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ECHOCARDIOGRAPHY (JM GARDIN, SECTION EDITOR)

Congenital Heart Disease in the Adult: What Should the Adult Cardiologist Know?

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Abstract There is an increasing population of adults with congenital heart disease (CHD) due to successful pediatric medical and surgical intervention, and commensurate with this increase is a rise in heart failure, hospital admissions, and hospital costs among adult CHD patients. This group of patients requires careful long-term evaluation and follow-up of the residua and sequelae of their cardiac anomalies that arise in adulthood to prevent late complications. This article addresses congenital heart defects that are encountered in a general adult cardiology practice and reviews clinical, anatomic, and imaging features of each lesion, fundamental management issues, indications for interventions (and often re-interventions), issues related to endocarditis prophylaxis, pregnancy, and appropriateness of referral to a dedicated adult CHD program for long-term care.

Keywords Echocardiography · Adult congenital heart disease · Pregnancy and heart disease

Abbreviations

2D	Two-dimensional
3D	Three-dimensional
ASD	Atrial septal defect
AVSD	Atrioventricular septal defects
BAV	Bicuspid aortic valve
CCTGA	Congenitally corrected transposition of the great arteries
CHD	Congenital heart disease
CMR	Cardiac magnetic resonance imaging
CW	Continuous wave
DCRV	Double chamber right ventricle
D-TGA	D-transposition of the great arteries
IVC	Inferior vena cava
LV	Left ventricle
PA	Pulmonary artery
PDA	Patent ductus arteriosus
PFO	Patent foramen ovale
PR	Pulmonic regurgitation
PS	Pulmonic stenosis
PV	Pulmonic valve
PW	Pulsed wave
RV	Right ventricle
RVOT	Right ventricular outflow tract
SVC	Superior vena cava
TOF	Tetralogy of Fallot
TR	Tricuspid regurgitation
VSD	Ventricular septal defect

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Introduction

There is a significant increase in the number of adults living with congenital heart disease (CHD) in the general population as a result of advancements in neonatal and pediatric medical and surgical care. The prevalence of CHD in the adult population is now estimated at 3.0 per 1000 adults [1–4] and there

has been a commensurate rise in heart failure [5•], hospital admissions and health care costs among adult CHD patients [6•, 7]. Hence, the practicing adult cardiologist should be facile with issues that develop in adults with CHD, either unrepaired or modified by surgery or transcatheter techniques. Echocardiography remains the primary diagnostic imaging modality for assessing and following these patients; however, advanced imaging with Cardiac CTA and MRI are often additive. This review will address common CHD lesions encountered in a general adult cardiology practice; review anatomic, clinical, and echocardiographic and advanced imaging features of each lesion; and highlight management pearls and appropriate timing of referral to a tertiary adult CHD program. Endocarditis prophylaxis and pregnancy considerations will also be briefly discussed.

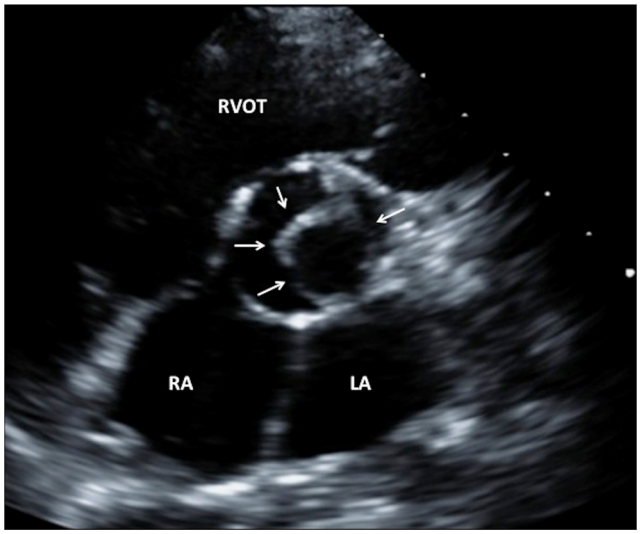

Fig. 1 Parasternal short-axis view of young adult with a unicuspid AV. Note the "keyhole" appearance with an open commissure between the non and left coronary cusps (*arrows*) (*LA* left atrium, *RA* right atrium, *RVOT* right ventricular outflow tract)

Congenital Heart Defects in Adulthood: Anatomic Features, Management and Long-Term Issues Pre and Post Repair

Left Ventricular Outflow Obstruction

Left ventricular (LV) outflow obstruction can exist at the subvalvular, valvular, supravalvular, and distal aortic levels either in isolation or in combination. A *bicuspid aortic valve* (BAV) represents the most common congenital cardiac abnormality with an estimated occurrence in 1–2 % of the population, and demonstrates multiple morphologic variants. Unicuspid unicommissural aortic valve disease, in which the only open commissure is located between the left and noncoronary leaflets, rarely presents in adulthood (see Fig. 1, Video 1). The congenitally abnormal aortic valve may be stenotic and/or regurgitant and is often accompanied by dilation of the ascending aorta due to abnormal aortic wall architecture [8, 9]. This aortopathy presents an increased risk for aortic aneurysm and dissection in patients with BAV, particularly those >50 years of age and those with significantly dysfunctional valves [9, 10].

Aortic coarctation is associated with BAV in a small percentage of cases, and in patients with Shone's complex, there may be multiple levels of left-sided obstruction, including supramitral ring, discrete subaortic membrane, congenitally abnormal aortic valve and aortic coarctation. BAVs may also be associated with atrial septal defects, ventricular septal defects and mitral prolapse, and less commonly is inherited, but familial clusters have been described [11]. Screening of first-degree relatives for BAV is recommended. Physical exam in young patients may demonstrate a prominent mid systolic ejection click, and the murmur of aortic stenosis or regurgitation. Auscultation of the scapular region is important to evaluate for concomitant coarctation. See Appendix 1 for comprehensive echocardiographic assessment.

Percutaneous aortic valvuloplasty is the therapy of choice for children and young adults with congenital aortic stenosis without significant regurgitation. After the fourth decade, the aortic valve may become thickened and calcified (commensurate with loss of the systolic ejection click) and may be less amenable to valvuloplasty. Complications of percutaneous aortic valvuloplasty include significant increase in aortic insufficiency (10–30 %), and stroke or other embolic complication if aortic calcification is present.

Discrete subaortic stenosis develops during later childhood or adulthood either de novo or following repair of membranous ventricular septal defect (VSD) or atrioventricular septal defects. It is composed of a fibromuscular ring of tissue, which may create significant obstruction to LV emptying and often causes aortic regurgitation due to valvular damage from turbulent subaortic flow. Regrowth and recurrence of LV outflow obstruction is common following surgical resection mandating regular clinical and echocardiographic monitoring. Physical exam in the adult will demonstrate a systolic LV outflow murmur, absence of systolic ejection click, and the murmur of aortic regurgitation. See Appendix 1 for echocardiographic assessment. Percutaneous balloon dilation is not recommended. Surgical membrane resection is recommended for a peak gradient of 50 mmHg or mean gradient of 30 mmHg or more by echocardiography [12] or for lesser gradients if progressive aortic regurgitation and LV dilation occur. However, each case must be considered independently as significant heterogeneity exists.

Aortic coarctation may be caused by a discrete membranous shelf or long-segment hypoplasia of the distal

transverse arch and isthmus. Half of the cases will be associated with BAV. While the initial presentation is often during infancy or childhood, later development of obstruction may also occur with clinical findings of resting or exercise-related hypertension in adult years. Re-coarctation following surgical repair or balloon dilation and/or stenting is common. Evidence of aortic obstruction may be audible in the left scapular region, and a systolic ejection click may be audible if a BAV is present. Two-dimensional (2D) echo imaging of the aortic arch with Doppler interrogation should allow detection of coarctation, especially when accompanied by Doppler sampling of flow in the abdominal aorta. Typical Doppler flow patterns in the descending thoracic aorta show some delay in systolic upstroke, turbulent high-velocity systolic flow, and continued anterograde gradient in diastole (see Fig. 2). Long-segment narrowing of the aorta, either de novo or following stent or surgical repair may cause significant systolic flow acceleration but the estimate of pressure gradient from the peak velocity is unreliable due to the pressure recovery phenomenon. See Appendix 1 for echocardiographic assessment. Cardiac magnetic resonance imaging (CMR) or CT angiography is usually necessary for full assessment of the aorta, brachiocephalic branches, and collaterals in the adult patient, and evaluation of the coarctation repair site by CMR or CT angiography should be done at least every 5 years to monitor for late sequelae including aneurysm, dissection, or re-obstruction [12].

