Chapter 11
Common Adult Congenital Heart Disease Issues

Erica O. Miller and James P. Eichelberger

Epidemiology

- Advances in diagnosis, surgical techniques, and medical management have dramatically altered the survival of children born with CHD and consequently the demographics of the CHD population.
- Over the last several decades, survival to adulthood for babies born with CHD has improved to greater than 85% [1].
- Heart failure, sudden death, arrhythmias, and vascular complications account for approximately 80% of deaths in all ACHD patients [2].
- Since 1990 myocardial infarction has been the leading cause of death in patients with acyanotic CHD, emphasizing the importance of prevention and management of atherosclerotic cardiovascular disease in patients with CHD [3].
- There are more adults living with congenital heart disease than children. In 2010, there were an estimated 1.4 million adults and 1 million children with CHD in the United States [4].
- The adult CHD population is growing more quickly than the pediatric CHD population. Data from the United States are unavailable, but in Canada between 1985 and 2005, the number of children living with CHD increased by 22%, while the number of adults living with severe CHD increased by 85% [5].
- In 2004, there were 46,500 hospitalizations for cardiac and circulatory congenital anomalies, with an aggregate cost of $1.4 million [6].

E. O. Miller (✉) · J. P. Eichelberger
University of Rochester Medical Center/Cardiology, Rochester, NY, USA
e-mail: Erica_Miller@URMC.Rochester.edu; James_eichelberger@urmc.rochester.edu

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Access to Healthcare

- One of the most important tasks of the adult cardiology consultant providing care for patients with CHD is improving access to care (Table 11.1). For a variety of psychosocial, financial, and infrastructural reasons, many young adults with congenital heart disease become disconnected with cardiology care and often with the healthcare system.
- According to 2008 American College of Cardiology (ACC) and American Heart Association (AHA) guidelines on the management of ACHD patients, all ACHD patients, even those with simple CHD such as isolated aortic valve disease or a small atrial septal defect, should be seen at least once in an ACHD center (Class I Recommendation). However, fewer than 30% of adults with CHD are seen in a specialized ACHD center [7, 8].

Transition from Pediatric to Adult Approach to Care

- Medical transition is the process of patients moving from a pediatric system of healthcare to an adult one.
- Transition involves both the process of pediatric patients becoming adults responsible for their healthcare decisions, depending on their capacity, and transferring care to adult healthcare providers.
- Goals of the transition process include optimizing health, minimizing disruptions in care, and helping youths reach their full potential [9].
- Transition planning has demonstration improvements in medical complications, cost, quality of life, functional status, perceived health status, adherence,

Table 11.1 Summary of consultant's role in improving access to care for ACHD patients [8]

ACC/AHA guideline	Role of cardiology consultant
Academic adult cardiology and cardiac surgery centers should have access to a regional ACHD center for consultation and referral	• Be aware of regional ACHD center • Contact ACHD center for consultation • Refer patients to ACHD center
All ACHD patients should be seen at least once in an ACHD center	• Document need for follow-up with an ACHD specialist in each patient's chart • Discuss the need for follow-up with an ACHD specialist with each patient
Diagnostic and interventional procedures for adults with complex and moderate CHD should be performed at a regional ACHD center	• Refer patients to ACHD center for diagnostic and interventional procedures
Surgical procedures requiring general anesthesia or conscious sedation should be performed at a regional ACHD center by an anesthesiologist familiar with ACHD	• Refer patients to ACHD center when general anesthesia or conscious sedation is required
Patients with complex or high-risk CHD should be transferred to an ACHD center for urgent or acute noncardiac problems	• Assist primary team in transfer of patients with complex or high-risk CHD to regional ACHD center when appropriate

continuity, and patient experience in chronic illnesses such as diabetes, cystic fibrosis, and juvenile idiopathic arthritis [9].
- In 2004, it was estimated that only 48% of adolescents with CHD underwent successful transition [10].
- Cardiology consultants can improve the transition process by recommending follow-up with an ACHD specialist to the patient and healthcare team. One series demonstrated that documentation in the chart recommending follow-up with an ACHD specialist was associated with a successful transition process (odds ratio 8, 95% confidence interval 4.72–16.41) as well as patient belief that follow-up should be with an ACHD specialist [10].

Hypertension

- ACHD patients may be particularly vulnerable to the effects of hypertension due to underlying altered hemodynamics.
- Blood pressure should be monitored and hypertension treated with a similar approach as for those without CHD, with special considerations listed in Table 11.2.

Dyslipidemia

- Lipids should be monitored and dyslipidemia treated for primary and secondary prevention of atherosclerotic cardiovascular disease in patients with ACHD similar to the general population, with special considerations listed in Table 11.3.

Table 11.2 Special considerations for the management of hypertension in patients with ACHD

ACHD population	Special hypertension considerations
Single ventricle Systemic right ventricle	Particularly sensitive to increased afterload Focus on afterload reduction Consider lower blood pressure goal [11]
Marfan syndrome with aortopathy	Use beta-blockers to reduce the rate of aortic dilation [12] Add angiotensin receptor blocker as tolerated
Coarctation of the aorta	Angiotensin-converting enzyme (ACE) inhibitors can precipitate renal failure in the setting of severe coarctation of the aorta due to restricted renal artery flow mimicking renal artery stenosis [11] Intervention (surgical or percutaneous) is recommended for coarctation with peak-to-peak gradient greater than or equal to 20 mmHg or at lower gradients with radiological evidence of significant collaterals [8]
Cyanotic heart disease	Risk for nephropathy Use caution with ACE inhibitors, angiotensin receptor blockers, and diuretics (Class I, Level of Evidence C) [11]
Eisenmenger physiology	Risk for increased right-to-left shunting with systemic vasodilators Use hydralazine with caution [11]

Table 11.3 Special considerations for the management of dyslipidemia in patients with ACHD [11]

ACHD population	Special dyslipidemia considerations
Transposition of the great arteries with a history of arterial switch operation	Coronary arteries have been translocated Translocated coronary arteries have abnormal vasoreactivity and increased intimal thickness Consider earlier and aggressive lipid management due to higher risk for coronary events (Class IIa, Level of Evidence: C)
Coarctation of the aorta	Increased risk for coronary events through unclear mechanisms, potentially hypertension or primary vasculopathy Consider earlier and more aggressive lipid management

Table 11.4 Indications for cardiac transplantation in patients with CHD [13]

New York Heart Association functional class IV HF not amenable to palliative or corrective surgery
Severe symptomatic cyanotic heart disease not amenable to palliation
Post-Fontan procedure with refractory HF, persistent protein-losing enteropathy, and/or plastic bronchitis despite optimal medical and surgical therapy
Pulmonary hypertension with the potential risk of developing fixed, irreversible elevation of pulmonary vascular resistance (PVR) that could preclude heart transplantation in the future

Heart Failure and Transplantation

- Unfortunately, medications demonstrating improved outcomes in non-ACHD patients with heart failure have not shown the same benefits in ACHD patients.
- Patients with moderate to complex CHD and heart failure should be managed by ACHD specialists (Class I, Level of Evidence: C) [11].
- Indications for cardiac transplantation in CHD patients are listed in Table 11.4.
- ACHD patients have longer transplant waitlist times than patients without CHD [14].
- When compared with non-ACHD heart transplant patients, ACHD heart transplant patients have higher 1-year mortality but lower 5- and 10-year mortality when adjusted for 1-year mortality [14].

Arrhythmias

- Arrhythmias are a major cause of morbidity and mortality in the ACHD population.
- ACHD patients may have arrhythmias related to their underlying anatomy, procedures, or both.
- Intra-atrial reentrant tachycardia (IART), including typical atrial flutter and often unusual circuits, is the most common arrhythmia in adults over the age of 40 years with CHD. Since atypical flutter is common and can have different flutter rates and flutter wave morphologies, it may be mistaken for sinus rhythm, so a high index of suspicion is needed [11].

- The more common arrhythmias and associated native and surgical anatomy are reviewed in Table 11.5.
- The most important step in management of arrhythmias in patients with ACHD is referral to an ACHD center for expert consultation in necessary further evaluation, medical management, and catheter and surgical interventions.
- Patients with ACHD are generally considered to be at higher risk for thromboembolic complications of atrial fibrillation, so anticoagulation is recommended even in the absence of traditional stroke risk factors such as those represented in the CHADS2VASC scoring algorithm. Warfarin is generally used since direct acting oral anticoagulants have not been well studied in patients with ACHD.
- Other guidelines for management of arrhythmias in ACHD patients are listed in Table 11.6.

