Evaluation of the Single Ventricle

10

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Abstract

The terms "single ventricle" and "functional single ventricle" apply to a broad group of congenital cardiac lesions that, despite their marked anatomic heterogeneity, share a common, unifying characteristic: a biventricular intracardiac repair cannot be performed to separate the systemic and pulmonary circulations. In essence, a single functional ventricular pumping chamber supplies both the systemic and pulmonary vascular beds; oxygenated and deoxygenated blood blend together, and all patients exhibit some degree of systemic arterial desaturation. In the management of these patients, the ultimate goal is to separate the systemic and pulmonary circulations such that deoxygenated systemic venous blood returns passively to the pulmonary arteries without the assistance of a separate pumping chamber. The resultant physiology, known as Fontan physiology, forms the basis for the current management of the single ventricle patient. Transesophageal echocardiography (TEE) can play an important role in the diagnostic evaluation of single ventricle patients. The most common application of TEE is in the intraoperative setting—preoperatively, to verify anatomy and physiology, and postoperatively, to evaluate the results of cardiac surgery. However, there are other non-operative settings in which TEE can be utilized to evaluate the detailed anatomy of the single ventricle patient, and to examine some of the possible problems and complications in operated patients. This chapter explores the use of TEE in the single ventricle patient.

Keywords

Single ventricle • Univentricular heart • Congenital heart disease • Echocardiography • Transesophageal echocardiography • Fontan operation • Pediatric cardiac surgery • Palliative surgery • Hypoplastic left heart syndrome • Tricuspid atresia • Heterotaxy

Introduction

The term "single ventricle", as used by specialists in congenital heart disease (CHD), is recognizably broad and nonspecific, encompassing a wide spectrum of cardiac lesions with differing pathologic anatomy and embryologic derivations. In its strictest sense, the term single ventricle refers to the class of lesions known as univentricular heart, in which one ventricular chamber is rudimentary or absent, and therefore unable to function as an independent, separate chamber [1, 2]. However from a surgical (and more utilitarian) sense, the use of the term "single ventricle" has been expanded to include all manner of cardiac lesions in which a biventricular

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intracardiac repair cannot be performed or may not be considered the optimal approach to separate the systemic and pulmonary circulations, and therefore only one functional ventricular pumping chamber supplies the systemic circulation [3, 4]. Systemic and pulmonary venous blood must, of necessity, commingle, producing mixing physiology and obligate systemic arterial hypoxemia. Because of the wide anatomic diversity of cardiac defects that can produce this physiology, the more generic term "functional single ventricle" is also used to describe such patients.

The most common application of transesophageal echocardiography (TEE) for single ventricle evaluation is in the intraoperative setting—preoperatively, to verify anatomy and physiology, and postoperatively, to evaluate the results of cardiac surgery. However, there are other non-operative settings in which TEE can be utilized to evaluate the detailed anatomy of the single ventricle patient. For example, in the adolescent or adult patient with complex single ventricle anatomy, TEE can serve as a supplemental (or even initial) tool for diagnostic evaluation. Postoperative Fontan patients are notorious for having marginal to poor transthoracic echocardiographic windows, and in this setting TEE can be used for improved evaluation.

This chapter discusses the TEE evaluation of the single ventricle patient. The focus will be upon those patients who, from an anatomic standpoint, have only one truly functional ventricle. As will be evident, there is great diversity among this group of patients, with each class of defect having its own distinctive anatomic and physiologic characteristics. Knowledge of these distinguishing features greatly facilitates their TEE assessment.

Author's note: As previously mentioned in Chap. 4, some of the standardized TEE terminology defined in that chapter might seem confusing when applied to the abnormal and unusual anatomy found in many single ventricle defects—for example, using the mid esophageal *four* chamber view for a double inlet ventricle with only *three* principal chambers. Nonetheless, for consistency's sake we will continue to utilize the standardized views and nomenclature defined in Chap. 4, and will denote modifications to those views when applicable. It is recommended that the reader have a reasonable degree of familiarity with the TEE views, probe positions and probe manipulations described in that chapter.

General Considerations for TEE Evaluation

As mentioned above, the term "single ventricle" comprises a heterogeneous group of lesions, and the TEE evaluation must be tailored to the anatomic considerations specifically associated with each cardiac lesion. Moreover, as is true with all CHD, considerable anatomic variation exists within any single group of cardiac defects, and the examiner must anticipate and prepare for this. The TEE study will rarely be the first diagnostic examination performed on a single ventricle patient; prior imaging and diagnostic information will usually be available, and the examiner should be thoroughly familiar with this information. Nonetheless the examiner should approach the TEE examination with a fresh perspective, because the TEE study will occasionally reveal new findings and/or modify previous ones. Moreover, in some instances the study provides important additional information that is considered vital in the formulation of a surgical plan or other therapeutic strategy. Whenever possible, a complete and systematic examination should be performed (see Chap. 4); the more complex the anatomy, the more meticulous and complete the TEE examination needs to be. In the sections that follow, the different types of single ventricle defects will be discussed, along with their most common anatomic features. Certain anatomic variations and abnormalities (such as a left superior vena cava to coronary sinus, pulmonary venous anomalies, patent ductus arteriosus) are common to a number of different pathologies, and these will not be repeated with each type of cardiac defect. However they again underline the importance of performing as complete a study as possible.

Single Ventricle: Anatomic Types

Hypoplastic Left Heart Syndrome

Hypoplastic left heart syndrome (HLHS) comprises a spectrum of lesions characterized by varying degrees of left sided hypoplasia [5, 6]. The classic constellation of associated cardiac defects includes mitral stenosis/mitral atresia, aortic valve atresia, a diminutive left ventricle (LV), a markedly hypoplastic ascending aorta, and hypoplastic aortic arch and aortic isthmus with a posterior aortic shelf (coarctation). However, the spectrum of HLHS can vary from mitral and aortic valve atresia, a tiny ascending aorta, and virtually no LV cavity, to mild mitral and aortic valve hypoplasia, antegrade flow across the aortic valve, and mild ascending aortic and LV hypoplasia. Nonetheless despite the anatomic variability, all cases of HLHS share a defining characteristic: an inability of the left sided structures to independently support the systemic circulation. Thus the right ventricle (RV) assumes the primary role as the systemic pumping chamber, providing blood to the systemic circulation via forward flow through the main pulmonary artery and patent ductus arteriosus (in the unoperated patient) or, if surgically palliated, via a Norwood-type procedure (see below).

The TEE evaluation of HLHS patients is relatively straightforward, focusing on the key features relevant for the patient's physiology and anatomy. Most of these features can be evaluated from the level of the mid esophagus; the most

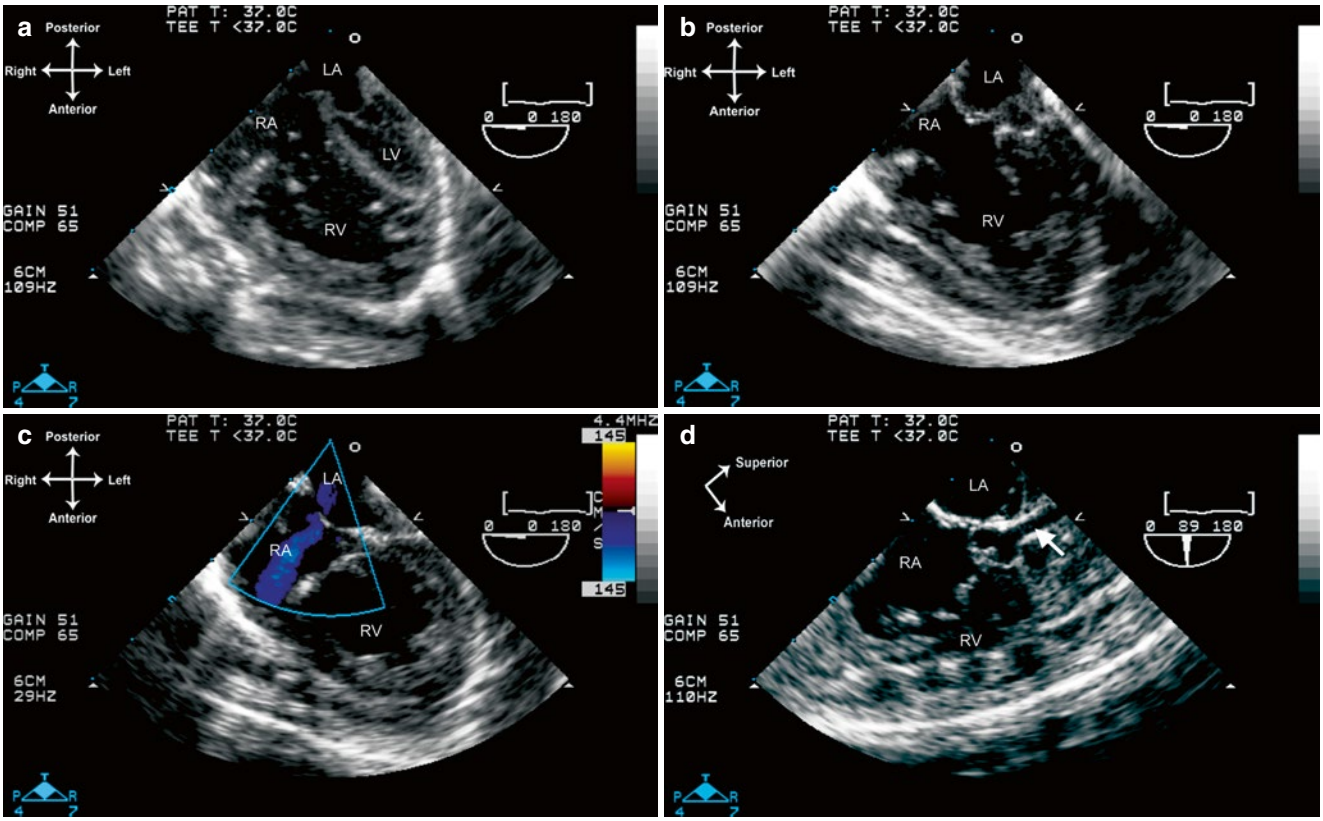

Fig. 10.1 Hypoplastic left heart syndrome, as viewed from the mid esophageal four chamber view. There can be significant anatomic variation ranging from mitral stenosis and left ventricle hypoplasia (**a**) to virtual absence of these structures (**b**). The atrial septum can be adequately visualized and interrogated by color Doppler also in this plane (**c**). The mid esophageal long axis view (**d**) allows for visualization of the aortic valve and diminutive proximal ascending aorta (*arrow*). *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

useful initial view is the mid esophageal four chamber (ME 4 Ch) view, multiplane angle 0°. From here, the multiplane angle can be varied as needed for optimal visualization of the different cardiac structures. Specific TEE evaluation should include systemic and pulmonary venous return, atrial septal patency, tricuspid valve anatomy and regurgitation (if present), RV morphology, function, and wall motion abnormalities (if present), and pulmonary valve function (Fig. 10.1, Video 10.1). If mitral and/or aortic valve are patent, flow across these valves can also be evaluated. Coronary artery origins can be viewed from a modified mid esophageal aortic valve short axis (ME AV SAX) view, with probe anteflexion and multiplane angles between 0 and 45°. Other intracardiac anomalies associated with HLHS, such as anomalous pulmonary venous return and left superior vena cava (SVC) to a dilated coronary sinus can also be seen by TEE using the mid esophageal views. The hypoplastic ascending aorta can best be visualized in the mid esophageal aortic valve long axis (ME AV LAX) view with a multiplane angle approximately 80–90° (Fig. 10.1c); an accurate ascending aortic diameter can also be measured. Ductal patency can be evaluated by imaging and color flow Doppler in the upper esophageal aortic arch short axis (UE Ao Arch SAX) and upper esophageal pulmonary artery long axis (UE PA LAX) views; there will usually be low velocity right to left shunting in systole, and left to right shunting in diastole. However the hypoplastic transverse aortic arch and proximal descending thoracic aorta are generally difficult to visualize fully by TEE; evaluation of these structures is better accomplished by other modalities such as transthoracic echocardiography. When HLHS is associated with a very small or absent atrial communication, a small decompressing vein can sometimes be present, connecting the left atrium/pulmonary veins to the innominate vein. Known as a *levoatrial cardinal vein*, it serves as an alternate pathway to re-route pulmonary venous blood back to the right heart [7]. If present, it can sometimes be visualized with an UE Ao Arch SAX and UE PA LAX views, using leftward rotation to demonstrate the vein as it returns to the innominate vein.

Hypoplastic Right Heart Syndrome

Hypoplastic right heart syndrome encompasses a spectrum of cardiac lesions characterized by pulmonary atresia (either a formed but imperforate pulmonary valve, also known as

membranous atresia, or long-segment muscular atresia), varying degrees of RV hypoplasia, and an intact ventricular septum. This class of cardiac defects is often referred to as pulmonary atresia/intact ventricular septum. The tricuspid valve is patent but hypoplastic, with the degree of tricuspid hypoplasia generally correlating with the extent of RV hypoplasia. In moderate hypoplasia, only two major portions of the RV (inflow and outflow) are present; the apical (trabecular) portion is either truncated or absent. This is termed a "bipartite" RV. In the most severe cases, only a tiny RV (usually the inflow portion) is present and this is termed a "unipartite" RV. Such patients have an obligate right to left shunt across the atrial septum, and a good-sized LV and aortic valve. Given the absence of antegrade flow from RV to pulmonary artery, patients require an alternate source of pulmonary blood flow, usually from a patent ductus arteriosus (PDA). As RV hypoplasia becomes more severe, there is an increasing likelihood of persistent embryonic sinusoidal connections (also known as ventriculo-coronary arterial communications) between the RV and the epicardial coronary arteries [8]. Some of these sinusoidal connections can contain stenoses that result in portions of the myocardium being perfused primarily or exclusively by the hypertensive RV; this situation is termed RV dependent coronary artery perfusion. In such cases, a two ventricle repair cannot even be considered, as the RV cannot be decompressed [9].

In the absence of RV-dependent coronary circulation, a determination must be made whether a hypoplastic right heart syndrome patient is a candidate for a two ventricle repair, or whether the patient should undergo single ventricle palliation. This determination is largely based upon an assessment of the degree of RV hypoplasia, which in turn is based primarily upon an evaluation of tricuspid valve annulus and RV size. The minimum tricuspid and RV dimensions that dictate the choice of one vs. two ventricle strategy vary among different institutions, and the published criteria continue to be evolve with time and increasing experience. Some centers use a tricuspid valve annulus z score anywhere between <−3 to −10 as a cutoff [10–13], some use the RV morphology (unipartite vs. bi/tripartite) [14], some use an estimation of RV diastolic volume [15], and some use a combination of these criteria. Whatever the method, if the tricuspid valve and RV are deemed too hypoplastic for a two ventricle circulation, then initial surgery is undertaken with the intention of single ventricle palliation—specifically, planned future bidirectional Glenn and Fontan procedures (discussed below). In this case, a systemic-pulmonary artery shunt (Blalock-Taussig shunt) is initially performed, often in conjunction with an atrial septectomy to assure unobstructed mixing of right and left atrial blood. In patients with milder forms of tricuspid valve and RV hypoplasia, a two ventricle (vs. "one and a half") approach can be considered. In these instances antegrade flow across the RV outflow tract is established in the newborn period, with the expectation that the increased blood flow through the RV will stimulate growth of this chamber. This can be accomplished surgically by reconstruction of the RV outflow tract, usually performed in combination with a systemic-pulmonary shunt (such as a Blalock-Taussig shunt) [16]. An alternative strategy utilizes interventional catheterization techniques to establish antegrade flow across the RV outflow tract, including catheter perforation of the pulmonary valve (using balloon, laser, or radiofrequency methods), along with transcatheter stenting of the ductus arteriosus [14, 17]. With either surgical or transcatheter approach, some patients experience enough growth of their RV that they can function effectively with two ventricle physiology [14, 18]. Eventually, these patients can undergo closure of the systemic-pulmonary shunt and, if an atrial defect is still present, it can also be closed. In other patients, the RV continues to be moderately hypoplastic and a "one and a half repair" is subsequently undertaken. In this type of operation, a bidirectional Glenn procedure (discussed below) is performed to allow SVC blood to drain directly to the pulmonary arteries, while leaving inferior vena cava (IVC) blood to return to the RV and then be ejected directly into the main and branch pulmonary arteries. At the time of the Glenn operation, the atrial septal defect, if present, is often closed [19].

