Chapter 27

Bidirectional Glenn

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Case Scenario

A 9-month-old male infant weighing 8.4 kg presents for a complex hypospadias repair. His past medical history is significant for hypoplastic left heart syndrome. He has undergone two previous surgeries: a Stage I palliation (Norwood procedure) shortly after birth and three months earlier a bidirectional Glenn procedure or Stage II palliation. The history obtained from the parents reveals that he is doing well and developing appropriately and has no other significant medical problems. He is regularly seen as an outpatient by his pediatric cardiologist and his only current medication is acetylsalicylic acid. Current vital signs are: SpO_2 79% on room air, heart rate 115 beats/minute, blood pressure 80/48 mm Hg, and temperature 36.8°C. The operative time is expected to be approximately 3 hours.

His last transthoracic echocardiogram revealed the following:

- *A mildly dilated right ventricle with normal function*
- *Trivial atrioventricular valve regurgitation*

Key Objectives

- Understand the anatomy and physiology of a Stage II palliation or bidirectional Glenn procedure.
- Describe the path of blood flow in these patients.
- Understand how the manipulation of pulmonary and systemic vascular resistances affect systemic oxygen delivery.
- Understand the potential effects of different anesthetic perioperative management plans.
- Discuss the importance of ventilatory strategies in patients with a bidirectional Glenn procedure.

Pathophysiology

What is hypoplastic left heart syndrome?

Hypoplastic left heart syndrome (HLHS) encompasses an array of cardiac anomalies in which the development of the left ventricle (LV) and/or the mitral valve, systemic outflow tract, and ascending aorta are hindered in utero. While some institutions treat this lesion with primary heart transplantation, it is most commonly treated via a three-stage palliative pathway. Stage I is often referred to as the Norwood procedure and includes aortic reconstruction along with creation of a systemic-to-pulmonary artery shunt to provide pulmonary blood flow (PBF). Stage II palliation involves the creation of either a bidirectional Glenn shunt or a hemi-Fontan, replacing the systemic-to-pulmonary artery shunt with a superior cavopulmonary anastomosis (SCPA) to provide PBF. The third or final palliative stage is known as the Fontan procedure, and it routes inferior vena cava (IVC) flow directly to the pulmonary vascular bed. (See Chapter 26.) After completion of the Fontan procedure all PBF is now passively supplied to the pulmonary vascular bed, allowing the single ventricle to provide systemic blood flow.

What is a bidirectional Glenn shunt?

The Glenn shunt is a form of SCPA named for William Glenn, the surgeon who first performed the procedure in 1958. It routes deoxygenated blood from the superior vena cava (SVC) to the right pulmonary artery. A modification of the Glenn shunt was developed in 1973 by Dr. Gaetano Azzolina to facilitate flow to both the left and right pulmonary arteries, leading to the term "bidirectional" Glenn (BDG) shunt. This procedure is now the second stage of the three-stage palliation for HLHS.

Why are single-ventricle palliative surgeries staged in this way?

Following Stage I Norwood palliation for HLHS the RV has a volume burden, as it supports both the pulmonary and systemic circulations in parallel. Over time, this can adversely affect ventricular function and manifest as RV dysfunction accompanied by tricuspid regurgitation (TR) due to malcoaptation of the valve leaflets secondary to distortion of the tricuspid annulus. During Stage I palliation the pulmonary

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vasculature is also exposed to excessive pressures due to blood flow from the shunt: either systemic arterial (modified Blalock–Taussig shunt) or ventricular (Sano shunt).

Low pulmonary vascular resistance (PVR) is required for passive PBF via either the BDG or hemi-Fontan to be successful, and therefore the Stage II procedure is usually performed around 3–6 months of age. At this point in the infant's development, the pulmonary vasculature has matured and PVR has fallen to sufficiently low levels to permit adequate passive PBF. Completion of this stage reduces the volume loading on the heart and wall stress on the RV, which over time allows for RV remodeling. Subsequently, improvement of RV function should occur along with reduction of TR from improved tricuspid valve leaflet coaptation. These factors are important for the long-term function of the single RV.

