Chapter 26

Stage I Palliation, Hypoplastic Left Heart Syndrome

Bishr Haydar

Case Scenario

A 6-week-old infant weighing 4 kg is scheduled for laparoscopic Nissen fundoplication with gastrostomy tube placement. She was born at term with hypoplastic left heart syndrome, for which she underwent a Stage I palliation (Norwood procedure) with a Sano modification at age 5 days. Her initial postoperative course was uncomplicated, but she has had persistent difficulty feeding and has failed to gain weight. She was discharged from the hospital on furosemide and aspirin.

Transthoracic echocardiogram at the time of discharge 3 weeks earlier showed the following:

- *Mild-to-moderate tricuspid regurgitation*
- *Mildly diminished right ventricular systolic function*
- *A patent Sano (right ventricular to pulmonary artery) shunt with a 40 mm Hg peak gradient*

Despite placement of a nasogastric tube for feeding, she has gained little weight and has persistent reflux. She appears alert, although small and thin, with a heart rate of 150 beats/minute, respiratory rate of 40 breaths/minute, and SpO_2 86% on room air. Her cardiologist would like surgery to take place as soon as possible.

Key Objectives

- Describe the anatomy and physiology of hypoplastic left heart syndrome.
- Describe available options for the initial palliation of hypoplastic left heart syndrome.
- Describe the anatomy and physiology of a patient who has undergone surgical Stage I palliation.
- Describe an appropriate plan for perioperative anesthetic management of this infant.
- Identify markers of inadequate pulmonary or systemic blood flow in infants after Stage I palliation.
- Describe the physiology of laparoscopy as it pertains to this population.
- Discuss perioperative complications related to this population.

Pathophysiology

What is hypoplastic left heart syndrome?

The term "hypoplastic left heart syndrome" (HLHS) is used to describe a spectrum of congenital cardiac abnormalities involving underdevelopment of left-sided heart structures. Findings may include mitral valve stenosis or atresia, aortic stenosis or atresia, hypoplasia or absence of the left ventricle (LV), and hypoplasia of the ascending aorta and aortic arch. Hypoplastic left heart syndrome occurs relatively commonly (approximately 1 in 4000 births).

The combination of these defects results in single-ventricle physiology, wherein a single ventricle, in this case the right ventricle (RV), is required to support both pulmonary and systemic circulations. Single-ventricle physiology requires complete intracardiac mixing of pulmonary venous and systemic venous blood that is then supplied to parallel pulmonary and systemic circuits.

Clinical Pearl

Hypoplastic left heart syndrome is a spectrum of congenital cardiac abnormalities involving underdevelopment of left-sided heart structures and resulting in physiology wherein a single ventricle, in this case the RV, is required to support both pulmonary and systemic circulations.

What is the circulatory pattern in patients with HLHS after birth and what does "ductal-dependent" mean?

In patients with HLHS, due to the inadequacy of the left-sided heart structures, blood flow through the native LV outflow tract is limited and therefore flow to the systemic circulation after birth is dependent on flow provided via the patent ductus arteriosus (PDA). In patients with aortic atresia, the child is completely dependent on the PDA for blood flow to the coronary and cerebral circulations and

https://doi.org/10.1017/9781108657341.027 Published online by Cambridge University Press

significant hypotension may be poorly tolerated. It is therefore necessary for patients with HLHS to be placed on continuous prostaglandin E_1 (PGE_1) infusions after birth to assure continued patency of the ductus arteriosus and flow to the systemic circulation. Hypoplastic left heart syndrome is therefore known as a "ductal-dependent" cardiac lesion. (See Figure 26.1.)

Clinical Pearl

In patients with HLHS, due to the inadequacy of the left-sided heart structures, flow to the systemic circulation after birth is dependent on flow provided via the PDA.

What is a "series" circulation as opposed to a "parallel" circulation?

In a normal two-ventricle heart the systemic and pulmonary circulations exist in series, with each circulation supported by its own ventricle. In HLHS patients, prior to and immediately after Stage I palliation, blood flow to both systemic and pulmonary circulations is supplied in parallel by the single ventricle. The single ventricle therefore has to perform increased work in order to maintain flow to both the systemic and the pulmonary circulations.

Once patients undergo Stage II palliation (hemi-Fontan or bidirectional Glenn, see Chapter 27) followed by the Fontan procedure (see Chapters 28–30) their systemic and pulmonary circulations then exist in series, meaning that blood pumped by the single ventricle will pass through both circulations before returning to the heart.

Clinical Pearl

In HLHS patients, prior to and immediately after Stage I palliation, blood flow to both systemic and pulmonary circulations is supplied in parallel by the single ventricle.

When do patients undergo Stage I palliation?

The use of PGE_1 is associated with a variety of dose-dependent side effects which can commonly include hypotension and apnea, and thus the lowest effective dose is utilized to minimize these side effects. Additionally, the natural decrease in pulmonary vascular resistance (PVR) after birth compared to systemic vascular resistance (SVR) means that pulmonary overcirculation will occur as blood preferentially flows to the circulation with lower resistance. In an infant with parallel circulations this increase in pulmonary blood flow (PBF) will come at the expense of systemic blood flow and oxygen delivery. For these reasons, unless other issues delay surgery it is preferable to proceed with the Stage I palliative surgery within the first week of life when possible.

