Anesthetic Management of the Adult Patient with Congenital Heart Disease

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Because continued improvement in the outcomes of congenital cardiac surgery has created a growing population of adults with repaired or palliated congenital heart disease (CHD), equal numbers of adults and children now have CHD. Almost all children with CHD are operative candidates, and surgery results in approximately 95% survival. Adult CHD patients may present for primary repair of their congenital lesion, final repair after previous palliation, surgical revision because of failure or lack of growth of a bioprosthetic material, conversion of an earlier repair to a more modern one, and noncardiac surgery.

Providing the optimal perioperative management for these patients can be challenging. They have congenital heart defects, such as shunt lesions and single-ventricle physiology, which are most familiar to pediatric specialists. However, these patients may also have coexisting diseases of adulthood, such as coronary artery disease and chronic renal insufficiency, which are best managed by a specialist in adult diseases. For this reason, many regionalized centers that treat adults with CHD have a multidisciplinary team that provides perioperative and follow-up care.

Despite the growing number of operations performed on adults with CHD and the development of an entire subspecialty devoted to the care of these patients, limited data exist concerning their anesthetic, intraoperative, and perioperative course. Most reviews of operations for adults or teenagers with CHD have focused on long-term results.

CARDIAC COMPLICATIONS

Arrhythmias

Adults with CHD have an increased incidence of arrhythmias, which may be classified as intra-atrial re-entrant tachycardia, ventricular tachycardia, heart block, or sinus node dysfunction. Usually, intra-atrial re-entrant tachycardia affects patients who have had extensive atrial dissection and repair, including the Mustard or Senning procedure (see Transposition of the Great Vessels, later) or the Fontan procedure (see Single Ventricle, later). More than 40% of these patients develop intracardiac thrombi, and the sudden death rate is 6% to 10%. The risk for ventricular tachycardia increases with advancing age and poorer hemodynamic status. Antiarrhythmic therapy suppresses symptomatic ventricular arrhythmias but does not prevent sudden death. Bradyarrhythmias may result from surgical injury of the sinus node or atrioventricular conducting tissue or from spontaneous complete heart block. Sinus node injury most commonly occurs during the Mustard, Senning, or Fontan procedures. In contrast, atrioventricular conduction injuries tend to occur during ventricular septal defect (VSD) closure, left ventricular outflow resection, or Tetralogy of Fallot repair. The incidence of complete heart block increases by about 2% per year in patients with ventricular inversion.

Pulmonary Hypertension

Patients with lesions that involve left-to-right shunts are at risk for pulmonary hypertension and pulmonary vascular obstructive disease. Exposure of the pulmonary vasculature to systemic pressure or excessive flow causes a progressive morphologic change in the microvasculature, which initially manifests as medial hypertrophy, and then it progresses to necrotizing arteritis. Pulmonary vascular resistance increases, eventually leading to right-to-left shunting with associated hypoxia and erythrocytosis (Eisenmenger syndrome). This syndrome develops earlier and more commonly among patients with high-flow, high-pressure, systemic-to-pulmonary shunts, as seen in truncus arteriosus. Without surgical repair, Eisenmenger physiology will develop in approximately 50% of patients with a VSD or patent ductus arteriosus (PDA) but in only 10% of those with an atrial septal defect (ASD). If Eisenmenger syndrome is not diagnosed until adulthood, the prognosis is generally poor, the average life expectancy being <6 years from the time of diagnostic catheterization. Cardiac catheterization should be performed in these patients to determine the angiographic appearance of the pulmonary vascular bed and to determine whether pulmonary vascular resistance decreases with 100% oxygen or other pulmonary vasodilators, such as nitric oxide or prostacyclin. In patients with fixed, irreversible pulmonary hypertension, attempted surgical repair of the shunt yields substantially increased morbidity and mortality. Lung transplantation should be considered in patients with good cardiac function, and combined heart-lung transplantation may be suitable for those with deteriorating cardiac function or complex CHD.

After heart-lung transplantation for pulmonary hypertension, the survival rate is 72% at 1 year and 42% at 5 years; the survival rate is similar after a single or double lung transplant.

Ventricular Dysfunction

Congenital heart anomalies place a pressure or volume burden on the heart, causing the myocardium to become hypertrophied or dilated. If the hemodynamic load is relieved by means of surgery or interventional cardiac catheterization, myocardial remodeling occurs, permitting normal or near-normal myocardial performance. However, hemodynamic abnormalities that are corrected later, after age 10 years, commonly produce long-standing ventricular dysfunction. Chronic cyanosis exacerbates the abnormalities of ventricular performance.

