Chapter 16

Congenital Heart Disease in Adults

Victor C. Baum, MD

Advances in perioperative care for children with congenital heart disease (CHD) over the past several decades has resulted in an ever-increasing number of these children reaching adulthood with their cardiac lesions palliated or repaired. The first paper on adult CHD was published in 1973; the field has grown such that there is now a text devoted to it, and even a specialty society dedicated to it, the International Society for Adult Congenital Cardiac Disease (http://www.isaccd.org).[1] There are estimated to be about 32,000 new cases of CHD each year in the United States and 1.5 million worldwide. More than 85% of infants born with CHD are expected to grow to adulthood. It is estimated that there are more than 500,000 adults in the United States with CHD; 55% of these adults remain at moderate to high risk, and more than 115,000 have complex disease.[2,3] There are as many adults with CHD as there are children, and the number of adults will only continue to increase. These patients can be seen by anesthesiologists for primary cardiac repair, repair after a prior palliation, revision of repair due to failure or lack of growth of prosthetic material, or conversion of a suboptimal repair to a more modern operation (Box 16-1). In addition, these adults with CHD will be seen for all the other ailments of aging and trauma that require surgical intervention. Although it has been suggested that teenagers and adults can have repair of congenital cardiac defects with morbidity and mortality approaching that of surgery done during childhood, these data are limited and may reflect only a relatively young and acyanotic sampling.[4] Other data suggest that, in general, adults older than 50 years of age represent an excessive proportion of the early postoperative mortality encountered, and the number of previous operations and cyanosis are both risk factors. Risk factors for noncardiac surgery include heart failure, pulmonary hypertension, and cyanosis.[5]

These patients bring with them anatomic and physiologic complexities of which physicians accustomed to caring for adults may be unaware and medical problems

BOX 16-1 ***Indications for Cardiac Surgery in Adults with Congenital Heart Disease***

- Primary repair
- Total correction following palliation
- Revision of total correction
- Conversion of suboptimal obsolescent operation into more modern repair

associated with aging or pregnancy that might not be familiar to physicians used to caring for children. This problem has led to the establishment of the growing subspecialty of adult CHD. An informed anesthesiologist is a critical member of the team required to care optimally for these patients. A specific recommendation is that noncardiac surgery on adult patients with moderate to complex CHD be done at an adult congenital heart center (regional centers) with the consultation of an anesthesiologist experienced with adult CHD. In fact, one of the founding fathers of the subspecialty wrote: "A cardiac anesthesiologist with experience in CHD is pivotal. The cardiac anesthesiologist and the attending cardiologist are more important than the noncardiac surgeon."[2] Centers may find it helpful to delegate one attending anesthesiologist to be the liaison with the cardiology service to centralize preoperative evaluations and triage of patients to an anesthesiologist with specific expertise in managing patients with CHD, rather than random consultations with generalist anesthesiologists.

GENERAL NONCARDIAC ISSUES WITH LONG-STANDING CONGENITAL HEART DISEASE

A variety of organ systems can be affected by long-standing CHD; these are summarized in Table 16-1. Because congenital cardiac disease can be one manifestation of a multiorgan genetic or dysmorphic syndrome, all patients require a full review of systems and examination.[6,7]

CARDIAC ISSUES

The basic hemodynamic effects of an anatomic cardiac lesion can be modified by time and by the superimposed effects of chronic cyanosis, pulmonary disease, or the effects of aging. Although surgical cure is the goal, true universal cure, without residua, sequelae, or complications, is uncommon on a population-wide basis. Exceptions include closure of a nonpulmonary hypertensive patent ductus arteriosus (PDA) or atrial septal defect (ASD), probably in childhood. Although there have been reports of series of surgeries on adults with CHD, the wide variety of defects and sequelae from prior surgery make generalizations difficult, if not impossible. Poor myocardial function can be inherent in the CHD but can also be affected by long-standing cyanosis or superimposed surgical injury, including inadequate intraoperative myocardial protection. This is particularly true of adults who had their cardiac repair several decades ago when myocardial protection may not have been as good and when repair was undertaken at an older age. Postoperative arrhythmias are common, particularly when surgery entailed long atrial suture lines. Thrombi can be found in these atria,

Table 16-1 Potential Noncardiac Organ Involvement in Patients with Congenital Heart Disease

Potential Respiratory Implications

- Decreased compliance (with increased pulmonary blood flow or impediment to pulmonary venous drainage)
- Compression of airways by large, hypertensive pulmonary arteries
- Compression of bronchioles
- Scoliosis
- Hemoptysis (with end-stage Eisenmenger's syndrome)
- Phrenic nerve injury (prior thoracic surgery)
- Recurrent laryngeal nerve injury (prior thoracic surgery; very rarely from encroachment of cardiac structures)
- Blunted ventilatory response to hypoxemia (with cyanosis)
- Underestimation of $Paco_2$ by capnometry in cyanotic patients

Potential Hematologic Implications

- Symptomatic hyperviscosity
- Bleeding diathesis
- Abnormal von Willebrand factor
- Artifactually elevated prothrombin/partial thromboplastin times with erythrocytic blood
- Artifactual thrombocytopenia with erythrocytic blood
- Gallstones

Potential Renal Implication

- Hyperuricemia and arthralgias (with cyanosis)

Potential Neurologic Implications

- Paradoxic emboli
- Brain abscess (with right-to-left shunts)
- Seizure (from old brain abscess focus)
- Intrathoracic nerve injury (iatrogenic phrenic, recurrent laryngeal, or sympathetic trunk injury)

precluding immediate cardioversion. Bradyarrhythmias can be secondary to surgical injury to the sinus node or conducting tissue or can be a component of the cardiac defect.

