Care of Children Who Have Had Surgery for Congenital Heart Disease

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Children who have had surgical correction for congenital heart disease can present to the ED with an acute illness that could be associated with their cardiac lesion. There is no data available to summarize complications that could be associated with surgically corrected congenital heart disease. This work was undertaken to describe the common procedures used, list known complications of these procedures, and review general management principles in caring for the acutely ill child who has had heart surgery. (Am J Emerg Med 2003;21:318-327. © 2003 Elsevier Inc. All rights reserved.)

The major texts in EM[1,2] and pediatric EM[3,4] center discussions of congenital heart disease around diagnosis and acute management of patients with suspected cardiac lesions. These texts focus on relevant anatomy, presentation, and stabilizing therapy. There is little literature for the EP focusing on the long-term outcome and complications seen in patients who have had palliative, temporizing, or definitive procedures. Most pediatric cardiology textbooks[5,6] and literature focus on immediate complications of the presently performed procedure. This body of literature has typically been directed at cardiologists, intensivists, and surgeons. There is little literature to assist the EP in understanding the late postsurgical complications that can develop subacutely or acutely. This review was undertaken to provide a description of the basic details of surgical repairs (definitive or palliative) and to describe postoperative complications that can present after discharge from the hospital. Our goal is to give EPs a better understanding of anatomy and a list of general principles to use when evaluating these patients. Throughout this work, the figures are drawn from standard schematic methods used by pediatric cardiologists. Hopefully, allowing the reader to become familiar with these drawing styles will allow easier interpretation of patient records.

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PROCEDURES

The following are descriptions of procedures that can be performed on children with a variety of heart lesions. In this section, the procedures are described, the indications for such procedures are listed, and known late complications that can cause acute symptoms are included.

Shunt Procedures

Most surgical procedures listed in this review are designed to reestablish "normal" circulation patterns. Shunt procedures, on the other hand, seek to increase mixing or further complicate flow patterns as a temporizing procedure to allow for a delayed definitive procedure. Ideally, this delay until a definitive procedure is performed will allow the child to grow, minimize technical difficulty, and retard the onset of pulmonary hypertension, which will adversely affect long-term survival. The 3 main types of aorta-to-pulmonary artery shunts are Potts, Waterston, and central shunts. The Potts anastomosis is between the descending aorta and the left pulmonary artery (Fig 1). The Waterston procedure is an anastomosis between the ascending aorta and the right or main pulmonary artery (Fig 1). The central shunt uses a graft from the ascending aorta to the pulmonary artery (Fig 2).

The most common shunt performed is a Blalock-Taussig (B-T) shunt or an anastomosis between a subclavian artery and an ipsilateral pulmonary artery. In the classic B-T shunt, the subclavian artery is sacrificed for this procedure. The modified B-T shunt, the most common technique used presently, is performed by using a Gore-tex conduit between the subclavian and pulmonary artery. This procedure spares the subclavian artery (Fig 3). The B-T shunt is used most commonly to reestablish pulmonary blood flow in children with atretic or stenotic pulmonary arteries and inadequate pulmonary blood flow. Pulmonary artery size at birth is determined by in utero pulmonary blood flow. Lesions impeding pulmonary blood flow result in inadequate-sized pulmonary arteries. Lesions in this category include tetralogy of Fallot, tricuspid atresia, pulmonary atresia, and right ventricular hypoplasia.[7]

Complications of B-T shunts can include injury to the phrenic or recurrent laryngeal nerves or injury to the sympathetic chain with resultant Horner syndrome. Currently, the ipsilateral vertebral artery is ligated during the modified B-T shunt to prevent a basilar steal phenomenon.[8] However,

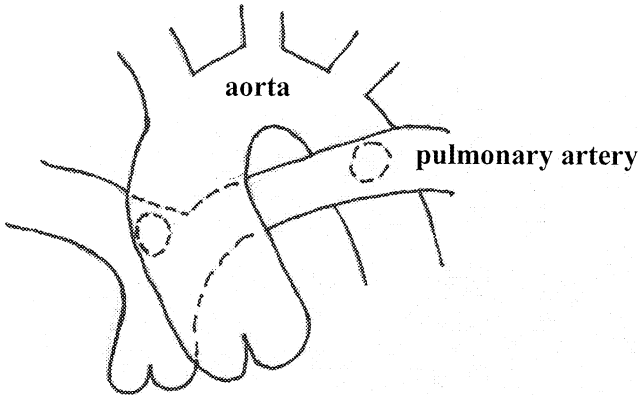

FIGURE 1. Example of Waterston shunt (ascending aorta to right pulmonary artery) and Potts shunt (descending aorta to left pulmonary artery).

older shunts could not have had this ligation. Long-term complications from vertebral artery ligation are unknown. Complications can also arise from the shunt size. If the shunt is too small, the child could have residual cyanosis from inadequate pulmonary blood flow. If the shunt is too large, pulmonary edema can result. Excessive flow complications after B-T shunts are usually limited by the size of the subclavian artery. However, excess pulmonary blood flow is the most common complication after modified B-T shunt, occurring in 28% of patients in one series.[7] Like with all procedures, stenosis at the anastomosis can occur.[9] Additionally, a thrombosis can develop in shunts formed with grafts. The shunt can clot when a child gets dehydrated as a complication of a viral illness, such as respiratory syncytial virus (RSV) or rotavirus. An uncommon complication recently noted is an aneurysm of the shunt that is diagnosed as an infiltrate on chest x-ray.[10]

