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ECHOCARDIOGRAPHY (JM GARDIN AND AH WALLER, SECTION EDITORS)

The Adult with Post-operative Congenital Heart Disease: a Systematic Echocardiographic Approach

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Abstract

Purpose of Review This article attempts to review some of the commonly seen repaired congenital heart defects in the adult population (ACHD), with a focus on important echocardiographic findings that may assist a practitioner in recognizing and managing this group of patients.

Recent Findings The prevalence of ACHD population is increasing, and currently, there are over 1 million adults with congenital heart disease. At the current time, the total number of ACHD patients exceeds the total number of pediatric patients with CHD. The recently released 2018 American College of Cardiology/American Heart Association guidelines for the management of adults with congenital heart disease (ACHD) recommends transthoracic echocardiography for all ACHD patients for initial assessment and serial assessment as needed (class of recommendation: I) and echocardiography remains the mainstay for diagnosing and managing these patients in adjunct with other available imaging modalities.

Summary It is imperative for all cardiologists to be familiar with the echocardiographic features of the commonly seen repaired CHD in adult population.

Keywords Adult congenital heart disease · Echocardiography

Introduction

The incidence of congenital heart disease (CHD) is estimated to be approximately 1 per 100 live births [1]. Due to the multiple advances in pediatric cardiology/cardiac surgery, up to 90% of children with CHD now survive into adulthood with average life expectancy of 75 years [2, 3]. A comprehensive echocardiographic evaluation with careful attention to segmental anatomy remains the cornerstone in (i) establishing a de novo diagnosis in the unrepaired adult patients with CHD and (ii) identifying the long-term hemodynamic complications associated with the surgical palliation/repair. This may need to be supplemented by additional imaging modalities (cardiac CT/MRI) and review of past cardiac surgical records whenever applicable.

The scope of this review is limited to congenital heart lesions that are commonly encountered in the adult echocardiographic laboratory and will be presented as two separate articles. The current article focuses on echocardiographic evaluation of adult patients with repaired/palliated CHD lesions.

Tetralogy of Fallot

TOF is a common form of cyanotic heart disease and most likely to survive into adulthood. It has an incidence of 0.34 per 1000 live births [1]. The primary pathology associated with

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TOF is the anterior-leftward and cephalad displacement of the conal/infundibular septum. The constellation of abnormalities that result from this pathology is the usual 'tetrad' of a ventricular septal defect (VSD) (usually peri-membranous), an aorta (Ao) overriding the ventricular septum, subvalvular (and often valvular) pulmonary stenosis, and right ventricular (RV) hypertrophy. Depending upon the degree of RV outflow tract obstruction, a wide phenotype of pre-operative presentations is possible, from the so-called "pink tet" infants who appear asymptomatic, to severely cyanotic neonates requiring early intervention (Supplemental Fig. 1). Indeed, it is the pulmonary valve and arteries which dictate both the surgical approach and the outcome in TOF.

In the current era, TOF is frequently not significantly cyanotic early in life and is repaired typically and semi-electively at 4–6 months of life. Repair consists of patch closure of the VSD to the overriding Ao and resection of the infundibular stenosis. If the pulmonary valve is hypoplastic/stenotic/dysplastic, the pulmonary annulus is incised and a patch is placed across the annulus to enlarge it, the so-called transannular patch. This style of surgery is frequent and serves to relieve the RV outflow obstruction at the cost of often significant pulmonary regurgitation. Pulmonary regurgitation is generally well tolerated in children, but the inevitable right ventricular dilation, even in the absence of symptoms, creates a clinical dilemma in adults. A RV to pulmonary artery (PA) conduit may be placed if there is an extreme form of TOF such as pulmonary atresia or if there is an anomalous left coronary artery which crosses the RV outflow tract and precludes the use of a transannular patch (Supplemental Fig. 2). Complete repair of TOF may be preceded in some cases by a Blalock-Taussig-Thomas (BTT) shunt, which is a shunt between one of the innominate/subclavian arteries and a branch PA. The BTT shunt provides temporary additional pulmonary blood flow until the complete repair is undertaken. Classically, a BTT shunt involved sacrificing one of the subclavian arteries and creating an end to side anastomosis to the respective PA. More commonly in the modern era, a prosthetic polytetrafluroethylene (PTFE) tube is interposed between these structures to accomplish the goal of increasing pulmonary blood flow. In either case, the operation may result in distortion of the recipient PA.

