Evaluation of Conotruncal Abnormalities

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Abstract

Conotruncal malformations encompass a group of congenital heart defects with abnormal ventriculo-arterial connections. The conotruncal abnormalities are often seen in association with other cardiac defects, and the intracardiac anatomy can be quite complex. Cardiac surgery is almost always required to repair the anatomic abnormalities and restore normal physiology. As such, transesophageal echocardiography (TEE) is often required in the intraoperative setting in order to evaluate and confirm preoperatively the cardiac abnormalities, and to assess the cardiac repair postoperatively. Moreover, patients with a conotruncal malformation (operated and sometimes unoperated) will survive into adulthood, and in the ambulatory setting, TEE can provide important anatomic and physiologic information superior to that available by transthoracic echocardiography. This chapter discusses the TEE evaluation of the most common and important conotruncal defects, including tetralogy of Fallot, double outlet right ventricle, truncus arteriosus, transposition of the great arteries (also known as complete, or D-transposition of the great arteries), and congenitally corrected transposition of the great arteries.

Keywords

Transesophageal echocardiography • Heart defects • Congenital • Tetralogy of Fallot • Double outlet right ventricle • Transposition of the great arteries, complete • Truncus arteriosus • Transposition of the great arteries, corrected

Introduction

Conotruncal malformations encompass a group of defects with abnormal ventriculo-arterial connections. Cardiac surgery is almost always required for conotruncal abnormalities and therefore transesophageal echocardiography (TEE) is essential in the evaluation of most patients with these defects.

This chapter will review the application of TEE in the evaluation of conotruncal anomalies including: tetralogy of Fallot, double outlet right ventricle, truncus arteriosus, transposition of the great arteries (also known as complete or D-transposition of the great arteries), and congenitally corrected transposition of the great arteries. These cardiac defects comprise the vast majority of conotruncal abnormalities encountered by congenital heart disease specialists; extremely rare conditions including anatomically corrected malposition of the great arteries and double outlet left ventricle will not be addressed.

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The online version of this chapter (doi:10.1007/978-1-84800-064-3_12) contains supplementary material, which is available to authorized users.

P.C. Wong, W.C. Miller-Hance (eds.), *Transesophageal Echocardiography for Congenital Heart Disease*,
DOI 10.1007/978-1-84800-064-3_12,

General Comments

TEE is an important diagnostic tool in patients diagnosed with conotruncal abnormalities in a variety of settings. Conotruncal malformations are usually diagnosed prenatally, or in the neonatal period, therefore diagnostic echocardiography is generally performed using transthoracic echocardiography (TTE). TEE becomes a powerful tool in the intraoperative setting. Preoperative TEE often provides additional anatomic information that allows the echocardiographer to determine the best views for imaging after surgical intervention; this is particularly important when complex baffling procedures are being considered. Postoperative TEE is an invaluable tool used to assess the adequacy of surgical repair for many of these lesions. This includes evaluation of: residual intracardiac shunts, residual outflow obstruction, evaluation of atrioventricular (AV) and semilunar valve function, and assessment of ventricular performance. TEE is also essential in the evaluation of patients (particularly adults) with poor transthoracic acoustic windows. Moreover, TEE is largely used in the evaluation of infective endocarditis, suspicion for intracardiac thrombus, and suspected aortic dissection [1], as discussed in Chap. 16. TEE is also helpful to assist in catheter-directed interventions (Chap. 17).

Specific Lesions

In this section, we discuss the anatomy, echocardiographic evaluation, and surgical considerations for: tetralogy of Fallot, double outlet right ventricle, truncus arteriosus, transposition of the great arteries, and congenitally corrected transposition of the great arteries.

Tetralogy of Fallot

Anatomy

The four morphologic abnormalities that constitute the term "tetralogy" of Fallot (TOF) include a ventricular septal defect (VSD), override of the aorta over the ventricular septum, subpulmonary stenosis, and right ventricle (RV) hypertrophy. Yet the fundamental anatomic defect in TOF is anterior malalignment of the infundibular (conal) septum. This malalignment results in a large, typically unrestrictive VSD with concurrent override of the aorta over the ventricular septum. Furthermore, the infundibulum (conal muscle under the pulmonary valve) is hypoplastic with variable degrees of subpulmonary and pulmonary stenosis. RV hypertrophy is a consequence of the large VSD (the RV being at systemic pressure) [2].

TOF displays wide anatomic variation: from mild pulmonary stenosis to the most severe form, pulmonary atresia. In TOF with pulmonary atresia (TOF/PA), the anatomy of the pulmonary arteries has a wide spectrum: from normal-sized, confluent pulmonary arteries to diminutive or even nonexistent pulmonary arteries with pulmonary blood flow supplied by way of aorto–pulmonary collateral vessels.

Known genetic abnormalities occur in 20 % of patients with TOF. 22q11.2 deletion syndrome (including the DiGeorge and velo-cardio-facial syndromes) is seen in 15 % of patients with TOF and is particularly common in those with associated arch sidedness abnormalities. TOF is the third most common lesion in children with Down syndrome (trisomy 21) and is sometimes seen in association with endocardial cushion defect in those patients. Other conditions associated with TOF include VACTERL association and CHARGE syndrome. Patients with TOF and chromosomal anomalies have worse outcome than those without genetic defects [3].