Indications for intervention for coarctation of the aorta include symptoms related to the coarctation (leg claudication or exertional headaches), refractory hypertension or resting peak-to-peak gradient of >20 mmHg (or <20 mmHg if collaterals are present) [12]. Among patients with unoperated coarctation, balloon dilation or percutaneous stent placement may be acceptable alternatives to surgical intervention based on anatomic features. Percutaneous balloon dilation is typically employed for postsurgical restenosis at the site of prior repair. Post procedure complications include aortic dissection, very rarely acute aortic rupture, and late aneurysm formation. Patients must be followed longitudinally for development of resting or exercise hypertension (even in well-repaired coarctation patients), complications of prior intervention, and, importantly, premature coronary artery disease [12, 13]. These patients should be followed periodically at a tertiary Adult CHD Center [14].

Right Ventricular Outflow Obstruction

Valvular pulmonic stenosis (PS) represents 80–90 % of the lesions which cause right ventricular (RV) outflow obstruction. The pulmonary valve may be bicuspid or dysplastic. There is often dilation of the distal main pulmonary artery (PA). In patients with Noonan's syndrome, there may be hypoplasia of the pulmonary annulus and suprapulmonary narrowing.

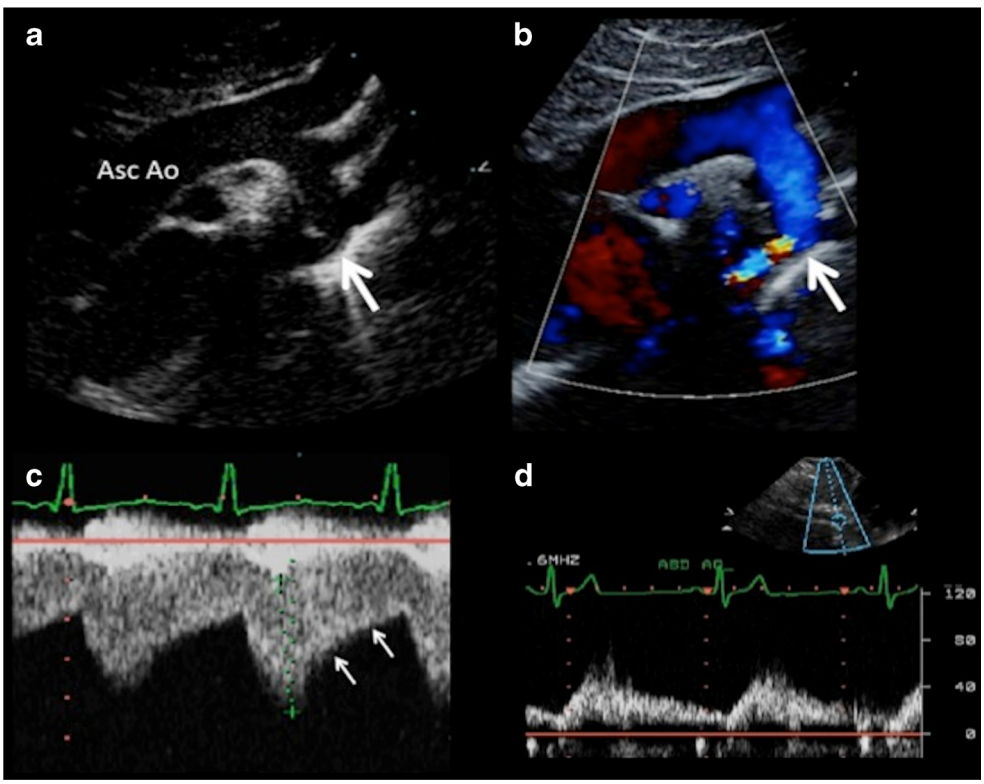

Fig. 2 Echocardiographic image obtained from the suprasternal notch. Panel **a** demonstrates narrowing and elongation of the transverse arch with discrete narrowing at the isthmus (*single arrow*). Panel **b** shows color Doppler in this view demonstrating narrowing and turbulence at the site of the discrete coarctation (*single arrow*). In panel **c**, a continuous wave Doppler profile of the descending thoracic aorta demonstrate high-velocity systolic flow through the coarctation with a persistent anterograde gradient in diastole (*double arrows*). Panel **d** is a pulsed Doppler profile obtained in the abdominal aorta which shows the typical pattern of delayed systolic upstroke, nonlaminar systolic flow and continued anterograde flow in diastole (*Asc Ao* ascending aorta)

Physical exam may reveal a systolic ejection click that decreases with inspiration, and a murmur of RV outflow obstruction, or pulmonic regurgitation. Percutaneous pulmonary balloon valvuloplasty has become the treatment of choice for patients with isolated valvular PS if the leaflets are not heavily calcified or dysplastic (i.e., systolic click is still present), and if there is no significant pulmonic regurgitation. Indications for intervention include peak transpulmonic valve gradient of ≥60 mmHg (or ≥50 mmHg in the presence of symptoms otherwise not explained) or a mean gradient of ≥40 mmHg (or ≥30 mmHg in the presence of symptoms) and absence of significant pulmonic regurgitation (PR) [12]. Postprocedural PR may develop early or years later, and serial echocardiography is indicated for monitoring of regurgitation and presence of RV dilation or dysfunction due to PR. Surgical pulmonic valve replacement is indicated for dysplastic or heavily calcified valves with moderate or more PR, and if significant PA dilation is present, pulmonary arterioplasty may be necessary.

Infundibular pulmonary stenosis is a key feature of Tetralogy of Fallot (TOF), which will be discussed in a subsequent section. Hypertrophy of the RV infundibulum may accompany stenosis at the valvular or supravalvar level and should be suspected when there is increased wall thickness and dynamic late-peaking Doppler systolic velocities in the subvalvular region. One particular form of subpulmonic obstruction is the so-called low-lying infundibular stenosis or double-chambered right ventricle (DCRV). In this entity, a fibromuscular collar develops between the RV inflow sinus and the RV outflow tract. This portion of the RV is often poorly visualized in standard echocardiographic planes, requiring a high index of suspicion to insure a correct diagnosis. A subcostal short-axis view which visualizes the right ventricular outflow tract (RVOT) as well as the main PA and branches may be the best view to obtain a Doppler gradient. DCRV can occur in association with TOF, or in combination with a membranous VSD and discrete subaortic membrane.

Supravalvular PS or branch PS is uncommon but occurs in congenital Rubella syndrome, Williams Syndrome, TOF, and Takayasu arteritis. Branch obstruction may also occur iatrogenically from previous aortopulmonary palliative shunts. Balloon dilation or stenting may be successful if the lesion is amenable.

Cardiac Shunt Lesions

Atrial septal defects are common adult congenital heart defects. Defects may occur at one or more locations within the septum (see Fig. 3). Left-to-right shunting at atrial level leads to right heart enlargement and signs of RV volume overload. Sufficiently large shunts which are uncorrected into adulthood may cause pulmonary arterial (PA) hypertension. Associated abnormalities are present in up to 30 % of patients with atrial septal defect

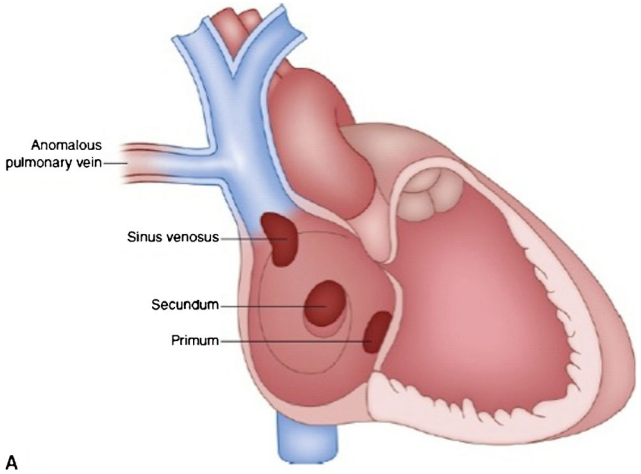

Fig. 3 Schematic diagram of the interatrial septum as seen from the right atrial surface showing the location of atrial septal defects and a typical location for partial anomalous connection of a right pulmonary vein (reprinted with permission from [47])

(ASD) which are specific to the ASD type: secundum ASD—valvular PS, BAV; primum ASD—cleft mitral valve; sinus venosus ASD—partial anomalous pulmonary venous return; coronary sinus ASD—persistent left superior vena cava (SVC), partial anomalous pulmonary venous return. Physical exam will commonly demonstrate wide and fixed splitting of S2, soft pulmonary flow murmur, precordial lift, and, if pulmonary hypertension is present, a prominent P2 at the apex. See Appendix 2 for transthoracic echocardiographic assessment.

