Congenital Heart Disease in the Adult Presenting for Non-Cardiac Surgery

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Introduction: Advancement of surgical techniques and medical management patients with congenital heart lesions has allowed this patient population to survive into adulthood, even with the most complex congenital lesions. As a result, patients with congenital heart lesions are increasingly encountered in the adult perioperative setting for laparoscopic, obstetric, plastic, spine, and other non-cardiac procedures. In the adult population, repaired or chronic non-palliated congenital heart lesions offer unique challenges. Therefore, adult anesthesiologists must be familiar with the anatomy, long-term manifestations, and unique perioperative management of adults with congenital heart disease.

Shunting Lesions: Intracardiac shunts, or communications between atria (atrial septal defects, ASD), ventricles (ventricular septal defects (VSD), or great vessels (patent ductus arteriosus, e.g.) are the most common type of congenital heart lesions. These communications allow shunting of blood from areas of high to low pressure, typically left to right. While many are repaired surgically, these lesions may go unrecognized until adulthood. The common consideration for shunting lesions is structural changes in the pulmonary vasculature resulting in pulmonary hypertension with subsequent right ventricular failure. Chronic severe pulmonary hypertension in the context of shunting lesions may result in Eisenmenger syndrome with reversal of shunt (right to left) and hypoxemia.

Preoperative evaluation should include a thorough history and physical, with particular attention to pulmonary hypertension and right ventricular failure. Right ventricular failure is heralded by elevated jugular venous pressure, hepatomegaly, hepatic dysfunction, or peripheral edema. 12-lead electrocardiogram is useful to detect atrial fibrillation or flutter, common with atrial enlargement in these patients. Patients with previous VSD repair may have heart block from patching proximal to the conduction system. These patients may have a permanent pacemaker, which requires interrogation and proper programming in the perioperative setting. Right heart catheterization useful for determining preoperative pulmonary artery pressures.

The primary goals for the anesthesiologist caring for these patients include avoidance of intravascular air, decreasing pulmonary vascular resistance, and maintaining adequate right ventricular function. Adequate ventilation and oxygenation is paramount to the care of these patients. Hypercarbia secondary to excessive premedication with midazolam or narcotics can result in right ventricular failure and cardiovascular collapse in patients with severe pulmonary hypertension. Similar effects may be seen with induction-induced apnea. Avoidance of sympathetic-
induced pulmonary vasoconstriction with adequate anesthesia, analgesia, and normothermia is recommended. Inhaled prostaglandins or nitric oxide are useful for reversible pulmonary hypertension. Right ventricular dysfunction in this setting may require inodilator drugs, such as milrinone or dobutamine.

Complex lesions of the great vessels: Lesions of the great vessels include transposition of the great arteries (TGA) and truncus arteriosus (TA). Critical for survival, these lesions require intracardiac shunting for mixing of blood and adequate oxygenation. Left unrepaired, these lesions typically result in severe pulmonary hypertension and are usually fatal.

TGA is a discordance of the ventricles and great vessels, resulting in a parallel circulation. To simplify, oxygenated blood from the lungs takes the following route: pulmonary veins—left atrium—left ventricle—pulmonary artery—lungs. Deoxygenated blood enters right atrium—right ventricle—aorta. Early repairs, such as the Mustard or Sennig procedures, involved an intra-atrial baffle or tunnel that redirects blood to the anatomically correct side. In these patients, the right ventricle is remains the systemic ventricle. Considerations for these patients are right ventricular failure and atrial arrhythmias from the baffle suture lines. Repair of TGA since the mid-1980s is via the arterial switch procedure, where the pulmonary artery and aorta are surgically reversed. Unique considerations include stenosis at the anastomotic sites (PA or aorta) and pulmonary valve or aortic insufficiency.
Similarly, repair of truncus arteriosus involves separation of the great vessels to their respective ventricles and VSD repair. Considerations are anastomotic strictures, aortic valve insufficiency, persistent VSD, and ventricular arrhythmias. Note that congenitally corrected TGA (left atrium—right ventricle—aorta; right atrium—left ventricle—pulmonary artery) does not require surgical correction. However, the anatomic right ventricle serves as the systemic ventricle, resulting in an increased incidence of heart failure over time.

Cardiac MRI is a useful tool for delineating the anatomy and identification of conduit or vessel abnormalities.

**Tetralogy of Fallot:** TOF is one of the most commonly encountered adult congenital heart lesions. TOF is defined by the presence of a VSD, overriding aorta, right ventricular outflow tract obstruction (or pulmonary valve stenosis), and right ventricular hypertrophy. Most patients encountered have received a definitive repair of this lesion, though there exists a rare population of unrepaired TOF. Unrepaired lesions, unlike our previous categories, obstruction to pulmonary flow is protective against pulmonary overload and pulmonary hypertension. However, right ventricular failure secondary to RVOT obstruction is common. Further, bidirectional or right to left shunting through the VSD may result in profound hypoxemia. This is treated by intravascular volume expansion, beta blockade, and phenylephrine. These maneuvers decrease RVOT obstruction and increase systemic vascular resistance, promoting left to right shunting.