With the most common form of TOF (malalignment VSD and subvalvar/valvar pulmonic stenosis), surgery is usually performed within the first 6 months of life. Some patients with only a mild to moderate amount of pulmonary outflow tract obstruction will have relatively balanced shunting across the VSD, with only a mild degree of cyanosis, and acceptable oxygen saturations that can sometimes be higher than 90 % (so-called "pink TOF"). In this group, complete, one-stage surgical repair can be electively performed at any time, usually 5–6 months, sometimes even later. In other patients, surgical intervention is necessary at a younger age; indications for this include significant cyanosis and/or "*tetralogy spells*" (acute, severe hypercyanosis). For such patients, many institutions utilize a staged surgical approach. Surgical palliation is first performed to augment pulmonary blood flow, usually with a modified Blalock-Taussig shunt. When pulmonary stenosis is primarily at pulmonary valve level, a few institutions will utilize balloon dilation of the pulmonary valve (performed in the catheterization laboratory) as an alternative form of palliation to increase pulmonary blood flow [4–6]. In either case, complete repair is undertaken several months later. However some centers prefer to perform a complete, one-stage repair of TOF at a very young age, even as a neonate [7–9].

The surgical management of patients with TOF/PA is variable. Most will require some type of surgical intervention, and the timing of surgery can vary from the neonatal period to infancy or even older. The surgical intervention is dependent on the anatomical subtype. Some patients are staged to complete repair: a modified Blalock-Taussig shunt or other type of aorto-pulmonary connection is performed in the neonatal period, followed by a complete repair (often with an RV to pulmonary artery conduit) within the first year of life. In some cases, initial palliation to establish reliable pulmonary blood flow can be performed without surgery. This is achieved by interventional catheterization methods

such as stenting of the ductus arteriosus [10–12] and, when membranous pulmonary valve atresia is present, radiofrequency perforation of a pulmonary valve [13]. In more severe cases such as TOF/PA with multiple aorto-pulmonary collateral arteries (MAPCAS), complex staged surgical palliation is required including *unifocalization* of the pulmonary arteries and collateral vessels to a central location with a reliable source of pulmonary blood flow (Blalock-Taussig shunt or RV to pulmonary artery conduit). It should also be noted that some patients with TOF/PA and MAPCAS do not undergo any intervention throughout their lifetime. They continue to have lower than normal arterial oxygen saturations, but these remain compatible with survival because the aorto-pulmonary collaterals provide enough pulmonary blood flow to maintain adequate systemic arterial oxygenation.

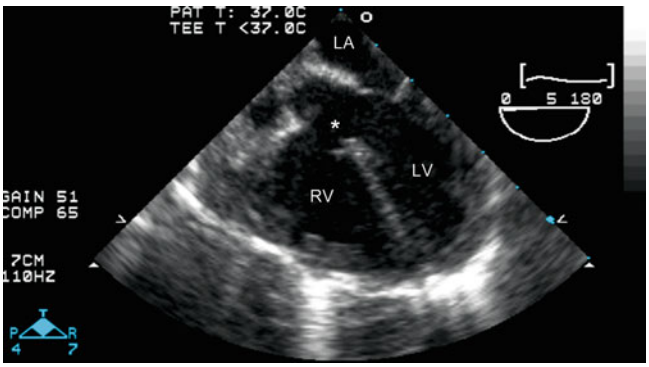


Fig. 12.1 Tetralogy of Fallot. Mid esophageal four chamber view showing right ventricular hypertrophy, the large malalignment ventricular septal defect (*asterisk*), and overriding aorta. *LA* left atrium, *LV* left ventricle, *RV* right ventricle

TEE Evaluation

There are pertinent findings in TOF to keep in mind when performing a TEE:

- Additional VSDs, which are typically seen in the muscular septum. Some patients may have absence or hypoplasia of the conal septum in addition to malalignment.
- Restrictive VSD due to accessory tricuspid valve tissue
- Coronary anomalies, particularly a coronary artery crossing the RV outflow tract, which occur in approximately 5 % of TOF patients. This includes variations such as origin of the left anterior descending coronary artery from the right coronary artery, and right coronary artery from the left anterior descending [14, 15].
- Persistent left superior vena cava (SVC), seen in 9–12 % of patients [16, 17]. In these cases the left SVC usually drains into a dilated coronary sinus, but in rare instances it can return directly to the left atrium.
- Discontinuous pulmonary arteries, whereby one pulmonary artery originates from a ductus arteriosus or collateral vessel.
- Common AV canal defect (complete AV septal defect), seen almost exclusively in Down syndrome.
- Absent pulmonary valve syndrome, A subtype of TOF, the main features of this condition include an unguarded pulmonary valve annulus with massively dilated main and proximal branch pulmonary arteries. Free pulmonary insufficiency is the norm, as well as compression of the right and left mainstem bronchi and/or as distal bronchioles.
- Atrial septal defect or patent foramen ovale (the so-called pentalogy of Fallot).