The TEE evaluation of the patient with hypoplastic right heart syndrome focuses on the adequacy of an interatrial communication, as well as the degree of tricuspid valve and RV hypoplasia. The atrial septum can be assessed from the ME 4 Ch and mid esophageal bicaval (ME Bicaval) views. The tricuspid valve and hypoplastic RV are best seen using a combination of MV 4 Ch (Fig. 10.2a, Video 10.2) and mid esophageal right ventricle inflow-outflow (ME RV In-Out) views (Fig. 10.2b, c, Video 10.2). Tricuspid and pulmonary valve annular diameters can also be measured using these views; as mentioned above, the tricuspid valve annulus diameter plays a very important role in determining whether intervention should be undertaken for a one or two ventricle strategy [15, 19, 20]. Coronary artery sinusoidal flow can sometimes be seen with color flow Doppler, but coronary artery stenoses cannot be diagnosed by echocardiography and therefore must be evaluated by angiography. Left ventricular and mitral valve function should be evaluated carefully, also from the midesophageal views, specifically the ME 4 Ch, mid esophageal two chamber (ME 2 Ch), mitral commissural (ME Mitral), and long axis (ME LAX) views. Additional information regarding LV and mitral valve function can be provided by the transgastric views: the transgastric basal and mid short axis (TG Basal SAX, TG Mid SAX) and transgastric two-chamber (TG 2 Ch) and transgastric long axis (TG LAX) views. Some patients can have significant LV outflow tract obstruction due to bowing of the ventricular septum that produces mid-cavitary obstruction, and this should also be evaluated carefully with a combination of mid esophageal (ME 4 Ch, ME LAX), transgastric (TG LAX), and deep transgastric long axis (DTG LAX) and deep

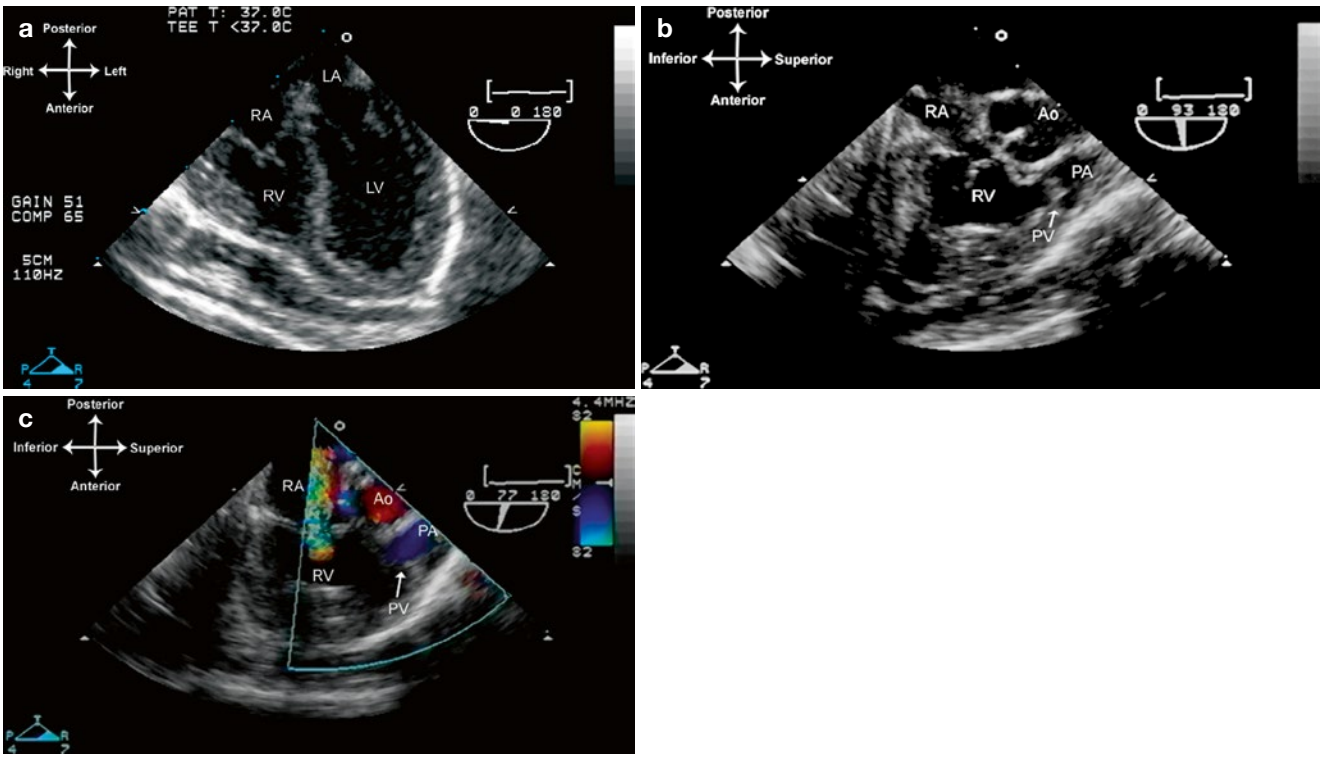

Fig. 10.2 Pulmonary atresia/intact ventricular septum, also known as hypoplastic right heart. The right ventricular cavity is small, muscular, and non-apex forming as displayed in the mid esophageal four chamber view (**a**). The trabecular portion of the right ventricle is typically underdeveloped, as shown in the mid esophageal right ventricular inflow-outflow views (**b**, **c**). Note the presence of an imperforate pulmonary valve without antegrade flow. There is significant tricuspid insufficiency. *Ao* ascending aorta, *LA* left atrium, *LV* left ventricle, *PA* main pulmonary artery, *PV* pulmonary valve, *RA* right atrium, *RV* right ventricle

transgastric sagittal (DTG Sagittal) views. The pulmonary valve, if present, can be evaluated and its annulus measured in the ME RV In-Out view, using a multiplane angle of 60–90° (Fig. 10.2b). The branch pulmonary arteries and ductus arteriosus are not always well seen, but ductal left to right flow can often be detected by color flow Doppler in the UE PA LAX and UE Ao Arch SAX views, using leftward probe rotation (when there is a left aortic arch).

Tricuspid Atresia

Patients with tricuspid atresia have no functional tricuspid valve. Often, there is complete absence of the tricuspid valve, with only a muscular shelf seen in the tricuspid position. In some instances the tricuspid valve is formed and present but imperforate (so-called "membranous" tricuspid atresia). In either case, there is obligate right to left shunting across the atrial septum, and complete mixing of deoxygenated and oxygenated blood at atrial level. There is generally a small RV cavity, with marked hypoplasia of the inflow portion but variable trabecular and infundibular portions, and usually (but not always) a communication between the LV and the hypoplastic RV. This communication has variously been termed a ventricular septal defect (VSD), or alternatively a *bulboventricular foramen* (BVF) [21, 22]—it generally is located between the LV and RV inflow portion, but can also be seen closer to the apex or infundibulum of the small RV. In the majority of cases, this defect will be the site of obstruction to LV to RV shunting, though obstruction can occur at the outlet valve itself (the valve arising from the RV) or in the subvalvar infundibulum [21]. The ventriculo-arterial connection can vary, and one of the most recognized classification systems for tricuspid atresia is based upon the relationship of the great arteries [23]. A summary of this classification system is given in Table 10.1. In general, this system's nomenclature is not widely used when clinicians describe and discuss tricuspid atresia, but it still serves to underline the most important anatomic features relevant to this anomaly, namely (a) great artery origin/relationship, (b) VSD/BVF size, and (c) whether aortic or pulmonary outflow obstruction is present. In Type I, seen in 70–80 % of patients with tricuspid atresia [24], the aorta arises in normal position from the LV, there is ventriculo-arterial concordance, and the aortic valve is of normal size and function. The pulmonary artery arises from the small RV chamber, and the amount of pulmonary blood flow depends upon the size of the VSD/BVF and the degree of subpulmonary stenosis (if present). Type I can be further classified into three subtypes, based upon the size of the VSD and degree of pulmonary outflow obstruction: Type Ia

(no VSD, pulmonary atresia), Type Ib (small VSD, pulmonary stenosis); Type Ic (large VSD, no pulmonary stenosis). If the ventricular defect is large and there is no subpulmonary stenosis, the patient can have excessive pulmonary blood flow leading to pulmonary overcirculation and congestion. If there is obstruction at the ventricular defect or subpulmonary level, there can be reduced antegrade flow leading to systemic arterial hypoxemia (Fig. 10.3, Video 10.3). The patient is then dependent upon an alternative source of pulmonary blood flow such as a patent ductus arteriosus or surgically created shunt. With Type II, seen in 12–25 % of patients with tricuspid atresia [24], there are transposed great arteries (ventriculo-arterial discordance), and the aorta arises from the small RV outflow chamber, generally to the right of the pulmonary artery, or D-transposed. Type II is also classified into three subgroups, based on the degree of pulmonary outflow obstruction: Type IIa (pulmonary atresia), Type IIb (pulmonary stenosis), Type IIc (no pulmonary stenosis). In general, with Type II tricuspid atresia there is either subaortic stenosis or subpulmonary stenosis, depending on the size of the VSD/BVF (through which blood must pass from LV to RV to aorta) and position of the infundibular septum. Type IIc is the most common form: in such cases the pulmonary valve (which arises from the LV) is usually of normal size and function. The ventricular defect is often surrounded by a muscular rim, and if small enough, there can be marked subaortic stenosis, often in association with hypoplasia of the aorta and aortic coarctation or interruption (in many cases requiring a patent ductus arteriosus for systemic perfusion). Such patients invariably require surgery to alleviate or bypass the subaortic stenosis, as well as repair the aortic arch. Conversely, Types IIa and IIb (which are much less common) have pulmonary outflow obstruction, usually due to posterior malalignment of infundibular septum that can produce significant subpulmonary stenosis and decreased pulmonary blood flow [25]. Such patients might require an alternative source of pulmonary blood flow, such as a ductus arteriosus or surgically created shunt. In these instances the VSD/BVF and aorta are generally of good size, and there is no obstruction to systemic blood flow. Types I and II are by far the most common forms of tricuspid atresia.

Table 10.1 Classification of tricuspid atresia

Type I	**Normally related great arteries**
Ia	Intact ventricular septum and pulmonary atresia
Ib	Small ventricular septal defect and pulmonary stenosis
Ic	Large ventricular septal defect, no pulmonary stenosis
Type II	**Transposed great arteries**
IIa	Ventricular septal defect with pulmonary atresia
IIb	Ventricular septal defect with pulmonary stenosis
IIc	Ventricular septal defect, no pulmonary stenosis
Type III	**Transposition or malposition of the great arteries**
	Associated complex lesions, including L-transposition of the great arteries, double outlet right ventricle, double outlet left ventricle, truncus arteriosus

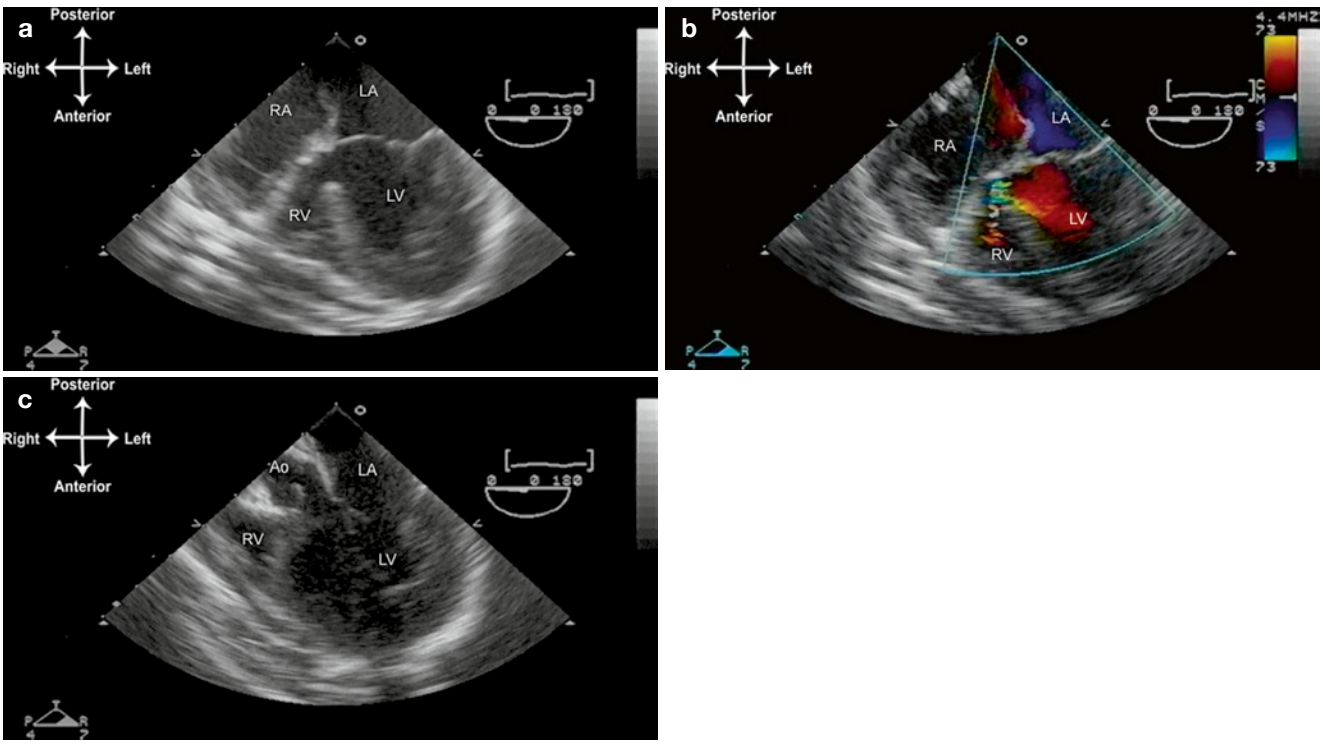

Fig. 10.3 Tricuspid atresia, normally related great arteries. (**a**) The mid esophageal four chamber view demonstrating lack of visible tricuspid valve; only a muscular shelf is seen, as well as a hypoplastic right ventricular chamber. (**b**) There is a small ventricular septal defect or bulboventricular foramen with turbulent left to right shunting. (**c**) Withdrawal and slight anteflexion of the TEE probe demonstrates the aortic valve (*Ao*) arising from the left ventricle. *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle.

Type III is much more rare, seen in about 3–6 % of patients with tricuspid atresia, and it is a less specific category that includes a number of other defects such as L-transposition of the great arteries, truncus arteriosus, etc. [24].

The TEE evaluation of tricuspid atresia focuses upon atrial septal patency and shunting, mitral valve and LV function, the position of the great arteries and ventriculo-arterial relationship, and outflow tract patency including the size of the VSD/BVF. Much of this evaluation can be performed from the level of the mid esophagus, particularly the ME 4 Ch view for evaluation of atrial septum, mitral valve, LV and great arteries as well as VSD/BVF. Supplemental evaluation of these structures can also be provided by other mid esophageal views: ME Bicaval (systemic venous return, atrial septum), ME LAX (mitral valve, semilunar valves, VSD/BVF), ME RV In-Out (VSD/BVF). The deep transgastric views (DTG LAX, DTG Sagittal) can also provide good visualization of the mitral valve and VSD/BVF, as well as the aortic and pulmonary outflow tracts. The DTG views are also useful for spectral Doppler evaluation of the outflow tracts. In patients with transposed great arteries, the size of the ventricular defect, and potential for subaortic stenosis, should be evaluated carefully (Fig. 10.4, Video 10.4). If the VSD/BVF and/or the subaortic area are felt to be potentially obstructive, the pulmonary valve should be evaluated with the anticipation of a possible Damus-Kaye-Stansel (DKS) operation. The evaluation of the VSD/BVF and the DKS operation are discussed below. In tricuspid atresia patients with transposed great arteries, left juxtaposition of the atrial appendages can sometimes be encountered [26, 27]. Care should be taken to recognize this entity and not confuse it with a large atrial septal defect; in fact, the orientation of the true atrial septal defect is often different from what is normally encountered [28]. This entity is best evaluated from a modified ME AV SAX view first retroflexed, then slowly anteflexed to demonstrate the right atrial appendage just anterior to the left atrial appendage. An example of left juxtaposition of the atrial appendages is given in Chap. 12—Conotruncal Malformations.

In tricuspid atresia patients with normally related great arteries, the size of the interventricular communication and determination of the presence and degree of pulmonary outflow tract obstruction are essential. If pulmonary blood flow is unobstructed, limitation by means of pulmonary artery banding is usually undertaken; if blood flow is reduced, interventions to increase pulmonary blood flow will need to be considered. Some patients have just enough restriction to pulmonary blood flow that they are neither overcirculated nor excessively cyanotic—in other words, they are well balanced.

