Chapter

28

Lateral Tunnel Fenestrated Fontan

Wenyu Bai

Case Scenario

A 14-year-old girl, weighing 53 kg, with severe thoracic scoliosis (Cobb angle 54°) is scheduled to undergo a posterior spinal fusion from level T4 to L1. She was born with tricuspid atresia, and initially underwent placement of a modified Blalock–Taussig shunt, followed by a bidirectional Glenn procedure and ultimately completion of her palliation with a lateral tunnel, fenestrated Fontan procedure. During a recent cardiac catheterization her Fontan pressure was 14 mm Hg and two stents were placed in her left pulmonary artery. Current medications include iron supplements 65 mg once a day, enalapril 5 mg once a day, and acetylsalicylic acid 81 mg once a day, with the latter discontinued 5 days prior to surgery. Recent laboratory evaluation revealed a hemoglobin of 12.5 g/dL, a hematocrit of 35%, and a platelet count of 165 K/μL. Coagulation studies (prothrombin time, international normalized ratio, and partial thromboplastin time) were within normal ranges. Two 250-mL aliquots of autologous blood were collected on two occasions prior to her spine surgery.

Transthoracic echocardiography 4 weeks prior to surgery showed the following:

- *Normal left ventricular function*
- *A patent Fontan fenestration with minimal right-to-left shunting*
- *An unobstructed Fontan pathway*

Key Objectives

- Understand Fontan physiology and the major determinants of Fontan circulation.
- Describe the anesthetic management of patients with Fontan physiology for noncardiac surgery.
- Discuss blood-saving strategies during posterior spinal fusion in patients with Fontan physiology.
- Describe perioperative complications in patients with Fontan physiology for spinal fusion.

Pathophysiology

What is tricuspid atresia?

Tricuspid atresia is a cyanotic congenital heart defect in which the tricuspid valve (TV) is underdeveloped, resulting in complete obstruction of blood flow from the right atrium (RA) into either ventricle, (See Figure 28.1.)

Characteristics of TA include the following:

- A lack of communication between the RA and the morphologic right ventricle (RV)
- An interatrial communication
- An enlarged left-sided atrioventricular (AV) valve
- Absence of the RV inlet with varying deficiency of the trabecular RV

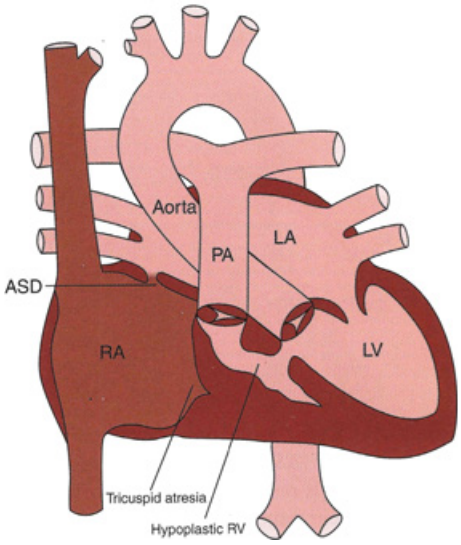

Figure 28.1 Tricuspid atresia. Diagram illustrating tricuspid atresia, atrial septal defect, and hypoplastic right ventricle. ASD, atrial septal defect; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle. In V. G. Nasr and J. A. DiNardo, *The Pediatric Cardiac Anesthesia Handbook*, 1st ed. John Wiley & Sons, 2017; 195–8. With permission.

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

The RV is often underdeveloped, ranging from near total absence to varying degrees of hypoplasia. Other abnormalities including pulmonary stenosis (PS) may be present as well. A bulboventricular foramen or ventricular septal defect (VSD) is present in nearly all cases. Depending on the size of the VSD and the degree of PS (if present), antegrade pulmonary blood flow may occur via the VSD from the left ventricle (LV) to the RV to the pulmonary artery.

After birth an unrestrictive interatrial communication, in the form of either a patent foramen ovale (PFO) or an atrial septal defect (ASD), is crucial to provide mixing between systemic and pulmonary venous blood, as systemic venous return cannot proceed from the RA to the RV. If not present, an urgent atrial septostomy must be performed to open the atrial septum and allow blood flow from the RA to LA; this is generally performed in the cardiac catheterization laboratory.

Tricuspid atresia has three major variants based on the associated relationship of the great vessels. Type I is characterized by normally related great arteries; pulmonary obstruction is most common in type I TA. Type II is characterized by dextro (d)-transposition of the great vessels and these patients generally have unobstructed pulmonary blood flow. Type III TA is characterized by levo (l)-transposition of the great arteries.

Clinical Pearl

Tricuspid atresia is a cyanotic congenital heart defect in which the tricuspid valve is underdeveloped, resulting in complete obstruction from the right atrium into either ventricle.

What are the initial sources of pulmonary blood flow in patients with TA?

The pathophysiology of TA is determined by the amount of pulmonary blood flow (PBF). Initial sources of PBF include flow via the patent ductus arteriosus (PDA) and antegrade flow provided via the VSD to the RV and pulmonary arteries. Depending on the size of the VSD and the degree of PS (if any) initial PBF may be decreased, balanced, or increased. Patients with decreased PBF will require placement of a systemic-to-pulmonary artery shunt as their initial surgical procedure. (See Figure 28.2.)