Though the diagnosis of TOF is generally made using TTE, TEE can be used for confirmation of diagnosis or in those patients with poor transthoracic acoustic windows. This includes the rare adult with unrepaired TOF. The starting point is generally the mid esophageal four chamber (ME 4 Ch) view which displays the four cardiac chambers and two AV valves along with the large malalignment VSD. This view also confirms RV hypertrophy. In addition, the atrial septum can be inspected for any defects. As the probe is advanced, the coronary sinus is viewed; it can be enlarged when it is connected to a left SVC. As the probe is withdrawn, the VSD is again seen with the aorta overriding the ventricular septum (Fig. 12.1, Video 12.1). Further withdrawal demonstrates the hypoplastic pulmonary outflow tract. The atrial septum is interrogated for a communication from inferior to superior in the 0–30° ME 4 Ch view. The mid esophageal bicaval (ME Bicaval) 90° view establishes the systemic venous connections (in a sagittal plane) and confirms the presence of an atrial communication, if present. As the probe sweeps to the left (counterclockwise), the VSD is seen with the anterior malalignment of the conal septum, in a view known as the mid esophageal right ventricular inflow-outflow (ME RV In-Out) view, multiplane angle 60°–90°. (Fig. 12.2, Video 12.2). From here, forward rotation of the multiplane angle setting to 90°–110° produces the mid esophageal long axis (ME LAX) and mid esophageal aortic valve long axis (ME AV LAX) views. Both of these views can also be used to evaluate the relationship of the overriding aorta to the VSD. If the TEE probe can be advanced into the stomach and anteflexed, the deep transgastric long axis (DTG LAX) and deep transgastric sagittal (DTG Sagittal) views provide excellent visualization of the overriding aorta, malalignment VSD, anteriorly malaligned infundibular septum, and narrowed RV outflow tract (Fig. 12.3, Video 12.3). These views are similar to those obtained with the standard transthoracic subcostal coronal and sagittal sweeps.

Color Doppler evaluation prior to surgical intervention includes assessment of flow across the atrial septum (typical flow pattern is from the left atrium to the right atrium). The ventricular septum can be interrogated in all views to assess for additional ventricular communications in the muscular septum. Spectral Doppler assessment of the gradient across the RV outflow tract is best performed in the mid esophageal

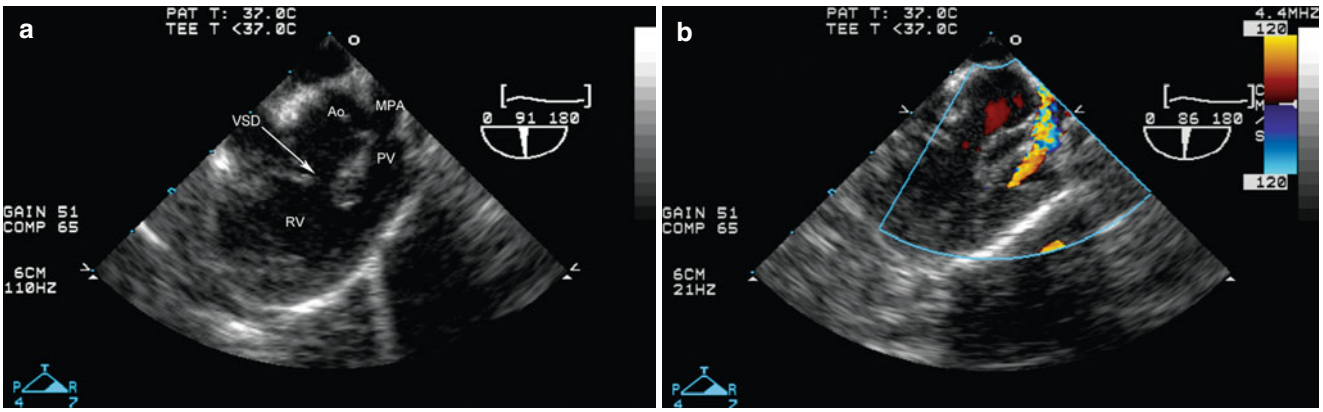


Fig. 12.2 Tetralogy of Fallot. (**a**) Modified mid esophageal right ventricular inflow-outflow view (multiplane angle about 90°) showing the malalignment ventricular septal defect (*VSD*), as well as narrowing of the right ventricular outflow due to a malaligned conal septum. (**b**) Color flow Doppler shows aliasing beginning at subpulmonary level, and continuing across the hypoplastic pulmonary valve. *Ao* aorta, *MPA* main pulmonary artery, *PV* pulmonary valve, *RV* right ventricle

ascending aortic short axis (ME Asc Ao SAX), ME RV In-Out or DTG LAX and Sagittal views—whichever yields a more favorable angle for Doppler evaluation (Figs. 12.2 and 12.3, Videos 12.2 and 12.3). To evaluate flow in the branch pulmonary arteries, and to search for a possible patent ductus arteriosus, color Doppler can be performed using the upper esophageal pulmonary artery long axis (UE PA LAX) and upper esophageal aortic arch short and long axis (UE Ao Arch SAX, UE Ao Arch LAX) views. It should be remembered that a significant proportion of TOF patients with a right-sided aortic arch (which occurs in about 25 % of TOF) will have a *left* sided ductus arteriosus originating from the left subclavian or innominate artery, and entering the main/left pulmonary artery [18].

Surgical Considerations

The operative repair of tetralogy of Fallot involves closure of the ventricular septal defect (Fig. 12.4, Video 12.4) and relief of the RV outflow tract obstruction. The outflow tract surgery is dependent upon the severity of infundibular and pulmonary valvar hypoplasia. A transannular outflow tract patch is