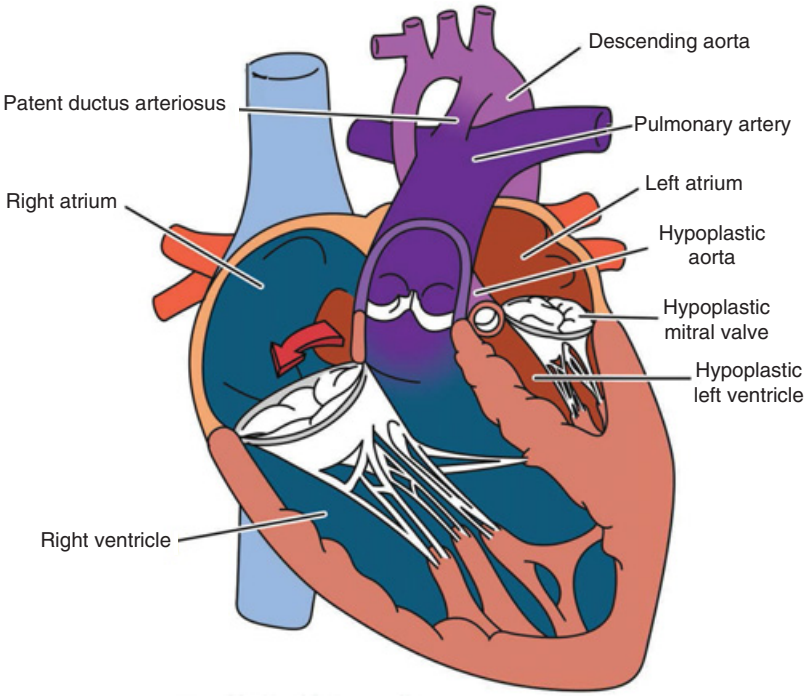

Figure 26.1 **Hypoplastic left heart syndrome.** Drawing by Ryan Moore, MD, and Matt Nelson.

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What is a "Stage I" palliation and what are the surgical goals?

A Stage I palliation for HLHS is often referred to as the "Norwood procedure," after surgeon William I. Norwood, who first successfully performed the procedure in Boston in 1983 [1]. Although a Stage I palliation for HLHS is a complicated procedure, it may be more easily understood if broken down into the individual goals the surgery must achieve.

1. ***Creation of an unobstructed source of systemic blood flow (aortic reconstruction)***: In HLHS patients the aorta is diminutive or hypoplastic, and prior to Stage I palliation distal flow to the aorta is provided via the PDA. Initial surgical palliation will utilize the patient's native pulmonary outlet, and utilizing autologous tissue supplemented with patch material for augmentation will adjoin it to the patient's hypoplastic aortic outlet, creating a single ventricular outlet, or "neo-aorta." Going forward this will serve as the systemic outflow tract. To utilize the pulmonary root for aortic reconstruction the main pulmonary artery will be transected and oversewn, so it is then necessary to create a source of PBF.
2. ***Creation of a PBF source***: As the main pulmonary artery is transected and oversewn in order to utilize the pulmonary outlet for reconstruction of the hypoplastic aorta, a new source of PBF must be established, with the goal of providing adequate but not excessive PBF. This systemic-to-pulmonary shunt may take one of several forms.
   a. *Modified Blalock-Taussig (mBT) shunt)*. A GORE-TEX® tube graft (generally 3.5 mm) is interposed between the right subclavian or innominate artery and the pulmonary artery (PA) to provide PBF. As PA diastolic pressures are lower than aortic diastolic pressure, this shunt is associated with increased flow during diastole, leading to lower aortic diastolic pressure. This may result in diastolic hypotension and coronary ischemia. (See Figure 26.2.)
   b. *Sano shunt*. An alternate approach preferred by some centers involves creation of a graft between the single RV and the PA, known as a Sano shunt. The RV–PA shunt maintains higher diastolic pressure with presumably better coronary blood flow and is associated with better early surgical outcomes in certain types of HLHS [2] (See Figure 26.3.)
   c. *Central shunt*. Aortic-to-pulmonary shunts are less commonly utilized.
3. ***Ensure complete atrial mixing***: Going forward the patient's heart will function as a single atrium and a single ventricle. Therefore, an atrial septectomy is necessary to allow complete mixing of oxygenated pulmonary venous return and deoxygenated systemic

