J Anesth (2011) 25:247–256
DOI 10.1007/s00540-010-1081-4

REVIEW ARTICLE

Anesthetic management of noncardiac surgery for patients with single ventricle physiology

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Received: 21 June 2010 / Accepted: 5 December 2010 / Published online: 1 January 2011

Abstract Patients with congenital heart diseases are a growing population, and noncardiac surgeries will become an important health care issue. Patients with single ventricle physiology are a particularly challenging population who will undergo staged, palliative repair toward a final step of Fontan circulation. Although Fontan surgery creates a serial circulation in which the ventricle pumps blood to the systemic circuit, pulmonary blood flow occurs without a dedicated ventricle. Despite progress in outcomes, this abnormal circulation remains associated with various co-morbidities such as ventricular dysfunction, arrhythmias, protein losing enteropathy, and plastic bronchitis. Health care professionals must prepare for these patients to present to noncardiac surgery at any stage of intervention, possibly with complications. Given that staged, palliative repair has undergone multiple modifications, patients who present for surgery can vary in types and timing of the repair. Anesthesiologists who care for them must be familiar with perioperative issues to optimize outcomes, especially because congenital heart disease is a risk factor for increased mortality for noncardiac surgery.

Keywords Congenital heart disease · Single ventricle physiology · Noncardiac surgery · Anesthesia

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Introduction

Congenital heart defects are the most common birth defects, occurring in approximately 4–9 of 1,000 live births [1]. Major advances in diagnosing and treating congenital heart disease (CHD) during the past 30 years have resulted in expected survival to adulthood of 85% [2]. Now estimates suggest that more adults than children are living with CHD in the US and Canada [2, 3], and a similar phenomenon will be observed in other countries. As a consequence, the number of this population presenting for noncardiac procedures will be likely to increase.

Many lesions will never be physiologically normal following "definite" surgical repair. This is particularly true for lesions with single ventricle physiology, in which only one anatomical or functional ventricular chamber pumps blood to the pulmonary and systemic vascular beds [4] (Table 1). These lesions require staged procedures to be converted into serial circulation where the ventricle pumps blood to the systemic circuit, whereas only pulmonary blood flow (PBF) occurs without a dedicated ventricle. Although long-term outcomes have improved during 2 decades, various co-morbidities remain. CHD adds a significant risk of mortality in children requiring noncardiac surgery [5, 6].

We review management of lesions with single ventricle physiology; their anesthetic implications for noncardiac surgery are discussed.

Management of lesions with single ventricle physiology

Neonatal period

Various anatomic lesions may be classified as single ventricle anatomy or physiology—a single ventricular chamber,

two ventricles (of which one ventricle is small with minimal contribution to cardiac ejection) and two normal-sized ventricles [which cannot be separated because a ventricular septal defect (VSD) is remote either from the great vessel or because of straddling of atrioventricular valve attachments over the VSD that cannot be separated].

Initially, one should determine the amount of pulmonary blood flow (PBF) and dependency of the pulmonary circulation on the ductus arteriosus. Patients with ductal-dependent circulation, initially on PGE_1, require a reliable source of PBF by creation of a systemic to pulmonary shunt. The most common shunt is a modified Blalock-Taussig (BT) shunt, in which GORE-TEX tube graft connects the innominate or subclavian artery to the pulmonary artery (PA). Some will have unrestricted flow to both the PA and the aorta. Such patients usually develop progressive, congestive heart failure (CHF) as pulmonary vascular resistance (PVR) falls in infancy, and require PA banding to limit PBF and protect the pulmonary circulation from high flow and pressure that can eventually cause irreversible pulmonary disease. Patients with systemic outflow obstruction may have a Damus-Kay-Stansel anastomosis/aortic arch reconstruction with BT shunt. Finally, some will have a combination of abnormalities, providing the appropriate amount of PBF and obviating the need for immediate surgical intervention in neonatal period (Table 2).

Infant period

In patients with single ventricle physiology, the ventricle(s) is pressure- and volume-loaded. The ventricle(s) becomes hypertrophied over time and is at risk of subendocardial ischemia. Therefore, the goal of surgical intervention is to reduce the volume load on the ventricle(s).

