Anesthetic Management of Adults with Congenital Heart Disease

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Abstract

Congenital heart defects constitute the most common type of birth defects and occur in approximately 8 in 1000 live births. With the exclusion of bicuspid aortic valve disease, the majority of untreated patients born with congenital heart disease (CHD) expire in childhood with approximately 15–25% surviving into adulthood. Significant advances in prenatal diagnosis, interventional cardiology techniques, congenital heart surgery, anesthesiology, and critical care have allowed approximately 90% of these patients to survive into adulthood. The profile of patients with congenital heart disease has evolved, and now there are estimates to suggest that in the United States, there are more adults than children living with congenital heart disease. Most of these patients will require additional interventional cardiology or cardiac surgical procedures either palliative or curative during adulthood. Although major studies evaluating this population have not been done, adults with CHD are a medically fragile group, which have an increased risk for perioperative morbidity and mortality. Formal guidelines which direct the management of these patients have not been developed; however, the American College of Cardiology recommends that adult patients with moderate to severe CHD be referred to a center specializing in the care of these patients and receive expert consultation with the appropriate care providers such as adult congenital heart cardiologists, surgeons, and anesthesiologists prior to undergoing procedures.

Keywords

Congenital heart disease · Right ventricle
Patent ductus arteriosus · Ventricular septal defect · Bicuspid aortic valve

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Congenital heart defects constitute the most common type of birth defects and occur in approximately 8 in 1000 live births. With the exclusion of bicuspid aortic valve disease, the majority of untreated patients born with congenital heart disease (CHD) expire in childhood with approximately 15–25% surviving into adulthood. Significant advances in prenatal diagnosis, inter-

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ventional cardiology techniques, congenital heart surgery, anesthesiology, and critical care have allowed approximately 90% of these patients to survive into adulthood Marelli et al. (2007). The profile of patients with congenital heart disease has evolved, and now there are estimates to suggest that in the United States, there are more adults than children living with congenital heart disease. Most of these patients will require additional interventional cardiology or cardiac surgical procedures either palliative or curative during adulthood. Although major studies evaluating this population have not been done, adults with CHD are a medically fragile group, which have an increased risk for perioperative morbidity and mortality. Formal guidelines which direct the management of these patients have not been developed; however, the American College of Cardiology recommends that adult patients with moderate to severe CHD be referred to a center specializing in the care of these patients and receive expert consultation with the appropriate care providers such as adult congenital heart cardiologists, surgeons, and anesthesiologists prior to undergoing procedures.

When caring for adult patients with CHD undergoing procedures requiring anesthesia, it is imperative that the providers appreciate the anatomy and physiology which may be relatively unique to each patient. A team approach is required, and the providers should communicate regarding the risks of the procedure, the procedural techniques anticipated, and the ability or lack thereof to compensate for any hemodynamic or physiologic perturbations such as hypovolemia/hypervolemia, anemia, and hypotension.

Epidemiology of Adult Congenital Heart Disease

Approximately 25% of adult patients with structural heart disease have not undergone repair of their lesion because they were not considered to have hemodynamic compromise worthy of intervention. Other patients may be diagnosed in adulthood when seeking medical attention for another condition such as pregnancy. Another subset of adult patients with unrepaired CHD have incurred severe physiologic perturbations as sequelae of their CHD and are no longer candidates for repair of their structural heart lesions. Of the patients with mild unrepaired CHD, the most common lesions include mild aortic valve stenosis secondary to a bicuspid aortic valve, atrial septal defects, small restrictive ventricular septal defects, mild pulmonary valve stenosis, mitral valve prolapse, and isolated congenitally corrected transposition of the great arteries. The majority of patients evaluated in the outpatient setting have previously undergone surgical or interventional catheter-based procedures. Adults who have undergone repair of what are considered uncomplicated lesions such as atrial septal defects (ASDs) or patent ductus arteriosus (PDA) may be indiscernible from unaffected patients. These same lesions if left unrepaired may result in pulmonary vascular disease/pulmonary hypertension or Eisenmenger syndrome (Baum & Perloff 1993). Patients with pulmonary vascular disease/pulmonary hypertension or Eisenmenger syndrome especially with cyanosis constitute an extremely high-risk population.

Cyanotic heart disease includes structural heart defects that result in a decrease in pulmonary blood flow or result in mixing of oxygenated and deoxygenated blood. These conditions lead to reduced blood oxygen content and cyanosis. The majority of patients with adult cyanotic congenital heart disease will have had previous surgical interventions as children. The most frequently encountered cyanotic defects managed in the adult congenital heart patient population include tetralogy of Fallot, D-transposition, single-ventricle anatomy, truncus arteriosus, total anomalous pulmonary venous return, and double outlet right ventricle (Table 1).

Table 1 Most commonly encountered adult congenital heart lesions

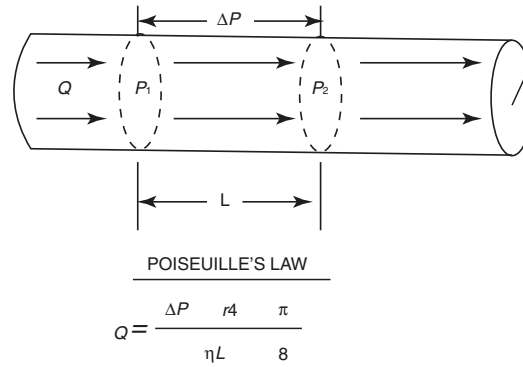
Tetralogy of Fallot, truncus arteriosus, and double outlet right ventricle with conotruncal anomalies postrepair
Secundum atrial septal defect
Ventricular septal defect
Complex single ventricles post-Fontan procedure
Coarctation of the aorta postrepair
Congenital aortic valve stenosis
Atrioventricular canal defects postrepair
Congenitally corrected transposition of the great arteries
Sinus venosus atrial septal defects with partial anomalous pulmonary venous return
Transposition of the great arteries after atrial or arterial switch

Major Categories of Congenital Cardiac Structural Defects

Adult congenital heart disease (ACHD) is composed of a variety of structural anomalies with hemodynamic consequences, which result from both the native anatomy and compensatory physiology. Due to the multitude of structural malformations, including complex forms with combined lesions, a classification system based on physiologic effects is the most useful. *The four categories, which are most commonly used, include shunt lesions, mixing lesions, obstructive lesions, and regurgitant lesions.* Over time, a compensatory physiology will evolve and produce aberrant loading conditions and potential impairment in ventricular function. In some patients, the compensatory changes are imperceptible and may cause little impairment in functions, while other lesions or their combined anatomy may cause significant cardiovascular compromise.

Shunt Lesions

Shunt lesions result in an aberrant communication between cardiac structures. The direction and volume of blood flow depends on the diameter and length of the communication and its relative position in the heart. The shunt lesion can occur at the level of the atria, ventricles, and great vessels or even be extra-cardiac. The flow of blood across the shunt or a vascular structure is determined by a number of factors, including the pressure drop across the structure, length, radius, resistance, and fluid viscosity. Poiseuille's law describes this relationship in the following equation:



where Q = blood flow, ΔP = pressure change in the fluid as it flows across the shunt, r = radius of the structure, L = length of the structure, and \ (n\) = the viscosity of blood. From this equation, it is seen that blood flow increases dramatically with increases in radius, increases to a lesser degree with increase in the pressure gradient across the structure, and decreases with an increase in length of the structure and blood viscosity. With a decrease in radius, a consequential decrease in blood flow to the fourth power is observed. Ohm's law describes the change in pressure, blood flow, and vascular resistance by the following equation:

$$Q = \Delta P / R$$

In this equation, P = the pressure generated within the cardiac chamber, Q = flow, and R = vascular resistance.

The direction and magnitude of shunt defects at the ventricular level and at the level of the great vessels, for example, a PDA, are determined by the radius of the defect and the difference in pulmonary and systemic vasculature resistance. For shunts at the level of the atrium, the ventricular diastolic pressure must also be taken into account. For small shunts, which persist into adulthood,

the indication for repair is based on the potential risk of strokes from paradoxical emboli and the occurrence of bacterial endocarditis. Moderate to large shunt defects can lead to ventricular failure due to volume overload. Shunt lesions that result in chronic increases in pulmonary blood flow may result in Eisenmenger syndrome. Eisenmenger syndrome is an irreversible increase in pulmonary vascular resistance (PVR) that ultimately leads to pulmonary hypertension, right-sided heart failure, and right-to-left shunting. An anesthetic consideration regarding right-to-left shunting is that inhalation inductions may be delayed. However, these delays may be more marked with larger shunts and insoluble anesthetic gases.

Mixing Lesions

Mixing lesions occur when blood from the pulmonary and systemic circulations combines to create a mixture of oxygenated and deoxygenated blood. As with shunt lesions, mixing defects may occur at the level of the atria, ventricles, or great vessels. The ratio of pulmonary and systemic flow is again based on vascular resistance. Increased pulmonary blood flow results with elevated systemic vascular resistance (SVR). This flow pattern results in ventricular volume overload and pulmonary hypertension. Vascular medial hypertrophy of the pulmonary vessels occurs and results in increased PVR and pulmonary hypertension. In severe cases of pulmonary overcirculation, systemic malperfusion occurs with resulting metabolic acidosis, shock, and cardiovascular collapse. Conversely, elevated PVR results in increased systemic flow, decreased pulmonary flow, hypoxemia, and cyanosis. Depending on the underlying anatomy, most patients with mixing defects receive palliation or complete repair in the neonatal period. Patients with ductal-dependent lesions frequently require a mixed circulation before repair. D-Transposition of the great vessels is a defect in which a mixing lesion is required for survival and an atrial septal defect may need to be created by the Rashkind procedure (balloon atrial septostomy) to allow mixing at the atrial level and prevent a circulation in parallel. Depending on the degree of systemic hypoxemia secondary to mixing of saturated and desaturated blood, deficits in oxygen delivery may exist with resulting compensatory mechanisms, including erythrocytosis and polycythemia with resulting hyperviscosity. Patients with mixing lesions are also at risk for bacterial endocarditis and paradoxical emboli and stroke. Other considerations for patients with mixing lesions include the degree of biventricular dysfunction and loading conditions as well as the function of other organ systems Brickner et. al. (2000).