Stage III (Fontan completion) surgery is completed usually around the age of 18 months to 4 years, and baffles IVC flow into the pulmonary circulation such that all systemic venous return now directly enters the pulmonary bed.

Is a superior cavopulmonary anastomosis performed only for HLHS patients?

Congenital cardiac patients with functionally univentricular hearts that are not suitable for biventricular repair can also be palliated via the single-ventricle pathway. In these patients, one of the ventricles is hypoplastic. The dominant ventricle could be either the LV, the RV, or of indeterminate morphology. All of these groups may undergo single-ventricle pathway palliation.

Why is a Stage II palliation performed and how are the goals achieved?

Physiologic objectives of second stage palliation are twofold:

- Creation of a lower pressure source of PBF that will grow with the patient
- Reduction of the volume burden on the single ventricle

Following this surgery, the single ventricle only needs to support the systemic cardiac output as blood flow to the lungs is now passive.

The systemic-to-pulmonary artery shunt (high-pressure arterial) that was created during the Stage I palliation is ligated. The SVC is then anastomosed to the pulmonary artery (PA), establishing the new source of PBF. The resulting circulation establishes passive flow of deoxygenated blood from the upper body via the SVC to the pulmonary arteries as the source of PBF.

Clinical Pearl

Anastomosis of the SVC to the PA establishes passive flow of deoxygenated blood from the upper body via the SVC to the pulmonary arteries as the source of PBF.

How does the BDG procedure differ from the hemi-Fontan?

Although the physiologic goals are the same, with provision of SVC blood to the pulmonary vascular bed, the BDG and hemi-Fontan differ anatomically. (See Figure 27.1.)

- ***Bidirectional Glenn procedure*** (see Figure 27.2):

1. The SVC is divided from the RA at the superior cavoatrial junction and the atrial end is oversewn.
2. The RPA is incised, and the posterior wall of the SVC is anastomosed to the superior edge of the PA.

Following a BDG, the completion Fontan operation typically involves the creation of an ***extracardiac conduit*** wherein the IVC is anastomosed to the PA or distal SVC.

- ***Hemi-Fontan procedure*** (see Figure 27.3):

1. The natural SVC-to-RA confluence is preserved, and a side-to-side anastomosis is used to join the SVC and RPA.
2. Homograft tissue is used to augment the branch pulmonary arteries, creating a dam across the superior cavoatrial junction, and preventing blood flow between the SVC and RA.
3. The SVC-RA junction is also enlarged to match the size of the IVC.

The hemi-Fontan addresses any PA hypoplasia, and later simplifies the completion of a ***lateral tunnel Fontan***. At the time of Fontan completion, the dam is excised, and a polytetrafluoroethylene patch is used to create a division in the atrium such that IVC flow is tunneled toward the pulmonary arteries and pulmonary venous return is directed across the atrioventricular valve.

What is the circulatory path for blood after Stage II palliation?

Deoxygenated blood from the lower body enters the common atrium via the IVC. Deoxygenated blood from the upper extremities, head, and neck passively drains via the SVC into the pulmonary artery. It then traverses the pulmonary vasculature and becomes oxygenated. This oxygenated blood drains via the pulmonary veins into the common atrium and mixes with the deoxygenated blood from the IVC. Blood then passes via the atrioventricular valve into the ventricle. The dominant ventricle (the RV in