Fontan and Baudet reported an operation to separate systemic and pulmonary circulations in series in 1971. Initially, the Fontan procedure was done without staged interventions. This strategy was accompanied with a high operative mortality (16–40%) [7, 8].

In the late 1980s, a staged approach to the Fontan procedure was introduced, which shortened the duration of excess in ventricular volume, thus reducing late operative mortality following palliative repair [9]. Bidirectional cavopulmonary shunt surgery (Fig. 1a) is often done typically between 4 and 6 months of age; a hemi-Fontan procedure (Fig. 1b) can be done instead. These procedures allow the ventricles to remodel and function at lower end-diastolic ventricular pressure before the Fontan procedure. Patients who undergo staged procedures show better contractility with smaller ventricular volume after a Fontan procedure [10].

Bidirectional cavopulmonary shunt

A bidirectional cavopulmonary shunt directs systemic venous blood from the superior vena cava (SVC) directly to pulmonary circulation. The SVC is transected, and the cranial end is sewn end-to-side to the right PA and the cardiac end over sewn. The SVC continues with bilateral PAs, thereby producing bidirectional PBF. The main PA is over sewn. Unless the inferior vena cava (IVC) is interrupted with azygous continuation, the azygous vein is ligated so that the SVC does not decompress retrograde to the IVC. It allows the ventricle(s) to reduce the volume load on itself and remodel, maintaining cardiac output at a lower ventricular end-diastolic pressure.

Hemi-Fontan surgery

The right atrium is anastomosed to the PA with patch augmentation. A GORE-TEX baffle is placed within the right atrium, redirecting blood flow from the SVC atriocaval junction across the atriopulmonary anastomosis. Proponents of this surgery over a bidirectional cavopulmonary shunt feel that it allows supplementation of a central PA area to optimize flow to the left lung. However,

Table 1 Lesions with single ventricle physiology

Tricuspid atresia
Pulmonary atresia/intact ventricular septum
Double inlet left ventricle
Unbalanced complete atrioventricular canal
Hypoplastic left heart syndrome
Double outlet right ventricle

Table 2 Initial surgical approach for lesions with single ventricle physiology

Anatomy	Surgical intervention
Two semilunar valves of adequate size, normal aortic arch	Pulmonary artery band
One semilunar valve, normal aortic arch	BT shunt
One semilular valve, hypoplastic aortic arch	Aortic arch reconstruction with BT shunt
Two semilunar valves, aortic stenosis	DKS anastomosis with BT shunt
Two semilunar valves with pulmonary stenosis	No initial intervention required