Clinical Pearl

The pathophysiology of TA is determined by the amount of PBF. Initial sources of PBF include flow via the PDA and antegrade flow provided via the VSD to the RV and pulmonary arteries.

What was the path of blood flow in this patient with TA at birth?

Systemic venous return from the vena cavae entered the RA and traveled via a PFO or ASD to the LA, where it mixed with pulmonary venous blood. From there it flowed via the mitral valve into the left ventricle (LV). It then traveled either via a small VSD to the RV or was ejected via the aorta. The majority of PBF was initially provided via the PDA. Ductal patency was maintained with a prostaglandin (PGE_1) infusion in the neonatal period until a modified Blalock–Taussig (mBT) shunt was placed surgically to provide PBF.

What cardiac surgical procedures do patients with TA undergo?

In tricuspid atresia the RV is often hypoplastic, leaving the LV as the systemic ventricle. Since the left ventricular outflow tract is normal in TA, a traditional Norwood procedure including aortic reconstruction is not necessary as it would be for a patient with hypoplastic left heart syndrome. A stable source of PBF is required, and thus the

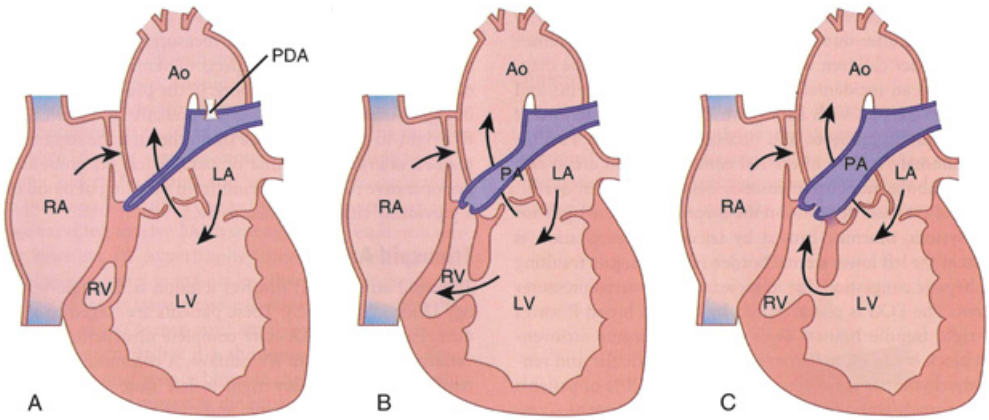

Figure 28.2 Anatomy and blood flow in tricuspid atresia Type I. Type 1A: Pulmonary atresia with pulmonary blood flow via patent ductus arteriosus or major aortopulmonary collateral arteries (MAPCAs). Type 1B: Small ventricular septal defect and pulmonary stenosis. Type 1C: Large ventricular septal defect and no pulmonary stenosis. From DiNardo J. A., Shukla A. C., and McGowan, F. X., Jr. Anesthesia for congenital heart surgery. In Davis P. J. and Cladis F. P., eds. *Smith's Anesthesia for Infants and Children*, 9th ed. Elsevier; 2017: 635–98. With permission.

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

first surgery for these patients is generally a systemic-to-pulmonary artery or mBT shunt.

Initial palliation with an mBT shunt allows the infant to grow while pulmonary vascular resistance (PVR) falls in the months following birth. Ultimately the pulmonary and systemic circulations are separated using two-staged operations for single-ventricle palliation. The first, performed at 3–6 months of age, is a hemi-Fontan or a bidirectional Glenn (BDG) procedure, which connects the superior vena cava (SVC) to the pulmonary artery, taking the place of the mBT shunt. Venous return from the SVC will now flow directly to the pulmonary circulation while venous return from the inferior vena cava (IVC) continues to mix with oxygenated pulmonary venous return in the atrium. At age 2–4 years this is followed by the Fontan procedure, which completes the separation of the pulmonary and systemic circulations by baffling IVC blood directly into the pulmonary vascular bed. (See Chapters 27, 29, and 30 for details of single-ventricle staged palliative procedures.)

What is the Fontan operation?

The Fontan operation completes the separation of the pulmonary and systemic circulations in a patient with single-ventricle physiology. Usually performed at 2–4 years of age, the Fontan procedure connects IVC blood directly to the pulmonary arteries or to the previously created superior cavopulmonary anastomosis (SCPA), allowing all systemic venous blood to flow directly to the lungs. The single ventricle (for patients with TA, the LV) then pumps oxygenated blood systemically. In addition to providing better systemic oxygenation (usual $SpO_2 > 90\%$), the Fontan procedure also reduces volume loading of the systemic ventricle and reduces the risk of paradoxical embolism.

There have been several modifications to the original Fontan procedure and currently the connection between the vena cavae is established either via placement of an extracardiac conduit (***extracardiac Fontan***) or an intraatrial baffle (***lateral tunnel Fontan***; see Figure 28.3). A communication or fenestration may be created between the Fontan conduit or baffle and the common atrium. This serves as a "pop-off" during times of high PVR, allowing right-to-left shunting to occur, augmenting ventricular filling and maintaining cardiac output, albeit at the cost of desaturation.

Clinical Pearl

A communication or fenestration may be made between the Fontan conduit or baffle and the common atrium. This serves as a "pop-off" during times of high PVR, allowing right-to-left shunting to occur, augmenting ventricular filling and maintaining cardiac output, albeit at the cost of desaturation.