The number of cardiac lesions and subtypes, together with the large number of contemporary and obsolescent palliative and corrective surgical procedures, makes a complete discussion of all CHD impossible. The reader is referred to one of the current texts on pediatric cardiac anesthesia for more detailed descriptions of these lesions, the available surgical repairs, and the anesthetic implications during primary repair.[8,9] Some general perioperative guidelines to caring for these patients are offered in Table 16-2.

Aortic Stenosis

Most aortic stenosis in adults is due to a congenitally bicuspid valve that does not become problematic until late middle age or later, although endocarditis risk is lifelong. Once symptoms (angina, syncope, near-syncope, heart failure) develop, survival is markedly shortened. Median survival is 5 years after the development of angina, 3 years after syncope, and 2 years after heart failure. Anesthetic management of aortic stenosis does not vary whether the stenosis is congenital (most common) or acquired.

Table 16-2 General Approaches to Anesthesia for Patients with Congenital Heart Disease

General

The best care for both cardiac and noncardiac surgery in adult patients with CHD is afforded in a center with a multidisciplinary team experienced in the care of adults with CHD and knowledgeable about both the anatomy and physiology of CHD and also the manifestations and considerations specific to adults with CHD.

Preoperative

- Review most recent laboratory data, catheterization, and echocardiogram and other imaging data. The most recent office letter from the cardiologist is often most helpful. Obtain and review these in advance.
- Drawing a diagram of the heart with saturations, pressures, and direction of blood flow often clarifies complex and superficially unfamiliar anatomy and physiology.
- Avoid prolonged fast if patient is erythrocytotic to avoid hemoconcentration.
- No generalized contraindication to preoperative sedation.

Intraoperative

- Large-bore intravenous access for repeat sternotomy and cyanotic patients.
- Avoid air bubbles in all intravenous catheters. There can be transient right-to-left shunting even in lesions with predominant left-to-right shunting (filters are available but will severely restrict ability to give volume and blood).
- Apply external defibrillator pads for repeat sternotomies and patients with poor cardiac function.
- Use appropriate endocarditis prophylaxis (orally or intravenously before skin incision).
- Consider antifibrinolytic therapy, especially for patients with prior sternotomy.
- Transesophageal echocardiography for cardiac operations.
- Modulate pulmonary and systemic vascular resistances as appropriate pharmacologically and by modifications in ventilation.

Postoperative

- Appropriate pain control (cyanotic patients have normal ventilatory response to hypercarbia and narcotics).
- Maintain hematocrit appropriate for arterial saturation.
- Maintain central venous and left atrial pressures appropriate for altered ventricular diastolic compliance or presence of beneficial atrial level shunting.
- Pao_2 may not increase significantly with the application of supplemental oxygen in the presence of right-to-left shunting.

Aortopulmonary Shunts

Depending on their age, adult patients may have had one or more of several aortopulmonary shunts to palliate cyanosis during childhood. These are shown in Figure 16-1. Although life saving, there were considerable shortcomings of these shunts in the long term. All were inherently inefficient, because some of the oxygenated blood returning through the pulmonary veins to the left atrium and ventricle would then return to the lungs through the shunt, thus volume loading the ventricle. It was difficult to quantify the size of the earlier shunts such as the Waterston (side-to-side ascending aorta to right pulmonary artery) and Potts (side-to-side descending aorta to left pulmonary artery). If too small, the patient was left excessively cyanotic; if too large, there was pulmonary overcirculation with the risk of developing pulmonary vascular disease. The Waterston, in fact, could on occasion result in a hyperperfused, hypertensive ipsilateral pulmonary artery and a hypoperfused contralateral pulmonary artery as it could direct flow to the ipsilateral side. There were also

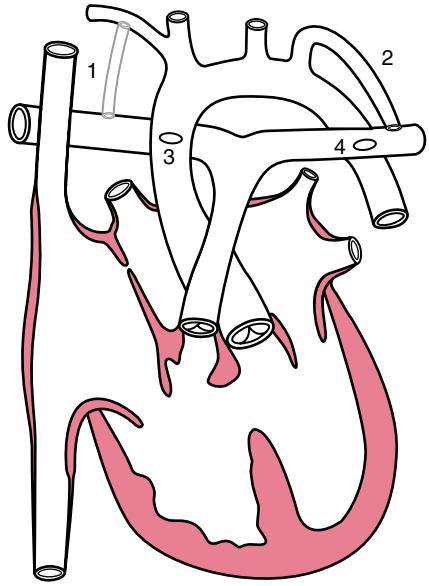

Figure 16-1 The various aortopulmonary anastomoses. The illustrated heart shows tetralogy of Fallot. The anastomoses are 1, modified Blalock-Taussig; 2, classic Blalock-Taussig; 3, Waterston (Waterston-Cooley); and 4, Potts. (Reprinted with permission from Baum VC: The adult with congenital heart disease. J Cardiothorac Vasc Anesth 10:261, 1996.)