Transesophageal echo (TEE) is often more useful for accurate visualization of ASD size and location, particularly for the superior sinus venosus ASD. Specific measurement of the septal rims around the defect and three-dimensional (3D) imaging is important if transcatheter closure is planned.

TEE, cardiac CTA, or CMR interpreted by a congenital heart disease-trained imager should also be considered for patients with unexplained RV volume overload to exclude a sinus venosus ASD or anomalous pulmonary venous return.

Most patients who undergo surgical ASD closure during childhood will have a successful return to normal cardiac size and function. However, a number of potential residua and sequelae remain after surgical ASD repair, especially when correction occurs later in life (see Appendix 2).

Device closure of ASD has become the treatment of choice for uncomplicated secundum defects with sufficient rims. Primum or sinus venosus atrial septal defects should not be closed percutaneously [12]. Occasionally, ASDs are multiple (example: concurrent secundum and sinus venosus defects) and prior to closure, the atrial septum must be carefully screened for other defects. Indications for closure include right heart enlargement, orthodeoxia-platypnea, or a history of paradoxical embolism in the presence of predominantly left-to-right shunting and less than moderate pulmonary hypertension [12]. Follow-up echocardiographic assessment is required to

assure appropriate device positioning, determine presence and degree of residual shunting, and detect complications such as thrombus formation. Aortic erosion is rare, but device proximity to aortic root must be assessed following intervention in all cases. See Appendix 2.

Ventricular septal defects are less commonly encountered in the adult population because of spontaneous closure of smaller defects or surgical repair of larger communications during childhood. Thus, the adult with a VSD either has a small, hemodynamically insignificant shunt, a large shunt which is restricted by some form of RVOT obstruction, or a large shunt with irreversible pulmonary hypertension. Associated lesions may include a membranous ventricular septal aneurysm, aortic valve prolapse into the VSD with aortic insufficiency (see Fig. 4), discrete subaortic membrane and DCRV, conal septal malalignment with RV or LV outflow obstruction, valvular PS and/or acquired infundibular hypertrophy (see Appendix 2 for echocardiographic assessment). Development of a new diastolic murmur among patients with isolated small VSDs must always be carefully evaluated for new AR or ruptured sinus of Valsalva aneurysm (see Fig. 5).

VSD closure is generally recommended when LV volume overload unexplained by alternative mechanism is present (Qp/Qs>1.5), when accompanied by progressive aortic regurgitation or sinus of Valsalva aneurysm or fistula, or when there is a history of bacterial endocarditis [12].

Muscular VSDs can occasionally be closed percutaneously. Percutaneous closure of perimembranous VSD is more challenging due to proximity of the aortic and tricuspid valve, as well as the conduction system, with postprocedural complete heart block occasionally seen. This technique is not yet approved by the United States Federal Drug Administration for perimembranous VSDs.

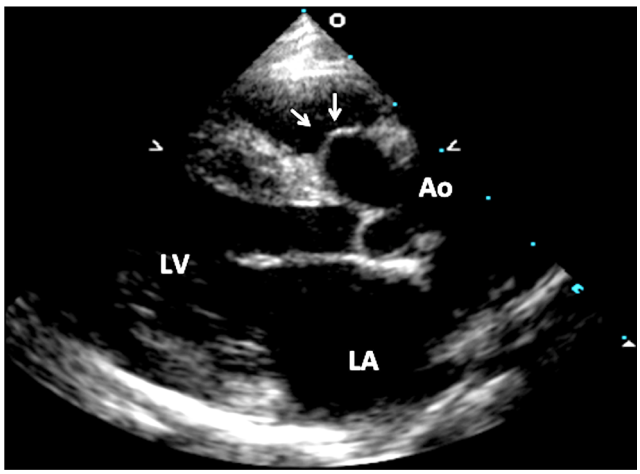

Fig. 4 Echocardiographic image of a parasternal long axis view demonstrating enlargement and prolapse of the right aortic sinus (*arrows*) into a supracristal (conal septal) VSD. The prolapsed aortic sinus obscures the VSD (*Ao* aorta, *LA* left atrium, *LV* left ventricle)

Atrioventricular septal defects (AVSD) (also termed AV canal defects or endocardial cushion defects) are a specific complex of abnormalities caused by abnormal formation of the crux of the heart, resulting in absence of the atrial and ventricular septum at the crux, and abnormalities of the atrioventricular valves. The AV valve inflow may be malaligned over the ventricular septum which results in underdevelopment of the RV or LV. AVSD is commonly seen in patients with Down syndrome. Partial or incomplete forms of this disorder exist with either an atrial or ventricular septal communication accompanied by a cleft in the mitral valve. The papillary muscle anatomy is frequently abnormal and parachute mitral valve or double-orifice mitral valve may be seen. While AV septal defects are often an isolated abnormality, they may also be associated with TOF, Ebstein's malformation of the right AV valve, and heterotaxy syndromes. See Appendix 2.

Surgical repair of AVSD consists of patch repair of the central AV septal defect and subdivision of the atrioventricular septal bridging leaflet with approximation of the cleft in the mitral valve.

Patent ductus arteriosus (PDA) is a communication between the descending thoracic aorta and the PA which is present in nearly all newborns but usually closes spontaneously within the first few months of life. Occasionally, a small PDA will persist into adult life and be detected incidentally on exam or by echocardiography. In rare instances, a large PDA may be undiagnosed during childhood and cause irreversible PA hypertension. Thus, an unrepaired PDA should always be considered in the differential diagnosis of adult patients who present with pulmonary hypertension. Shunt flow through a small PDA is readily detected by color flow Doppler as a continuous or diastolic flow stream entering the main PA near the PA bifurcation. The peak PDA jet velocity is a good indicator of the aortopulmonary pressure gradient. In the case of severe PA hypertension, the PDA flow velocities are very low and may be difficult to detect. Cardiac CTA or CMR can be utilized to demonstrate the ductal communication in these cases.

In adult patients, in the absence of severe pulmonary hypertension, closure of a PDA is reasonable to prevent LV volume load and endarteritis [12]. Repair of PDA can be accomplished by surgical ligation or division of the ductus. However, transcatheter closure using coils or devices is now the technique of choice for isolated PDAs of the appropriate geometry (see Fig. 6). Currently, for adults with a PDA, the Amplatzer ductal occluder is the most commonly used device, with coil embolization employed for smaller PDAs measuring <2 to 3 mm or for residual leaks. See Appendix 2 for echocardiographic evaluation.

Conotruncal Abnormalities

Tetralogy of Fallot is the most common of the cyanotic congenital heart lesions. Caused by anterior deviation of the conal