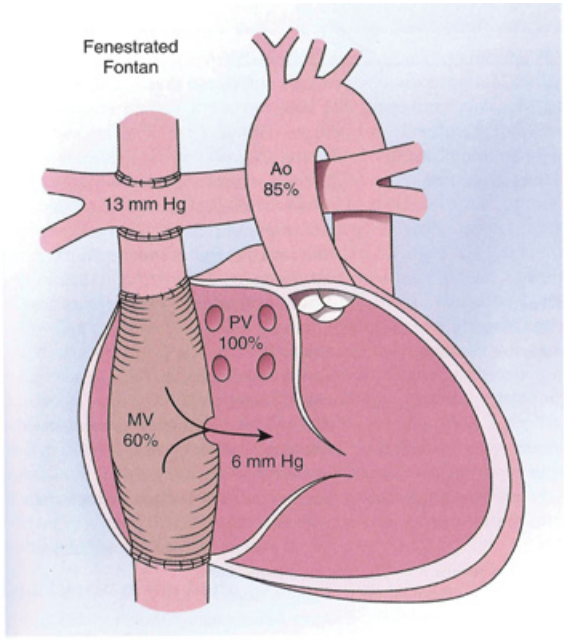

Figure 28.3 Lateral tunnel Fontan with patent fenestration. Ao, Aortic saturation; MV, mixed venous saturation; PV, pulmonary vein saturation. From B. D. Kussman, A. J. Powell, and F. X. McGowan Jr. Congenital cardiac anesthesia nonbypass procedures. In Davis P. J. and Cladis F. P., eds. *Smith's Anesthesia for Infants and Children*, 9th ed. Elsevier; 2017: 699–743. With permission.

What are the major determinants of the Fontan circulation?

The ***transpulmonary gradient*** (TPG) may be defined as the pressure difference between the systemic venous system and the common atrial pressure. After Fontan completion, the TPG across the Fontan pathway is the driving force to maintain blood flow through the pulmonary vasculature to maintain adequate oxygenation and cardiac output.

Important determinants of the function of the Fontan circulation include the following:

- Systemic ventricular function
- Atrioventricular valve competency
- Cardiac rhythm
- Systemic venous pressure and volume status
- Pulmonary vascular pressure and resistance

Alterations in any of these factors, alone or in combination, may compromise systemic cardiac output. One of the key features of a Fontan circulation is the higher central venous pressure (CVP), usually 10–14 mm Hg, required to maintain the pulmonary circulation and cardiac output.

Survival has significantly improved in patients with a Fontan circulation over the past two decades; a recently published study demonstrated a 26-year survival rate of

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

89% in patients with a lateral tunnel Fontan [1]. This success has created a growing population of children and adults with single-ventricle physiology who may require additional surgical procedures to address other comorbid conditions, including spine surgeries.

Clinical Pearl

The transpulmonary gradient is defined as the pressure difference between the central venous system (PA pressure) and the common atrial pressure. Patients with Fontan circulation have higher CVPs (10–14 mm Hg) which are required to maintain pulmonary circulation and cardiac output.

What is scoliosis and what are the different types and incidence of scoliosis in children?

Scoliosis is defined as a lateral curvature of the spine >10° on an anterior–posterior standing radiograph. The location of the scoliotic curve varies and can be thoracic, lumbar, or thoracolumbar. Scoliosis is classified as idiopathic, congenital, or neuromuscular, based on the etiologies.

Idiopathic scoliosis is the most common subtype. However, multiple studies have reported the incidence of scoliosis in patients with CHD and a history of cardiothoracic surgery to be higher, and a prevalence of 9.8% has been reported in patients with Fontan circulation [2, 3]. Congenital scoliosis arises as a result of congenital malformations of the spine. Neuromuscular scoliosis occurs due to neurologic or muscular disorders. These patients, who often have impaired respiratory and cardiac function, are more clinically complex than patients with idiopathic scoliosis due to their neurologic disease.

How is the severity of scoliosis measured and what are the surgical treatment options?

The Cobb angle, a measurement of the degree of side-to-side spinal curvature, is used to define the severity of the scoliosis and determine the appropriate treatment. Spinal fusion is recommended for patients with a Cobb angle above 45–50 degrees at mature skeletal growth to decrease the curve progression and ongoing impairment of pulmonary function.

Early onset of congenital scoliosis, and some types of neuromuscular scoliosis, can cause significant adverse effects on lung development, predisposing these patients to impaired gas exchange and pulmonary hypertension. In these patients the use of growing rods or vertical expandable prosthetic titanium ribs provides the option to correct spine and chest wall abnormalities while allowing the spine to continue to grow.

Anesthetic Implications

How should this patient be assessed preoperatively?

Preoperatively, the anesthesiologist should focus on past medical history, changes in recent health status, exercise capacity, pulmonary function, and current medications. Any history of changes in recent health or exercise capacity in Fontan patients warrant additional investigation.

A recent visit with pediatric cardiology is essential, and electrocardiogram, echocardiogram, cardiac catheterization, and/or cardiac magnetic resonance imaging findings should be reviewed. Important considerations include the evaluation and review of cardiac anatomy, single-ventricle function, Fontan pathway, transpulmonary gradient, the status of the fenestration (open vs. closed), and baseline oxygen saturation. For patients with severe scoliosis, pulmonary function testing is often helpful to guide intraoperative and postoperative ventilatory management. A preoperative consultation with a pediatric electrophysiologist is recommended for patients who have a history of arrhythmias or a pacemaker.