BT Blalock-Taussig, *DKS* Damus-Kay-Stansel

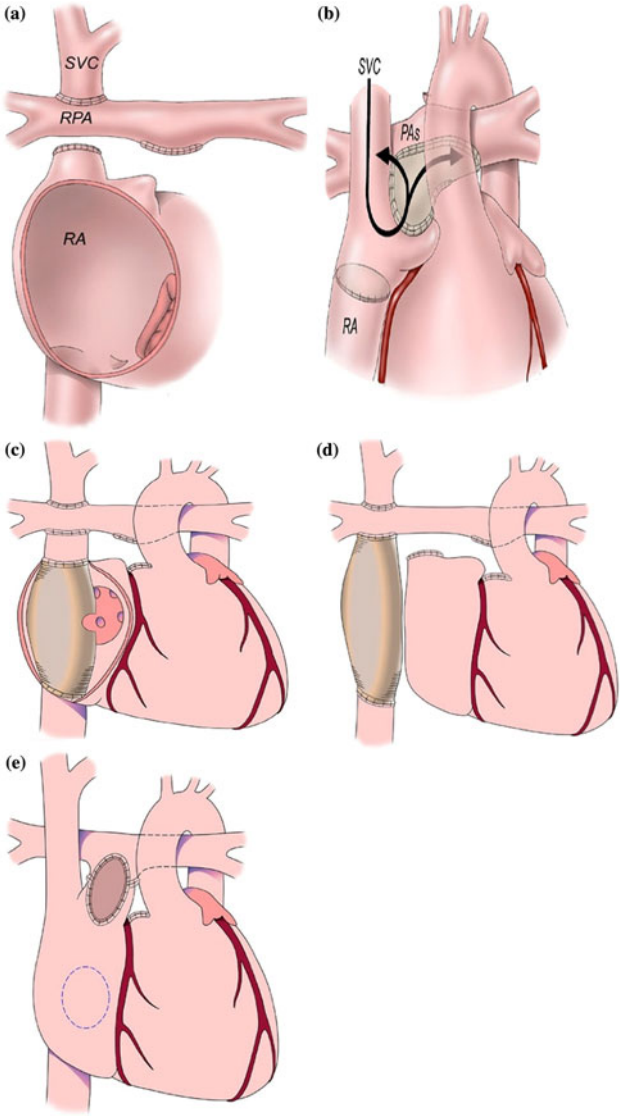

Fig. 1 Heart diagram of palliative repair for single ventricle physiology. **a** Bidirectional cavopulmonary shunt, **b** hemi-Fontan, **c** lateral tunnel Fontan, **d** extracardiac Fontan, **e** atriopulmonary connection Fontan are depicted. *SVC* superior vena cava, *RPA* right pulmonary artery, *PA* pulmonary artery, *RA* right atrium

this procedure involves extensive surgery near the sinus node and sinus node artery, which might result in a higher incidence of late sinus node dysfunction [11].

Beyond infancy

PBF in a bidirectional cavopulmonary shunt is dependent on the SVC flow. In an observational study of healthy children by Salim et al. [12], the SVC flow accounted for 49% of cardiac output in newborn infants, reached a maximum of 55% at 2.5 years old and then gradually decreased to the adult value of 35% by 6.6 years old [13]. This shunt palliation appears to be good for the first 5–7 years, but then deteriorates after 7 years with progressive cyanosis secondary to pulmonary arteriovenous malformation [14–16]. Therefore, a Fontan operation is typically performed at 1.5–4 years old.

Perfect Fontan circulation requires no impediment across the pulmonary vascular bed (no PA stenosis or hypoplasia, no pulmonary venous obstruction, no increased PVR, no atrioventricular valve stenosis or regurgitation), good systolic/diastolic function, normal sinus rhythm and no systemic outflow/inflow obstruction. Additional interventions might be necessary during a Fontan operation to optimize Fontan circulation.

Modified Fontan operation

This involves inclusion of the IVC blood flow into the PA. Since the initial report in 1971 [17], several modifications have been undertaken. Total cavopulmonary connection (TCPC), which directly diverts the SVC and the IVC flow to the PA without the right side of the heart, is now common. This can be achieved by placing a conduit in the atrium (lateral tunnel Fontan; Fig. 1c) or placing a conduit outside the atrium (extracardiac Fontan; Fig. 1d). Advocates of an external conduit approach have stressed its superiority because the incidence of atrial arrhythmias may decrease with fewer sutures on the atrium. Advocates of lateral tunnel Fontan are concerned about the possibility of increased thromboses or stenoses with an external conduit. Fenestration is often made in the baffle to provide adequate cardiac output at the cost of desaturation. Several reports demonstrated that a fenestration reduces morbidity, particularly pleural effusions, the duration of hospitalizations and possibly mortality [11]. Fenestration can be closed spontaneously or with a device called umbrella during cardiac catheterization.

Although TCPC is standard, atriopulmonary connection was created in the distant past (Fig. 1e). It is energy-insufficient because the SVC and the IVC blood collide within the atrial chamber, creating turbulence [18], often leading into chronic atrial distension and late supraventricular arrhythmias. Some patients with this type of Fontan undergo conversion into TCPC to attempt to decrease arrhythmias.

Complication

Various morbidities are associated with the Fontan operation, such as ventricular dysfunction, arrhythmias, thromboembolism, protein losing enteropathy and plastic bronchitis [19].

Ventricular dysfunction

For patients born with single ventricle physiology, the short duration of volume load is preferable. However, patients

who underwent a Fontan procedure before the 1990s might have experienced long-standing, large ventricular volume-load because bidirectional cavopulmonary shunt was not part of staged palliation. Many patients have residual dilation, cardiac overgrowth and hypertrophy.