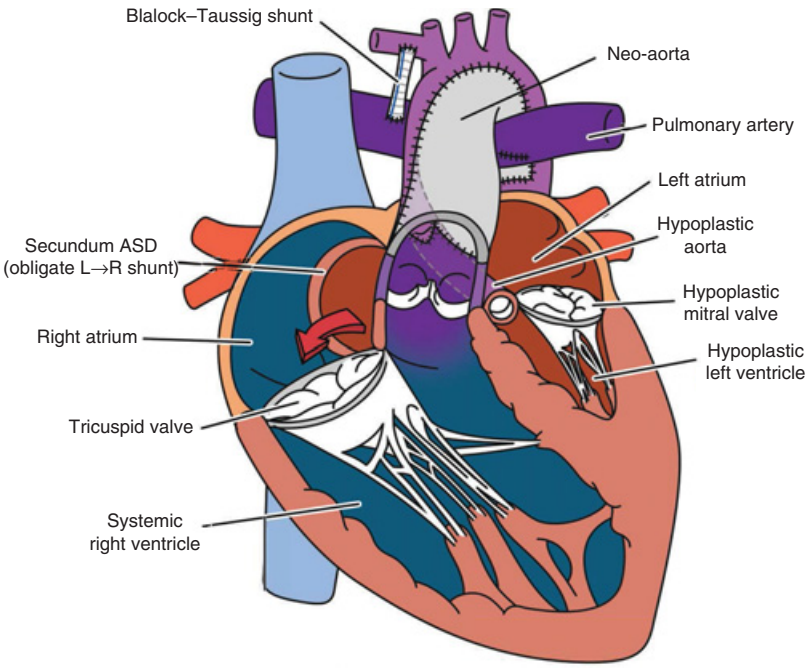

Figure 26.2 HLHS Stage I palliation with modified Blalock-Taussig shunt. Drawing by Ryan Moore, MD, and Matt Nelson.

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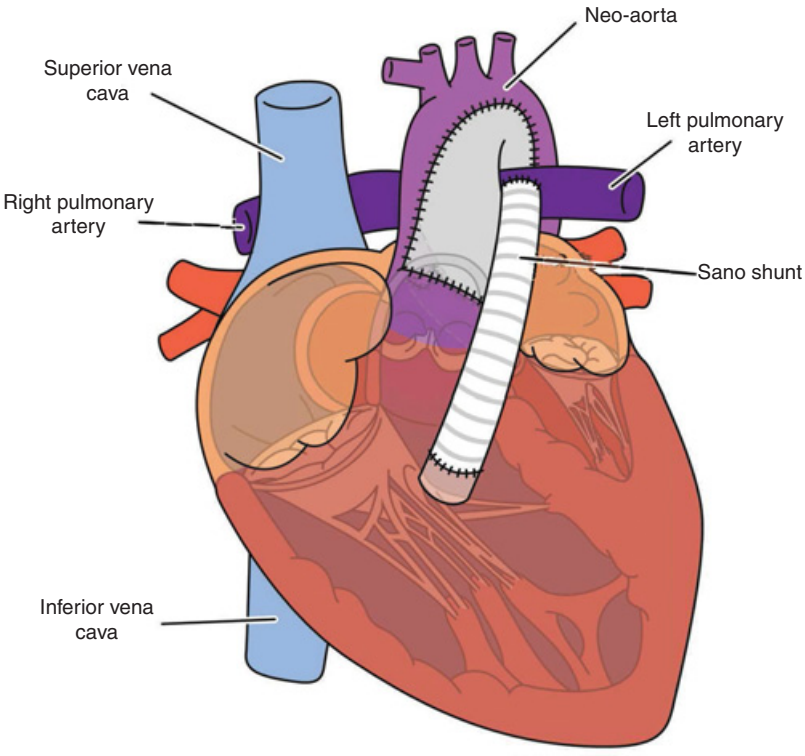

Figure 26.3 HLHS Stage I palliation with Sano shunt. Drawing by Ryan Moore, MD, and Matt Nelson.

venous return. Because of this mixing, the patient's expected oxygen saturations will be 75–85% after Stage I palliation.

Clinical Pearl

Due to mixing of pulmonary and systemic venous return in the common atrium, the patient's expected oxygen saturations will be 75%–85% after Stage I palliation.

What is the new pattern of blood flow in a patient after a Stage I procedure?

- Systemic and pulmonary venous return mix in the common atrium.
- Blood flows through the atrioventricular valve (tricuspid valve in the case of HLHS).
- The single systemic ventricle pumps blood into the newly constructed "neo-aorta."
- Systemic blood flow is provided via the neoaorta.
- A surgically created shunt (either an mBT or Sano) provides PBF.

Therefore, blood ejected from the single ventricle can enter ***either*** the pulmonary or systemic circulation. The ratio of pulmonary to systemic blood flow (Q_p:Q_s) therefore varies depending on resistance in each circulation (PVR and SVR). As PVR is generally lower than SVR, the risk for excessive PBF is high. Mechanical restriction to pulmonary blood flow is achieved by using an appropriately sized shunt.

What is the "hybrid approach" to Stage I palliation? When is it utilized?

The hybrid approach refers to the combined use of surgical and interventional cardiology techniques to achieve the desired goals and is performed in the cardiac catheterization laboratory or in a specially designed "hybrid" operating room. It is performed via sternotomy but without the use of cardiopulmonary bypass (CPB).

The hybrid procedure is preferred for patients who may be at higher than normal risk for a traditional Stage I procedure and might be expected to have a poor outcome. This often includes patients weighing <2 kg, including those who are premature and small for gestational age. It is also utilized in patients whose families wish to avoid blood transfusion, such as Jehovah's Witnesses. Hybrid procedures may also be used to limit PBF in other forms of congenital heart disease. This approach allows the patient to grow until their size is more appropriate for surgical repair or until cardiac transplantation, depending on their disease.

