Chapter

71

Noncardiac Surgery in a Glenn Patient

Jaime Bozentka and Laura Diaz-Berenstain

A three-year-old female is scheduled for laparoscopic appendectomy. She has been vomiting for the past 24 hours and has had minimal oral intake. She has a history of hypoplastic left heart syndrome and her last cardiac surgery was a Glenn procedure performed at six months of age. Her mother states that she is normally active and has no trouble keeping up with her five-year-old brother. They are regularly seen by a cardiologist at a hospital two hours from home.

Her current vital signs are: heart rate 140 beats/min; respiratory rate 20 breaths/min; SpO_2 78% on room air. Her abdomen is tender, but not distended. Her hemoglobin is 15 and her hematocrit is 45%. All other laboratory work is within normal limits. A bedside echocardiogram in the emergency department shows normal ventricular function and trace atrioventricular valve regurgitation.

What Is the Anatomical Name and Description of a Bidirectional Glenn Procedure? Why Is It Described as "Bidirectional?"

First introduced in 1958 by William Glenn, superior cavopulmonary anastomosis (SCPA) is a procedure in which the superior vena cava (SVC) is disconnected from the atrium and anastomosed to the right pulmonary artery (Figure 71.1). The main pulmonary artery, if not atretic, is disconnected from the heart, and other surgically created sources of pulmonary blood flow, such as an existing systemic to pulmonary shunt, are removed. It is referred to as "bidirectional" because blood from the SVC flows to the right pulmonary artery as well as across to the left pulmonary artery, thereby supplying both lungs.

What Type of Cardiac Lesions Require SCPA?

SCPA is typically the second stage in the palliation of single ventricle cardiac lesions. Patients may require single ventricle palliation for a variety of pathologies including hypoplastic right heart lesions (tricuspid atresia, pulmonary atresia with intact ventricular septum, severe forms of Ebstein's anomaly with tricuspid dysplasia), hypoplastic left heart lesions (aortic atresia or stenosis, mitral atresia, or stenosis), or lesions which do not permit surgical septation of the heart into two ventricle physiology (unbalanced atrioventricular canal defects, double inlet ventricle, some forms of heterotaxy).

The Flow of Blood to the Heart in the Glenn Patient

All blood flow to the pulmonary circulation is passive and supplied from the upper body via the SVC anastomosis to the pulmonary arteries. Because patients have had a complete atrial septectomy as part of their first stage procedure, when oxygenated blood returns to the heart from the pulmonary veins, it mixes with deoxygenated blood returning from the lower body via the inferior vena cava (IVC) and then proceeds to the single ventricle.

What Are Typical Hemoglobin–Oxygen Saturations for a Patient with SCPA Physiology?

Typical hemoglobin–oxygen saturations for a patient with Glenn physiology are between 75 and 85%.

What Is the Typical Hematocrit in a Patient with SCPA Physiology?

Due to the cyanotic nature of the physiology and the need to maximize oxygen delivery, the hematocrit of

https://doi.org/10.1017/9781108668736.072 Published online by Cambridge University Press