Obstructive Lesions

Obstructive or stenotic lesions impose a ventricular pressure load proximal to the defect that results in increased intracavitary pressure and concentric ventricular hypertrophy. Initially, the wall stress is preserved; however, over time the vascular supply becomes inadequate to support the hypertrophied ventricle. The ventricle becomes relatively ischemic and dilated with increased wall tension and decreased contractility. Chronic obstructive lesions also lead to decreased ventricular compliance and diastolic dysfunction, even in cases with preserved systolic function. Patients with obstructive lesions are extremely preload dependent for adequate diastolic filling and cardiac output. Lesions, which are severely obstructive, can present with inadequate cardiac output and perfusion of the pulmonary, coronary, cerebral, and peripheral tissue beds. Significant decreases in exercise tolerance and cardiovascular reserve may be seen in patients with obstructive lesions.

Regurgitant Lesions

In general, regurgitant lesions do not usually exist as primary defects. They more commonly develop as sequelae or as a result of intervention secondary to CHD. However, worsening valvular incompetence or regurgitation results in decreases in ventricular ejection volume and function. The

compensatory mechanisms to increase stroke volume cause ventricular dilation with increased wall tension and a propensity for ventricular failure. A significant number of interventional cardiac catheter-based procedures and even cardiac surgical interventions are performed to correct lesions such as pulmonary regurgitation after tetralogy of Fallot repair in the adult congenital heart population.

Acyanotic Congenital Heart Disease

Dextrocardia

Dextrocardia is usually described as the heart having a mirror-image location in the right hemithorax. Dextrocardia with situs inversus is a mirror-image location of the intrathoracic structures as well as the intra-abdominal structures. This cardiac anomaly causes the heart to lie in the right hemithorax with the liver and gallbladder under the left portion of the diaphragm. The stomach is located under the right portion of the diaphragm and the appendix is located in the left lower quadrant. Dextrocardia with situs inversus may be functionally normal. Isolated dextrocardia with normally related abdominal viscera frequently indicates the presence of CHD. In some patients, the stomach bubble may be indistinguishable from large bowel gas because the stomach is not in its usual position, either left or right, and the liver appears to be uniformly distributed across the right and left diaphragm. This suggests visceral heterotaxy or situs ambiguous, associated with either polysplenia or asplenia and a high incidence of congenital heart malformations. In these patients, the apex may point to the left, right, or anteriorly and is referred to as mesocardia or atrial isomerism.

Dextrocardia with situs inversus usually exists in otherwise structurally and functionally normal hearts. These patients will generally have their congenital malpositioning noted when they present for noncardiac-related medical attention. The risk of noncardiac surgery in patients with dextrocardia with situs inversus and otherwise structurally normal hearts is the same as for patients with situs solitus (normally positioned abdominal viscera) and normally positioned hearts. The importance of diagnosing this anomaly lies in the correct interpretation of symptoms related to the underlying anatomic location of the organ in question. For example, patients with dextrocardia with situs inversus will experience the pain of acute appendicitis in the left lower quadrant rather than the right lower quadrant. The diagnostic importance of recognizing this anomaly has obvious surgical implications.

Congenitally Corrected Transposition of the Great Vessels

In uncorrected D-transposition of the great vessels (D-TGV), the aorta arises from the anatomic right ventricle; in corrected L-transposition of the great vessels (L-TGV), the venae cavae drain into the right atrium, which empties into an anatomic left ventricle through a mitral valve. The anatomic left ventricle ejects into the pulmonary artery (PA). In this form of transposition, the atrioarterial connections are concordant, and therefore systemic venous return enters the lung for oxygenation, and pulmonary venous return enters the left atrium and passes into an anatomic right ventricle through a tricuspid valve. Saturated blood then enters the aorta from the anatomic right ventricle. Without another major structural anomaly such as a ventricular septal defect (VSD) with or without pulmonic stenosis, patients with L-TGA may be entirely asymptomatic. There may be people with L-TGA who remain undiagnosed. However, there are a number of known associations with L-TGA, including complete heart block, VSD, pulmonic stenosis, left-sided atrioventricular (AV) valve regurgitation, Ebstein's anomaly of the tricuspid valve, and other complex lesions. Complete heart block may result from the abnormal alignment of the ventricles and interventricular septum. This malalignment of the connection between the bundle of His and the AV node may worsen as the heart grows with a loss of continuity between the two structures and resulting third-degree or complete heart block. Eventually, these patients may

require a permanent pacemaker. With L-TGA, the morphologic right ventricle (systemic ventricle) is at risk of depressed function. There is also a risk of bacterial endocarditis secondary to left AV valve regurgitation. The long-term function of the anatomic right ventricle as the systemic ventricle remains controversial. By 45 years of age, 67% of adults with L-TGA with associated anomalies such as ASD or VSD have developed congestive heart failure (CHF). Even without associated anomalies, approximately 25% of patients with L-TGA will have developed CHF by 45 years of age. To decrease complications from L-TGA, the "double switch" procedure can be performed in which the venous return is transposed using an atrial switch procedure and either the ventricular or arterial connections are transposed via a Rastelli procedure or an arterial switch procedure. In caring for the patient with L-TGA undergoing noncardiac surgery, one must consider the possibilities for disturbances in AV conduction, the ventricular function, and the risk of endocarditis. A preoperative ECG should be obtained, with intraoperative and postoperative ECG monitoring performed. For patients with 2:1 heart block (second-degree block), it may be prudent to place a temporary transvenous or transesophageal pacemaker. If the ventricular function is impaired, arterial line and/or transesophageal echocardiographic monitoring may be warranted.

Congenitally Bicuspid Aortic Valve

A congenitally bicuspid aortic valve may result from abnormal differentiation of the primitive truncal valve, leaving two of the aortic valve cusps fused together, producing a bicuspid instead of tricuspid valve. The bicuspid aortic valve orifice is eccentric and opens in an abnormal fashion. The abnormal orifice produces varying degrees of obstruction, which results in stenosis and increased flow velocity with a systolic ejection murmur. The risk to the patient with a bicuspid aortic valve undergoing noncardiac surgery is determined by the degree of stenosis of the valve and the ventricular function. Bacterial endocarditis is also a risk factor and prophylactic antibiotics should be considered. For patients with severe bicuspid aortic stenosis, balloon valvuloplasty may substantially reduce the gradient across the valve and offer significant symptomatic relief. Balloon valvuloplasty is only an option if the valve is thin and mobile. Adult patients with severe fibrocalcific bicuspid aortic stenosis account for 50% of the surgically significant cases of aortic stenosis. These patients are similar to patients with acquired aortic stenosis with calcific disease of a trileaflet valve and incur similar surgical risks and anesthetic management. Calcific bicuspid aortic stenosis is even less amenable to balloon valvuloplasty than a calcified, stenotic trileaflet valve. However, in the age of transcatheter aortic valve replacement (TAVR), these patients may be candidates for a minimally invasive replacement which leaves them open in the future for a valve in a valve replacement with the deferral of open cardiac surgical intervention. As with all adults with congenital heart disease, it is important to consider the possibility of acquired coronary artery disease (CAD), and it is advisable to screen these patients for CAD. It needs to be determined whether angina is secondary to CAD or solely to the aortic stenosis with the increased oxygen demands of the hypertrophied left ventricle. For patients with bicuspid aortic stenosis, appropriate preoperative evaluation and optimization will help decrease the perioperative risk factors. Intraoperative management should be guided by the patient's functional status and the complexity of the surgical procedure. Conventional monitoring and the use of arterial line, pulmonary artery catheter (PAC), and transesophageal echocardiography may be useful in guiding perioperative therapy. Patients must be monitored for signs of ischemia and ventricular dysfunction. Also, acute decreases in SVR will not be compensated by an increase in stroke volume secondary to the fixed left ventricular outflow tract (LVOT) obstruction.

Volume losses and hypotension must be promptly treated. Volume replacement should be pursued cautiously to avoid pulmonary edema which may result depending on the functional sta-

tus of the left ventricle or systemic ventricle. Pharmacologic support of SVR and ventricular dysfunction may be necessary with inotropes. The requirement of inotropic support is best guided by the previously mentioned monitoring modalities.

Patients with bicuspid aortic stenosis are also at risk for aortic regurgitation secondary to the thickened, calcified leaflets that may become incompetent secondary to their immobile nature. The bicuspid aortic valve may also become incompetent secondary to poststenotic aortic root dilatation. Aortic regurgitation imposes a volume load on the already pressure-overloaded systemic ventricle, which may further impair ventricular function and increase the risk of bacterial endocarditis. In patients with impaired ventricular function, the mode of aortic valve replacement needs to be considered with either surgical replacement or TAVR.