Table 16-3 Aortopulmonary Shunts

Waterston	Ascending aorta → right pulmonary artery	No longer done
Potts	Descending aorta → left pulmonary artery	No longer done
Classic Blalock-Taussig	Subclavian artery → ipsilateral pulmonary artery	No longer done
Modified Blalock-Taussig	Gore-Tex tube subclavian artery → ipsilateral pulmonary artery	Current
Central shunt	Gore-Tex tube ascending aorta → main pulmonary artery	Current

surgical issues when complete repair became possible. Takedown of Waterston shunts often required a pulmonary arterioplasty to correct deformity of the pulmonary artery at the site of the anastomosis, and the posteriorly located Potts anastomoses could not be taken down from a median sternotomy. Patients with a classic Blalock-Taussig shunt almost always lack palpable pulses on the side of the shunt. Even if there is a palpable pulse (from collateral flow around the shoulder), blood pressure obtained from that arm will be artifactually low. Even after a modified Blalock-Taussig shunt (using a piece of Gore-Tex tubing instead of an end-to-side anastomosis of the subclavian and pulmonary arteries), there can be a blood pressure disparity between the arms. To ensure a valid measurement, preoperative blood pressure should be measured in both arms (Table 16-3).

BOX 16-2 ***Complications of Atrial Septal Defect in Adulthood***

- Paradoxic emboli
- Effort dyspnea
- Atrial tachyarrhythmias
- Right-sided failure with pregnancy
- Pulmonary hypertension
- ↑ Right-sided failure with ↓ left ventricular compliance with aging
- Mitral insufficiency

Atrial Septal Defect and Partial Anomalous Pulmonary Venous Return

There are several anatomic types of ASD. The most common type, and if otherwise undefined the presumptive type, is the secundum type located in the midseptum. The primum type at the lower end of the atrial septum is a component of the endocardial cushion defects, the most primitive of which is the common atrioventricular canal. The sinus venosus type, high in the septum near the entry of the superior vena cava, is almost always associated with partial anomalous pulmonary venous return, most frequently drainage of the right upper pulmonary vein to the low superior vena cava. An uncommon atrial septal-type defect is when blood passes from the left atrium to the right via an unroofed coronary sinus. Only secundum defects are considered, although the natural histories of all of the defects are similar (Box 16-2).

Because the symptoms and clinical findings of an ASD can be quite subtle and patients often remain asymptomatic until adulthood, ASDs represent approximately one third of all CHD discovered in adults. Although asymptomatic survival to adulthood is common, significant shunts ($\dot{Q}p/\dot{Q}s > 1.5:1$) will probably cause symptoms over time, and paradoxic emboli can occur through defects with smaller shunts. Effort dyspnea occurs in 30% by the third decade, and atrial flutter or fibrillation in about 10% by age 40. The avoidance of complications developing in adulthood provides the rationale for surgical repair of asymptomatic children. The mortality for a patient with an uncorrected ASD is 6% per year over 40 years of age, and essentially all patients over 60 years of age are symptomatic. Large nonrepaired defects can cause death from atrial tachyarrhythmias or right ventricular failure in 30- to 40-year-old patients. With the decreased left ventricular diastolic compliance accompanying the systemic hypertension or coronary artery disease that is common with aging, left-to-right shunting increases with age. Pulmonary vascular disease typically does not develop until after the age of 40, unlike ventricular or ductal level shunts, which can lead to it in early childhood. Mitral insufficiency can be found in adult patients and is significant in about 15% of adult patients. Paradoxic emboli remain a lifelong risk.

Late closure of the defect, after 5 years of age, has been associated with incomplete resolution of right ventricular dilation. Left ventricular dysfunction has been reported in some patients having defect closure in adulthood, and closure particularly in middle age may not prevent the development of atrial tachyarrhythmias or stroke.[10] Survival of patients without pulmonary vascular disease has been reported to be best if operated on before 24 years of age, intermediate if operated on between 25 and 41 years of age, and worst if operated on thereafter. However, more recent series have shown that even at ages over 40, surgical repair provides an overall survival and complication-free benefit compared with medical management.[11] Surgical morbidity in these patients is primarily atrial fibrillation, atrial flutter, or junctional rhythm.

Current practice is to close these defects in adults in the catheterization laboratory via transvascular devices if anatomically practical (Fig. 16-2). For example, there needs to be an adequate rim of septum around the defect to which the device can attach. Device closure is inappropriate if the defect is associated with anomalous pulmonary venous drainage. The indications for closure are the same as for surgical closure.[12]

An otherwise uncomplicated secundum ASD, unlike most congenital cardiac defects, is not associated with an increased endocarditis risk. Presumably this is because the shunt, although potentially large, is low pressure and unassociated with jet lesions of the endocardium.