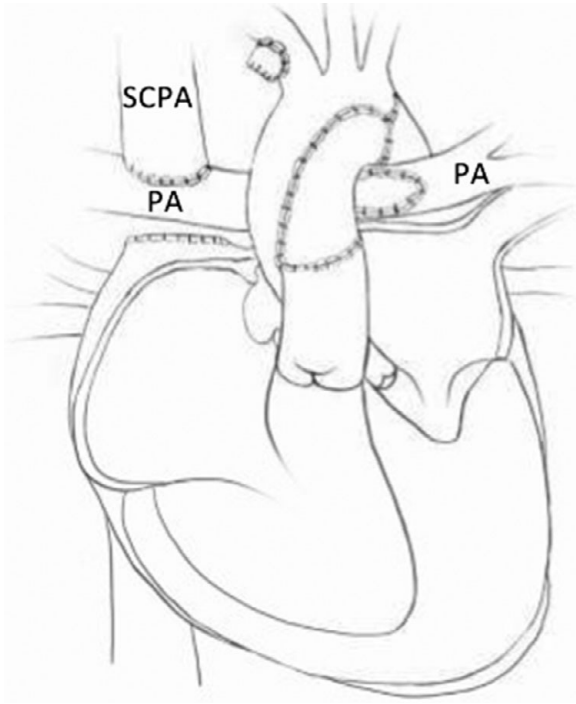

Figure 71.1 A schematic of hypoplastic left heart syndrome following superior cavopulmonary anastomosis (SCPA) or the Glenn procedure highlighting connection of the SVC and pulmonary artery (PA). Printed with permission from Texas Children's Hospital

SCPA patients is generally expected to be maintained between 40% and 45%.

At What Age Is SCPA Generally Performed? Why Is It Not Performed on Newborns?

The SCPA/Glenn procedure is usually performed at two to six months of age, as pulmonary vascular resistance (PVR) continues to fall after birth and passive pulmonary blood flow becomes possible. It cannot be performed earlier due to high PVR in newborns that would preclude successful passive pulmonary blood flow.

What Physiologic Advantage Does SCPA Offer Over Native Circulation or a Systemic-to-Pulmonary Arterial Shunt?

Prior to SCPA, blood exits the single ventricle via an outflow tract that then provides flow to both the systemic and pulmonary vascular circulations, with their relative resistances determining the amount of flow to each. This arrangement, known as *parallel circulation*, places a volume burden on the ventricle. SCPA places the two vascular beds in series by making pulmonary blood flow a diversion of systemic venous return. This effectively reduces the volume load on the ventricle, which in turn allows ventricular remodeling to occur, improving ventricular function and lowering end diastolic pressure. In addition, the transition to a *series circulation* also eliminates reliance on the balance between systemic and pulmonary vascular resistance to provide adequate flow to each system.

In comparison to a modified Blalock–Taussig (BT) shunt, which is made of synthetic Gore-Tex, the native tissue of the SVC provides a more stable source of pulmonary blood flow that is able to grow with the patient and is less likely to be compromised by thrombosis.

What Is the Final Stage in Single Ventricle Palliation?

At two to five years of age, the patient will undergo total cavopulmonary anastomosis (TCPA), also known as the Fontan procedure. This surgery results in all systemic venous return being diverted passively to the pulmonary system by connecting the inferior vena cava (IVC) to the pulmonary artery, most often via an extracardiac conduit. After undergoing the Fontan procedure most single ventricle patients will have hemoglobin-oxygen saturation in the high 90s.

Why Is SCPA Considered More Stable for Noncardiac Surgery than TCPA?

In comparison to TCPA, cardiac output after SCPA is not entirely dependent on pulmonary blood flow because the IVC remains directly connected to the heart, providing venous return. Therefore, should pulmonary blood flow be compromised by an increase in PVR, such as during bronchospasm or light anesthesia, cardiac output can be maintained via IVC flow. After TCPA, an abrupt increase in PVR could result in a precipitous decrease in pulmonary blood flow and preload to the single ventricle, unless a fenestration was created at the time of surgery, which allows IVC blood to potentially divert from the extracardiac conduit to the atrium. In TCPA patients with a fenestration, blood can shunt from the conduit to the single ventricle, as the resultant decrease in hemoglobin-oxygen

https://doi.org/10.1017/9781108668736.072 Published online by Cambridge University Press

saturation is better tolerated than low cardiac output. These patients may have hemoglobin-oxygen saturation in the 85–95% range.

If SCPA Is More Stable, Why Is Completion of the TCPA Necessary?