Other Systemic Diseases Associated with Congenital Heart Disease

Pulmonary disease

Patients with cyanotic CHD require increased minute ventilation to maintain normocapnia. This requirement is because venous blood, which is rich in carbon dioxide, bypasses the lungs because of an intracardiac right-to-left shunt (i.e., there is an increase in physiologic dead space). Because of this dead space, end-tidal carbon dioxide monitoring will underestimate the amount of arterial carbon dioxide. Although cyanotic patients have normal increases in minute ventilation caused by increased carbon dioxide, they have a blunted ventilatory response to hypoxemia that improves after surgical correction of hypoxemia.

Renal disease

In CHD, chronic cyanosis produces a renal histopathology characterized by hypercellular glomeruli with basement membrane thickening, focal interstitial fibrosis, tubular atrophy, and hyalinization of afferent and efferent arterioles. In one study, more than 13% of surgical patients with cyanotic CHD developed postoperative acute renal failure after undergoing cardiopulmonary bypass.

Hematologic disease

Long-standing cyanotic CHD leads to erythrocytosis and blood hyperviscosity. As the hematocrit and viscosity increase, oxygen delivery eventually decreases because of the diminished cardiac output. Severe polycythemia also leads to coagulation and platelet abnormalities. At the time the hematocrit exceeds 65%, hemorrhagic and thrombotic complications will be reduced if a prophylactic isovolemic phlebotomy is performed before elective surgery.

Neurologic disease

Patients with an intracardiac shunt are at risk for cerebral emboli and brain abscesses. Children with severe polycythemia are vulnerable to cerebral venous and arterial thrombosis. Patients with CHD may have coexisting anomalies of the central nervous system. Moreover, adult patients who had cardiac surgery during the early years of extracorporeal circulation may have undergone prolonged deep hypothermic arrest and had air or particulate emboli. Such patients may have residual neurologic deficits or seizures.

Other congenital anomalies

More than 25% of patients with CHD have associated noncardiac anomalies, most commonly orthopedic and genitourinary malformations. Thirteen percent of CHD patients have a chromosomal abnormality, and 5% have a syndrome or heritable disorder. To detect such anomalies, especially malformations of the airway, these patients should undergo a thorough review of their various systems and a focused airway examination.

SPECIFIC CARDIAC LESIONS

Atrial Septal Defect

Because many such patients are asymptomatic, they may present for primary repair in adulthood. Because complications such as tachyarrhythmias and paradoxical emboli increase in frequency with aging, however, repair is ideally performed in childhood. Surgical closure after 5 years of age is associated with incomplete resolution of right ventricular hypertrophy, and survival is worse when ASD is closed after 24 years of age.

Ventricular Septal Defect

Most small-to-moderate VSDs will close in the first decade of life. Children with larger VSDs will have congestive heart failure and may develop irreversible pulmonary hypertension if closure is delayed. Delayed surgical closure may also place the child at risk for ventricular dysfunction years after surgical repair. Adults are at increased risk for aortic insufficiency resulting from valve prolapse into the defect.

Patent Ductus Arteriosus

Persistence of a PDA leads to a high-pressure shunt from the aorta to the pulmonary artery. This left-to-right shunt places a volume burden on the heart and, if the PDA is large, will lead to end-stage pulmonary hypertension. In adulthood, the ductus may become calcified or undergo aneurysmal dilation, and cardiopulmonary bypass or deep hypothermic circulatory arrest may be necessary to control blood flow to the ductus during repair.

Coarctation of the Aorta

In coarctation of the aorta, long-term left ventricular obstruction leads to ventricular hypertrophy, premature coronary atherosclerotic disease, and a poor long-term outcome after repair in adulthood. In patients undergoing repair after the 40th year of age, the 15-year survival rate is 50%, and half of the patients have persistent hypertension. Patients operated on

during childhood may develop an aneurysm at the repair site or have recoarctation requiring repeat surgery; 5 years after primary repair, the incidence of recoarctation approaches 20%.

Valvular Anomalies

Congenital abnormalities of the semilunar or atrioventricular valves may cause stenosis or regurgitation, and the anesthetic considerations are similar to those for acquired valvular disease.