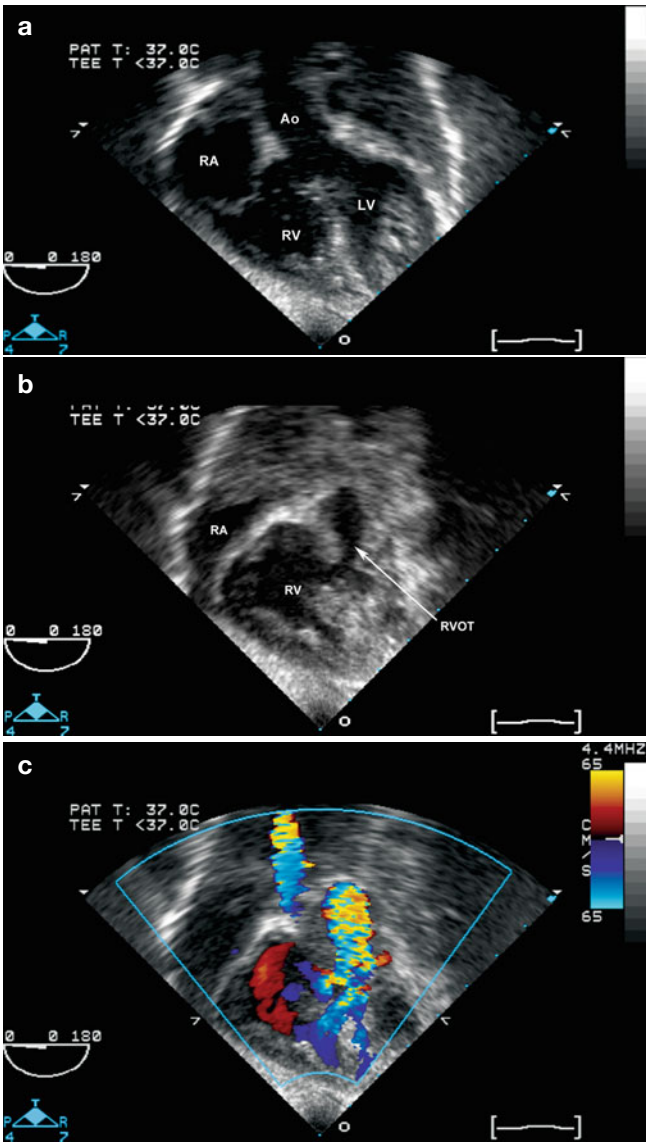
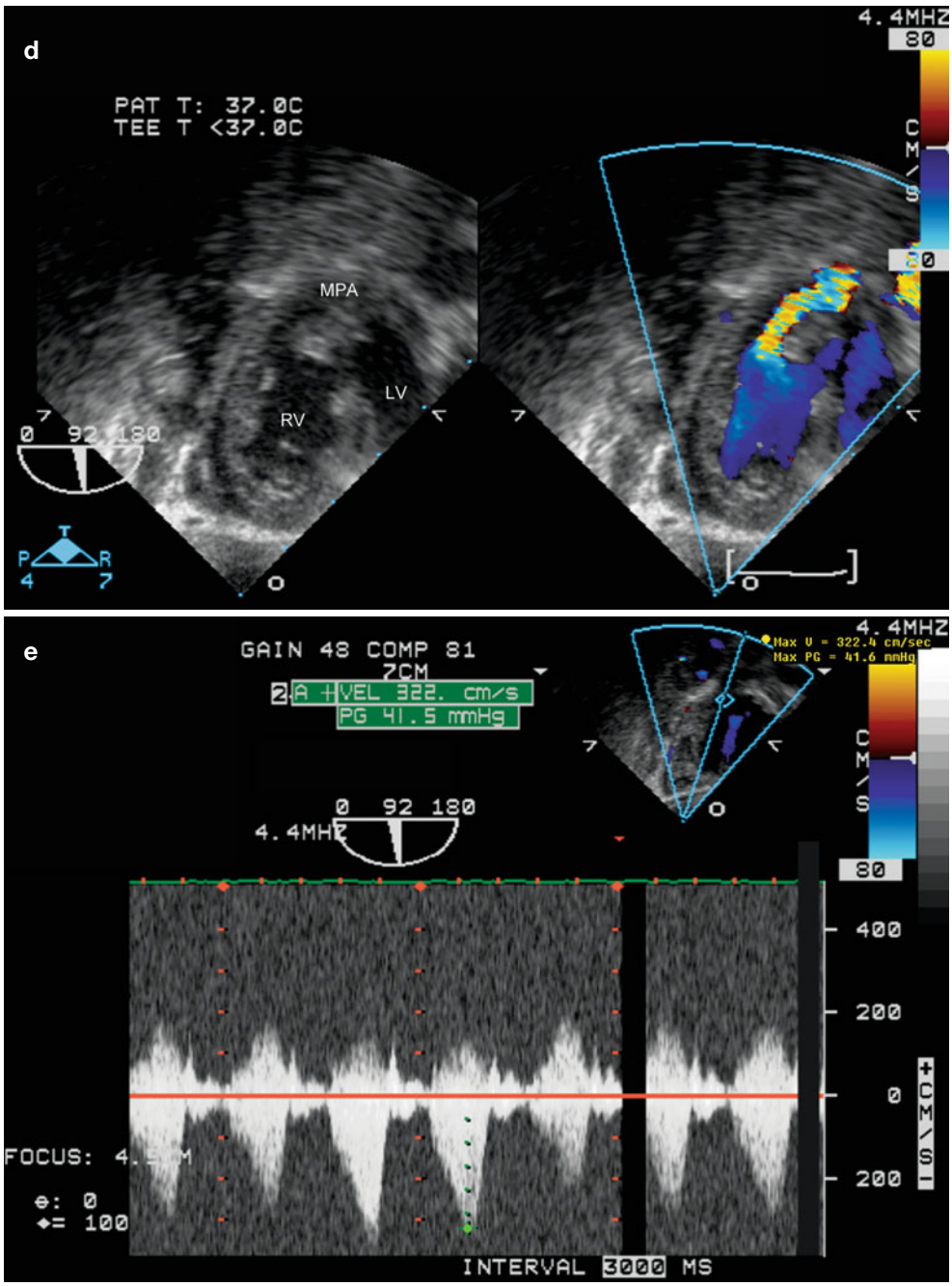