Table 11.5 Trends in arrhythmia occurrence among patients with CHD [11]

Arrhythmia	At-risk population
Intra-atrial reentrant tachycardia (IART) or atrial flutter	Mustard or Senning repair for D-transposition of the great arteries (TGA) Fontan procedure for single ventricle physiology Atriotomy for atrial septal defect (ASD) or tetralogy of Fallot (TOF)
Atrial fibrillation	Mitral valve disease Congenital aortic stenosis (AS) Palliation procedure for single ventricle physiology
Ventricular tachycardia (VT)	TOF Ventriculotomy Ventricular septal defect (VSD) patch Congenital AS D-TGA or L-TGA Severe Ebstein's anomaly Single ventricle physiology Pulmonary arterial hypertension
Atrioventricular (AV) block	L-TGA Atrioventricular septal defect

Table 11.6 ACC/AHA guidelines for the management of arrhythmias in patients with ACHD [8, 11]

Class I
1. Effective anticoagulation (generally with warfarin) is recommended in older ACHD patients with sustained atrial fibrillation, whether or not those patients meet the usual criteria for anticoagulation of patients with atrial fibrillation in acquired heart disease (e.g., CHADS2VASC score) (Level of Evidence: C) [11]
Class IIa
1. It is reasonable to recommend the use of an implantable cardioverter defibrillator for any patient who has had a cardiac arrest or experienced an episode of hemodynamically significant or sustained VT (Level of Evidence: C) 2. Pacemaker implantation can be beneficial in ACHD patients with bradyarrhythmias and may be helpful in overdrive pacing in patients with difficult-to-control tachyarrhythmias (Level of Evidence: B) [8]
Class IIb
1. Pacemaker implantation may be beneficial for asymptomatic adult patients with resting heart rates of less than 40 beats per minute or abrupt pauses in excess of 3 s (Level of Evidence: C) [8]

Table 11.7 ACC/AHA guidelines for the evaluation and management of liver disease [11]

Class I
1. Serial evaluation of liver function should be performed for all patients with a history of previous palliation with the Fontan procedure (Level of Evidence: B)
2. All ACHD patients with a history of previous surgical palliation of CHD before 1992 should undergo screening for hepatitis C (Level of Evidence: B)
3. There is an increased frequency of gallstones and need for cholecystectomy in ACHD, especially in the cyanotic and Fontan populations. Vigilance should be high for diagnosis (Level of Evidence: B)

Liver Disease

- Patients with right-sided heart failure are at risk for congestive hepatopathy.
- Patients with a history of Fontan palliation procedure are at risk for liver disease including cirrhosis. The mechanism for this is not clearly defined but may include hypoxia, low cardiac output, perioperative insults, and elevated central venous pressures [11].
- Additional guidelines for liver disease are listed in Table 11.7.

Hyperviscosity

- Patients with cyanosis develop secondary erythrocytosis as a physiologic compensation mediated by erythropoietin to improve tissue oxygenation.
- Typically, patients with cyanosis develop a new equilibrium with a higher hematocrit and are considered compensated, especially if they are iron replete. Other patients have decompensated erythrocytosis with rapidly increasing hematocrit and symptoms of hyperviscosity (Table 11.8) [15].
- Symptoms of hyperviscosity include headache, fatigue, dizziness, visual disturbances, paresthesias, irritability, myalgias, anorexia, loss of concentration, and muscle weakness.
- Volume depletion can mimic and worsen symptoms of hyperviscosity and must be ruled out and treated.
- The symptoms of iron deficiency are also similar to those of hyperviscosity, and iron deficiency also worsens hyperviscosity due to decreased oxygen carrying capacity and increased rigidity of iron-deficient microspherocytes [15].
- ACC/AHA guidelines for the use of phlebotomy are listed in Table 11.9.
- Phlebotomy is only recommended for symptomatic hyperviscosity as frequent phlebotomy increases the risk of stroke for reasons that are poorly understood, as well as iron deficiency.
- Phlebotomy may also be considered in select patients undergoing noncardiac surgery who are at significantly increased risk of bleeding, as phlebotomy may decrease perioperative bleeding risk.
- Phlebotomy is performed by withdrawing blood and replacing the removed volume with isotonic saline [16].

Table 11.8 Compensated and decompensated erythrocytosis

Compensated erythrocytosis	Decompensated erythrocytosis
Stable hematocrit	Unstable, increasing hematocrit
Iron replete	Iron deficiency may be present
Absent or mild hyperviscosity symptoms	Hyperviscosity symptoms present

Table 11.9 ACC/AHA guidelines for the use of phlebotomy in patients with cyanotic CHD [8]

Class I
Therapeutic phlebotomy is indicated for: • Hemoglobin >20 g/dL and hematocrit >65% • Symptoms of hyperviscosity: headache, increasing fatigue, or others • Absence of dehydration or anemia/iron deficiency (Level of Evidence: C)
Class III
Repeated routine phlebotomies are not recommended due to the risk of iron depletion, decreased oxygen carrying capacity, and stroke (Level of Evidence: C)

Preoperative Risk Assessment

- While patients with CHD were not excluded from the major perioperative risk assessment studies from which the revised cardiac risk index (RCRI) and the National Surgical Quality Improvement Program (NSQIP) were derived, CHD is not specifically addressed in either model [17, 18]. One series demonstrated increased risk of death, perioperative cardiac arrest, myocardial infarction, stroke, respiratory complications, renal failure, sepsis, venous thromboembolism, perioperative transfusion, and reoperation in young adults aged 18–39 years with a history of prior heart surgery who were then undergoing noncardiac surgery, as compared with those without a history of prior heart surgery [19].
- Overall rates of complications are likely relatively low with one series including both children and adults up to age 50 years with a history of congenital heart disease reporting an overall event rate of perioperative morbidity and mortality of 5.4%. Factors associated with perioperative events in this series included cyanosis, current treatment for congestive heart failure, poor general health, and procedures performed on the respiratory or nervous systems [20]. Other features associated with increased perioperative risk are included in Table 11.10.
- In addition to consideration of these risk features, preoperative risk assessment for the ACHD patient should include the basic diagnostic tests listed in Table 11.11.
- Consultation with an ACHD expert regarding assessment of perioperative risk is also recommended [8].
- In the absence of an emergency, surgeries should be performed at an ACHD center with an anesthesiologist familiar with ACHD.

Table 11.10 CHD lesions and features associated with increased perioperative risk [8]

High risk	• Prior Fontan procedure • Primary or secondary pulmonary hypertension, especially if severe • Cyanotic CHD • Complex CHD with residual valvular dysfunction or the need for anticoagulation • New York Heart Association (NYHA) functional class III or IV • Severe systemic ventricular dysfunction (ejection fraction <35%) • Malignant arrhythmias • Severe left-sided obstructive lesions
Moderate risk	• Prosthetic valve or conduit • Intracardiac shunt • Moderate left-sided obstruction • Moderate systemic ventricular dysfunction

Table 11.11 Components of preoperative evaluation for ACHD patients [8]

• Systemic arterial oximetry
• ECG
• Chest X-ray
• Transthoracic echocardiogram
• Complete blood count
• Coagulation screen

Imaging

- Noninvasive imaging modalities for patients with ACHD include transthoracic and transesophageal echocardiography, cardiac magnetic resonance imaging (MRI), and cardiac computed tomography (CT). The benefits and limitations of each of these imaging modalities are described in Table 11.12.
- Transthoracic echocardiography is the primary imaging technique.

Pregnancy

- The recommendations for care of pregnant women with CHD are outlined in Table 11.13 [8].
- CHD is the most common type of heart disease encountered in pregnant women, and among women with CHD who become pregnant, 11% of pregnancies are affected by cardiac complications, most commonly heart failure and arrhythmias [22].
- Heart failure is related to the 30–50% increase in plasma volume that occurs in pregnancy, which is typically less well tolerated in the setting of myocardial dysfunction or obstructive lesions such as a stenotic valve.
- Arrhythmias during pregnancy are likely a result of multiple factors, including chamber dilation in the setting of volume overload and adrenergic receptor hyperexcitability related to estrogen and progesterone [23].