Transposition of the Great Vessels

Without surgical intervention, patients with transposition of the great vessels die in infancy. Before 1980, the most common surgical treatment for this defect was the atrial switch (Mustard or Senning) procedure, in which systemic blood returning to the right atrium is directed to the mitral valve, and pulmonary venous blood is directed to the tricuspid valve. With this procedure, the right ventricle remains the systemic pump. Postoperatively, however, the incidence of atrial arrhythmias and sinus node dysfunction increases with time because of extensive atrial suturing and possible injury to the sinus node artery during surgery. In addition, tricuspid insufficiency and right ventricular failure develop in some patients, who are candidates for valve replacement or surgical conversion to the arterial switch procedure. Because the left ventricle is the pulmonary pump, patients must undergo progressive arterial banding to "train" the left ventricle to become the systemic pump or must use a left ventricular assist device as a bridge to recovery. In the 1980s, the arterial switch procedure became the operation of choice for most patients. Its long-term outcome is believed to be better, but concern remains about growth of the coronary anastomotic sites and development of aortic valve insufficiency.

Congenitally Corrected Transposition of the Great Vessels (L-Transposition, Ventricular Inversion)

In this congenital malformation, the physiologic left ventricle is the pulmonary ventricle, and the physiologic right ventricle is the systemic ventricle. This anomaly is associated with a VSD in 80% of patients. In the absence of associated lesions, the malformation may be unrecognized in childhood. With increasing age, the systemic (right) ventricle tends to fail, leading to tricuspid valve insufficiency. Also, arrhythmias develop in about one third of patients, heart block being the most common form. A newer surgical correction, the double switch procedure, may be performed in adolescence or early adulthood. In this procedure, an atrial switch (the Senning baffle procedure) and an arterial switch are performed, making the anatomic left ventricle the systemic pump. Before this procedure is undertaken, the patient must undergo pulmonary artery banding to "train" the left ventricle.

Ebstein's Anomaly

In Ebstein's anomaly, the attachment of the tricuspid valve is displaced downward, creating a malformed (small) right ventricle, with an atrialized portion of the right ventricle between the tricuspid valve annulus and the attachments of that valve's posterior and septal leaflets. Patients may present in infancy with cyanosis or later in life with congestive heart failure or cyanosis; some adults are asymptomatic and have a normal life expectancy. Almost half of the patients have arrhythmias, most commonly supraventricular ones, and many patients have the accessory pathway of Wolf-Parkinson–White. In adults with this disease, the most common surgical procedure is tricuspid repair or replacement. Atrioventricular block is common after tricuspid replacement.

Tetralogy of Fallot

Most patients with Tetralogy of Fallot undergo surgical repair in childhood, although primary repair in adults can also yield a good outcome. Currently, the VSD associated with Tetralogy of Fallot is closed through a right atrial incision; however, adults who have undergone repair may have had a right ventriculotomy for VSD closure. These patients commonly develop ventricular arrhythmias many years after surgical repair and are also at risk for sudden death; however, ventricular arrhythmias may not be the major cause of sudden death. The most common reasons for reoperation in adults are related to the right ventricular outflow tract—pulmonary insufficiency and right ventricular-to-pulmonary artery conduit failure.

Atrioventricular Canal

Atrioventricular canal results from incomplete closure of the endocardial cushions and is associated with a primum ASD, a VSD, and a common atrioventricular valve, usually occurring in patients with trisomy 21. Surgical correction involves closure of the ASD and VSD, division of the atrioventricular valve, and closure of a cleft in the anterior leaflet of the mitral valve. Repair is usually performed in infancy because of the risk for end-stage pulmonary vascular disease. Postoperatively, residual or progressive mitral valve regurgitation is common, with 10% to 30% of patients requiring repeat surgery.

Truncus Arteriosus

In truncus arteriosus, a common semilunar (truncal) valve gives rise to the systemic, pulmonary, and coronary circulations, and a VSD is present below the truncal valve. Surgical repair is usually performed in infancy and typically involves VSD closure, routing the left ventricular blood through the truncal valve, and inserting a right ventricle-to-pulmonary artery conduit to provide pulmonary blood flow. Almost all patients with this type of repair will later need replacement of the pulmonary conduit. In addition,

some patients develop truncal (neo-aortic) valve insufficiency requiring valve repair or replacement.