Although some discussion is given to onset times with intravenous or inhalation induction agents, clinical differences are hard to notice with modern low-solubility volatile agents. Thermodilution cardiac output reflects pulmonary blood flow, which will be in excess of systemic blood flow. Pulmonary arterial catheters are not

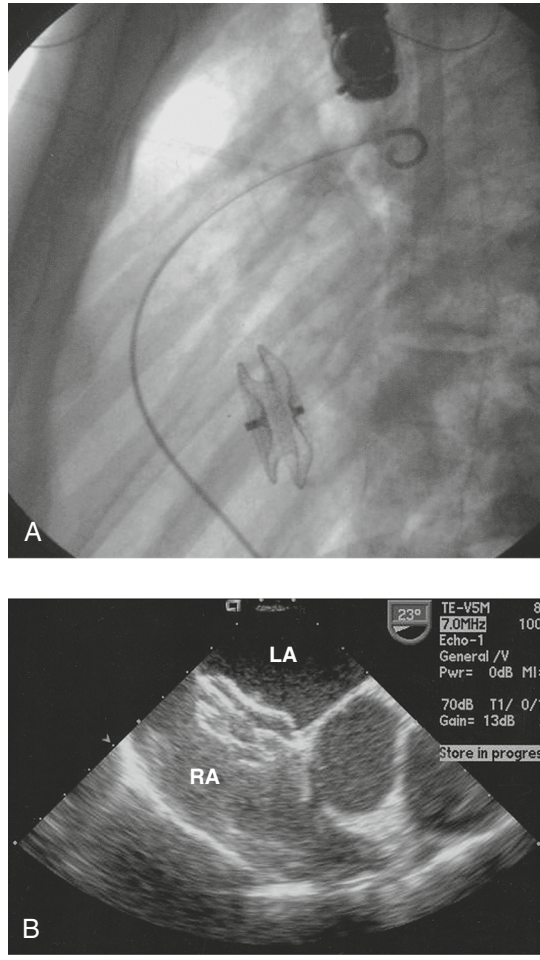

Figure 16-2 Closure of an atrial septal defect in an adult with use of a transvascular device (the Amplatzer septal occluder). **A,** Radiograph. **B,** Transesophageal echocardiogram. The device is clearly visualized spanning and occluding the atrial septal defect. RA = right atrium; LA = left atrium. (Courtesy of Dr. Scott Lim.)

routinely indicated. Patients generally do tolerate any appropriate anesthetic; however, particular care should be taken in patients with pulmonary arterial hypertension or right-sided failure.

Coarctation of the Aorta

Nonrepaired coarctation of the aorta in the adult brings with it significant morbidity and mortality. Mortality is 25% by age 20, 50% by age 30, 75% by age 50, and 90% by age 60. Left ventricular aneurysm, rupture of cerebral aneurysms, and dissection of a postcoarctation aneurysm all contribute to the excessive mortality. Left ventricular failure can occur in patients over 40 with nonrepaired lesions. If repair is not undertaken early, there is incremental risk for the development of premature coronary atherosclerosis. Even with surgery, coronary artery disease remains the leading cause of death 11 to 25 years after surgery. Coarctation is accompanied by a bicuspid aortic valve in the majority of patients. Although endocarditis of this abnormal valve is a lifelong risk, these valves often do not become stenotic until middle age or later. Coarctation can also be associated with mitral valve abnormalities (Box 16-3).

Aneurysms at the site of coarctation repair can develop years later, and restenosis as well can develop in adolescence or adulthood. Repair includes resection of the coarctation and end-to-end reanastomosis. Because this sometimes resulted in recoarctation when done in infancy, for many years a common repair was the Waldhausen or subclavian flap operation, in which the left subclavian artery is ligated and the proximal segment opened and rotated as a flap to open the area of the coarctation. Aneurysms in the area of repair are a particular concern in adolescents and adults after coarctectomy. Persistent systemic hypertension is common after coarctation repair. Adult patients require continued periodic follow-up for hypertension. A pressure gradient of 20 mmHg or more (less in the presence of extensive collaterals) is an indication for treatment. Recoarctation can be treated surgically or by balloon angioplasty with stenting.[13] Surgical repair of recoarctation or aneurysm in adults is associated with increased mortality and can be associated with significant intraoperative bleeding. It requires lung isolation for optimal surgical exposure and placement of an arterial catheter in the right arm.

Half of patients operated on after age 40 have persistent hypertension, and many of the remainder have an abnormal hypertensive response to surgery. Long-term survival is worse for patients having repair later in life. Patients older than 40 having repair have a 15-year survival of only 50%.

Blood pressure should be obtained in the right arm unless pressures in the left arm or legs are known to be unaffected by residual or recurrent coarctation. Postoperative hypertension is common after repair of coarctation and often requires treatment for some months. Postoperative ileus is also common, and patients should be maintained NPO for about 2 days.