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What are the goals of a hybrid procedure?

Goals of a hybrid procedure are similar to the Stage I Norwood procedure. (See Figures 26.2 and 26.3.)

- ***Systemic blood flow provision***: Rather than continuing to rely on PGE_1 to maintain ductal patency, the surgeon and interventional cardiologist ***place a stent directly into the PDA*** via a sheath in the PA. The stented PDA will allow continued provision of systemic blood flow via the distal aorta. Surgical reconstruction of the aorta will take place at the time of the Stage II palliative procedure.
- ***Pulmonary blood flow*** is supplied via the pulmonary arteries, and in order to prevent pulmonary overcirculation, ***restrictive bands are placed on the left and right pulmonary arteries.*** The degree of restriction is titrated based on oxygen saturation and direct pressure measurements measured via the pulmonary artery sheath and transesophageal echocardiography.

What surgical stages are generally involved in complete palliation of an HLHS patient?

The ultimate goal of single-ventricle palliation is utilization of the single ventricle to provide systemic blood flow. Pulmonary blood flow is supplied by passive blood flow of systemic venous return directly to the lungs.

Stage I palliation involves the creation of a single atrium by performing an atrial septectomy, reconstructing the aorta as the outflow tract of the single ventricle and creating a surgical shunt for PBF. Because this circulatory arrangement involves complete mixing of systemic and pulmonary venous return the patient remains cyanotic after Stage I pallation.

As the fixed size of either an mBT or a Sano shunt means that the infant will become increasingly cyanotic with growth, ***Stage II palliation*** is usually performed between 3 and 6 months of age, allowing PVR to continue to decrease prior to this surgery. In Stage II palliation the initial mBT or Sano shunt is taken down, and a new source of PBF is created either by anastomosis of the superior vena cava (SVC) directly to the PA via a bidirectional Glenn procedure (BDG) or through creation of an atrial baffle with a hemi-Fontan procedure. After the Stage II surgery deoxygenated blood from the inferior vena cava (IVC) still mixes with oxygenated blood from the pulmonary veins in the atrium, so the patient continues to be cyanotic. (See Chapter 27.)

The Fontan procedure – the final stage – creates a connection between the IVC and the PA, either through an extracardiac tube graft or by creating a lateral tunnel via revision of the atrial baffle [3]. The systemic and pulmonary circulations are now separated and oxygen saturations are usually in the 90s. (See Chapters 28–30.)

Clinical Pearl

After Stage II palliative surgery deoxygenated blood from the IVC still mixes with oxygenated blood from the pulmonary veins in the atrium, so the patient remains cyanotic, but they now have a source of PBF that will grow as they grow. After the Fontan procedure hemoglobin–oxygen saturations should be in the 90s.

What is the Q_p:Q_s ratio and what is its significance?

Pulmonary blood flow is denoted by Q_p, and systemic blood flow by Q_s. The Q_p:Q_s ratio represents the ratio of pulmonary blood flow to systemic blood flow. In a normal biventricular heart this ratio is 1.

How do changes in the balance of Q_p and Q_s affect the patient?

In a single-ventricle (parallel) circulation, blood ejected by the ventricle can flow through either the systemic (Q_s) or pulmonary arterial (Q_p) system.

- A marked decrease in PVR or a sudden increase in SVR will cause increased Q_p at the cost of Q_s, resulting in hypotension and signs of poor perfusion.
- A marked decrease in SVR will cause increased Q_s at the cost of Q_p and result in increased cyanosis.

Diminished cardiac output, whether from myocardial dysfunction, ischemia, or atrioventricular (AV) valve regurgitation, can result in reduced flows to both circulations, causing both hypotension and desaturation.

What is the optimal Q_p:Q_s in a patient with Stage I physiology and what oxygen saturation does this typically reflect?

An oxygen saturation of 75%–85% as measured by pulse oximetry appears to be optimal for most patients with Stage I physiology, as higher saturations may indicate pulmonary overcirculation, placing increased demands on the single ventricle as it provides flow to both circulations. These saturations typically reflect a Q_p:Q_s ratio of 0.7–1.0 [4].

Clinical Pearl

Following Stage I palliation, the ideal systemic oxygen saturation is 75%–85%. This reflects a Q_p:Q_s ratio of 0.7–1.0. Although some patients may demonstrate higher resting oxygen saturations, they should be monitored carefully for signs and symptoms of inadequate perfusion.

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What factors allow optimization of Stage I palliation physiology?

Optimization of the patient's physiology focuses on maximizing cardiac output and oxygen delivery. Atrioventricular valve regurgitation and dysrhythmias are poorly tolerated as they reduce cardiac output. Marked changes in either pulmonary or systemic vascular resistance may cause an imbalance of Q_p:Q_s. As increased oxygen carrying capacity is desirable, a higher hematocrit is utilized to maintain adequate oxygen delivery. Cyanotic patients typically require a hematocrit of 40% or greater.

Anesthetic Implications

What is the significance of this patient's failure to thrive?