Echocardiographic evaluation of patients with repaired TOF should focus on assessment of RV function, size, and pressure, as well as pulmonary and tricuspid regurgitation (Table 1). Patients with a transannular patch are expected to have free pulmonary insufficiency which should be noted (Fig. 1). The parameters used to identify patients for elective pulmonary valve replacement (PVR) are based on cardiac MRI (CMR) study. CMR is utilized in the characterization of RV and left ventricular (LV) systolic and diastolic indexed volumes and function, pulmonary regurgitation percentage, as well as degree of tricuspid regurgitation. Per the recent American College of Cardiology (ACC)/American Heart Association (AHA) guidelines for patients with adult congenital heart disease (ACHD), it is appropriate to consider elective

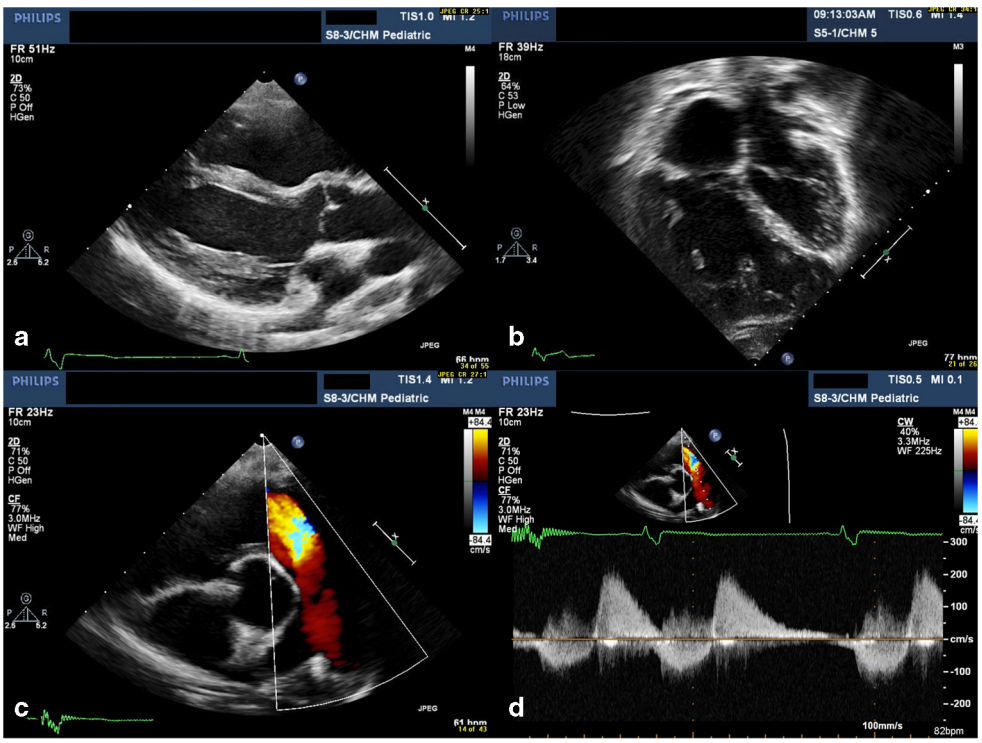

Fig. 1 A 23-year-old patient post repaired Tetralogy of Fallot. (**a**) Parasternal long axis view demonstrating dilated RV, overriding aortic valve with a VSD patch. (**b**) Apical 4 chamber view showing dilated, apex-forming RV. (**c**) Parasternal short axis view showing free PI. (**d**) Continuous wave Doppler of the RVOT showing free PI and no pulmonary stenosis