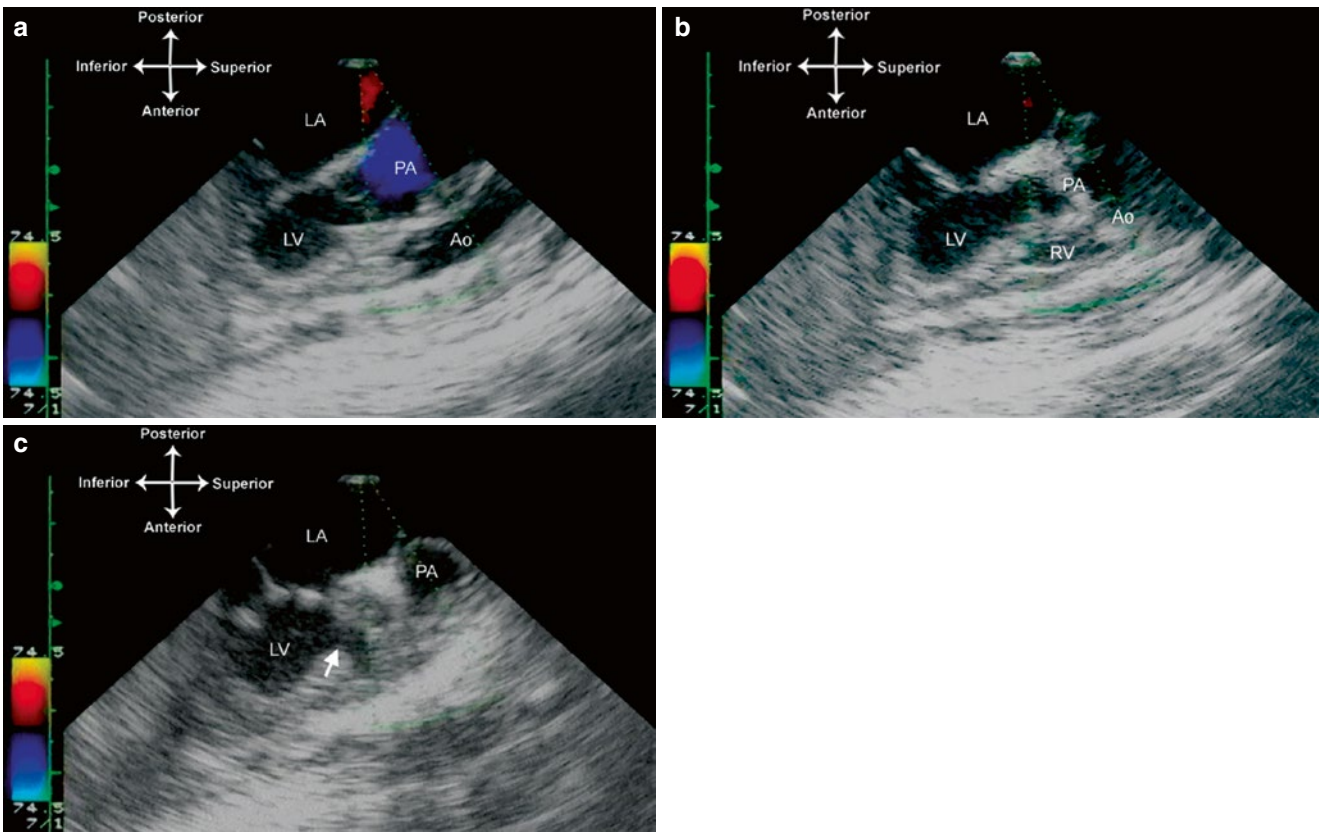

Fig. 10.4 Tricuspid atresia with D-transposed great arteries. Images taken from the mid esophageal aortic valve long axis view, multiplane angle 90°, showing the relationship of the great arteries with the pulmonary artery arising from the left ventricle, and the aorta arising from a small anterior right ventricular outflow chamber (**a**). The ventricular septal defect (bulboventricular foramen) is noted by the *arrow* (**b**). *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *Ao* Aorta

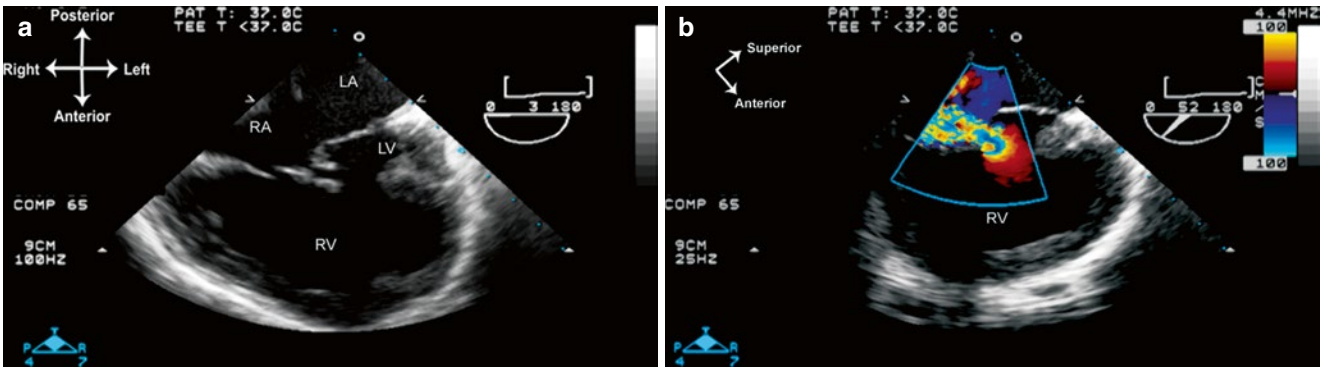

Fig. 10.5 Unbalanced atrioventricular (*AV*) septal defect. (**a**) Mid esophageal four chamber view demonstrates an extremely right dominant AV septal defect with a tiny left ventricle. The large common AV valve empties almost completely into the large right ventricle. (**b**) Significant central AV valve regurgitation as documented with a multiplane angle of 52°. *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

Such patients might not require any intervention for the first few months of their life, although ultimately they will require the same surgeries used for all single ventricle patients—namely, the Glenn and Fontan procedures (discussed below).

Univentricular Atrioventricular (AV) Connection

This category of single ventricle includes those defects in which both atria empty completely (or almost completely) into one large ventricle. A second ventricle might or might not be present; if present, it is generally quite hypoplastic. This group of defects includes unbalanced AV septal defect, double inlet ventricle (DILV), as well as the rare double inlet right ventricle (DIRV) and true single (or solitary) ventricular chamber of indeterminate morphology.

Patients with unbalanced AV septal defects have similar characteristics to a large AV septal defect (Chap. 8—AV septal defects and AV valve anomalies), but the common AV valve—instead of being balanced equally over both ventricles—is oriented significantly toward one ventricle. Thus, both atria empty primarily or almost completely into that chamber, and there is usually some degree of hypoplasia of the other ventricle (Fig. 10.5). In some patients, the degree of AV valve unbalance is so pronounced, and/or the degree of ventricular hypoplasia so apparent, that it is clear that the defect cannot be septated — i.e. the patient must be treated as a functional single ventricle. In others, this assessment is more difficult, and various criteria are utilized to determine whether a biventricular repair is feasible [29, 30]. Unbalanced AV septal defect is a common accompanying feature of heterotaxy syndrome (discussed below). The TEE evaluation of such patients is very much the same as for any patient with an AV septal defect, with particular attention paid to the degree of common AV valve distribution or commitment to the underlying ventricular chambers, AV valve function, ventricular hypoplasia, and possible aortic or pulmonary outflow tract obstruction. A combination of mid esophageal views (ME 4 Ch, ME 2 Ch, ME LAX) are best for evaluating the common AV valve and ventricular chambers, and also the degree of AV valve regurgitation (if present). When available, the deep transgastric views (DTG LAX, DTG Sagittal) can be used to evaluate the AV valve *en face* and its balance over both ventricles, similar to the views obtained with a subcostal sagittal view [29, 31]. The TEE evaluation of AV septal defects is discussed extensively in Chap. 8.

Double inlet ventricles are characterized by both atria and AV valves emptying into a large ventricle of either left or right ventricle morphology. The vast majority of these are DILV: both AV valves empty into a large ventricle of LV morphology, and there is generally a smaller RV outflow chamber located anteriorly. In many cases of DILV, it is very difficult to determine which AV valve is tricuspid, and which is mitral. There are three general classes of DILV: (1) normally related great arteries; (2) D-transposed great arteries; (3) L-transposed great arteries. The patient with DILV and normally related great arteries, also known as a Holmes' heart [32], has a normal aortic valve and aorta arising from the large LV, with the pulmonary artery arising from the RV outflow chamber with a variable degree of subpulmonary stenosis (Fig. 10.6, Video 10.5). The patient with DILV and D-transposed great arteries has the aorta arising from a rightward, RV outflow chamber; with L-transposed great arteries the aorta arises from a leftward RV outflow chamber. Of the three types of DILV, the L-transposed is the most common type (60–65 % of all DILV), followed by the D-transposed form (20–25 %), and lastly the normally related great arteries variation (15 %). In both forms of DILV with transposed great arteries, there is either significant subaortic narrowing (often with ascending aorta and aortic arch hypoplasia) or subpulmonary narrowing, depending on the size of the VSD/BVF and position of the infundibular septum [1, 33]. This is analogous to tricuspid atresia and transposed great arteries, as described above.

DIRV is a rare defect in which both AV valves empty into a chamber of RV morphology [34]. In these instances, there

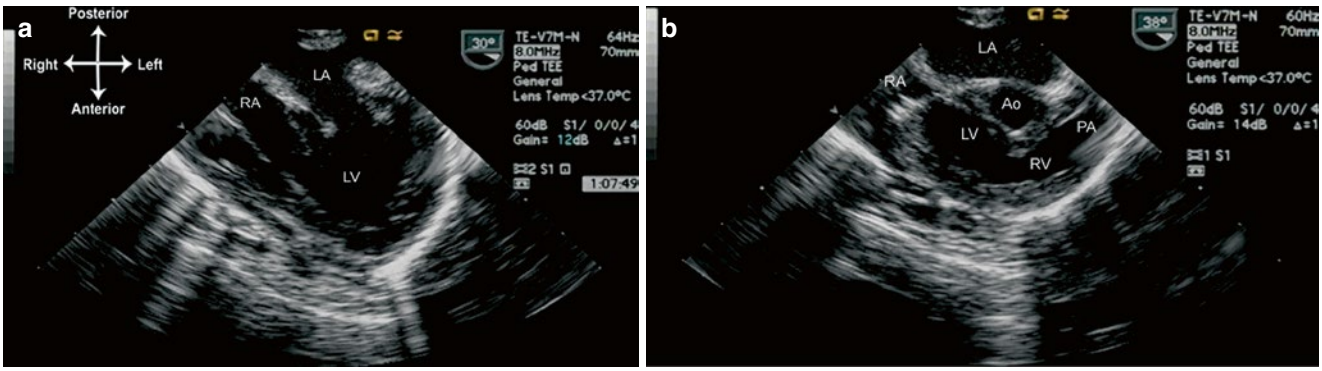

Fig. 10.6 Double inlet left ventricle, normally related great arteries (Holmes' heart) as viewed from the mid esophageal four chamber view. (**a**) A multiplane angle of 30° is used to visualize both atrioventricular valves. (**b**) Rotation of multiplane angle to approximately 40° and probe tip anteflexion demonstrate the origin of the pulmonary artery from the hypoplastic right ventricular outflow chamber (*RV*). *Ao* aorta, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium

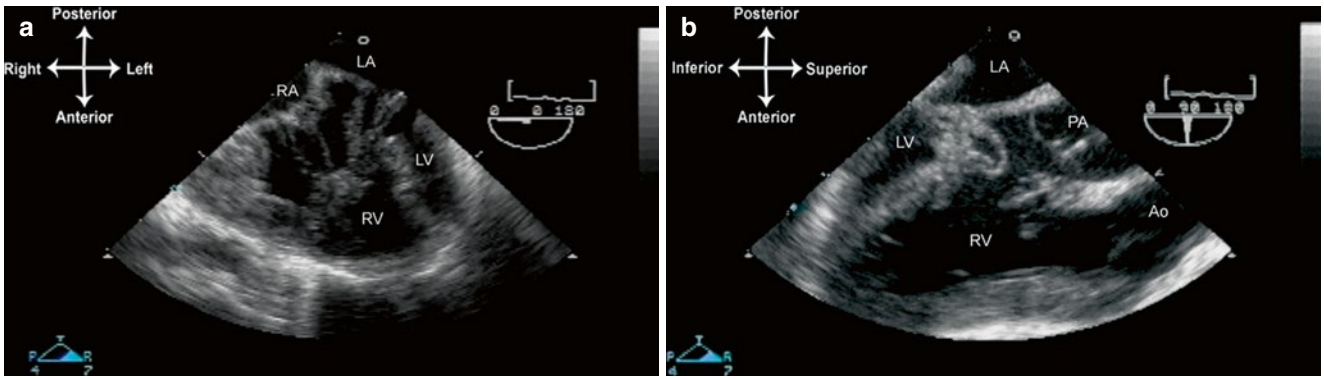

Fig. 10.7 Double inlet, double outlet right ventricle. (**a**) This image is obtained from the mid esophageal four chamber view. Both right and left atrioventricular valves drain into the large right ventricle (*RV*), inserting into a large, bizarre RV papillary muscle. There is a hypoplastic left sided left ventricle (*LV*). (**b**) Equivalent of the mid esophageal aortic valve long axis (90°) view. Both great arteries are seen to arise from the RV, with the aorta anterior to pulmonary artery. *Ao* aorta, *LA* left atrium, *PA* pulmonary artery, *RA* right atrium

are prominent muscle bundles in the large RV chamber; occasionally a large muscle bar is present that can be confused for a remnant of ventricular septum [35]. Usually, both great arteries also emerge from the same large RV, thus this lesion is often termed a double inlet, double outlet RV [35]. However unlike DILV, no anterior outflow chamber is visible. Instead, a rudimentary LV chamber may or may not be present; when present, it is located posteriorly, with no outflow to either great artery (Fig. 10.7, Video 10.6). Varying degrees of obstruction to one of the great arteries (generally the pulmonary trunk) can also be seen [36].

In the true single ventricle, there is no visible second chamber, and the large solitary ventricle has a very primitive morphology that eludes definitive identification as either an RV or LV [4, 33, 37]. There can be either one or two AV valves emptying into the large chamber, or there can even be one large common AV valve (i.e. a large common AV canal). Often, the heart has a very primitive appearance, with bizarre and loose muscular trabeculations seen in the large ventricular cavity. The great arteries have a variable relationship, generally with either stenosis of the aortic or pulmonary artery outflow tracts. There is a higher prevalence of atrial isomerism with indeterminate ventricular morphology [38].

Evaluation of the univentricular heart by TEE is similar to the evaluation of tricuspid atresia. The views of the inflows are best obtained beginning with the lower esophageal situs short axis (LE Situs SAX), first using a multiplane angle of 0–20°, then withdrawing the probe to visualize the junction between IVC and right atrium. Further withdrawal of the probe to the ME 4 Ch view demonstrates the AV valve(s) by imaging. Both color flow and spectral Doppler are necessary to evaluate AV valve function. When two AV valves are present, if either AV valve is stenotic or atretic, patency of the atrial septum must be assessed both by imaging and color flow Doppler. The relationship of the great arteries must also be determined; if one of these arises from a smaller outflow chamber, the VSD/BVF size and patency also need to be evaluated, preferably from several multiplane angles between 0 and 90°. Any subpulmonary or subaortic stenosis can be well visualized from a ME RV In-Out, ME AV LAX, and ME LAX views, multiplane angles between 70° and 120°, optimized to yield the best longitudinal view of

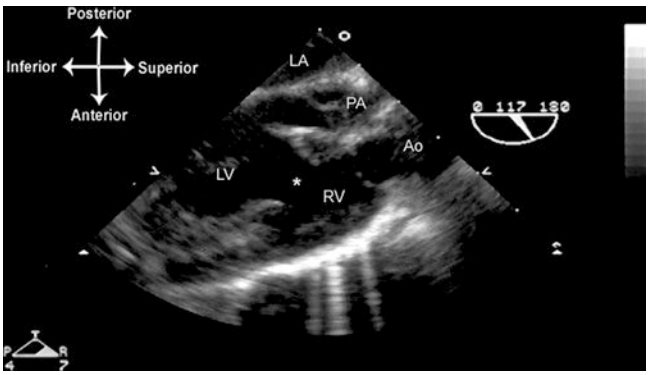

Fig. 10.8 Double inlet left ventricle, D-transposed great arteries, following pulmonary artery band placement. The mid esophageal aortic valve long axis view, multiplane angle of 117°, optimally demonstrates the long axis of both outflow tracts, and also shows the ventricular septal defect/bulboventricular foramen (*). This view also captures the left atrium and both ventricles in the same cross-sectional plane as the two outflow tracts. *Ao* aorta, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *RV* right ventricle

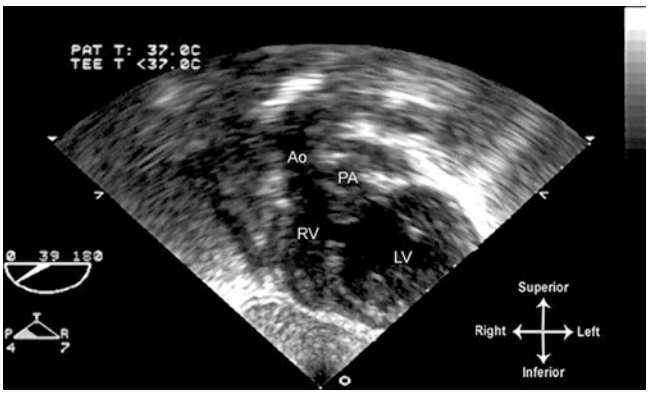

Fig. 10.9 Image from same patient as depicted in Fig. 10.8 obtained from a deep transgastric long axis view, multiplane angle of 39°. Both outflow tracts are displayed. This view offers an excellent angle for spectral Doppler evaluation of the outflow tracts. *Ao* aorta, *LV* left ventricle, *PA* pulmonary artery, *RV* right ventricle

the outflow tract (Fig. 10.8, Video 10.7). When available, the deep transgastric windows (DTG LAX, DTG Sagittal) provide good visualization of a number of structures, including the VSD/BVF, as well as excellent angles of Doppler interrogation for gradients across either the aortic or pulmonary outflow tracts (Fig. 10.9).

As with tricuspid atresia, left juxtaposition of the atrial appendages is also associated with DILV and D-transposed great arteries, and care should be taken not to confuse this entity with a large atrial septal defect.

Heterotaxy

Despite an external appearance of right-left symmetry, much of the human body's internal organ layout is in fact asymmetric, and this asymmetry leads to a definition of normal (and abnormal) sidedness. In *situs solitus* (which typifies most humans), the liver and IVC are on the right, the stomach and aorta on the left. The heart, being an asymmetric organ, also lateralizes to lie in the left chest. In *situs inversus*, the organs lateralize to the opposite side, producing a "mirror image" layout of the heart, lungs, and abdominal viscera. The term *heterotaxy* denotes an abnormality of laterality, or sidedness, in which there is (paradoxically) bilateral visceroatrial symmetry. Hence this condition is known as *situs ambiguous* or *atrial isomerism* and involves multiple organ systems in the body, including the heart. From a genetic standpoint, it is now known that heterotaxy belongs to a class of abnormalities known as "ciliopathies", characterized by abnormalities or dysfunction of primary cilia. During cardiac embryogenesis, primary cilia on the embryonic node are required for generation of the left-right axis. Ciliary dysfunction abnormalities are responsible for disorders of left-right axis determination during early embryonic development. This results in a disorder of laterality, manifested by abnormalities of any internal organ that is asymmetrically position—including the heart. Significant phenotypic variation can result [39, 40].

Patients with heterotaxy generally have significant cardiac abnormalities, often affecting multiple segments of the heart [41–43]. However despite the myriad potential variations in cardiac anatomy, heterotaxy patients tend to fall into one of two categories: (1) right isomerism (bilateral right-sidedness), or **asplenia**; (2) left isomerism (bilateral left-sidedness), or **polysplenia**. The visceral and cardiac variants most commonly seen with each category are listed in Table 10.2. It is important to note, however, that this table lists only the most commonly associated features seen with each category, and a number of exceptions have been reported, as well as considerable overlap between the two categories.