Fig. 12.3 Tetralogy of Fallot: deep transgastric long axis (**a**–**c**) and sagittal views (**d**, **e**) that simulate transthoracic subcostal coronal and sagittal views. (**a**–**c**) Shows the overriding aorta and malalignment ventricular septal defect (**a**). With slight probe withdrawal, the anteriorly malaligned infundibular septum is seen (**b**), producing subpulmonary stenosis, with color flow aliasing at this level (**c**). (**d**, **e**) Also show the anteriorly malaligned infundibular septum with color flow Doppler aliasing, as well as the ventricular septal defect with *right* to *left* shunting. The spectral Doppler tracing shows the typical "dagger" shape seen with subpulmonary stenosis. *Ao* aorta, *LV* left ventricle, *MPA* pulmonary artery, *RA* right atrium, *RV* right ventricle, *RVOT* right ventricular outflow tract

Fig.12.3 (continued)



used when the outflow tract obstruction is severe, with significant pulmonary valve annular hypoplasia. In milder forms of TOF, patch enlargement of the hypoplastic infundibulum (a non transannular patch), if required, is performed while the pulmonary valve is preserved to avoid long-term severe pulmonary regurgitation. In some centers, a transatrial approach is employed, in which the VSD is closed and RV muscle is resected from the right atrium, thereby minimizing or avoiding a right ventriculotomy [19]. Conduits from the RV to the pulmonary artery are required in many cases of pulmonary atresia or when a coronary (typically a left anterior descending from the right coronary) crosses the RV outflow tract. The coronary arteries can be evaluated preoperatively using the mid esophageal aortic valve short axis (ME AV SAX) view, as discussed in Chap. 4. With probe anteflexion and careful evaluation, it is possible to visualize a coronary crossing the RV outflow tract (Fig. 12.5, Video 12.5).

Postoperative TEE is indicated to evaluate for (1) residual RV outflow tract obstruction, (2) residual VSDs (Figs. 12.6 and 12.7, Video 12.6), (3) assessment of ventricular performance, (4) severity of pulmonary regurgita-

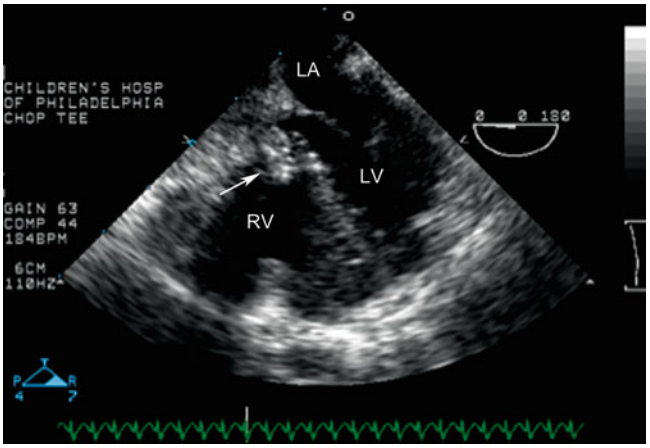


Fig. 12.4 Mid esophageal four chamber view demonstrating right ventricular hypertrophy in a patient with tetralogy of Fallot. Also note the presence of a ventricular septal defect patch (*arrow*), in good position

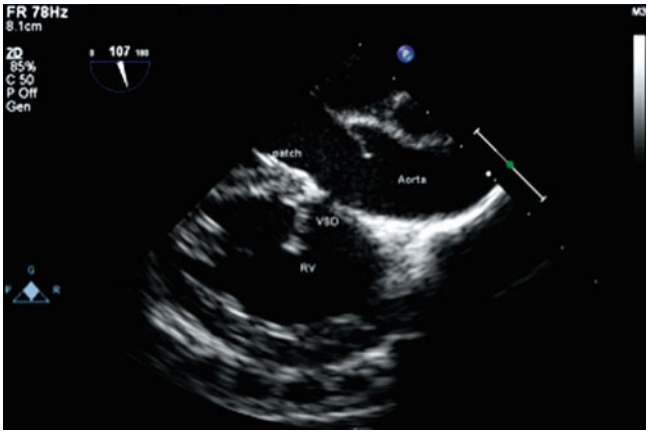


Fig. 12.6 Modified mid esophageal long axis view, with multiplane angle 107° in a patient with tetralogy of Fallot demonstrating a residual ventricular septal defect (*VSD*). The defect is located in the superior aspect of the VSD patch just under the aortic valve. *RV* right ventricle

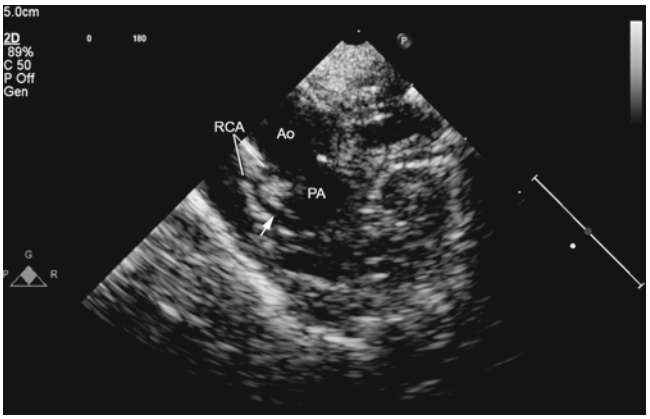


Fig. 12.5 Preoperative study in a patient with tetralogy of Fallot. A prominent anterior descending coronary artery (*arrow*) arises from the right coronary artery (*RCA*) and courses anterior to the right ventricular outflow tract, thereby precluding a transannular patch. *Ao* aorta, *PA* main pulmonary artery