Coarctation of the Aorta

Coarctation of the aorta occurs in 8–10% of all congenital heart defects. It is more common in males than females, with a ratio of 2:1. Patients with Turner's syndrome have a 30% incidence of coarctation. Coarctation is an obstructive lesion that may present early and acutely in the neonatal period as critical coarctation (approximately 9% of coarctation cases) or may be detected later in life as an incidental finding, manifesting as right upper extremity hypertension with diminished lower extremity pulses. The majority of coarctations are located close to the insertion of the ductus arteriosus into the aorta. These juxtaductal coarctations are thought in some way to be produced by contraction of the arterial wall musculature that occurs with constriction of the ductus early in the neonatal period. This may account for the acute presentation of systemic hypoperfusion in affected neonates. Other forms of coarctation are not as easily explained. Coarctation may have only mild gradients at rest, but the gradient will significantly increase with any hyperdynamic state. Unrelieved coarctation leads to upper extremity hypertension, and the consequence of sustained hypertension secondary to coarctation is that the arteries of the head and neck as well as the coronary arteries are exposed to an increased risk of developing atherosclerotic changes. The ventricular myocardium also hypertrophies, increasing the demands on the coronary arteries. The diminished pulsatility of the blood flow distal to the coarctation promotes renal secretion of renin and angiotensin, which may further promote hypertension. The most common sequelae are systolic hypertension, recurrent or residual coarctation, aortic aneurysm, aortic dissection, intracranial aneurysm, and intracranial hemorrhage secondary to intracranial aneurysm formation and sudden death. Due to the significant long-term sequelae and potential disastrous comorbidities, it is of the upmost importance to repair coarctation in a timely fashion. The longer the delay in treatment, the greater the risk of persistent hypertension after the repair. Additionally, the actual life expectancy is significantly reduced, the longer the lesion is left untreated. The overall survival after repair is 91% at 10 years, 84% at 20 years, and 76% at 30 years. For patients who have undergone surgical repair, 30% will have hypertension with a higher incidence than those repaired within the first years of life. If a coarctation is treated within the first few years of life and no significant gradient persists, the patients are expected to be able to lead essentially normal lives.

The surgical approaches to coarctation repair include resection of the stenotic area with direct end-to-end anastomosis, subclavian flap aortoplasty, or aortoplasty with a homograft or synthetic graft material. All of these techniques require the use of aortic cross clamping around the area of coarctation, with the surgical risks of paraplegia or paresis secondary to spinal cord ischemia. Residual obstruction or re-coarctation occurs in 6–33% of all patients. The manner in which residual obstruction or re-coarctation is treated is dictated by the severity and anatomic location of the stenosis. Minimally invasive, catheter-based techniques have become the standard of care and are frequently required more than once in a given patient. Stented graft placement and balloon angioplasty of the aorta are the most common therapy required after the initial surgical repair.

Patients with a history of coarctation both unrepaired and repaired also require the following issues to be considered perioperatively:

- The potential presence and functional status of a concomitant bicuspid aortic valve (as many as 85% of patients with coarctation will have a bicuspid aortic valve)
- The presence of hypertension
- Systemic ventricular function
- The potential for premature or concomitant CAD
- The need for lifelong bacterial endocarditis prophylaxis

In addition to a complete history and physical examination, an ECG, echocardiogram, and ventricular stress test will provide a preoperative assessment of myocardial function, ventricular hypertrophy, and potential for ischemia. The patient with coarctation at the highest risk undergoing a procedure is the older patient with coexisting stenotic or an incompetent bicuspid aortic valve, depressed systemic ventricular function, and acquired CAD. If the patient is to undergo a surgical procedure and it is not urgent, intervention including coarctation repair, aortic valve replacement, coronary artery bypass or angioplasty/stenting, and medical management of the hypertension and depressed ventricular function may be undertaken to decrease the operative risk and provide optimal patient management. In patients with sequelae secondary to coarctation including systemic hypertension and depressed ventricular function, an arterial line and PAC placement are recommended to guide perioperative management. Depending on the degree of ventricular function impairment, intraoperative transesophageal echocardiography (TEE) may be helpful.

Pulmonic Stenosis

Pulmonic stenosis (PS) is an obstructive lesion of the right ventricular outflow tract that produces a pressure overload situation and compensatory changes associated with such lesions. Pulmonic stenosis may be subvalvular, valvular, or supravalvular. PS may also occur near the bifurcation and involve the branch pulmonary arteries. PS may be an isolated lesion or occur as an associated lesion with other complex malformations (tetralogy of Fallot) or as part of a syndrome such as Williams syndrome or Rubella syndrome and others. Mild PS generally causes no major hemodynamic perturbations. However, if the stenosis is severe, right ventricular hypertrophy and dysfunction may ensue. Most importantly for the patient undergoing a procedure are the limitations of compensatory responses of the systemic circulation to critical hemodynamic changes such as hypovolemia. In the case of mild PS, the risk of endocarditis exists with little hemodynamic compromise. However, for patients with severe PS and with impaired right ventricular function, their hemodynamics may be significantly improved with balloon valvuloplasty if the lesion is amenable to intervention. For emergent procedures in patients with severe pulmonic stenosis, perioperative arterial line monitoring and central venous pressure monitoring may be beneficial. Intraoperative transesophageal echocardiography (TEE) should be used for patients with PS and right ventricular or biventricular dysfunction in which loading conditions and ventricular function monitoring are required but in whom a PAC is not an option.

Primary Pulmonary Hypertension and Pulmonary Vascular Disease

Primary pulmonary vascular disease or primary pulmonary hypertension (PPH) is characterized by a decrease in the cross-sectional area of the pulmonary vascular bed caused by pathologic changes in the vascular tissue. PPH is characterized by progressive, irreversible vascular changes similar to those seen in Eisenmenger syndrome but without intracardiac anomalies. PPH is extremely rare in pediatric patients and is primarily a condition of adulthood and is more prevalent in women. It has a poor prognosis and may progress to a cyanotic condition with biventricular heart failure. Pulmonary vascular disease results

from any process that produces prolonged elevation of PA pressure, such as large left-to-right shunts, obstructive airway disease, and chronic lung disease with hypoxia, and causes progressive medial hypertrophy of the pulmonary vasculature with an ultimate decrease in the cross-sectional area. The cause of PPH is not fully understood, but endothelial dysfunction of the pulmonary vascular bed may be an important factor. If severe pulmonary hypertension develops suddenly in the face of an unprepared or non-hypertrophied right ventricle (RV), right-sided heart failure will result. In patients with chronic pulmonary hypertension, gradual hypertrophy and dilatation of the RV develop and the RV pressure may ultimately exceed the systemic pressure.

A decrease in cardiac output may result from at least two mechanisms: (1) A volume and pressure overload of the RV impairs cardiac function, primarily by impaired coronary perfusion of the hypertrophied and dilated RV and decreased left ventricular (LV) function resulting from the dramatic leftward shift of the interventricular septum caused by increasing RV volume. The latter also alters LV structures and decreases LV compliance, resulting in an increase in both LV end-diastolic pressure and left atrial (LA) pressure. (2) The second mechanism may result if a sudden increase in PVR occurs with decreased pulmonary venous return to the LA and hypotension and circulatory shock results as a consequence. Pulmonary edema can occur with or without elevation of the LA pressure. Direct disruption of the walls of the small arterioles proximal to the hypoxic/constricted arterioles may be responsible similar to the mechanism which is proposed for high-altitude pulmonary edema.

Irrelevant of the cause, the clinical manifestations of pulmonary hypertension are similar when significant hypertension exists. The patient's history will often reveal dyspnea, fatigue, and syncope on exertion. A history of CHD or CHF in infancy is present in most cases of Eisenmenger syndrome. Some patients may have a history of angina and headache. Hemoptysis is a late and occasionally fatal manifestation. Cyanosis with or without clubbing may be present. The neck veins may be distended with signs of right-sided heart failure such as hepatomegaly and ankle edema. The ECG will likely show right-axis deviation secondary to RVH, and right atrial enlargement may manifest as arrhythmias occurring in the later stage.

PPH and other types of pulmonary vascular disease are difficult to treat and nearly impossible to reverse unless the etiology is eliminated. Measures to remove or treat the underlying cause include timely surgical correction of the congenital heart defects such as ventricular septal defects (VSDs) or patent ductus arteriosus (PDA) before obstructive anatomic changes occur in the pulmonary vessels. The anesthetic management of pulmonary hypertension is guided by the underlying cause. Strategies for the intraoperative management including cases requiring cardiopulmonary bypass (CPB) and non-bypass cases need individualized assessment. The incidence of symptomatic pulmonary hypertension after repair of an intracardiac lesion does not often justify the placement of pulmonary artery catheters (PACs); however, centers that are familiar with their use have low associated morbidity. The most common strategies to manage pulmonary hypertension include avoiding maneuvers which elicit vasoconstrictive responses, maintaining oxygenation to lower pulmonary vascular resistance (PVR), maintaining alkaline pH, minimizing tidal volumes or using spontaneous modes of ventilation, and the use of pulmonary vasodilators such as nitric oxide, inhaled prostacyclin (PGI_2), and, in the ICU, oral sildenafil. Nitric oxide is a direct pulmonary vasodilator with no significant systemic effect as it is rapidly metabolized by red blood cells. Nitric oxide is administered by inhalation with a special delivery system which can be added to a ventilator or anesthesia machine.

Noncardiac surgery or procedures for patients with PPH, PH, or PVD can be formidable. Fixed or elevated PVR may limit perioperative hemodynamic compensation. The potential for hypotension, RV or biventricular dysfunction, and hemodynamic instability is significant. An abrupt fall in systemic vascular resistance (SVR) may precipitate intense cyanosis. An intraoperative arterial line and PAC monitoring are recom-

mended. In patients in whom placing a PAC is not feasible or in whom the risk of arrhythmia, RA/RV perforation, or PA rupture is believed to be unacceptably high, a TEE may be important in accessing loading conditions, in evaluating biventricular function and the magnitude of right-to-left shunting and air embolism, and in estimating PA pressures from the tricuspid regurgitant jet. TEE must be performed and interpreted by a qualified practitioner in all settings. The perioperative assessment of volume status and necessity for circulatory with inotropes is critically important in the patient with PH. The use of regional anesthesia for patients with pulmonary hypertension is recommended if appropriate for a given procedure as long as the patient does not have associated sedation which incurs significant hypoventilation. In the case of a general anesthetic, appropriate hemodynamic monitoring, airway and ventilatory management along with an appropriate depth of anesthesia will decrease the risk of perioperative instability.