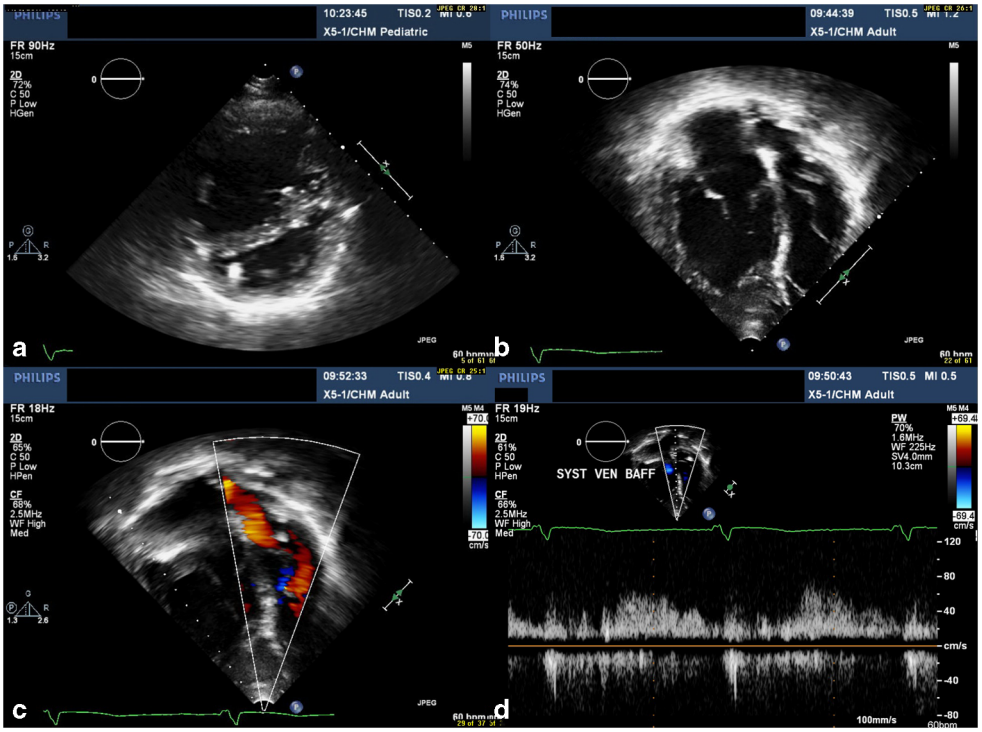

Fig. 2 A 30-year-old male with D-TGA post atrial switch operation. (**a**) Parasternal short axis view showing dilated, hypertrophied systemic RV resulting in bowing of the intraventricular septum into the LV. (**b**) Apical 4 chamber view showing the systemic venous baffle (SVB) in the left atrium and a dilated, hypertrophied RV. (**c**) Apical 4 chamber view showing flow across the SVB. (**d**) Continuous wave Doppler across the SVB showing low velocity, phasic flow suggesting no stenosis

PVR when at least two of the following criteria are met: "(i) ≥ mild RV or LV ventricular dysfunction, (ii) severe RV dilation (RV end-diastolic volume ≥ 160 mL/m^2 or RV end-systolic volume index ≥ 80 mL/m^2, RV end-diastolic volume ≥ 2 times the LV end-diastolic volume), and (iii) RV systolic pressure ≥ 2/3rd systemic pressure, or progressive objective reduction in exercise capacity" [4••].

Additionally, the aortic root should be assessed on echocardiography, as adult patients with TOF can develop aortic root dilation [5]. Guidelines recommend surveillance echocardiograms to be done at least every 2 years, with more frequent echocardiograms indicated based on severity of condition and patient symptoms [4••].