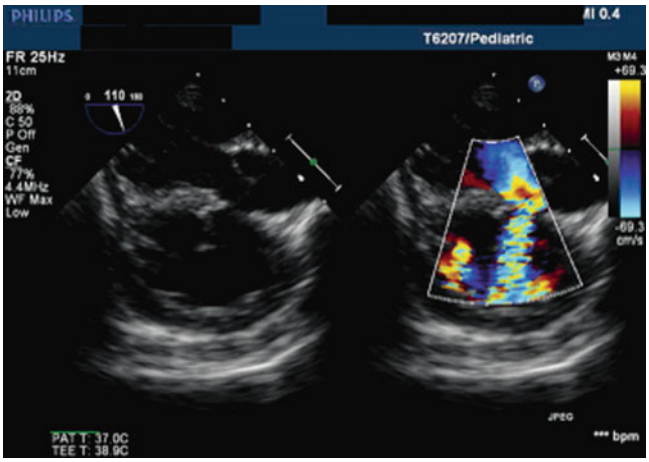


Fig. 12.7 Modified mid esophageal long axis view, with multiplane angle 110°. This color compare image illustrates the presence of a residual ventricular septal defect after repair of tetralogy of Fallot; the defect is located immediately inferior to the aorta. The etiology of the defect is attributed to patch dehiscence. Here, the ventricular shunt direction is left to right

tion, and (5) flow direction across the atrial septum (if an intentional small communication is present). Branch pulmonary artery stenosis can be difficult to diagnose using TEE [20], but in some instances useful information about the branch pulmonary arteries can be obtained using the mid and upper esophageal views, using the techniques and views discussed in Chap. 4 (Fig. 12.8, Video 12.7). As in all forms of conotruncal malformations, all TEE views should be used to assess for residual VSD. The most common type of residual defect is a peri-patch leak between sutures (Figs. 12.6 and 12.7). Defects less than 3 mm tend to have minimal hemodynamic impact, though some defects of that size can be a concern, particularly in very small infants [21]. Determination of RV pressure either by Doppler interrogation of the tricuspid regurgitation jet (if present) or by the residual VSD jet, particularly when the RV outflow tract is unobstructed, is of utmost importance in assessment of hemodynamic significance of the residual VSD. By definition, a restrictive VSD is associated with RV pressure that is subsystemic. If RV pressure cannot be estimated by TEE, an assessment of pulmonary to systemic blood flow ratio (Qp/Qs), in which the surgeon directly obtains blood samples from the SVC and pulmonary and systemic arteries for oximetry, is helpful to determine the significance of the residual VSD.

One of the most challenging types of residual VSDs to assess is the intramural defect. This type of defect occurs in the setting of conotruncal malformations, when the VSD