Table 11.12 Imaging modalities for patients with ACHD

Imaging modality	Benefits	Limitations
Transthoracic echocardiography (TTE)	• Define anatomy • Quantify ventricular function • Assess severity of valvular lesions • Quantify right ventricular and pulmonary artery pressure • Identify arterial and venous vascular anomalies • Assess volume status • Contrast agents help with assessment of left ventricular size and function • Agitated saline can be used to identify intracardiac or transpulmonary right-to-left shunts (ASD, patent foramen ovale, baffle leak), anomalous venous connections (e.g., persistent left-sided superior vena cava), and acquired intrapulmonary shunts (e.g., arteriovenous malformations) • Strain imaging may be helpful in identifying ventricular dysfunction	• Image quality affected by prior surgeries, obesity, and lung disease • Limited assessment of the right ventricle due to geometric shape and location • Interpretation requires expertise in both congenital and acquired heart diseases • Ultrasound contrast agents aside from agitated saline are not approved for use in patients with right-to-left shunts or bidirectional shunts • Strain imaging not uniformly available
Transesophageal echocardiography (TEE)	• Overcome some imaging limitations of TTE (e.g., obesity, lung disease) • Excellent imaging of the thoracic aorta, bioprosthetic valves, intra-atrial septum, and potential right and left atrial thrombi • Superior image resolution than TTE for identification of vegetations in the workup of endocarditis	• Invasive procedure requiring sedation • Performance and interpretation require expertise in both congenital and acquired heart diseases
Cardiac MRI	• Three-dimensional real-time imaging with high spatial and temporal resolution • Unparalleled visualization of the right ventricle • Excellent visualization of myocardium for tissue characterization (e.g., scar and fibrosis) • Can be used to quantify severity of regurgitant lesions and shunt fractions	• Local and regional expertise varies • Unable to reliably assess coronary arteries • Longer image acquisition times • Patient tolerance may be limited by claustrophobia and difficulty with breath holding • Arrhythmias affect gating and limit image quality • Patients may have metallic implants (pacemakers, surgically placed epicardial pacing leads, mechanical heart valves) making MRI contraindicated • Gadolinium contraindicated in the setting of significant renal dysfunction due to the risk of nephrogenic systemic fibrosis
Cardiac CT	• Noninvasive imaging of the coronary arteries • Qualitative LV and RV assessments for patients with inability to tolerate MRI • Faster image acquisition time	• Radiation exposure [11, 21]

Table 11.13 ACC/AHA guidelines for pregnancy management in patients with ACHD [8]

Class I
1. Patients with CHD should have consultation with an ACHD expert before they plan to become pregnant to develop a plan for management of labor and the postpartum period that includes consideration of the appropriate response to potential complications. This care plan should be made available to all providers (Level of Evidence: C)
2. Patients with intracardiac right-to-left shunting should have fastidious care of intravenous lines to avoid paradoxical air embolus (Level of Evidence: C)
3. Prepregnancy counseling is recommended for women receiving chronic anticoagulation with warfarin to enable them to make an informed decision about maternal and fetal risks (Level of Evidence: B)
Class IIa
1. Meticulous prophylaxis for deep venous thrombosis, including early ambulation and compression stockings, can be useful for all patients with intracardiac right-to-left shunt. Subcutaneous heparin or low-molecular-weight heparin is reasonable for prolonged bed rest. Full anticoagulation can be useful for the high-risk patient (Level of Evidence: C)
2. It is reasonable to consider antibiotic prophylaxis against endocarditis before vaginal delivery at the time of membrane rupture in select patients with the highest risk of adverse outcomes: prosthetic cardiac valve or prosthetic material used for cardiac valve repair and unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits (Level of Evidence: C)

Table 11.14 CARPREG risk factors for cardiac complications in pregnancy [24]

1. Prior cardiac event, including heart failure, transient ischemic attack, or stroke or arrhythmia preceding pregnancy
2. New York Heart Association Class III (marked limitation of physical activity) or IV (symptoms of heart failure at rest) or cyanosis
3. Significant obstructive lesions including mitral or aortic stenosis
4. Left ventricular systolic dysfunction with ejection fraction <40%

- Cardiac risk in pregnancy can be estimated using the CARPREG score, in which four risk factors are assessed (Table 11.14) [24].
- If no risk factors are present, the risk for adverse cardiac events during pregnancy including pulmonary edema, arrhythmia, embolic stroke, or cardiac death in a woman with known cardiac disease (CHD or acquired) is 5%. If one risk factor is present, the risk increases to 25%, and if two risk factors are present, the risk increases to 75% [24].
- The World Health Organization (WHO) has developed a well-validated strategy to quantify risk during pregnancy according to type of CHD, as described in Table 11.15 [23].
- Thromboembolism is another important risk for women with CHD during pregnancy. Two percent of pregnancies of women with CHD are affected by thromboembolic events, while these occur in only 0.5–0.1% of uncomplicated pregnancies [22]. Pregnancy is a hypercoagulable state due to decreased free protein S and increased vitamin K-dependent clotting factors. Women with CHD at particularly high risk for arterial and venous thrombosis are those with grafts, mechanical heart valves, arrhythmias, and cardiac chamber dilatation [23].

- Guidelines recommend prepregnancy counseling for women receiving warfarin to enable them to make an informed decision regarding maternal and fetal risks [8]. Continuation of warfarin during pregnancy is controversial, with some advocating avoidance since it crosses the placenta and, at doses higher than 5 mg per day and during weeks 6–12 of gestation, is associated with central nervous system teratogenicity. If used during pregnancy and vaginal delivery is planned, warfarin should be discontinued at around 36 weeks of gestation due to risk of fetal intracranial hemorrhage [25]. If enoxaparin is used in place of warfarin during pregnancy, anti-Xa levels must be followed carefully, drawn 4–6 h after injection with a goal level of 0.8–1.2 U/mL [26].
- Common cardiac medications that are generally avoided in pregnancy are angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, statins, and amiodarone [23].

Contraception

- Many women with congenital heart disease are not offered appropriate contraception, likely due to a combination of cardiologists' lack of familiarity with contraception and patients' incorrect perception that they may not be candidates for hormonal contraception [27]. Cardiologists as well as patients should understand that the risk for complications from pregnancy is significantly higher than the risk of contraception-related adverse events.
- Contraception should especially be emphasized for patients whose pregnancy would be very high risk, including those with Eisenmenger's syndrome, cyanotic congenital heart disease with resting oxygen saturation less than 85%, severe coarctation of the aorta, and Marfan syndrome with aortic dilation (Table 11.15).
- Other important considerations are the risk for hypertension and thrombosis with estrogen-containing contraceptives and potential drug interactions between hormonal contraceptives and cardiac medications [28].

Exercise

- Patients with CHD have lower exercise tolerance than those without CHD, but CHD patients tend to be less aware of exercise limitations [29].
- Many noncardiac factors may contribute to exercise intolerance, including deconditioning, exercise limitations during childhood, and misperceptions about exercise limitations [8].
- Given the physical and psychosocial benefits of exercise, the importance of exercise should be emphasized to all CHD patients. Patients should have an individualized exercise prescription that is updated regularly by their ACHD provider. Evidence to support specific exercise recommendations in patients with CHD is limited.