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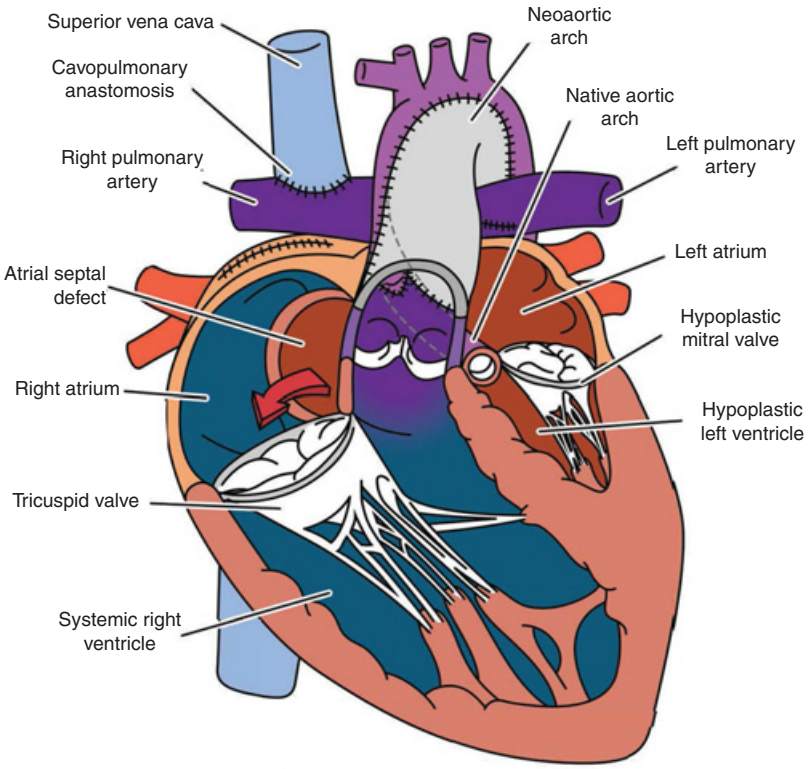

Figure 27.1 Hypoplastic left heart syndrome, Stage II palliation, superior cavopulmonary anastomosis (bidirectional Glenn). Drawing by Ryan Moore, MD, and Matt Nelson.

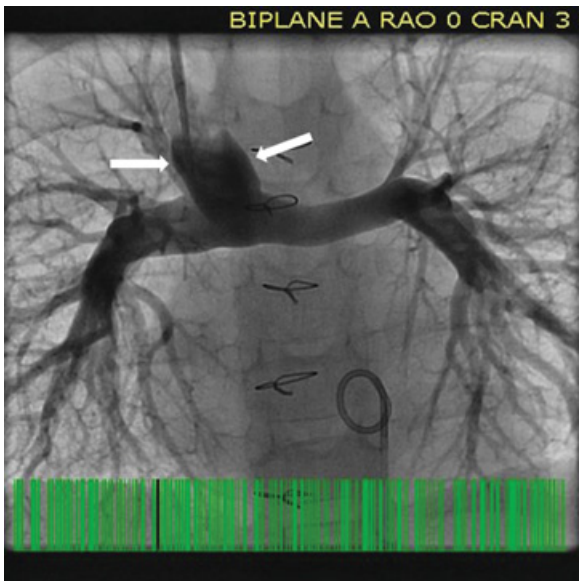

Figure 27.2 Bidirectional Glenn procedure. An angiogram is performed in AP projection in the SVC in a patient with a Glenn shunt. The direct anastomosis of the SVC to the pulmonary artery is noted (arrows). Courtesy of Russel Hirsch, MD.

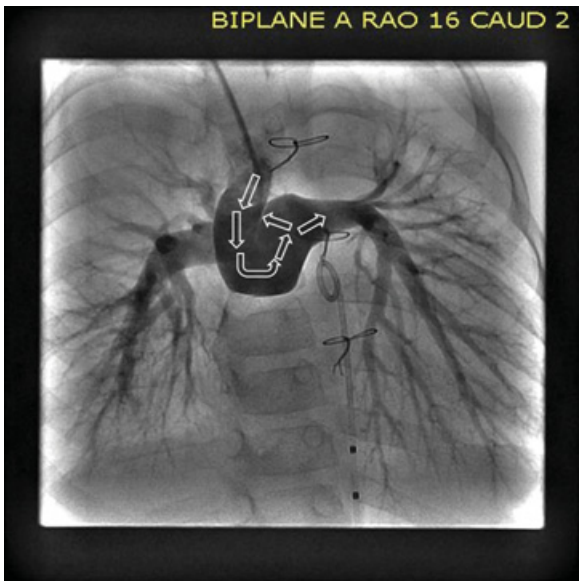

Figure 27.3 Hemi-Fontan. An angiogram is performed in AP projection in the SVC in a patient with a hemi-Fontan. The angiogram demonstrates the anterior course of the SVC with reversed curve anastomosis into the pulmonary arteries. The arrows demonstrate the direction of blood flow. Courtesy of Russel Hirsch, MD.