Left-to-Right Shunt Lesions

Patent Ductus Arteriosus

A patent ductus arteriosus (PDA) is a persistent communication between the left PA and the descending aorta, which is approximately 5–10 mm distal to the origin of the left subclavian artery. It is a normal fetal structure and generally undergoes functional closure in the early neonatal period and becomes the ligamentum arteriosum. If the PDA does not close, persistent shunting occurs between the aorta and the PA. A PDA occurs in 5–10% of all congenital heart lesions, excluding premature infants. It is more common in females than males, with a male-to-female ratio of 1:3. A PDA is an extremely common problem among premature infants as a component of persistent fetal circulation and may require interventional catheter-based closure or surgical ligation if it results in respiratory failure or CHF. Patients are generally asymptomatic if the ductus is small. However, a large shunt may predispose patients to lower respiratory tract infections, atelectasis, pulmonary hypertension, bacterial endocarditis, and CHF. Adult patients with PDAs may have developed pulmonary hypertension and pulmonary vascular disease with right-to-left shunting and cyanosis. Paradoxical emboli and stroke are also significant risks for patients with PDA. Therefore, the existence of a PDA is an indication for closure either by transcatheter approach or surgical ligation. The presence of severe pulmonary vascular disease and PH may be a contraindication for closure.

Atrial Septal Defects

Atrial septal defects (ASDs) or ostium secundum ASD occurs as an isolated anomaly in 5–10% of all CHD. It is more common in females than males with a male-to-female ratio of 1:2. Thirty to fifty percent of pediatric patients with CHD have an ASD as an associated defect. Three types of ASDs exist, including secundum, primum, and venosus defects. A patent foramen ovale (PFO) in ordinary circumstances does not cause intracardiac shunting although in certain conditions such as high right-sided heart pressures it may. Ostium secundum defects are the most common type of ASD, accounting for 50–70% of all ASDs. This defect allows for left-to-right shunting of blood from the LA to the RA. Primum defects occur in approximately 30% of all ASDs, including those existing as complete endocardial cushion defects. Isolated primum defects occur in approximately 15% of ASDs. Sinus venosus defects occur in about 10% of all ASDs and are most commonly located at the entry of the superior vena cava (SVC) into the RA. The right pulmonary veins may anomalously drain into the RA and rarely at the entry site of the inferior vena cava (IVC). Spontaneous closure occurs before 18 months of age in more than 80% of patients with defects less than 8 mm. An ASD with a diameter greater than 8 mm rarely closes spontaneously. If a defect is left untreated, CHF and pulmonary hypertension may develop in adults in their 20s and 30s. Patients are at risk for atrial arrhythmias such as atrial fibrillation or flutter, paradoxical emboli and stroke, and rarely bacterial endocarditis.

Device or surgical closure is indicated for patients with left-to-right shunts with a (Q_p/Q_S) of more than 1.5:1. Some physicians consider a smaller shunt to be an indication for intervention because of a risk for paradoxical emboli and stroke. Severe pulmonary vascular disease or high PVR is considered a contraindication for closure. An isolated secundum ASD occurring in an otherwise healthy young adult without pulmonary vascular disease/pulmonary hypertension incurs little additional risk during noncardiac surgery. However, there are two conditions in which difficulties could arise: paradoxical embolization of air or other materials which could stream across the ASD into the system and potentially cerebral circulation. Also, in response to hemorrhage or hypovolemia, there is a compensatory increase in SVR with the decrease in venous return; this situation could worsen a left-to-right shunt.

With time, the large left-to-right shunt decreases pulmonary compliance and increases the work of breathing. The right side of the heart becomes enlarged with both diastolic and systolic dysfunction developing. The LV becomes less distensible and this may worsen the left-to-right shunt. In the fourth decade of life, there is an increase in the incidence in atrial arrhythmias especially atrial fibrillation, atrial flutter, and supraventricular tachycardia. A baseline ECG should be obtained in patients with a history of ASD. The majority of adults older than 35 years of age will frequently be symptomatic from a secundum ASD with at least moderate PH or PVD and RV dysfunction as a result of the chronic left-to-right shunt. For adult patients undergoing cardiac surgery for repair of an ASD, an arterial line, central venous line, and TEE monitoring are recommended. For patients undergoing device closure of their ASD, sedation with intracardiac echo may be recommended or general anesthesia with TEE monitoring may also be undertaken. An arterial line and central line are generally not necessary for device closure. For patients undergoing noncardiac surgery, the monitoring required is generally dictated by the severity of their pulmonary hypertension. For patients with severe PH undergoing a major surgical operation, an arterial line, central venous pressure monitoring, and TEE are recommended. TEE in this situation would be valuable in accessing loading conditions, biventricular function, and the presence and magnitude of shunting. PACs are generally not recommended due to the propensity to cross the atrial septal defect into the left heart. Also, patients with ASDs frequently have valvular regurgitant lesions of the tricuspid, pulmonary, and mitral valve and require bacterial endocarditis prophylaxis.

Ventricular Septal Defects

Ventricular septal defects (VSDs) are the most common form of congenital heart disease and account for 15–20% of congenital heart defects, not including those VSDs which coexist with cyanotic heart lesions. Perimembranous defects are the most common defects accounting for 70% of VSDs. With a small restrictive defects, patients may remain relatively asymptomatic until early adulthood. With moderate to large unrestrictive defects, patients will have delayed growth and development and decreased exercise tolerance, recurrent pulmonary infections, and CHF. Spontaneous closure occurs in 30–40% of patients with membranous VSDs during the first 6 months of life. CHF develops in infants with large unrestrictive VSDs at approximately the first 6–8 weeks of life. Pulmonary vascular occlusive disease may begin to develop as early as 6–12 months of life in patients with unrestrictive defects, but the resulting right-to-left shunt or Eisenmenger syndrome usually does not develop until the teenage years. As mentioned earlier, VSD is the most common form of CHD in childhood. However, certain unrepaired forms of CHD are rarely found in adulthood and VSDs are among them. There are occasional instances when patients with small to moderate-sized, restrictive VSDs present in adulthood.

The repair of VSDs in the adult population may be undertaken in an open surgical fashion or more commonly with a device closure. The degree of pulmonary hypertension or pulmonary vascular occlusive disease will dictate whether the clo-

sure can be performed. For noncardiac surgery the risk is low and related to the magnitude of the left-to-right shunt and to the compensatory response of the LV to the volume overload. Patients with unrestrictive VSDs, who have survived to adulthood, usually do so because an increase in PVR reduces the left-to-right shunt and the volume overload to the LV but achieves a reversed shunt. Very few patients with unrepaired, unrestrictive VSDs survive to adulthood due to the rapid development of PVD, Eisenmenger syndrome, and biventricular failure. Patients with repaired or unrepaired VSDs should be managed and monitored as mentioned previously in the discussion of pulmonary vascular disease. Bacterial endocarditis prophylaxis should be given to both patients with repaired and unrepaired VSDs. Patients with VSDs may have valvular heart disease as a consequence of compensatory intracardiac changes or surgical interventions. Arrhythmias and varying degrees of heart block may be present, including right bundle branch block (RBBB), which occurs in less than 10% of patients and may be a cause of sudden death. Complete heart block occurs in less than 5% of patients. Residual shunts occur in 20% of repaired patients with VSDs and if hemodynamically significant may result in pulmonary hypertension Canneson et. al (2009).

Cyanotic Congenital Heart Disease

Patients with cyanotic CHD are hypoxemic with arterial desaturation which results from shunting of systemic venous blood into the arterial circulation. The severity of hypoxemia and desaturation is determined by the magnitude of the shunting. Most patients with cyanotic CHD do not survive to adulthood without surgical intervention or palliation. In adults with cyanotic CHD, the most common causes are tetralogy of Fallot and Eisenmenger syndrome.

Tetralogy of Fallot

Tetralogy of Fallot (TOF) occurs in 10% of all CHD. It is the most common cyanotic heart defect seen in children after infancy and is the most common form of adult cyanotic CHD. TOF is characterized by a large subarterial VSD, an aorta that overrides the left and right ventricles, right ventricular outflow tract (RVOT) obstruction, and right ventricular hypertrophy (RVH). The RVOT obstruction may be subvalvular, valvular, and supravalvular or in the branch pulmonary arteries. There are also several associated anomalies with TOF, including pulmonary atresia, a right-sided aorta, and ASD, which occurs in 10% of patients and is referred to as pentalogy of Fallot. Coronary anomalies occur in 10% of patients. Patients with TOF are cyanotic secondary to right-to-left shunting. Secondary to the large VSD, the right and left ventricles have equalized pressures. Right-to-left shunting of the venous blood occurs because of increased resistance to pulmonary blood flow secondary to the RVOT obstruction. The severity of the RVOT obstruction determines the magnitude of shunting and therefore cyanosis. The resistance to flow across the RVOT is relatively fixed, and thus changes in SVR affect the magnitude of right-to-left shunting. A decrease in SVR worsens the right-to-left shunt, whereas an increase in SVR decreases the right-to-left shunting. In managing a patient with TOF, it is imperative that the SVR be maintained so the hypoxemia is not worsened.