Table 1 Echo check list: post-op TOF

✓Degree of RV dilation
✓RV function
✓Residual RV outflow tract (RVOT) obstruction
✓Tricuspid regurgitation: estimate right ventricular systolic pressure (RVSP)
✓Pulmonary insufficiency
✓Doppler across RVOT
✓Evaluate branch pulmonary arteries for size/narrowing
✓Look for residual VSD
✓Look for aortic insufficiency

D-Transposition of the Great Arteries

D-Transposition of the great arteries (D-TGA) is a common cyanotic CHD. It represents 5–7% of all CHD with an incidence of 0.3 per 1000 live births [1, 6]. The pathology consists of atrioventricular (AV) concordance and ventriculoarterial (VA) discordance. There is normal atrial situs with normally (D-) looped ventricles. The morphological RV is the systemic ventricle and is connected to the Ao while the morphological LV is the sub-pulmonary ventricle and is connected to the PA. This abnormal connection leads to a "parallel circulation", in which the de-oxygenated blood circulates to the systemic circulation (RA– > RV– > Ao and back to RA) while the oxygenated blood circulates in the pulmonary circulation (LA– > LV– > PA and back to LA). Infant survival in D-TGA requires adequate mixing between the systemic and pulmonary circulations through an atrial septal defect (ASD), VSD, or a patent ductus arteriosus (PDA). In babies born without adequate mixing between the parallel circulations, a balloon atrial septostomy is performed to allow necessary mixing of de-oxygenated with oxygenated blood for survival while awaiting definitive surgery.

In the previous era and up until the mid to late 1980s, surgical palliation of D-TGA was accomplished via Mustard or Senning procedure, also known as an "atrial switch" operation (Supplemental Fig. 3). This included creation of an atrial baffle, which directs the systemic venous blood from the inferior vena cava (IVC) and superior vena cava (SVC) across the atrium and into the sub-pulmonary LV through the mitral

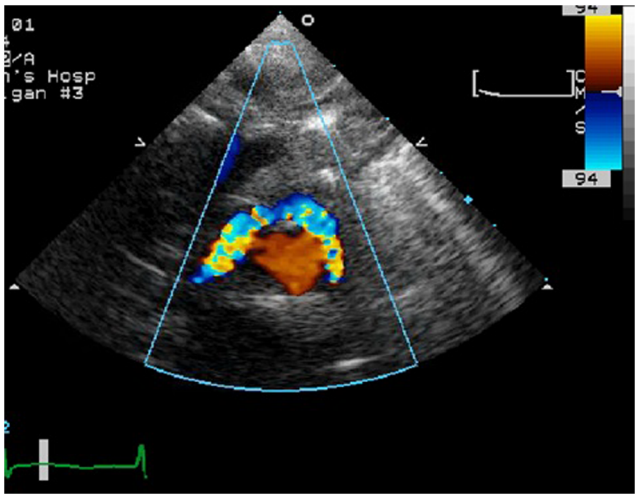

Fig. 3 A 21-year-old male with D-TGA status post arterial switch surgery. High parasternal view showing typical pulmonary artery branches straddling anterior/superior to the aorta due to the LeCompte maneuver (branch pulmonary arteries pulled anteriorly) with flow acceleration

valve. This then results in a pulmonary venous baffle, connecting the pulmonary veins and LA to the systemic RV through the tricuspid valve [7]. Although this form of palliation achieved physiological correction of the circulation (from parallel to series), the anatomy remained uncorrected. The RV is retained as the systemic ventricle with some long-term adverse hemodynamic consequences which included tachy-and-brady arrhythmias given the manipulation of the sinus node region and multiple suture lines in the atrial tissue, failure of the systemic RV often with systemic atrioventricular (tricuspid) valve regurgitation, and baffle obstructions or leaks [4••]. Atrial switch patients frequently require a pacemaker.

In the current era, but now for over 30 years, the atrial switch operation has been replaced by the "Jatene" procedure, or arterial switch operation. This repair is usually done in the neonatal period [8]. It consists of resecting the trunks of the main PA and Ao above the level of the semilunar valves and re-attaching them to the anatomically correct trunks (Supplemental Fig. 4). The coronary arteries are removed from the original aortic trunk and re-implanted into the native pulmonary valve/sinuses. Thus, with this operation, both anatomical and physiological correction is achieved. The patient's native pulmonary valve becomes the "neo-aortic valve" while the native aortic valve becomes the "neo-pulmonary-valve" [8].