Table 11.15 WHO categories of risk for cardiac complications during pregnancy in patients with ACHD [23]

WHO Class	Risk for cardiac complications in pregnancy	Type of CHD
I	Low: risk of maternal morbidity or mortality no higher than in general population	• Uncomplicated, small, or mild: pulmonary stenosis, ventricular septal defect, patent ductus arteriosus • Successfully repaired simple lesions: atrial septal defect, ventricular septal defect, patent ductus arteriosus, total anomalous pulmonary venous return
II	Moderate: small increase in risk of maternal morbidity or mortality	• Unrepaired atrial septal defect • Repaired tetralogy of Fallot
II or III		• Hypertrophic obstructive cardiomyopathy • Marfan syndrome without aortic root dilatation
III	High: significant increase in risk of severe maternal morbidity or mortality	• Unrepaired cyanotic heart disease • Complex congenital heart disease • Marfan syndrome with bicuspid aortic valve
IV	Very high: pregnancy contraindicated due to very high risk of severe maternal morbidity or mortality	• Eisenmenger's syndrome • Cyanotic congenital heart disease with resting oxygen saturation <85% • Severe coarctation of the aorta • Marfan syndrome with aortic dilation

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- Patients with cyanosis should be advised to avoid dehydration due to risk for hyperviscosity (see "Hyperviscosity" section above).
- Patients with Marfan syndrome should be counseled to avoid high-impact activity due to the risk of aortic injury.
- Patients on therapeutic anticoagulation should also be counseled to avoid high-impact activity due to the risk of bleeding.
- Vigorous exercise should also be avoided in those at risk for sudden death, for example, patients with hypertrophic obstructive cardiomyopathy [29].

Infective Endocarditis Prevention and Evaluation

- Patients with congenital heart disease are at increased risk for endocarditis due to native valvular abnormalities and shunts including ventricular septal defect and patent ductus arteriosus, as well as surgical implantation of prosthetic material. The risk for endocarditis must be discussed with patients.
- Administration of antibiotics for endocarditis prophylaxis is reasonable for dental procedures in CHD patients with the conditions listed in Table 11.16, because these patients are at highest risk for infective endocarditis.

Table 11.16 Cardiac conditions and types of CHD with significantly increased risk for endocarditis for which administration of antibiotics for endocarditis prophylaxis with dental procedures is reasonable [8]

ACC/AHA Class IIa recommendations (Level of Evidence: B)
Previous infective endocarditis
Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
Unrepaired cyanotic CHD including palliative shunts and conduits
CHD completely repaired with prosthetic material or device, during the first 6 months after the procedure
Repaired CHD with residual defects at or adjacent to the site of repair inhibiting endothelialization

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Table 11.17 ACC/AHA guidelines for the evaluation of infective endocarditis [8]

Class I
When patients with ACHD present with an unexplained febrile illness, blood cultures should be drawn before antibiotics are administered to avoid delay in diagnosis of potential infective endocarditis (Level of Evidence: B)
Transthoracic echocardiography should be performed when the diagnosis of native valve infective endocarditis is suspected (Level of Evidence: B)
Transesophageal echocardiography should be performed if transthoracic echocardiography images or inadequate or equivocal, in the presence of a prosthetic valve or material or surgically corrected shunt or complex CHD anatomy, or to define possible endocarditis complications (abscess, valvular destruction or dehiscence, embolism, or hemodynamic instability) (Level of Evidence: B)
ACHD patients with evidence of infective endocarditis should have early consultation with a surgeon with expertise in ACHD due to potential for rapid deterioration and possible infection of prosthetic material (Level of Evidence: C)

- Endocarditis prophylaxis is not indicated for nondental procedures such as gastrointestinal endoscopy or sterile urologic procedures [8].
- In patients with CHD and fever without another clear source, clinicians must maintain a high index of suspicion for infective endocarditis. The ACC/AHA guidelines for the evaluation of infective endocarditis are listed in Table 11.17 [8].

Anatomy of CHD Lesions

- More commonly encountered acyanotic and cyanotic CHD lesions are listed in Table 11.18.
- Cyanosis is defined as concentration of deoxygenated hemoglobin greater than 5 gm/dL, which correlates with a peripheral oxygen saturation less than 85% [21].

Table 11.18 Acyanotic and cyanotic CHD lesions

Acyanotic	Cyanotic
Atrial septal defect	Tetralogy of Fallot
Patent foramen ovale	Ebstein's anomaly
Ventricular septal defect	Transposition of the great arteries
Patent ductus arteriosus	Eisenmenger's syndrome [30]
Pulmonary stenosis	
Aortic coarctation	

Atrial Septal Defect (ASD)

- An ASD allows blood to pass from the left atrium to the right atrium resulting in right ventricular volume overload and pulmonary overcirculation.
- Small defects (less than 0.5 cm in diameter) generally are not associated with significant shunting. Large defects (greater than 2 cm in diameter) often have associated large shunts [30].
- Types: ostium secundum (75%) near the fossa ovalis, ostium primum (15–20%) near the crux of the heart, sinus venosus (5–10%) near the superior or inferior vena cava, and rarely coronary sinus (less than 1%) near the ostium of the coronary sinus.
- Symptoms: dyspnea, palpitations, fatigue, exercise intolerance, and frequent pulmonary infections.
- Signs:
  - Physical examination: fixed splitting of the second heart sound on auscultation and relative pulmonic stenosis murmur secondary to increased blood flow across the pulmonic valve. Flow across the atrial septal defect does not create a murmur.
  - ECG: right axis deviation and incomplete right bundle branch block for ostium secundum ASDs and left axis deviation for ostium primum ASDs.
  - Chest X-ray: cardiomegaly secondary to right ventricular and right atrial enlargement, enlarged pulmonary artery, and increased pulmonary vascularity.
- Diagnosis is with echocardiography. Sinus venosus ASDs may be missed on transthoracic echocardiography. Agitated saline contrast echocardiography increases sensitivity for detection of ASDs. Transesophageal echocardiography may be needed.
- Cardiac catheterization may be necessary to quantify the shunt fraction.
- Indications for interventional or surgical treatment of ASDs are listed in Table 11.19.
- All types of ASDs may be repaired surgically; percutaneous repair is for ostium secundum ASDs only.
- Complications: atrial arrhythmias (atrial flutter, atrial fibrillation, sick sinus syndrome) and paradoxical embolism from venous thromboses or intravenous infusions [8, 21, 30].

Table 11.19 ACC/AHA guidelines for interventional or surgical treatment of ASDs [8]

Class I recommendation
Closure of an ASD either percutaneously or surgically is indicated for right atrial and right ventricular enlargement with or without symptoms (Level of Evidence: B)
Class IIa recommendation
Closure of ASD, either percutaneously or surgically, is reasonable in the presence of paradoxical embolism (Level of Evidence C) or documented platypnea-orthodeoxia (Level of Evidence: B)
Class IIb recommendation
Closure of an ASD, either percutaneously or surgically, may be considered in the presence of net left-to-right shunting, pulmonary artery pressure less than two thirds systemic levels, pulmonary vascular resistance less than two thirds systemic vascular resistance, or when responsive to either pulmonary vasodilator therapy or test occlusion of the defect (patients should be treated in conjunction with providers who have expertise in the management of pulmonary hypertensive syndromes) (Level of Evidence: C)
Class III recommendation
Patients with severe irreversible PAH and no evidence of a left-to-right shunt should not undergo ASD closure (Level of Evidence: B)

Patent Foramen Ovale (PFO)

- A PFO is a flap-like communication between the right and left atria and may have associated fenestrations or aneurysmal changes in the septum primum or secundum.
- Many do not consider PFO an atrial septal defect since no tissue is missing from the atrial septum but it can result in a shunt, hemodynamic consequences, symptoms, some signs, and complications similar to those of atrial septal defects.
- Found in 27% of normal hearts on autopsy [31].
- Valsalva maneuver greatly increases sensitivity of echocardiography in diagnosis of PFO. PFO is found in 5% of general population with saline contrast at rest and 25% with Valsalva (similar to prevalence in autopsy study) [32].
- Data are mixed on association between PFO and cryptogenic stroke. Risk of paradoxical embolism (RoPE) score can be used to estimate the probability that a PFO discovered in a patient with a cryptogenic stroke is incidental or pathogenic and the PFO-attributable fraction of stroke [33].
- Antiplatelet therapy is recommended for patients with stroke or transient ischemic attack (TIA) and PFO. Anticoagulation is recommended for patients with stroke, PFO, and venous thrombosis. Additional recommendations from the AHA and American Stroke Association (ASA) are listed in Table 11.20 [34].
- Invasive management such as PFO closure for patients with stroke and PFO is controversial, with recent studies showing reduction in the risk of recurrent stroke in patients treated with percutaneous PFO closure devices but an increased risk of atrial fibrillation and procedure and device-related complications [35, 36].
- The most recent American Academy of Neurology practice update recommends counseling all patients with PFO and stroke or TIA that having a PFO is common and it is uncertain if their PFO contributed to their stroke or TIA. The practice

Table 11.20 AHA and American Stroke Association (ASA) guidelines for the management of PFO in the setting of stroke [34]

Class I recommendations
For patients with an ischemic stroke or transient ischemic attack (TIA) and both a PFO and a venous source of embolism, anticoagulation is indicated, depending on stroke characteristics (Level of Evidence: A)
For patients with an ischemic stroke or TIA and a PFO who are not undergoing anticoagulation therapy, antiplatelet therapy is recommended (Level of Evidence B)
Class IIa recommendation
For patients with an ischemic stroke or TIA and both a PFO and a venous source of embolism in whom anticoagulation is contraindicated, an inferior vena cava filter is reasonable (Level of Evidence: C)
Class IIb recommendations
There are insufficient data to establish whether anticoagulation is equivalent or superior to aspirin for secondary stroke prevention in patients with PFO (Level of Evidence: B)

update also recommends against routinely offering percutaneous PFO closure to patients with cryptogenic stroke and in rare circumstances such as patients having recurrent strokes despite medical therapy, considering closure with the Amplatzer PFO Occluder [37].