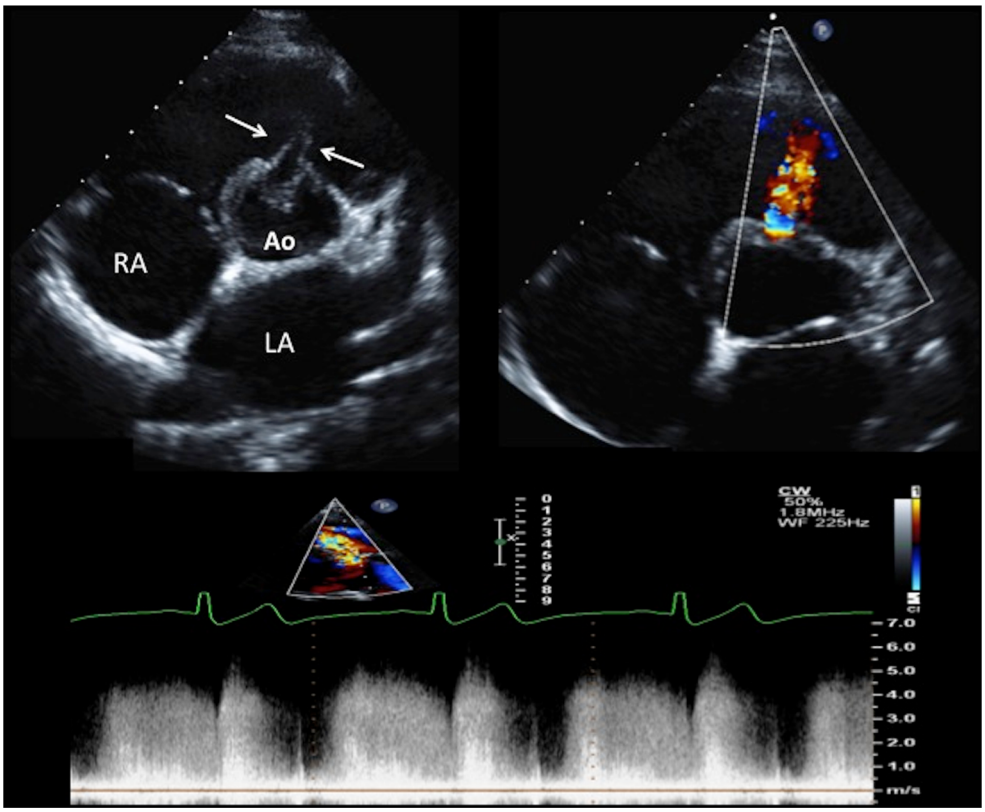

Fig. 5 Echocardiographic parasternal short axis image of a sinus of Valsalva aneurysm in a patient with a supracristal (conal septal) VSD. In the *upper left panel*, a channel protrudes into the RV outflow tract (*arrows*). The color Doppler image in the upper right panel shows flow passing through the aneurysm into the RVOT. In the *lower panel*, the CW Doppler profile shows a continuous high-velocity flow indicating that this is a fistula from an aortic source (*Ao* aorta, *LA* left atrium, *RA* right atrium)

septum, the features of the tetrad include a subaortic VSD, infundibular and valvular PS, overriding aorta, and RV hypertrophy. The degree of conal septal deviation determines a morphologic and clinical spectrum ranging from severe cyanosis in infancy to a noncyanotic patient with VSD and mild PS. Tetralogy of Fallot usually requires surgical palliation or correction in infancy; however, uncorrected TOF in the adult population may be seen in individuals who have not had access to surgical care or in cases where the degree of RVOT obstruction is mild. Associated lesions include ASD, right aortic arch (25 %), anomalous origin and course of the left coronary artery (3 %), branch PS, occasionally AVSDs, and very rarely Ebstein's anomaly. This lesion may be heritable in patients with DiGeorge syndrome (22q11 deletion). See Appendix 3 for echocardiographic assessment.

Among adults with TOF, surgical repair may have included a palliative aortopulmonary connection such as a Waterston, Potts, or Blalock-Taussig shunt (see Fig. 7). Complete repair consists of patch closure of the VSD and augmentation of the RVOT with an outflow tract patch. Although done less

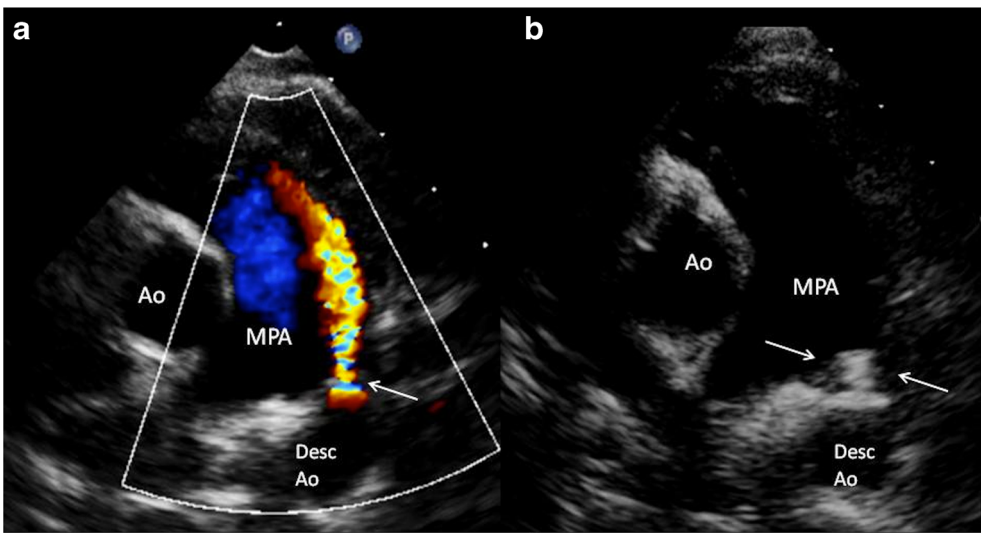

Fig. 6 Echocardiographic parasternal short-axis images of a patent ductus arteriosus before (panel **a**, *arrow*) and after device closure (panel **b**, *arrows*) (*Ao* aorta, *Desc Ao* descending aorta, *MPA* main pulmonary artery) (modified with permission from [48])

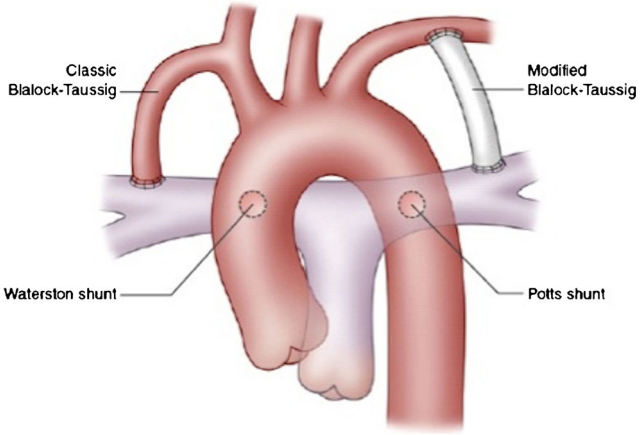

Fig. 7 Schematic diagram showing the various locations of palliative systemic to pulmonary shunts (reprinted with permission from [47]

frequently now, transannular extension of the RVOT patch had been the standard surgical practice, resulting in significant pulmonary regurgitation (see Fig. 8). In the adult patient post repair, physical examination may reveal a short diastolic murmur of severe PR, and there may be a systolic ejection murmur or cannon A waves if there is residual PS. A harsh VSD murmur may be heard if there is a residual patch-related defect. An RV lift or gallop may also be present. The longstanding RV volume overload that PR imposes leads to development of RV dilation and dysfunction, and if PR is not intervened upon in a timely way, RV dilation and dysfunction will become irreversible [15], increasing a patient's risk for chronic right heart failure, and development of cirrhosis from chronic congestion. Echocardiography (see Appendix 3) and cardiac MRI play an important role in evaluating the severity of PR [16] (see Fig. 8) and RV size and resting function, helping to determine the

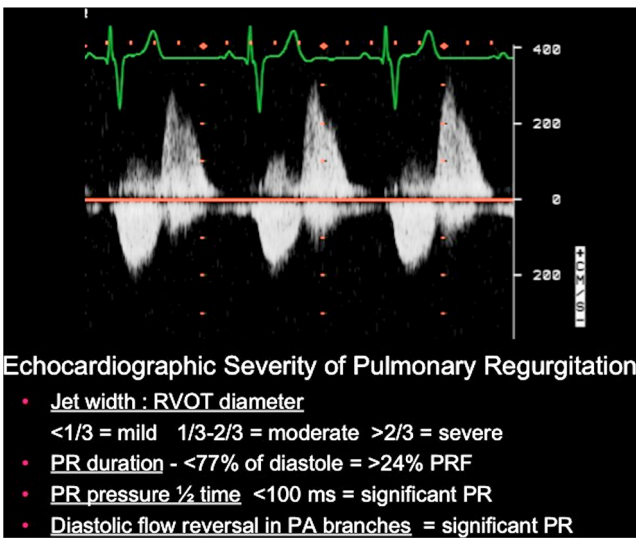

Fig. 8 Continuous Wave Doppler profile from a patient with severe pulmonary regurgitation following repair of Tetralogy of Fallot. Several echocardiographic measures of the severity of pulmonary regurgitation are shown. *PRF* pulmonic regurgitation fraction

optimal timing of re-operation for pulmonary valve replacement [17••]. Additionally, functional exercise testing may be helpful to determine right ventricular reserve, as poor RV reserve may correlate with adverse outcomes of heart failure and arrhythmia. Risk of sudden cardiac death increases with QRS duration >180 ms [18], RV dilation, and dysfunction, and if ambient ventricular arrhythmias are present [19]. Most importantly, patients with TOF should be followed at a dedicated tertiary ACHD Center at least annually, and imaging reviewed by a congenitally trained echocardiographer or radiologist [20] (see Appendix 3).