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patients with HLHS) then ejects blood into the systemic circulation.

What are the expected oxygen saturations in patients with a bidirectional Glenn shunt?

The superior systemic venous circulation and the pulmonary circulation are in series; therefore only blood from the SVC travels to the lungs and is oxygenated. Due to the atrial septectomy that has been previously performed in these patients, oxygenated blood returning to the atrium via the pulmonary veins mixes with deoxygenated systemic venous return from the IVC. This mixing results in systemic oxygen saturations in the range of 75%–85%. Therefore, oxygen saturations after Stage II palliation do not differ significantly from saturations prior to Stage II palliation.

Over time, patients with SCPA connections may form veno-venous collaterals and pulmonary-venous connections, which can result in worsening cyanosis [1]. Prior to an anesthetic it is important to be aware of these as possible causes when greater than expected levels of hypoxemia are observed in BDG patients. In addition to the formation of collaterals, a persistent left SVC to coronary sinus connection may also be a cause of cyanosis. Cardiac catheterization and echocardiography data should be assessed in patients who have lower than expected systemic saturations to elucidate the cause.

Clinical Pearl

Oxygenated blood returning to the common atrium via the pulmonary veins mixes with deoxygenated systemic venous return from the IVC. This mixing results in systemic oxygen saturations ranging from 75% to 85%. Therefore, oxygen saturations after Stage II palliation do not differ significantly from saturations prior to Stage II palliation.

Anesthetic Implications

What specific areas should be evaluated in the preoperative assessment?

In addition to a thorough anesthetic preoperative assessment, attention should focus on establishing an understanding of the primary congenital cardiac lesion, the anatomy, and the level and direction of both intracardiac and extracardiac shunts. Furthermore, it is crucial to successful perioperative management to understand and appreciate the effects on systemic tissue oxygen delivery of manipulating both the pulmonary and systemic vascular resistances.

Previous anesthetic records, if available, should also be reviewed with particular attention to the airway management history. Previous intubations and the duration of time spent intubated after previous surgeries, particularly if prolonged, can increase the risk of subglottic stenosis.

How is functional status assessed?

Functional assessment involves evaluating appropriate development, weight gain, attainment of milestones, and reviewing for signs of heart failure. Heart failure can result when the heart is either pressure and/or volume overloaded. These signs differ with age. An infant in heart failure may present with poor feeding, failure to gain weight, and hepatomegaly. Signs that are common to all ages include tachypnea, tachycardia, diaphoresis, and cool extremities.

What should be done if the patient has a respiratory tract infection?

Pediatric patients with recent or active respiratory tract infections are at increased risk for perioperative respiratory complications [2]. In patients with a BDG, the presence of upper or lower respiratory tract infections can have additional hemodynamic ramifications, as infections can increase airway reactivity and reduce pulmonary compliance. Both are poorly tolerated and can lead to intraoperative complications that may arise from difficulty maintaining adequate PBF. Acute increases in PVR resulting from bronchospasm in a patient with an SCPA will present as increased cyanosis, coupled with hypotension from reduced preload. Elective surgery should not be undertaken in the presence of a significant upper or lower respiratory infection, and the case should be rescheduled for 4–6 weeks after resolution of symptoms, allowing airway reactivity to return to normal.

Clinical Pearl

In patients with a BDG, the presence of upper or lower respiratory tract infections can have additional hemodynamic ramifications as they can increase airway reactivity and reduce pulmonary compliance. Both are poorly tolerated and can lead to complications intraoperatively that may arise from difficulty maintaining adequate PBF.

What medications might the child be taking? Which medications should be withheld prior to surgery?

Polycythemia resulting from chronic cyanosis increases whole blood viscosity, reducing blood flow in small arterioles and capillaries. This places these patients at an

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increased risk of thrombus formation. As a result, patients with a BDG shunt are often taking antiplatelet agents or anticoagulants.