The complications that affect shunt flow will most commonly present with worsening of the child's baseline hypoxia. The goal of the treating physician is to determine whether this change is the result of an increase or a decrease in pulmonary blood flow. Chest radiographs should be examined for subtle increases as well as subtle decreases in

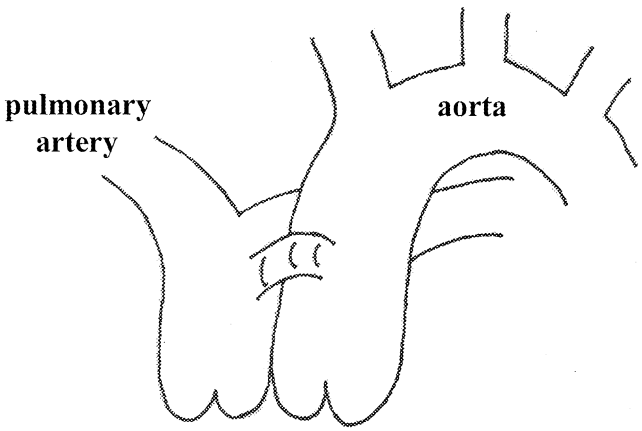

FIGURE 2. Central shunt (ascending aorta to Main pulmonary aorta).

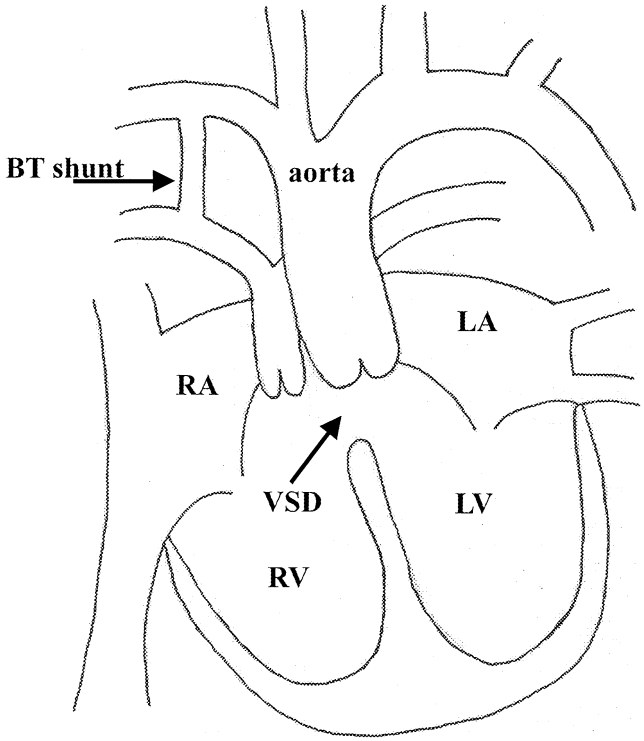

FIGURE 3. Blalock-Tausig shunt in patient with tetralogy of Fallot (right subclavian artery to right pulmonary artery).

pulmonary blood flow. Increases in liver span could suggest excessive shunt flow resulting in increased central venous pressure in a common atrium or increased end diastolic volume in a common ventricle. Uncommonly, the child with a shunt can present with an increase in room air oxygen saturations. This could be the result of a decrease in pulmonary blood flow (lessening degree of pulmonary edema) or an increase in pulmonary blood flow.

Complications of Potts, Waterston, and central shunts usually depend on the relative size of the shunt. As seen in patients with B-T shunts, too small a shunt could not relieve cyanosis, whereas too large a shunt could produce increased pulmonary blood flow, which can lead to pulmonary hypertension.[11,12] Potts and Waterston shunts are rarely used today because of frequent difficulties with pulmonary hypertension.

In summary, all of the shunts described here are placed as a temporizing procedure. Complications of these shunts, regardless of the location, are the same. The clinician should assess shunt function by performing a physical examination, including auscultating for a continuous murmur, measuring liver span, identifying any increase or decrease in pulse oximetry, and evaluating a chest radiograph for an increase or decrease in pulmonary vascularity.

Fontan

The Fontan procedure is designed to allow any available ventricular anatomy to pump blood systemically, whereas the pulmonary system is perfused directly through the vena cava. Typically, the superior vena cava drains directly into the right pulmonary artery and the inferior vena cava drains

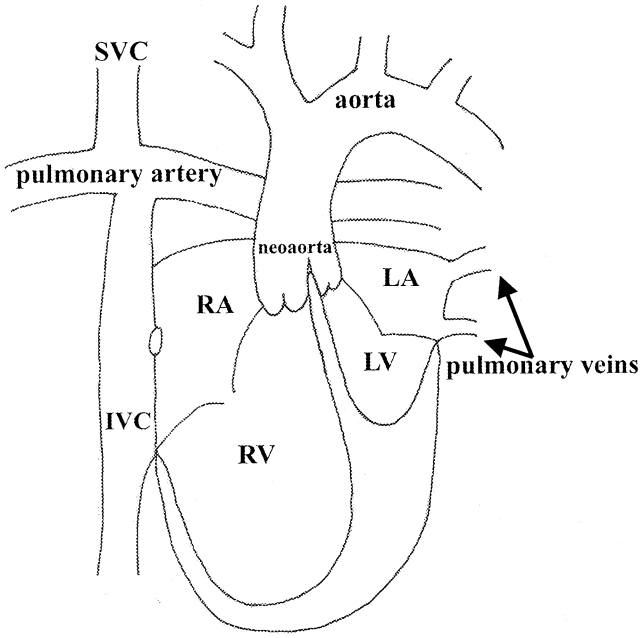

FIGURE 4. Fenestrated Fontan in hypoplastic left heart syndrome (also after Norwood-neoaorta and Blalock-Tausig shunt) (IVC and SVC right pulmonary artery, with fenestration into right atrium).