Feeding and digestion require relatively high metabolic demand in infants as compared to their resting state and so both failure to thrive and gastroesophageal reflux are frequently seen in this patient population. They can be manifestations of the inability to mount an appropriate cardiac or pulmonary response to increased metabolic demand. Vocal cord dysfunction can occur as a result of recurrent laryngeal nerve injury during aortic arch reconstruction or as a result of prolonged intubation and may also result in feeding intolerance. Approximately 15%–30% of patients with HLHS have other genetic abnormalities, and reflux and failure to thrive may also be symptoms of other congenital abnormalities or syndromes.

What preoperative assessment should be performed for this patient?

A thorough preoperative chart review, interview, and physical exam are essential. Initial assessment should include review of a recent echocardiogram, focusing on ventricular function, AV valve regurgitation and shunt patency. Anesthetic records and hospital discharge summaries should be reviewed for complications associated with Stage I surgery and hospitalization. Perioperative and postoperative complications can also include nervous system injury, shunt malfunction, arterial or central venous occlusion, and renal dysfunction. The most recent cardiology notes and laboratory studies, especially hematocrit, should be reviewed as well. Blood type and antibody screening should be up to date, as patients have usually been previously transfused and may require transfusion to maintain a hematocrit in the preferred range above 40%.

Preoperative evaluation should include assessment for adequacy of cardiac output. Infants experience peak metabolic demand while feeding and digesting which can produce transient symptoms of inadequate cardiac output. These can include tachypnea, interruption of feeding, sweating, and worsened cyanosis. This may result in poor weight gain; severely inadequate cardiac output may result in lethargy. Home oxygen saturation values should be reviewed. In this patient, a baseline oxygen saturation of 86% may reflect pulmonary overcirculation, especially in this child with poor feeding. Aspirin is often continued in the perioperative period while diuretics are typically held during fasting.

Clinical Pearl

Specific echocardiographic findings indicate patients at increased risk for adverse outcomes. These include impaired ventricular function, significant valvular regurgitation and coronary artery pathology. Patients with mBT shunts are at greater risk for diastolic hypotension and myocardial ischemia than those with Sano shunts.

How should parents be counselled about anesthetic risk?

Infants with Stage I physiology are at markedly increased risk for perioperative complications compared to healthy infants and older patients. They have reduced respiratory reserve owing to their baseline desaturation, as well as limited cardiac reserve. Hypotension necessitating intraoperative inotropic support is, unfortunately, not uncommon. The presence of a common atrium may allow any intravenous (IV) air bubble to enter the arterial tree and result in stroke. Because of the risk of perioperative hemodynamic and respiratory instability, most Stage I patients are routinely taken to an intensive care unit (ICU) for postoperative management. At some centers they are routinely transferred to the ICU while still intubated, allowing the intensivists to carefully monitor for adequate perfusion, ventilation, and full awakening prior to extubation.

An increase in afterload or decrease in cardiac output may result in sluggish shunt flow, which can result in shunt thrombosis. This may require emergent initiation of extracorporeal membrane oxygenation (ECMO) for survival. Intensive care units allow for more rapid initiation of ECMO compared to the recovery room or hospital ward.

Parents should therefore be counseled on the potential need for inotropic support and blood transfusion, and the likelihood of postoperative intubation following the procedure. The risk of stroke, cardiac arrest, and shunt thrombosis requiring ECMO cannulation are significantly higher compared to children without heart disease, though they are still uncommon.

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Clinical Pearl

Parents should be counseled preoperatively on the potential need for inotropic support and blood transfusion, and the likelihood of postoperative intubation following the procedure.

What vascular access and monitoring is advisable for this procedure?

For this procedure, a peripheral IV line in the upper extremity is preferred, in case of IVC obstruction due to insufflation or injury during dissection. However, it is best to avoid internal jugular or subclavian central venous access, as this may cause venous stenosis that could subsequently negatively impact successful Stage II palliation. If upper extremity central access is needed, a peripherally inserted central catheter along with large-bore peripheral IV access would be preferable and may prove beneficial if use of vasopressor and/or inotropic therapy is likely.

It is generally advisable to have two peripheral IV lines at a minimum to administer fluids, blood if necessary, and vasoactive drugs if needed. The gradient between $PaCO_2$ and $ETCO_2$ increases during laparoscopic procedures, and this increase is magnified in patients with cyanotic heart disease. Measuring instantaneous blood pressure with an arterial catheter is advantageous and placement of an arterial line is generally prudent. However, this must be balanced against the difficulty of placing arterial access in an infant who has recently had an arterial line and will require future arterial lines; patient status and the expected length of surgery can aid the practitioner in making this decision. Other factors to consider when determining the need for invasive monitors include potential blood loss, patient-specific bleeding and/or transfusion risk factors, postoperative monitoring needs, long-term venous access, and postoperative disposition.

What method of induction is preferable and what are the hemodynamic goals during induction?

Although induction can be achieved via inhalational or intravenous routes, an IV induction would be safest for this patient. For any patient with poor cardiopulmonary reserve, IV induction provides optimal hemodynamic control, the option for correction of fluid deficits prior to induction and immediate access for resuscitation. It also makes rapid sequence induction possible, if deemed necessary. Preferred induction medications may include ketamine, fentanyl, and etomidate along with neuromuscular blocking agents.