Patients with TOF generally have cyanosis starting within the first year of life. These children exhibit what is referred to "tet spells" or hypercyanotic spells. These sudden hypoxic episodes are characterized by tachypnea and severe cyanosis with occasional loss of consciousness, seizures, cerebrovascular accidents, and even death. Tet spells do not occur in adolescents or adults. Adults with TOF have dyspnea and very limited exercise tolerance. They also have multisystemic complications of chronic cyanosis, including erythrocytosis, hyperviscosity, renal dysfunction, uric acid metabolism disturbances/gout, hemostatic derangements, cerebral abscesses, strokes, and endocarditis. Most patients with TOF who have not undergone surgical repair or palliation die in childhood. The survival rate is 66% at 1 year of age, 40% at 3 years of age, 11% at 20 years of age, 6% at 30 years of age, and 3% at 40 years of age.

In the past, infants with TOF underwent one of the three palliative procedures to increase pulmonary blood flow with a direct arterial-to-venous shunt (systemic to PA). This serves to increase pulmonary blood flow, decrease cyanosis, and improve exercise tolerance. These shunt procedures included the Waterston procedure (ascending aorta to PA anastomosis), the Potts procedure (descending aorta to left PA anastomosis), and the Blalock–Taussig shunt (subclavian artery to PA anastomosis). These procedures were often associated with long-term consequences such as pulmonary hypertension, PA distortion, and LV volume overload. At present, complete surgical repair with closure of the VSD and relief of the RVOT obstruction is usually performed. Palliative shunting, balloon valvuloplasty, and RVOT stenting are generally only performed on children with unfavorable PA anatomy or those too small or ill to undergo complete repair.

Adult and pediatric patients who have undergone repair of TOF are at risk for other long-term sequelae. Pulmonary regurgitation and/or stenosis may develop as a consequence of surgical repair of the valve or the RVOT with result RVH and RV dysfunction. Repair or replacement of the pulmonary valve may be required either by a surgical approach or more commonly by a transcatheter or hybrid approach. Aneurysm formation at the site of the RVOT repair is not uncommon in repaired TOF patients and has a risk of rupturing. Patients also frequently experience residual or recurrent obstruction of the RVOT and require re-intervention either by surgical or interventional cardiology technique. Ten to twenty percent of patients with repaired TOF have residual VSDs and may be at risk for developing pulmonary hypertension and require repeat surgery. Aortic regurgitation is also a common postrepair finding secondary to the location of the subarterial VSD. Right bundle branch block and other conduction disturbances are common after TOF repair. Complete heart block is a rare complication that requires a permanent pacemaker. All patients with TOF, both repaired and unrepaired, are at risk for endocarditis and should receive SBE prophylaxis before dental or surgical procedures Perloff & Warner (2001).

Ebstein's Anomaly

Ebstein's anomaly of the tricuspid valve occurs in less than 1% of all congenital heart defects. Ebstein's anomaly is a congenital defect of the tricuspid valve in which the septal leaflet and occasionally the posterior leaflet are displaced into the right ventricle and the anterior leaflet is usually redundant, malformed, and abnormally adherent to the right ventricular free wall. As a result, a portion of the right ventricle is "atrialized" in that it is incorporated into the RA and a functional hypoplasia of the RV results. Redundant tricuspid valve tissues can occasionally obstruct the RVOT, and tricuspid regurgitation (TR) or tricuspid stenosis is frequently present. The RA is dilated and hypertrophied and patients are prone to arrhythmias. Eighty percent of patients with Ebstein's anomaly have an ASD or patent foramen ovale (PFO) by which right-to-left shunting may occur.

In the case of Ebstein's anomaly, the severity of the hemodynamic compromise is dependent on the functional status of the tricuspid valve leaflets. Patients with mild apical displacement of the tricuspid leaflets have virtually normal valvular function, whereas those with severe tricuspid leaflet displacement or abnormal anterior leaflet attachment, with valvular dysfunction, have elevated RA pressure and right-to-left interatrial shunting. Likewise, the clinical presentation of Ebstein's anomaly varies from severe cyanosis and heart failure in a fetus or neonate to the absence of symptoms in an adult in whom it is discovered as an incidental finding.

Intrauterine mortality is high for the fetus with Ebstein's anomaly. Neonates with severe disease have cyanosis with heart failure and a murmur noted in the first few days of life. As PVR decreases, there may be a transient improvement, but the condition worsens after the ductus arteriosus closes, thereby decreasing pulmonary blood flow. Older children with Ebstein's anomaly often come to medical attention when a murmur is found on physical exam. Adolescents and adults frequently present with supraventricular tachycardia (SVT) and other arrhythmias. For patients with paroxysmal atrial tachycardia, cya-

nosis, and cardiomegaly, these are the factors which are most closely associated with poor outcome. In patients with Ebstein's anomaly with ASD or PFO, there is a risk of paradoxical embolism, brain abscess, and sudden death. On physical exam patients may have severe cyanosis, hepatomegaly due to passive congestion and elevated RA pressures. Frequently, these patients have large P waves on ECG as well as right bundle branch block (RBBB) and first-degree heart block. Approximately 20% of patients with Ebstein's anomaly have Wolff–Parkinson–White (WPW) syndrome, which is a ventricular preexcitation arrhythmia through an accessory pathway between the atrium and the ventricle. A delta wave may be present on ECG.

For adult patients with Ebstein's anomaly who are symptomatic despite medical management, it is recommended to undergo repair or replacement of the tricuspid valve and closure of the ASD. Surgery may also be recommended for patients with less severe symptoms but whom also have cardiomegaly. For patients undergoing both cardiac surgical intervention and noncardiac surgery, it is recommended that all patients have complete perioperative evaluation and optimization including appropriate imaging such as echocardiogram and/or MRI as well as ECG. Perioperative arterial line and central venous pressure monitoring are recommended as well as intraoperative TEE. TEE monitoring is recommended to evaluate anatomy, shunting, loading conditions, biventricular function, the presence of RVOT obstruction, the presence of associated anomalies, loading conditions, and the integrity of the repair in the case of cardiac surgery. PAC monitoring is not recommended as it is likely to cross the ASD into the left heart.

Transposition of the Great Arteries

D-Transposition of the great arteries (D-TGA) occurs in about 5% of all CHD with a male predominance of three males to one female. D-TGA exists when there is an anatomic discordance between the ventricles and the great arteries, in that the morphologic RV gives rise to the aorta and the morphologic LV gives rise to the PA. The physiologic consequence, surgical plan, and long-term sequelae are contingent on the age at presentation and the associated anomalies. If the systemic and pulmonary venous returns are normal, the alternatives for surgical correction have traditionally been "switching" the circulation at the great arterial level (arterial switch or Jatene procedure) or at the atrial level (Senning or Mustard procedure). For patients with D-TGA, VSD, and subpulmonic stenosis, the Rastelli procedure realigns the ventricular septum so that the appropriate venous return is associated with the correct great vessel and the subpulmonic stenosis is resected. Patients with associated defects such as aberrant venous return and RVOT or LVOT obstruction (occurs in 30% of patients) require a customized plan for correction or palliation. Additionally, 30% of patients with D-TGA have coronary anomalies including the origins of the coronary arteries and the sinuses of Valsalva, which are displaced to the posterior portion of the transposed aorta. This represents a risk for coronary compromise and ischemia as a short- and long-term consequence of arterial switch procedures. VSDs of hemodynamic significance are also associated with 25% of D-TGA patients Hucin et al. (2000).

Prior to the introduction of the atrial baffle procedures (Senning and Mustard operations), the 1-year mortality for patients with D-TGA was approximately 90%. The Senning and Mustard interatrial baffle procedures were developed in the late 1950s and early 1960s and significantly improved survival for patients with D-TGA. Many of these patients now comprise a significant portion of the adult congenital population. The procedures attempt to correct the circulation which runs parallel by redirecting venous return at the atrial level. Systemic venous blood enters the RA and is directed across the atrial septum through the baffle and across the mitral valve to the LV and into the PA. Similarly, pulmonary venous return is directed across the atrial septum through the tricuspid valve and into the RV and ejected into the aorta. The difference between the two procedures involves aspects related to the atrial septum construction and placement of the suture lines, which have had long-term sequelae.

Atrial switch procedures have a number of long-term consequences including arrhythmias, sudden death, RV (systemic ventricle) dysfunction, baffle leaks, and obstruction which impairs venous return from the respective circulation. SVT, atrial fibrillation, atrial flutter, and sinus node dysfunction are the most common arrhythmias. Pulmonary hypertension, ventricular dysfunction, and junctional rhythm are risk factors for SVT. Approximately, one third of patients with atrial switch procedures will ultimately require a permanent pacemaker secondary to sinus node dysfunction. Ten percent of patients will experience baffle obstruction of either the systemic or pulmonary venous return. The function of the RV as the systemic ventricle deteriorates with time, and sudden death is experienced in 5% of patients and is attributable to persistent heart failure. Other causes of sudden death in these patients are attributable to arrhythmias and pulmonary hypertension.

For patients with D-TGA, VSD, and subpulmonic stenosis, the Rastelli procedure is frequently performed. In this procedure, the VSD is closed in a manner that redirects venous return to the appropriate great vessel, and an RV to PA conduit is placed to relieve the subpulmonic stenosis. Long-term follow-up in these patients reveals 74% survival for patients operated on in the 1980s. The majority of patients require further intervention both surgical and catheter based. There is also a 5% risk for sudden death in these patients due to ventricular dysfunction and arrhythmias.