Some patients may have residual infundibular, valvular, or branch pulmonary stenosis, and low-lying infundibular stenosis (DCRV) can develop postoperatively. Aortic root enlargement may be a progressive problem, particularly in patients with prior aortopulmonary shunts or with pulmonary atresia; however, aortic dissection is rare. Left ventricular dysfunction is also a postoperative problem in cases with longstanding aortopulmonary shunts, earlier era of surgical repair, severe RV dilation, or unrecognized coronary anomalies. Patients with pulmonary atresia or those with a coronary artery crossing the RVOT may need an RV to PA conduit, or Rastelli procedure. Subsequent conduit obstruction or deterioration is common, requiring stenting or conduit replacement [21]. Transcatheter pulmonary valve implantation with the Melody Valve (Medtronic Inc., Minneapolis, MN) is currently approved for use among patients with dysfunctional Rastelli conduit and recent data suggests very good short and medium term success [22]. A growing population of postoperative TOF patients includes those who have had mechanical or biologic prosthetic pulmonary valve replacements, with subsequent issues related to prosthetic valve dysfunction or infection. Placement of a transcatheter pulmonary valve within a failed bioprosthesis has also been evaluated in a multicenter experience recently and can be accomplished with high rate of success, a low procedural morbidity and mortality, and excellent short-term results [23•].

Complete transposition of the great arteries (D-TGA) is another common cause of cyanosis in the newborn period. In this entity, the aorta arises from the morphologic RV and the PA arises from the morphologic LV (see Fig. 9a). There is nearly always an interatrial communication or PDA and a VSD is present in 45 % of cases. Subpulmonary LV outflow tract obstruction occurs in about 25 % of patients. Anomalies of coronary artery origin and course also occur. Survival into adulthood without surgical repair is very poor. Patients with D-TGA should have regular follow-up at a dedicated tertiary ACHD Center [14].

Surgical repair for D-TGA prior to the 1980s was directed at reversing the systemic and pulmonary venous inflow to the heart by placing baffles within the atria to re-direct flow (Mustard or Senning procedure) (see Fig. 9b). This technique had low operative mortality and good early clinical outcomes.

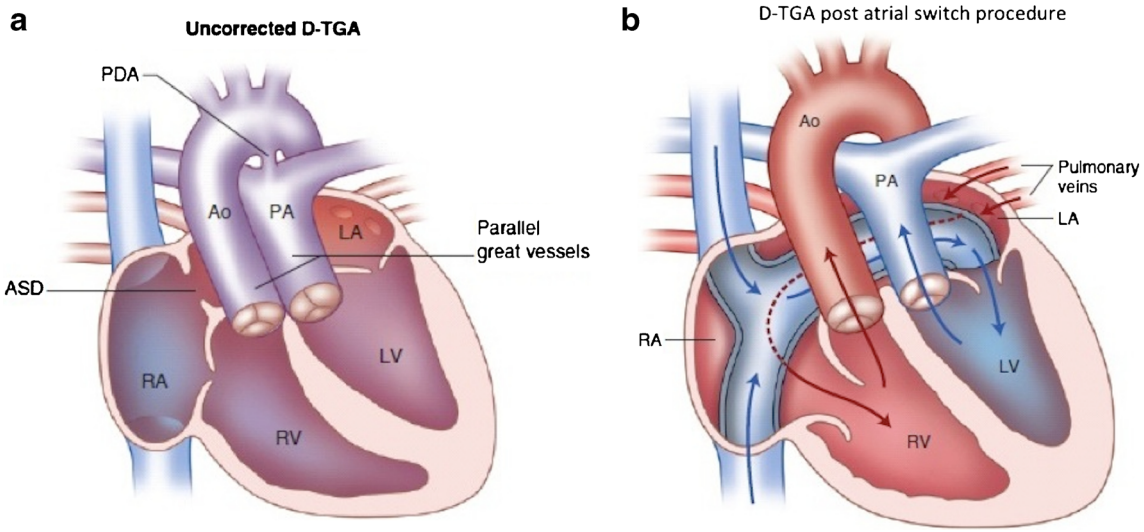

Fig. 9 **a** Schematic diagram of the native anatomy in D-transposition of the great arteries. (*Ao* aorta, *ASD* atrial septal defect, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *PDA* patent ductus arteriosus, *RA* right atrium, *RV* right ventricle) (reprinted with permission from [47]). **b** Schematic diagram of an atrial switch procedure for D-transposition of the great arteries. The atrial septum is resected and a baffle is fashioned to direct systemic venous flow (*blue arrows*) from the caval entries to the mitral valve. The pulmonary venous flow (*red arrows*) passes behind the baffle to cross the tricuspid valve (*Ao* aorta, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle) (reprinted with permission from [47])

ECG post atrial switch procedure will demonstrate a wide RBBB, with RVH and strain pattern consistent with systemic RV. However, among patients with D-TGA who have undergone an atrial switch operation, up to 25 % will demonstrate late baffle leaks, and those with large defects may undergo percutaneous closure (see Appendix 3). Baffle obstruction is observed in up to 15 % of those with prior atrial switch procedure, and balloon angioplasty can result in long-term improvement. In experienced hands, stent deployment is highly successful in relieving obstruction with low complication rates [24]. Patients with atrial switch procedure and systemic right ventricles are at high risk for heart failure. Systemic hypertension and systemic tricuspid regurgitation (TR) may provide pressure and volume loads that the systemic RV does not tolerate well in the long term, therefore counseling regarding sodium restriction and maximal afterload reduction is of paramount importance at an early age to prevent RV deterioration. Trials with angiotensin receptor blockers have not definitely shown an improvement in short-term outcomes (3 years) but have shown a trend towards improved RV mass and volumes [25].

The arterial switch operation is now the repair of choice for D-TGA and involves resection and transfer of the aorta and PA to their appropriate ventricle of origin, with translocation of the coronary arteries to the neo-aortic root. This results in a corrected circulation with a near normal echocardiographic appearance. ECG postarterial switch may look entirely normal with a narrow QRS. Postoperative problems may result from supra-AS or supra-PS at the anastomosis site, neo-aortic valve regurgitation, tethering or branch PS due to stretch on the main PA as it is relocated anteriorly, and coronary artery stenosis or occlusion following translocation (see Appendix 3).

Congenitally corrected transposition (CCTGA) is an uncommon congenital abnormality in which inversion of the ventricles and transposition of the great arteries creates an anatomically abnormal but physiologically corrected cardiac state. The systemic venous return passes through the mitral valve into the anatomic LV and from there into the PA. Pulmonary venous blood passes through the tricuspid valve into the anatomic RV and thence into the aorta. About 95 % of patients have situs solitus and the heart is often mesocardic. Associated abnormalities are present in 90 % of cases and include VSD (70 %), valvar or subvalvar PS (40–50 %), and abnormalities of the systemic tricuspid valve (90 %) including an Ebstein-like displacement [26] (see Appendix 3). Heart block is common due to the abnormal course of the conducting system, occurring at a rate of approximately 2 % per year [27, 28]. The coronary arteries supply their anatomic ventricle and thus are inverted: left coronary artery from the right facing aortic sinus, right coronary artery from the left-facing sinus. Similarly, the electrical bundles are also inverted leading to the classic CCTGA electrocardiographic pattern (see Fig. 10) and coronary anomalies can also occur including single and commissural coronary origins.

Patients with CCTGA and no associated anomalies may be undiagnosed until later in life and have no cardiovascular symptoms. Those with an associated VSD, PS, tricuspid valve abnormality, or heart block usually come to medical attention during childhood or early adulthood. The major clinical problem in CCTGA relates to long-term function of the morphologic RV as it assumes the role of systemic pump impacted by volume

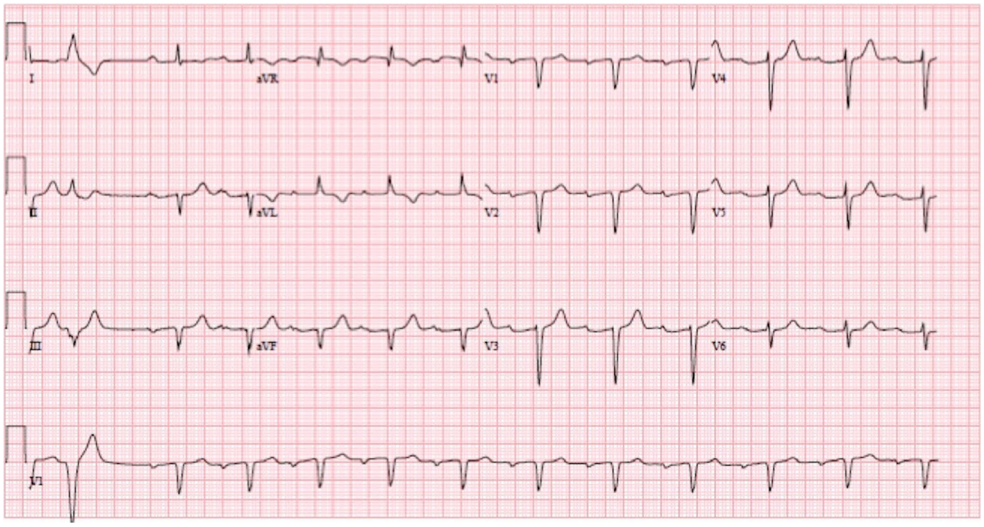

Fig. 10 Electrocardiogram of a patient with congenitally corrected transposition of the great arteries. Left and right bundles are inverted. Note Q waves V2-V4, S waves in V5-V6, Q waves inferiorly, prolonged PR interval

overloading from VSD or TR. As with patients with D-TGA and atrial switch procedures with systemic RVs, patients with CCTGA should be strongly counseled regarding Na restriction and maximal afterload reduction starting at an early age, and should be regularly followed at a tertiary ACHD center.