BOX 16-3 ***Complications of Aortic Coarctation in Adulthood***

- Left ventricular failure
- Premature coronary atherosclerosis
- Rupture of cerebral aneurysm
- Aneurysm at site of coarctation repair
- Complications of associated bicuspid aortic valve
- Exacerbation of hypertension during pregnancy

Eisenmenger's Syndrome

Eisenmenger described a particular type of large VSD with dextroposition of the aorta. In a general way, the term *Eisenmenger's syndrome* has come to describe the clinical setting in which a large left-to-right cardiac shunt results in the development of pulmonary vascular disease. Although early on the pulmonary vasculature remains reactive, with continued insult pulmonary hypertension becomes fixed and does not respond to pulmonary vasodilators. Ultimately, the level of pulmonary vascular resistance is so high that the shunt reverses and becomes right-to-left. The development of pulmonary vascular disease is dependent on shear rate. Lesions with high shear rates, such as a large VSD or a large PDA, can result in pulmonary hypertension in early childhood. Lesions such as an ASD with high pulmonary blood flow but low pressure may not result in pulmonary vascular disease until late middle age. Pulmonary vascular disease progression is also accelerated in patients living at altitude.

Eisenmenger physiology is compatible with survival into adulthood. However, reported rates of survival after diagnosis vary, probably based on the relatively long life expectancy and variability in the time of diagnosis. Cantor and coworkers reported median survival to 53 years but with wide variation. Others reported survival of 80% at 10 years after diagnosis and 42% at 25 years; or survival of 77% at 5 years and 58% at 10 years. Syncope, increased central venous pressure, and arterial desaturation to less than 85% are all associated with poor short-term outcome.[14,15] Most deaths are sudden cardiac deaths. Other causes of death include heart failure, hemoptysis, brain abscess, thromboembolism, and complications of pregnancy and noncardiac surgery. These patients face potentially significant perioperative risks. Findings of Eisenmenger's syndrome are summarized in Table 16-4.

Surgical closure of cardiac defects with fixed pulmonary vascular hypertension is associated with very high mortality. Lung or heart-lung transplantation is a surgical alternative. Although there are several surgical series reporting survival after heart-lung or single- or double-lung transplantation performed for primary pulmonary hypertension, it is unclear if this cohort of patients is similar to patients with Eisenmenger physiology.

Fixed pulmonary vascular resistance precludes rapid adaptation to perioperative hemodynamic changes. Changes in systemic vascular resistance are mirrored by changes in intracardiac shunting. A decrease in systemic vascular resistance is accompanied by increased right-to-left shunting and a decrease in systemic oxygen saturation. Systemic

Table 16-4 Findings in Eisenmenger's Syndrome

- *Physical examination:* loud pulmonic component of the second heart sound, single or narrowly split second heart sound, Graham Steell murmur of pulmonary insufficiency, pulmonic ejection sound ("click")
- *Chest radiography:* decreased peripheral pulmonary arterial markings with prominent central pulmonary vessels ("pruning")
- *ECG:* Right ventricular hypertrophy
- Impaired exercise tolerance
- Exertional dyspnea
- Palpitations (often due to atrial fibrillation or flutter)
- Complications from erythrocytosis/hyperviscosity
- Hemoptysis from pulmonary infarction, rupture of pulmonary vessels or aortopulmonary collateral vessels
- Complications from paradoxical embolization
- Syncope from inadequate cardiac output or arrhythmias
- Heart failure (usually end stage)

vasodilators, including regional anesthesia, need to be used with caution, and close assessment of intravascular volume is important. Epidural analgesia has been used successfully in patients with Eisenmenger physiology, but the local anesthetic needs to be delivered slowly and incrementally with close observation of blood pressure and oxygen saturation. Postoperative postural hypotension can also increase the degree of right-to-left shunting, and these patients should change position slowly.

Placement of pulmonary artery catheters in these patients is problematic for a variety of reasons and of less utility than might be expected. Pulmonary arterial hypertension is a risk factor for pulmonary artery rupture from a pulmonary artery catheter. Rupture is particularly worrisome in these cyanotic patients who can also have hemostatic deficits associated with erythrocytosis. Abnormal intracardiac anatomy and right-to-left shunting can make successful passage into the pulmonary artery difficult without fluoroscopy. Relative resistances of the pulmonary and systemic beds are reflected in the systemic oxygen saturation, readily measured by pulse oximetry, so measures of pulmonary artery pressure are not required. In addition, in the presence of right-to-left shunting, thermodilution cardiac outputs do not accurately reflect systemic output. Thus, the value of pulmonary artery catheters in these patients is minimal at best, and they essentially are never indicated. The one potential exception is the patient with an ASD who is at risk to develop right ventricular failure if suprasystemic right ventricular pressure develops.

Fixed pulmonary vascular resistance is by definition unresponsive to pharmacologic manipulation. That said, it would seem prudent to avoid factors known to increase pulmonary vascular resistance, including cold, hypercarbia, acidosis, hypoxia, and α-adrenergic agonists. Although the last of these is commonly listed to be avoided, it seems that in the presence of pulmonary vascular disease due to intracardiac shunting, the systemic vasoconstrictive effects predominate and systemic oxygen saturation increases.

