

Caring for the Adult with Congenital Heart Disease: Management of Common Defects

By Reema Chugh, MD, FACC

Introduction

Congenital heart defects (CHDs) are common birth defects, with major ones occurring in nearly 1% of live births. Many are diagnosed with CHD in infancy and childhood, whereas others are not diagnosed until adulthood. Advances in pediatric medicine and surgery since the 1940s have made it possible for nearly 85% of infants born with CHDs to survive into adulthood.

Primary care physicians, obstetricians, and cardiologists are now seeing patients in transition to adulthood from the pediatric cardiology clinics. In addition, there are those who were lost to follow-up monitoring for several years because of problems with insurance or the belief that they did not need follow-up care because their defect was “repaired.” All patients do need follow-up monitoring because there are long-term residua and sequelae associated with repair of nearly all the defects. Routine follow-up care allows prevention, early identification, and appropriate management of these problems. Women with CHDs may be at risk for maternal and fetal complications during pregnancy and should have preconception counseling.

Since 1980, the care of CHD in adults has developed into a subspecialty of cardiovascular diseases.¹ The Bethesda conferences held in 1990 and 2000 and the Canadian Cardiovascular Society Consensus Conference 2001 updated established guidelines for the care of the adult with congenital heart disease.²⁻⁵

The first part of this article reviews some of the challenges posed by some of the relatively common “simple” heart defects seen in my practice. The second part details management of the more “complex” CHDs (Table 1). The aim of this article is to provide an outline of practice guidelines for primary care physicians, nurses, and allied health professionals providing care for adults with CHDs.

Patent Foramen Ovale

Patent foramen ovale (PFO) is a slit-shaped tunnel-like defect in the atrial septum, a residual from the fetal circulatory system caused by the failure of the primum and secundum atrial septa to fuse postnatally. It is the most common CHD, with an autopsy-derived incidence of about 27% for probe-patent PFO.

Atrial septal aneurysm is another defect of atrial morphogenesis and is characterized by a redundant, undulating, interatrial membrane in the region of the fossa ovalis. The diameter of the base exceeds 15 mm, and the amplitude of the interatrial septum excursion is 10 mm to 15 mm (Figure 1). Atrial septal aneurysm is associated with PFO in 50% to 85% of cases.

The PFO has caught recent clinical interest because of its clinical manifestations (Table 2). Of special importance is the association with paradoxical embolism, especially in adults age <55 years with cryptogenic stroke. However, the clinical diagnosis of paradoxical embolism is presumptive and is based on the presence of a right-to-left shunt in the absence of a left-side thromboembolic source. An adequate Valsalva maneuver is essential while assessing the presence of a right-to-left shunt by an agitated saline contrast study with transtho-

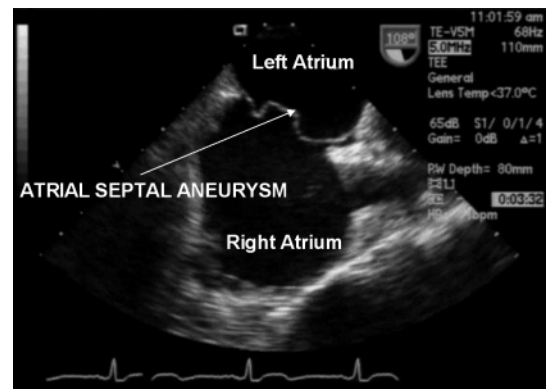


Figure 1. Atrial septal aneurysm.



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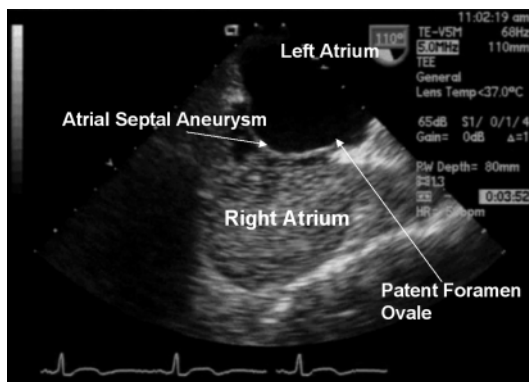


Figure 2. Transesophageal echocardiogram with an agitated saline contrast “bubble” study for evaluation of right-to-left interatrial shunt.

racic or transesophageal echocardiography (Figure 2).