The physical examination should encompass the airway, heart and lungs, overall perfusion, and evaluation for potential difficulties with vascular access. The physical examination of a well-functioning Fontan patient should be notable for a baseline $SpO_2 > 90$. It is also important to document preoperative neurologic function due to the risk of perioperative neurologic complications.

Laboratory tests should include a complete blood count (including platelet count), basic metabolic panel, liver panel, and coagulation profile. Normal results are anticipated in well compensated Fontan patients who are not on anticoagulant medications.

Clinical Pearl

Important considerations include the evaluation and review of cardiac anatomy, single-ventricle function, Fontan pathway, transpulmonary gradient, fenestration status, and baseline oxygen saturation. Normal laboratory results are anticipated in well compensated Fontan patients who are not on anticoagulant medications.

Is multidisciplinary preoperative collaboration important for this patient?

Due to the complex perioperative management considerations involved in patients with Fontan physiology who are

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

undergoing a major surgery such as a PSF, close collaboration among pediatric orthopedists, cardiologists, neurophysiologists, anesthesiologists, and postoperative intensivists is essential. Pediatric cardiologists can clarify specific issues regarding the anatomy and physiology of the Fontan patient, as well as consult and assist postoperatively in medical management. Pediatric orthopedic surgeons should be involved in discussions about potential intraoperative blood loss, which can be higher than normally anticipated due to the higher CVPs and coagulation issues that exist in Fontan patients. The need to optimize blood pressure to minimize surgical blood loss must be balanced with maintaining adequate cardiac output, and this should also be discussed with the surgeons. The surgeons may opt to minimize operative time and choose a moderate curve correction.

Neurophysiology specialists should be involved in preoperative discussions regarding the effects of anesthetic medications on neuromonitoring, as well as plans to treat potential decreases or loss of neuromonitoring signals, including the necessity for an intraoperative wake-up test. It is important to rehearse the potential intraoperative wake-up test with the patient during the preoperative visit to psychologically prepare the patient and family.

What medications are generally taken by patients with Fontan physiology and how should they be managed perioperatively?

Acetylsalicylic acid (ASA), warfarin and angiotensin converting enzyme inhibitors (ACEi) are the most commonly administered medications in patients with well-compensated Fontan physiology.

Most patients receive ASA or warfarin for chronic thrombotic prophylaxis. Cardiologists recommend stopping ASA or warfarin 5–7 days before the surgery to facilitate a normally functioning coagulation system. For individuals who are advised to continue ASA preoperatively, the surgeon and anesthesiologist should be made aware of this due to the potential effects on platelet function. Appropriate blood products should be prepared preoperatively.

Angiotensin converting enzyme inhibitors are used to reduce afterload and minimize strain on the single ventricle. However, with general anesthesia they may result in severe hypotension refractory to treatment with adrenergic agonists. It is recommended that the patient not take ACEi on the morning of surgery. Refractory intraoperative hypotension may be treated with intravenous vasopressin.

What premedication options exist for this patient?

A benzodiazepine may be administered either orally or intravenously for anxiolysis and should be well tolerated in a patient with Fontan physiology, but should probably be utilized judiciously and as a single agent. Heavy sedation or combinations of multiple sedative medications may cause hypoventilation and hypercarbia with subsequent increases in PVR, which is undesirable in patients with a Fontan circulation. In a teenager, the administration of oral midazolam prior to placement of a peripheral intravenous (IV) line may be helpful if the patient is excessively anxious.

Acetaminophen (10–15 mg/kg, maximal dose of 1 g) may be administered orally in the preoperative area. Gabapentin, an anticonvulsant medication used to treat partial seizures and neuropathic and perioperative pain, may also be used as an adjunct for perioperative pain control in a dosage of 10 mg/kg administered orally.

What considerations exist for anesthetic induction?

Although either inhalational or IV techniques can be safely used for anesthetic induction in a well-compensated Fontan patient, ideally IV medications with minimal cardiac depressive effects (etomidate, ketamine, midazolam, and/or opioids) should be used for patients with Fontan physiology. Although propofol may be used, in larger doses it may cause myocardial depression, afterload reduction and venous dilation which may not be well tolerated by these patients. If propofol is used, it is imperative to titrate to effect with careful monitoring during induction. When IV access is not obtained preoperatively, careful titration of sevoflurane is commonly used for anesthetic induction. A fentanyl bolus given IV before intubation can blunt the increase in PVR associated with laryngoscopy and endotracheal tube placement. Either a short-acting neuromuscular blocking agent or rocuronium, an agent that can be reversed by sugammadex prior to initiating neuromonitoring, should be administered to facilitate intubation. The anesthesiologist should carefully plan the induction, choosing the approach most likely to maintain hemodynamic stability in this patient with unique physiology and limited cardiac reserve.

Clinical Pearl

Either intravenous or inhalational agents can be safely used for induction in a well-compensated Fontan patient. Induction agents and techniques should not cause increases in PVR or depression of the systemic ventricle, and preload should be optimized prior to anesthetic induction.

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

How should intraoperative fluids be managed?