Antiarrhythmic agents, diuretics, and antihypertensives are also frequently used in patients with congenital heart disease (CHD). In particular, angiotensin converting enzyme inhibitors (ACEi) are frequently prescribed for patients with single-ventricle physiology. Evidence for preoperatively withholding these in children is lacking and as a result most guidelines regarding the perioperative administration of ACEi in pediatric patients have been adapted from adult practice. As such, most ACEi are withheld on the morning of surgery.

When considering which medications to continue or withhold, it is advisable to review each case on an individual basis. Advice from the child's pediatric cardiologist should be sought should questions arise.

What preoperative investigations are required?

All available preexisting cardiac data for the patient should be reviewed, including a recent echocardiogram and any cardiac catheterization data. Specific areas of interest on the echocardiogram include assessment of ventricular systolic function (ejection fraction) and atrioventricular valve competence. The patient should also be in compliance with requested cardiology follow-up visits.

An electrocardiogram (ECG) should be evaluated to assess for normal sinus rhythm and conduction abnormalities. Arrhythmias are generally poorly tolerated in patients with CHD, particularly those with single-ventricle physiology. Furthermore, single-ventricle patients with ventricular ectopy have an approximately 30% higher incidence of intraoperative arrythmias resulting in mortality and adverse outcomes [3]. Therefore, the patient with an abnormal ECG should be considered at an increased risk of developing intraoperative arrhythmias.

Available laboratory data should be reviewed. If not recently performed, it is reasonable to obtain a basic metabolic profile and a complete blood count prior to surgery. Although significant blood loss would not be anticipated in this surgery, it is booked for 3 hours.

What is the expected hematocrit and why might it be higher than normally expected?

Polycythemia as a result of ongoing cyanosis is expected, and therefore a hematocrit in the range of 40%–45% is often seen. The transfusion threshold for patients with single-ventricle physiology should be lower than that in a noncyanotic patient; in order to preserve oxygen carrying capacity and tissue oxygen delivery the desired hematocrit is greater than 40%. Maintaining a higher hematocrit also increases the arterial oxygen saturation in a child with this physiology by improving the mixed venous saturation.

Clinical Pearl

The target hematocrit for patients with Stage II physiology is 40%–45%.

Are there specific preoperative fasting considerations?

While no specific changes should be made to the institutional and nationally accepted preoperative fasting times in a patient with a BDG, normal preload is important for maintenance of stable hemodynamics. Therefore, fasting for clear fluids should be minimized, and if prolonged fasting times are unavoidable, the preoperative institution of intravenous (IV) fluids should be considered. Dehydration should be avoided, as cyanotic patients become polycythemic as an adaptive response, which can cause hyperviscosity and increased risk for thrombus formation.

Should anxiolytic premedication be utilized?

At 9 months of age, infants are not generally likely to experience separation anxiety from the parents or caregivers and often respond well to simple maneuvers such as being held and hearing soothing voices. Therefore, routine premedication in this age group may not be necessary. However, cases should be assessed on an individual basis. If premedication is deemed necessary, patients with BDG shunts can be safely premedicated with an oral or intranasally administered benzodiazepine.

The use of premedication can be helpful in the extremely anxious child as it can aid in decreasing oxygen consumption. In addition, it may reduce the dose of induction agent required, which in turn limits the decrease in systemic vascular resistance (SVR) associated with some anesthetic induction agents.

Is endocarditis prophylaxis required?

In 2017, a focused update of the 2014 guidelines released by the American College of Cardiology and American Heart Association Task Force stated that in the absence of active infection there is no evidence to suggest any benefit from the routine use of antimicrobial agents for infective endocarditis (IE) prophylaxis in gastrointestinal or genitourinary procedures. However, it is their consensus opinion that the use of antimicrobial prophylaxis in patient

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populations who are either at increased risk of developing IE or at a higher risk of adverse outcomes from contracting IE is not unreasonable. Patients with CHD, either unrepaired or repaired, who have shunts or valvular regurgitation at or adjacent to a prosthetic patch or device would be considered appropriate for IE prophylaxis when undergoing certain dental procedures [4].

What intraoperative monitors should be used? Are invasive pressure monitors required?

Stable BDG patients with normal ventricular function do not require invasive monitors for hypospadias repairs, and the use of standard recommended American Society of Anesthesiologists monitors is appropriate.