Arrhythmias

Twenty years of follow-up of atriopulmonary connection patients has shown that as many as 50% developed atrial tachycardia, usually in association with significant hemodynamic abnormalities [20]. The most common arrhythmia was intra-atrial re-entry tachycardia or atrial flutter. Ventricular arrhythmias were rare. The latter is probably related to poor ventricular function, if present. Given that arrhythmias do not favor this circulation, various interventions are attempted (e.g., pharmacological therapy, catheter ablation, pacemaker implantation, surgical Fontan revision into TCPC and the MAZE procedure).

Protein losing enteropathy

This is defined as persistent hypoalbuminemia (<3.0 mg/dl) in the absence of liver or renal disease, associated with clinical features such as abdominal pain, diarrhea, edema and ascites. Patients frequently have a limited hemodynamic reserve, demonstrated as increased systemic venous pressure, decreased cardiac index and increased end-diastolic ventricular pressure. The incidence is 3–14% on long-term follow-up. The development of protein-losing enteropathy is associated with a poor clinical course after a Fontan operation. Mortality is 50% within 5 years once the diagnosis is made.

Plastic bronchitis

Plastic bronchitis is a recurrent formation of branching bronchial casts that can cause partial or complete obstruction of the tracheobronchial tree. Their sizes vary from small segmental casts in the bronchi to large casts filling the entire lung. Often they are associated with dyspnea and desaturation. It might be necessary to remove the casts with rigid bronchoscopy [21].

Anesthetic considerations for noncardiac surgery

Presurgical repair

As shown in Table 2, cardiac lesions other than balanced parallel circulation need surgical intervention shortly after birth. However, some patients are required to have noncardiac surgeries even before cardiac intervention. Several case reports have described anesthetic management for these patients [22–24]. Noncardiac surgeries for this population occur mostly on emergent basis. Surgical indications are mostly related to gastrointestinal or airway issues. Necrotizing enterocolitis (NEC) is often a cause requiring surgical intervention. Neonates with inadequate systemic blood flow, whether it results from excessive PBF and CHF, or late diagnosis with a restrictive patent ductus arteriosus and shock, are at risk for NEC. Underlying disease might provide cardiovascular insight.

Because preoperative assessment is done within a limited period, adequate information must be obtained by physical examination, test results and discussion with primary team. An echocardiogram is especially important as a part of the workup. Surgical conditions necessitating neonatal emergencies are often associated with abnormalities in other organ systems [25]; thorough assessment of multiple organs is required.

Preoperatively, PGE_1 is infused continuously for suspected ductal-dependent circulation. PGE_1 can be discontinued in any patient once an echocardiogram determines a reliable nonductal source of systemic or pulmonary blood flow. Precise attention is necessary in case of PGE_1 infusion because it is associated with the risk of apnea. Surgical procedures must be undertaken whenever indicated, but it is crucial to keep in mind that the neonatal heart just after birth is fairly immature. Although contractility is enhanced in neonates by increased production and release of thyroid hormone, cortisol and catecholamine immediately after birth, only 30% of the myocardial mass comprises contractile tissue versus 60% in the mature myocardium.

Neonates have a lower velocity of shortening, a diminished length-tension relationship and a reduced ability to respond to afterload stress [26], so it is important to choose anesthetic management with less cardiodepressant effect. This is especially true for single ventricle physiology with the excess volume load to sustain parallel circulation. Compared with inhalational induction, intravenous induction is preferable with less cardiodepressant effect. The primary goal of hemodynamics is optimization of systemic and pulmonary circulations. Given that emergent gastrointestinal surgeries are the majority, placement of an arterial line, possibly a central line, is advisable. Inotropic support should be readily available during the procedure. Postoperative disposition should be made in advance.

Timing of cardiac surgery after these noncardiac surgical interventions is not well defined, and decisions must be made case by case, based on the patient's clinical status.

Precavopulmonary shunt patients

Patients will undergo cardiac intervention shortly after birth as in Table 2 other than ones with balanced parallel

circulation. Still at this stage, the ventricle(s) is operating with an excess in volume load required to sustain parallel circulation. When possible, elective, noncardiac surgery should be scheduled after superior cavopulmonary shunt. When necessary, gastrointestinal interventions seem common. The study by Jeffries et al. [27] showed that 41% of patients developed gastrointestinal complications following Norwood palliation for hypoplastic left heart syndrome. Some required a surgical gastrostomy tube for better feeding, and about one-fifth of them developed NEC.