Adequate preload is essential to maintain normal cardiac output and compensate for possible peripheral vasodilatation during induction. A patient with Fontan physiology should avoid excessive preoperative fasting and should be encouraged to drink clear liquids up to 2 hours before surgery. Fluids should be administered as soon as IV access is established, if possible before induction of anesthesia. A crystalloid bolus of 5–10 mL/kg before or during induction may be given to maintain hemodynamic stability. Intraoperative fluid management should be guided by patient's hemodynamics, fasting time, blood and third space fluid loss, urine output, and laboratory measurements. It is crucial to have adequate IV access with at least two large-bore peripheral IV lines, as rapid fluid or blood administration is often necessary during this surgery. It is important to remember that the maintenance of adequate CVP is critical for providing PBF, and that hypovolemia is poorly tolerated. Meticulous attention to removal of bubbles from IV lines is absolutely necessary as Fontan patients may have an open atrial fenestration or other arterial-venous connections that put them at risk for systemic embolism of air.

Clinical Pearl

Meticulous removal of bubbles from IV lines when administering medications is absolutely necessary as Fontan patients may have an open atrial fenestration or other arterial-venous connections that put them at risk for systemic embolism of any air.

What are the effects of spontaneous versus mechanical ventilation in a patient with a Fontan circulation?

During most uncomplicated surgeries spontaneous ventilation is preferentially used whenever possible in Fontan patients. Negative intrathoracic pressure during inspiration enhances venous return and hemodynamic performance. Mechanical positive pressure ventilation (PPV) can lead to less favorable hemodynamics due to increased intrathoracic pressure leading to decreased central venous return. However, during a spinal fusion, when the patient is prone for several hours, spontaneous ventilation is not possible. Hypoventilation with associated hypercarbia, hypoxia, and atelectasis, all of which can lead to increased PVR, should be avoided in patients with Fontan physiology. Mechanical ventilation with maintenance of low mean airway pressures and normocarbia should be the goal. Tidal volumes of 6–8 mL/kg along with physiologic use of positive end-expiratory pressure (PEEP) is generally appropriate.

Clinical Pearl

Mechanical ventilation with maintenance of low mean airway pressures and normocarbia should be the goal. Tidal volumes of 6–8 mL/kg along with physiologic use of PEEP is generally appropriate.

What routine monitoring is utilized for patients undergoing PSF?

Constant vigilance to hemodynamics, respiratory mechanics, and the surgical field to rapidly identify and treat derangements during spinal surgery is essential, particularly in a single-ventricle patient. Potential difficulties include significant blood loss with hemodynamic derangements, fluid shifts, and neurologic injury. In addition to standard American Society of Anesthesiologists recommended monitors, an arterial line, urinary catheter, and neuromonitoring should be used. Neuromonitoring may include somatosensory-evoked potential (SSEP), transcranial motor-evoked potentials (TcMEPs), and electromyography (EMG). Somatosensory-evoked potentials monitor the somatosensory pathway and TcMEPs monitor the corticospinal tract or motor activity. A decrease in amplitude of 50% and increase in latency of >10% in SSEPs are considered significant changes that may require anesthetic or surgical intervention. Many anesthetic medications, including both IV and inhalational agents, prolong latency or reduce amplitude at varying doses. Electromyography is used to evaluate spinal nerve root injury and is frequently used during pedicle screw placement. Both EMG and TcMEPs are inaccurate with the use of neuromuscular blocking agents, so these should be avoided except during induction. Preferences for anesthetic management should be discussed with the neuromonitoring team in advance, particularly as institutional preferences may vary slightly. In general, a balanced anesthetic or total intravenous anesthetic technique is preferred.

Is there a need for additional monitoring?

Intraoperative CVP monitoring, transesophageal echocardiography (TEE) and near-infrared spectroscopy may be utilized as well for optimal hemodynamic monitoring, depending on the status of the patient.

While a CVP line may not always be utilized for a PSF in a healthy two-ventricle patient with idiopathic scoliosis, it is advisable to place a CVP line in patients with Fontan physiology undergoing PSF. The ability to follow CVP

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

trends is not only useful for guiding volume and blood replacement therapy but can also be helpful for administration of vasoactive drugs if necessary. Central venous pressure increases when a patient is prone due to IVC compression, decreased cardiac compliance, and an increase in intrathoracic pressure, with one study showing an average increase in CVP when prone of 9 mm Hg [4]. The CVP in a patient with Fontan physiology may need to be in the high teens or low 20s to provide adequate cardiac output in the prone position. It is critical to monitor CVP trends, especially once the patient has been turned prone.

Although CVP monitoring can provide valuable information, it is not without risks. It should be noted that the CVP in a Fontan patient actually reflects mean pulmonary artery pressure, since the SVC and IVC are directly connected to the pulmonary arteries. Placement of a central venous catheter can result in pulmonary artery injury, paradoxical air emboli, Fontan pathway thrombus, and central line associated blood stream infection. The length of central venous line in the internal jugular vein must be carefully assessed. To reduce complication risks the CVP catheter should be removed postoperatively as soon as hemodynamics are stabilized. Alternatively, TEE is an excellent tool for intraoperative monitoring of ventricular function, ventricular preload, the Fontan pathway, any cardiac shunting (baffle leak or fenestration), and venous air embolism, but its use often depends on the availability of a pediatric cardiologist to perform and interpret the TEE. Near-infrared spectroscopy may be a useful tool to monitor cardiac output, cerebral perfusion, and tissue oxygenation in Fontan patients, especially when CVP is elevated due to prone positioning during posterior spinal fusion [5].