Total cavopulmonary anastomosis becomes necessary because of a progression in cyanosis that is inevitable following SCPA. The cause of this increased cyanosis is two-fold. Firstly, as the child ages and begins walking, the ratio of blood flow through the SVC to that in the IVC begins to favor the IVC, resulting in a greater proportion of deoxygenated blood return to the heart. Secondly, due to the lack of hepatic blood flow to the lungs, all SCPA patients form pulmonary arteriovenous malformations (AVM) which result in increased intrapulmonary shunting.

What Should I Tell Parents about Anesthetic Risk?

According to the Pediatric Perioperative Cardiac Arrest Registry data, children with heart disease, including those who have undergone SCPA, have a higher incidence of cardiac arrest during anesthesia than those without heart disease. In addition, more than half of these cardiac arrests occurred in the general OR rather than the cardiac OR or cardiac catheterization laboratory. For this reason, a comprehensive preoperative evaluation is critical in order to identify and anticipate any situations that would incur additional risk in this already compromised population.

Are Patients with SCPA Physiology Appropriate for Outpatient Surgery, or Surgery at a Surgery Center?

An SCPA patient who is developing normally, without functional restrictions, and undergoing regular evaluation by a pediatric cardiologist can be an appropriate candidate for same day surgery for low-risk procedures where there is minimal expectation for postoperative pain or functional limitations. However, common post-anesthetic complications will be less well-tolerated in this patient population compared to healthy children. Even mild bleeding or vomiting can lead to dehydration and a reduction in pulmonary blood flow. Similarly, increased pulmonary vascular resistance with a resultant increase in hypoxemia can result from unrecognized upper respiratory infection (URI), bronchospasm, or surgical complications. Therefore, planned outpatient procedures should be done at a hospital which allows postoperative monitoring and admission if necessary, rather than at a surgery center.

What Information Should Be Gathered in a Preoperative Cardiac Evaluation in an SCPA Patient?

The history should focus on determining whether any degree of functional limitation exists due to the cardiac disease. In younger patients, this may manifest as tachypnea, diaphoresis with feeds, or failure to thrive. In older children dyspnea on exertion or the inability to keep up with peers may be noted. Preoperative vital signs, in particular the hemoglobin-oxygen saturation while on room air, will help to establish baseline values to maintain during the perioperative period. The most recent report from the patient's cardiologist should be sought, as well as a recent electrocardiogram and echocardiogram. Gather a complete medication history as patients may be on a variety of agents such as antihypertensives, diuretics, or anticoagulants, and occasional patients may be on anti-arrhythmic medications.

When Reviewing the Electrocardiogram and Echocardiogram, What Are the Most Significant Indicators of Cardiac Status?

Single ventricle patients generally do not tolerate cardiac arrhythmias. Therefore, normal sinus rhythm should be ensured on electrocardiogram. When reviewing the echocardiogram, the two most important pieces of information are the systolic function of the single ventricle and the atrioventricular valve competence. Other things of importance include the presence of any residual ventricular outflow tract obstruction and any previously noted or currently abnormal pulmonary artery anatomy.

What NPO Considerations Exist?

The American Society of Anesthesiology NPO guidelines of eight hours for solids, six hours for formula, four hours for breastmilk, and two hours for clear liquids should still be followed. However, as these patients are reliant on preload to drive pulmonary blood flow, emphasis should be placed on ensuring

https://doi.org/10.1017/9781108668736.072 Published online by Cambridge University Press

that clear liquid intake is continued until the two-hour cutoff. If fasting is prolonged, strong consideration should be given to preoperative institution of intravenous fluids in order to minimize dehydration and its adverse effects.

How Should Upper Respiratory Tract Infection (URI) Symptoms Be Evaluated in These Patients?

The increased PVR and airway hyperreactivity that accompanies URIs can result in life-threatening cyanosis and hypoxemia in these patients that rely on passive flow of blood through the pulmonary system. Therefore, extra caution should be utilized in the presence of any signs of URI and strong consideration should be given to rescheduling the case, if at all possible, after resolution of symptoms and return to baseline status has occurred.