The arterial switch operation or Jatene procedure involves transplanting the coronary arteries to the PA and connecting the proximal great vessels to the distal ends of the other vessel in an attempt to restore anatomically correct circulation. This procedure is considered superior to the atrial switch procedures in that it is physiologically correct and results in fewer long-term complications such as arrhythmias, RV failure, baffle obstruction, and TR. For patients with uncomplicated D-TGA, normal sinus rhythm is usually present, and providing good coronary blood flow, LV function is preserved. However, PA stenosis at the site of reconstruction occurs in 5–10% of cases. Complete heart block is a risk in 5–10% of patients, and aortic regurgitation (AR) is a late complication and occurs in 20% of patients especially in patients who underwent PA banding in the neonatal period. An important cause of AR may be unequal size of the pulmonary valve cusps that leads to eccentric valvular coaptation. The most important life-threatening complication associated with arterial switch is the possibility of coronary obstruction which leads to myocardial ischemia, infarction, and death. Long-term outcomes continue to be accessed with many patients who have previously undergone the arterial switch procedure entering their third decade of life. The overall conclusion is that the arterial switch is the procedure of choice with preservation of physiologic circulation but that coronary obstruction, pulmonary stenosis, and aortic insufficiency remain significant problems.

Eisenmenger Syndrome

Eisenmenger syndrome develops over time as a consequence of significant, uncorrected left-to-right shunting in association with a VSD, ASD, or PDA. As discussed previously in this chapter, morphologic transformations occur in the small PAs and arterioles, leading to pulmonary hypertension and the resultant reversal of the intracardiac shunt creating cyanosis. In the small PAs and arterioles, medial hypertrophy, intimal cellular proliferation, and fibrosis lead to narrowing or closure of the vessel lumen. With sustained pulmonary hypertension, insidious atherosclerosis and calcification often develop in large PAs. The initial morphologic alterations, medial hypertrophy of the pulmonary arterioles, intimal proliferation and fibrosis, and occlusion of capillaries are potentially reversible. However, as the disease progresses, the morphologic changes including plexiform lesions and necrotizing arteritis are irreversible and result in obliteration of the pulmonary vascular bed, which leads to increased PVR. As the PVR approaches or exceeds systemic resistance, the shunt is reversed. The morphologic changes in the pulmonary vasculature that occur with Eisenmenger syndrome usually

begin in childhood, but symptoms don't usually start to appear until adolescences or early adulthood when the right-to-left shunt is sustained and cyanosis appears.

Most patients have decreased exercise tolerance and dyspnea and these symptoms may be compensated for a time period. Atrial arrhythmias frequently develop such as atrial fibrillation and atrial flutter. Erythrocytosis develops due to arterial desaturation, and symptoms of hyperviscosity such as visual disturbances, fatigue, headache, dizziness, and paresthesias frequently develop. Hemoptysis secondary to rupture or infarction of dilated PAs can occur and be life-threatening. Patients who have developed cyanosis also have abnormal hemostasis and are at risk for cerebrovascular accidents which can occur due to paradoxical emboli, venous thrombosis, or intracranial hemorrhage. Patients with Eisenmenger are also at risk for brain abscess and syncope. Syncope may occur secondary to inadequate cardiac output or arrhythmias. Symptoms of heart failure are not common until the disease is in advanced stages and implies a very poor prognosis. Patients with Eisenmenger are at risk for sudden death.

Medical management of Eisenmenger syndrome is similar to that of pulmonary hypertension and includes inhaled oxygen, nitric oxide, treprostinil, and IV vasodilators such as epoprostenol. Other dilators such as sildenafil may be taken orally and used in conjunction with calcium channel blockers, endothelin receptor antagonists, anticoagulants, diuretics, and antiarrhythmics. For some patients, the ultimate therapy may be heart–lung transplantation providing other organ systems have preserved function. The effectiveness of these therapies is limited by the severity of the disease. Patients with Eisenmenger syndrome should avoid high-altitude and excessive exertion, dehydration, and the use of systemic vasodilators.

Pregnancy is associated with high maternal and fetal morbidity and mortality. The fixed PVR precludes any compensation for acute fluctuations in SVR, cardiac output, and blood volume during labor, delivery, and the peripartum period. Any acute decrease in SVR precipitates increased right-to-left shunting and results in severe cyanosis. Valsalva maneuvers during labor may acutely increase SVR, depress systemic and cerebral perfusion, and trigger fatal syncope.

Patients undergoing both cardiac and noncardiac surgery should be cared for in centers experienced in the management of such patients and require a multidisciplinary approach. The anesthesia care required is meticulous management of fluid status and both systemic and pulmonary vascular resistance. Maintenance of SVR and contractility, minimizing blood loss and intravascular volume depletion, as well as preventing paradoxical embolization are imperative. Most patients are admitted for overnight IV placement and mild hydration. Prophylactic phlebotomy is generally not done in most centers as the volume shifts may be poorly tolerated. The intraoperative management is frequently dictated by the nature of the procedure; however, most general anesthetic including cardiac procedures will necessitate arterial line and central venous pressure monitoring and TEE monitoring. PACs are not indicated due to the likelihood of crossing the shunt lesion such as an ASD or the potential for PA rupture.

Single-Ventricle Physiology and Complex Cyanotic Congenital Heart Disease

Advances in palliative and corrective heart surgery, catheter-based interventions, anesthesia, and imaging have significantly increased the number of patients with complex, cyanotic heart disease who have reach adulthood. Patients who have undergone single-ventricle palliation with a Fontan procedure illustrate the cumulative risks of dysrhythmias, decreased ventricular function, and chronic hypoxemia as well as other multiorgan system sequelae.

Many complex congenital heart lesions result in anatomic malformations that are not amenable to surgical intervention and result in a functioning biventricular circulation. These lesions include severe Ebstein's anomaly, tricuspid atresia, hypoplastic left heart syndrome (HLHS), unbalanced atrioventricular canal (AV canal), and various

defects associated with heterotaxy syndrome. These forms of CHD require staged procedures that attempt to use an existing ventricle as the systemic ventricle and create a mechanism for pulmonary blood flow that may not necessarily require a pumping chamber and will likely be passive flow. The surgical plan for these patients is somewhat individualized and dependent on the native anatomy with the ultimate conformation relatively similar among patients with single-ventricle physiology. The Fontan circuit functions to direct systemic venous return via a conduit into the PA system. The circuit does not require a right-sided heart chamber, and pulmonary venous blood returns into a common atrium and flows into the systemic ventricle Hosking & Beynen (1992).

The first palliative operation for patients with single-ventricle physiology usually requires a shunt from an arterial source that provides pulmonary blood flow or a ductal stent to maintain an open PDA as the source for pulmonary blood flow in the first-stage hybrid palliation procedure. Adequate mixing at the atrial level is required with an unrestrictive ASD along with unobstructed systemic ventricular outflow. In the past, the Blalock–Taussig (BT) shunt (right subclavian to PA anastomosis) or a central shunt (aorta to PA shunt) was performed. Presently, the modified BT shunt is frequently used with a Gore-Tex conduit from the right subclavian artery to the PA to provide pulmonary blood flow. Historically, the classic BT shunt permanently compromised arterial blood flow to the right arm. The modified BT shunt is generally ligated at the time of the subsequent procedure and does not permanently compromise flow. This is relevant in that patients who have undergone a classic BT shunt will not have an accurate blood pressure measured in the right upper extremity. The BT shunt in the case of HLHS provides the pulmonary blood flow while the creation of a neoaorta (Norwood procedure) from the proximal PA creates the manner in which systemic blood flow leaves the heart. The subsequent procedures in the palliative process attempt to route systemic blood flow to the PA vascular bed. The second-stage procedure (bidirectional Glenn shunt) involves creating a superior vena cava to PA anastomosis. The final stage, or Fontan procedure, results in the creation of an inferior vena cava to PA connection and may be undertaken in a number of ways. Fenestration of the Fontan conduit creates a functional ASD that serves as a pressure outlet in the setting of pulmonary hypertension and maintains LA filling and systemic perfusion Khairy et al. (2007).

Modifications and revisions of the single-ventricle palliative path have been made since their initial performance in the 1970s and decreased the incidence of adverse sequelae such as arrhythmias, pulmonary hypertension, ventricular dysfunction, and heart failure. For patients who underwent the Fontan procedure in the 1970s, the 15-year survival rate was 50–80%. The majority of Fontan patients have NYHA class I or II symptoms and exhibit decreased exercise tolerance. Approximately 40% of these patients require reoperation for conduit obstruction, permanent pacemaker, or AV valve repair for valvular regurgitation. Atrial arrhythmias are very common among Fontan patients, and loss of the atrial contribution to cardiac output can result in significant impairment, and patients will frequently require permanent pacing.

Another significant adverse sequela of Fontan physiology is protein-losing enteropathy (PLE). PLE develops in 5–10% of patients and is manifested by gastrointestinal protein malabsorption, ascites, peripheral edema, pleural effusions, and low serum protein levels. Elevated systemic venous pressure, abnormal mucosal blood flow patterns, and glycosylation of enteric proteins have been implicated as the cause of this condition. Patients with fenestrated versus non-fenestrated Fontan conduits have a decreased incidence of protein-losing enteropathy. The therapies include fenestration of the Fontan conduit, anticoagulation, dietary modifications, corticosteroids, and ultimately heart transplantation.

Following Fontan procedures, the formation of arteriovenous malformations (AVMs) has been observed, especially pulmonary AVMs in the lungs. Pulmonary AVMs produce a significant volume load on the ventricle and impair function. Shunting may also occur through the AVMs and induce parenchymal changes, hypoxia, and poor lung compliance.