For most heterotaxy patients, especially those with right isomerism (asplenia), the preponderance of cardiac abnormalities—particularly hypoplasia of one ventricle—dictates that a single ventricle pathway be chosen [44, 45]. However in a small percentage of patients (mainly those with left isomerism, or polysplenia), a two ventricle approach can be contemplated [46, 47]. Therefore when a TEE evaluation of either right or left isomerism is performed, a very careful and comprehensive evaluation is recommended in consideration of the implications regarding patient management. Because of potential for anatomic abnormalities at multiple levels, a complete, segmental examination (Chap. 4) should be performed, particularly in those patients with unknown or uncertain anatomy. Using the many available TEE views discussed in Chap. 4 as well as this chapter, careful attention should be paid to systemic and pulmonary venous drainage, number and function of the AV valves, ventricular size and function, great artery position, and possible outflow tract stenosis. In the intraoperative setting, the examination also needs to be tailored to the important surgical questions that must be answered for the proposed surgery. For example, if Fontan surgery is contemplated, evaluation of systemic and pulmonary venous drainage is paramount (Fig. 10.10, Video 10.8)

Table 10.2 Right vs. left isomerism: common anatomic features

	Right isomerism (asplenia)	Left isomerism (polysplenia)
Liver	Midline, symmetric	Asymmetric or symmetric
Lungs	Bilateral tri-lobe	Bilateral bi-lobe
	Eparterial bronchi	Hyparterial bronchi
Spleen	Usually absent, but rarely can be multiple	Usually multiple, but can be only one or absent
Cardiac position	Dextrocardia (36–41 %)	Dextrocardia (33–42 %)
Systemic venous drainage	Bilateral SVCs are common	Interrupted hepatic segment of IVC; azygous or hemiazygous continuation to right or left SVC
	Coronary sinus usually unroofed	One or both SVCs can be present; if two SVCs, one generally drains into a coronary sinus
	IVC usually present; IVC and hepatic veins return together to common atrium	Separate hepatic drainage to atrium
	There can be separate drainage of hepatic veins	
Pulmonary venous drainage	Anomalous pulmonary venous drainage to systemic vein (62–88 %)	Usually to back of common atrium; abnormal position of atrial septum results in right veins to right side, left veins to left side
	Can return directly to heart, often via a common collector	
Atria and atrial septum	Bilatral right atrial appendages	Bilateral left atrial appendages
	Usually common atrium; small remaining atrial septal remnants	Usually common atrium; if atrial septum present, usually primum atrial septal defects
AV valve	Large common AV valve, often RV dominant	Two AV valves (common AV valve can also be seen)
Ventricles	Usually large RV, hypoplastic LV	Can be two balanced ventricles or one dominant ventricle
Ventricular outflow tracts	Often subvalvar/valvar pulmonary stenosis or atresia (88–96 %)	Valvar/subvalvar pulmonary stenosis (about 40 %) and aortic obstruction/atresia (17–22 %)
	Rarely, aortic stenosis/atresia/coarctation	
Ventriculo-arterial relationships	DORV (82 %)/TGA (9 %); often subvalvar/valvar pulmonary stenosis or atresia (88–96 %)	Normally related great arteries (50–84 %); DORV or ventriculo-arterial discordance (16–37 %);
	Rarely, aortic stenosis/atresia/coarctation	

AV atrioventricular, *DORV* double outlet right ventricle, *IVC* inferior vena cava, *LV* left ventricle, *RV* right ventricle, *SVC* superior vena cava

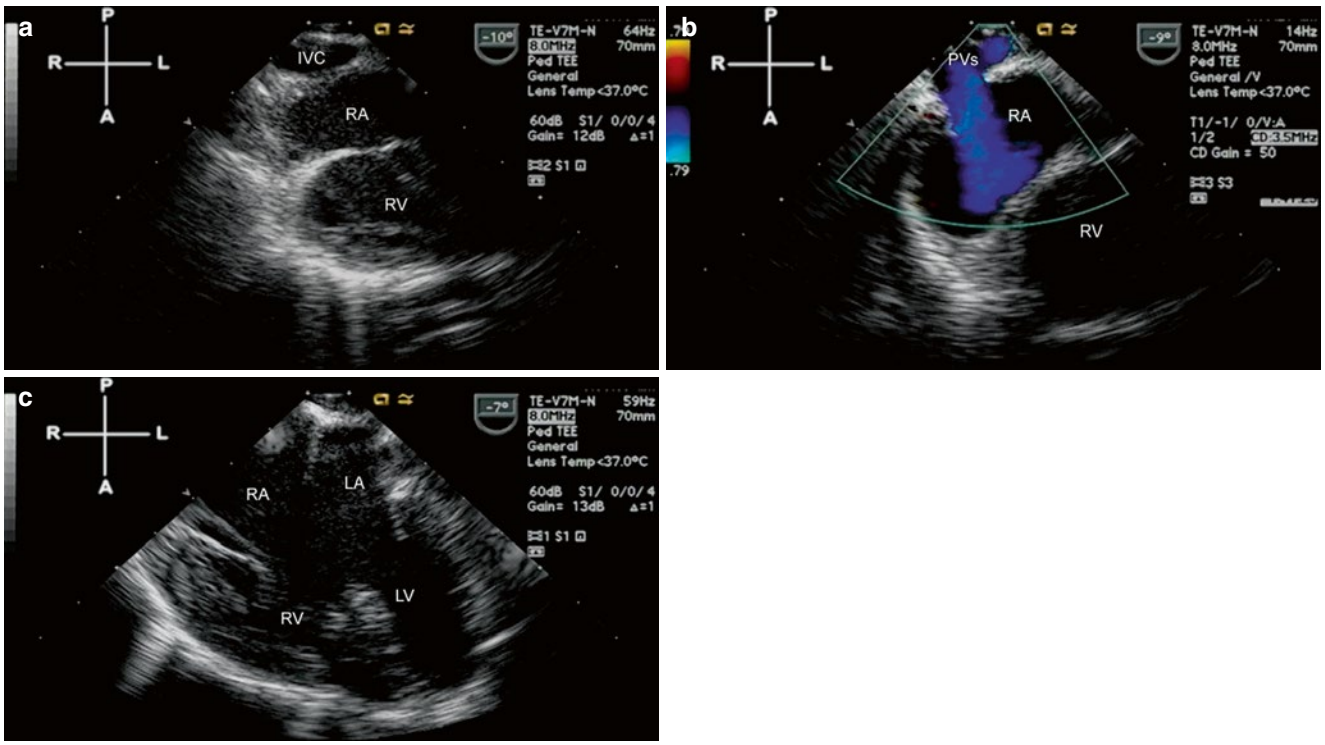

Fig. 10.10 Pre-Fontan study in a patient with right isomerism (asplenia) and double outlet right ventricle/pulmonary atresia. (**a**) Is obtained using multiplane angle 0° after withdrawal of the TEE probe from a lower to mid esophageal position. The inferior vena cava (*IVC*) receives the right hepatic veins and returns to the right atrium. In (**b**) the pulmonary veins are shown as they also return to the right sided atrium. (**c**) Demonstrates the presence of an unbalanced atrioventricular septal defect, right dominant. *LA* left atrium, *LV* left ventricle, *PVs* pulmonary veins, *RA* right atrium, *RV* right ventricle

Other Types of Single Ventricle

There are a number of other forms of complex congenital heart disease that—for one reason or another—fall under the "single ventricle" umbrella, even if they have two AV valves and two ventricles. In some patients, both ventricles are well-formed but other intracardiac features make a two ventricle repair unfeasible or undesirable, and therefore these patients are placed on a single ventricle pathway. Such complicating features include straddling/overriding AV valves, or double outlet RV with a VSD that is remote from either semilunar valve and makes two ventricle repair a high-risk proposition [48–50]. There are also some rare, complex cardiac defects that historically do not lend themselves well to a two ventricle repair. These include some forms of double outlet LV [51], superior-inferior ventricles with criss-cross AV relations [52, 53], etc. In many of these cases, there exist significant intracardiac abnormalities in addition to right or left ventricular hypoplasia.

When TEE is performed in patients with one of these complex cardiac lesions, a complete evaluation should be performed, with specific attention paid to AV valve size and orientation, ventricular chamber size and location of ventricular septal defect(s), and great artery relationships. In some cases, information from the TEE will be utilized to render an assessment as to whether a two ventricle repair is feasible. Currently, the bias for most clinicians caring for CHD patients is to perform a two ventricle repair whenever possible. However, the question as to whether a good one ventricle repair is better than a marginal two ventricle repair remains unanswered and is, in many instances, controversial [54–56].

Surgery for the Single Ventricle

With a single ventricle and single ventricle physiology, there is only one functioning pump that must serve both systemic and pulmonary circulations. Because of this, obligatory mixing of systemic and pulmonary blood flow occurs, resulting in systemic arterial hypoxemia. The eventual goal of surgical management of the single ventricle is to restore normal systemic arterial oxygenation by separating the circulations; this is performed by rerouting systemic venous (deoxygenated) blood directly into the pulmonary arteries, thereby bypassing the heart. Deoxygenated blood is expected to flow directly to the pulmonary arteries without the aid of a cardiac pumping chamber [57]. This is known as the Fontan procedure, which is named and based upon the original surgery described in 1971 by Fontan and Baudet for treatment of tricuspid atresia [58]. Over the past 40+ years the Fontan procedure has undergone a number of modifications and improvements, and has gained widespread acceptance as the preferred surgical management strategy for all forms of single ventricle [59]. It has been clearly established that, even without the aid of a right-sided pumping chamber, Fontan physiology is compatible with long-term survival and good functional status [60]. There are still a number of ongoing questions regarding long-term morbidity (arrhythmias, protein-losing enteropathy, liver dysfunction, ventricular dysfunction, plastic bronchitis, etc.) and survivability into late adulthood; these topics are beyond the scope of the current chapter but are fully discussed in many other references [59, 61–64].

While the ultimate goal for all single ventricle patients is the Fontan procedure and achievement of Fontan physiology, the complete Fontan operation cannot be performed in neonates or young infants because of a still immature pulmonary vascular bed, and still developing neonatal myocardium. The ideal timing for the Fontan procedure is generally felt to be after 2–3 years of age [65, 66]. Therefore, assuming single ventricle patients are correctly diagnosed in early infancy, almost all will require one or more staging surgeries prior to the Fontan procedure. The purpose of these staging surgeries is to ensure that patients are optimally prepared for their eventual Fontan procedure. For successful Fontan physiology, certain key anatomic and physiologic factors should ideally be present, including adequate size pulmonary arteries, low pulmonary arteriolar resistance, no more than mild AV valve regurgitation, and normal ventricular systolic and diastolic function [65, 66]. A full discussion of these important factors can be found in many references [59, 63, 67–70].

Different institutions utilize TEE to varying degrees for the various surgical procedures performed as part of single ventricle surgical pathway. Nonetheless, TEE can provide useful and important information. Preoperative evaluation of single ventricle patients by TEE is generally excellent due to its superior imaging capabilities. However, most preoperative single ventricle patients will have already undergone extensive imaging by several different modalities, hence TEE is used primarily as a complementary imaging technique, e.g. when assessment of AV valve function is desired in a larger patient with poor transthoracic imaging. For postoperative assessment, TEE can play a more prominent role. It is well known that postoperative single ventricle patients, particularly Fontan patients, are notoriously difficult to image by transthoracic echocardiography. Therefore TEE can provide a much greater contribution in the assessment of such patients, most notably in the immediate postoperative period. The following section discusses the important preoperative and postoperative features that can and should be assessed by TEE, whenever it is performed in single ventricle patients.

Preoperative Assessment

The preceding sections of this chapter detailed individual defects considered part of the single ventricle spectrum, including specific anatomic characteristics that should be evaluated. While considerable anatomic heterogeneity exists among the different classes of defects, what becomes evident is that certain common anatomic and functional characteristics must be explored whenever a TEE is performed for preoperative assessment. These characteristics are relevant not only for the patient's current clinical condition, but also for any future palliative surgeries that might be contemplated. The list of potential surgeries for single ventricle palliation is limited (as will be discussed below), thus certain key anatomic features carry great weight—and therefore assume particular relevance—in the preoperative evaluation of these patients. Some of the features were mentioned previously for the individual cardiac lesions, but the following list summarizes the essential elements that should be addressed during preoperative assessment of all such patients:

- **Systemic venous return**. The goal of single ventricle palliation is the return of all systemic venous blood directly to the pulmonary arteries. Thus, detailed knowledge of systemic venous return is essential. Although in most cases systemic venous drainage will have been adequately delineated prior to surgery, in many instances TEE can provide helpful confirmatory information. The presence/absence of the hepatic portion of IVC and the drainage of the hepatic veins should be verified. In some patients, particularly those with visceral heterotaxy, some hepatic veins can drain separately from the IVC and directly into the atrium (Fig. 10.11, Video 10.9). In others, particularly those patients with polysplenia (left isomerism), the hepatic portion of the IVC can be absent (interrupted IVC with azygos continuation) and there can be separate entrances of right and left hepatic veins into the atria. Advancing the TEE probe to the lower esophageal position (LE Situs SAX, multiplane angle 0–20°), with careful withdrawal and clockwise/counterclockwise turning of the TEE probe, can demonstrate the sites of IVC and hepatic vein drainage. Identification of the SVC drainage is also very important: both the number of SVCs (one vs. two), and their sites of drainage, should be determined. This can be performed by withdrawing the TEE probe to the mid/upper esophagus, including the mid esophageal ascending aortic short axis (ME Asc Ao SAX) and UE PA LAX views (both using a multiplane angle 0°), and rotating clockwise and counterclockwise. It is important to search directly for a separate right and left SVC and not rely upon the presence/absence of a dilated coronary sinus, because the coronary sinus could be unroofed or absent, allowing left SVC return directly to the atrium. Sagittal views of both SVCs can then be obtained in the mid to upper esophageal position, multiplane angle at approximately 80–90°, using a combination of views including ME Bicaval, mid esophageal ascending aortic long axis (ME Asc Ao LAX) with rightward rotation to visualize a right SVC, and ME 2 Ch, ME Asc Ao LAX, and UE Ao Arch (all with leftward rotation) to visualize a left SVC. With these sagittal views, azygos or hemiazygos drainage to the SVC can sometimes be seen coursing posteriorly and over the corresponding branch pulmonary artery.
- **Pulmonary venous return**. In general, pulmonary venous drainage will have been established with imaging studies performed prior to the TEE. Nonetheless, in some single ventricle patients the drainage of the pulmonary veins might not be completely clear. The role of the preoperative TEE is to confirm the drainage of all pulmonary veins, and establish that this drainage is unobstructed. Because of the close proximity of the esophagus to the atria, TEE generally provides excellent visualization of pulmonary venous return, both for imaging and color

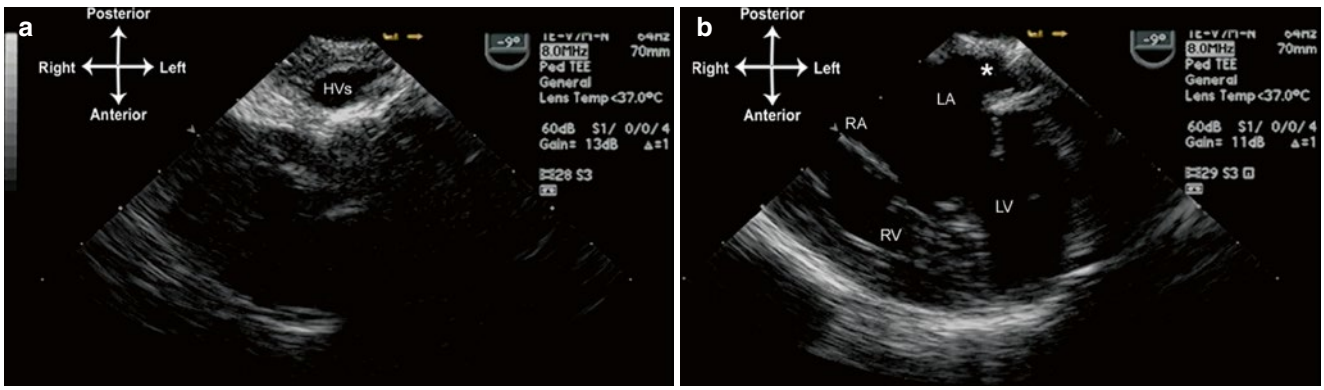

Fig. 10.11 Image from patient in Fig. 10.10. The left sided hepatic veins return separately to the left sided atrium (**a**). The entrance of these veins into the left atrium is marked by an asterisk (**b**). *HVs* left hepatic veins, *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

flow/spectral Doppler evaluation (Chaps. 4 and 6). Any obstruction to pulmonary venous flow must be noted, because it will have negative consequences for further single ventricle palliation [71, 72]. The relationship of the pulmonary venous return to the systemic veins should also be noted, as the placement of a Fontan conduit could potentially interfere with pulmonary venous return, and surgical adjustments might be required [73].

- **Patency of the atrial septum**. An unrestrictive atrial septal defect is essential in those patients in whom systemic or pulmonary venous return empties into an atrial chamber associated with a stenotic or absent AV valve, or a hypoplastic/nonfunctional ventricular chamber (e.g. tricuspid atresia or HLHS). In such cases, the only pathway for egress of blood from the chamber is through the atrial communication. Using TEE, this can be evaluated both by imaging as well as color flow/spectral Doppler interrogation. Midesophageal views with a multiplane angle of 0–90° (ME 4 Ch, ME Bicaval) can be used, combined with other probe maneuvers such as advancement/withdrawal and left/right rotation. There are two types of restrictive atrial communication: with left to right shunting, and with right to left shunting. A restrictive *left to right* atrial communication is found in those defects in which there is left sided obstruction, such as HLHS and DILV with left AV valve stenosis/atresia. In these cases, atrial restriction is characterized by a number of findings, including: (a) small size (diameter <3 mm); (b) color flow Doppler aliasing of the atrial shunt and a mean gradient >5 mmHg [74]; (c) dilated pulmonary veins, along with prominent flow reversal in these structures; (e) dilated left atrium, with an atrial septum bowing toward the right (although a large left atrium does not always indicate a restrictive atrial communication); (f) in HLHS patients, a small left atrium with thick atrial septum [75]. The incidence of atrial septal restriction in HLHS patients is estimated between 6 and 20 % [75–78]. When the atrial septal communication is felt to be restrictive, it often requires enlargement, either by interventional cardiology procedures such as balloon dilation/stenting of the septum, or an atrial septectomy at the time of surgery. In HLHS patients with a restrictive atrial septal defect, emergent atrial septostomy is sometimes necessary, though mortality is much higher in this subset of patients [76, 78, 79]. This has led to development of *in utero* atrial septoplasty in HLHS patients prenatally diagnosed as having a restrictive atrial septal defect [80].