Ventricular Septal Defect (VSD)

- VSD is the most common congenital heart defect in children, with many (especially muscular VSDs) closing spontaneously in childhood.
- Nomenclature has been complicated by multiple synonyms used to describe different types of VSDs, with perimembranous VSDs being the most common (80%). Muscular VSDs are next most common (20%), followed by VSDs just below the aortic valve (5%) and near the junction of the mitral and tricuspid valves (5%).
- Symptoms and signs depend on the type and size of VSD.
- Signs:
  - Physical examination: systolic murmur loudest over the left lower sternal border. Holosystolic in the setting of low right ventricular pressures and can be early systolic as RV pressure increases or with very small defects that close with muscular contraction
  - ECG: right and left or isolated right ventricular hypertrophy
  - Chest X-ray: left atrial and left ventricular enlargement with increased pulmonary vascular markings, which will all resolve if significant pulmonary arterial hypertension develops
- Complications: infective endocarditis, aortic regurgitation, and pulmonary vascular disease.
- Diagnosis is by echocardiography; typically transthoracic echocardiography is sufficient.

Table 11.21 ACC/AHA guidelines for the management of VSDs [8]

Class I recommendations
Closure of a VSD is indicated when there is a Qp/Qs (pulmonary-to-systemic blood flow ratio) of 2.0 or more and clinical evidence of LV volume overload (Level of Evidence: B)
Closure of a VSD is indicated when the patient has a history of IE (Level of Evidence C)
Class IIa recommendations
Closure of a VSD is reasonable when net left-to-right shunting is present at a Qp/Qs greater than 1.5 with pulmonary artery pressure less than two thirds of systemic pressure and PVR less than two thirds of systemic vascular resistance (Level of Evidence: B)
Closure of a VSD is reasonable when net left-to-right shunting is present at a Qp/Qs greater than 1.5 in the presence of LV systolic or diastolic failure (Level of Evidence: B)
Class IIb recommendation
Device closure of a muscular VSD may be considered, especially if the VSD is remote from the tricuspid valve and the aorta, if the VSD is associated with severe left-sided heart chamber enlargement, or if there is PAH (Level of Evidence: C)
Class III recommendation
VSD closure is not recommended in patients with severe irreversible pulmonary arterial hypertension. (Level of Evidence: B)

- Cardiac catheterization may be helpful to quantify shunting, to assess for pulmonary arterial hypertension including response to vasodilators, and to look for other lesions [8, 21, 30].
- Recommendations for VSD closure are included in Table 11.21 [8].

Patent Ductus Arteriosus (PDA)

- The ductus arteriosus is a communication between the left pulmonary artery and the descending aorta just distal to the left subclavian artery, which allows blood to bypass the lungs in the fetal circulation. It typically closes in the days following birth.
- If the ductus arteriosus remains open, there is a left-to-right shunt.
- Symptoms and signs depend on size and degree of shunting.
- Symptoms may include fatigue, dyspnea, palpitations, and those of infective endocarditis, endarteritis, and congestive heart failure.
- Signs:
  - Physical examination: widened pulse pressure, lower oxygen saturation with cyanosis or clubbing in the lower extremities, bounding peripheral pulses, continuous “machine-like” murmur loudest in the left infraclavicular area
  - ECG: left atrial and left ventricular hypertrophy
  - Chest X-ray: left atrial and left ventricular enlargement, enlarged proximal pulmonary artery, prominent ascending aorta, PDA which may be visible as an opacity at where the descending aorta and aortic knob meet
- Complications: infective endocarditis and arteritis.

- Diagnosis may be by echocardiography and often in combination with cardiac catheterization to quantify the shunt, evaluate for pulmonary arterial hypertension including response to vasodilators, and angiography to determine the size and shape of the ductus.
- Recommendations for closure of PDAs are listed in Table 11.22. Closure is often percutaneous in the cardiac catheterization laboratory and is safer for calcified PDAs. Larger PDAs or those distorted by aneurysm or prior endarteritis may need to be closed surgically [8, 21, 30].

Pulmonic Stenosis

- Pulmonic stenosis can be valvular, supravalvular, or subvalvular. Valvular is the most common, comprising 80–90% of all congenital causes of right ventricular outflow tract obstruction. There are three types of valvular pulmonic stenosis, including the most common dome-shaped form and less commonly a dysplastic, unicuspid, or bicuspid valve [8].
- Pulmonic stenosis may be associated with other forms of congenital heart disease. Valvular pulmonic stenosis is a part of Noonan syndrome, which demonstrates autosomal dominant inheritance with variable penetrance. Other features of Noonan syndrome include short stature, intellectual disability, low set ears, and webbed neck. Supravalvular pulmonic stenosis is associated with Williams syndrome, with other features including an infantile hypercalcemia, outgoing personality, intellectual disability, and a broad forehead with full cheeks.
- Many patients are asymptomatic. Symptoms, if present, may include dyspnea, presyncope, syncope, and anginal chest pain related to an enlarged pulmonary artery causing left main coronary artery compression.
- Signs:
  - Physical examination: pulmonary ejection sound ("click") that decreases with inspiration, early systolic murmur that increases with inspiration, wide splitting of S2, and signs of right heart failure that occur late

Table 11.22 ACC/AHA guidelines for PDA closure [8]

Class I recommendation
Closure of a PDA either percutaneously or surgically is indicated for left atrial and/or left ventricular enlargement or if pulmonary arterial hypertension is present or in the presence of net left-to-right shunting (Level of Evidence C) or in the setting of prior endarteritis (Level of Evidence: C)
Class IIa recommendations
It is reasonable to close an asymptomatic small PDA by catheter device (Level of Evidence: C)
PDA closure is reasonable for patients with PAH with a net left-to-right shunt (Level of Evidence: C)
Class III recommendation
PDA closure is not indicated for patients with PAH and net right-to-left shunt (Level of Evidence: C)

  - ECG: usually normal, with severe pulmonic stenosis, and may see right axis deviation, right atrial enlargement, and right ventricular hypertrophy
  - Chest X-ray: may see increased vascular fullness in the left lung base as compared with the right (Chen's sign) and dilation of the main pulmonary artery in some forms

- Complications: right ventricular failure
- Diagnosis is by echocardiography. Often transthoracic echocardiography is sufficient. In some cases, transesophageal echocardiography may be helpful to better visualize the right ventricular outflow tract. Cardiac MRI and CT may be helpful to define pulmonary artery anatomy and quantify associated lesions like pulmonic regurgitation and tricuspid regurgitation. Cardiac catheterization is rarely necessary.
- Treatment for pulmonic stenosis may be surgical or percutaneous with balloon valvotomy. ACC/AHA guidelines for management are listed in Table 11.23 [8].