More commonly encountered is the patient with repaired TOF with a VSD patch and augmentation of the RVOT or pulmonic valve. A common surgical method to augment the RVOT is transannular patching. These repairs have resulted in severe pulmonary valve insufficiency, RV volume overload, and right ventricular failure. The combination of RV enlargement and RV scarring at the ventriculotomy site causes localized slowing of the conduction system and malignant ventricular arrhythmias. A QRS interval $> 180$ ms is predictive of re-entrant monomorphic ventricular tachycardia. It is not uncommon for patients with repaired TOF to have an implantable cardioverter defibrillator (ICD). In addition, the overriding aorta may result in aortic insufficiency over time. VSD patch leak is also a consideration. Intravascular air should always be avoided in congenital heart patients, even with repaired lesions.

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**Figure 3:** Sequelae of TOF repair. A. TEE modified RV outflow tract view with dilated right ventricle, RVOT and pulmonic valve. B. Same TEE view with severe pulmonic insufficiency by color Doppler. C. Left ventricular outflow tract TEE view with dilated RV, VSD patch, and overriding aorta.
Complex congenital cardiac lesions: Several types of complex congenital heart lesions exist where preservation of biventricular function is not feasible. These lesions include hypoplastic left heart syndrome, tricuspid atresia, unbalanced atrio-ventricular septal defects, double inlet left ventricle, double outlet right ventricle, and some forms of heterotaxy syndrome.

While the initial anatomy of complex congenital heart lesions is rather variable, surgical treatment often results in the common endpoint, Fontan palliation. Regardless of the initial congenital lesion, there are two essential components of Fontan physiology: 1) the presence of a single systemic ventricle (SV), either an anatomic right or left ventricle, and 2) systemic venous return enters the pulmonary artery without ventricular assistance.
The second point is the result of surgical reconstruction in a staged fashion. Over the past several decades, a total cavopulmonary connection (TCPC) is the method of choice. TCPC involves surgical anastomosis of the superior vena cava (SVC) and the inferior vena cava (IVC) to the pulmonary artery.

**Evaluation of the adult with Fontan physiology involves a thorough history and physical examination. Medical records, most recent catheterization, cardiac MRI, and laboratory data are invaluable to elucidate cardiac anatomy, oxygen saturation, pressure data, and SV function. Medical history should focus on current and changes in health status, exercise capacity, recent hospitalization, and current medications. Physical examination of a well-functioning Fontan patient should be relatively unremarkable. Fontan patients are expected to be normothermic and acyanotic; peripheral arterial pulses are palpable and precordial auscultation is devoid of murmurs. Normal oxygen saturation by pulse oximetry ranges from 90 to 95% in patients with a well-functioning Fontan, due to a small shunt via the coronary sinus. Chest radiography should reveal a normal cardiac silhouette; normal sinus rhythm on electrocardiography (ECG) is expected.

Without the presence of a ventricle, pulmonary blood flow is dependent upon the maintenance of a transpulmonary gradient (TPG)—a 5 to 8 mmHg gradient between the central venous system and the left atrium. Decreases in transpulmonary gradient occur with inadequate preload, depressed ventricular function, or increases in pulmonary or systemic vascular resistance. Decreased TPG results in decreased cardiac output, systemic hypotension, and poor tissue perfusion. Induction of anesthesia, hypoventilation / hypoxemia, postoperative pain, inadequate anesthesia / sympathetic stimulation, prolonged NPO status may all result in a decreased TPG.

Standard intraoperative monitoring is adequate for patients with a well-functioning Fontan, particularly for procedures where minimal hemodynamic derangements or fluid shifts are expected. Upper extremity blood pressure measurements should be taken on the opposite side of a previous Blalock-Taussig (subclavian artery to pulmonary artery) shunt to avoid an artificially low blood pressure. Central venous access risks TCPC thrombosis, endocarditis, and vascular or intrathoracic injury. Achieving adequate cardiac output is the mainstay of anesthetic management of patients with Fontan physiology. Conservative administration of intravenous fluids prior to induction of anesthesia may be prudent to counteract NPO status and venodilation from anesthetic agents. While there is predilection of volatile anesthetics and positive pressure ventilation to cause hypotension in Fontan patients, general anesthesia must be adequate to prevent sympathetic stimulation leading to detrimental increases in pulmonary vascular resistance. Similarly, inadequate