Echocardiographic evaluation of the patient who has undergone the atrial switch should include visualization of the systemic and pulmonary venous baffles for obstruction or leaks [6] (Table 2). The baffles are best seen in the apical views, but this may be difficult in some patients (Fig. 2). If the baffles cannot be visualized on echocardiography, the patient may require additional imaging such as cardiac MRI. If there is baffle obstruction or leak, interventional cardiac catheterization is required to dilate/stent the baffles (usually SVC-systemic venous baffle in case of Mustard procedure) or device closure of the baffle leak.

Additionally, the systemic RV should be assessed, looking particularly for function, hypertrophy, and dilation [6]. CMR

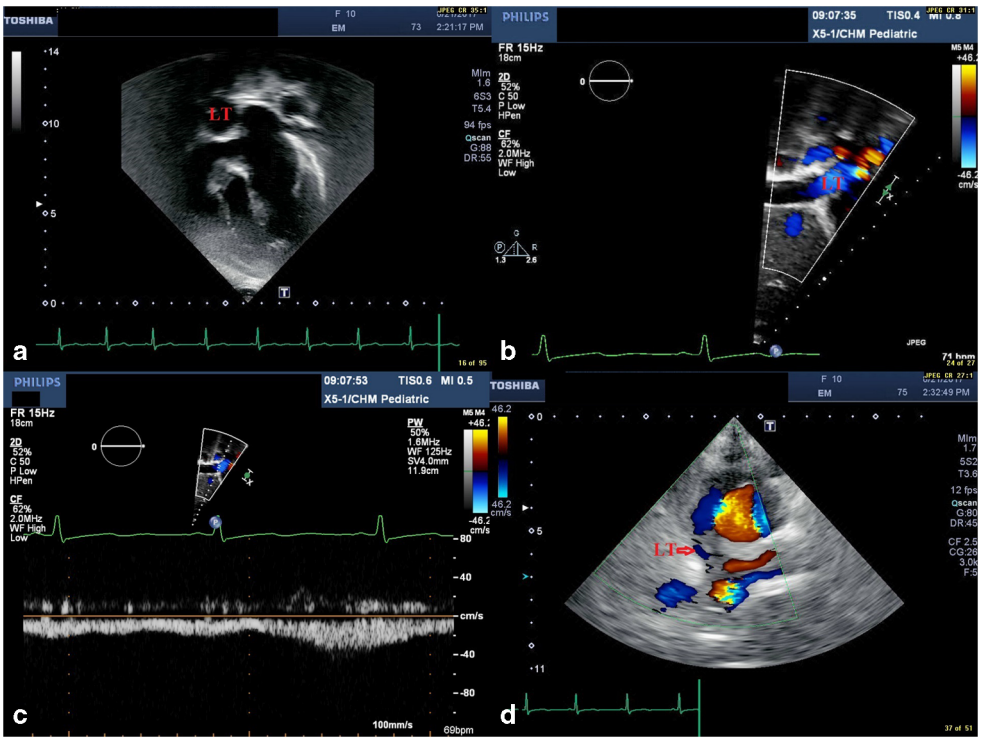

Fig. 4 A 33-year-old female with tricuspid atresia post Fontan surgery. (**a**) Apical 4 chamber view showing a single AV valve with single, systemic ventricle, and the lateral tunnel. (**b**) Subcostal bicaval view demonstrating color flow in the IVC end of the Fontan circuit. (**c**) Doppler assessment of flow in the Fontan circuit: phasic and low velocity. (**d**) Suprasternal short axis view showing a patent SVC connection to the RPA. LT = lateral tunnel

Table 2 Echo check list: post-op D-TGA/atrial switch operation (Mustard/Senning)

- ✓Degree of RV dilation
- ✓RV function
- ✓Degree of Tricuspid regurgitation
- ✓Evaluate systemic and pulmonary venous baffles for stenosis or leak

can assist in delineating RV function [6]. The severity of tricuspid valve regurgitation (TR) often correlates with the degree of RV dysfunction as the tricuspid valve is now the systemic AV valve [6].