Preoperative evaluation is essential to determine the balance of pulmonary and systemic blood flow, presence of cardiac failure and possible end-organ injury from reduced systemic perfusion. This includes a thorough history, cardiovascular status, respiratory status, functional status, vital signs, an electrocardiogram, an echocardiogram, chest radiograph and blood work. An echocardiogram assesses the patient's current ventricular function, atrioventricular valve function, arch obstruction and shunt patency. Input from the patient's cardiologist is also valuable. A respiratory illness must be evaluated because this can cause decreased SpO_2 because of decreased PBF from increased PVR and an increased A-a gradient. If surgery is not emergent, it may be advisable to postpone surgery until symptoms recover.

Anesthetic induction method may be affected by the patient's current status warranting surgery, but overall intravenous induction is advisable. These patients are at risk for subendocardial ischemia because volume overload increases ventricular end-diastolic volume and pressure. Under these conditions, a small increase in heart rate may reduce subendocardial perfusion enough to induce ischemia and ventricular fibrillation. The arterial oxygen saturation usually increases once the patient is anesthetized and paralyzed due to increased mixed venous oxygen saturation caused by reduced peripheral O_2 extraction, and improved cardiac output that is secondary to reduced myocardial work and afterload on the ventricle. However, increased pulmonary/systemic blood flow (Qp/Qs) with overventilation must be kept in mind as a cause of increased oxygen saturation after the induction of anesthesia. The degree of invasiveness in monitoring should be dictated by the patient's clinical condition and proposed procedure. However, an arterial line is useful for optimizing hemodynamics.

The primary goal in managing patients with single ventricle physiology is optimizing systemic oxygen delivery and perfusion pressure. This is achieved by balancing systemic and pulmonary circulations. Qp/Qs is a well-described tool to optimize systemic oxygen delivery. Based on the mathematical model, maximal oxygen delivery (the product of systemic oxygen content and systemic blood flow) is achieved for a ventricular output when Qp/Qs is at or just below 1 [28]. Qp/Qs is calculated as $(SaO_2 - SmvO_2)/(SpvO_2 - SpaO_2)$ [$SmvO_2$, $SpvO_2$ and $SpaO_2$ represent mixed venous, pulmonary vein and pulmonary artery saturation, respectively]. In patients with intracardiac complete mixing, SaO_2 is equal to $SpaO_2$, and Qp/Qs can be calculated using SaO_2 and $SmvO_2$ with the assumption of $SpvO_2$ 95–96% at room air, and $SmvO_2 = SsvcO_2$, However, this assumption is sometimes not accurate. Therefore, there is some limitation of estimation of Qp/Qs. $SmvO_2$ is the useful clinical indicator because oxygen delivery is parallel to $SaO_2/(SaO_2 - SmvO_2)$ at the same cardiac output [28]. At the same SaO_2, oxygen delivery goes up when $SmvO_2$ goes up and vice versa. Overall the reference of management is listed in Table 3. Changes in systemic vascular resistance (SVR) have a more dramatic influence on hemodynamics than changes in PVR [29]. Hypothermia should be avoided because it could increase SVR, leading into lower cardiac output.

The mainstay of anesthetic maintenance is by volatile anesthetics and opioids. Emergency cardiac resuscitation drugs and inotropic support should be readily available.

Although pneumoperitoneum would be not in favor for this physiology, the safety of laparoscopic surgery has been described [30, 31]. Pneumoperitoneum will reduce functional residual volume. Qp/Qs will be affected by systemic vascular resistance elevation, and CO_2 elevation with pneumoperitoneum. Therefore, vigilant observation as well as low threshold of converting open procedures may be advisable.

Postcavopulmonary shunt patients

In this physiology, hemodynamic performance tends to be most resilient because the single ventricle is no longer operating with the excess in volume load required to sustain parallel circulation; Qp/Qs is not subject to a wide variation in relation to manipulation of PVR; cardiac output is not entirely dependent on the PBF, as it is in the Fontan circulation [32]. When possible, it may be prudent to defer elective, major, noncardiac surgery until the child has recovered from cavopulmonary shunt procedures. However, any potential impact that a moderate elevation in SVC pressure has on the surgery should be weighed against the benefits of hemodynamic stability.