Nerve blocks offer an attractive alternative to general anesthesia if otherwise appropriate. If patients have general anesthesia, consideration should be given to postoperative observation in an intensive or intermediate care unit. Because of the increased perioperative risk, patients should be observed overnight, particularly if they have not had recent surgery or anesthesia, because their responses will be unknown. Ambulatory surgery is possible for patients having uncomplicated minor surgery with sedation or nerve block.

Patent Ductus Arteriosus

Beyond the neonatal period, spontaneous closure of a PDA is uncommon. The risk of a long-standing moderate to large PDA is volume overloading of the left atrium and left ventricle with the risk of development of pulmonary vascular disease. With time, the ductus can become calcified or aneurysmally dilated with a risk of rupture. Ductal calcification or aneurysm increases the risk of surgery, which rarely requires cardiopulmonary bypass. Nonrepaired, the natural history is for one third of patients to die of heart failure, pulmonary hypertension, or endocarditis by 40 years of age and two thirds by age 60. Although small PDAs are of no hemodynamic consequence, even small PDAs carry relatively high endocarditis risk. Surgical closure should be considered for all adults with PDA, and transvascular closure by means of one of several devices is possible.[16]

Tetralogy of Fallot

The classic description of tetralogy of Fallot includes (1) a large, nonrestrictive malaligned VSD, with (2) an overriding aorta, (3) infundibular pulmonic stenosis, and (4) consequent right ventricular hypertrophy, all derived from an embryonic

anterocephalad deviation of the outlet septum. However, there is a spectrum of disease, with more severe defects including stenosis of the pulmonary valve, stenosis of the pulmonary valve annulus, or stenosis and hypoplasia of the pulmonary arteries in the most severe cases. Pentalogy of Fallot refers to the addition of an ASD. With advances in genetics, up to one third or more of cases of tetralogy have been ascribed to one of several genetic abnormalities, including trisomy 21, the 22q11 microdeletion, the genes *NKX 2.5* and *FOG 2.4*, and others. Tetralogy of Fallot is the most common cyanotic lesion encountered in the adult population. Nonrepaired or nonpalliated, approximately 25% of patients survive to adolescence, after which the mortality is 6.6% per year. Only 3% survive to age 40.[17] Unlike children, teenagers and adults with tetralogy do not develop "tet spells." Long-term survival with a good quality of life is expected after repair. The 32- to 36-year survival has been reported to be 85% to 86%, although symptoms, primarily arrhythmias and decreased exercise tolerance, occur in 10% to 15% at 20 years after the primary repair (Box 16-4).

It is uncommon to encounter an adolescent or adult with nonrepaired tetralogy. However, it can be encountered in immigrants or in patients whose anatomic variation was considered to be inoperable when they were children. In tetralogy, the right ventricle "sees" the obstruction from the pulmonic stenosis. Pulmonary vascular resistance is typically normal to low. Right-to-left shunting is unaffected by attempts at modulating pulmonary vascular resistance. Shunting is minimized, however, by pharmacologically increasing systemic vascular resistance. Increases in the inotropic state of the heart increase the dynamic obstruction at the right ventricular infundibulum and worsen right-to-left shunting. β-Blockers are often used to decrease inotropy. Although halothane was the historic anesthetic of choice in children with tetralogy due to its myocardial depressant effects and ability to maintain systemic vascular resistance, current practice is to use sevoflurane, without undue consequence from a reduction in systemic vascular resistance. Anesthetic induction in adults can easily be achieved with narcotics.[18]

Patients require closure of the VSD and resolution of the pulmonic stenosis. Although current practice is to repair the VSD through the right atrium in an effort to maintain competence of the pulmonary valve and limit any ventriculotomy, older patients will likely have had repair via a right ventriculotomy. A large right ventriculotomy increases the risks of arrhythmias and sudden death. Patients who have had a right ventriculotomy will have an obligate right bundle-branch block pattern on the ECG. However, unlike the more usual bundle-branch block in adults, this represents disruption of the His-Purkinje system only in the right ventricular outflow, in the area of the right ventricular incision. Because the vast majority of His-Purkinje conduction is intact, it does not carry increased risk for the development of complete heart block. These patients can have an abnormal response to exercise.

BOX 16-4 ***Risk Factors for Sudden Death after Repair of Tetralogy of Fallot***

- Repair requiring ventriculotomy
- Older age at repair
- Severe left ventricular dysfunction
- Postoperative right ventricular hypertension (residual outflow tract obstruction)
- Wide-open pulmonary insufficiency
- Prolongation of the QRS

Some patients require repair of pulmonic stenosis by placement of a transannular patch, with obligate residual pulmonary insufficiency. Isolated mild to moderate pulmonary insufficiency is generally well tolerated, but in the long term it can contribute to right ventricular dysfunction with a risk of ventricular tachycardia and sudden death. Atrial tachyarrhythmias occur in about one third of adults late after repair and can contribute to late morbidity.[19] The development of atrial flutter or atrial reentrant tachycardia is often a harbinger of hemodynamic compromise. The substrate is usually an atrial surgical scar and the trigger is atrial dilation, such as from tricuspid insufficiency with right ventricular dysfunction. The mechanism for the development of ventricular arrhythmias is presumably the same, namely dilation superimposed on surgical scar.