Surgical management for the adult with CCTGA is often a nonanatomic repair directed towards closing the VSD and reducing the volume load on the systemic right ventricle from a regurgitant tricuspid valve. This may involve tricuspid valve repair or replacement [29, 30].

The MitraClip (Abbott Vascular, Santa Clara, CA) procedure is currently approved for patients with acquired functional mitral regurgitation as an alternative to surgical mitral valve repair [31]. This procedure has promise for potential percutaneous repair of systemic TR among patients with CCTGA [32].

An alternative surgical repair for CCTGA is an anatomic approach to allow the LV to become the systemic ventricle. This requires a so-called double switch procedure to baffle the venous return to the opposite ventricle and an arterial switch or a Rastelli procedure to connect the great arteries to the appropriate ventricle [33]. Although this involves a considerable degree of re-structuring, it can be accomplished successfully in infancy before the LV becomes deconditioned. Use of this technique in later years requires that the LV be functioning at systemic pressure or prepared for pressure loading with a PA band. Subsequent LV dysfunction may be problematic in follow-up. Mortality and morbidity are higher for this procedure in adult patients [33, 34].

In some patients with CCTGA and severe systemic RV dysfunction, cardiac transplantation may be the only surgical option.

Ebstein's Malformation

Ebstein's anomaly of the tricuspid valve is a malformation with a widely variable anatomic and clinical spectrum. Failure of delamination of the tricuspid leaflets during embryologic formation of the AV valves results in a tethered anterior leaflet and apically displaced septal and posterior leaflets (see Fig. 11, Video 2). Significant TR is a common clinical problem. The right atrium and atrialized RV may become markedly dilated. The true RV is often quite small, myopathic and RV contractility may be impaired. LV function may also be compromised from impingement of the dilated right heart. The majority of patients have either an ASD or patent foramen ovale (PFO). Associated lesions include PS or pulmonary atresia, and atrial

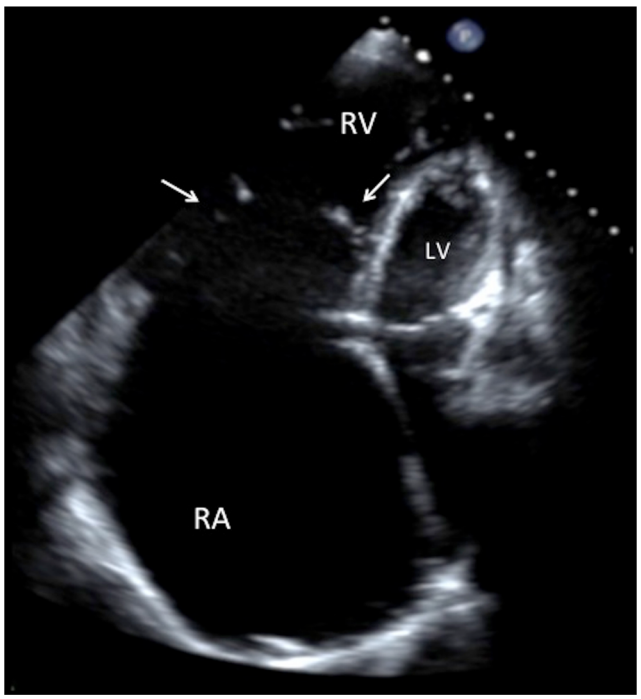

Fig. 11 Apical four-chamber view in a patient with Ebstein's anomaly of the tricuspid valve. The image shows the tethering and apical displacement of the tricuspid valve leaflets (*arrows*) with dilated right atrium and atrialized RV (*LV* left ventricle, *RA* right atrium and atrialized RV, *RV* right ventricle)

arrhythmias are common. See Appendix 3 for echocardiographic assessment. Patients with Ebstein anomaly should be followed regularly at an adult CHD center [14].

Transcatheter device closure of the ASD or PFO may be indicated in some patients with sizable L-R shunts or paradoxical embolus if the degree of TR is mild-to-moderate and test occlusion indicates that the right heart will tolerate an intact atrial septum [35].

Surgical treatment of patients with Ebstein's malformation is indicated for progressive exercise intolerance, cyanosis, right heart failure, or paradoxical embolus; however, this surgery is high risk and must be performed at an ACHD center by a surgeon with experience in Ebstein's anatomy. The goals of surgery are closure of the atrial communication, arrhythmia treatment by surgical interruption of bypass tracts or a Maze procedure, and tricuspid valve repair or replacement. If RV function is significantly impaired, it may be helpful to unload the ventricle with a concomitant Glenn (SVC to PA) shunt. Complete unloading with a Fontan procedure, or a cardiac transplant, may be required in cases with severe RV dysfunction (see Appendix 3). Percutaneous tricuspid valve replacement with the Melody Valve has also been described in a small cohort of CHD patients with failure of a bioprosthesis in the tricuspid position, either due to stenosis or regurgitation [36].

Complex Congenital Defects with Single Ventricle Physiology

Complex congenital heart disease represents a very small proportion of adults with CHD (3–5 %) [1] but a larger percentage of these patients are surviving with current medical and surgical management, and are at high risk of hospital admission and resource utilization [37, 38]. Evaluation of the patient with complex CHD relies heavily upon knowledge of the initial anatomy and any interventional or surgical treatment. This should be obtained from patient history and complete medical and operative records. Imaging these patients is complex and must be approached systematically (see Appendix 3). Additionally, all patients with complex congenital diagnosis should be referred and followed at a tertiary ACHD center for routine follow-up and imaging [14].

Many of the complex congenital lesions result in single ventricle physiology: tricuspid atresia, mitral atresia, double-inlet LV, single ventricle, unbalanced AV canal, hypoplastic left heart, hypoplastic right heart, and heterotaxy syndromes. These lesions are not amenable to a two-ventricle surgical repair and would be considered for a *Fontan procedure* to directly route systemic venous return to the pulmonary circulation, allowing the functional ventricle to perform as the systemic pump. A number of different versions of this procedure have been employed since its inception: right atrial appendage-to-PA connection, lateral interatrial tunnel with cavopulmonary anastomosis, and extracardiac pathway with cavopulmonary connection (see Fig. 12) [39]. A fenestration is often created in the lateral or extracardiac tunnel to allow decompression of the high central venous pressures in the immediate postoperative period (see Videos 3 and 4). The fenestration is usually repaired at a later time by device closure (see Fig. 13).