During other major surgical procedures, should the use of invasive monitoring be required, placement of arterial access in the upper extremity on the ipsilateral side as a previous modified Blalock–Taussig shunt should be avoided. Residual stenosis may exist at the site of the previous shunt that can cause an erroneously low blood pressure reading. Should central venous access be required it should be remembered that placement of an internal jugular line will yield PA and not atrial pressures. Institutional preferences exist, and some centers may prefer to avoid utilizing the internal jugular and subclavian vessels to mitigate the risk of causing thrombosis or stenosis and reducing SVC drainage into the pulmonary arteries.

Clinical Pearl

Should central venous access be required in a patient with an SCPA it should be remembered that placement of an internal jugular line will yield PA and not atrial pressures.

What are the intraoperative management goals for a child with a BDG?

General goals for the intraoperative management of HLHS patients post Stage II palliation are:

- ***Maintain adequate preload.*** Should hypotension be encountered intraoperatively, this should be treated initially with IV fluids. Insensible fluid losses, fluid deficit from preoperative fasting, and ongoing blood loss should be accounted for and replaced.
- ***Maintain low PVR*** to maximize passive PBF. In addition, a high-normal pCO_2 results in cerebral vasodilatation and therefore augments cerebral blood flow, in turn increasing venous return to the pulmonary vasculature via the SVC. When using mild hypoventilation, it is important to avoid significant acidosis, which increases PVR.
- ***Avoid acute increases in PVR and SVR*** from sympathetic surges resulting from noxious stimuli.
- ***Preserve cardiac contractility*** and maintain a normal or low SVR. Using a balanced anesthetic technique is preferable when aiming to achieve stable hemodynamics.
- ***Maintain coronary perfusion pressure*** and preserve myocardial O_2 supply/demand balance.
- ***Avoidance of air embolus*** (paradoxical air embolus). All IV administration lines and connectors should be deaired, and medication administration sites should be free of air.
- ***Maintain normal sinus rhythm*** and avoid arrhythmias.

Clinical Pearl

A high normal pCO_2 results in cerebral vasodilatation and therefore augments cerebral blood flow, in turn increasing venous return to the pulmonary vasculature via the SVC. When using mild hypoventilation, it is important to avoid significant acidosis, which increases PVR.

Does the SCPA affect cerebral perfusion pressure in single-ventricle patients?

In single-ventricle patients, cerebral autoregulation remains intact. However, the modification to the venous return pathway from brain to the heart via the SVC to PA anastomosis has implications for cerebral perfusion pressure (CPP). Cerebral perfusion pressure has typically been described as mean arterial pressure minus intracranial pressure (ICP); jugular venous pressure or CVP can be used in place of ICP if their numerical values are higher. In the SCPA circulation, the following equation more accurately depicts CPP (if PAP is higher than ICP):

$$\mathbf{CPP = MAP - mPAP,}$$

where CPP = cerebral perfusion pressure; MAP = mean arterial pressure; and mPAP = mean pulmonary arterial pressure. It can be seen from this equation that factors that decrease MAP or increase ICP and PAP will decrease CPP. Infants with SCPA usually have low PVR and therefore low PA pressures. In infants with a BDG or hemi-Fontan who have low pulmonary pressures, CBF can be increased by maintaining slight hypercarbia, which dilates cerebral blood vessels and promotes greater blood flow in the SVC, thus increasing oxygenated venous return to the heart. However, in all Stage II single-ventricle palliation patients, to maintain adequate CPP and adequate systemic

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oxygenation, PAP should be kept low and MAP maintained near normal.

Occasionally a patient is unable to progress past the BDG circulation to Stage III (Fontan) palliation due to elevated PA pressures. In these patients, the presence of elevated PA pressures and low MAP places them at significant risk of cerebral ischemia intraoperatively from inadequate cerebral perfusion. In this scenario, it is crucial to avoid decreases in SVR and PVR should be kept as low as possible. Older patients with SCPA should be assumed to have elevated PA pressure and should be cared for in specialist centers.