A multicenter European study of 581 patients with ischemic stroke demonstrated that the association of recurrent stroke rate for patients with atrial septal aneurysm and PFO was 15.2% compared with 4.2% in the absence of these two defects.⁶ However, the PFO in Cryptogenic Stroke Study (PICSS), a prospective population-based study, suggested that after correction for age and comorbidity, an isolated PFO is not an independent risk factor for future cerebrovascular events in the general population.⁷ It must be noted that the patients in this study were not representative of the patient population with PFO and ischemic stroke. Therefore, although all the PFOs in the general population need not be medically treated or closed, adults age <55 years with cryptogenic stroke still need evaluation for consideration of medical therapy or closure, now possible by a percutaneous device.

With respect to selection of medical therapy versus device closure, the present data are even more controversial. In the Warfarin-Aspirin Recurrent Stroke Study, 2206 patients were randomized to aspirin or warfarin (international normalized ratio, 1.4 to 2.8), with no significant benefit shown from either treatment at two years.⁸ Another prospective study evaluated 140 consecutive patients with transcranial Doppler after PFO device closure for cryptogenic stroke and demonstrated a residual detectable shunt in 9% at one-year follow-up evaluation after successful implantation.⁹ Thus, there is controversy about the choice of management options for PFO. New trials are being conducted to address these questions.

Meanwhile, current guidelines do not specifically recommend one therapy over another. There is consensus that long-term anticoagulation is favored in most pa-

tients with high-risk features, including atrial septal aneurysm, a PFO with major shunt, Eustachian valve anatomy favoring right-to-left shunt, presence of venous thrombus or hypercoagulable states, and stroke involving multiple territories. Percutaneous closure is indicated for high-risk patients with recurrence despite therapeutic anticoagulation or in high-risk patients in whom long-term anticoagulation is contraindicated.

Bicuspid Aortic Valve

Bicuspid aortic valve (BAV) is defined as an aortic valve with (effectively) two instead of three valve leaflets. Of many variations in the pattern,¹⁰ the most common has fusion of the right and left aortic valve cusps.

Table 1. “Simplified” classification of congenital heart defects

Abnormal intracardiac communications: <i>shunt lesions</i>
<ul style="list-style-type: none"> • Patent foramen ovale (PFO) • Atrial septal defect (ASD) • Ventricular septal defect (VSD) • Patent ductus arteriosus (PDA)
Congenital valvular defects
<i>Stenotic lesions</i>
<i>Aortic</i>
<ul style="list-style-type: none"> • Valvular—bicuspid, unicuspid, or quadricuspid aortic valve • Subvalvular stenosis or membrane • Supravalvular stenosis
<i>Pulmonary</i>
<ul style="list-style-type: none"> • Pulmonary stenosis • Subpulmonic/infundibular stenosis
<i>Mitral</i>
<ul style="list-style-type: none"> • Mitral stenosis • Cor triatriatum
<i>Regurgitant lesions</i>
<ul style="list-style-type: none"> • Aortic regurgitation • Pulmonary regurgitation • Mitral—cleft mitral valve • Tricuspid—Ebstein’s anomaly
Great vessel lesions
<ul style="list-style-type: none"> • Patent ductus arteriosus • Coarctation of aorta
Conotruncal or major septation defects
<ul style="list-style-type: none"> • Tetralogy of Fallot • Endocardial cushion defects • Abnormal chamber and great vessel communications <ul style="list-style-type: none"> • D transposition of the great arteries (DTGA) • Congenitally corrected transposition of the great arteries (CCTGA) • Double-outlet right ventricle • Total or partial anomalous pulmonary venous return • Truncus arteriosus
Atresias
<ul style="list-style-type: none"> • Pulmonary atresia • Tricuspid atresia • Mitral atresia • Hypoplastic left heart syndrome

