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The Postoperative Cardiac Patient

William Tsai, MD,* Bruce L. Klein, MD†

The number of children surviving medical management and surgical repair of congenital heart lesions has steadily increased. Children with complex heart defects are therefore seen with increasing frequency in emergency departments. This article reviews the epidemiology of congenital heart disease, some common types of surgical repair, and a few postoperative complications and their management.
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The number of children surviving surgical repair of complex congenital heart defects is increasing. These children with complex heart defects are being seen with increasing frequency in the emergency department (ED). Management of complications in these children can be difficult, often provoking anxiety in the ED physician. Knowing the types of congenital heart operations and their associated postoperative complications is valuable in the initial diagnosis, stabilization, and treatment for these children. This article discusses the epidemiology of congenital heart disease, some common types of surgical repair, and a few postoperative complications and their management.

Epidemiology

Congenital heart defects are diagnosed in 8 to 9 of 1000 live births. Currently, 1 million people in the United States live with a congenital heart defect [1]. In 2000, 25000 congenital cardiovascular operations were performed and 130000 hospitalizations were necessary, generating charges of 6.5 billion dollars [2]. Although congenital heart defects remain a major cause of death in infancy and childhood, mortality has declined 39% from 1979 through 1997 [3].

Types of Cardiac Lesions

Several main problems distinguish the pathophysiology of congenital heart defects. There may be (1) excess volume load in which one or both ventricles are forced to pump an excess volume of blood, (2) excess pressure load to one or both ventricles resulting from obstructed outflow from the cardiac chambers, (3) cyanosis due to reduced pulmonary blood flow or inadequate mixing between 2 parallel circulations, (4) diminished systemic blood flow, or (5) mixed lesions with features from more than 1 category. Table 1 categorizes the different types of cardiac lesions based on this physiologic classification.

Common Types of Repair

Dr Robert E. Gross completed the first successful surgical procedure for a congenital heart defect when he ligated a patent ductus arteriosus in 1938. In 1948, Blalock and Taussig reported the surgical creation of a systemic-to-pulmonary-artery shunt, pioneering the surgical era for the treatment of cyanotic congenital heart disease. John Gibbon performed the first successful open heart procedure using artificial cardiopulmonary bypass on a young woman with an atrial septal defect in 1953, and thus, all of the necessary building blocks for the effective surgical

*Pediatric Critical Care and Emergency Medicine, Children's National Medical Center, NW, Washington, DC.
†Pediatric Transport Service, Children's National Medical Center, NW, Washington, DC 20010.
Reprint requests and correspondence: Bruce L. Klein, MD, Pediatric Transport Service, Children's National Medical Center, 111 Michigan Ave., NW, Washington, DC 20010.
(E-Mails: m.j.kate@hotmail.com, bklein@cnmc.org, brucelklein@comcast.net)

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Table 1 Classification of congenital heart defects. Some illustrative examples.

Volume Load	Pressure Load	Cyanotic, Diminished Pulmonary Flow	Diminished Systemic Flow	Mixed Categories
Anomalous pulmonary venous drainage	Aortic valve stenosis	Pulmonary atresia with intact ventricular septum	Coarctation of the aorta	Double outlet right ventricle
Aortopulmonary window	Pulmonary valve stenosis	Pulmonary atresia with ventricular septal defect	Hypoplastic left heart syndrome	Tetralogy of Fallot
Atrial septal defect	Subaortic stenosis	Tricuspid atresia with pulmonary stenosis	Interrupted aortic arch	Transposition of the great arteries
Atrioventricular canal defect	Supravalvular aortic stenosis		Tricuspid atresia with aortic stenosis	Truncus arteriosus
Patent ductus arteriosus	Supravalvular pulmonic stenosis			
Valvular insufficiency				
Ventricular septal defect				

management of children with congenital heart defects were in place. The last several decades have seen refinements in both surgical procedures and cardiopulmonary bypass techniques that have enabled surgical teams to accomplish complete or palliative repairs on younger and younger children. Furthermore, improvements in preoperative diagnostic accuracy and the ability to stabilize critically ill patients before surgery have improved outcomes significantly.