into the left pulmonary artery, either directly or through the right atrium. A Glenn shunt (superior vena cava to pulmonary artery) is performed as a palliative procedure or staging procedure until the final Fontan can be performed (Fig 4). Initial anatomy and right ventricular size can lead surgeons to perform some modification of the classic procedure, but the basic flow patterns and complications are the same for any variation. The Fontan is most commonly used when a child has single ventricle physiology. Anomalies of this type include tricuspid atresia, severe Ebstein's anomaly, hypoplastic left heart syndrome (HLHS), and some types of complex congenital heart disease.

Short-term complications of the Fontan include thromboembolism, thrombosis, and stenosis of the anastamosis.[13,14] Long-term complications can result from longstanding increased central venous pressure: fluid retention with ascites, atrial arrhythmias, pulmonary effusions, pericardial effusions, sudden death, and a protein-losing enteropathy with peripheral edema.[15,16] A cavopulmonary isolation procedure can decrease atrial arrhythmias and pulmonary effusions.[16] This isolation procedure involves direct anastomosis of the inferior vena cava to the right pulmonary artery through external conduits or lateral tunnels. A pacemaker could be required to control atrial arrhythmias[15,17] in some patients. Asymptomatic laboratory abnormalities can include a mildly hypercoagulable state and cholestasis,[16,18] although an activated coagulation system has not been implicated as a risk factor for thrombotic complications.[13] In that series, thrombotic complications were noted within 4 years of surgery in 25% of survivors.[13] Twenty-five percent of thrombi were in the systemic arterial circulation, whereas 75% were in the peripheral venous or pulmonary arterial circulation.[13] Stroke was noted in 1 of 107 patients reported less than 6 years out from their surgery.[19] Table 1 lists the complications associated with the Fontan repair.

SPECIFIC ANOMALIES

The following section includes a description of specific anomalies and the surgical techniques involved with the repair. Also, complications that can cause acute symptoms are included in each section.

Atrial Septal Defects

Surgical repair of an atrial septal defect is performed to prevent the development of arrhythmias and pulmonary hypertension with subsequent Eisenmenger physiology. An atrial septal defect should be considered in a child with a pulmonary flow murmur or a child with fixed splitting of the second heart sound. This defect may not be suspected until the child is school-aged. Repair is accomplished through a midline sternotomy with an incision into the superior vena cava or the right atrium. Septal closure is achieved through patching or suturing the defect. Complications from the surgical procedure can arise if flow from the vena cava or pulmonary vein is obstructed by the repair, or if the sinoatrial (SA) or arteriovenous (AV) node is injured during repair. Early complications from this repair include obstruction of the inflow from the vena cava or pulmonary veins or arrhythmias resulting from injury to the SA node during repair. Venous obstruction can lead to either hepatic congestion or pulmonary edema. Atrial arrhythmias are the most common delayed-onset complication of this procedure. Atrial fibrillation and flutter are more common than bradyarrhythmias. These arrhythmias are more common in patients who are older at the time of their repairs.[20-22]

Ventricular Septal Defects

Unrepaired ventricular septal defects allow mixing of blood at the ventricular level. Initially, shunting is from left to right, resulting in increased pulmonary blood flow. If this persists for several years, pulmonary hypertension can develop and right-to-left shunting (Eisenmenger's physiology) can result. Therefore, young children with this lesion will become desaturated only if they experience pulmonary edema from the excessive pulmonary blood flow. Older children can be cyanotic in the face of Eisenmenger's physiology. The severity of the lesion could not be able to be determined by the type of murmur present. Immediate closure is not imperative, because many small ventricular septal defects (VSDs) will close spontaneously. The goal of medical therapy is to control congestive heart failure; al-

TABLE 1. Complications Seen After the Fontan Procedure

Thromboembolism
Thrombosis
Increased central venous pressure
Ascites
Pulmonary effusion
Pericardial effusion
Protein-losing enteropathy
Atrial arrythmia
Sudden death

lowing the child to grow facilitates the surgical repair, because it is technically easier in a larger child. Children are referred for surgical closure when they fail medical therapy or the cardiology team thinks the lesion will not close spontaneously. Failure of medical therapy is often defined as uncontrolled congestive heart failure, poor weight gain in infancy, or development of pulmonary hypertension. Aortic insufficiency (secondary to prolapse of an aortic cusp into the defect) is another indication for surgical closure. The surgical approach depends on the type and size of the VSD. Defects can be accessed through the tricuspid valve, through the pulmonary artery, through the aortic valve, or through a right ventriculotomy.