Table 11.23 ACC/AHA guidelines for the management of pulmonic stenosis [8]

Class I recommendations
Balloon valvotomy is recommended for asymptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 60 mmHg or a mean Doppler gradient greater than 40 mmHg (in association with less than moderate pulmonic valve regurgitation) (Level of Evidence: B)
Balloon valvotomy is recommended for symptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 50 mmHg or a mean Doppler gradient greater than 30 mmHg (in association with less than moderate pulmonic regurgitation) (Level of Evidence: C)
Surgical therapy is recommended for patients with severe pulmonic stenosis and an associated hypoplastic pulmonary annulus, severe pulmonary regurgitation, subvalvular pulmonic, or supravalvular pulmonic stenosis. Surgery is also preferred for most dysplastic pulmonary valves and when there is associated severe tricuspid regurgitation or the need for a surgical Maze procedure (Level of Evidence: C)
Class IIb recommendations
Balloon valvotomy may be reasonable in asymptomatic patients with a dysplastic pulmonary valve and a peak instantaneous gradient by Doppler greater than 60 mmHg or a mean Doppler gradient greater than 40 mmHg (Level of Evidence: C)
Balloon valvotomy may be reasonable in selected symptomatic patients with a dysplastic pulmonary valve and peak instantaneous gradient by Doppler greater than 50 mmHg or a mean Doppler gradient greater than 30 mmHg (Level of Evidence: C)
Class III recommendations
Balloon valvotomy is not recommended for asymptomatic patients with a peak instantaneous gradient by Doppler less than 50 mmHg in the presence of normal cardiac output (Level of Evidence: C)
Balloon valvotomy is not recommended for symptomatic patients with pulmonic stenosis and severe pulmonary regurgitation (Level of Evidence: C)
Balloon valvotomy is not recommended for symptomatic patients with a peak instantaneous gradient by Doppler less than 30 mmHg (Level of Evidence: C)

Coarctation of the Aorta

- Coarctation of the aorta is a discrete narrowing of the descending aorta at the ligamentum arteriosus, which is near the origin of the left subclavian artery.
- May be associated with other congenital lesions such as bicuspid aortic valve, subvalvular aortic stenosis, VSD, and hypoplasia of the aortic arch.
- Usually asymptomatic, but patients may have headaches and leg fatigue, or with severe coarctation, there may be lower extremity claudication.
- Signs:
  - Physical examination: hypertension in the right arm (and often the left arm as well) with lower blood pressures in the lower extremities, decreased femoral pulses, brachio-femoral delay, hyperdynamic carotid pulses, continuous mammary artery murmurs if significant collaterals have developed
  - ECG: left ventricular hypertrophy
  - Chest X-ray: dilated ascending aorta, "3 sign" due to indentation at the coarctation site, notching on the underside of ribs due to collateral vessels
- Complications: systemic hypertension even after treatment, aortic dissection or rupture (not necessarily at the site of the coarctation) due to associated aortopathy, accelerated coronary artery disease, stroke, congestive heart failure, endocarditis, endarteritis, and intracerebral hemorrhage.
- Diagnosis is by echocardiography in the suprasternal notch view, with turbulent flow in the proximal descending aorta and characteristic forward flow in diastole.
- Additional imaging: every patient with coarctation, including those who have had reparative procedures, should have at least one MRI or CT scan to completely evaluate the thoracic aorta and intracranial vessels (ACC/AHA Class I Recommendation, Level of Evidence: B) [8].
- Medical management: first-line medications for the treatment of hypertension in the setting of coarctation of the aorta include beta-blockers, ACE inhibitors, or angiotensin receptor blockers (with monitoring of renal function).
- ACC/AHA guidelines for the management of coarctation of the aorta are listed in Table 11.24.

Tetralogy of Fallot

- As the name implies, tetralogy of Fallot has four components:
  - Right ventricular outflow tract obstruction
  - Right ventricular hypertrophy
  - VSD
  - Aorta that overrides the right and left ventricles (Fig. 11.1)
- Cyanosis results from right-to-left shunting across the VSD.
- There is a spectrum of severity including very mild cyanosis.

Table 11.24 ACC/AHA guidelines for the management of coarctation of the aorta [8]

Class I recommendations
Intervention for coarctation is recommended for peak-to-peak coarctation gradient greater than or equal to 20 mmHg or peak-to-peak coarctation gradient less than 20 mmHg in the presence of anatomic imaging evidence of significant coarctation with radiological evidence of significant collateral flow (Level of Evidence: C)
Choice of percutaneous catheter intervention versus surgical repair of native discrete coarctation should be determined by consultation with a team of ACHD cardiologists, interventionalists, and surgeons at an ACHD center (Level of Evidence: C)
Percutaneous catheter intervention is indicated for recurrent, discrete coarctation and a peak-to-peak gradient of at least 20 mmHg (Level of Evidence: B)
Surgeons with training and expertise in CHD should perform operations for previously repaired coarctation and long recoarctation segment or concomitant hypoplasia of the aortic arch (Level of Evidence: B)
Class IIb recommendation
Stent placement for long-segment coarctation may be considered, but the usefulness is not well established, and the long-term efficacy and safety are unknown (Level of Evidence: C)

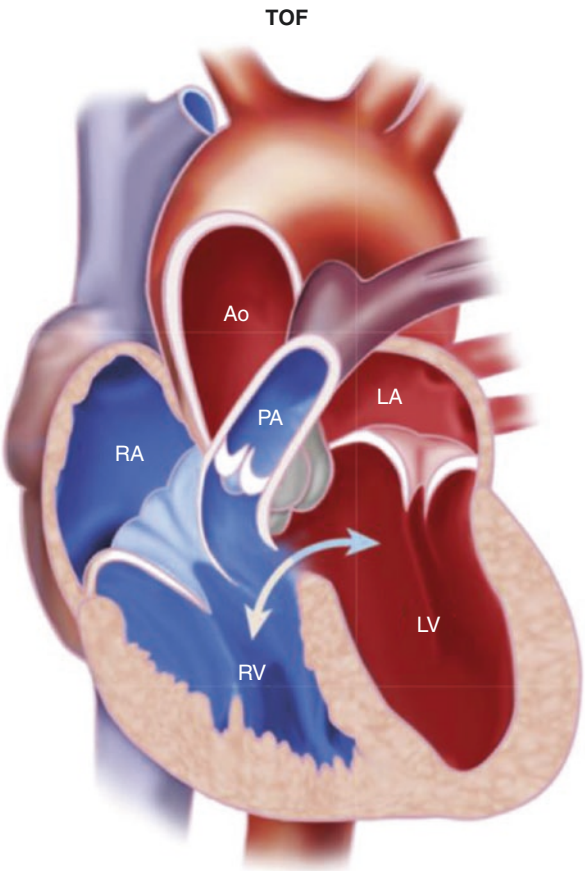

Fig. 11.1 Tetralogy of Fallot. Reproduced with permission from Otto (2013) [32]

- Associated congenital lesions include pulmonary artery hypoplasia and stenosis, ostium secundum ASD, atrioventricular septal defect (more commonly in patients with Down syndrome), right-sided aortic arch, and anomalous coronary arteries (left anterior descending artery arising from the right coronary artery and crossing the right ventricular outflow tract).
- Most patients with tetralogy of Fallot seen as adults in the United States will have had prior surgery.
- Historically, patients underwent palliative procedures to increase pulmonary blood flow, including Blalock-Taussig, Potts, and Waterston shunts (see Table 11.26).
- Currently, patients with tetralogy of Fallot undergo complete repair at a young age, including relief of right ventricular outflow tract obstruction with patch augmentation, pulmonary valve replacement, VSD closure, and surgeries for associated anomalies.
- Symptoms include cyanosis including episodic worsening ("spells") in children, dyspnea, and exercise intolerance.
- Signs:
  - Physical examination: digital clubbing, right ventricular lift or tap, systolic ejection murmur potentially with a thrill due to right ventricular outflow tract obstruction with shorter, softer murmur consistent with more severe obstruction, and a single second heart sound (due to inaudible pulmonic component)
  - ECG: right axis deviation and right ventricular hypertrophy
  - Chest X-ray: "boot-shaped" heart with an upturned right ventricular apex and concave main pulmonary artery segment