Clinical Pearl

Factors that decrease MAP or increase ICP and PA pressures will decrease cerebral perfusion pressures in the patient with a BDG. To maintain adequate cerebral perfusion pressure and adequate systemic oxygenation, PA pressures should be kept low and MAP maintained near normal.

Which pharmacologic agents are suitable for induction of anesthesia?

Either inhalational induction or IV induction of anesthesia is suitable. The exact choice of inhalational or IV induction agent matters less than the expertise by which it is administered.

Inhalational induction with sevoflurane and an air/oxygen mixture is appropriate. The use of nitrous oxide is not contraindicated, but many anesthesiologists avoid its use in congenital cardiac patients with shunts. First, it can cause gas bubble expansion, which can potentiate the harmful effects of inadvertent intravenous gas embolus. Second, it can cause constriction of pulmonary vascular smooth muscle, and so may increase PVR.

Propofol decreases SVR and MAP. In children with CHD, this can worsen right-to-left shunting and reduce systemic oxygen saturations. Furthermore, the reduction in MAP can decrease coronary perfusion pressure, which, if significant enough, can impair myocardial function and precipitate arrhythmias. However, with awareness of the potential detrimental effects and the use of smaller doses, it can be used safely in this patient group in patients with normal ventricular function.

Other induction agents such as etomidate and ketamine, barbiturates, and volatile agents such as isoflurane and desflurane can be used safely in this patient population with awareness of their mechanisms of action, influence on PVR and SVR, and adverse effects.

How can PVR be manipulated?

In patients with single-ventricle physiology who have undergone a BDG, the sole source of PBF is passive drainage from the SVC. As a result, an understanding of how to manipulate PVR is of the utmost importance intraoperatively as increases in PVR or intrathoracic pressure will negatively impact PBF.

Pulmonary vascular resistance is influenced by pCO_2, pO_2, pH, temperature, mean airway pressure, atelectasis, and sympathetic stimulus. Factors increasing PVR include acidosis, high pCO_2, high mean airway pressure, hypothermia, basal atelectasis, increased sympathetic stimulus, and a low pO_2. To maintain a low PVR use a higher inspired O_2 concentration, avoid significant hypoventilation and acidosis, maintain low mean airway pressures and normothermia, and avoid sympathetic surges.

Clinical Pearl

Optimal pulmonary blood flow in patients with a BDG occurs when the PVR is low. Factors negatively impacting PVR include acidosis, high pCO_2, high mean airway pressure, hypothermia, basal atelectasis, increased sympathetic stimulus, and a low pO_2.

How should ventilation be managed?

The key to successful management of ventilation in HLHS patients with a BDG shunt is understanding that in this patient population, PBF occurs passively. With this in mind, it becomes clear that factors that influence PVR and intrathoracic pressure gradients have a major role in influencing PBF.

Spontaneous ventilation is beneficial as the negative intrathoracic pressure generated by inspiration augments venous return via the SVC to the PA, improving PBF. However, the use of a spontaneous breathing technique may be limited by the duration and type of operative procedure and patient positioning. The eventual loss of alveolar recruitment and basal atelectasis over long cases needs to be considered when formulating an appropriate ventilation strategy.

The use of positive pressure ventilation (PPV) is possible in individuals with a BDG. Although using this technique will eliminate the augmentation of PBF associated with spontaneous inspiration, it allows for greater control over the end-tidal carbon dioxide ($ETCO_2$) concentration and enables the anesthesiologist to optimize positive end-expiratory pressure (PEEP) and maintain alveolar recruitment. Ideally, low peak inspiratory pressures should be maintained by ventilating on the optimal (steep) part of the compliance curve. Maintaining

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low peak inspiratory pressures and using a slightly prolonged expiratory phase will help to facilitate PBF, which occurs during expiration with PPV [5].

In the patient with a BDG, maintaining slight hypercarbia ($ETCO_2$ concentration of 45–55 mm Hg) results in vasodilation of cerebral blood vessels and increases cerebral blood flow [6], which in turn increases the volume of deoxygenated blood returning to the PAs via the SVC. It appears that the additional PBF that results from mild hypercarbia induced cerebral vasodilation outweighs the negative effects of the mild acidosis on increasing the PVR.