  A restrictive *right to left* atrial communication is found in those defects in which there is obstruction to right AV valve inflow, such as tricuspid atresia, hypoplastic right heart syndrome, or DILV with right AV valve stenosis/atresia. In these cases the restriction can be more challenging to determine; unlike HLHS and other left sided obstruction, the atrial septum is often redundant and it can be difficult to define accurately the margins of the atrial defect. Moreover, in some patients restriction might not be apparent immediately after birth, but can increase with time [81]. This type of atrial septal restriction occurs in 5–15 % of tricuspid atresia patients [82, 83]. Overall, less data is available in this group of patients. One study has shown that an atrial septal aneurysm (defined as bulging >6 mm beyond the plane of the atrial septum) and atrial septal defect diameter <5 mm (measured on the 2D echocardiographic image) places tricuspid atresia patients at higher risk for atrial septal restriction [83].
- **Atrioventricular valve (AV) function**. The AV valve(s) should be examined structurally in terms of the leaflets, support apparatus, and annular size. The function of the valves should be assessed as well. This is accomplished by two-dimensional imaging as well as color flow mapping and spectral Doppler. In single ventricle patients, the presence and degree of AV valve regurgitation can have significant bearing on clinical outcomes [84]. In patients with two AV valves (e.g. double inlet LV), the function of both valves should be examined. If one valve is excessively regurgitant, and the other valve competent and of adequate size, the regurgitant valve can be repaired or even oversewn. If one AV valve is significantly stenotic or atretic, a large atrial defect is mandatory to allow atrial blood to pass unobstructed to the other AV valve (as noted above). For the TEE evaluation of the AV valves, the mid esophageal views (ME 4 Ch, ME 2 Ch, ME Mitral, ME LAX) are useful to evaluate valve morphology and function. Additional information regarding chordal and papillary muscle architecture can be provided by the transgastric views: the "short axis" transgastric views (TG Basal SAX, TG Mid SAX) and "long axis" transgastric views (TG 2 Ch, TG LAX, and transgastric RV inflow or TG RV In) views. Given the unusual orientation of the AV valve(s) in many forms of single ventricles, variation in multiplane angle between 0° and 120° in many of these views might be necessary to facilitate optimal visualization. In some instances the deep transgastric views (DTG LAX, DTG Sagittal) can be useful for *en face* evaluation of the AV valves.
- **Ventricular systolic and diastolic function**. Ventricular function plays an essential role in the short and long-term viability of the patient with a single ventricle. However given the unusual and often distorted geometry of many forms of single ventricle, it can be difficult if not impossible to provide accurate quantification of ventricular systolic function, particularly within the time constraints of the intraoperative setting. Nonetheless some type of qualitative assessment of ventricular systolic function should be performed, even if it falls under a somewhat subjective assessment range of "*normal*" to "*mild-moderate-severely depressed*". Any regional wall motion abnormality, including segmental ventricular hypokinesis or dyskinesis,

should be recorded and noted. If time permits, assessment of diastolic and global cardiac function can be performed by analysis of AV valve inflow and tissue Doppler analysis, and other measurements such as the myocardial performance (Tei) index [85–88]. Again, given the abnormal geometry in many single ventricle patients, the utility of standard two-dimensional echocardiographic measurements remains to be determined. The TEE analysis of ventricular function is discussed in detailed in Chap. 5.

- **Ventricular septal defect (VSD) or bulboventricular foramen (BVF)**. Very rarely does a single ventricle patient actually possess only one ventricular chamber. More commonly, there is a large functional ventricular chamber and a smaller, nonfunctional chamber. The connection between these two chambers occurs via a VSD of variable size. In many instances the small ventricular chamber derives solely from the embryologic bulbus cordis, and accepts no AV valve tissue, hence the communication is termed the "bulboventricular foramen" or BVF as previously noted. In those cases in which the aorta arises from the smaller ventricular chamber (e.g. tricuspid atresia or DILV with transposed great arteries), a small VSD/BVF can result in significant subaortic obstruction which, if not addressed, can lead to significant ventricular hypertrophy as well as systolic/diastolic dysfunction (all negative risk factors for subsequent Fontan physiology). Progression of VSD/BVF obstruction can also occur over time after palliative interventions and even after the Fontan procedure [89]. Therefore a careful assessment of the VSD/BVF and subaortic anatomic region must be performed to determine whether this issue needs to be addressed, either by enlargement of the VSD/BVF, a palliative arterial switch operation, or a Damus-Kaye-Stansel procedure (see below). The TEE assessment of the VSD/BVF consists of anatomic evaluation from the mid esophageal window (Fig. 10.8), and linear measurements of diameter should be obtained in at least two orthogonal planes (for example, ME 4 Ch at 0° and ME 2 Ch at 90°). Using measurements of a major and minor axis, Matitiau et al. found that the cross-sectional area of the VSD/BVF can be estimated using the formula of an ellipse (major diameter × minor diameter × π/4); a cross-sectional area <2 cm^2/m^2 was found to be predictive of subsequent VSD/BVF obstruction [90]. However, this formula might be simplistic because in many cases, the cross-sectional profile of the defect is asymmetric and irregular. Depending upon the location and orientation of the defect, the Doppler gradient across the VSD/BVF can also be obtained by the mid esophageal views (ME 4 Ch, ME LAX, ME AV LAX, etc.), generally with multiplane angles approximating 80–90°, or from the deep transgastric window (DTG LAX, DTG Sagittal) with probe anteflexion. The Doppler-derived gradient can be quantified using the simplified Bernoulli equation: **maximum instantaneous pressure gradient = $4V^2$,** with *V* equal to the maximum velocity (meters/second) obtained by continuous wave Doppler (see also Chaps. 1 and 5). However spectral Doppler evaluation across a VSD/BVF can be misleading, because in the presence of a large unobstructed pulmonary valve, there might not be increased flow velocity across the VSD/BVF, even with a potentially restrictive ventricular communication. The reason for this is that an elevated pressure will not develop in the ventricular chamber proximal to the VSD/BVF, as blood flow "decompresses" through the large pulmonary valve. Thus 2D anatomic evaluation of the VSD/BVF is much more valuable for assessment. The native aortic valve annulus and ascending aorta (if visible) can also be visualized and measured using ME AV LAX, ME Asc Ao LAX and UE Ao Arch SAX views (multiplane angle 80–110°). Concurrent aortic and aortic arch anomalies (hypoplasia, interruption) are associated with significantly smaller VSD/BVF areas, however other imaging modalities (transthoracic echocardiography, MRI) are often needed to assess these [91].
- **Outflow tract and semilunar valve patency and function**. The size and patency of the outflow tracts and semilunar valves should be assessed during the TEE examination. Standard mid-esophageal views—ME 4 Ch, ME LAX, ME AV LAX, ME AV SAX, ME RV In-Out—are usually adequate to provide evaluation of these structures. Deep transgastric views (DTG LAX, DTG Sagittal) are useful for Doppler interrogation of flow across the valves. If one semilunar valve is hypoplastic or stenotic, the flow across the valve should be evaluated by color flow Doppler and the Doppler-derived gradient quantified by spectral Doppler, using the simplified Bernoulli equation described above. It should be noted that the gradient estimated by continuous wave Doppler represents a maximum instantaneous gradient, and therefore tends to overestimate the true peak to peak gradient as obtained by direct manometric measurement. Nonetheless it serves as a reasonable approximation. With the heterogeneity of single ventricle anatomy, it is impossible to stipulate a "best" view for spectral Doppler; the best window is the one that provides the most favorable Doppler angle of interrogation. Color flow Doppler should also be used for evaluation of the presence/degree of regurgitation of either semilunar valve, particularly in those cases in which a Damus-Kaye-Stansel or Norwood repair is contemplated, or in those patients who have already undergone these procedures.
- **Main and branch pulmonary artery architecture**. Pulmonary artery size and anatomy are two of the most important factors related to the eventual outcome of the single ventricle patient [65, 69, 92, 93]. Unfortunately the visualization of the pulmonary arteries by TEE is quite variable, and therefore TEE should not be the expected or preferred method for evaluation of pulmonary artery size or stenoses. When TEE does provide visualization of the

branch pulmonary arteries, features such as ductus arteriosus flow, and the sizes of the proximal right and left branch pulmonary arteries can be measured and compared. However, one cannot expect to rely consistently upon TEE assessment of pulmonary artery size and anatomy when planning surgical management.

Surgical Interventions for Single Ventricle and Their Assessment by TEE

Modified Blalock-Taussig and Central Shunts

The modified Blalock-Taussig shunt and its related variant, the central shunt, are mainstays of palliation for both one and two ventricle repairs. The purpose of these procedures is to augment pulmonary blood flow in patients with significant pulmonary stenosis or pulmonary atresia, thereby enhancing systemic arterial saturation. The original or classic Blalock-Taussig shunt involved ligation and division of the right subclavian artery, with the proximal portion turned down and anastomosed to the right pulmonary artery [94]. The modified Blalock-Taussig shunt was subsequently designed to preserve the blood flow to the distal right subclavian artery [95]. This modification involves a tube made of prosthetic material (generally 3.5–5 mm) anastomosed between the right subclavian or innominate artery at one end, and one of the branch pulmonary arteries at the other. The central shunt utilizes the same approach, but the connection of the shunt to the pulmonary artery is much more centrally placed; generally it is located on the main pulmonary artery at the bifurcation point of the two branch pulmonary arteries.

As noted previously, TEE visualization of the branch pulmonary arteries is highly variable. Moreover, visualization of the aortic head and neck vessels by TEE can be difficult. Therefore, TEE may have limited utility in the evaluation of the Blalock-Taussig or central shunt. The upper esophageal window must be used to visualize the shunt, using the upper esophageal aortic arch long axis (UE Ao Arch LAX) and UE PA LAX views; continuous color flow Doppler into a main or branch pulmonary artery confirms shunt patency (Fig. 10.12, Video 10.10). Continuous wave Doppler can be used to evaluate the gradient across the shunt, using the simplified

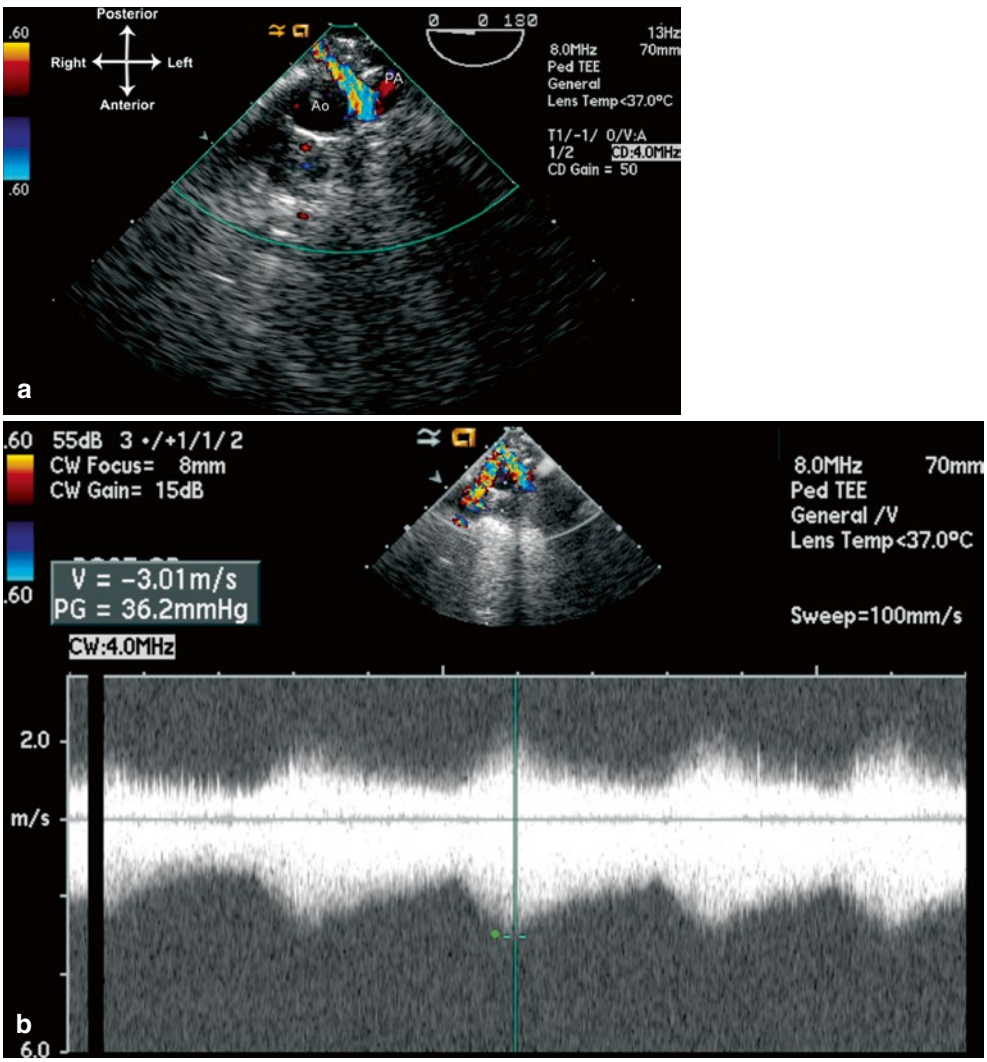

Fig. 10.12 Right modified Blalock-Taussig shunt, visualized from an upper esophageal pulmonary artery long axis view, multiplane angle 0°. (**a**) Flow entering the small right pulmonary artery by color Doppler. (**b**) Continuous flow as displayed by spectral Doppler. *Ao* ascending aorta, *PA* pulmonary artery

Bernoulli equation described above. The systolic pulmonary artery pressure can be estimated by the following equation:

$$\mathbf{PASP} = \mathbf{SBP} - 4V^2$$

PASP = Pulmonary artery systolic pressure (mmHg)
SBP = Systemic systolic pressure (mmHg)
V = Peak velocity (meters/second) across the shunt

As noted previously, the gradient estimated by continuous wave Doppler represents a maximal instantaneous gradient, and therefore tends to overestimate the true peak to peak gradient as obtained by direct manometric measurement. Moreover, because of sometimes unfavorable angles of interrogation, the calculated gradient might underestimate the true gradient across the shunt. Nonetheless with an optimally positioned spectral Doppler angle of interrogation, it can provide a reasonable approximation of the systolic pulmonary arterial pressure. A high gradient is generally desirable because it indicates a low pulmonary artery pressure (i.e. no pulmonary hypertension), while a low gradient could be indicative of elevated pulmonary artery pressure. With a change of the multiplane angle to 80–90° (UE Ao Arch LAX with rightward probe rotation), the length of the shunt can sometimes be visualized by a combination of imaging and color flow Doppler.

Pulmonary Artery Band

Banding of the pulmonary artery can be utilized in the surgical management of patients with biventricular circulations as well as those with a single ventricle. This procedure is performed to limit pulmonary blood flow in single ventricle patients with unobstructed antegrade flow into the main and branch pulmonary arteries. Such patients generally have pulmonary overcirculation leading to congestive heart failure, as well as high pressures in the pulmonary arteries. These two factors can lead to significant muscularization of the pulmonary arterioles and elevation of the pulmonary vascular resistance and pulmonary arterial pressures; these are known risk factors for subsequent single ventricle palliation [57, 70, 89]. Thus, the pulmonary artery band is placed both to limit pulmonary blood flow and to "protect" the pulmonary arteries from potentially detrimental exposure to high pressure and shear stress. The technical aspects of pulmonary artery band placement are relatively straightforward: a small length of material is placed around the main pulmonary artery, between the valve and pulmonary artery bifurcation, and restricts flow across the artery. Pressure gradients across the pulmonary artery band can be estimated by echocardiography or measured directly. One of the benefits of this surgical technique is that it can be performed quickly, without the use of cardiopulmonary bypass.

Band adjustments can be guided by TEE. This is best performed in the mid esophageal window (ME LAX, ME AV LAX, ME AV SAX, ME Asc Ao SAX, ME Asc Ao LAX), and upper esophageal window (UE Ao Arch SAX, UE Ao Arch LAX), with probe anteflexion and variable multiplane angles of interrogation from 0° to 90° depending upon the anatomy (Fig. 10.13a, b, Video 10.11). Alignment of the spectral Doppler cursor guided by two-dimensional and color flow mapping allows for estimation of the band gradient by continuous wave Doppler, using the simplified Bernoulli equation given above (peak velocity across the band is used for *V*). The deep transgastric window (DTG LAX, DTG Sagittal) can also be used for this evaluation (Fig. 10.13c–e, Video 10.11) Depending upon the spatial position of pulmonary artery in relation to the aorta (anterior vs. posterior), the most effective TEE view to obtain a band gradient will vary, and therefore the TEE probe will need to be manipulated accordingly to optimize Doppler interrogation of the band. Effective pulmonary artery banding produces a distal pulmonary artery systolic pressure about 30–50 % of systemic systolic arterial pressure [96]. Obviously the gradient across the band will depend upon systemic arterial pressure, but in general the gradient in a neonate will be between 40 and 70 mmHg (Doppler velocity about 3.2–4.2 m/s) [97]. For balanced single ventricle physiology (pulmonary: systemic flow ratio about 1:1), an optimal systemic arterial oxygen saturation is between 75 and 85 % [97, 98]. Also, with effective banding, there is often a shift in ventricular septal position that can be noted (if sufficient ventricular septum is present). Other features to evaluate include ventricular and AV valve function, which can be influenced by loading conditions. Longer-term issues to be assessed following pulmonary artery band placement include pulmonary valve competency (which can be adversely affected by a pulmonary artery band in close proximity), and possible migration of the pulmonary artery band distally, potentially impinging on one or both branch pulmonary arteries.