It is important to know whether the patient had a previous mBT shunt, and if so, where it was located (right or left side; right-sided mBT shunt being most common). A noninvasive blood pressure cuff or an arterial catheter placed on or in the ipsilateral side where the previous mBT shunt existed may measure falsely low systemic blood pressure due to vessel stenosis near the shunt take-down. If there is no alternative but to use the arm on the ipsilateral side of the previous mBT shunt, the blood pressure in that arm should first be verified to be the same as in other extremities.

Clinical Pearl

Central venous pressure in a patient with Fontan circulation may need to be in the high teens or low 20s to provide adequate cardiac output in the prone position. It is critical to monitor the CVP trends, especially when the patient is turned prone. It should be noted that the CVP in a Fontan patient reflects mean pulmonary artery pressure as the SVC and IVC are directly connected to the pulmonary arteries.

What are the general anesthetic principles for managing a patient with Fontan physiology for noncardiac surgery?

Guiding principles include maximizing preload, minimizing myocardial depression caused by medications and/or inhalational agents, maintaining sinus rhythm, and avoiding increases in PVR which can be caused by hypoxia, hypercarbia, acidosis, high peak inspiratory pressures with mechanical ventilation, and poorly controlled pain or surgical stimulation.

How should temperature be managed intraoperatively for this patient?

It is challenging to keep a PSF patient normothermic during surgery, but intraoperative hypothermia can lead to increases in PVR and decreases in cardiac output which are particularly detrimental to a Fontan patient. Lower temperatures shift the oxyhemoglobin dissociation curve to the left, reducing oxygen availability to the tissues. Therefore, active warming should begin during induction of anesthesia. There is potential for the patient to lose significant heat, particularly during prone positioning and in the time before surgical incision. An underbody convection warmer may be used under the patient in the preoperative area and turned on during induction. Core temperature should be monitored during the surgery using a nasal, esophageal, or bladder temperature probe.

What issues are associated with surgery in the prone position in a Fontan patient?

Prone positioning during anesthesia can elicit a variety of respiratory and cardiovascular responses. There may be a decrease in cardiac index and ventricular compliance, varying degrees of increased pressure in the IVC, and increased peak airway pressures (dependent on the type of operating frame used) when patients are in the prone position [6]. While these changes may be tolerated in otherwise healthy patients they often present challenges in patients with Fontan physiology.

Extensive padding is necessary to prevent a wide spectrum of compression and stretch injuries as well as to keep the abdomen free from impeding ventilation. In Fontan patients, it is of particular importance to ensure free diaphragmatic movement as increases in intraabdominal pressure may impair ventilation and cause IVC compression. In turn this can result in a decrease in systemic venous return to the Fontan circuit and a reduction in preload to the systemic ventricle. Impaired ventilation can lead to increases in PVR and further elevations in systemic venous

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

pressure, and then, as a consequence, increases in epidural venous pressure and intraoperative bleeding. Careful positioning is essential to minimize these risks. A Jackson table with abdominal free 4-point pads has the least effect on cardiac performance and if available should be considered for patients with Fontan physiology.

Clinical Pearl

Care should be taken during positioning to ensure that lung excursion is unimpeded and padding of pressure points is optimal. Increases in intraabdominal pressure may impair ventilation, cause IVC compression, and decrease preload and cardiac output. Increased intraabdominal pressure may also further elevate systemic venous pressure, which may in turn increase epidural venous pressure and intraoperative bleeding.

What is the differential diagnosis and treatment for intraoperative hypoxemia in this patient?

The differential diagnosis for acute oxygen desaturation in a Fontan patient includes the common airway-related causes such as accidental extubation during prone positioning, endobronchial intubation, bronchospasm, endotracheal obstruction, and atelectasis. Additionally, in a Fontan patient, an acute elevation of PVR causing right-to-left shunting across a fenestration or baffle leak should be considered. Intraoperative TEE is a valuable tool to assist in this clinical diagnosis and guide treatment. Techniques to decrease PVR and improve oxygenation include optimizing mechanical ventilation (increasing FiO_2, correcting hypercarbia, and avoiding excessive positive pressure), correcting hypothermia and acidosis, and administering phosphodiesterase inhibitors such as milrinone and/or utilizing inhaled nitric oxide.

What techniques can be used to minimize intraoperative blood loss and use of allogeneic transfusion in this patient?

Intraoperative bleeding during PSF is primarily venous, with the average blood loss in the range of 800–1200 mL for children and adolescents with idiopathic scoliosis. Children with Fontan physiology may have significantly higher intraoperative blood loss, which may be in excess of an entire blood volume [7, 8].

Techniques to minimize intraoperative blood loss and allogeneic transfusion in patients having a PSF include preoperative or intraoperative autologous blood donation, appropriate prone positioning, an appropriate operating table, intraoperative blood salvage, controlled hypotension, and use of intravenous antifibrinolytic agents (ε-aminocaproic acid or tranexamic acid). Due to the higher baseline CVP required to maintain the Fontan circulation and diminished cardiac functional reserve, controlled hypotension is not recommended in single-ventricle patients. Goals include keeping hematocrit ≥30% and mean arterial pressure ≥60–65 mm Hg.

Clinical Pearl

Keep hematocrit ≥30% and mean arterial pressure ≥60–65 mm Hg. Controlled hypotension is not recommended in single-ventricle patients.