There are 2 categories of surgery for congenital heart defects. Ideally, complete correction of the defect results in a normal 4-chamber heart and restoration of normal blood flow to the pulmonary and systemic circulations. If complete correction is not feasible, the alternative is palliation. Rather than correcting the abnormality, palliative surgery seeks to circumvent the adverse consequences of the defect. Palliation is sometimes used as a temporary solution while awaiting complete correction at a later time. Other abnormalities are only amenable to palliation, with no option for correction. Table 2 lists examples of complete repairs.

Table 2 Examples of surgeries that result in complete repair.

Procedure	Diagnosis	Description
Arterial switch	D-transposition of the great arteries	Aorta and pulmonary artery moved to proper ventricles, coronary arteries reimplanted
Atrial septal defect closure	Atrial septal defect	Closes atrial septal defect
Coarctation repair	Coarctation of the aorta	End-to-end anastomosis
Patent ductus arteriosus ligation	Patent ductus arteriosus	Interruption of patent ductus arteriosus
Valve replacement	Valvular disease	Replaces any valve
Ventricular septal defect closure	Ventricular septal defect	Closes ventricular septal defect

Palliative Repairs

Numerous palliative repairs exist, ranging from pulmonary artery banding for control of excessive pulmonary blood flow to the Mustard and Senning procedures for transposition of the great arteries. Although a full description of all palliative procedures is beyond the scope of this article, systemic-to-pulmonary-artery shunts and Fontan procedures will be discussed in greater detail. Table 3 lists common palliative repairs.

Systemic-to-pulmonary-artery shunts include the Blalock-Taussig (BT) shunt, modified BT shunt, and central shunts; classic Waterston and Potts shunts are less common. Their purpose is to provide a secure source of pulmonary blood flow in cyanotic patients with decreased pulmonary blood flow. Variations of the BT shunt and its modifications can be performed on infants and children and may provide good long-term palliation. Systemic-to-pulmonary-artery shunts are usually used for temporary palliation while awaiting more definitive repair.

In 1971, Fontan and Baudet described a palliative procedure that resulted in complete separation of systemic and pulmonary circuits despite having only 1 functional ventricle. In the original procedure, superior vena caval blood was directed to the right pulmonary artery (Glenn shunt), and inferior vena caval flow was directed into the left pulmonary artery via the right atrial appendage; this left no communication between the pulmonary arteries. The operation is no longer done this way. Currently, the Fontan is usually performed in stages. The superior vena cava is anastomosed directly to the right pulmonary artery, and the right and left pulmonary arteries remain in continuity. The inferior vena cava return to the right atrium is directed toward the lower

Table 3 Common palliative surgical procedures.

Procedure	Diagnosis	Description
BT, modified BT, central, Cooley, or Waterston shunt	Lesions with cyanosis and diminished pulmonary blood flow	Systemic-to-pulmonary-artery shunt (eg, subclavian artery to pulmonary artery communication). Designed to provide reliable source of pulmonary blood flow
Glenn, bidirectional Glenn, hemi-Fontan, or Fontan procedure	Lesions with single ventricle physiology (eg, double inlet left ventricle, pulmonary atresia with intact ventricular septum, tricuspid atresia, among others)	Bidirectional Glenn or hemi-Fontan anastomosis results in a connection from superior vena cava to pulmonary artery. The full Fontan procedure results in a total cavopulmonary anastomosis, so that flow from both the superior vena cava and the inferior vena cava go to the pulmonary artery. All blood flow to the lungs is passive
Norwood repair (generally refers to Stage 1)	Hypoplastic left heart syndrome	Stage 1: Construction of neoaorta from the main pulmonary artery and the ascending aorta (usually hypoplastic). The main pulmonary artery is over sewn and a BT shunt is created Stage 2: Placement of a Glenn shunt and takedown of the BT shunt Stage 3: Complete cavopulmonary connection (Fontan repair)

portion of the superior vena cava either by means of a baffle within the right atrium or with an extracardiac conduit; this flow then passes through the lower superior vena cava to the pulmonary arteries. The end result of this total cavopulmonary connection is to establish passive blood flow from the systemic venous circulation directly into the pulmonary arteries, bypassing the ventricle entirely. The single functional cardiac ventricle pumps oxygenated blood returning from the lungs to the systemic arterial circulation.