Cardiac output is best maintained through ensuring adequate preload and judicious dosing of anesthetic agents. Decreases in PVR must be avoided and supplemental oxygen should be used sparingly, immediately before laryngoscopy or when a patient becomes hypoxemic. Oxygen saturations should be maintained at or near the baseline preoperative value.

The initiation of positive pressure ventilation (PPV) and/or administration of high-dose sevoflurane can lead to anesthetic overdose. Volatile anesthetic agents can cause myocardial depression and should be used judiciously. Even with the use of neuromuscular blockade and reduced dosing of inhaled anesthetics hemodynamic instability is not uncommon.

Clinical Pearl

Priorities during anesthetic induction are to maintain the appropriate Q_p:Q_s balance and to minimize reductions in cardiac output. Oxygen saturations should be maintained at or near the baseline preoperative value and inspired oxygen concentrations should be used as needed to maintain baseline saturations.

What are the anesthetic considerations for a patient whose PBF is provided via a systemic-to-pulmonary shunt?

Inhalational induction may be prolonged in patients with cyanotic heart disease and an IV induction of anesthesia would be recommended for this patient. These patients can be extremely sensitive to reductions in PVR that result in excess Q_p, which in turn compromises Q_s. Efforts should be made to maintain oxygen saturations at the preoperative baseline level. After endotracheal intubation this often means utilizing an inspired oxygen concentration (FiO_2) at or near 0.21. Once insufflation has commenced it is generally necessary to adjust ventilator settings to compensate for the increase in $ETCO_2$ and it may also be necessary to increase FiO_2 at that time if oxygen saturations have decreased appreciably from baseline.

Patients with Stage I single-ventricle physiology have increased ventricular volume work and afterload relative to normal biventricular hearts. Alterations in preload due to prolonged preoperative fasting and/or the effects of decreased cardiac contractility due to anesthetic agents may therefore be more deleterious. Hypotension is very common and may require the judicious replacement of

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fluids, administration of blood products, or in some cases inotropic therapy. Paradoxical emboli are a major concern, as anything that enters systemic venous return can be immediately transmitted to the systemic arterial circulation due to the complete mixing of the circulations.

Patients are frequently taking antiplatelet or anticoagulant medications to prevent shunt thrombosis or treat previous vascular occlusions due to cardiac catheterization or prolonged central venous or arterial catheter use. If present, long-standing polycythemia can result in a reduction in plasma clotting factor compensation which may further increase bleeding risk.

Cyanotic patients and those with surgical grafts are also at increased risk for infective endocarditis and should receive appropriate antibiotic prophylaxis.

Lastly, any precipitous fall in SpO_2 that fails to respond to the appropriate manipulations of increased FiO_2, volume administration and vasoactive medications necessitates an emergent echocardiogram. Shunt thrombosis, particularly in a patient with an mBT shunt, is a catastrophic occurrence and can require the emergent initiation of ECMO.

Clinical Pearl

Any precipitous fall in SpO_2 that fails to respond to the appropriate manipulations of increased FiO_2, volume administration and vasoactive medications necessitates an emergent echocardiogram to rule out shunt thrombosis.

How can anesthetics and PPV alter the balance of systemic and pulmonary blood flow?

In patients with normal pulmonary compliance, PVR can be readily reduced with mechanical ventilation. Accidental hyperventilation and hyperoxia are, unfortunately, common. Volatile anesthetics reduce blood pressure in a dose-dependent fashion through reductions in contractility, stroke volume, cardiac output, and SVR. This may result in a reduction in both Q_s and Q_p. Dexmedetomidine may reduce the need for anesthetics implicated in possible anesthetic neurotoxicity to the developing brain, but it should be noted that rapid boluses may result in a greater increase in SVR relative to PVR as well as bradycardia.

What are signs of inadequate Q_s? What are the likely causes and best initial treatments?

Inadequate Q_s can be identified by hypotension, mottled appearance, hepatomegaly, lactic acidosis, and low mixed venous oxygen content. This can result from either low PVR resulting in excess Q_p at the cost of Q_s, diminished total cardiac output, or both. Pulmonary blood flow may be reduced with ventilatory maneuvers such as decreasing FiO_2 and permissive hypercarbia to increase PVR. Cardiac output can be augmented by use of volume expansion and inotropic agents. Intraoperatively the most likely causes of inadequate systemic flow are diminished cardiac output due to anesthetic agents and excess reduction in PVR from excess ventilation or hyperoxia.

Clinical Pearl

Inadequate Q_s can be identified by hypotension, mottled appearance, hepatomegaly, lactic acidosis, and low mixed venous oxygen content. This can result from low PVR causing excess Q_p at the cost of Q_s, or through diminished total cardiac output, or both.

What are signs of inadequate Q_p? What are the likely causes and best initial treatments?

Inadequate Q_p is often marked by cyanosis and desaturation, as well as a possible increase in the gradient between $ETCO_2$ and $PaCO_2$. This may be related to globally reduced cardiac output, increased PVR, or reduced SVR. Treatments can include PVR reduction, increasing cardiac output and treatment of reduced SVR.