In some cases, the right ventricular outflow tract patch needs to be extended onto the branch pulmonary arteries to relieve obstruction. Patients with abnormal coronary arteries may have required repair using a right ventricle-to-pulmonary-artery conduit to avoid doing a right ventriculotomy in the area of the coronary artery. Repair at a younger age (<12 years) results in better postoperative right ventricular function. Because there is an unrestrictive VSD, in the nonrepaired adult systemic hypertension developing in adult life imposes an additional load on both ventricles, not just the left. The increase in systemic vascular resistance decreases right-to-left shunting and diminishes cyanosis but at the expense of right ventricular or biventricular failure.

Sudden death or ventricular tachycardia requiring treatment can occur in up to 5.5% of postoperative patients over 30 years, often years postoperatively. The foci for these arrhythmias are typically in the right ventricular outflow tract in the area that has had surgery, and they can be ablated in the catheterization laboratory. Older age at repair, severe left ventricular dysfunction, postoperative right ventricular hypertension from residual or recurrent outflow tract obstruction, wide-open pulmonary insufficiency, and prolongation of the QRS are all predictors of sudden death.[20] Premature ventricular contractions and even nonsustained ventricular tachycardia are not rare but do not seem to be associated with sudden death, making appropriate treatment options difficult. It has been suggested that QRS prolongation to longer than 180 milliseconds is a risk factor. This marker, although highly sensitive, has a low positive predictive value. The impact of this risk factor in the current group of younger patients who have not had ventriculotomies is unclear, as their initial postoperative QRS durations are shorter than in patients who had a right ventriculotomy.

Although for many years it was thought that moderate to severe pulmonary insufficiency in these patients was well tolerated, it has become apparent from a number of series that right ventricular dysfunction and both atrial and ventricular arrhythmias can be common long-term sequelae. For this reason, patients with symptomatic pulmonary insufficiency from a transannular patch or aneurysm formation at the site of a right ventricular outflow tract patch can require reoperation to replace a widely incompetent pulmonary valve with a bioprosthetic valve with or without a tricuspid annuloplasty. Interestingly, the incidence of atrial arrhythmias may not be diminished when adult patients have a pulmonary valve placed, although the incidence of ventricular arrhythmias is decreased. Right ventricular dysfunction improves in a variable number of adults, suggesting that pulmonary valve placement be done sooner rather than later. There is very early experience with a pulmonary valve that can be inserted transvenously in the catheterization laboratory, although currently it is appropriate only for a limited range of pulmonary artery sizes.

Additional possible late-term complications include residual VSD, patch dehiscence, progressive aortic insufficiency, left ventricular dysfunction from surgical injury to an anomalous coronary artery or long-standing preoperative cyanosis, and heart block from VSD closure (uncommon today). Because patients who have had

repairs using a conduit require multiple sternotomies and the valved conduit tends to lie immediately behind and in close proximity to the sternum, sternotomy carries with it significant potential risk for laceration of the conduit. On occasion, the femoral vessels are cannulated for bypass prior to sternotomy.

Most adult patients require reoperation to repair the right ventricular outflow tract or to insert or replace a valve in the pulmonic position. Other reasons for reoperation include repair of an outflow tract aneurysm at the site of a patch, repair of a residual VSD, or repair of an incompetent tricuspid valve. These patients often have diminished right ventricular diastolic compliance and require higher-than-normal central venous pressure. Postoperative management includes minimizing pulmonary vascular resistance and maintaining central venous pressure. Patients often require treatment post-bypass with an inotrope and afterload reduction.[21]

Transposition of the Great Arteries (D-Transposition)

In D-transposition of the great arteries, there is a discordant connection of the ventricles and the great arteries. The aorta (with the coronary arteries) arises from the right ventricle, and the pulmonary artery arises from the left ventricle. Thus, the two circulations are separate. Postnatal survival requires interchange of blood between the two circulations, typically via a patent foramen ovale and/or a PDA. With a 1-year mortality approximating 100%, all adults with D-transposition have had some type of surgical intervention. Adults will have had atrial-type repairs (Mustard or Senning), whereas children born after the mid-1980s will have had repair by arterial switch (the Jantene operation). Some will also have had repair of D-transposition with a moderate to large VSD by means of a Rastelli operation.

Atrial repairs function by redirecting systemic venous blood to the left ventricle (and thence to the transposed pulmonary artery) and pulmonary venous blood to the right ventricle (and thence to the aorta). The Mustard operation uses an intra-atrial conduit of native pericardium, whereas the Senning operation uses native atrial tissue to fashion the conduit. The arterial switch operation transposes transected aorta and pulmonary artery such that they now arise above the appropriate ventricle. This operation also requires transposing the coronary arteries from the aorta to the pulmonary root, which, following the procedure, becomes the aortic root. The Rastelli procedure closes the VSD on a bias such that the left ventricle empties into the aorta and connects the right ventricle to the pulmonary artery by means of a valved conduit.