BAV occurs in 1% to 2% of the population, and diagnosis is usually suspected on physical examination (Table 2) and confirmed by an echocardiogram. In the parasternal short-axis view, a BAV opens as an oval (football) in contrast to the triangle of a trileaflet valve. A transesophageal echocardiogram may be required when the aortic valve morphology cannot be visualized clearly in the parasternal short-axis view in a transthoracic echocardiogram.

Unfortunately, BAV is diagnosed in a substantial proportion of affected adults only when they present with infective endocarditis (Figure 3), which is responsible for many cases in which the presentation is severe aortic regurgitation. Because of the high incidence of endocarditis and its associated complications, antibiotic prophylaxis is recommended in all patients, even in the absence of associated stenosis or regurgitation.

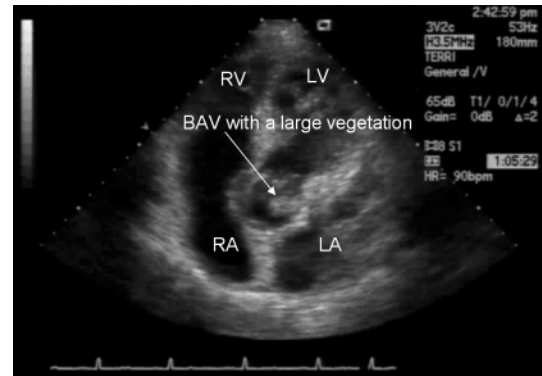


Figure 3. Transthoracic echocardiogram showing bicuspid aortic valve (BAV) with infective endocarditis.

LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

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Table 2. Findings, manifestations, and associated defects of congenital heart defects			
Heart defect	Salient findings on cardiac examination	Clinical manifestations	Commonly associated defects
Patent foramen ovale	None	Paradoxical embolus Stroke	Atrial septal aneurysm
Bicuspid aortic valve	Systolic ejection click Early peaking systolic flow murmur	Infective endocarditis Aortic dissection Aortic stenosis Aortic insufficiency	Coarctation of aorta Aortic root dilation Sub- and supra-aortic stenosis Ventricular septal defect
Secundum atrial septal defect	Prominent right ventricle Left parasternal impulse Wide fixed splitting of S2 Pulmonary ejection systolic at left upper sternal edge Accentuated P2	Atrial arrhythmias Right heart failure Pulmonary hypertension Left ventricular dysfunction Paradoxical embolism/stroke	Anomalous pulmonary venous return
Ventricular septal defect	Pansystolic murmur Precordial thrill Accentuated P2 (in pulmonary hypertension)	Infective endocarditis Left ventricular dysfunction Conduction defects (heart block) Aortic insufficiency Tricuspid regurgitation	Atrial septal defect Aortic root dilation
Patent ductus arteriosus	Continuous “machinery” murmur at the left upper sternal border with radiation to the back	Infective endocarditis/endarteritis Pulmonary hypertension Heart failure	
Coarctation of aorta	Thrill in the suprasternal notch Loud aortic closure sound	Hypertension Infective endocarditis/endarteritis Recoarctation Aortic aneurysm Premature coronary artery disease	Bicuspid aortic valve Aortic aneurysm Intracranial aneurysms Subaortic stenosis Mitral stenosis
Pulmonary stenosis	Systolic ejection click at left upper sternal border louder during expiration Harsh crescendo-decrescendo systolic ejection murmur radiating to the back and varying with respiration	Right ventricular hypertrophy Pulmonary regurgitation Pulmonary artery dilation/aneurysm	Patent foramen ovale Atrial septal defect Peripheral pulmonary stenosis