Postsurgical risks include residual VSD, resultant aortic insufficiency, and electrocardiographic abnormalities. Electrocardiographic abnormalities can occur as a result of surgical disruption of the right conducting bundle during ventriculotomy or swelling near the AV node or His conducting system during patch placement. Residual complete atrioventricular block is uncommon after repair,[23] whereas right bundle branch block is common after a ventriculotomy. The most feared late complication of this repair is late-occurring fatal arrhythmia. In 1 series of patients experiencing sudden death after congenital heart disease, none of the patients had a corrected VSD.[24] However, another series followed patients with VSD repair and pulmonary hypertension. In that series, cases of sudden death did occur.[25] Therefore, the true incidence and predisposing factors for this complication cannot be clearly defined. Preexisting pulmonary hypertension can increase a patient's risk of late arrhythmia; otherwise, these patients do very well. Most survivors of VSD repair are asymptomatic or have mild exercise intolerance. In addition, those with no residual VSD do not require endocarditis prophylaxis.

Atrioventricular Septal Defects

Children with atrioventricular septal defects (AV canal, endocardial cushion defect) have a varied degree of clinical severity. Patients almost always have a murmur detected in the newborn nursery. The clinical spectrum can range from asymptomatic to severe congestive heart failure. Clinical presentation depends on the size of the defects and the amount of regurgitation from the AV valves. Additionally, the timing of surgery, the type of surgical repair, and the risks of surgery are related to the severity of the initial anatomy, especially if there is pulmonary hypertension from left-to-right shunting, pulmonary outflow tract obstruction (tetralogy of Fallot physiology), or complex congenital heart disease (eg, heterotaxy). Surgical techniques continually improve to treat this condition. The late complications generally depend on the severity of the presurgical clinical condition and presurgical anatomy. Atrial arrhythmias or heart failure can occur postoperatively if the patient has residual increased atrial pressures. These complications can also occur if the child has a residual ventricular septal defect, pulmonary hypertension, or residual AV valve regurgitation. Heart block requiring a pacemaker can occur postoperatively. When treating a child who has had this type of anatomy, it is important to inquire about any of the listed residual anatomic abnormalities that could predispose a child to these complications.

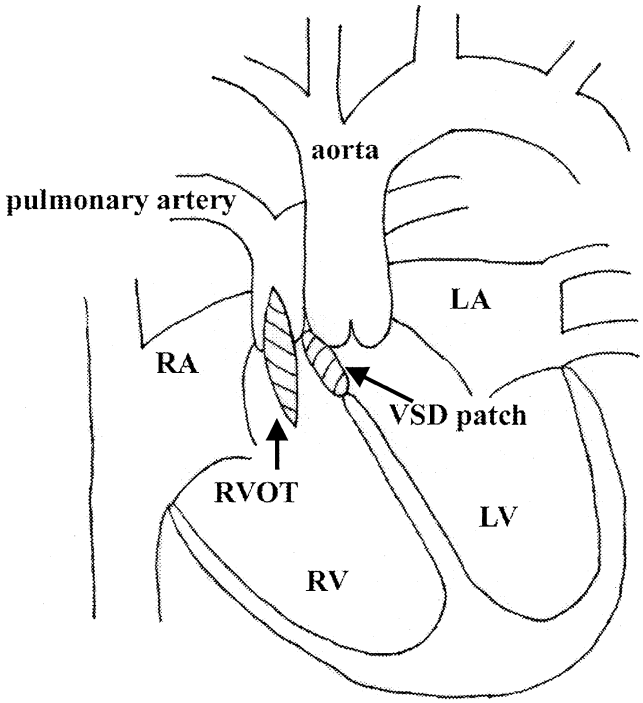

FIGURE 5. Corrected tetralogy of Fallot (VSD patch and right ventricular outflow tract patch).

Artificial AV valves, with the resultant expected complications, can be necessary if functional valves cannot be fashioned surgically.

Tetralogy of Fallot

Tetralogy of Fallot is a spectrum of illness in which patients experience variable pulmonary blood flow and oxygen saturation, depending on right ventricular outflow tract anatomy. These children are also susceptible to hypercyanotic episodes or "tet spells." Although oxygen therapy for a hypoxic child is appropriate, children experiencing hypercyanotic episodes do not respond briskly to oxygen therapy because the hypoxia is caused by an acute decrease in pulmonary blood flow.

Timing of repair and complication rates after repair depends on initial anatomy and patient size. Initial definitive repair is preferred over a temporizing shunt followed by definitive repair. Surgical approach and technique vary with patient size and lesion anatomy. In patients with a small degree of pulmonary stenosis, a transatrial repair is possible, eliminating the risks associated with ventriculotomy. Patients with more severe lesions can require ventriculotomy and a reconstructed pulmonary outflow tract in cases in which native pulmonary arteries are inadequate (Fig 5).

The most common complications after this procedure include congestive heart failure (right or left), right ventricular failure, or conduction abnormalities. Any of these complications can result in increased risk for arrhythmias. Congestive heart failure can result when there is a residual, uncorrected shunt. This can occur with a residual intracardiac shunt, a significant systemic to pulmonary collateral artery, or a residual systemic-to-pulmonary palliative shunt.