Recent respiratory illness must be addressed because of the passive nature of PBF. The expected systemic arterial oxygen saturation typically ranges from 75 to 85%. Preoperative laboratory testing is guided by the proposed surgical procedure and status of the patient. Recent electrocardiogram and echocardiogram will be indicated. Chest radiograph may be indicated, depending on the type of surgical procedure and clinical status. Cardiac catheterization may be considered when any deterioration of functional status is noted. NPO should be well coordinated to minimize the risk of dehydration.

General anesthesia seems to be the mainstay of anesthetics. While most of these patients will tolerate an

Table 3 Management of lesions with single ventricle physiology

Physiology	Management
Balanced flow	
Qp: Qs = 0.7–1.5: 1	No intervention
SaO_2 75–80%	
SaO_2–$SmvO_2$ 25–30%	
Overcirculated	
Qp: Qs > 2–3: 1	Raise PVR: Controlled hypoventilation, mild acidosis, Low FiO_2
Causes: Low PVR, large MBTS or Sano shunt, residual arch obstruction	Increase systemic O_2 delivery: Afterload reduction, inotropic support, hematocrit > 40%
SaO_2 > 85–90%	
SaO_2–$SmvO_2$ 35–40%	
Undercirculated	
Qp: Qs <0.7: 1	Lower PVR: Controlled hyperventilation, alkalosis, sedation/paralysis, aggressively treat atelectasis, nitric oxide
SaO_2 < 65–75%	Increase systemic O_2 delivery: inotropic support
SaO_2–$SmvO_2$ 25–30%	
Causes: High PVR, small MBTS or Sano shunt, pulmonary venous desaturation with underestimation of actual Qp: Qs	
Low cardiac output	
SaO_2 < 70–75%	Minimize O_2 consumption: Sedation/paralysis, inotropic support, afterload reduction
SaO_2–$SmvO_2$ 35–40%	
Causes: Ventricular dysfunction, myocardial ischemia, depressed contractility, afterload mismatch (Residual arch obstruction), AV valve regurgitation	

PVR pulmonary vascular resistance, *MBTS* modified Blalock-Taussig shunt, *AV* atrioventricular

inhalation induction from a cardiovascular standpoint, a few things must be appreciated. First, there may be venous congestion of the head and tongue due to a relatively high SVC pressure. Second, with Qp/Qs of 0.5–0.7, the speed of an inhalational induction will be slow. Third, anything causing high intrathoracic pressure such as coughing or breath holding will cause a significant reduction of PBF, and therefore significant desaturation. Whatever induction method is chosen, premedication is preferable because these patients have had and will have multiple interventions.

The goal of intraoperative management is to maintain adequate intravascular volume to enhance PBF, and minimize PVR. To accomplish the latter, maintaining adequate tidal volume with short inspiratory time, that is, minimizing mean airway pressure is preferred. While minimizing PVR is important, hyperventilation should be avoided since it is associated with a decrease in cerebral, SVC and PBF [33]. Intraoperative monitoring depends on the type of surgery and the patient status.

Limiting a significant increase in pulmonary vascular tone should be applied during emergence and postoperatively. Adequate pain control is necessary. Regional anesthesia may be advisable in large, open gastrointestinal surgeries. The control of postoperative agitation is important. Dexmedetomidine has been suggested to give stable hemodynamics over postoperative agitation [34, 35]. Also, in the same context, antiemesis prophylaxis is advisable because vomiting and retching not only potentially cause dehydration, but also impair respiratory mechanics, leading to cyanosis.

A laparoscopic procedure has been performed in this physiology as well. Similar considerations should be given as in precavopulmonary shunt patients.

Post-Fontan patients

Various case reports have been reported with their successful anesthetic management [36–46], including a case for failing Fontan [47].