The perioperative management of these patients requires attention to maintaining SVR, not increasing PVR and attention to loading conditions and fluid status. High positive pressures with ventilation or other maneuvers which decrease venous return are not recommended. The type of invasive monitoring is dictated by the procedure. For patients undergoing cardiac surgery or procedures, arterial line and central pressure monitoring is recommended. TEE is required to access ventricular function, AV valve regurgitation, potential paradoxical emboli, loading conditions, and the integrity of the repair. For patients undergoing noncardiac surgery, similar monitoring may be advised depending on the procedure. Attention needs to be paid to bleeding or other volume losses with appropriate fluid management or blood replacement. Patients are at high risk for antibodies and type-specific blood may take extra time to prepare.

Perioperative Concerns and Anesthetic Management

The perioperative strategy and management require a complete assessment and consider factors relevant to both the patient's underlying physiology as well as factors related to the proposed procedure. The patient's baseline cardiovascular reserve and underlying multi-organ system impairment must be evaluated in conjunction with the concerns specific to the CHD and potential acquired comorbidities related to aging. The techniques required for the procedure need to be considered and balanced in light of what the patient's cardiovascular reserve will tolerate. For example, a patient with a failing Fontan physiology is unlikely to tolerate high-pressure abdominal insufflation associated with numerous laparoscopic procedures currently being performed. It is recommended that adult patients with CHD be cared for in a center with a dedicated team familiar with their unique physiology and that a multidisciplinary approach be used when faced with complicated medical decisions regarding these patients Landzberg et al. (2001).

Syndromes and Associated Anomalies

Many forms of CHD occur in association with other congenital syndromes and congenital anomalies which are noncardiac. Endocardial cushion defects such as AV canal, ASDs, and VSDs are common among patients with Down's syndrome. These patients have characteristic anomalies which impact management such as developmental delay, hypotonia, large tongue, short necks, potential for atlanto-occipital dislocation, and other features related to the syndrome. DiGeorge syndrome is another association in which conotruncal anomalies occur with abnormal calcium homeostasis, immunodeficiency, and multiple midline craniofacial anomalies. VACTERL association includes vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities. Patients with VACTERL association typically have at least three of these defects and require attention to potential other systemic involvement. However, in the adult congenital population, it is likely that other noncardiac defects would have been previously diagnosed Perloff & Child (2008).

Perioperative Monitoring

Perioperative monitoring must be carefully considered and appropriate for the level of baseline impairment and anticipated demands of the procedure. Arterial line, central venous pressure monitoring, and TEE monitoring may be required to ensure the most accurate assessment of loading conditions, biventricular function, direction and magnitude of shunting, and potential for paradoxical embolism. The standard monitoring equipment such as ECG, pulse oximetry, and temperature and noninvasive blood pressure monitors may also require a little extra consideration. For example, the question may arise as to where to place the ECG pads for a patient with dextrocardia or heterotaxy syndrome. The answer is that a 5-lead and 3-lead ECG placement is done in the standard fashion

for all patients (baseline ECGs are also performed in the standard position). For patients who have undergone a coarctation repair incorporating the left subclavian artery, the right upper extremity will most accurately reflect the systemic blood pressure. There may also be a residual gradient between the upper and lower extremity blood pressures in these patients. For patients who have previously undergone a classic BT shunt, the ipsilateral extremity may not accurately reflect the systemic blood pressure as well as oxygen saturation, and an alternative site should be selected. For patients undergoing cardiac surgery, cerebral oximetry is also recommended and considered standard of care.

Airway and Ventilatory Management

Patients with CHD may have associated anomalies including those involving the airway. In the adult population, these anomalies may already be well documented and/or known to the patient or their families. In addition, patients who have undergone previous surgery may have experienced prolonged periods of intubation and mechanical ventilatory support, and the potential for subglottic stenosis and a history of tracheostomy are possibilities. The transition from spontaneous ventilation to positive pressure ventilation can significantly impair preload, secondary to decrease systemic venous return. This is especially important in the scenario of hypovolemia.

Patients with single-ventricle physiology are extremely sensitive to positive pressure ventilation because their pulmonary blood flow is preload dependent. Episodes of desaturation may be an indication that pulmonary blood flow is being compromised. If standard interventions are attempted such as increasing tidal volume or adding positive end-expiratory pressure (PEEP), the result may further decrease in the saturation, secondary to further decrease in pulmonary blood flow. In the case of single-ventricle physiology, maintaining adequate intravascular volume status, limiting the peak inspiratory pressure, and avoidance of PEEP will facilitate pulmonary blood flow and preserve oxygenation.

Intravenous Access Considerations

Intravenous access can often present a significant problem in patients with CHD, especially those who have undergone multiple procedures or who have significant peripheral collateralization secondary to cyanotic heart disease or venous obstruction. A careful examination of the patient may reveal previously cannulated or cut-down sites as well as a review of previous operative reports and cath reports. Ultrasound of the potential sites as well as angiography in the cath lab will help in identifying sites available for vascular cannulation. Patients with right-to-left shunts are at risk for paradoxical embolization of air or particles, even when infused into peripheral lines and vigilance and the use of infusion filters is recommended to decrease the possibility of emboli.

Perioperative Fluid Management and NPO Intervals

Standard NPO intervals in healthy adult patients with normal cardiovascular reserve generally have little impact. However, for patients with cardiac anatomy dependent on adequate filling pressures, even minor decreases in preload can have adverse effects. Patients with single-ventricle physiology, cyanotic patients with erythrocytosis, patients with Eisenmenger physiology, and patients with volume-dependent obstructive lesions are extremely sensitive to acute decreases in preload. Decreased preload and the vasodilating effects of anesthetic agents can lead to acute circulatory collapse even in patients who seem well compensated prior to induction. Ensuring adequate preload prior to induction and judicious titration of anesthetic agents can facilitate preservation of cardiac output in the previously described patients. Management of perioperative volume losses should be guided by the monitoring of vital signs, arterial line pressure, central

venous pressure, hematocrit, urine output, acid–base status, and electrolytes.

Third-space losses, especially in the case of patients with protein-losing enteropathy, must be considered. PLE results in hypoalbuminemia, ascites, and pericardial and pleural effusion and is associated with high systemic venous pressures and ventricular dysfunction of the single ventricle. These patients are frequently total volume fluid overloaded with third-space sequestered volume and intravascular space depleted with poor cardiovascular reserve. This clinical situation implies extremely poor cardiac reserve, and the ability to tolerate or compensate for intravascular depletion, vasodilating anesthetics, and positive pressure ventilation is exceptionally limited. Medication doses should be reduced to account for the effect of hypoalbuminemia, and all medications should be titrated to effect with hemodynamic monitoring. Appropriate first-case scheduling, minimizing NPO intervals, or preoperative hospital admission with IV hydration should be considered in this group of patients, to avoid the risks of intravascular volume depletion.

Hematocrit and Perioperative Transfusion Management

Patients with chronic cyanosis will compensate for their hypoxemia with a baseline erythrocytosis in an attempt to increase oxygen delivery. If a chronically cyanotic patient is found to have a normal range hematocrit, a coexisting anemia, usually iron deficiency, should be considered. Erythrocytosis predisposes patients to complications of hyperviscosity. A hematocrit of greater than 40% is generally considered to provide an adequate red cell mass for oxygen delivery. However, the patient's own baseline hematocrit will frequently serve as a guide, providing they are not anemic.

Extra consideration must be given to the situation in which a transfusion is required in that many patients with CHD require special preparation of blood products. Patients with DiGeorge syndrome or who are immunosuppressed require cytomegalovirus (CMV)-negative blood that has been irradiated and leukocyte filtered. This is done to decrease the potential transmission of CMV and prevent graft-versus-host disease, along with other leukocyte-mediated transfusion reactions. If a question arises regarding the type of blood products required for a specific condition, a hematologist or blood bank pathologist should be consulted. Additionally, the majority of adult congenital heart patients have undergone previous heart surgery with the concomitant exposure to blood products and previous transfusion. It is extremely common for these patients to have developed antibodies that may complicate the cross-match process, and extra time must be allowed in order to prepare type-specific blood products. Attention must also be paid to patients that may in the future require heart transplantation. Limiting transfusions and therefore antibody formation is vital for these patients. Adult congenital heart patients should also receive leukocyte-depleted blood to also help decrease antibody formation.

Antibiotic Prophylaxis

The requirement for SBE prophylaxis is dictated by the underlying cardiac disease and anatomy as well as the type of procedure being performed. The American Heart Association offers a comprehensive list with specific recommendations for patients with CHD on their website www.americanheart.org. Most patients with CHD have significant graft material and prosthetic valves as well as intracardiac shunts and regurgitant or stenotic valves. They are at high risk for endocarditis and it is a frequent complication for many patients with CHD.

Pacemakers and Arrhythmias

Arrhythmias are among the most prevalent and complex sequelae for patients with adult CHD. The arrhythmias and common rhythm disturbances associated with each defect have been previously discussed and can be found with

the respective lesion discussion. Many patients with adult CHD require antiarrhythmic therapy that should be reviewed before surgery. The side effects and interactions of these medications should be known. A baseline ECG is recommended before all procedures, and if arrhythmias do arise in the perioperative period, a rapid diagnosis should be made and therapy should be instituted. The underlying etiology should be considered in each case with respect to the circumstances and underlying anatomy and physiology. For example, D-TGA with atrial switch anatomy is known to be associated with atrial arrhythmias. Electrolyte disturbances especially secondary to diuretic therapy, hypercarbia, acidosis, severe hypoxia, and antiarrhythmic agents can also produce arrhythmias. Medications that blunt vagal tone should be used with extreme caution.

Many patients with adult CHD have permanent pacemakers and implantable cardiac defibrillators (ICD). The settings and the underlying rhythm should be known. A magnet for conversion to the asynchronous mode should be available, and the therapy mode of the ICD may need to be deactivated to avoid triggering inappropriately in the operative setting.