Damus-Kaye-Stansel Procedure

The Damus-Kaye-Stansel (DKS) procedure is considered for a single ventricle patient when there is potential or actual obstruction to blood flow through the native ascending aorta [99–101]. This could occur in patients such as those with transposition of great arteries and origin of the aorta from a small outflow chamber in the presence of a restrictive BVF (e.g. patients with tricuspid atresia or DILV), or in those with origin of the aorta from the larger ventricle, but with significant aortic or subaortic stenosis, or aortic atresia [102]. The DKS procedure involves transection of the native main pulmonary artery, and end to side anastomosis of the proximal main pulmonary artery to the ascending aorta, enabling unobstructed systemic output. The distal main pulmonary artery is oversewn, and pulmonary blood flow provided by either an aortopulmonary shunt or cavopulmonary connection (see below).

A standard preoperative TEE study is performed to evaluate intracardiac anatomy and the position and function of the semilunar valves, using the mid esophageal windows (ME

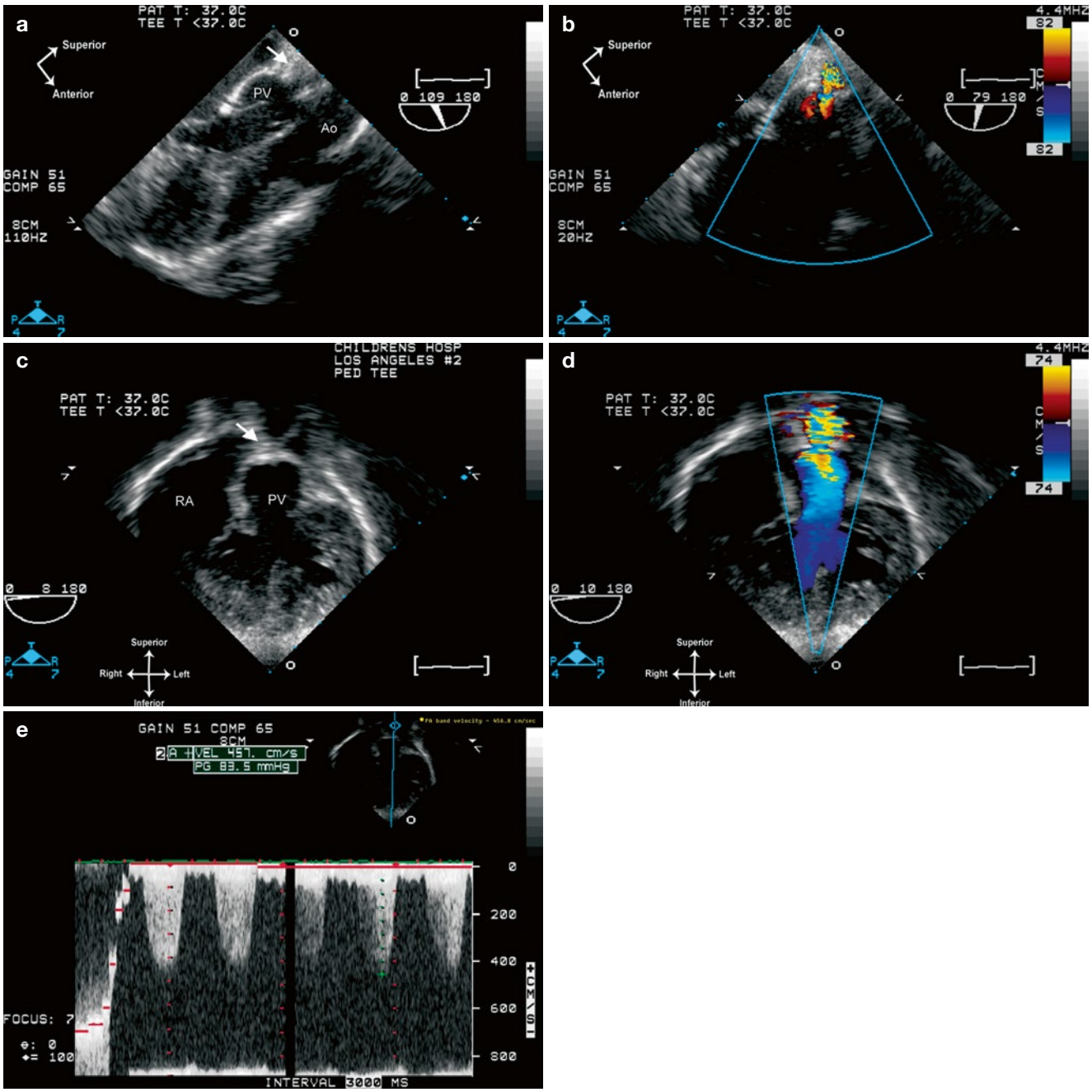

Fig. 10.13 Evaluation of pulmonary artery band. Two-dimensional image and color flow Doppler as seen from the mid esophageal long axis view (**a**, **b**) and deep transgastric long axis view (**c**, **d**). In both views, the location of the band above the pulmonary valve is indicated by the *arrow*. Note the excellent angle provided by the deep transgastric view for Doppler interrogation of the pulmonary band (**e**). *Ao* aorta, *PV* pulmonary valve, *RA* right atrium

LAX, ME AV SAX, ME AV LAX, ME LAX, ME RV In-Out) and varying multiplane angle to best display semilunar valve motion and function. Postoperatively, TEE can be used to assess patency of the great artery anastomosis as well as semilunar valve competence, which can sometimes be compromised during the procedure. Mid to upper esophageal views (ME Asc Ao LAX, ME Asc Ao SAX, UE PA LAX, UE Ao Arch SAX) obtained by multiplane imaging are useful for this evaluation (Fig. 10.14, Video 10.12) Significant semilunar valve regurgitation can prove detrimental to patient survival [103] (Fig. 10.15, Video 10.13). Ventricular and AV valve function should also be assessed.

As an alternative to the DKS procedure, a palliative arterial switch can be performed. This procedure, which is applicable when there are transposed great arteries with the aorta arising from the small outflow chamber (usually in the setting of tricuspid atresia or DILV), involves performing an arterial switch operation including a LeCompte maneuver to

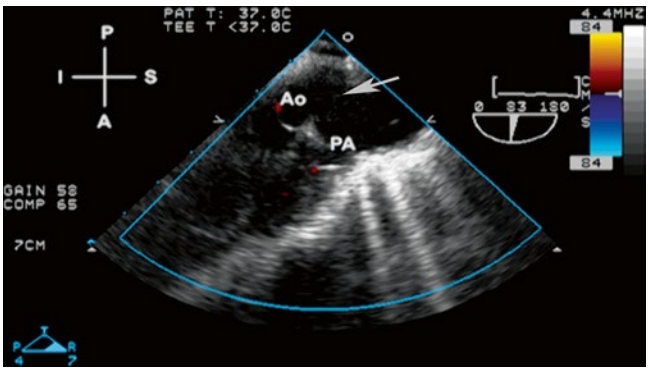

Fig. 10.14 Damus-Kaye-Stansel anastomosis as viewed from a mid esophageal ascending aortic long axis view, multiplane angle 80–90°. The widely patent anastomosis (shown by the *arrow*) lies just above the semilunar valves. *Ao* native aorta, *PA* native pulmonary artery

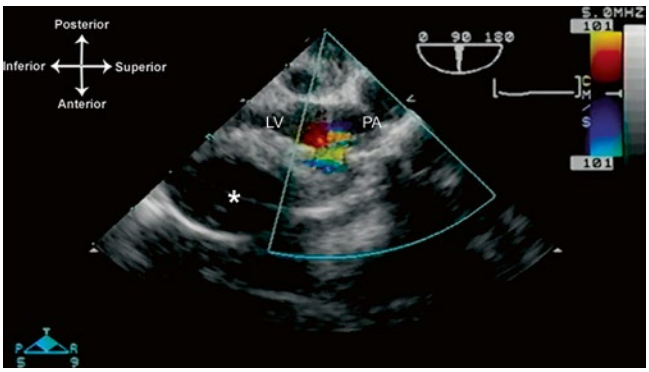

Fig. 10.15 Problematic Damus-Kaye-Stansel (DKS) in a patient with situs inversus, congenitally corrected transposition of the great arteries and hypoplastic right ventricle, as viewed from the equivalent of the mid esophageal aortic valve long axis view, multiplane angle about 90°. The creation of the DKS anastomosis resulted in distortion of the native pulmonary valve, producing significant regurgitation as shown by the color Doppler signal. The ventricular function was severely depressed. A large subpepicardial hematoma (*) occurred as the result of external cardiac massage. *LV* left ventricle, *PA* native pulmonary artery

"relocate" the aorta back to the larger ventricle [104, 105]. Control of pulmonary blood flow can be either from an aorto-pulmonary shunt (e.g. Blalock-Taussig or central shunt), or pulmonary artery band, as dictated by oxygen saturations. In some instances neither is required, as the degree of subpulmonary stenosis after the palliative arterial switch (formerly native subaortic stenosis) is adequate to provide enough pulmonary blood flow for acceptable systemic oxygenation, while at the same time maintaining low pulmonary arterial pressure.

Norwood Procedure

The Norwood procedure utilizes and extends the principle of the DKS procedure to those patients with significant hypoplasia of the ascending aorta and aortic arch. This procedure was initially developed for hypoplastic left heart syndrome patients [106], and has undergone a number of different modifications. The basic principle, however, remains as follows: the main pulmonary artery is disconnected

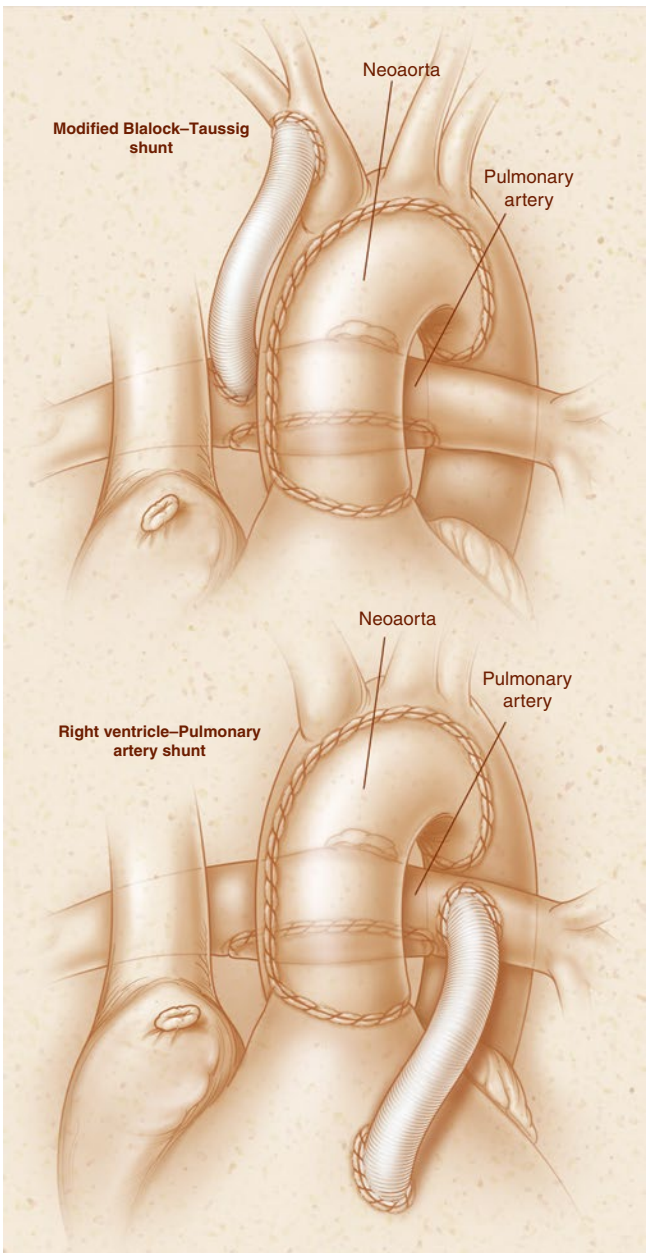

Fig. 10.16 Graphic of the Norwood procedure displaying modified Blalock-Taussig shunt in the *upper panel* and right ventricular to pulmonary artery (Sano) conduit in the *lower panel* (From: Ohye et al. [109]; Reprinted by permission, Massachusetts Medical Society)

from its branches and utilized to augment the hypoplastic ascending aorta and aortic arch, in essence creating a "neo-aorta". An atrial septectomy is performed, the disconnected branch pulmonary arteries are repaired, and pulmonary blood flow provided by either by a modified Blalock-Taussig shunt or a conduit placed between RV infundibulum and the main pulmonary artery (Sano modification) (Fig. 10.16) [107, 108]. Currently, institutions vary in their preference for the type of shunt (Blalock-Taussig vs. Sano modification). A recent randomized multicenter study comparing both shunt types showed better transplantation-free survival

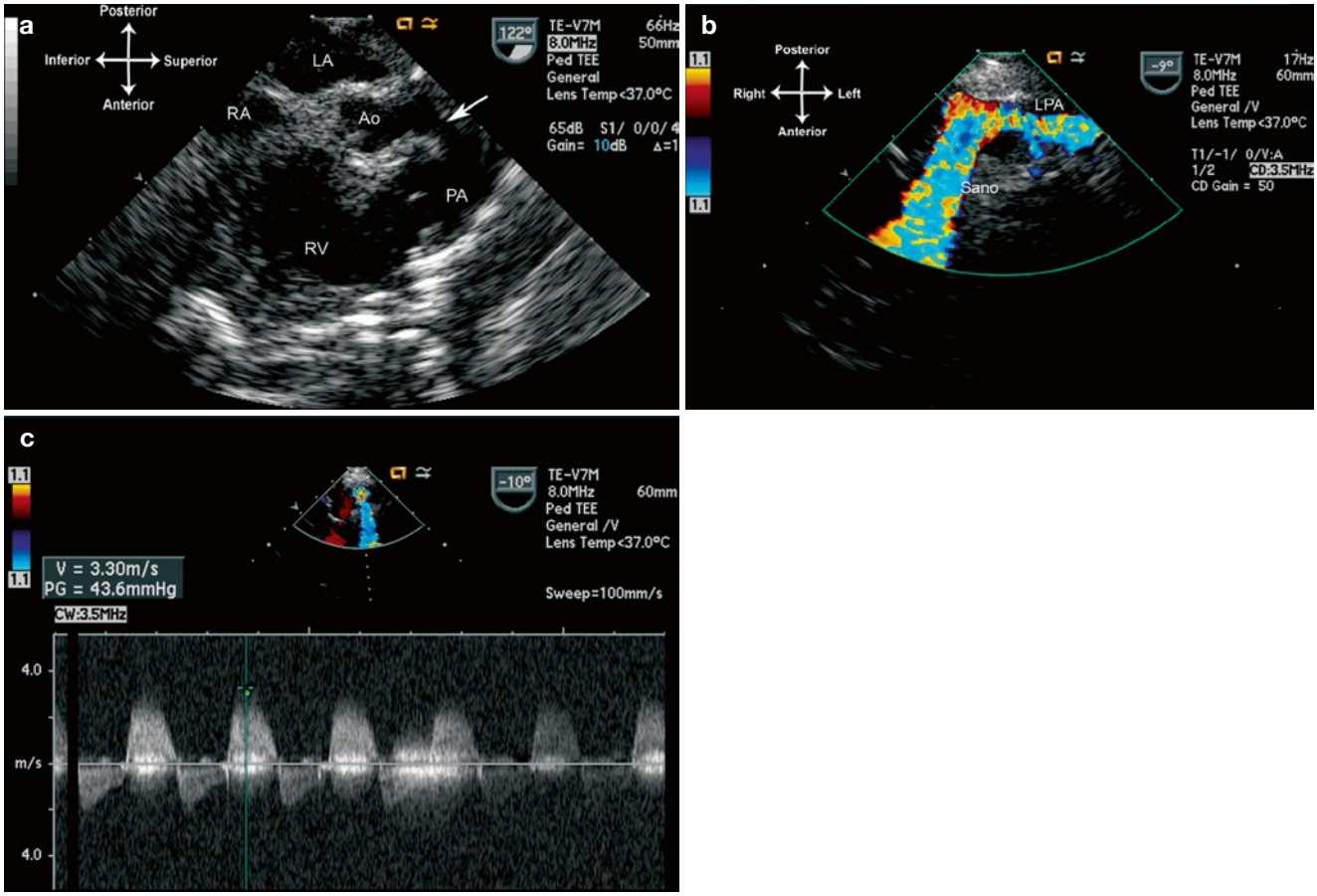

Fig. 10.17 Image of a patient with hypoplastic left heart syndrome variant following Norwood-Sano procedure. (**a**) The anastomosis (*arrow*) between a smaller native aorta (*Ao*) and native pulmonary artery (*PA*) from mid esophageal long axis view. (**b**, **c**) Images obtained from the upper esophageal pulmonary artery long axis view, demonstrating color Doppler aliasing in the distal Sano and to and fro flow by the spectral Doppler tracing of the Sano shunt. *Ao* native aorta, *LA* left atrium, *PA* native pulmonary artery (now neo-aorta), *RA* right atrium, *RV* right ventricle, *LPA* left pulmonary artery

for the Sano modification group within the first 12 months following surgery, but no significant difference between the two groups after that period [109].