The physiology of Fontan circulation

Because circulation is in series in Fontan patients, cardiac output is completely dependent on PBF. However, the loss of the right ventricle in this circulation could cause the following: First, the essential function of the right ventricle is not only to provide pulsatile flow through the pulmonary artery system, but also to maintain low pressure in the highly compliant systemic venous system, particularly the splanchnic bed. However, the lack of the right ventricle requires that systemic venous pressure should be elevated. Normally 70% of the total blood volume is contained on the venous side of the circulation, with the venous circulation having a capacitance 19 times that of the arterial circulation. Fontan patients have increased venous stiffness and decreased splanchnic venous capacitance [48, 49]. Fontan patients adapt to reduce venous capacitance (reduced unstressed volume) so that elevated systemic pressure can be maintained with a normal systemic venous volume (increased stressed volume). Therefore, stimuli that reduce stressed volume (venodilation from any source, blood loss and dehydration) do no good. Second, nonpulsatile PBF due to the loss of the right ventricle increases PVR by approximately 100% than that seen with pulsatile flow because recruiting distal pulmonary vasculature is lost without pulsatile flow energy, thereby reducing the area of the pulmonary vascular bed [50]. In addition, long-term loss of pulsatile PBF is accompanied by reduction of endothelial release of nitric oxide [51]. Therefore, Fontan patients are vulnerable to further increase in afterload.

PBF and cardiac output are exquisitely affected by respiratory mechanics. PBF is reduced, stagnant or reversed when intrathoracic pressure is positive [52, 53]. Also, a reduction in cardiac output with positive end expiratory pressure (PEEP) has been demonstrated [54]; therefore PEEP should be used with caution. In contrast, negative intrathoracic pressure at inspiration during spontaneous ventilation assists antegrade flow in the pulmonary arteries and the SVC. Penny and Redington [55] demonstrated that total forward PBF in normal inspiration increased by 64%. However, hypoventilation and hypoxia can increase PVR, which are disadvantageous. So, overall it is important to maintain adequate ventilation as well as minimize mean airway pressure.

Overall, the maintenance of effective PBF and cardiac output following the Fontan procedure depends on the pressure gradient between the pulmonary artery and pulmonary venous atrium (transpulmonary gradient, TPG). Typically, a systemic venous pressure of 10–15 mmHg and pulmonary venous atrium pressure of 5–10 mmHg, TPG of 5–10 mmHg is ideal for Fontan circulation.

Anesthetic management

Baseline exercise tolerance is typically reduced in Fontan patients, and their exercise reserve capacities are usually less than 50% of age-matched, control subjects. Recent illness should be addressed, especially respiratory tract infections, because changes in airway resistance and PVR are detrimental, and may pose an unacceptable risk for elective surgery in Fontan patients, as PBF and cardiac output will decrease if respiratory complications (e.g., laryngospasm or bronchospasm) occur. Arterial oxygen saturation should be high 90 s without fenestration, and high 80 s with fenestration, although it will not be 100% because the coronary sinus typically is incorporated into

the systemic side. Additionally, Fontan patients often have the aforementioned chronic complications.

An echocardiogram will help to assess ventricular function, atrioventricular valve function, Fontan pathway patency and status of fenestration. A chest radiograph may be indicated, depending on the type of surgical procedures and clinical status. Cardiac catheterization may be indicated before surgery if there has been a change in symptoms or deterioration in function. Hemodynamic assessment and fenestration device closure should be considered for surgeries anticipating significant volume requirements. Although there is no study addressing the necessity of preoperative fenestration closure, the risk of embolus with right-to-left shunt is theoretically high because baffle pressure is higher than common atrial pressure in this circulation.

Premedication is often desirable because these patients have multiple previous interventions. Often these patients may need a relatively higher amount, and previous anesthetic records are helpful. No specific anesthetic agents or regimens are recommended. However, sympathectomy with neuraxial blockade may result in a reduction of preload and be detrimental in the Fontan patient, although successful regional anesthesia has been documented [44–46].