A complete echocardiographic examination should define the morphology, determine the degree of aortic stenosis and regurgitation, and include assessment of associated defects that may occur in 20% to 50% of patients (Table 2). Because of inherent aortic structural wall abnormalities, BAV is associated with aortic root dilation and risk of aortic dissection.¹¹ The risk of aortic aneurysm and dissection is markedly increased when BAV is associated with coarctation of the aorta. Hypertension control and avoidance of heavy lifting and isometric exercises are essential in preventing progression of aortic root dilation. Although aortic root dilation occurs more commonly in Marfan syndrome, there are more patients with aortic dilation associated with BAV because it is a more common defect. Serial echocardiography assesses aortic root dimensions (Figure 4). Computed tomography scanning or magnetic resonance imaging (MRI) is required when aneurysm of the arch or descending aorta is suspected in patients with coexistent coarctation of the aorta.

Aortic stenosis due to calcification develops at an earlier age in men with BAV than in women with BAV. On average, the aortic valve gradient increases approximately by 18 to 27 mm Hg for each decade of life, depending on the anatomy of the cusps as well as acquired risk factors. The development of left ventricular dysfunction may mask the degree of stenosis assessed by valve gradients. The risk factors for atherosclerosis, such as hyperlipidemia, obesity, and smoking, may contribute to the age-related deterioration of the aortic valve. Hence, especially intensive cardiovascular risk reduction should be advised at an early age.

Aortic valvuloplasty is preferred in children, whereas

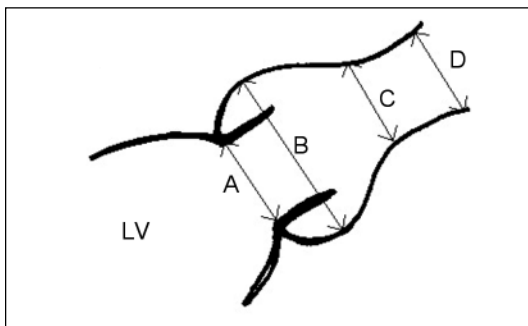


Figure 4. Aortic root. Schematic representation of the aortic root in the long axis view showing the points of measurement during TTE or TEE.

A = aortic annulus; B = mid-sinus level; C = sinotubular junction; D = 2 cm distal to the sinotubular junction; TEE = transesophageal echocardiography; TTE = transthoracic echocardiography.

aortic valve surgery (repair or replacement) is performed in adults. Emergency surgery is often required for those presenting with infective endocarditis and new-onset severe aortic regurgitation or aortic root abscess.

Long-term follow-up monitoring is mandatory for this possibly preventable and potentially life-threatening condition. The rate of familial recurrence is approximately 9%, with an autosomal-dominant pattern of inheritance and incomplete penetrance and variable expression. Echocardiographic screening of first-degree relatives is recommended.

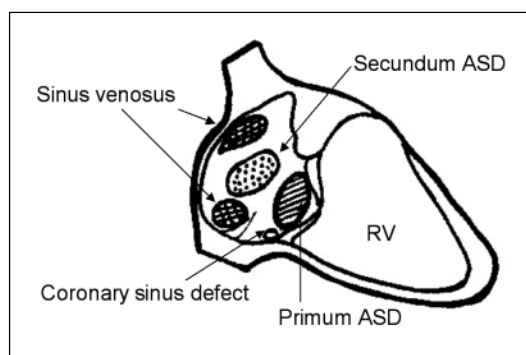


Figure 5. Types of atrial septal defects (ASDs).

Atrial Septal Defect

Atrial septal defect (ASD) is a direct communication between the cavities of the atrial chambers that permits shunting of blood.¹ The secundum defect is the most common form of ASD (Figure 5). Associated defects occur in nearly 30% of cases (Table 2). In one in six patients, ASD is likely to be first diagnosed in adulthood and present with palpitations due to atrial arrhythmias or increasing shortness of breath. On physical examination, the characteristic findings are a prominent right ventricular parasternal lift, persistent or wide fixed splitting of the second heart sound, and a pulmonary systolic ejection murmur at the left upper sternal edge due to increased pulmonary flow. In the presence of pulmonary hypertension, there is accentuation of the pulmonary component of the second heart sound. The electrocardiogram may show sinus rhythm or atrial fibrillation, right axis deviation (left axis deviation in primum ASD), and a bifid notch on the ascending limb of the R wave in inferior leads (“crochetage”).