Single Ventricle

A wide variety of anatomic abnormalities will produce single-ventricle physiology. Most of these patients have atresia of an atrioventricular or a semilunar valve, with complete mixing of systemic venous and pulmonary venous blood. Ventricular ejection into the pulmonary or systemic circulation is based largely on the vascular resistance of these circuits. Most patients require surgical palliation in infancy with either a shunt that provides pulmonary blood flow from the systemic circulation or a pulmonary artery band that limits flow into the pulmonary circuit. These patients will undergo staged repair of their heart disease, eventually culminating in the Fontan procedure. This operation routes systemic venous blood directly to the pulmonary circulation, and central venous pressure becomes the driving force for pulmonary blood flow. With this physiology, negative intrathoracic pressure generated by spontaneous ventilation promotes pulmonary blood flow. Conversion from spontaneous ventilation to positive-pressure ventilation increases the intrathoracic pressure, thereby decreasing pulmonary blood flow, which reduces the ventricular preload and may significantly decrease the cardiac output. The Fontan procedure was originally performed via an atriopulmonary connection. With this anatomy, the right atrium frequently becomes massively dilated, leading to atrial dysrhythmias. Atrial contraction also creates turbulence in the blood stream, reducing effective forward flow. Failing Fontan repairs have been successfully converted to an extracardiac, or lateral-tunnel, Fontan repair. Consequently, systemic blood flows to the lungs directly from the superior vena cava and by way of a large venous conduit or intracardiac tunnel that connects the inferior vena cava and pulmonary arteries, minimizing arrhythmias and improving function.

ANESTHETIC MANAGEMENT

On the basis of the data mentioned earlier and our experience, the following management strategy for adult patients undergoing surgery for CHD is recommended:

1. Preoperative Preparations

a. Patient data should be presented to a multidisciplinary group of cardiologists, surgeons, and anesthesiologists. Data analysis should include the results of laboratory testing, cardiac catheterization, echocardiography, Holter monitoring, chest radiography, and magnetic resonance imaging. The multidisciplinary group should arrive at a consensus about surgical options, which may include cardiac transplantation, and about the timing of surgery.
b. The patient's cardiac rhythm should be assessed, with particular emphasis on the functioning of pacemakers (if present) and on the underlying cardiac rhythm in case of pacemaker failure.
c. An anesthetic plan should be developed in accordance with the patient's unique pathophysiology and anticipated response to anesthetic interventions. This is particularly important for patients with single ventricle and poor ventricular function, who may not tolerate myocardial depressants, positive-pressure ventilation, or loss of sinus rhythm.

2. General Operating Room Care

a. Large-bore intravenous access should be established, and provisions should be made for the rapid infusion of volume. A pressurized rapid-infusion system, capable of delivering at least 500 mL/min of warmed fluid or blood, is recommended. If massive bleeding occurs, rapid infusion can be established with the cardiopulmonary bypass machine: tubing from the venous reservoir is passed through a roller-pump head and connected to a large-bore venous access device. Heparin is administered, and large volumes can be transfused while preparing to institute bypass rapidly via the femoral route.
b. Multifunction external pacing, defibrillating, and cardioversion pads should be applied, and antiarrhythmic drugs should be immediately available. In pacemaker-dependent patients who have very slow or nonexistent underlying ventricular escape rhythms, preoperative insertion of a transvenous pacemaker should be considered.
c. In a preoperative discussion, the surgeon, anesthesiologist, and perfusionist should plan for emergency femoral bypass if necessary.
d. Preparations should be made to treat postoperative hemorrhage. Tranexamic acid, ε-aminocaproic acid, and aprotinin can reduce bleeding in these patients. Adequate blood products, including platelets, fresh frozen plasma, and cryoprecitipate, should be available. Cell salvage, with reinfusion of washed autologous red blood cells, is appropriate. During cardiopulmonary bypass, thromboelastography with heparinase and Celite® (International Technidyne Corporation, Edison, NJ) added to neutralize heparin and speed results may be useful to predict the need for blood products after cardiopulmonary bypass, particularly in patients with a baseline coagulopathy related to cyanosis.
e. Transesophageal echocardiography is indicated for congenital heart operations in infants and children and is equally applicable to congenital heart operations in adults.
f. Neurologic monitoring with transcranial Doppler ultrasonography (to assist in detecting and

limiting cerebral emboli), bispectral index electroencephalography and near-infrared spectroscopy may be helpful in minimizing neurologic complications.

SUMMARY

As the number of CHD repairs in adults continues to increase, these operations will be performed in a wider variety of institutions and systems. Unfortunately, not all of these centers will have an optimal environment for correcting CHD in adults. This type of surgery is best accomplished in a facility specifically designed for treating adults with CHD. Optimal care of these patients is provided by cardiologists who are trained and experienced in pediatric and adult cardiology, by surgeons who are trained and experienced in treating CHD, and by anesthesiologists who are experienced in caring for adults with CHD. Whatever the setting, cardiac anesthesiologists involved in these cases must be thoroughly aware of the anesthetic implications for the unique pathophysiology of each patient, and they must not rely on their "usual" expectations of either true pediatric CHD or acquired adult heart disease.

FURTHER READING

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