Pulmonary vascular resistance can be reduced with lung recruitment, increased FiO_2, and treatment of pain and/or acidosis. Treatment of low SVR often begins with vasopressors such as epinephrine or phenylephrine that increase SVR more than PVR. However, vasopressors must be used cautiously, as they may reduce end-organ perfusion, resulting in lactic acidosis and further impairing myocardial performance. Cardiac output can be increased through volume expansion and inotropic therapies.

What are other causes of desaturation besides decreased Q_p?

In addition to decreased Q_p, other contributing causes to desaturation can include pulmonary derecruitment and atelectasis, congestive heart failure with pulmonary edema, and inadequate oxygen delivery leading to low mixed venous oxygen saturation. Increasing oxygen delivery either via transfusion and/or increasing cardiac output can raise mixed venous oxygen saturation, resulting in increased arterial oxygen saturation. Anatomic obstruction to Q_p must also be considered. Shunt stenosis due to kinking or thrombosis is, unfortunately, common. Another less likely cause is obstruction to pulmonary venous return,

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which can be caused directly by pulmonary venous stenosis or downstream by a restrictive atrial septum resulting in high left atrial pressure.

Clinical Pearl

Increasing oxygen delivery either via transfusion and/or increasing cardiac output can raise mixed venous oxygen saturation, resulting in increased arterial oxygen saturation.

What are signs of simultaneously inadequate systemic and pulmonary blood flow and what treatments are indicated?

Patients may manifest any combination of the symptoms mentioned earlier. Treatment will require increased flow which may involve volume expansion including transfusion, along with inotropes and possibly inodilators such as milrinone to maximize perfusion. Veno-arterial extracorporeal membrane oxygenation (VA-ECMO) may be required if the above fail to improve oxygen saturation and perfusion.

What are the likely causes and initial treatment for hypoxemia and *hyper*tension?

Intraoperatively the onset of hypoxemia without hypotension or signs of reduced cardiac output is often the result of atelectasis. When hypoxemia is associated with hypertension, elevation in PVR causing a reduction in Q_p relative to Q_s is likely. Elevated PVR may result in sluggish shunt flow and should therefore be addressed. Reduction in PVR through utilization of anesthetic or analgesic agents and treatment of acidosis and hyperthermia may be helpful. Oxygen supplementation should be used judiciously in order to avoid hyperoxia and excessive Q_p. Hypoxemia may also reflect low mixed venous oxygen saturation. In this situation volume expansion, transfusion and inotropic therapy may also be required. Should hypoxemia and hypertension persist despite therapy, consideration should be given to the occurrence of a potential pulmonary hypertensive crisis, in which case inhaled nitric oxide would prove useful.

What physiologic changes occur with laparoscopy?

In healthy children, insufflation pressures of 6 mm Hg are not associated with hemodynamic changes by transesophageal echocardiography. However, insufflation pressures of 10 mm Hg reduced aortic blood flow and increased SVR without changes in blood pressure. Insufflation with pressures of 12 mm Hg reduced systemic ventricular systolic function and cardiac index. Insufflation also decreases lung compliance in children [5, 6].

During insufflation carbon dioxide (CO_2) uptake into the circulation occurs at a greater rate in children as compared to adults. Minute ventilation typically needs to be increased 20%–30% to offset some of this uptake. However, some of the CO_2 is buffered and released postoperatively, resulting in a sustained need for increased minute ventilation to prevent hypercapnia postoperatively [7]. Cyanotic patients normally have a greater difference between $PaCO_2$ and $ETCO_2$ than acyanotic patients due to reduced PBF and increased alveolar dead space. Insufflation worsens both of these factors. This results in an approximate doubling of the $PaCO_2$-$ETCO_2$ difference during laparoscopy [8].

Clinical Pearl

Cyanotic patients normally have a greater difference between $PaCO_2$ and $ETCO_2$ than acyanotic patients due to reduced PBF and increased alveolar dead space. Insufflation worsens both of these factors. This results in an approximate doubling of the $PaCO_2$-$ETCO_2$ difference during laparoscopy.

Would it be safer to perform this procedure with an open laparotomy?

In patients with CHD, multiple retrospective studies have shown fewer complications and lower mortality in patients undergoing laparoscopic procedures [9]. A study using national surgery quality data revealed lower morbidity and a trend toward lower mortality in patients with minor CHD undergoing laparoscopic procedures, but no difference in those with severe or major CHD. Laparoscopic procedures were associated with shorter length of stay and less transfusion. Patients with CHD have a higher rate of reintubation, infection, readmission, and mortality, with a roughly fourfold higher rate as compared to healthy patients. Laparoscopy in all patients with CHD appears to have a mortality of 20%–30% [10–13].