This complication should be noted postoperatively before discharge. Right ventricular dysfunction can occur as a result of persistent pulmonary outflow obstruction, pulmonary hypertension from a palliative procedure, pulmonary regurgitation through a conduit or dysfunctional pulmonary valve, or volume overload from a residual VSD. Acutely, if the child's right ventricle is preload-dependent as a result of increased afterload, the child can be less tolerant of dehydration. Conduction abnormalities and arrhythmias can occur after surgical correction of tetralogy of Fallot.[26] The bundle of His can be injured during repair leaving a residual right bundle branch block, bifascicular block, or complete AV block. A pacemaker is necessary in the case of complete AV block.[23] Additionally, patients with persistent premature ventricular contractions could need cardiology evaluation, especially if these are multifocal or provoked by exertion. Patients who have had a repaired tetralogy of Fallot are at increased risk for sudden death from arrhythmias.[24]

Transposition of the Great Vessels

The arterial switch procedure was not a viable technique until described by Jatene et al in 1976.[27] Before that, the only viable surgical options were the intraatrial baffle procedures described by Mustard and Senning. In cases of transposition of the great vessels with other congenital heart lesions, such as a VSD, the Rastelli procedure is performed. All of the procedures are described here.

Arterial Switch

Transposition of the great vessels is a congenital anomaly in which the aorta and coronary arteries arise from the anatomic right ventricle and the pulmonary artery arises from the anatomic left ventricle. Multiple surgical techniques have been attempted to manage this condition. Presently, the most commonly performed procedure is the arterial switch repair. In this, the aorta and coronary arteries are surgically attached to the left ventricle and the pulmonary artery is surgically attached to the right ventricle. One recent series suggests that another procedure will be required in over one fourth of patients.[28] True reintervention and complication rates will undoubtedly be higher, but this procedure has not been performed long enough to give long-term risks. Described long-term complications include sudden death, stenosis at the anastomosis in the pulmonary artery or aorta (supravalvular aortic or pulmonary stenosis), branch pulmonary artery stenosis, left ventricular failure, and caval vein thrombosis.[29] The most common complication requiring reintervention is pulmonary stenosis.[28] Because of the anatomic relationships of this condition, the repair includes moving the coronary arteries separately from the aorta. Immediately postoperative myocardial infarction can occur if the artery is injured during repair. Coronary artery stenosis can occur after arterial switch,[30] although the rates and types of risks to the coronary arteries are unknown.

Atrial Baffle Procedures

Before a viable arterial switch procedure, baffle techniques were the most effective technique to care for patients with transposition of the great vessels. The Mustard and Senning procedures were the most common of these baffle

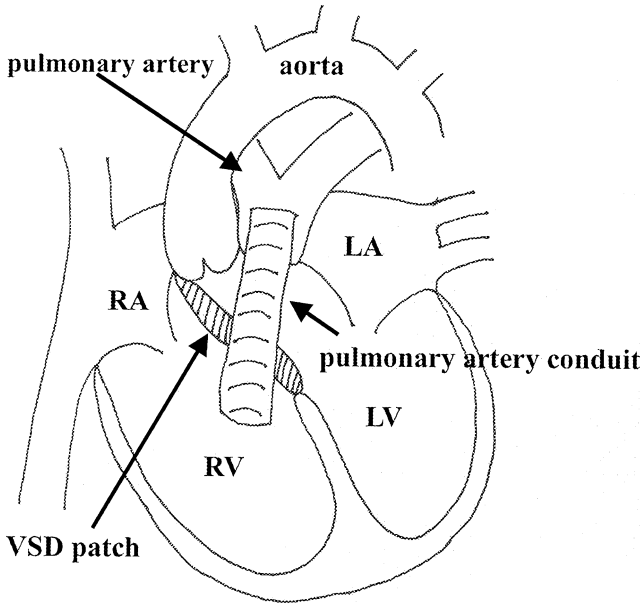

FIGURE 6. Transposition of the Great Arteries with Rastelli procedure (ventricular septal defect patch and right ventricle to pulmonary artery conduit).

procedures. With both of these procedures, intraatrial baffles redirect flow across an open atrial septum to the ventricle supplying the desired artery. After this procedure, the anatomic right ventricle supplies the work to pump blood to the aorta, whereas the anatomic left ventricle pumps blood to supply the pulmonary artery. Complications include syncope,[31] atrial arrhythmias, junctional rhythms, and baffle obstructions: stenosis or thrombosis of pulmonary or systemic venous drainage. Atrial arrhythmias include atrial fibrillation, atrial flutter, and sick-sinus syndrome. Atrial flutter has been noted as a cause of sudden death.[15] Baffle obstruction can manifest with superior or inferior vena cava syndrome, ascites, protein-losing enteropathy, or peripheral edema. The right ventricle seems to be able to sustain the systemic circulation into adulthood.[32] Some decreased exercise tolerance can be described by adult survivors of this procedure. The cause of this complaint is unclear and is likely multifactorial. Factors include a smaller increase in stroke volume and heart rate than in normal control subjects.[32] Other researchers have found a decreased forced vital capacity and decreased forced expiratory flow volume in 1 second compared with normal control subjects.[33]

Rastelli Procedure

The Rastelli procedure is used in cases of a complex transposition. Complex transpositions include, but are not limited to, those with a subvalvular pulmonary stenosis and ventricular septal defect. In this procedure, the anatomic left ventricle pumps blood to the aorta through a patch across the VSD. The main pulmonary artery is ligated and the anatomic right ventricle pumps to the pulmonary artery through a homograft or conduit (Fig 6). The complications of this procedure include sudden death, left ventricular

dysfunction,[34] pulmonary stenosis with right ventricular dysfunction and ventricular arrhythmias, and conduction abnormalities.