In summary, when PPV is employed, the following principles should be used:

- Maintain low PVR.
- Maintain low intrathoracic pressure.
- Maintain a target $ETCO_2$ of 40–45 mm Hg.
- Use a slightly prolonged expiratory time to allow extra time for PBF to occur.
- Optimize tidal volumes and PEEP to achieve optimal compliance of gas exchanging units.
- Maintain adequate minute ventilation.

Clinical Pearl

Although spontaneous ventilation can be beneficial, the use of a spontaneous breathing technique may be limited by the duration and type of operative procedure and patient positioning. The eventual loss of alveolar recruitment and basal atelectasis over long cases needs to be considered when formulating an appropriate ventilation strategy.

Can a laryngeal mask airway be used?

General anesthesia with spontaneous ventilation via a laryngeal mask airway (LMA) can be effectively used in HLHS patients with BDG shunts. It provides the advantage of maintaining the negative intrathoracic pressure associated with inspiration which in turn helps augment PBF in patients with a BDG shunt. However, this needs to be balanced against the duration of surgery and the inevitable derecruitment and atelectasis that can occur over prolonged cases with spontaneous ventilation and LMA usage. In addition, many anesthesiologists prefer not to use supraglottic airway devices in infants.

What happens if laryngospasm or bronchospasm occurs?

In BDG patients the source of preload for the heart is derived from two sources: deoxygenated blood from the IVC and blood from the SVC which has become oxygenated after passing through the pulmonary vasculature. Laryngospasm or bronchospasm will cause an acute and abrupt increase in PVR with a resultant decrease in PBF. In the patient with a BDG, both hypotension and marked oxygen desaturation will be evident, resulting from the reduction in oxygenated preload. However, in the BDG circulation, the heart will still receive some preload in the form of deoxygenated blood from the IVC. Thus, both laryngospasm and bronchospasm should be aggressively managed in order to restore the passive flow of oxygenated blood from the pulmonary vasculature.

What are the analgesic options for this patient?

Intravenous analgesics are suitable, preferably a multimodal strategy, which will help reduce the total opiate dose. Postoperative respiratory depression in children with BDG shunts should be avoided, as it will lead to hypoxia from alveolar hypoventilation and hypercarbia. This in turn will raise the PVR and further impede PBF.

Local anesthetics are also a very useful adjunct. Caudal or penile blocks are both appropriate for hypospadias repairs and provide excellent intraoperative and postoperative analgesia. Prior to considering any regional or neuraxial analgesic technique, the anticoagulant medication history should be reviewed to ensure there are no contraindications to performing the procedure. Local and national guidelines regarding safety of regional anesthesia in patients on anticoagulants and antiplatelet agents should be followed.

Are there any specific considerations for emergence from anesthesia?

While tracheal extubation is generally performed with the patient awake, if an LMA was utilized it may often be safely removed with the child still in a deep plane of anesthesia. As with the initial choice of airway utilized for the procedure, decision making regarding the timing of airway removal at the end of the procedure should be individualized and based on the child's airway characteristics as noted during induction, the duration of surgery, and the intraoperative course. Should the decision be made to remove either an endotracheal tube or an LMA with the patient "deep," most practitioners then elect to remain in the operating room until the patient has emerged from anesthesia. If doubt exists regarding any of these factors, the airway should be removed with the patient awake in the operating room environment.

What are the discharge criteria for patients with Stage II palliation?

Standard local guidelines should be used when determining if pediatric patients meet discharge criteria following surgical intervention. Additional considerations for BDG

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patients that could result in an adverse outcome include residual sedation and poor tolerance of hypovolemia from emesis and/or poor oral intake. If the infant has returned to baseline, has good analgesia, and is tolerating feeds appropriately, they may be appropriate for discharge. However, the length of the surgical procedure is an important factor to consider, and with a complex repair of 3 or more hours duration it would be reasonable to admit for overnight observation following the procedure. Of note, a careful handoff of care should be completed with recovery staff and expected vitals and ranges for blood pressure, heart rate, and oxygen saturations (75%–85%) should be discussed.

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