Table 11.26 Commonly encountered shunts

Shunt	Purpose	Details	Lesions treated	Years used
Blalock-Taussig	Increase pulmonary blood flow	Connect subclavian artery to pulmonary artery via direct anastomosis or with graft	Tetralogy of Fallot Pulmonary atresia Tricuspid atresia	1945–present
Glenn	Provide pulmonary blood flow without utilizing a ventricle in single ventricle physiology	Connect superior vena cava to right pulmonary artery. Classic: right pulmonary artery is no longer connected to main pulmonary artery. Bidirectional: right pulmonary artery remains connected to main pulmonary artery	Single ventricle anatomy Tricuspid atresia Double-inlet ventricle Hypoplastic left heart syndrome	Classic, 1959–1980s Bidirectional, 1985–present
Potts	Increase pulmonary blood flow	Connect descending aorta to left pulmonary artery	Tetralogy of Fallot Pulmonary atresia Tricuspid atresia	1940s–1960s
Waterston	Increase pulmonary blood flow	Connect ascending aorta to right pulmonary artery	Tetralogy of Fallot Pulmonary atresia Tricuspid atresia	1960s–1980s [32]

- Complications: ventricular tachycardia and fibrillation with increased risk of sudden death, atrial arrhythmias, severe pulmonic insufficiency following reparative surgery with resulting right ventricular failure, endocarditis, and complications of chronic cyanosis.
- Diagnosis is by echocardiography. Cardiac catheterization can provide additional hemodynamic data and define coronary and pulmonary artery anatomy [8, 21, 30].
- Repeat intervention for pulmonary regurgitation is often required, with ACC/AHA guidelines for interventions listed in Table 11.25 [8].
- Patients with tetralogy of Fallot should be followed closely by an ACHD specialist [8].

Ebstein's Anomaly

- Rare form of congenital heart disease.
- Tricuspid valve leaflets displaced into the right ventricle resulting in a dysfunctional tricuspid valve (often regurgitant, may be stenotic) as well as a small functional right ventricle (Fig. 11.2).
- Eighty percent of patients have an associated intra-atrial communication (ASD or PFO), and cyanosis results from right heart and tricuspid valve dysfunction, increased right atrial pressure, and right-to-left shunting across the intra-atrial communication.
- Approximately 25% of patients have one or more accessory conduction pathways (Wolff-Parkinson-White syndrome).

Table 11.25 ACC/AHA guidelines for surgery for adults with previous repair of tetralogy of Fallot [8]

Class I recommendation
Pulmonary valve replacement is indicated for severe pulmonary regurgitation and symptoms or decreased exercise tolerance (Level of Evidence: B)
Class IIa recommendation
Pulmonary valve replacement is reasonable in adults with previous tetralogy of Fallot, severe pulmonary regurgitation, and moderate to severe right ventricular dysfunction (Level of Evidence: B), moderate to severe right ventricular enlargement (Level of Evidence: B), development of symptomatic or sustained atrial and/or ventricular arrhythmias (Level of Evidence: C), or moderate to severe tricuspid regurgitation (Level of Evidence: C)
Surgery is reasonable in adults with prior repair of tetralogy of Fallot and residual right ventricular outflow tract (RVOT) obstruction (valvular or subvalvular) with peak instantaneous RVOT echocardiography gradient greater than 50 mmHg (Level of Evidence: C), right ventricle to left ventricle pressure ratio greater than 0.7 (Level of Evidence: C), progressive and/or severe dilatation of the right ventricle with dysfunction (Level of Evidence: C), residual VSD with a left-to-right shunt greater than 1.5:1 (Level of Evidence: B), severe aortic insufficiency with associated symptoms or more than mild left ventricular dysfunction (Level of Evidence: C), or a combination of multiple residual lesions (e.g., VSD and RVOT obstruction) leading to right ventricular enlargement or reduced right ventricular function (Level of Evidence: C)

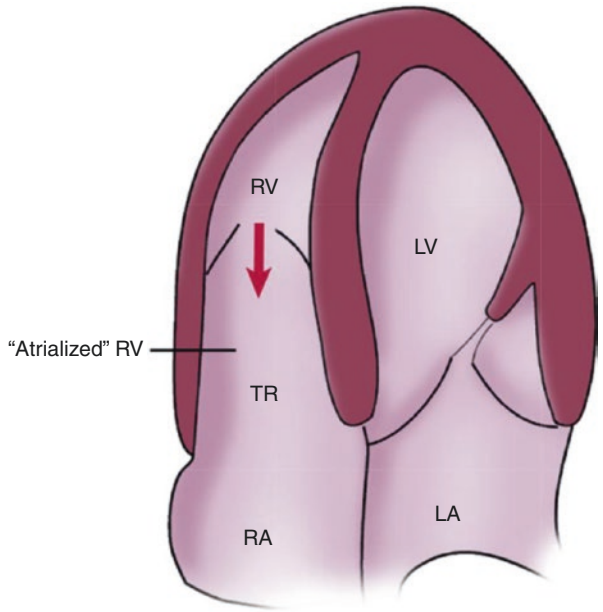

Fig. 11.2 Ebstein's anomaly. Reproduced with permission from Otto (2013) [32]

- Spectrum of severity from severe heart failure and death in the fetus or infant to minimal hemodynamic derangements with a normally functioning tricuspid valve.
- Symptoms include palpitations, syncope, dyspnea, exercise intolerance, fatigue, and right-sided congestive heart failure.
- Signs:
  - Physical examination: jugular venous pressure often normal due to large and compliant right atrium and holosystolic murmur at the left lower sternal border due to tricuspid regurgitation
  - ECG: delta wave due to preexcitation, tall and peaked ("Himalayan") P waves, right bundle branch block
  - Chest X-ray: may be normal in mild cases or demonstrate significant cardiomegaly ("wall-to-wall heart") with right atrial enlargement in severe cases
- Complications include atrial arrhythmias, sudden death, and, if an intra-atrial communication is present, paradoxical embolism and brain abscess.
- Diagnosis is by echocardiography, and transesophageal echocardiography may be required to evaluate for ASD or PFO.
- Management of Ebstein's anomaly in adults is generally aimed at prevention and treatment of complications. Tricuspid valve repair or replacement and ASD closure if present should be performed by congenital heart disease surgeons in the setting of symptoms, decreasing exercise tolerance, oxygen saturation less than 90%, paradoxical embolism, progressive cardiomegaly on chest X-ray, or progressive RV dilation or reduction of RV systolic function (Class I ACC/AHA Recommendation, Level of Evidence: B) [8].

Dextro-transposition of the Great Arteries (D-TGA)

- The aorta arises anteriorly from the right ventricle and the pulmonary artery arises from the left ventricle (see Fig. 11.3).
- Creates parallel circuits of blood flow with deoxygenated blood returning to the right heart and then being pumped into the systemic circulation via the aorta and oxygenated blood returning to the left heart and then being pumped into the pulmonary circulation. For a baby with transposition of the great arteries to survive, there must be a communication between the circuits, via a patent ductus arteriosus, patent foramen ovale, ASD, or VSD.
- Untreated, mortality is 90% by 6 months of age [30].
- Repaired patients will have had an atrial switch procedure (Senning or Mustard procedures) in the 1960s–1990s or an arterial switch procedure performed more recently.

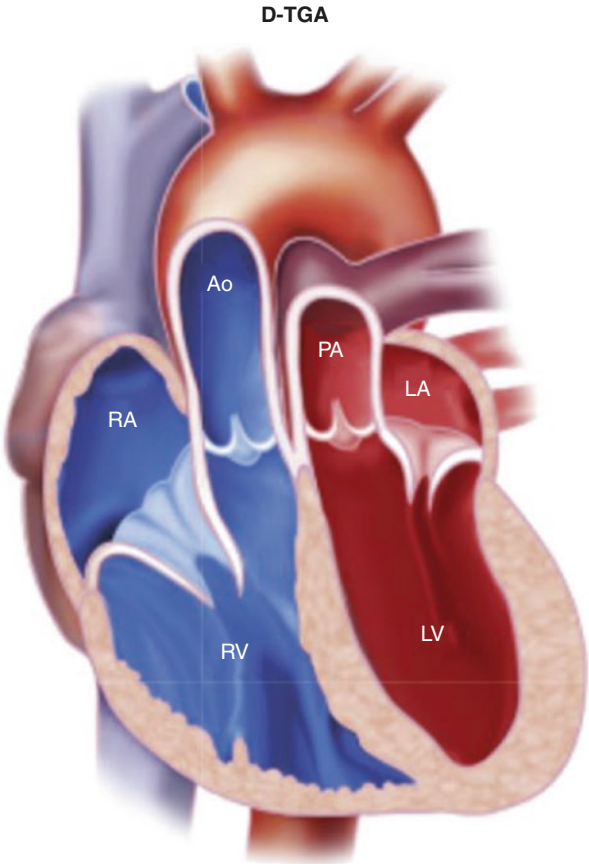

Fig. 11.3 Dextro-transposition of the great arteries. Reproduced with permission from Otto (2013) [32]