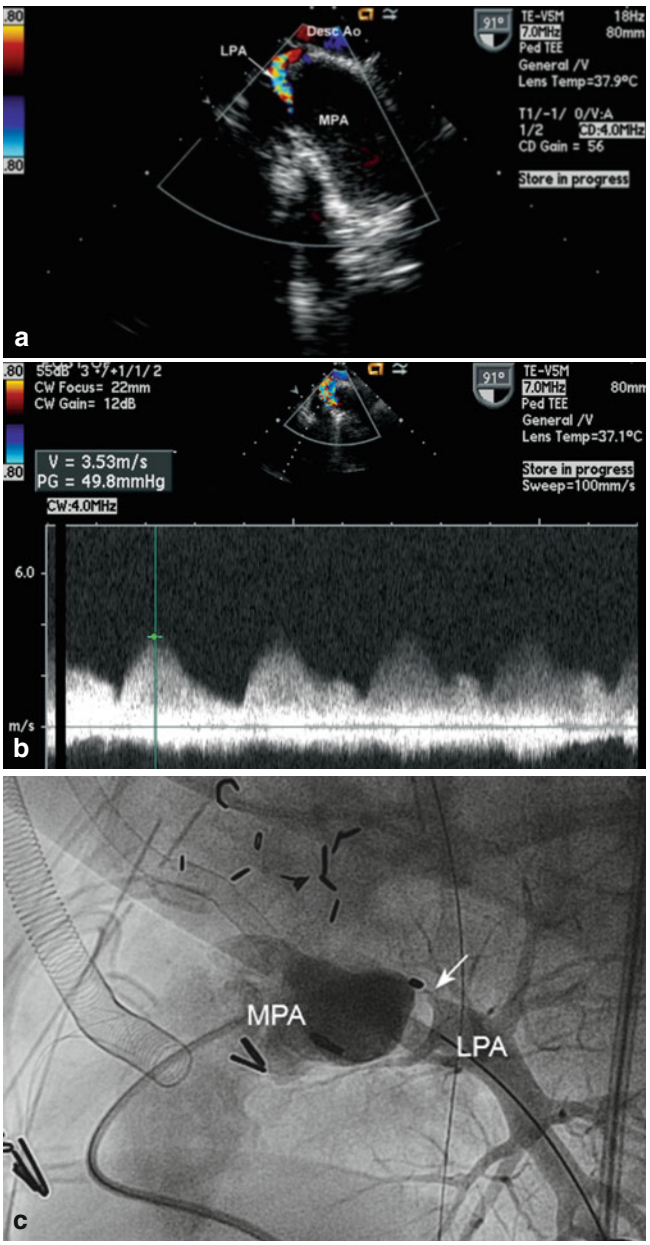


Fig. 12.8 (**a**) Upper esophageal aortic arch short axis with counterclockwise rotation, demonstrating significant left pulmonary artery stenosis in this patient with tetralogy of Fallot who underwent complete correction, including unifocalization of discontinuous pulmonary arteries. (**b**) Spectral continuous wave Doppler tracing showing the significant gradient across the stenotic area. (**c**) Postoperative pulmonary artery angiogram confirming the marked stenosis (*arrow*) of the proximal left pulmonary artery. *Desc Ao* descending aorta, *LPA* left pulmonary artery, *MPA* main pulmonary artery

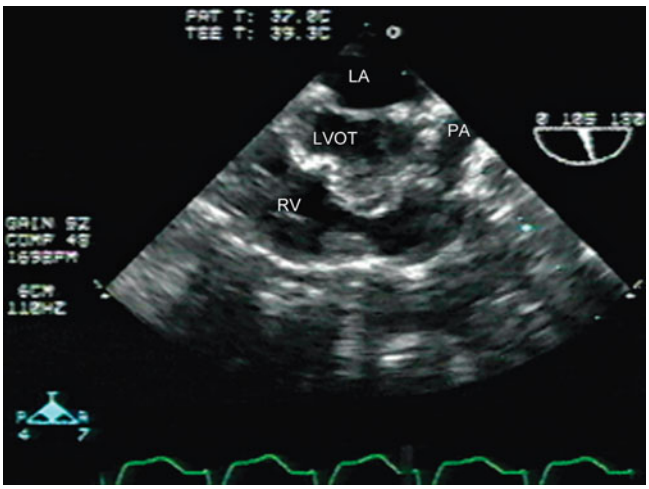


Fig. 12.9 Residual right ventricular outflow tract narrowing is seen in a patient after tetralogy of Fallot repair in a modified mid esophageal right ventricular inflow-outflow view, with slight probe withdrawal to display the outflow tract more clearly *LA* left atrium, *LVOT* left ventricular outflow tract, *PA* pulmonary artery, *RV* right ventricle

patch attaches to the trabeculated right ventricular free wall related to the ventriculoinfundibular fold (parietal band), creating a communication through the intertrabeculated spaces to the right ventricular cavity [21–23]. This area of the heart is difficult for the surgeon to visualize during the repair. Importantly, these defects can increase in size over time. The ME 4 Ch view highlights this type of residual defect well but it can also be seen with a modified ME RV In-Out view, in which the multiplane angle has been rotated to about 90°. Intramural residual VSDs should be looked for in all patients undergoing repair of conotruncal anomalies that include VSD closure.

Assessment of the RV outflow tract for residual obstruction is best performed in a modified ME RV In-Out view with rotation of the multiplane angle to 90–110° (Figs. 12.9 and 12.10, Video 12.8). Doppler interrogation of the RV outflow tract can be performed with this view, and withdrawal of the probe enables evaluation of the more distal portion of the outflow tract. In some cases where the outflow obstruction is more inferior, the DTG LAX and DTG Sagittal views can be used for optimal Doppler evaluation. A peak Doppler velocity that exceeds 3 m/s (36 mmHg) is considered potentially significant [24]. However, it should be noted that a hypercontractile state exists immediately following TOF repair, due in part to inotropic support and hypovolemia, which can accentuate a dynamic RV outflow tract gradient. Indeed, a study by Kaushal et al. evaluating 166 TOF patients immediately following transatrial repair (median age 7 years) found that 35 % had "significant" residual obstruction (gradient >40 mmHg, right:left ventricular pressure ratio >85 %). Most of these patients (88 %) had a dynamic, rather than fixed, obstruction as defined by TEE; patients with fixed obstruction underwent immediate surgical revision, while those with dynamic obstruction did not. Operative mortality and morbidity were not related to

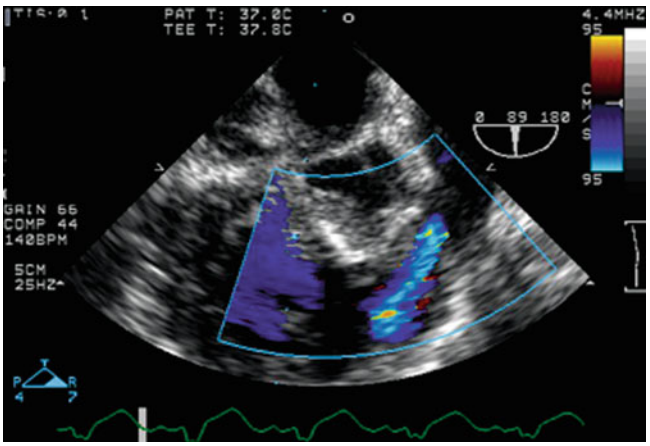


Fig. 12.10 Residual right ventricular outflow tract obstruction seen following tetralogy of Fallot surgery, with color Doppler interrogation in a modifed mid esophageal right ventricular inflow-outflow view. Following surgery, increased velocity in the right ventricular outflow tract is present (manifested by color flow Doppler aliasing), mostly related to dynamic subpulmonary obstruction