The surgeon reports profuse oozing in the surgical field. The patient's current blood pressure is 79/40 mm Hg (mean 48), with a CVP trending from 20 to 16 mm Hg. What are the differential diagnoses and potential therapies?

This clinical presentation strongly suggests intravascular volume depletion which is a common scenario in a Fontan patient having a PSF. Sequential arterial blood gases (ABGs) with hematocrit should be obtained to provide information to help confirm the diagnosis and guide appropriate therapy and fluid management. Posterior spinal fusion in Fontan patients is associated with major intraoperative blood loss with means of 64–84 mL/kg or higher [7]. Appropriate volume replacement given expeditiously to maintain adequate preload and cardiac output is absolutely fundamental. Balanced salt solutions, followed by 5% albumin, are suitable to replace blood loss initially. The transfusion threshold is lower in patients with Fontan physiology and it is recommended to keep the hematocrit ≥30 [5, 7]. In addition to transfusing autologous blood and blood from intraoperative cell savage, allogeneic whole blood or packed red blood cells are commonly used to replace intraoperative volume depletion caused by surgical bleeding. Other blood products such as fresh frozen plasma, platelets, and cryoprecipitate are also frequently needed, guided by intraoperative coagulation testing, including rotational thromboelastometry or thromboelastography. Massive intraoperative bleeding may require use of a rapid transfusion system and/or activation of the massive transfusion protocol.

Diminished ventricular function may also be a cause of hypotension, especially if anesthetics with cardiac depressant effects are used. Ventricular function and cardiac

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

filling can be evaluated by CVP trends and TEE. The dynamic visualization of cardiac function and venous return by TEE is more helpful than CVP monitoring [9]. If cardiac function is compromised, the use of inotropic medications should be considered. Drugs such as the phosphodiesterase inhibitor milrinone can be used to improve both systolic and diastolic function without increasing PVR. Catecholamine inotropic agents, which improve systolic function, may worsen diastolic function by impairing ventricular relaxation. However, epinephrine in low doses (0.02–0.04 mcg/kg/minute) can be considered for refractory hypotension.

Cardiac arrhythmias are not uncommon in patients with a Fontan circulation. The onset of atrial tachycardia may produce hemodynamic compromise and necessitate antiarrhythmic therapy immediately, including direct current cardioversion. Should this occur the electrophysiology team should be contacted immediately. Defibrillator pads should be placed on the patient before incision.

Clinical Pearl

Massive intraoperative bleeding may require the use of a rapid transfusion system and activation of the massive transfusion protocol. If cardiac function is compromised, the phosphodiesterase inhibitor milrinone should be considered. Epinephrine in low doses (0.02–0.04 mcg/kg/minute) should be considered for refractory hypotension, assuming hypotension is not secondary to hypovolemia.

How should a sudden change in SSEPs be managed in this patient?

It is important to have a plan should sudden changes in neuromonitoring occur to prevent new-onset neurological deficits. When neuromonitoring signals are changed or lost, anesthetic techniques which could compromise oxygen delivery should be adjusted or corrected immediately. This may include transfusing blood to increase the hematocrit and raising the mean arterial blood pressure to optimize tissue perfusion and increase oxygen delivery to the spinal cord. An ABG should be measured in order to correct metabolic acidosis and adjust the partial pressure of carbon dioxide to help improve oxygen supply to the spinal cord. Also, medications administered just prior to the signal changes should be considered as a potential cause for the signal changes.

At a time of potential neurologic compromise it is critical for the entire patient care team to communicate and collaborate to lessen the risk of neurologic damage. Neuromonitoring specialists should recheck the signals, electrodes, and connections to determine the pattern and timing of signal changes. The surgeon should review the surgical steps that occurred prior the signal changes and consider adjusting the distraction, removing rods and screws, and using intraoperative imaging to evaluate and prevent the possibility of spinal cord compression or ischemia.

Although neuromonitoring can be helpful to prevent permanent spinal cord damage, an intraoperative wake-up test may be needed to make a formal diagnosis and conclusively rule out permanent neurologic damage. Ideally a wake-up test should have been carefully planned, discussed, and rehearsed with the patient before the surgery. Still, it can be associated with severe complications. A Fontan patient with a pacemaker was reported to have suffered ventricular tachycardia during a wake-up test that necessitated intraoperative CPR for resuscitation [10]. Continuous dexmedetomidine, a highly selective α_2-adrenergic receptor agonist, along with an amnestic such as midazolam, may provide adequate sedation for the patient during the wake-up test.

What perioperative analgesic plans could be suggested for this patient?

Inadequate pain control may cause anxiety, hypertension, and increases in SVR and PVR and should be avoided. However, pain relief with heavy sedative effects should also be avoided due to potential risks of hypoventilation, hypoxia, and hypercarbia.

Multimodal pain control combining opioids and a variety of nonopioid medications is optimal in patients with Fontan physiology. Opioids can be given orally, intravenously and via the epidural space. Medications such as acetaminophen, nonsteroidal antiinflammatory drugs (NSAIDs), gabapentin, ketamine, and methadone may be administered as adjuncts for pain control perioperatively.

Epidural analgesia with local anesthetics and opioid infusion has been used successfully in Fontan patients but the use of ongoing anticoagulant therapies in many Fontan patients may preclude or limit the use of epidural or other regional analgesia techniques. If utilized, local anesthetic dosing, with lidocaine or bupivacaine, should be titrated and monitored carefully to minimize sympatholytic effects and to facilitate postoperative neurologic examinations. Augmenting preload with IV fluids before epidural dosing is prudent. Patient-controlled analgesia is another option.