When TEE is performed for the Norwood procedure, the preoperative examination serves to provide a baseline assessment of the anatomy, as well as an evaluation of tricuspid valve competency and ventricular systolic function. The postoperative TEE focuses on the following: determination of atrial communication adequacy from ME 4 Ch and ME Bicaval views, reevaluation of tricuspid and "neo-aortic" valves using ME 4 Ch and ME LAX views using both imaging and color flow Doppler, and assessment of ventricular systolic function in the ME 4 Ch, ME 2 Ch, and transgastric views. The neo-aortic root, as well as the anastomosis of the native ascending aorta to the neo-aorta, are best visualized with a ME LAX or ME AV LAX view (Fig. 10.17a, Video 10.14). The reconstructed transverse aortic arch and descending aorta can be visualized by withdrawal of the probe from the mid to the upper esophageal position (ME Asc Ao LAX and SAX, UE Ao Arch LAX, and UE Ao Arch SAX) and concomitant leftward rotation. This can be done with multi-plane angles of 0° or 100°. It is rare to be able to visualize the entire arch in a single plane. Instead, the complete arch can be visualized in sections, with gradual leftward rotation of the TEE probe. However the evaluation of potential residual aortic arch gradients may or may not be feasible from these views, due to suboptimal angle of Doppler interrogation. The Blalock Taussig/central shunt can be visualized in the manner described above. If an RV to pulmonary artery conduit (Sano modification) was placed, this is best seen from the UE PA LAX or UE Ao Arch SAX positions, either 0° or 90°, with prominent anteflexion of the TEE probe (Fig. 10.17b, Video 10.15). Continuous wave Doppler can be used to interrogate the conduit (Fig. 10.17c). However its orientation can produce unfavorable Doppler angles of interrogation and sometimes an incomplete spectral signal (Fig. 10.17c).

A more recent alternative to the Norwood procedure, known as the "Hybrid procedure", has received increasing attention [110–112]. The name of this procedure derives from an approach that combines a catheter-based intervention with surgery. This procedure involves catheter-based stent placement across the ductus arteriosus in order to maintain

ductal patency and systemic output, and surgical bilateral pulmonary artery banding to limit pulmonary blood flow. A balloon atrial septostomy (and/or stenting of the atrial communication) is performed either at the same time or deferred for a few days. When a hybrid Norwood procedure is performed, definitive aortic arch reconstruction is delayed until a later time in infancy (usually 4–6 months later) at which time removal of the ductal stent, pulmonary artery debanding and possible pulmonary arterioplasties are performed. This operation constitutes the equivalent to the standard second stage (Stage II) of single ventricle palliation, and a bidirectional Glenn anastomosis (discussed below) is also done at that time. In most institutions, TEE is not performed during a Hybrid procedure. However, if a TEE is considered necessary, the evaluation should focus on the specific questions and relevant aspects of the anatomy, applying similar principles as for the standard Norwood procedure.

In neonates with HLHS and intact or a severely restrictive atrial communication, radiofrequency perforation of the atrial septum, followed by balloon dilation and possible stenting of the interatrial communication, can be guided by TEE imaging in combination with fluoroscopy.

Bidirectional Cavopulmonary (Glenn) Anastomosis

The bidirectional cavopulmonary anastomosis, also known as the bidirectional Glenn procedure, represents an intermediate stage (sometimes called "Stage II") for single ventricle palliation prior to the Fontan completion. It is generally performed about 6 months of age, though the exact timing can vary; some centers report successful results with an early bidirectional Glenn (2–4 months) [113]. The original or classic Glenn procedure involved an end to end anastomosis between upper SVC and right pulmonary artery (RPA) after transection of the lower SVC and division of the more proximal RPA, thus the SVC provided flow only to the RPA. In contrast, the bidirectional Glenn anastomosis connects the SVC to the RPA but no transection of the RPA is performed, allowing SVC blood to flow directly into both pulmonary arteries. When bilateral SVCs are present, each is anastomosed to its respective branch pulmonary artery, and the procedure is then known as a *bilateral* bidirectional Glenn anastomosis [59]. An alternative method of directing SVC blood to the right pulmonary artery is known as the hemi-Fontan procedure, in which the SVC blood is redirected using a portion of right atrium into the right pulmonary artery, and a barrier is created at the SVC/right atrial junction using an imperforate disc. This procedure was created to facilitate the completion of the lateral tunnel Fontan procedure.

The bidirectional Glenn can also be used as part of the "one and a half" repair, described in the section for hypoplastic right heart syndrome. The one and a half repair is used when there are two ventricles, but the pulmonary ventricle—generally the morphologic RV—is judged incapable of carrying a full cardiac output. Usually, this is due to RV hypoplasia or a tricuspid valve abnormality. The one and a half procedure retains IVC blood return to the RV, from which it is ejected into the pulmonary artery; however SVC blood is channeled directly to the pulmonary arteries via a bidirectional Glenn anastomosis. Hence, the volume load on the RV is reduced by 25–50 % [13]. In some patients, a small atrial communication is left in place and can be closed (via transcatheter approach) at a later time [114, 115]. The one and a half repair is pertinent not just for hypoplastic right syndrome, but for a number of other forms of CHD in which partial bypassing of the RV might be useful, such as Ebstein's anomaly of the tricuspid valve, straddling tricuspid valve, and hypoplastic tricuspid valve associated with other forms of CHD [115–117]. In addition, it has been described in association with the anatomic repair of congenitally corrected transposition of the great arteries, in which an inferior hemi-Mustard repair is performed in conjunction with a bidirectional Glenn procedure and arterial switch or Rastelli operation (discussed in Chap. 12) [118]. Whatever the indication, when contemplating a one and a half repair, the same physiologic considerations apply as for any prospective bidirectional Glenn or Fontan patient—namely, there must be low pulmonary artery pressure and pulmonary vascular resistance, low systemic ventricular end diastolic pressure, good systemic ventricular function, and low probability of post-Glenn pulmonary hypertension due to factors such extra pulmonary blood flow from multiple VSDs [117].

Another important variation of the bidirectional Glenn operation worth mentioning is the Kawashima procedure. This procedure is generally performed in the setting of left isomerism (polysplenia), when there is interruption of the hepatic portion of IVC and azygos or hemiazygos continuation to the right or left SVC. In the Kawashima procedure, a bidirectional Glenn (or bilateral bidirectional Glenn) is performed, but the azygos or hemiazygos is not ligated allowing systemic venous blood from the lower body to return to the pulmonary arteries [119]. This nearly creates a complete Fontan repair, with only hepatic venous and coronary sinus blood continuing to drain into the heart. It is estimated that, after a Kawashima procedure, 80–85 % of total systemic venous blood returns directly to the lungs [120]. In general, the Kawashima procedure is performed within 6–12 months of life. While it would seem that patients might be too young for the near-Fontan physiology that results at such a young age, in fact most institutions report excellent outcomes following the Kawashima procedure [120]. However this group of patients is notoriously prone to the development of pulmonary arteriovenous malformations, felt to be due to the lack of hepatic venous blood (containing an as yet unidentified "hepatic factor") returning directly into the pulmonary arteries [121–123]. Largely because of this complication, completion of the Fontan circuit is now generally performed at several years of age, with the hepatic veins redirected into the

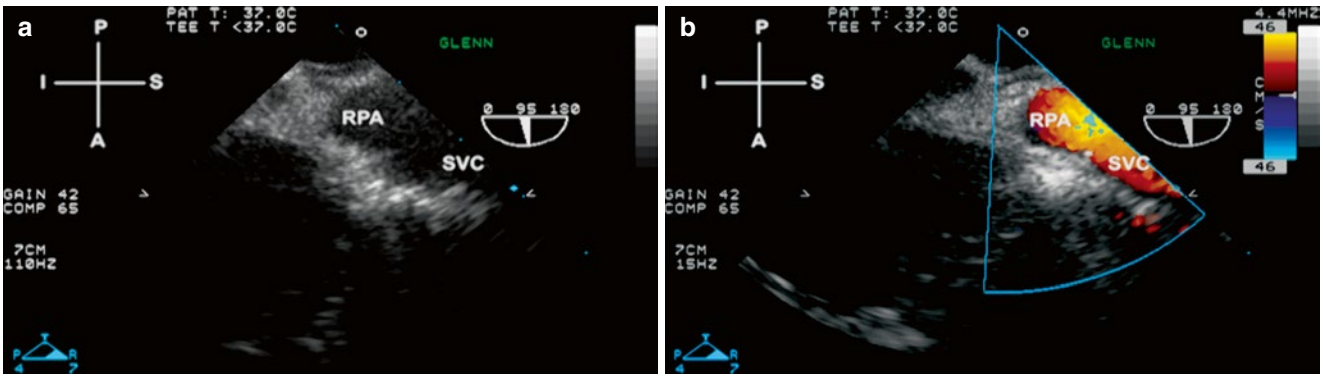

Fig. 10.18 Right bidirectional cavopulmonary (Glenn) anastomosis as viewed from the mid to upper esophageal position, multiplane angle 95°. A mid esophageal ascending aortic long axis view is first obtained, then the probe is turned to the right and slightly withdrawn. The anastomosis is well seen by two-dimensional imaging (**a**), and color flow Doppler (**b**). Potential narrowing at the anastomotic region and/or Doppler flow acceleration can be seen in this view. *RPA* right pulmonary artery, *SVC* superior vena cava

pulmonary artery in a manner similar to standard Fontan completion [120, 124]. This generally results in normal systemic oxygen saturations, likely due to (a) elimination of the hepatic venous right to left shunt into the atrium; (b) return of "hepatic factor" directly into the pulmonary arteries, leading to resolution of the arteriovenous malformations [125].

There is limited utility for TEE in patients with bidirectional Glenn surgery. Given the variability in pulmonary artery visualization by TEE, the Glenn anastomosis cannot always be optimally seen. Withdrawal to a mid to upper esophageal position (ME Asc Ao LAX, UE Ao arch SAX), along with rightward rotation of the TEE probe (in the case of a right sided Glenn) and slight anteflexion while using a multiplane angle of 90° will best show this connection. Color flow and spectral Doppler demonstrate the low velocity, undulating nature of the venous return into the pulmonary arteries (Fig. 10.18, Video 10.16). Any narrowing of the Glenn anastomosis will result in a narrowed appearance and turbulence at the site.

Fontan Procedure

The Fontan procedure was initially described for patients with tricuspid valve atresia [58]. Since then, it has undergone a significant number of changes and modifications, but the underlying, basic concept of Fontan physiology remains the same—diversion of systemic venous blood directly to the pulmonary arteries without the aid of an RV pumping chamber, thereby separating the systemic and pulmonary circulations. The Fontan procedure has become the cornerstone and ultimate goal in the management and surgical planning of any patient with single ventricle physiology, irrespective of ventricular anatomy. In this section, the most common variations of the Fontan are discussed, along with the applications of TEE for its evaluation.

The initial description and early modifications of the Fontan procedure were characterized by one important constant: the anatomic right atrium remained an integral part of the Fontan circuit. The atrial septum was closed, and all right atrial blood was directed in some manner to the pulmonary artery (the right atrium was connected either directly to the pulmonary artery or to the RV outflow tract) [126–128]. This was known as an *atriopulmonary connection* (Fig. 10.19a), or in the case of connection to the RV outflow tract, and *atrioventricular connection* [63]. Over time, problems arose in these patients due to the development of right atrial dilation, atrial arrhythmias, and sinus node dysfunction as well as systemic ventricular dysfunction. Even though it is no longer performed, it should be recognized that there continue to be a large a number of surviving adult CHD patients who have undergone this type of Fontan procedure. Some of these patients have required, or will require, surgical revision to a more contemporary Fontan modification, to allow for a more favorable hemodynamic result [129–132].

To improve upon the energetics of flow in the Fontan circuit, the concept of the *total cavopulmonary connection* was subsequently conceived [133, 134]. In this operation, an intra-atrial tunnel (lateral tunnel Fontan) is constructed directing flow from the inferior vena cava through the inferior portion of the SVC that is connected direct to the RPA, thereby channeling IVC blood into the pulmonary arteries. With the lateral tunnel Fontan, part of the patient's right atrial lateral wall serves as a portion of the tunnel (Fig. 10.19b). An alternate option more recently favored by many institutions consists of placement of an external conduit allowing for flow between the IVC and pulmonary arteries, but avoiding suture lines in the right atrium (a potential source of arrhythmias); this is known as the *extracardiac* Fontan [135, 136] (Fig. 10.19c).

An important added variation to the modified Fontan procedure involves placement of a small fenestration, usually between the Fontan pathway and the anatomic right atrium [137]. The original concept was to improve the immediate postoperative course in high risk Fontan patients by providing a controlled right to left shunt that helps to preserve systemic cardiac output in the presence of factors that may have resulted in elevated pulmonary vascular resistance. This

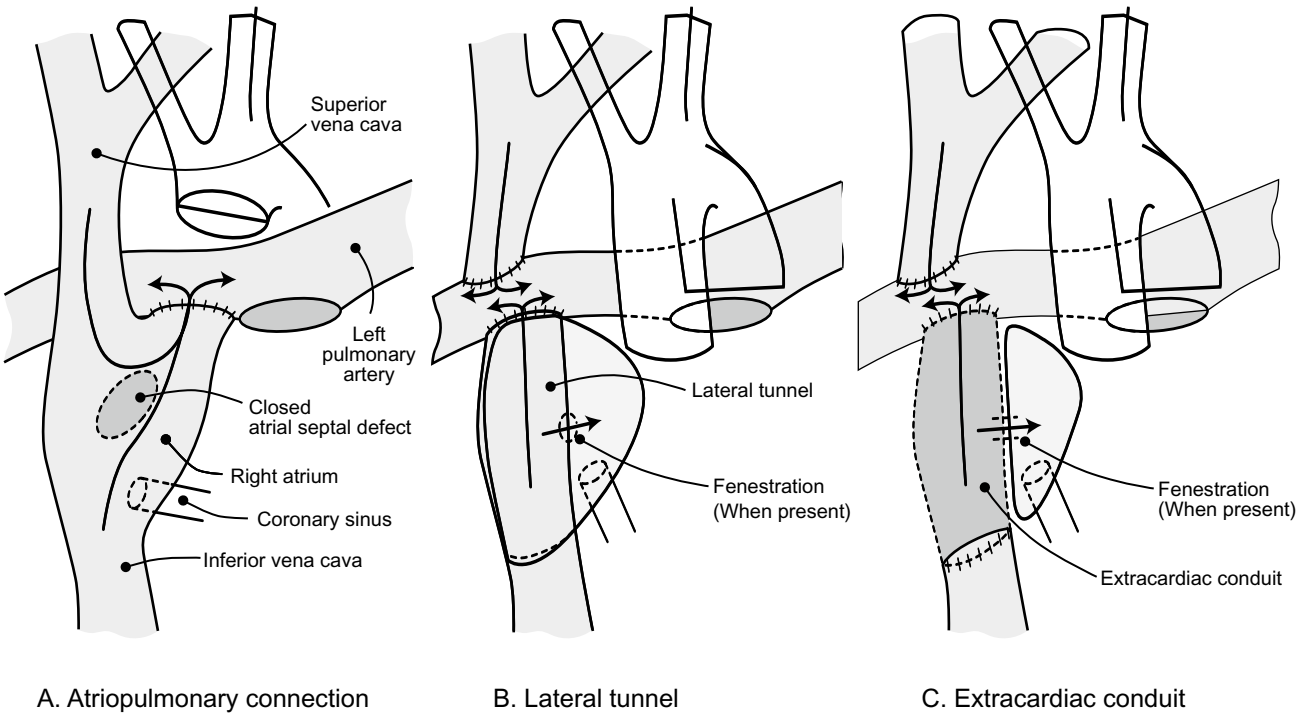

Fig. 10.19 The different types of Fontan circulation (Diagram modified from de Leval [67]. Permission obtained from Nature Publishing Group. Adapted by permission from Macmillan Publishers Ltd: copyright 2005)

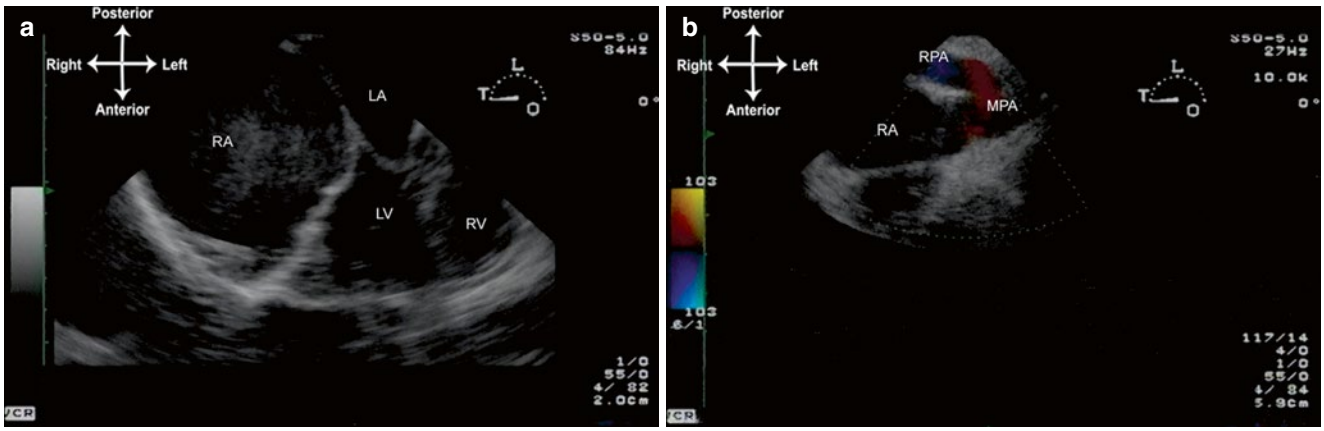

Fig. 10.20 Atriopulmonary Fontan connection, as seen from the mid esophageal four chamber view in a patient with double inlet left ventricle and L-malposed great arteries. The right atrioventricular valve was surgically obliterated as part of the original Fontan procedure. (**a**) The severely dilated right atrium with swirling of flow and spontaneous echo contrast seen. (**b**) The atriopulmonary connection by color Doppler. *LA* left atrium, *LV* left ventricle, *MPA* main pulmonary artery, *RA* right atrium, *RPA* right pulmonary artery, *RV* right ventricle

application was subsequently expanded to all Fontan patients at a number of CHD centers. Over time, the fenestration may close spontaneously, or patients can undergo catheter closure of the fenestration if it is no longer considered necessary [138]. Significant debate remains regarding the routine need for Fontan fenestration, patient selection, and eventual management of a patent fenestration (whether to leave or occlude) [62, 139–141].