Coarctation Repair

There are 3 main surgical procedures for repairing coarctation of the aorta: resection of the coarctation shelf with an end-to-end anastomosis of the descending aorta, subclavian artery flap aortoplasty, and synthetic patch aortoplasty. All 3 of these repairs are subject to the same late complications. The most common complication is recoarctation. Patients can also have aneurysm formation, aortobronchial fistula, or arrhythmia, which can cause sudden death.

A recent series recounts the need for reoperation in a group of patients who have had repair.[35] Of 580 patients, 383 were available for follow-up. Twenty-three of these 383 (6%) required reoperation. Recoarctation (stenosis) was present in 9. The surgical indication was symptoms or gradient of 20 mm Hg. Aneurysmal dilation was noted in 8, massive hemoptysis from aortobronchial fistula in 2 patients, intracardiac stenosis was seen in 3 patients, and 1 person had a ruptured sinus of Valsalva aneurysm.[35] Survivors of coarctation repair are also at an increased risk of sudden death from arrhythmias.[24] The electrocardiography of survivors can have a persistent pattern of left ventricular hypertrophy, even without increased left ventricular pressure.

Restenosis, the most common of the complications, occurs in patients who had any of the 3 types of initial surgical corrections. Restenosis can occur at the suture line, like with the end-to-end anastomosis, or along the length of the patch, like with the subclavian flap. The resection with end-to-end anastomosis can have a restenosis rate of up to 60%.[36] The cause of aneurysms in this population is uncertain, but there is data to suggest that the microscopic wall of the remaining aorta is abnormal at the level of the coarctation, predisposing these patients to aneursysms.[37]

When caring for patients who had coarctation of the aorta, it is important to remember the risk of associated anomalies. Of the 103 patients described by McElhinney et al,[38] 89 (86%) had another cardiac anomaly and 14 (14%) had other noncardiac anomalies. The most common cardiac anomalies were bicuspid aortic valve (83 patients) and small VSD (19 patients). The most common noncardiac anomaly was Turner syndrome (6 patients). Among other anomalies associated with Turner syndrome, one fourth to one third can have renal malformations. Additionally, dilation of the aortic root can occur, especially in those patients with bicuspid aortic valves.

Hypoplastic Left Heart Syndrome

The surgical therapy for children with hypoplastic left heart syndrome is often referred to as a staged reconstruction or Norwood procedure. This procedure is a multistep repair used primarily for HLHS, but also complicated left-sided cardiac lesions. The first step of this procedure is to create a neoaorta using the pulmonary artery. This replaces the atretic aorta and allows perfusion of the systemic circulation and coronary arteries. The pulmonary arteries are disconnected and perfused through a modified Blalock-Taussig shunt (Fig 7).

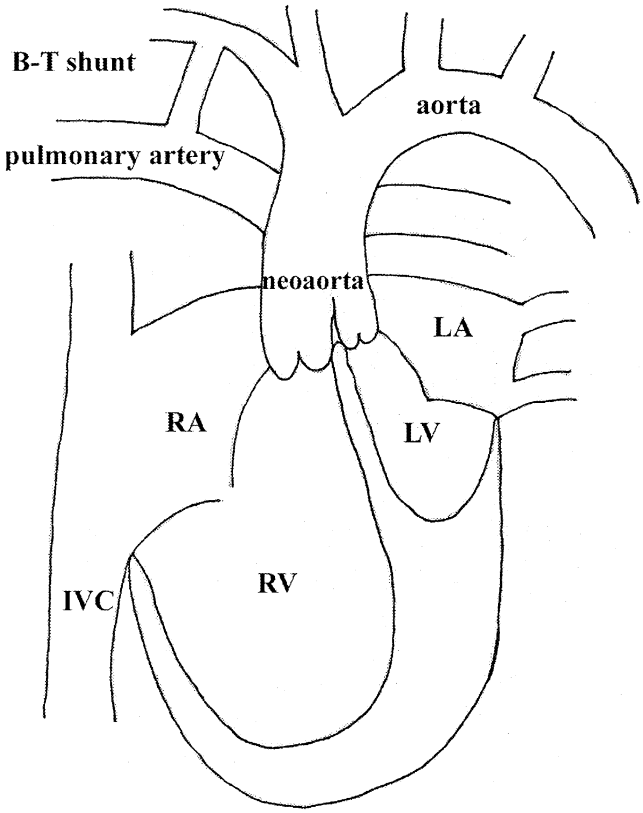

FIGURE 7. Hypoplastic left heart syndrome after Norwood procedure (creation of neoaorta, Blalock-Tausig shunt).

In the second stage, a Glenn shunt or hemi-Fontan is performed to perfuse the pulmonary circulation directly from the superior vena cava and the aortopulmonary shunt (typically modified B-T shunt) of step 1 is removed (Fig 8). The inferior vena cava and right atria are not yet isolated from the systemic perfusion. This second stage is performed at approximately 6 months of age. The superior vena cava is disconnected from the heart and anastomosed to the right pulmonary artery. Like with an isolated Fontan, noted complications include pleural effusions, pericardial effusions, phrenic nerve palsy, and transient superior vena cava syndrome.[39-42] The third and final stage, performed at approximately 18 months, involves completion of the Fontan. When complete, all systemic venous blood drains directly to the pulmonary system. Pulmonary venous blood enters the left atrium, crosses the open atrial septum into the right atrium, and enters the right ventricle and then exits through the constructed neoaorta. This technique is evolving, and long-term acute complications are unknown.