Overall, general anesthesia or sedation seems to be the mainstay. Mask induction can be performed in a child with well-compensated Fontan physiology. However, in patients with ventricular dysfunction and reduced cardiac output, induction should be accomplished intravenously with drugs known to have minimal myocardial depressant effects. General principles during intraoperative management are to maintain adequate preload and preserve low PVR, sinus rhythm and ventricular contractility and reduce afterload. NPO must be well coordinated to minimize the risk of dehydration because Fontan circulation is preload dependent. Avoiding dehydration preoperatively and maintaining good hydration intraoperatively is important. However, aggressive volume expansion is not advisable given that these patients can have diastolic dysfunction. This is especially important when one has to anesthetize patients with failing Fontan circulation. One reason why Fontan patients need large amounts of volume intraoperatively is the effect anesthetics have on stressed volume. Because Fontan patients have increased stressed volume, vasodilation from anesthetic agents can increase stressed volume significantly. Therefore, dopamine or some vasoconstrictors may potentially be beneficial in minimizing the increase in stressed volume and avoiding large volume administration. To minimize PVR, maintaining adequate ventilation and minimizing mean airway pressure are important. When positive ventilation is used, this will be established by moderately elevated tidal volumes (10–15 ml/kg), low respiratory rates, limiting inspiratory time and avoiding excessive PEEP. Pharmacological intervention of PVR is also considered. Maintaining end-diastolic ventricular pressure in the low range is necessary to accomplish good Fontan circulation. Therefore, the maintaining sinus rhythm and contractility is important. Afterload reduction with milrinone may prove helpful for improving contractility and decreasing atrioventricular valve regurgitation if any.

Placement of central venous line into the SVC will allow monitoring of baffle pressure (if no baffle obstruction) and mixed venous oxygen saturation.

Preparing for possible intraoperative bleeding is important. Bleeding can be from increased venous pressure as described in scoliosis surgery [37]. The effect of residual anticoagulants may play a role. Fontan patients seem to be prone to increased risk of thrombosis. Patients occasionally take aspirin and Coumadin. Fontan patients have a derangement of various coagulation factors such as lower concentration of protein C, factor II, V, VII, X, ATIII and plasminogen, but elevated factor VIII [56]. Anticoagulants should be stopped before elective surgery.

A limiting significant increase in pulmonary vascular tone should be applied during emergence and postoperatively. Postoperative pain control is important. Splinting from pain possibly could increase PVR. However, hypoventilation from an overdose of pain medication also increases PVR.

The effect of an operative approach on this physiology needs consideration. Increased intraabdominal pressure from insufflation and elevated arterial carbon dioxide tension in laparoscopic surgery will increase PVR and decrease cardiac output. Successful laparoscopic surgery has been described in functional Fontan [40, 41]. Limiting insufflation pressure to 10–12 mmHg seems to reduce the adverse effects of laparoscopy in these case reports. Adequate hydration, attention to the signs of decreasing PBF, communication with the surgeon and a low threshold to convert to an open procedure is essential when performing laparoscopic surgery in patients with Fontan physiology. In failing Fontan, these discussions might not be applicable.

Additional issues

Pregnancy with Fontan

With the improvement of long-term survival, patients with Fontan circulation have reached child-bearing age. The physiological change accompanied with pregnancy may potentially put a burden on the mother with Fontan physiology. The surplus of systemic venous return may lead to complications, such as atrial arrhythmias, and CHF. The risk of pregnancy is reported as limited for women with Fontan palliation who have adequate repair without significant long-term sequelae [57]. It seems that reports on patients with atriopulmonary connection have been a

majority, who frequently develops atrial arrythmias during pregnancy [57]. The information on patients with TCPC will increase in the future.

Anesthesiologists may encounter those patients earlier than their due dates, because the high incidence of premature labor and delivery has been observed in this population [57]. The timely insertion of an effectively working epidural catheter is advisable to avoid emergency situations in which anesthesiologists are forced to rapidly induce general anesthesia. Gradual titration of epidural anesthetics with hydration is preferable in non-emergent situations. Both successful general anesthesia and regional anesthetics have been described in emergent cesarian section [44–46]. The anesthetic considerations discussed above should be followed.

Heterotaxy syndrome

Despite the improvement of outcomes for patients with single ventricular physiology, patients with heterotaxy syndrome in association with single ventricular physiology in association with a functional single ventricle who undergo single ventricle palliation continue to exhibit a high mortality and morbidity [58]. Overall anesthetic management about this entity can be referred to the recent review [59]. These patients tend to have risk factors associated with poor Fontan outcome, including abnormal heart rhythm, atrioventricular valve regurgitation, pulmonary vein stenosis and increased pulmonary vascular resistance.

Acknowledgments This is supported in part by Grant-in-Aid for Scientific Research© (nos. 20591844 and 18591726) (S.Uezono).

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