Atrial repairs result in a systemic right ventricle, and these patients consistently have abnormal right ventricular function that can be progressive with a right ventricular ejection fraction of about 40%. Mild tricuspid insufficiency is common, but severe tricuspid insufficiency suggests the development of severe right ventricular dysfunction. There is an 85% to 90% 10-year survival with these operations, but by 20 years survival is less than 80%. Over 25 years, about half develop moderate right ventricular dysfunction and one third develop severe tricuspid insufficiency. Although it always remains abnormal, it has been suggested that earlier surgery minimizes right ventricular dysfunction. Because of the incidence of right ventricular dysfunction, some patients with atrial repairs have been converted to an arterial switch, following preparation of the left ventricle by a pulmonary artery band to prepare it to tolerate systemic arterial pressure.

Atrial repairs bring an incidence of late electrophysiologic sequelae including sinus node dysfunction (bradycardia), junctional escape rhythms, atrioventricular block, and supraventricular arrhythmias. Atrial flutter occurs in 20% of patients by age 20 with half having progressive sinus node dysfunction by that time. On occasion these tachyarrhythmias can result in sudden death, presumably from 1:1 conduction producing

ventricular fibrillation. The loss of sinus rhythm in the face of right ventricular (the systemic ventricle) dysfunction can also contribute to late sudden death. The risk of late death after an atrial repair is almost three times higher if there is an associated VSD. The incidence of tachyarrhythmias does decrease, however, after the tenth postoperative year.

An arterial switch operation can be done after a failed atrial repair in adults, but the outcome is generally poor. It is suggested that younger patients do better.

Very-long-term outcome after the arterial switch procedure is still not known. It does appear that there is essentially no mortality after 5 years postoperatively, and late surgical reintervention is mostly due to supravalvular pulmonic stenosis.[22] Although many of these children have abnormal resting myocardial perfusion, up to 9% can have evidence of exercise-induced myocardial ischemia. The implication for the development of premature coronary artery disease in adulthood is not known, and there is also some concern about the ultimate function of the neoaortic valve.

After an atrial or a Rastelli repair, pregnancy and delivery are generally well tolerated, although right ventricular failure and deterioration in functional capacity can occur. There is an increased incidence of prematurity and small-for-date infants in these women.

Ventricular Septal Defects

More than 75% of small and moderate VSDs close spontaneously during childhood by a gradual ingrowth of surrounding septum. Of those that close spontaneously, almost all have closed by 10 years of age. Other mechanisms for natural closure include closure by tricuspid valve tissue, closure by prolapsed aortic leaflet, and closure by endocarditis. Some VSDs result in the development of aortic insufficiency in adults from prolapse of the aortic valve into the defect. Although the risk of endocarditis is ongoing, there is no hemodynamic risk of a small VSD in the adult. If pulmonary vascular disease is present, it can progress if closure of a large VSD is delayed.

Although some studies have reported possible ventricular dysfunction years after surgical repair, these are older reports and patients were operated on later than by current standards. It does appear, though, that the ventricle successfully remodels from chronic volume overload if surgical correction is done by 5 years of age and perhaps up to 10 to 12 years of age.

Although some discussion is given to onset times with intravenous or inhalation induction agents, clinical differences are hard to notice with modern low-solubility volatile agents. Thermodilution cardiac output reflects pulmonary blood flow, which will be in excess of systemic blood flow. Pulmonary arterial catheters are not routinely indicated. In the patient with a moderate or large left-to-right shunt, low inspired oxygen and moderate hypercarbia avoid intraoperative decreases in pulmonary vascular resistance with pulmonary overcirculation and left ventricular dilation. However, unlike children, it would be rare to encounter adults with large left-to-right shunts. Adults with nonrepaired lesions would have either small shunts or would have had large shunts that caused Eisenmenger physiology.

SUMMARY

- Due to surgical successes in treating congenital cardiac lesions, there are currently as many or more adults than children with congenital heart disease (CHD).
- These patients may require cardiac surgical intervention for primary cardiac repair, repair after prior palliation, revision of repair due to failure or lack of growth of prosthetic material, or conversion of a suboptimal repair to a more modern operation.

- These patients will be encountered by noncardiac anesthesiologists for a vast array of ailments and injuries requiring surgery.
- If at all possible, noncardiac surgery on adult patients with moderate to complex CHD should be done at an adult congenital heart center with the consultation of an anesthesiologist experienced with adult CHD.
- Delegation of one anesthesiologist as the liaison with the cardiology service for preoperative evaluation and triage of adult CHD patients will be helpful.
- All relevant cardiac tests and evaluations should be reviewed in advance.
- Sketching out the anatomy and path(s) of blood flow is often an easy and enlightening aid in simplifying apparently very complex lesions.

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