GENERAL PRINCIPLES OF MANAGEMENT

Following is a list of general principles to consider when evaluating a child who has had surgery for congenital heart disease.

Complex Lesions or Lesions Not Described Here

The repair of lesions not described here usually involves some combination of procedures and, therefore, complica-

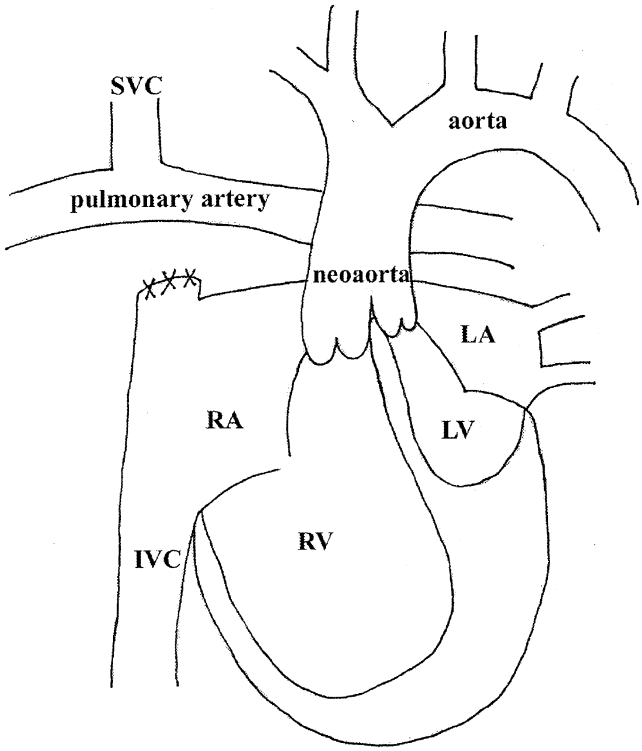

FIGURE 8. Hypoplastic left heart syndrome after Glenn shunt (SVC to right pulmonary artery) after Norwood procedure.

tions described in previous sections. As a general rule, the most common complication of any procedure is stenosis at the anastomosis. Therefore, if a child presents with this situation, their presentation should represent deterioration from baseline to presurgical physiology.

Evaluation of all children with corrected congenital heart disease with an acute deterioration should include a history and physical examination, oxygen saturation measurement, electrocardiogram, and a chest radiograph. The history of an infant in heart failure is one in which the child has not been feeding as well or becomes tachypneic and diaphoretic with feeding. Goals of the physical examination include identifying a change in murmur or liver span, and measuring pre- and postductal blood pressures. Oxygen saturations should include pre- and postductal measurements. The chest radiograph should be examined for changes (either increase or decrease) in heart size or pulmonary vascularity. Thoughtful evaluation of these results, combined with knowledge of the child's anatomy, should reveal if there is a cardiac cause for the patient's presenting condition.

Hypoxia

Worsening of baseline hypoxia is usually the result of a change in pulmonary blood flow. Flow can be decreased or increased. In children with significant mixing, such as a child who has had a palliative shunt awaiting definitive repair, hypoxia that responds dramatically to oxygen is not typically the result of changes in shunt flow or percent of cardiac output to the lungs. The diagnosis of the change in pulmonary blood flow can be suggested by increased or decreased pulmonary vascularity on a chest radiograph that is compared with a previous study.

Worsening hypoxia can also occur in children with tetralogy of Fallot. Tet spells, or paroxysmal hypoxic episodes or hypercyanotic episodes, manifest with abrupt onset of worsening hypoxia. Most commonly this will occur when there is a decrease in peripheral vascular resistance, like after exercise when a child has peripheral vasodilation or after a nap when a child has lower sympathetic tone. The children have an acute increase in the right to left shunt with a resulting decrease in pulmonary blood flow. In response to the decreased oxygen tension in the blood, there is further diffuse systemic vasodilation. This decrease in systemic vascular resistance further worsens the right to left shunt. Many tet spell are self-limited. Some require intervention. Treatment of tet spells require placing the child in the knee-to-chest position. Ideally, this position would increase venous return and increase systemic vascular resistance, which would decrease the right-to-left shunt and increase pulmonary blood flow. An intravenous fluid bolus can increase pulmonary blood flow. Supplemental oxygen should be applied. This should be discontinued if it further aggravates the child and worsens the clinical condition, because supplemental oxygen usually does not substantially increase the oxygen content of the arterial blood. Subcutaneous morphine injections have been used, although the exact mechanism of its effectiveness is unclear.

In cases of refractory spells, authors suggest propranolol or esmolol.[43,44] Propanol dosing has been suggested at a maximum of 0.1 mg/kg (to a maximum dose of 1 mg). Sodium bicarbonate (1 mEq/kg) can be effective.[43] Authors have suggested compression of the abdominal aorta through external compression of the abdomen as therapy.[45] Disappearance of the femoral pulse marks effective abdominal compression. Intravenous phenylephrine (2 to 5 μg/kg/min) has been used in refractory cases.[46]

ASSOCIATED ANOMALIES

Congenital heart lesions are common in children with common malformation syndromes. It is imperative to inquire about other anomalies when managing these patients. In children with Down syndrome, 40% to 50% will have congenital heart disease. These children can have other significant anomalies such as a small trachea, atlantoaxial subluxation, and macroglossia. Of children with Turner syndrome, 25% can have congenital heart disease. These children can also have renal anomalies. Some authors have suggested a high rate of airway anomalies in children with congenital heart disease[47,48]; however, the true rate of difficult airways is unclear in this population.