The recent ACC/AHA guidelines for patients with ACHD recommend that patients who have undergone the atrial switch operation receive annual cardiac imaging, including echocardiogram and CMR when indicated [4••].

Patients who underwent the arterial switch operation may do extremely well clinically and will frequently have near-normal hemodynamics. The most common complication following this operation is distortion or narrowing of the proximal pulmonary arteries. This area can be very difficult to visualize echocardiographically in the adult, but a high parasternal short axis view usually serves best (Fig. 3). There may also be obstruction of RV or LV outflow near the anastomotic sites where the PA and Ao were re-attached, regurgitation or stenosis of the neo-aortic valve, and coronary artery stenosis or obstruction [4••, 6, 9•]. Subsequently, imaging should be dedicated to visualizing these structures (Table 3). Recent guidelines recommend obtaining routine echocardiogram at a minimum of every 1–2 years [4••].

Single Ventricle/Fontan

Congenital heart defects with single ventricle physiology are individually uncommon. However, the surgical approach tends to create a unified group of patients with many similar characteristics and problems. Indeed, repair of single ventricle generally constitutes between 10 and 15% of the surgical volume in most CHD centers. This group of patients, once felt to be non-survivable, now constitutes a growing and extremely complex group of adult patients. The anatomic variations which are commonly seen include tricuspid atresia (and other causes of hypoplastic RV), hypoplastic left heart syndrome (HLHS), and unbalanced AV canal defects (with hypoplastic RV or LV). In this section, we will focus on tricuspid atresia (TA) and HLHS. Their estimated incidence is 0.10 and 0.27 per 1000 live births, respectively [10]. In TA, the tricuspid valve is not formed and deoxygenated blood shunts from RA to LA across a patent foramen ovale/ASD. A VSD may be present, allowing LV blood to flow via the RV to the branch PA to be oxygenated. In the absence of a VSD, a PDA is required to provide pulmonary blood flow. A minority of TA may also have transposition of the great arteries which then may result in unrestricted pulmonary flow but compromised flow to the systemic circulation.

For all patients born with single ventricle physiology, a staged palliation (two or usually three stages) is performed in the current era. The ultimate goal of single ventricle palliation is to allow the single ventricle to pump to the systemic circulation and create a passive flow circuit to the lungs. This separates the circulations into two and takes the volume load away from the single ventricle. The first stage, regardless of the underlying anatomy, requires the creation of a stable systemic blood flow from the single ventricle to the aorta and a controlled, low pressure flow to the lungs, usually via a BTT shunt or some variation. This stage is usually carried out in the newborn period. The second stage is usually performed at 6–8 months of age, where the superior caval blood is directly connected to the pulmonary arteries and the first stage shunt is taken down, still allowing inferior caval blood to pass through the single ventricle circuit. Third stage, or completion, of Fontan palliation usually takes place between 18 months and 3 years of age and involves directing the IVC blood into the pulmonary circuit.

The original, "classic" Fontan palliation was performed in patients with TA [11] as a single stage operation. The superior vena cava (SVC) was anastomosed to the distal right PA followed by connecting the right atrial appendage to the proximal end of the right PA [11]. Since Fontan's first description of this surgery in 1971, there have been several modifications in order to optimize the passive pulmonary blood flow from the systemic venous system. The final stage can have a

Table 3 Echo check list: post-op D-TGA/arterial switch operation (Jatene)

- ✓LV size and function
- ✓Supravalvular aortic or pulmonary stenosis
- ✓Neo-aortic insufficiency
- ✓Estimate RVSP if tricuspid regurgitation present
- ✓Evaluate branch pulmonary arteries (PA straddles Ao: LeCompte maneuver)
- ✓Evaluate coronary arteries (if possible)

Table 4 Echo check list: post-op single ventricle (Fontan)

- ✓Ventricular size and function
- ✓Degree of atrioventricular valve regurgitation
- ✓Evaluate ventricular outflow tract
- ✓Semilunar valve for stenosis or regurgitation
- ✓Evaluate Fontan pathway for patency/ thrombus/ fenestration