Although various modifications of the original operation exist, it is common to use the term *Fontan operation* to designate any operation that reroutes systemic venous return from the superior and inferior vena cavae directly into the pulmonary arteries passively, without power from a pumping ventricle. The single functional ventricle is recruited to pump blood to the systemic circulation. The Fontan procedure is potentially useful for any cardiac lesion with only a single functional ventricle.

History and Physical Examination

Evaluation of the postoperative cardiac patient begins with a complete history and physical examination. Specific questions regarding the original defect and type of repair are especially important, as are plans for future operations. In cyanotic patients, baseline arterial oxygen saturation, which can range from 65% to 90%, should be ascertained. The presence of residual defects as well as any history of prior complications (eg, persistent pleural or pericardial effusions or episodes of transplant rejection) should be elicited.

Several important clues may be detected during the physical examination. The extremities should be examined for edema or stigmata of poor perfusion. Signs of congestive heart failure, such as hepatomegaly, pulmonary edema, or cardiogenic shock, should be noted. The presence of a midline sternotomy or right or left thoracotomy scar can be a clue as to what procedure was performed (in case the patient or caretaker is unaware). Physical examination includes careful auscultation for outflow or regurgitant murmurs as well as pericardial or pleural rubs. Consultation with the patient's cardiologist or primary physician may be particularly helpful in understanding the patient's clinical status.

Postoperative Complications

Systemic-to-Pulmonary Shunts

Early complications may include excessive shunt flow, usually noted within the first several days after the surgery. Excessive flow results in symptoms typical of pulmonary edema and increased volume load of the heart (congestive heart failure). Inadequate shunt flow may also occur. This can be acute because of thrombosis of the shunt and presents with sudden cyanosis. Urgent intervention is usually necessary. Pharmacologic thrombolytic therapy may occasionally be successful in this setting; mechanical restoration of pulmonary flow is

usually required, either by catheter-directed therapy or surgical intervention.

Because these shunts are usually prosthetic tubes of fixed size, the amount of pulmonary flow does not increase as the patient grows. Therefore, the shunt will eventually become too small, resulting in gradually progressive cyanosis, which indicates the need for additional surgery (either more definitive surgery or placement of a second shunt).

Late Complications After Fontan Operation

Late complications after Fontan operation encompass entities that are, in part, due to the elevated systemic venous pressure required to maintain pulmonary blood flow after the operation. This increased central venous pressure may result in early postoperative pleural effusions (thought to be due to lymphatic congestion) that may persist for weeks. Supraventricular tachyarrhythmias, thromboembolic complications, protein-losing enteropathy, and pulmonary arteriovenous fistulae also occur.

The incidence of supraventricular tachyarrhythmias immediately after the modified Fontan procedure ranges from 20% to 37.5%, and late onset tachyarrhythmias approach 20% by 5 years post operation [4-6]. Types of supraventricular tachyarrhythmias include atrial flutter, atrial ectopic tachycardia, and atrioventricular re-entry tachycardia. Symptoms may include chest pain, palpitations, hypoxemia, and/or frank congestive heart failure in patients with compromised ventricular function, although occasional patients may be asymptomatic. The ED evaluation of the child presenting after the Fontan operation with chest pain, palpitations, worsening hypoxemia, congestive heart failure, or new pleural effusion should include an electrocardiogram (ECG) and consultation with a pediatric cardiologist. Atrial flutter and supraventricular tachycardia may be treated with medications such as digoxin or amiodarone. Direct-current cardioversion should be used in cases associated with acute hemodynamic compromise.