RESUSCITATION

In patients who have preexisting cyanosis, there are 3 principles to remember when intubating children.[49] First, cyanotic infants who may have a hypercyanotic spell with intervention (intravenous access, intubation, and so on) can benefit from premedication with a small dose of a narcotic. This group of children could include those with unrepaired tetralogy of Fallot or any child who has any desaturation or bradycardia with any stimulation in the ED. Second, posi-

tive pressure ventilation in cyanotic infants can further decrease pulmonary blood flow. Pulmonary blood flow in children with a shunt depends on the ratio of pulmonary vascular resistance to systemic vascular resistance. Positive pressure ventilation will increase pulmonary vascular resistance, upsetting this delicate balance.

Third, medication choices during urgent intubation can also affect shunt flow. Unfortunately, no literature is available that describes the hemodynamic effects of agents in children with cardiopulmonary instability. However, there is catheterization laboratory data for several agents. It is generally accepted that narcotics and benzodiazepines do not decrease systemic vascular resistance or myocardial contractility. Propofol can decrease systemic vascular resistance and can decrease cardiac contractility.[50,51] A sudden decrease in systemic vascular resistance will decrease pulmonary flow in cyanotic children with congenital heart disease in which pulmonary blood flow is through a shunt. Ketamine has been controversial. Some authors suggest that ketamine can increase pulmonary vascular resistance, decreasing shunt flow.[49] Others have found that ketamine caused no clinically significant decrease in pulmonary vascular resistance in children with congenital heart disease undergoing cardiac catheterization.[51-55] In addition, a small percent of patients will have an increase in systemic vascular resistance with ketamine, which could improve shunt (pulmonary) flow.[54]

There is no data on the effectiveness of cardiopulmonary resuscitation with chest compressions in children with anomalous cardiac anatomy. In patients with normal hearts, it is standard teaching that chest compressions will generate approximately 30% of baseline cardiac output. Systemic oxygenation will be minimal in a situation in which prearrest blood flow is only a fraction of cardiac output.

ENDOCARDITIS

Children with congenital heart disease are at increased risk for endocarditis. Endocarditis is uncommon in patients with surgically corrected septal defects or right-sided lesions.[56] Patients at highest risk are those having surgical manipulation of the aortic valve or aorta. These cases of endocarditis do not always present with classic predisposing causes. In a report detailing cases of endocarditis in patients with congenital heart disease, a predisposing event could be determined in only 87 of 214 cases.[57] Dental procedures accounted for 42 of the 87, whereas skin infections accounted for 10 of the 87. Unfortunately, patients received appropriate endocarditis prophylaxis in half of the cases of endocarditis after dental procedures.

The series by Li and Somerville[57] does not list any cases of endocarditis following traumatic injury. Current recommendations[58,59] do not suggest endocarditis prophylaxis for cases of traumatic injury, whether minor laceration or severe tissue injury. In the review by Cetta et al,[60] no cases of endocarditis occurred after tattooing, although only 6% of patients took prophylaxis before the procedure. One author[59] recommends endocarditis prophylaxis for urethral catheterization in the face of a urinary tract infection. However, other authors[58,59] do not recommend prophylaxis for urinary catheterization without infection or for endotracheal intubation.

TABLE 2. Corrected Congenital Heart Disease Lesions Associated With Sudden Death

High risk
Tetralogy of Fallot
Aortic stenosis
Transposition of the great arteries
Coarctation of the aorta
Atrioventricular septal defect
Pulmonary stenosis
Anomalies that undergo the Fontan procedure
Low-risk
Ventricular septal defect

Endocarditis prophylaxis is indicated for all patients with corrected congenital heart disease, except for the following cases: patients who are 6 months out from a surgical repair of a VSD or PDA, with no residual defect, do not require endocarditis prophylaxis; and patients with an isolated ASD do not require prophylaxis before repair or 6 months after a complete repair has been performed.[59,61]

The classic symptoms of Roth spots, Osler's nodes, Janeway lesions, and splinter hemorrhages are all uncommon. The most common symptoms are fever, petechiae, malaise, embolic events, and a new or changing murmur. Other nonfocal symptoms can occur, including splenomegaly, heart failure, gastrointestinal symptoms, and arthralgias. Fever is usually low grade. The enlarged spleen is usually nontender. Arthralgias usually involve the large joints. Additionally, laboratory findings are not very sensitive. Possible laboratory abnormalities include elevated erythrocyte sedimentation rate, anemia, and hematuria.

SYNCOPE

In addition to the usual causes of syncope in the pediatric patient, patients with corrected congenital heart disease are often at risk for pathologic causes of syncope. Corrected lesions that are at risk for sudden death are listed in Table 2. In addition, patients with a Blalock-Taussig shunt can be at risk for syncope from a basilar steal phenomenon if the vertebral artery was not ligated. Patients who have had a Fontan, including repair of HLHS, are at risk for embolic phenomenon. In addition, there is a report of a child having recurrent syncope in which the cause was found to be a residual shunt across a presumed repaired total anomalous pulmonary venous return.[62] This shunt was expected because the boy had a drop in transcutaneous pO_2 during exertion.

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