The TEE evaluation of the Fontan patient varies based upon the procedure performed. In the case of the atriopulmonary/atrioventricular Fontan connection, the systemic venous return to the right atrium can be visualized sagittally using the ME Bicaval view, or in cross section with probe withdrawal from LE Situs SAX to ME 4 Ch views along with slight anteflexion (multiplane angle 0°). In general, such patients will have a distended atrium, and sluggish blood flow often creates the appearance of "smoke" (spontaneous echo contrast due to red cell aggregation) in the atrium [142, 143] (Fig. 10.20a, Video 10.17). In the ME 4 Ch view (multiplane angle 0–30°), the atrial septum is well-seen; it usually

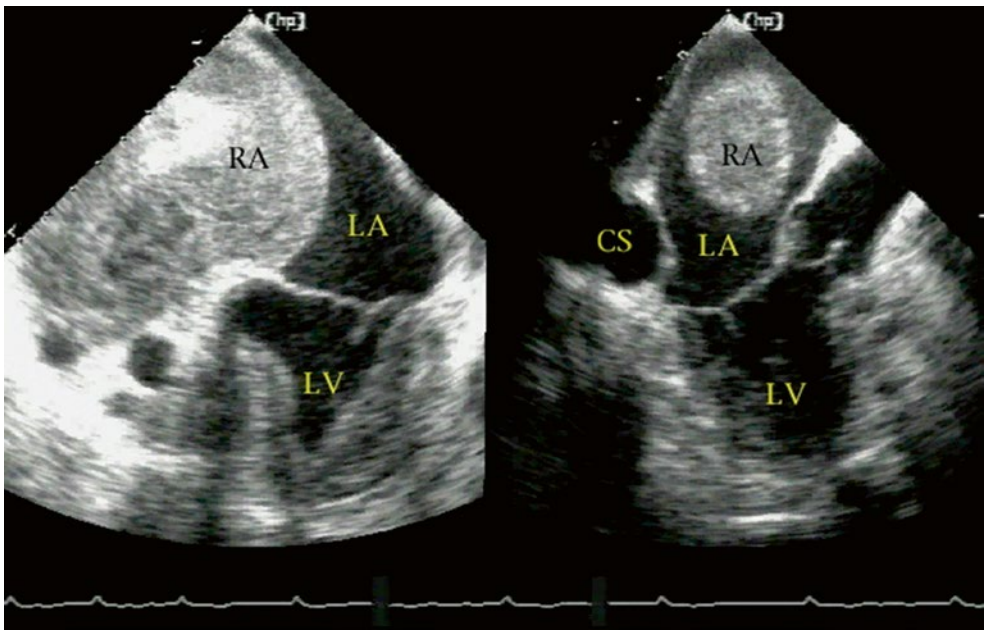

Fig. 10.21 Atriopulmonary Fontan in a patient with tricuspid atresia, showing a severely dilated right atrium completely filled with thrombus, in orthogonal views. Note the dilated coronary sinus (*CS*) due to elevated right atrial pressure resulting from the thrombus. *LA* left atrium, *LV* left ventricle, *RA* right atrium

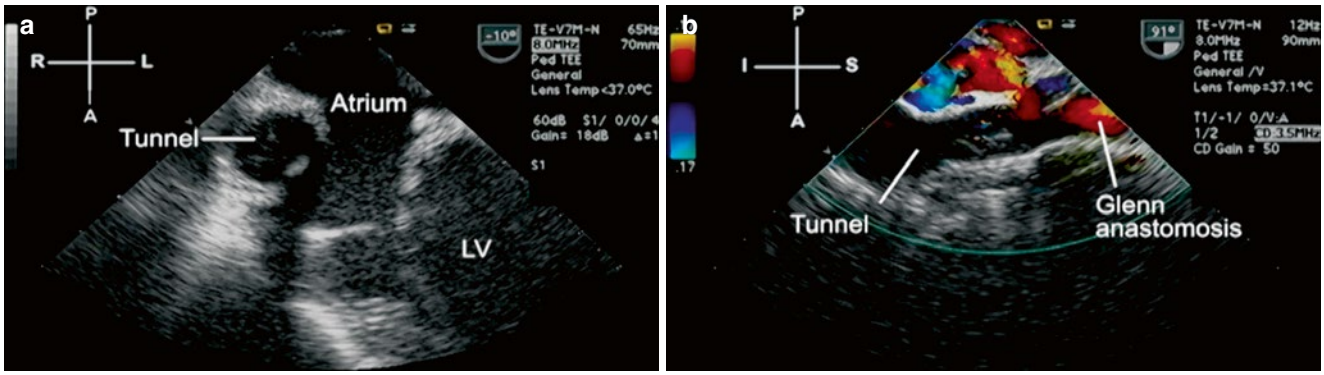

Fig. 10.22 Lateral tunnel Fontan in a patient with double inlet left ventricle, as seen from the mid esophageal views. (**a**) The tunnel in cross-section in the mid esophageal four chamber view. (**b**) The length of the tunnel and the Glenn anastomosis shown more superiorly by color flow Doppler. This view is the equivalent of the mid esophageal bicaval view, with the probe withdrawn slightly to visualize the tunnel and Glenn anastomosis. *LV* left ventricle

bows toward the left, and can be inspected for any residual right to left leaks by color flow Doppler. Depending on the type and location of the atriopulmonary/atrioventricular connection, the mid esophageal views (ME 4 Ch, ME RV In-Out, ME AV SAX, ME Asc Ao SAX) can be used to evaluate patency of the connection (Fig. 10.20b). The branch pulmonary arteries can be visualized in the manner described previously. Flow into the branch pulmonary arteries will be low velocity and phasic in nature. Atrioventricular valve and semilunar valve function, and ventricular systolic function, should be examined carefully. Intra-atrial thrombi can sometimes result from the sluggish blood flow in the right atrium, and these can be seen as irregular echogenic structures. In some cases, there can be near complete thrombosis of the right atrium (Fig. 10.21, Video 10.18)

For the total cavopulmonary connection (modified Fontan procedure), IVC and hepatic drainage should be inspected, and with either the lateral tunnel or extracardiac conduit, the anastomosis of the Fontan pathway to the IVC should be visualized. This is best initiated from the LE Situs SAX view, multiplane angle 0°, and often some probe anteflexion and rightward rotation is necessary (Fig. 10.22a, Video 10.19). The probe is withdrawn to the ME 4 Ch view, and a cross-sectional "sweep" of the Fontan pathway is inspected for any possible narrowing or masses/thrombosis. With further withdrawal to a higher mid esophageal position (ME Asc Ao SAX), the entrance into the pulmonary artery can be seen. If the pulmonary arteries can be visualized, the flow into both branches should be evaluated. Also in the mid-esophageal position the multiplane angle can be rotated to 80–90° to a ME Bicaval view and the probe rotated rightward to reveal a sagittal view of the Fontan pathway, and to evaluate for possible narrowing (Fig. 10.22b, Video 10.19). When a fenestration has been placed, the location can be highly variable. Generally, a fenestration is best seen in the ME 4 Ch view with the transducer angle about 0°

(Fig. 10.23, Video 10.20). Color flow Doppler demonstrates the site of fenestration flow and right to left shunting, and a gradient can be estimated using spectral and/or continuous wave Doppler. Again, as with any Fontan modification, it is important to evaluate overall ventricular function, as well as AV and semilunar valve function. In the patient with an external conduit, the entrance of the pulmonary veins should also be examined for possible compression by the conduit (Fig. 10.24, Video 10.21). However it is also important to remember that temporary pulmonary vein stenosis can be created by compression from the TEE probe, and therefore TEE findings of pulmonary vein stenosis should be corroborated with postoperative imaging studies as well as surgical findings [144, 145].

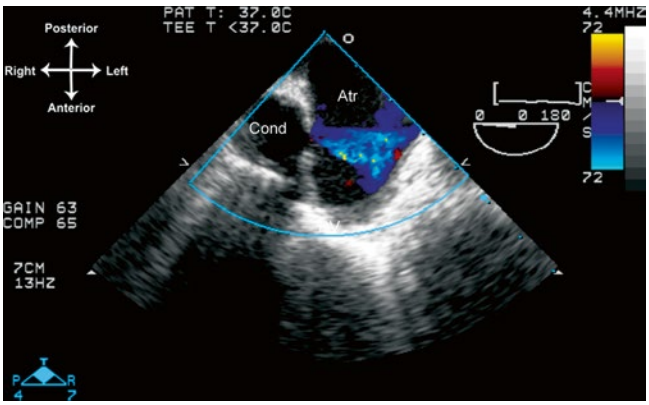

Fig. 10.23 Extracardiac Fontan with fenestration, as viewed from a mid esophageal four chamber view. The Fontan conduit (Cond) is located just outside and to the right of the physiologic "left" atrium, and seen in cross-section. Right to left shunting through the fenestration is clearly seen by color flow Doppler entering the atrium. *Atr* atrium

The use of TEE to evaluate Fontan patients has application in multiple settings [146]. In the operating room, preoperative TEE is used to confirm intracardiac anatomy and function (as noted above), and postoperatively to evaluate patency of the Fontan pathway, characterize flow across a fenestration if present, assess AV and semilunar valve function, and evaluate cardiac function. It should be noted that the extracardiac Fontan can be difficult to visualize immediately following surgery if a polytetrafluoroethylene (PTFE, or Gore-Tex) conduit is used, due to the shadowing and reverberation produced by the microscopic air contained in the walls of the conduit. Over time, the conduit becomes more readily visible. In the outpatient setting, TEE becomes

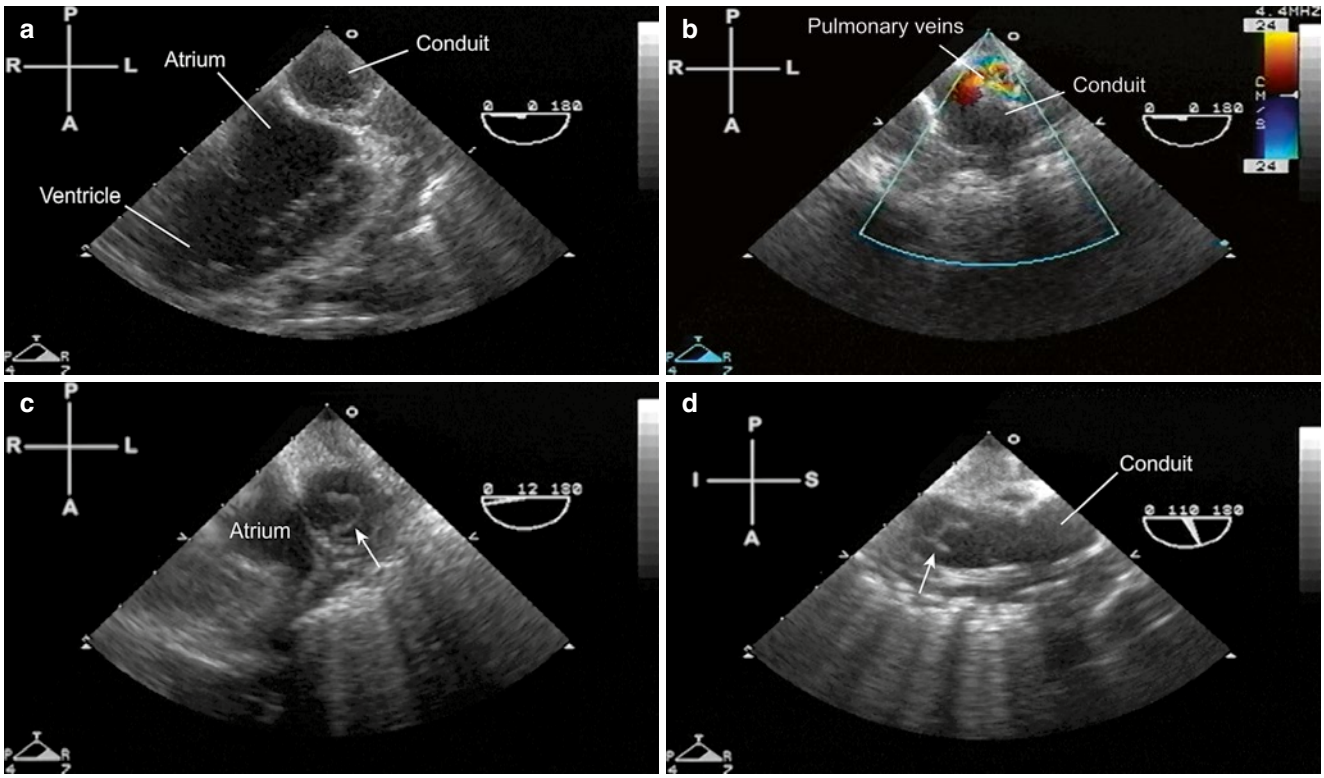

Fig. 10.24 Extracardiac Fontan conduit placed between a right sided IVC and the left pulmonary artery in a patient with heterotaxy and dextrocardia. (**a**) The equivalent of a mid esophageal four chamber view, with the probe rotated slightly rightward (clockwise). Dextrocardia is seen as well as the large conduit passing behind and slightly to the left of the atrium. As the conduit passes from right to left behind the heart, it compresses the pulmonary veins (**b**), causing pulmonary venous obstruction as shown by mosaic color flow next to the conduit. This resulted in a low cardiac output state that likely contributed to the development of a thrombus within the conduit, identified by the *arrow* (**c**). (**d**) The multiplane angle has been rotated to 110° in order to optimize visualization of the length of the conduit. The thrombus is seen toward the more inferior portion (*arrow*)

increasingly more useful in older, larger patients, in whom transthoracic echocardiographic imaging can be quite challenging. When atrial fibrillation is present, TEE is very useful for the assessment of possible thrombosis in the atrial appendage(s), analogous to its utility in the adult patient with atrial fibrillation [147]. Thus TEE can play an important role in ambulatory assessment of adults who have undergone a Fontan procedure. In the intensive care setting, TEE can be useful in those situations in which transthoracic imaging is compromised [148]. In addition, in patients undergoing transcatheter closure of a Fontan fenestration or creation of a fenestration in the patient with a failing Fontan circulation, TEE can be used for monitoring and procedural guidance.

Additional Evaluation

Excessive cyanosis following the bidirectional Glenn, hemi-Fontan, or non fenestrated Fontan procedures raises the possibility of a right to left shunt somewhere in the circulation. In such cases, the underlying cardiac anatomy and repair should be reviewed, and the TEE evaluation performed accordingly, for potential intracardiac shunts. For the atriopulmonary, atrioventricular, and lateral tunnel Fontan procedures, the possibility of a residual intracardiac shunt (across the intra-atrial baffle, atrial septum, or other structure) should be considered. Injection of agitated saline contrast into the systemic venous circuit can be helpful in revealing occult right to left shunts. The examiner should keep an open mind to all possibilities, because surprising findings are occasionally encountered [149]. In addition, excessive cyanosis following the Fontan procedure also raises the question of collateral vessels arising from the systemic venous system and returning directly to the heart (bypassing the lungs), or alternatively, pulmonary arteriovenous malformations. These possibilities can be evaluated during a TEE study by the use of an agitated saline contrast injection. Because agitated saline is filtered in the lungs, none or a minimal amount should return to the atrium [150]. For a possible venous collateral returning to the heart, agitated saline is injected into a systemic vein (preferably through a central line) at a site proximal to the potential site of the collateral. If contrast is seen returning directly to the atrium immediately after injection, the presence of a collateral should be strongly suspected. For pulmonary arteriovenous malformation, contrast will first be visualized in the pulmonary veins, and subsequently in the atrium. It should be noted that these techniques are useful but not fully diagnostic, and there can be false positive results [151]. Cardiac catheterization and angiography are often necessary to provide more definitive anatomic evaluation, particularly in the case of venous collaterals returning to the heart.

Summary

The terms "single ventricle" and "functional single ventricle" refer to a broad, heterogeneous group of congenital cardiac lesions differing markedly in terms of their anatomy and physiology. However despite their great diversity, all of these cardiac defects are distinguished by a simple, common, and unifying characteristic: a biventricular intracardiac repair cannot be performed. For these patients, the management is relatively straightforward: the ultimate goal is to separate the systemic and pulmonary circulations such that deoxygenated systemic venous blood returns passively to the pulmonary arteries without the assistance of a separate pump. Optimizing such patients for this unique physiology—known as Fontan physiology—forms the basis of all current management strategies for the single ventricle patient. This is a very important group of patients, and their diagnosis, evaluation, and management represents one of the most active and topical areas in contemporary pediatric cardiovascular medicine. Moreover, due to the great advances and successes with medical and surgical therapies, particularly with the Fontan procedure, a significant proportion of single ventricle patients are now reaching adulthood. Thus, this group constitutes an ever-growing and increasingly important proportion of the patients seen by adult CHD specialists. In this chapter we discussed the major types of cardiac defects producing single ventricle physiology, as well as their surgical management. For such patients, TEE is very useful both in terms of preoperative diagnosis and intraoperative assessment; moreover it can be helpful as well in a number of other clinical scenarios.

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