Thromboembolic complications in Fontan patients are being recognized with increased frequency. Thrombosis in the systemic venous circulation may be life-threatening and result in decreased cardiac output with its attendant morbidity and mortality. Furthermore, many Fontan procedures include a fenestration, a small communication between the systemic venous system and the systemic circulation. An embolism crossing at the fenestration would then reach the aorta and could cause arterial obstruction anywhere in the systemic distribution. Cerebrovascular events, inferior or superior vena cava syndrome, pulmonary embolus, hypoxemia, and chest pain can complicate the postoperative care of these single-ventricle patients.

Thromboemboli after Fontan procedures occur at a rate of 3.9 events per 100 patient-years and have been associated with significant morbidity and mortality [7]. Formation of thrombi probably results from decreased systemic venous flow rates due to elevated central venous pressures; in addition, coagulopathies may contribute to the formation of venous thrombi [8]. Dehydration and supraventricular arrhythmias also increase the risk of subsequent thrombosis.

Fontan patients with evidence of thrombus formation or significant hypoxemia should receive intravenous hydration to help improve pulmonary blood flow and cardiac output. A pediatric cardiologist should be consulted. Echocardiography should be performed and consideration given to ventilation-perfusion or computed tomographic scanning if pulmonary embolus is suspected. Cardiac catheterization may be indicated. Treatment may include heparinization or thrombolytic therapy.

Postpericardiotomy Syndrome

Postpericardiotomy syndrome may present as an acute illness with fever and a pericardial or pleural inflammatory reaction. It usually develops within the first 1 to 2 weeks after open heart surgery but may appear as late as several weeks after surgery.

The incidence of postpericardiotomy syndrome has remained stable at 30% (10%-60%) [9,10]. In children younger than 2 years, the incidence is 3.5% [11]. It most commonly occurs after open heart surgery but may be seen after pacemaker insertion and angioplasty. When associated with myocardial infarction, the postpericardiotomy syndrome is known as Dressler's syndrome.

The etiology of postpericardiotomy syndrome is unclear and may involve an autoimmune process and the development of antimyocardial antibodies [12]. Postpericardiotomy syndrome frequently is associated with a concomitant viral infection. Circulating antigen-antibody complexes may cause the subsequent development of symptoms.

Children may present with fever of 38°C to 39°C and generalized malaise. Young children have vague symptoms, displaying irritability, diminished appetite, and nonspecific chest pain. Postpericardiotomy syndrome can be particularly difficult to identify in infants and children not yet old enough to describe symptoms. Older children and adults have more specific chest pain and may complain of chest tightness that varies with the respiratory cycle. They may complain of pain that radiates to the neck, shoulder, or back. Sometimes, however, the symptoms may be ambiguous and nonspecific (eg, resembling a viral illness with vague gastrointestinal symptoms).

Fever, tachycardia, pericardial or pleural rubs, and fluid retention are noted during the illness. A pericardial friction rub may be absent because of the separation of pericardial surfaces by increased fluid volume if the effusion is large. Pericardial tamponade is rare, with an incidence of 0.1% to 6%, and appears to be more frequent in children receiving anticoagulation [13,14]. Pulsus paradoxus, weak and thready pulses, jugular

venous distension, and hepatomegaly indicate the possibility of tamponade.

A complete blood count may demonstrate leukocytosis and concomitant eosinophilia. Possible ECG changes include diffuse ST-T wave abnormalities, but the ECG may be normal. When there is a significant effusion, chest x-ray reveals an enlarged cardiac shadow. However, echocardiography specifically identifies pericardial fluid and remains the gold standard for diagnosis.

Treatment includes the use of salicylates such as aspirin [15]. If signs of tamponade are present, pericardiocentesis is indicated. Indomethacin and ibuprofen have been found to be effective in relieving and shortening the duration of symptoms in adults. Steroids are reserved for cases with large pericardial effusions or for those in whom salicylates are not effective.

