or end-organ injury should also be sought, as well as any evidence of an ongoing upper respiratory tract infection (URI). URIs have a greater impact on pulmonary vascular resistance in children with CHD$^{17}$.

The literature describes the successful use with careful titration of propofol, ketamine, fentanyl, midazolam, etomidate, and inhalational agents in this patient population for both induction and maintenance of anesthesia$^{8-9,17}$. Intravenous induction is typically preferred; fortunately, many of these patients present to the OR with central access. Intraoperative arterial access is prudent as the physiologic consequences of laparoscopy can cause abrupt changes in the patient’s hemodynamic status and because the ventilation cannot be adjusted based upon ETCO$_2$ evaluation alone$^{15}$. Hypercarbia and acidosis increase PVR and should be avoided through an effective ventilation strategy. Hyperoxia results in pulmonary over-circulation at the expense of systemic perfusion and too should be avoided. The hematocrit in this patient population should be maintained over 40% to maximize oxygen carrying capacity. Blood pressure should be supported with inotropic agents as necessary. This patient population is exquisitely prone to myocardial ischemia. Postoperative ventilation is often necessary$^8$.

The anesthesiologist must be knowledgeable and experienced in order to care for a patient with complex congenital heart disease for noncardiac surgery. A lack of understanding of the physiology of these unique patients can result in devastating consequences.

References:

Figure 1: The Norwood operation for first-stage palliation of hypoplastic left heart syndrome. The pulmonary artery is transected just proximal to its bifurcation, and the pulmonary artery is anastomosed in an end-to-side or side-to-side fashion to the ascending aorta, with reconstruction of the ascending and transverse aorta. Pulmonary blood flow is supplied by a modified Blalock-Taussig shunt (A) or right-ventricle to pulmonary artery conduit (B).16