The diagnosis is usually confirmed by transthoracic echocardiography, which may demonstrate a discontinuity of the interatrial septum on two-dimensional echocardiography and an intracardiac shunt at the atrial level with color Doppler. In addition, there may

The development of left ventricular dysfunction may mask the degree of stenosis assessed by valve gradients.

be right heart enlargement and elevated pulmonary artery pressure in patients with a long-standing right-to-left shunt. When the defect is not clearly defined by transthoracic echocardiography in the presence of right heart enlargement, further evaluation with transesophageal echocardiography is warranted.

The three major reasons for deterioration with age are as follows: 1) age-related decrease in left ventricular compliance may lead to augmentation of the left-to-right shunt; 2) heart failure may be precipitated by atrial arrhythmias; and 3) persistence of left-to-right shunt may lead to mild-to-moderate pulmonary hypertension and consequent right ventricular pressure and volume overload. Delayed closure has been shown to be associated with a higher likelihood of long-term complications, including atrial arrhythmias, pulmonary hypertension, right heart failure, paradoxical embolism, and stroke.

The consensus is to close defects associated with symptoms or right heart enlargement. Percutaneous transcatheter device closure is presently the method of choice for defects within the fossa ovalis, with an adequate rim (4–5 mm) between the defect and the aortic valve annulus, atrioventricular (AV) valves, and pulmonary and systemic veins, to avoid distortion of these surrounding structures. In the presence of pulmonary hypertension, the defect may be safely closed if pulmonary artery systolic pressure is less than 50% of the systemic arterial pressure. Right heart catheterization to assess the pulmonary vascular resistance may be required for those with higher pulmonary artery pressures. In general, all defects should be considered for early closure unless there are specific contraindications.

Ventricular Septal Defect

Ventricular septal defect (VSD) is a communication between the two ventricles resulting from failure of the components of the interventricular septum to fuse. Perimembranous VSD is the most common form¹ (Figure 6). Long-term complications depend on the size and the location of the VSD and any associated defects (Table 2).

The direction and the volume of a shunt are dictated by the size of the defect and the ratio of pulmonary vascular resistance to systemic vascular resistance. The left heart is volume-loaded, and depending on the size of the defect, the right heart may become pressure-loaded. In a small, restrictive VSD, right ventricular systolic pressure is less than half of the left ventricular systolic pressure, whereas in a large, unrestrictive VSD, there is equalization of right and left ventricular pressures due to free communication between the two chambers, leading to increased pressures to systemic

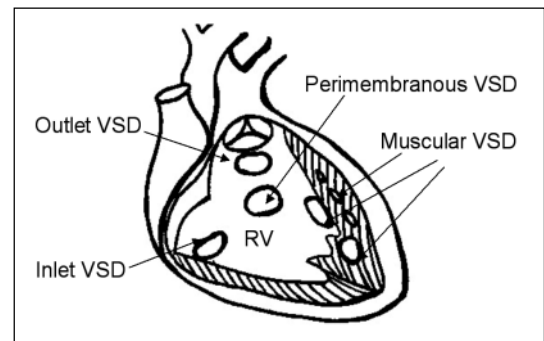


Figure 6. Types of ventricular septal defects (VSDs).

levels in the pulmonary arteries and the development of irreversible pulmonary vascular disease. This leads to bidirectional or reversed (right-to-left) shunt through the large VSD and severe pulmonary hypertension with systemic pulmonary artery pressures, originally described by Victor Eisenmenger in 1897. This was later described with its distinctive clinical and physiologic characteristics as Eisenmenger syndrome by Paul Wood in 1958.¹³ A VSD may usually be diagnosed on physical examination by the presence of a precordial thrill and a pansystolic murmur. The pulmonic component of the second heart sound may be accentuated in the presence of pulmonary hypertension.