Infective Endocarditis

Infective endocarditis is defined as a microbial infection of the endocardial surfaces of the heart. Mortality was universal before the introduction of sulfonamides and still remains high. Although less frequent than seen in adults, infective endocarditis accounts for 1 of 1280 to 1 of 4500 pediatric inpatient admissions a year [16]. Although the most common underlying heart defect was caused by rheumatic fever in the 1970s, now, nearly 70% of pediatric endocarditis cases occur in children with a congenital heart defect. Ventricular septal defect, tetralogy of Fallot, and aortic stenosis are the most common congenital defects associated with infective endocarditis [17]. Neonates sometimes develop right heart endocarditis, possibly because of indwelling vascular devices or long-term central venous catheter use [18].

Infective endocarditis typically develops on endocardial surfaces that are subject to pressure gradients and turbulent blood flow. The resulting damage to the endothelium predisposes to platelet adherence and fibrin activation. For example, infective endocarditis may occur on the low-pressure side of a ventricular septal defect or involve areas of the heart subject to turbulent flow, such as a bicuspid aortic valve. Foreign material such as prosthetic valves may also serve as foci for the development of infective endocarditis. When bacteremia occurs (eg, from dental procedures or spontaneously), these microorganisms can get entrapped within this meshwork and become a nidus of infection.

Clinical symptoms in infective endocarditis reflect activation of the immune response, local tissue damage, and cardioembolic phenomena. Patients present with fever, malaise, and myalgias, and they may be misdiagnosed with a viral syndrome or other intercurrent illness. Physical examination should assess for embolic sequelae, such as petechiae, splinter hemorrhages, or Roth's spots. A new or unexplained murmur suggests valvular involvement. Heart failure may result from damaged valves or chordae, as well as from the development of fistulous shunts.

Diagnosis in the emergency department is based largely on history, symptoms, signs, and clinical suspicion. The hallmark of diagnosis, however, is the persistence of positive blood cultures [19]. Therefore, multiple sets of blood cultures should be drawn. Other laboratory tests may be helpful but are nonspecific. Echocardiography is an important tool in the diagnosis of endocarditis and should be considered in consultation with a pediatric cardiologist.

Broad coverage with empirical antibiotics is necessary, pending identification of the organism and its sensitivities. In cases of congestive heart failure due to valve dysfunction, valve surgery may be necessary.

Cardiac Allogaft (Transplant) Rejection

The incidence of rejection in children is 1.5 to 2.5 episodes per patient and is highest in children aged 1 to 4 years. In patients with heart transplants, rejection is the second leading cause of death after infection and accounts for 19% of deaths [20].

History and physical examination may be of limited utility because of the often nonspecific symptoms and signs of acute rejection. Symptoms such as fatigue, nausea, vomiting, diarrhea, and/or fever, can be vague and mimic acute gastroenteritis or other viral syndromes. Moderate to severe rejection may manifest with symptoms and signs of congestive heart failure. There may be a new heart murmur or gallop. Rarely, unexplained persistent tachycardia may be the sole sign of acute rejection.

Blood tests are nonspecific and may reveal leukocytosis as well as an increase in creatine phosphokinase and troponin levels. Chest x-ray may demonstrate cardiomegaly, pulmonary edema, or pleural effusion. The ECG may have decreased voltages, nonspecific ST and T-wave changes, or ventricular ectopy such as premature ventricular contractions. Echocardiography may show subtle signs of systolic or diastolic dysfunction; however, the gold standard for diagnosis is transcatheter endomyocardial biopsy with histologic staging.

Suspected episodes of acute rejection should be managed aggressively, in consultation with a pediatric transplant cardiologist. Admission to the hospital is frequently necessary. The use of pulse dose steroids as well as biologic immunosuppressives, such as OKT-3 or antithymocyte globulin, should be considered. Infectious complications are similar to those experienced by patients receiving immunosuppressive regimens for other conditions but are beyond the scope of this review.

Summary

Management of postoperative cardiac surgery patients can be quite complex and challenging. However, under-

standing the common cardiac lesions, surgical interventions, and complications that may be encountered in the ED should result in timely and effective intervention and minimize morbidity and mortality in this sometimes fragile patient population.

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