Table 5 Post-op echo check list: truncus arteriosus

LV size and function
Truncal root size
Truncal/aortic valve stenosis or regurgitation
RV to PA conduit for stenosis or regurgitation
RV size and function
Estimate RVSP
Look for residual VSD

number of variations when the IVC is connected to the pulmonary circulation via an intra-cardiac lateral baffle/tunnel or via an extra-cardiac prosthetic conduit [12] (Supplemental Fig. 5). The baffle/conduit may also be created with a small fenestration to the atrium resulting in a right to left shunt. The purpose of this fenestration is to decompress the Fontan circuit if the pressures are elevated, accepting some degree of desaturation from the resulting right-to-left shunt.

Patients with HLHS will undergo a first stage neonatal palliation, known as the Norwood operation. The Norwood operation re-purposes the functional RV to provide the systemic circulation. A neo-aorta is created by first transecting the main PA, followed by creating an anastomosis between the main PA and the aorta to reconstruct the aortic arch [13]. Pulmonary blood flow is provided via a systemic-pulmonary shunt. This can be a modified BTT shunt (as described earlier), or a Sano shunt, which is a PTFE tube connecting the RV to the PA [14]. One potentially significant difference among single ventricle patients, as illustrated in the description of these two anatomic variations (HLHS and TA) is the morphology of the single systemic ventricle—a right ventricle in the case of HLHS and a left ventricle in TA.

Echocardiography should focus on evaluation of single ventricular function, as it is the only pumping chamber for the heart and degree of systemic AV valve regurgitation, which can lead to dilation of the single ventricle and eventual dysfunction [15]. The systemic semilunar valve should be evaluated carefully for obstruction/regurgitation. Imaging of the Fontan pathway, including the connection between the IVC and SVC and the pulmonary circulation should be attempted, but may be difficult in adult patients (Table 4). Flow in the Fontan pathway may be best visualized in the subcostal and/or suprasternal views (Fig. 4). Fontan patients are at risk of developing venous thromboembolism due to the slow flow state; hence, the presence of any clots or leaks within the Fontan circuit should be carefully evaluated. Additionally, attempts should be made to evaluate the branch PA's, particularly in and around the anastomotic site which may require supplemental cross-sectional imaging. The ACC/AHA guidelines for the