Echocardiography defines the VSD location, degree of shunt, and associated defects. Most large VSDs are diagnosed and closed in childhood. Although nearly 10% of small VSDs may spontaneously close, a persistence of these to adulthood is associated with a 25% likelihood of serious complications. These complications may include infective endocarditis in 11%, heart failure, conduction blocks, and arrhythmias.¹⁴ In patients with a supracristal (subpulmonic) VSD, there is progressive aortic regurgitation, due to prolapse of the aortic valve leaflet into the defect, resulting from

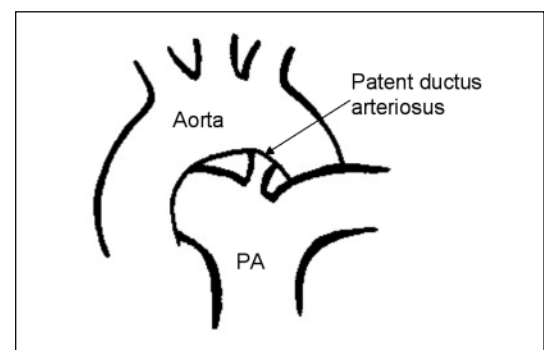


Figure 7. Patent ductus arteriosus.

PA = pulmonary artery.

the *Venturi effect*. Therefore, antibiotic prophylaxis and long-term follow-up monitoring are needed.

Patent Ductus Arteriosus

Patent ductus arteriosus (PDA) is a residual fetal communication between the proximal left pulmonary artery and the descending aorta distal to the left subclavian artery. During fetal life, the communication allows the diversion of blood from the right ventricle to the descending aorta, thus bypassing the pulmonary circulation¹ (Figure 7). Patients with a small PDA are usually asymptomatic, whereas those with a moderate to large PDA may present with dyspnea or palpitations. Sometimes PDA is diagnosed on physical examination by the continuous “machinery” murmur at the left upper sternal border with radiation to the back, whereas in others it is detected only with echocardiography.

The long-term residua and sequelae and the type of intervention depend on the size and shape of the PDA. The risk of endarteritis is present in all patients, and therefore antibiotic prophylaxis, early closure, or both are recommended. Heart failure and pulmonary hypertension are likely to develop with a moderate-size or larger PDA. Device closure is preferred for the majority of these PDA patients.¹⁵ Indications for surgical closure include a PDA that is too large for device closure or presence of ductal aneurysm. Increased pulmonary vascular resistance at the time of closure may also present with late pulmonary hypertension. Severe, irreversible pulmonary vascular disease is a contraindication for PDA closure. Endocarditis or endarteritis prophylaxis is recommended for six months after device closure and for life for residual PDA.

Coarctation of the Aorta

Coarctation of the aorta is congenital narrowing of the aorta at the junction of the distal aortic arch and the descending aorta, below the origin of the left subclavian artery¹ (Figure 8). It represents up to 8% of all CHDs. The discrete coarctation is not just limited to focal stenosis but is one variant of a diffuse arteriopathy and associated structural abnormalities of the great arterial walls. It is therefore no longer really considered to be a “simple” heart defect.¹¹ Moderate to severe coarctation is usually diagnosed in infancy or childhood. In adulthood, the diagnosis is suspected in a person presenting with hypertension. A brachial and femoral blood pressure recording may demonstrate upper-body arterial hypertension. Characteristic physical findings include weak, delayed femoral pulses; a prominent left ventricular impulse; a loud

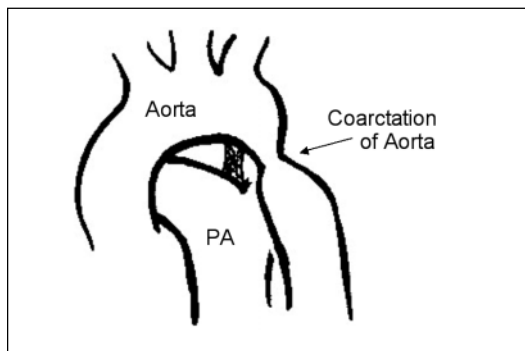


Figure 8. Coarctation of the aorta.