Intrathecal morphine (6 mcg/kg, maximal 600 mcg for patients receiving gabapentin and 8 mcg/kg, maximal 800 mcg for patients without gabapentin) administered by the anesthesiologist after induction, before incision, may also be utilized. The patient could also receive oral gabapentin preoperatively, IV acetaminophen intraoperatively and in

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press

the first 24 hours postoperatively, followed by oral opioids and NSAIDs during the next 2–3 days.

What is an appropriate plan for extubation in this patient?

Extubation in the operating room at the end of the surgery is optimal for a patient with Fontan physiology, assuming an uneventful intraoperative course. The shift from mechanical to spontaneous ventilation should improve hemodynamics and decrease the risks of ventilator-associated sedation. An awake patient can facilitate accurate postoperative neurologic exams as well. When a Fontan patient has other severe comorbidities such as severe restrictive lung disease or unstable hemodynamics, delayed extubation may be necessary, and appropriate pain management and sedation strategies should be utilized. Hypertension associated with inadequate pain control may add extra strain to the systemic ventricle and should be avoided. Intravenous midazolam and opioids are commonly used medications for postoperative pain control and sedation. Additionally, dexmedetomidine has been shown be an excellent sedative in the intensive care unit.

What are the possible postoperative complications in patients with Fontan physiology undergoing PSF?

Postoperative complications can be severe in these patients and can include continuous bleeding with hypotension requiring vasoactive medications, prolonged intubation, acute renal dysfunction, pleural effusions, Horner syndrome, superior mesenteric artery syndrome, and delayed neurologic damage [7, 8, 10]. Postoperative bleeding may be significant and death has been reported due to hypovolemic shock and dysrhythmias which started during surgery and continued in the immediate postoperative period [8]. This emphasizes the need for continuous close monitoring with early intervention or resuscitation of Fontan patients in the immediate postoperative period either in a pediatric intensive care unit or a cardiac intensive care unit.

As Fontan patients are at risk for thromboembolic events, cardiologists often recommend continuation of anticoagulation therapies, particularly ASA or low-molecular-weight heparin throughout the perioperative period. Pneumatic compression boots for deep vein thrombosis prophylaxis are also important, especially in the immediate postoperative period for patients with limited ambulation.

References

1. C. Schilling, K. Dalsiel, R. Nunn, et al. The Fontan epidemic: population projections from Australia and New Zealand Fontan registry. *Int J Cardiol* 2016; **219**: 14–19.
2. J. A. Herrera Soto, K. L. Vander Hare, P. Barry-Lane, et al. Retrospective study on the development of spinal deformities following sternotomy for congenital heart disease. *Spine* 2007; **32**: 1998–2004.
3. M. Kadhim, C. Pizarro, L. Holmes, et al. Prevalence of scoliosis in patients with Fontan circulation. *Arch Dis Child* 2013; **98**: 170–5.
4. D. E. Soliman, A. D. Maslow, P. M. Bokesch, et al. Transesophageal echocardiography during scoliosis repair: comparison with CVP monitoring. *Can J Anaesth* 1998; **45**: 925–32.
5. C. Walker, D. Martin, J. Klamar, et al. Perioperative management of a patient with Fontan physiology for posterior spinal fusion. *J Med Cases* 2014; **5**: 392–6.
6. Z. E. Brown, M. Görges, E. Cooke, et al. Changes in cardiac index and blood pressure on positioning children prone for scoliosis surgery. *Anaesthesia* 2013; **68**: 742–6.
7. M. B. Rafique, E. A. Stuth, J. C. Tassone. Increased blood loss during posterior spinal fusion for idiopathic scoliosis in an adolescent with Fontan physiology. *Pediatr Anesth* 2006; **16**: 206–12.
8. C. P. C. Macarrón, E. S. Ruiz, J. B. Flores, et al. Spinal surgery in the univentricular heart – is it viable? *Cardiol Young* 2014; **24**: 73–8.
9. D. Vischoff, L. P. Fortier, E. Villeneuve, et al. Anaesthetic management of an adolescent for scoliosis surgery with a Fontan circulation. *Paediatr Anaesth* 2001; **11**: 607–10.
10. C. I. Leichtle, M. Kumpf, M. Gass, et al. Surgical correction of scoliosis in children with congenital heart failure (Fontan circulation): case report and literature review. *Eur Spine J* 2008; **17**: 312–17.

Suggested Reading

Edgcombe H., Carter K., and Yarrow S. Anaesthesia in the prone position. *Br J Anaesth* 2008; **100**: 165–83.

Kadhim M., Pizarro C., Holmes L., et al. Prevalence of scoliosis in patients with Fontan circulation. *Arch Dis Child* 2013; **98**: 170–5.

Macarrón C. P. C., Ruiz E. S., Flores J. B., et al. Spinal surgery in the univentricular heart – is it viable? *Cardiol Young* 2014; **24**: 73–8.

Walker C., Martin D., Klamar J., et al. Perioperative management of a patient with Fontan physiology for posterior spinal fusion. *J Med Cases* 2014; **5**: 392–6.

https://doi.org/10.1017/9781108657341.029 Published online by Cambridge University Press