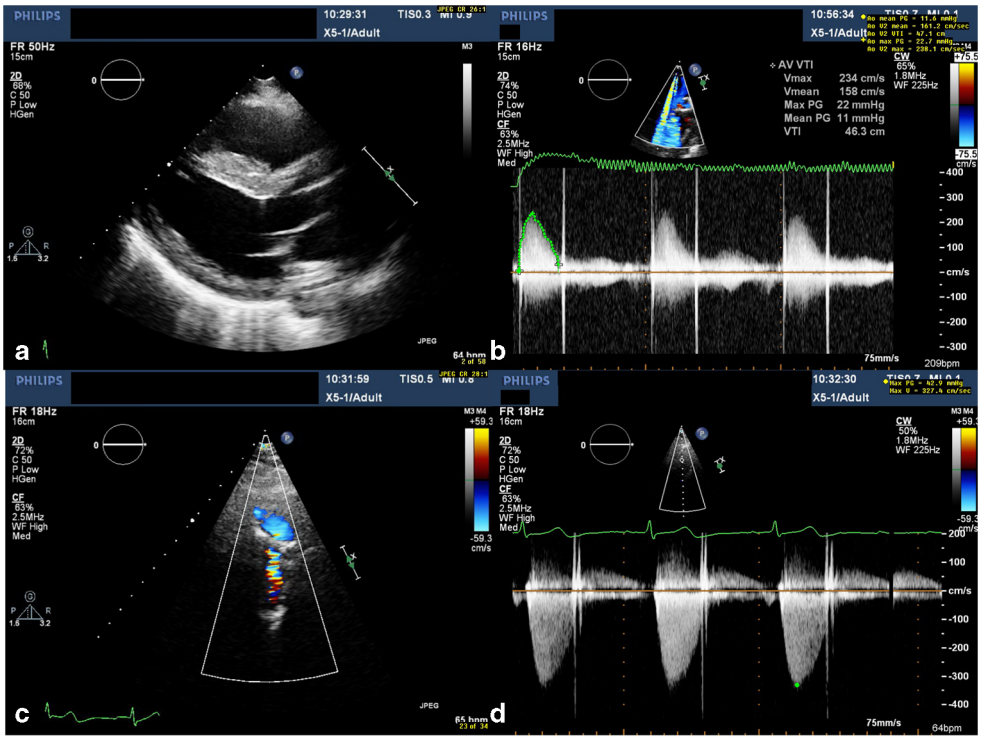

Fig. 5 A 25-year-old female post repaired truncus arteriosus now with a prosthetic aortic valve. (**a**) Parasternal long axis view showing prosthetic aortic valve. (**b**) Continuous wave Doppler across the aortic valve. (**c**) High parasternal view demonstrating color flow across the RV to PA conduit. (**d**) Doppler evaluation of RV-PA conduit showing mild stenosis

care of ACHD patients recommends annual echocardiography for patients with Fontan physiology [4••].

Truncus Arteriosus

Truncus Arteriosus is a less common form CHD with an estimated incidence of approximately 0.10 per 1000 live births [10]. This defect is associated with DiGeorge's syndrome in up to 40% of cases [15]. It also represents an example of patients who require repair with the use of an extra-cardiac RV to pulmonary artery conduit.

In patients with truncus arteriosus, the right and left ventricle pump into a common arterial trunk with a single semilunar valve (truncal valve) which then gives rise to the aorta and pulmonary arteries. This valve is often malformed. A mal-alignment ventricular septal defect, similar to that seen in TOF, is present. The common trunk provides flow to both the systemic and pulmonary circulations.

Surgical repair is typically performed in the neonatal period and leads to improved outcomes [16]. To repair this defect, the pulmonary arteries are separated from the common arterial trunk, allowing the trunk to become the neo-aorta which exclusively provides the systemic blood flow. A conduit is used to connect the RV to the pulmonary arteries which were detached from the common trunk. This operation follows the surgical concept proposed by Rastelli and carries his name [17]. The VSD is closed with a patch to the neo-aorta. If there is significant stenosis or insufficiency of the common truncal valve, it may require replacement during initial surgery or at a later time [17]. The RV to pulmonary conduit represents the clinical focal point in this repair. Conduits may fail because of fibrous deposition, lack of growth in a growing patient, or valvular failure. Patients who have undergone a successful repair of truncus arteriosus will require multiple conduit revisions in their lifetime.

Echocardiographic imaging should focus on establishing the patency and status of the RV-PA conduit and valve within, evaluating the branch pulmonary arteries and truncal root, truncal/neo-aortic valve for any stenosis or regurgitation and ventricular function (Table 5). The truncal valve may be visualized in the parasternal long axis views and RV-PA conduit may require some modification of the standard view to show RV outflow tract (Fig. 5). The ACC/AHA guidelines recommend consideration of annual echocardiograms for all patients who have a RV-PA conduit in place [4••]. The durable life expectancy of the conduit in an adult may be 15–20 years. Newer, catheter-based devices, stents, and catheter-deployed valves may hopefully extend that duration. [4••].

Conclusion

As the number of adults with repaired CHD is on a rise, it becomes imperative for cardiologists caring for adults to be familiar with the CHD, its surgical repair and echocardiographic features that will assist appropriate recognition and referral to an adult congenital program. Echocardiography is an easily available, non-invasive imaging tool in the cardiologists' office and further supplementation with other imaging modalities may be necessary in this group of patients. Experience in imaging congenital heart disease is essential for the management of these patients.

Compliance with Ethical Standards

Conflict of Interest Amrit Misra, Chenni Sriram, Pooja Gupta, and Richard Humes 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.

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