- The atrial switch procedure involves construction of intra-atrial baffles from atrial tissue (Senning) or pericardium (Mustard) to redirect systemic venous return to the left ventricle to be oxygenated in the lungs and pulmonary venous return to the right ventricle and onto the systemic arterial circulation (Fig. 11.4). The right ventricle continues to function as the systemic ventricle. Complications include congestive heart failure from systemic right ventricular dysfunction, tricuspid regurgitation, atrial arrhythmias including sinus node dysfunction, baffle leak and stenosis, and sudden death.
- Since the 1990s, patients have undergone the arterial switch procedure, which involves transecting the pulmonary artery and aorta above the semilunar valves, anastomosing the aorta to the native pulmonic valve with reimplantation of coronary arteries, and anastomosing the pulmonary artery to the native aortic valve. The arterial switch is performed in infants and has excellent long-term outcomes. Complications include coronary artery stenosis, valvular regurgitation, and stenosis in the great arteries.
- Coronary angiography is reasonable in all patients following arterial switch operation to rule out obstruction (Class IIa ACC/AHA Recommendation, Level of Evidence: C).
- Patients with D-TGA should be followed closely by an ACHD specialist [8, 21, 30, 32].

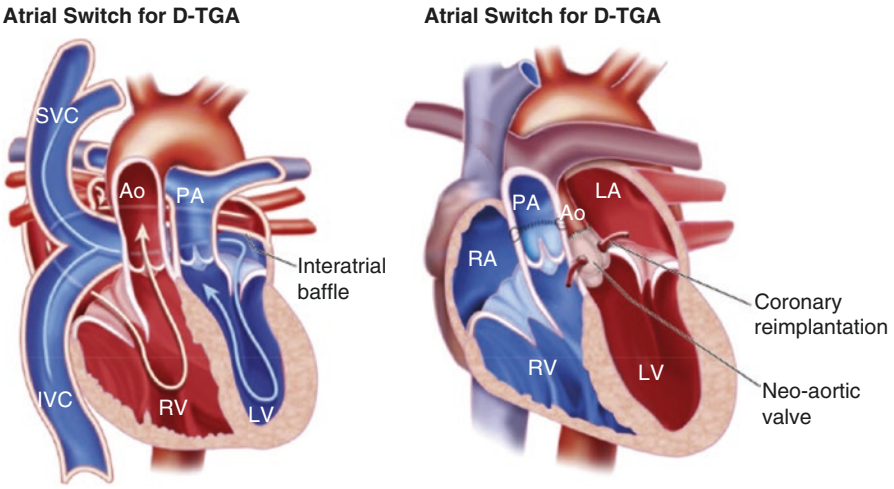

Fig. 11.4 Atrial switch operation and arterial switch operation. Reproduced with permission from Otto (2013) [32]

Eisenmenger's Syndrome

- Occurs in various forms of congenital heart disease with left-to-right shunts that result in severe and eventually irreversible pulmonary vascular disease and pulmonary arterial hypertension with subsequent reversal of shunt direction to create a right-to-left shunt.
- A murmur of childhood may disappear as left-to-right shunting ceases, which be falsely interpreted as the communication closing.
- Symptoms may not appear until late childhood or adulthood and include decreased exercise tolerance, exertional dyspnea, palpitations, hemoptysis, and symptoms of hyperviscosity (see "Hyperviscosity" section).
- Signs:
  - Physical examination: clubbing and cyanosis, palpable right ventricular heave, prominent second heart sound, right-sided fourth heart sound, clear lungs
  - ECG: right ventricular hypertrophy
  - Chest X-ray: prominent central pulmonary arteries with decreased vascular markings ("pruning")
- Complications include atrial arrhythmias, thrombosis as well as bleeding due to abnormal hemostasis, paradoxical embolization, brain abscess, and sudden death.
- Echocardiography is used to define anatomy, but cardiac catheterization is needed to quantify shunt and pulmonary vascular disease and to assess responsiveness to inhaled vasodilators.
- Treatment includes pulmonary vasodilators (due to potential to improve quality of life) and heart or combined heart and lung transplantation.
- The following exposures must be avoided: pregnancy, dehydration, moderate and severe strenuous exercise, excessive heat (hot tub or sauna), chronic high-altitude exposure (especially greater than 5000 ft above sea level), iron deficiency, air bubbles in intravenous tubing, and endocardial pacing.
- Patients with Eisenmenger's syndrome should be followed closely by an ACHD specialist [8, 30].

Prior Surgeries

- History and details or prior surgeries must be obtained.
- Commonly encountered shunts are listed in Table 11.26.

Fontan Palliation

- A palliative surgery performed for children with congenital heart disease that is not amenable to biventricular repair, for example, tricuspid atresia, double-inlet ventricle, and hypoplastic left heart syndrome.
- Goals of the Fontan procedure include providing adequate pulmonary and systemic blood flow, alleviating cyanosis, and decreasing ventricular volume overload.
- A direct connection is created from systemic venous return to the pulmonary artery without a ventricle in between. This may be done via an extra- or intracardiac conduit (see Fig. 11.5).
- Drawbacks of Fontan physiology include chronic systemic venous hypertension and decreased cardiac output and exercise tolerance with passive filling of the pulmonary circulation.
- A 12-year survival is approximately 70% [38].
- Complications include atrial arrhythmias, sudden death, atrial thrombus, hepatic congestion, cirrhosis, hepatocellular carcinoma, protein-losing enteropathy, and plastic bronchitis.
- Management includes regular imaging with echocardiography or cardiac MRI, cardiac catheterization for change in cardiac symptoms, electrophysiology consultation, warfarin for patients with atrial level shunting, atrial thrombus, atrial

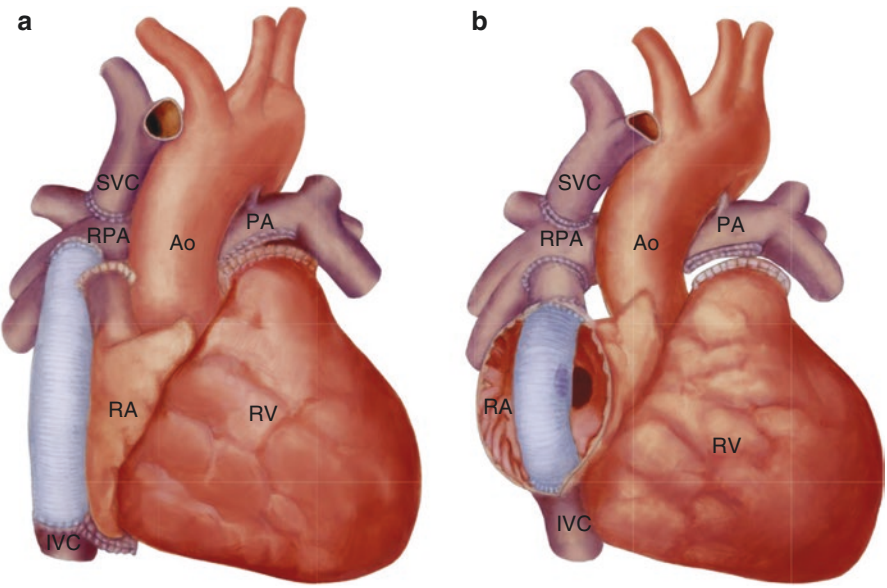

Fig. 11.5 Fontan anatomy with extra-cardiac (**a**) or intracardiac (**b**) conduit. Reproduced with permission from Otto (2013) [32]

arrhythmia, or history of thromboembolic event, ACE inhibitors and diuretics for systemic ventricular dysfunction (Class IIa ACC/AHA Recommendation, Level of Evidence: C), and in some cases reoperation or cardiac transplantation [8].

- Patients who have undergone Fontan should be followed closely by an ACHD specialist [8, 32].

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