**Figure 5: Fontan schematic in patient with hypoplastic left heart syndrome: Salient features are total cavopulmonary return, atrial communication, and right ventricle as systemic ventricle.**

SVC: superior vena cava; IVC: inferior vena cava; PA: pulmonary artery; PV: pulmonary veins; LA: left atrium; RA: right atrium; ASD: atrial septal defect; RV: right ventricle; Ao: aorta
tidal volumes lead to atelectasis, hypoxemia, and hypercarbia. Since the majority of venous return with intermittent positive pressure ventilation occurs during expiration, inspiratory time should be shortened accordingly. Caution must be taken with monitored anesthesia care (MAC) since hypoventilation leads to increased PVR. Epidurals have been used successfully in the perioperative management of Fontan patients. Epidural dosing is progressively titrated to minimize sympatholytic effects, venodilation, and decreased pulmonary return. Endocarditis prophylaxis is recommended Fontan patients, due to increased endocarditis risk with palliative shunts and conduits.

In spite of excellent survival rates, Fontan patients are at progressive risk for failing palliation. Preoperative findings indicative of a failing Fontan include fatigue, decreased activity level, weight gain or volume retention, palpitations, syncopal or pre-syncopal episodes, oxygen saturation below 90%, and dyspnea. Physical findings are often secondary to SV failure, atrial arrhythmias or sinus node dysfunction, AV valve dysfunction, infectious processes, protein-losing enteropathy and systemic-to-pulmonary artery collaterals. Abnormalities on chest radiography consistent with failing Fontan include cardiomegaly, pulmonary edema, and pleural effusions.

Failing Fontan: Organ System Involvement

- Pulmonary
  - elevated pulmonary vascular resistance
  - cyanosis
  - pleural effusions
  - plastic bronchi
- Hepatic
  - cirrhosis
  - hepatomegaly
  - edema
  - protein-losing enteropathy
  - mesenteric fibrosis
  - intestinal lymphangiectasia
  - esophageal varices
- Renal Failure
- Vascular
  - Venovenous occlusion
  - Collaterals
  - Veno-venous
  - Arteriovenous
  - Veno-arterial
- Cardiac
  - arrhythmias
  - SVT
  - Atrial fibrillation
  - Atrial flutter
  - IART
  - Ventricular
  - heart failure
  - cardiomegaly
  - peripheral or pulmonary edema
  - decreased exercise capacity
- Hematologic
  - thromboembolic
  - pulmonary embolus
  - CVA
  - coagulopathy
  - anemia
  - lymphopenia

The incidence of arrhythmias, particularly atrial tachyarrhythmias, increases with time. An arrhythmia somewhat unique to Fontan patients is intra-atrial reentrant tachycardia (IART. IART is a slow atrial flutter with 2:1 AV conduction. The diagnosis is often elusive as the second p wave is buried within the QRS complex or the T-wave. Protein-losing enteropathy (PLE) is an enigmatic problem with a significant mortality and no clear treatment modality. Pleural effusions, decreased SpO2, peripheral edema, and ascites are consistent findings in patients with PLE. Decreased levels of albumin and total protein, and stool positive for alpha-1 antitrypsin confirm the diagnosis of PLE.

Hepatic cirrhosis is a commonly associated with failing Fontan palliation. Coagulopathy and esophageal varices may be present in these patients. Thromboembolic events, including pulmonary embolism and cerebral vascular accidents, occur in up to 25% of patients, and are a source of significant morbidity and mortality for patients with Fontan physiology. Prior to elective cardioversion, intracardiac thrombi should be excluded by TEE.
Plastic bronchitis, a late finding after Fontan palliation, consists of obstructive bronchial casts within the tracheobronchial tree. Once new-onset or worsening failed Fontan physiology is evident, elective surgery should be postponed for further investigation.

Recently published guidelines for preoperative evaluation of adults with Fontan physiology are as follows:
Fontan: non-cardiac surgery preoperative evaluation

- Poor or declining functional status
  - Abnormal physical examination
    - cyanosis
    - clubbing
    - hepatomegaly
    - ascites
    - Oxygen saturation <90%

- Functional status >4 METS
  - Oxygen saturation > 90%
  - Unremarkable physical examination

Chose radiograph
Echocardiogram

Abnormal

- Cardiomegaly
- Pleural effusions
- Restrictive atrial septum
- Evidence of caval, branch pulmonary artery, or aortic obstruction
- Moderate-to-severe AV valve regurgitation
- Patent Fontan fenestration or residual shunts

YES
Cardiac catheterization

NO
Optimal medical status

Proceed with non-cardiac surgery

PT/PTT, CBC, electrolytes, BUN/Cr, LFTs
- ECG
- Chest radiograph

Normal
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