It is critical to know the type of Fontan procedure performed when trying to evaluate the patient echocardiographically. A well-functioning Fontan pathway is expected to have nondistended cavae with low velocity, respirophasic anterograde flow in caval, lateral tunnel or extracardiac pathways, and branch pulmonary arteries (see Appendix 3). Patients who have undergone a Fontan procedure may also develop conduit leaks or conduit obstruction. Device closure, balloon angioplasty, and stent placement are all employed to preserve conduit integrity [24]. Late postoperative issues also include arrhythmias (both atrial and ventricular), ventricular dysfunction, intracardiac thrombus formation and embolization, protein-losing enteropathy and hepatic dysfunction for which routine screening is recommended [40]. Arrhythmia therapy should avoid agents like calcium channel blockers with

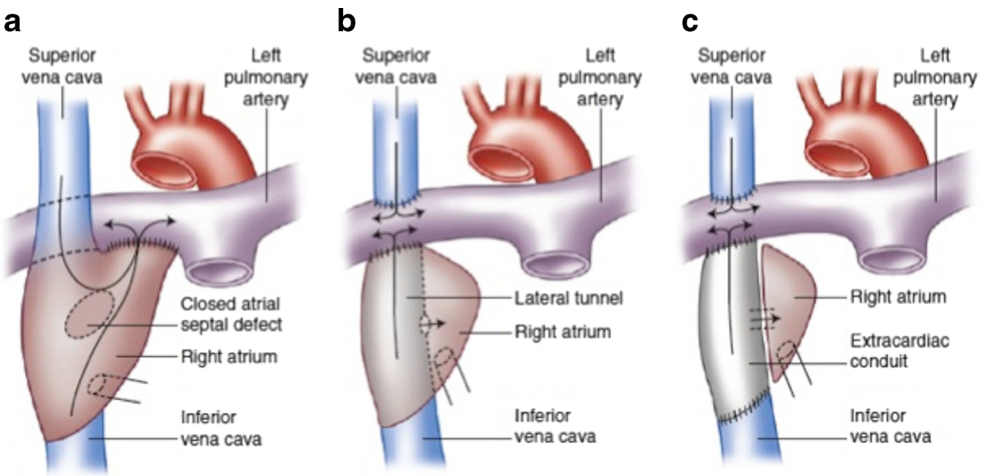

Fig. 12 Schematic diagram of the various types of Fontan repair. Panel **a** demonstrates the earlier type of repair with right atrial appendage anastomosis to the pulmonary artery. Panel **b** shows the lateral tunnel repair with cavopulmonary anastomosis. Panel **c** shows the extracardiac Fontan (reprinted with permission from [47])

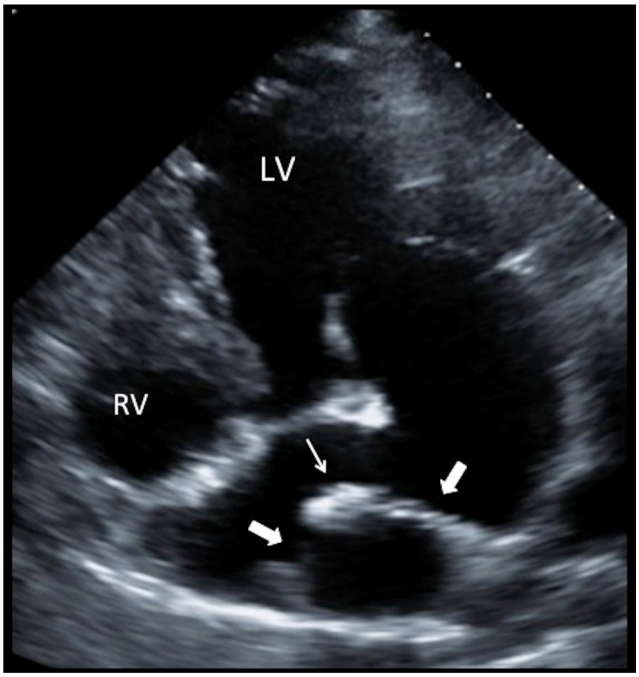

Fig. 13 Apical four-chamber view in a patient with tricuspid atresia post fenestrated lateral tunnel Fontan palliation. At the tricuspid annulus is a fibrous band of tissue, and the right ventricle is hypoplastic. The Fontan pathway through the atrium is seen as a circular structure within the right atrium (*large arrows*). There is an Amplatzer closure device within the baffle closing a fenestration (*small arrow*) (*LV* left ventricle, *RV* right ventricle)

negative ionotropic activity, as they may precipitate worsening heart failure. Sodium restriction and afterload reduction also remains of paramount importance.

Endocarditis Prophylaxis Among Adult Congenital Heart Patients

The risk of endocarditis among CHD patients is increased compared to the general population with an increase in associated mortality [41]. However, prophylactic antibiotics has not been definitively shown to change this rate of endocarditis among many patients and in 2007 revised guidelines recommend prophylaxis only for patients at highest risk of morbidity or mortality from infective endocarditis including patients with (1) prosthetic heart valves or prosthetic material used for cardiac repair, (2) prior history of endocarditis, (3) CHD including unrepaired cyanotic CHD, completely repaired congenital heart defects with prosthetic material or device during the 6 months post procedure and repaired CHD with residual defects at the site or adjacent to the site of prosthetic device or (4) patients post cardiac transplantation with valvulopathy [42]. These guidelines significantly changed prior recommendations for patients with simple congenital valvular lesions such as BAV, or unrepaired VSDs or ASDs. A subsequent study from the Dutch National CONCOR registry elucidates increased risk of developing infective endocarditis among adults with acyanotic, unrepaired CHD including VSDs, ASDs, BAVs, and coarctation of the aorta; however, they emphasize the critical importance of individually tailoring surveillance, prophylaxis and counseling among adolescents and adults with CHD [43]. Education regarding prompt attention to fevers, rapid treatment of skin infections, and early recognition of signs and symptoms of endocarditis is of paramount importance for all at-risk adult CHD patients.

Pregnancy in the CHD Patient

An increasing number of women with CHD are surviving to childbearing age. Maternal mortality is fortunately uncommon, with the exception of patients with Eisenmenger syndrome and Marfan syndrome with aortopathy, where pregnancy is contraindicated [44, 45]. During pregnancy, there is a significant increase in stroke volume, heart rate, and cardiac output and simultaneous decrease in systemic vascular resistance. These adaptations reach a maximum during labor and delivery and volume shifts persist post delivery. As a consequence of these physiologic changes, women who were asymptomatic prior to pregnancy may develop symptoms during pregnancy and may present with a new diagnosis of CHD [46]. Simple or moderate congenital heart defects may be uncovered, such as ASDs, PDAs, coarctation of the aorta or CCTGA. It is critically important that women with known underlying congenital heart disease are routinely counseled regarding the risks of pregnancy and potential cardiovascular complications that could occur during pregnancy or post delivery, including arrhythmia and heart failure, and are followed very closely by a congenital cardiologist comfortable with cardiac disease in pregnancy. Additionally, women with more complex CHD or cyanosis who present during pregnancy must be carefully assessed regarding both maternal and fetal risks of pregnancy and monitored closely, and deliver in a center with cardiology and cardiac surgical support [44].

Conclusions

Successful treatment of children with CHD has led to an increasing number of adults with unoperated, palliated, and repaired CHD entering the general adult cardiology practice. Echocardiography remains the mainstay for diagnosis and follow-up for this population. Knowledge of the specific anatomic and functional issues for the wide variety of congenital anomalies is critical to insure appropriate assessment and timely intervention or reintervention to prevent long-term complications including arrhythmia and heart failure. Importantly, all adult patients with CHD, even those with simple lesions, benefit from at least a

one-time visit in a tertiary ACHD center to ensure cardiac anatomy is appropriately delineated, associated lesions are not overlooked, and a long-term management plan is established. Patients with moderate and complex congenital disease should be referred to a tertiary ACHD center for more regular monitoring and imaging by a congenitally trained imager. Endocarditis prophylaxis should be individualized and finally, patients with CHD are now reaching childbearing age and careful cardiac follow-up during pregnancy and post delivery facilitates successful maternal and fetal outcomes.

Compliance with Ethics Guidelines

Conflict of Interest Doreen DeFaria Yeh and Mary Etta King declare that they have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

Appendix 1. Echocardiographic Essentials for Assessment of Left and Right Heart Obstruction

Bicuspid Aortic Valve:

1. Morphologic aortic valve features, location of commissural fusion and opening
2. Degree of stenosis and regurgitation
3. Estimated aortic valve area
4. Dimensions of the aortic annulus, sinuses of Valsalva, and ascending aorta
5. LV size, mass, and function
6. Associated left sided obstructive lesions including coarctation, subaortic stenosis, and supramitral ring, additionally ASDs, VSDs and mitral abnormalities

Discrete Subaortic Stenosis:

1. The nature and location of the membrane
2. The degree of LVOT obstruction and aortic valvular regurgitation
3. Aortic annular dimension and aortic valve morphology
4. LV size, mass, and function.
5. Associated defects such as membranous VSD and DCRV

Aortic Coarctation (native or recurrent coarctation):

1. Aortic annulus, sinuses, ascending aorta, transverse arch, isthmus, and descending thoracic aortic dimensions
2. Brachiocephalic branch anatomy
3. Pulsed wave (PW), continuous wave (CW) and color Doppler sampling of the transverse and descending thoracic aorta
4. Pulsed Doppler sampling of the abdominal aorta
5. LV size, mass, and function
6. Aortic valve anatomy and function
7. Associated lesions such as VSD, subaortic membrane, BAV and mitral valve anomalies (mitral prolapse, parachute mitral valve, supramitral ring, aberrant papillary muscle arrangement), aortic arch abnormalities and aberrant subclavian arteries