What Is the Minimum Workup That Should Be Ensured Prior to Emergency Surgery?

If a child presents emergently to a hospital other than the one where he or she normally receives cardiac care, every attempt should be made to obtain the most recent cardiac data from the home institution. If such information cannot be obtained, a cardiology consult should be obtained in addition to an ECG and echocardiogram. In many cases of emergency surgery the child may be dehydrated from decreased oral intake and vomiting prior to presentation. As pulmonary blood flow is dependent on adequate preload, particular attention should be paid to the volume status and induction should be delayed until adequate hydration is achieved.

What Are Current AHA Recommendations for Subacute Bacterial Endocarditis (SBE) Prophylaxis?

The AHA recommends SBE prophylaxis for cardiac conditions with the highest risk of adverse outcomes from endocarditis. These conditions include:

- Prosthetic cardiac valves or valve material
- Previous history of endocarditis
- Unrepaired cyanotic congenital heart disease (CHD)
- Completely repaired CHD with prosthetic materials during the first six months following repair
- Repaired CHD with residual shunts or valvular regurgitation adjacent to prosthetic material
- Cardiac transplant patients with valvulopathy

As a cyanotic lesion, SCPA meets the criteria for SBE prophylaxis prior to dental procedures that manipulate the oral mucosa as well as invasive respiratory tract procedures.

What Methods Are Appropriate for Induction of Anesthesia?

The child with SCPA physiology may be appropriate for either intravenous or inhalational induction. The decision should be based on the preoperative evaluation as well as the needs of the case being performed while keeping in mind caveats specific to SCPA patients. If the preoperative workup reveals any compromise in cardiac function, an intravenous induction should be performed. Additionally, if there is a possibility of dehydration, intravenous access should be obtained for fluid resuscitation prior to anesthetic induction. In a patient with no cardiac symptoms, inhalational induction is generally well tolerated. It should be noted that induction will be slower than in non-SCPA patients due to the interventricular mixing of pulmonary and IVC blood. In addition, any coughing or breath-holding will decrease pulmonary blood flow and lead to increased desaturation. Consideration should also be given to the fact that as these children have already had many interventions, intravenous access may be difficult and premedication may be necessary.

What Respiratory and Ventilatory Considerations Exist for These Patients?

SCPA patients function optimally during spontaneous ventilation, when negative intrathoracic pressures help to draw blood into the pulmonary circulation. When positive pressure ventilation is necessary, conservation of passive pulmonary blood flow requires maintenance of normal to low PVR. A ventilation strategy that uses moderate tidal volumes and short inspiratory times will result in

https://doi.org/10.1017/9781108668736.072 Published online by Cambridge University Press

low mean airway pressure and PVR. Positive end expiratory pressure (PEEP) should be utilized to maintain functional residual capacity, but high PEEP should be avoided as this will decrease pulmonary blood flow.

Maintain slight hypercarbia, targeting an end-tidal carbon dioxide level between 40 and 50 mmHg. This will increase cerebral blood flow, thereby increasing flow through the pulmonary vascular system. However, excessive hypercarbia should be avoided as the associated increase in pulmonary vascular resistance will begin to outweigh the benefits of increased cerebral blood flow. Inspired oxygen should be used as needed to maintain baseline preoperative oxygen saturation.

Can a Laryngeal Mask Airway (LMA) Be Used in SCPA Patients?

While the use of an LMA offers the advantage of allowing spontaneous ventilation, care must be taken to avoid hypoventilation which could lead to atelectasis and intrapulmonary shunting. Additionally, some anesthesiologists prefer not to use LMAs in younger patients. With vigilant observation an LMA may be employed for shorter surgeries, such as urologic procedures, where the anesthesiologist has full access to the airway and conversion to general endotracheal tube anesthesia is possible if necessary.

What Considerations Exist When Placing Invasive Lines in Patients with SCPA Physiology?