PA = pulmonary artery.

aortic closure sound; a thrill in the suprasternal notch; and a vascular murmur between the shoulder blades, beginning in midsystole and persisting beyond the second heart sound. Continuous murmurs due to collaterals may be present.

Echocardiography confirms the diagnosis and screens for commonly associated defects such as BAV (75%–85%), aortic root dilation/aneurysm, VSD, and mitral valve abnormalities. MRI shows the site of stenosis, the extent and degree of narrowing, the pressure gradient across the stenosis, aortic arch anatomy, and aortopulmonary collaterals and assesses left ventricular mass and function. It is especially of use for delineating aneurysms after surgery.

Patients with coarctation of the aorta are at increased risk for aortic aneurysms and dissection. Hypertension often persists after surgery, and ambulatory blood pressure monitoring may detect uncontrolled hypertension in many who are normotensive at rest during office visits. Adequate blood pressure control decreases the incidence of long-term complications such as premature coronary artery disease and heart failure that are common in these patients.^{16,17}

In addition, intracranial aneurysms occur even in normotensive patients with coarctation of the aorta, presenting as headaches or even hemorrhage due to rupture.¹⁸ All patients with coarctation of the aorta must receive endocarditis prophylaxis because they are at risk for endarteritis and endocarditis involving associated lesions.

Resection of the coarctation of the aorta with end-to-end anastomosis is the procedure of choice in most adults. Surgery done early on reduces long-term complications. Angioplasty with or without stenting is an option for coarctation, recoarctation, or residual stenosis in the absence of any paracoarctation aneurysms.

Device closure is preferred for the majority of these PDA patients.¹⁵

Percutaneous balloon valvuloplasty should be performed for moderate to severe pulmonary stenosis (characterized by a peak transvalvular gradient >50 mm Hg) even if the patient is asymptomatic.¹⁹

Pulmonary Stenosis

Pulmonary stenosis is the most common form of right-side obstruction. It results from fusion of valve leaflets and may occur as an isolated heart defect in up to 10% of patients with CHDs. On an echocardiogram, it may appear as a dome-shaped valve with fusion of the leaflets, narrow valve orifice, and increased likelihood of calcification in adulthood. Another form of pulmonary stenosis is a dysplastic valve with thickened leaflets. The former subtype is more likely to be associated with pulmonary artery dilation and aneurysms due to associated connective tissue disorder.¹¹

Pulmonary stenosis has been associated with maternal rubella, Noonan's syndrome, Williams syndrome, and Alagille syndrome. Most patients with mild stenosis are asymptomatic. Echocardiography confirms the diagnosis and assesses the severity of the transvalvular gradient, right ventricular systolic pressure, right ventricular size and function, and associated defects (Table 2). Percutaneous balloon valvuloplasty should be performed for moderate to severe pulmonary stenosis (characterized by a peak transvalvular gradient >50 mm Hg) even if the patient is asymptomatic.¹⁹ Surgical valvotomy or pulmonary valve replacement is considered when there is significant calcification, a thickened stiff dysplastic valve, and a concomitant pulmonary artery aneurysm. Pulmonary valve replacement should also be considered for severe pulmonary regurgitation following an intervention.

Conclusion

All adults with CHDs, unrepaired or repaired, require long-term follow-up monitoring for residua and sequelae. Long-term survival and quality of life depend on appropriate advice regarding bacterial endocarditis prophylaxis, risk-factor reduction, and early detection of complications. ♦

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