Valvular PS:

1. Pulmonary valve leaflet morphology and thickness
2. Annular, main PA, and branch dimensions
3. CW Doppler peak and mean gradient
4. Degree of regurgitation
5. Subvalvular hypertrophy or obstruction
6. RV size, wall thickness, and function
7. Doppler TR peak velocity
8. Associated ASD, PFO, VSD, other RV outflow obstruction

Infundibular PS:

1. 2D imaging of the mid and distal RV outflow tract in parasternal and subcostal views
2. Color and PW/CW Doppler sampling of the RV outflow tract proximal to the pulmonic valve
3. Pulmonary valve morphology
4. Doppler TR peak velocity

Appendix 2. Echocardiographic Essentials for Evaluation of Congenital Shunt Lesions

Atrial septal defects (native or preprocedural imaging):

1. ASD size and location from multiple windows
2. Septal rims
3. RV size and function
4. Estimated RV and PA pressure from TR and PR jet velocity
5. Associated lesions—PS, mitral valve prolapse, cleft mitral valve, anomalous pulmonary veins, persistent left SVC

Postoperative ASD assessment:

1. Residual shunting
2. Residual pulmonary hypertension
3. RV enlargement and dysfunction

4. RV or RA thrombus
5. Mitral regurgitation from cleft MV or mitral valve prolapse
6. SVC or pulmonary vein stenosis after sinus venosus ASD repair

Post device closure ASD assessment:

1. Device position
2. Residual shunting
3. Device impingement upon systemic or pulmonary venous inflow
4. Device impairment of aortic, mitral or tricuspid valve function
5. Device impingement upon posterior aortic wall
6. Thrombus or vegetation on right or left atrial device facets

Native Ventricular Septal Defect:

1. Size and location of VSD
2. Direction of shunt flow; LV to RV shunting, LV to RA shunting
3. LA size
4. LV size, mass, and function
5. VSD gradient by CW Doppler
6. Estimated RV and PA pressure from TR and PR velocities
7. Presence and degree of RVOT obstruction—DCRV, infundibular, valvar, or branch PS
8. Associated lesions—membranous septal aneurysm, aortic valve prolapse, or discrete subaortic membrane

VSD: Following surgical repair:

1. Residual VSD shunt
2. Residual pulmonary hypertension
3. LV dysfunction
4. TR due to surgical distortion of the septal leaflet
5. Aortic valve prolapse with aortic insufficiency
6. Aortic valve distortion and dysfunction due to VSD patch placement
7. Discrete subaortic membrane and DCRV

Unrepaired Atrioventricular Septal Defect:

1. Size and location of atrial and ventricular defects
2. Presence, direction, and size of ASD and VSD shunt
3. Estimate of RV and PA pressure from TR and PR velocities
4. Specifics of atrioventricular valve anatomy—common AV valve, septal attachments of anterior and posterior bridging leaflets (Rastelli Type A, B, or C); unbalanced alignment
5. Degree of left or right AV valve regurgitation
6. Papillary muscle anatomy and chordal attachments
7. LV size, mass, and function
8. RV size, wall thickness, and function
9. Associated defects—PDA, coarctation, subaortic obstruction

Postoperative AVSD:

1. Residual atrial or ventricular shunt; possible LV-right atrial shunting
2. Residual pulmonary hypertension
3. AV valve regurgitation
4. Discrete subaortic stenosis
5. Assessment of other co-existing preoperative issues

PDA pre- and postintervention:

1. Size and shape of the ductal channel
2. Size and direction of shunt,
3. Aortopulmonary gradient from peak systolic PDA jet velocity
4. LA and LV chamber size; LV function
5. Main PA and branch PA dimensions
6. Aortic arch and isthmus anatomy
7. Device or coil position with specific attention to residual shunt, branch PA stenosis or aortic protrusion

Appendix 3. Echocardiographic Essentials for Assessment of Complex Congenital Heart Disease

Unoperated patient with TOF:

1. VSD location, size and shunt direction; additional VSDs
2. Degree and nature of RV outflow tract obstruction
3. RVOT and branch PA gradients
4. Pulmonary annulus, main and branch PA dimensions
5. RV size, wall thickness, and quantitative assessment of function
6. Coronary artery origins and course
7. Atrial septal anatomy
8. Aortic arch situs
9. Aortopulmonary collaterals

Postoperative patient with TOF:

1. Residual VSD
2. Residual infundibular, valvular or branch PS; DCRV
3. Degree of PR (CW across RV outflow tract)
4. Residual atrial shunting
5. RV volumes, quantitative systolic, and diastolic function
6. Estimated RV pressure from Doppler peak TR jet velocity

7. Aortic valve anatomy and function
8. Aortic root dimensions
9. LV size, global and segmental function
10. Conduit location, size, and gradients, if present
11. Prosthetic pulmonic valve (PV) stenosis or regurgitation, if present

D-Transposition of the Great Arteries after Mustard or Senning procedure (atrial switch):

1. Systemic RV size and function
2. Subaortic and aortic stenosis
3. Subpulmonary LVOT obstruction
4. Estimated subpulmonary ventricular pressure from mitral valve regurgitant velocity
5. Tricuspid valve (systemic AV valve) regurgitation
6. Baffle pathway obstruction
7. Baffle leak (consider agitated saline study if leak not obviously present by Doppler)
8. Associated lesions—VSD, valvar or branch PS, PDA, coarctation

Postarterial switch operation for D-TGA:

1. Aortic root, supravalvar aortic dimensions, and gradient
2. Neo-aortic valve regurgitation
3. Neo-pulmonic valve function
4. Suprapulmonary stenosis in MPA or branches
5. LV size, mass, global and segmental function
6. RV size, wall thickness and function
7. Proximal coronary artery patency
8. Associated lesions—VSD, coarctation

Congenitally Corrected Transposition of the Great Arteries (CCTGA):

1. Identification of visceroatrial situs
2. Establishment of cardiac position—levo-, meso-, or dextrocardia
3. Demonstration of atrioventricular and ventriculoarterial discordance (RA-LV-PA; LA-RV-Aorta)
4. Location, size, and direction of VSD shunt if present
5. Nature and degree of PS or subpulmonary outflow obstruction
6. Anatomy and function of tricuspid valve (systemic AV valve)
7. Size and function of the RV (systemic ventricle)
8. Coronary artery anatomy

Postoperative patient with CCTGA:

1. Biventricular size and function
2. Residual VSD
3. Residual RV or LV outflow obstruction
4. Degree of TR
5. Atrial baffles if present
6. Great artery anastomoses if present
7. Conduit function if present
8. Neo-aortic size and insufficiency
9. Coronary artery patency if translocated

Ebstein's anomaly:

1. Tricuspid leaflet origin, position, size and tethering
2. Degree of TR
3. RA and atrialized RV size and function
4. True RV size and function
5. Pulmonary valve stenosis
6. Main and branch PA dimensions
7. Atrial septal communication and shunt direction
8. LV size and function

Postoperative Ebstein's anomaly:

1. Anatomic assessment of tricuspid valve repair or replacement
2. Degree of TR, presence of paravalvular leaking
3. RA size
4. RV size and function
5. LV size and function
6. Patency of Glenn shunt if present
7. Patency of Fontan pathway if present
8. ASD device position and residual shunt if present

Complex CHD: must be individualized to the specific lesion but includes the following basic features:

1. Visceroatrial situs—solitus, inversus, indeterminate
2. Cardiac position—levocardia, mesocardia, dextrocardia
3. Number of atrial chambers and morphology
4. Number of ventricular chambers and morphology
5. Atrioventricular relationship
6. Number of great arteries and their relationship
7. Ventriculoarterial relationship
8. Intracardiac shunts
9. Outflow tract obstruction
10. Other associated anomalies—coarctation, branch PS, systemic and pulmonary venous abnormalities

Complex CHD following the Fontan procedure:

1. Inferior vena cava (IVC) and SVC size and flow pattern
2. Right atrial size
3. Right atrial to PA anastomosis patency and flow patterns, if present
4. Interatrial or extracardiac venous conduit flow patterns

5. Branch PA size and flow pattern
6. Thrombus within the systemic venous pathway
7. Baffle fenestration flow and Doppler gradient
8. Pulmonary venous inflow and left atrial size
9. Atrioventricular valve function
10. Systemic ventricular size and function
11. Outflow tract obstruction
12. Aortic valve stenosis or incompetence

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