Postoperative Considerations

In the immediate postoperative period, extreme vigilance and similar levels of monitoring which were employed during the operative period should be continued. The level of postoperative monitoring should be appropriate for the patient's baseline physiology and the nature of the surgical procedure and its anticipated postoperative sequelae. Many patients with adult CHD will continue to need arterial line and CVP monitoring in the postoperative period and may require management in the intensive care unit that an otherwise normal cardiovascular status patient undergoing the same procedure would not require.

Pain management and postoperative nausea and vomiting are important considerations. Appropriate analgesia including narcotic medications, regional anesthesia which does not impair SVR, acetaminophen derivatives, and nonsteroidal anti-inflammatory medications are all used with success in patients with CHD. Analgesia facilitates improved respiratory mechanics and decreases oxygen consumption. However, respiratory depression, hypercarbia, and airway obstruction must be avoided secondary to their significant effects on increasing PVR. The management of postoperative nausea and vomiting is crucial for oral hydration and the ability to resume medication regimens especially antiarrhythmic and anticoagulation medications.

Conclusion

Adult patients with CHD represent an ever-increasing group of patients with complex cardiac anatomy and physiology as well as acquired comorbidities. These patients represent a challenge to the medical community and require a multidisciplinary team approach with comprehensive perioperative planning. At this time, there are no evidence-based guidelines for the perioperative management of adult patients with CHD. Large-scale clinical trials are required to demonstrate the optimal management strategies, and it is recommended that these patients be cared for in cardiac centers with congenital heart specialists who are familiar with their physiology. An appreciation of the previously discussed principles will assist the provider in evaluating a multitude of structural lesions based on their underlying physiologic effects. Ultimately, an understanding of the patient's unique physiology and multi-organ system sequelae will enhance the likelihood of perioperative cardiovascular stability.

Case Study: Anesthetic Management of a Hybrid Approach to Transcatheter Pulmonary Valve Replacement in a Previously Repaired Patient with Tetralogy of Fallot

A 15-year-old male with a history of TOF s/p transannular patch repair as an infant presents with severe pulmonary valve regurgitation (PR)

and right heart dilatation. Although asymptomatic from a cardiac standpoint, he did have severe right ventricular dilation on MRI (RVEDV = 180 mL/m ^{2}). After careful discussion about treatment options, the patient decided on a transcatheter Melody valve. A diagnostic angiogram showed an extremely enlarged and tortuous pulmonary artery not amenable to a Melody valve. 3D reconstruction of the patient's PA was used to further evaluate the anatomy for other possible interventions. After careful planning and several virtual implants, a hybrid procedure was decided upon in which a Melody valve could be placed within a pulmonary artery stent. In preparation for the hybrid procedure, the patient was premedicated with midazolam and induced with etomidate and cisatracurium for general endotracheal anesthesia. A radial arterial line and right internal jugular central line were placed. Both OR staffing and a perfusionist were on standby to go on cardiopulmonary bypass and convert to an open procedure if necessary. Access for stent delivery was achieved through a subxiphoid incision by a cardiothoracic surgeon. Under fluoroscopy and transesophageal echocardiogram (TEE), a covered stent and landing stent were placed with vascular plugs along the tortuous pulmonary artery taking care not to compromise the branch pulmonary arteries and ensuring good flow through the RVOT. After confirmation of proper seating of the stents, the Melody valve was delivered through the subxiphoid incision and deployed within the landing zone stent in the pulmonary artery. Postdeployment intracardiac echo (ICE), TEE, and angiogram showed no PR and no compromise to the branch pulmonary arteries. The patient was transferred to the PICU where he was extubated after several hours and discharged home the next day (Figs. 1, 2, 3, and 4).

Case Discussion

Surgical repair of TOF has been occurring for about 50 years with excellent outcomes. However, with longer survival, we are seeing more long-term complications of these repairs, specifically pulmonary valve regurgitation. Untreated PR in these patients can result in right-sided remodeling, which if left untreated can lead to right heart failure. Previously, that intervention was not recommended until the patient became symptomatic from the pulmonary regurgitation. However, several studies have shown that early repair of pulmonary valve regurgitation can prevent long-term right heart remodeling.

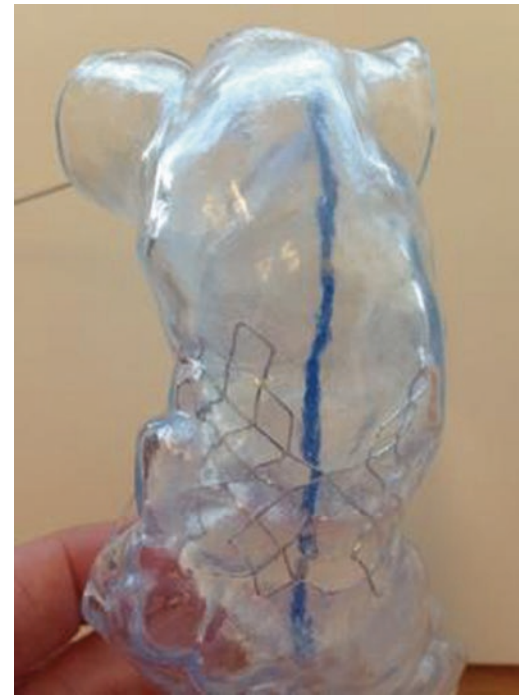


Fig. 1 3D reconstruction of pulmonary artery

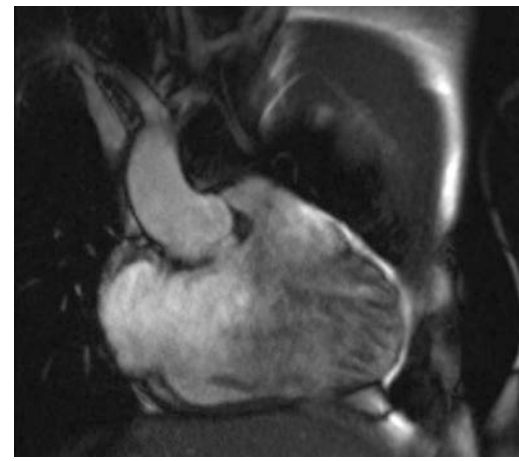


Fig. 2 Cardiac MRI showing dilated right ventricle

Currently, cardiac MRI is the gold standard for measuring right ventricular volume and pulmo-

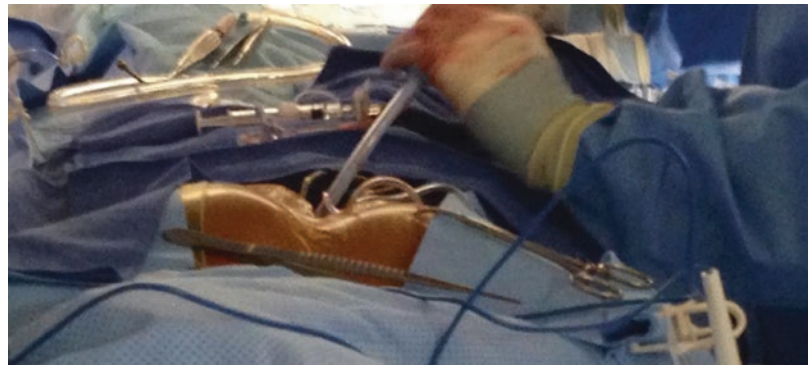


Fig. 3 Subxiphoid access for stent delivery

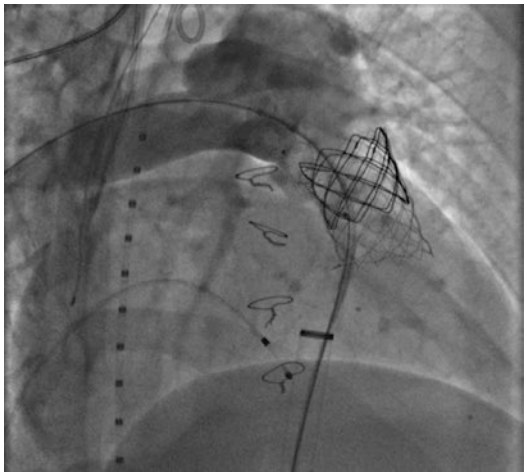


Fig. 4 Angiogram showing post-Melody valve deployment with no pulmonary regurgitation

nary regurgitation. The precise timing of when to intervene is unclear, but studies seem to show remolding of the RV when RVEDV >150 mL/m^2. Despite the overall good outcomes with surgical repair for these patients with pulmonary regurgitation, it is not without its risk factors. Most patients will have significant scar tissue formation with adherent right ventricle to the chest wall, making entering the chest extremely high risk. There are some limitations secondary to unfavorable PA anatomy which may require a hybrid approach with access through a subxiphoid incision to allow for multiple large stent placement and/or better trajectory of valve deployment. General anesthesia and arterial line are mandatory, while central access should be considered depending on the patient's venous access and baseline cardiac function. TEE is used to monitor function, access the pre- and postimplant anatomy, access loading conditions, guide the procedure, and monitor for pericardial effusion. The usual goals for PR physiology should be applied for these cases. Perfusionists and surgical staff should be readily available to go on bypass and convert to an open procedure should complications arise. Complications include bleeding, pulmonary artery rupture, cracked stents, compression of coronary vessels, arrhythmias, pulmonary regurgitation, and stent migration and embolism. Early extubation in the cath lab has been described; however, it may be prudent to leave the patient intubated if there are concerns for bleeding or if there are large fluid shifts related to volume resuscitation. The Melody valve and other transcatheter valves have made repair of pulmonary regurgitation in previously repaired TOF patients much less invasive and ultimately safer. It is important to be aware of the complications that can occur with the hybrid procedure.

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