Regarding patients with single-ventricle physiology in particular, an older case series with open fundoplication described instances of hemodynamic instability, some requiring VA-ECMO cannulation and/or resulting in postoperative death [14]. A subsequent case series evaluating laparoscopic procedures found no such major complications. The need for empiric blood transfusion to increase hematocrit to >45% and occurrence of infection were quite

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common. In another subsequent case series, hemodynamic instability was very common, with one patient requiring VA-ECMO cannulation for shunt thrombosis [15]. For Nissen fundoplication in particular, surgical dissection near the diaphragm results in increased mediastinal and pleural pressures, worsening lung compliance and potentially further increasing intraoperative $PaCO_2$ [11].

Clinical Pearl

Laparoscopic procedures in patients with Stage I physiology may be technically challenging. Abdominal insufflation and positioning changes should be applied in a graded fashion to ensure patient tolerance.

If the patient developed refractory hypoxemia, what additional cause should be considered?

Shunt thrombosis is a life-threatening emergency that results in marked reduction or cessation of PBF. The ensuing hypoxemia and hypercarbia cause rapid deterioration of cardiac performance, shock, and cardiac arrest. Initial hypoxemia is associated with marked reduction and then loss of $ETCO_2$. Auscultation of the heart may reveal the loss of the characteristic shunt murmur. Bradycardia and ST segment changes are often the first manifestations of inadequate myocardial oxygen delivery.

Immediate treatments include:

- Administration of heparin, typically 100 units/kg IV
- Stat call for echocardiography to assess shunt flow
- Epinephrine boluses to attempt to dislodge thrombus
- Emergent consultations to cardiology and cardiac surgery for shunt recannulation in the catheterization lab or revision in the operating room
- Initiation of VA-ECMO cannulation

In patients with an open chest or a recent sternotomy that can be safely and rapidly reopened, direct massage of the shunt may dislodge the thrombus. Patients who develop cardiac arrest related to complete shunt obstruction may have no PBF and cannot be resuscitated without restoration of shunt flow or an alternative means of oxygenation, namely VA-ECMO.

Clinical Pearl

Decreases in cardiac output can precipitate shunt thrombosis, which is life-threatening and may require initiation of VA-ECMO and emergent cardiac catheterization or surgery to restore blood flow. Anesthetics, positioning, and laparoscopic insufflation may each independently cause reductions in cardiac output.

What are the intraoperative and postoperative analgesic options for this patient?

After confirming the absence of anticoagulation, neuraxial techniques including single shot and continuous caudal analgesia have been successfully used for in this patient population. Abdominal wall blocks may also be considered, and, at a minimum, local anesthetic should be infiltrated near incisions. Intravenous acetaminophen may also reduce postoperative opioid analgesic requirements. In young infants, the combination of non-steroidal anti-inflammatory drugs and acetaminophen may not reduce pain scores or opioid consumption compared to acetaminophen alone. The use of nonsteroidal antiinflammatory drugs is controversial, owing to their effects on platelet and renal function. Repeated dosing in young infants has also been associated with increased postoperative bleeding [16].

If postoperative ventilation is required, dexmedetomidine may provide both sedation and analgesia.

Adequate analgesia is essential as inadequate pain control can cause increased oxygen consumption and subsequently desaturation.

What is the appropriate postoperative disposition for this patient?

A discussion including the patient's primary cardiologist, surgeon, the intensivist who may receive the patient, and the anesthesiologist should occur preoperatively regarding postoperative disposition.

Some hospitals require ICU admission following general anesthesia for all Stage I patients, owing to their fragile physiology and significantly elevated mortality as compared to other phases of single-ventricle palliation. At a minimum, patients with risk factors for increased morbidity and mortality or evidence of poor cardiopulmonary reserve must be scheduled for ICU. Any patient with Stage I physiology undergoing elective or semielective procedures under general anesthesia should have an ICU bed readily available for postoperative care, as perhaps 25% of patients experience an escalation of care such as unplanned ICU admission [12]. Even with an uneventful perioperative course postoperative observation in an ICU setting would be recommended for this patient.

Should this patient be immediately extubated at the conclusion of surgery?

After fundoplication, patients exhibit impaired diaphragmatic function, which reduces pulmonary reserve. After laparoscopy or thoracoscopy, patients have transiently increased pulmonary demand to eliminate CO_2

https://doi.org/10.1017/9781108657341.027 Published online by Cambridge University Press

that was absorbed and buffered during insufflation. Patients with tachypnea and other signs of inadequate cardiopulmonary reserve at baseline may benefit from mechanical ventilation until excess CO_2 is eliminated and adequate postoperative analgesia and hemodynamics have been proven. However, the negative aspects of mechanical ventilation are numerous. Positive pressure ventilation impairs venous return and requires sedation, which may result in hypotension. Admission to the ICU and postoperative mechanical ventilation is associated with increased cost and length of stay. As presented, this patient is a candidate for extubation at the end of surgery should the perioperative course prove uneventful although he should still be admitted to an ICU setting.

Can this patient have this surgery at a hospital that does not have ECMO capability?

A known complication for a patient with shunt-dependent single-ventricle physiology having a Nissen fundoplication is the need to resuscitate using ECMO in the event of shunt thrombosis. Therefore, it would be prudent to have the procedure performed in a center with ECMO capability, although the ability to put the patient on CPB during resuscitation might be an alternative.

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Suggested Reading

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https://doi.org/10.1017/9781108657341.027 Published online by Cambridge University Press