Due to the multitude of previous surgeries and catheterizations, both arterial and central venous access may prove difficult to obtain in the child with SCPA physiology. At the site of previous Blalock–Taussig (BT) shunt, the possibility of residual subclavian arterial narrowing can occasionally negatively impact arterial pressure readings in that extremity. For central venous access, institutional preferences vary, and many would advocate avoiding both internal jugular and subclavian catheters, as the potential for SVC thrombosis could prove catastrophic to pulmonary blood flow. A femoral line may be preferable if central venous access is necessary. Caution must be taken against the introduction of air bubbles through any lower extremity catheter as the IVC flows directly to the systemic circulation, introducing the risk of cerebral and coronary air embolisms.

Are SCPA Patients Candidates for Laparoscopic Surgery?

There is a risk that the reduction of functional residual capacity associated with pneumoperitoneum could compromise pulmonary blood flow. However, laparoscopic surgery has been shown to be safe in the single ventricle population. With the use of low insufflation pressures, constant vigilance, and a low threshold for conversion to open surgery, laparoscopic procedures may be attempted in the SCPA patient.

What Concerns Exist during the Maintenance of Anesthesia?

In a well-compensated SCPA patient, both inhaled and intravenous maintenance anesthetics may be well tolerated. If any cardiac compromise exists, a high-dose narcotic and muscle relaxant-based technique may provide a more stable hemodynamic profile. Care should be taken to maintain adequate intravascular volume. When appropriate, intraoperative hematocrit levels should be followed and kept above 40–45%; with ongoing blood loss these patients will require a more aggressive transfusion strategy than healthy children. It is particularly important to formulate a plan that will minimize nausea/vomiting, pain, and agitation in the postoperative period, as these all may lead to an increase in PVR and decreased pulmonary blood flow. Antiemetics should be administered prior to emergence. Consideration should be given to regional anesthetic techniques when possible, keeping in mind that the child may have previously been on anticoagulants. Postoperative agitation can be mitigated with dexmedetomidine, narcotics, and if necessary, benzodiazepines.

When Is It Safe to Discharge a Patient with SCPA Physiology?

Children with SCPA physiology poorly tolerate common postoperative issues such as vomiting, decreased oral intake, hypoventilation, and pain. It is therefore of utmost importance that these factors are well controlled prior to considering discharge from the day surgery/hospital environment. Additional aspects, such as the distance the family must travel

https://doi.org/10.1017/9781108668736.072 Published online by Cambridge University Press

(should the need to return to the hospital occur), and parental comfort with discharge should be considered. If any question exists as to the safety of discharge, the child should be admitted to the hospital overnight for observation and monitoring.

Suggested Reading

Gottlieb EA, Andropoulos DB. Anesthesia for the patient with congenital heart disease presenting for noncardiac surgery. *Current Opinion in Anaesthesiology*. 2013; 26(3):318–26. PMID: 23614956.

Ramamoorthy C, Haberkern CM, Bhananker SM, et al. Anesthesia-related cardiac arrest in children with heart disease: data from the pediatric perioperative cardiac arrest (POCA) registry. *Anesthesia & Analgesia*. 2010;110(5):1376–82. PMID: 20103543.

Veyckemans F, Momeni M. The patient with a history of congenital heart disease who is to undergo ambulatory surgery. *Current Opinion in Anaesthesiology*. 2013; 26(6):685–691. PMID: 24126690.

Wilson W, Taubert K, et al. Prevention of infective endocarditis: guidelines from the American Heart Association, by the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease. *Circulation*. 2007;116:1736–54. PMID: 17446442.

Yuki K, Casta A, Uezono S. Anesthetic management of noncardiac surgery for patients with single ventricle physiology. *Journal of Anesthesia*. 2011;25(2):247–56. PMID: 21197552.

https://doi.org/10.1017/9781108668736.072 Published online by Cambridge University Press