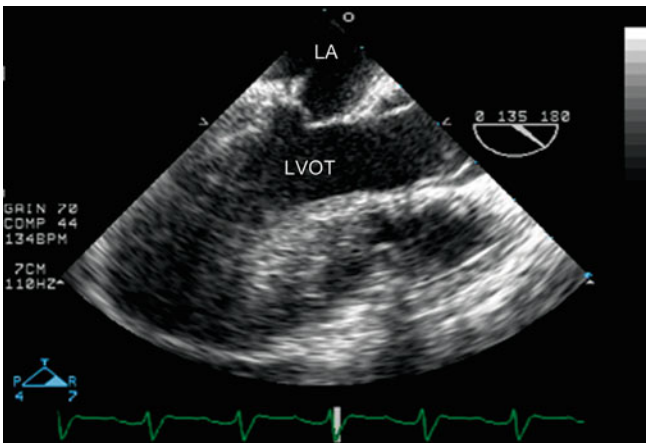


Fig. 12.11 Mid esophageal aortic valve long axis view demonstrating an unobstructed left ventricular outflow tract (*LVOT*) after repair of tetralogy of Fallot. The aortic valve is in the open position. *LA* left atrium

higher residual gradients, and on follow-up (mean 18.5 months) outflow gradients had declined sharply (mean 16 mmHg) irrespective of the severity of intraoperative gradients [25]. Thus a "significant" gradient in the immediate postoperative study should be viewed carefully; the nature of the obstruction (fixed vs. dynamic), and other intraoperative factors, should be considered when deciding to reoperate.

In the immediate postoperative hemodynamic assessment of the TOF patient, several other factors should be evaluated. Detection of intracardiac air and monitoring of cardiac de-airing can be performed using the ME 4 Ch and ME LAX views. Evaluation of right and left ventricular function can be performed with a number of mid esophageal and transgastric TEE views, including ME 4 Ch, mid esophageal two chamber (ME 2 Ch), ME RV In-Out, and the transgastric basal and mid short axis (TG Basal SAX, TG Mid SAX) views. The LV outflow tract is well visualized in both the ME AV LAX, multiplane angle 120–135° (Fig. 12.11, Video 12.9), and ME 4 Ch view (Fig. 12.12, Video 12.10) but this is rarely obstructed after TOF repair. Assessment of severity of pulmonary regurgitation can also be performed using the same views used to evaluate the RV outflow tract, including the ME RV In-Out view and the DTG Sagittal and LAX views. Some degree of pulmonary regurgitation can be expected after TOF repair, particularly following transannular patch. Several methods can be used to evaluate the degree of pulmonary regurgitation, including width of the color Doppler jet compared to outflow tract diameter (mild ≤1/3, moderate 1/3 to 2/3, severe ≥2/3) and presence of diastolic flow reversal in the branch pulmonary arteries [26]. However, changing hemodynamic and ventilatory conditions following TOF surgery can alter the amount of pulmonary regurgitation noted in the immediate postoperative period, compared to later in the postoperative course.

In some TOF patients, a small atrial communication (usually a patent foramen ovale) is left patent as a "pop off" to facilitate cardiac output in the immediate postoperative period. The Doppler flow pattern across the foramen ovale is typically from right atrium to left atrium after surgery as a result of poor RV compliance (from the right ventriculotomy and RV hypertrophy) (Fig. 12.13). The atrial septum can be evaluated by sweeping from the inferior vena cava (IVC) to the superior vena cava. This sweep is performed by starting with the lower esophageal situs short axis (LE Situs SAX) view to image the IVC and hepatic veins, then slowly withdrawing the probe to the ME 4 Ch view (with slight rightward probe rotation) to visualize the atria, atrial septum and superior vena cava, while maintaining the multiplane angle between 0 and 30°.

In the case of a patient with TOF and AV canal defect, both atrial and ventricular septal patches should be evaluated for residual shunts. As with all AV canal repairs (Chap. 8), both AV valves (particularly the mitral valve) must be evaluated for possible regurgitation (Fig. 12.14, Video 12.11). However in addition, the RV outflow tract must be evaluated in the same manner as other TOF patients

In older patients, TEE can be used to assess TOF repair including adults with poor acoustic windows. The long-term impact of severe pulmonary regurgitation can be gauged by evaluation of RV size and performance. When there appears to be significant negative impact of the regurgitation (manifested by marked RV dilation and/or deteriorating RV systolic function), prosthetic pulmonary valve implantation is indicated. This is usually performed

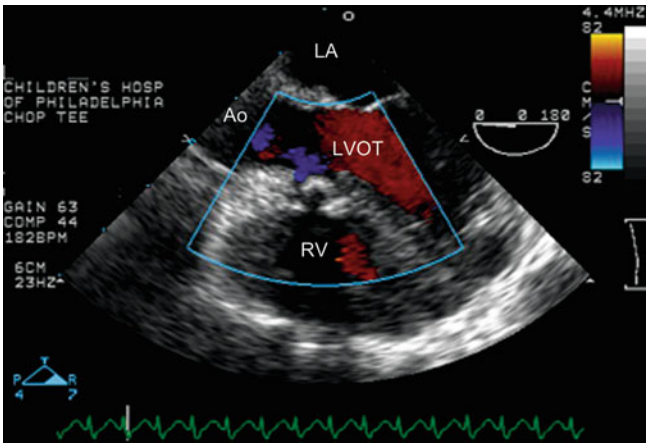


Fig. 12.12 Mid esophageal four chamber view with probe withdrawn to image the left ventricular outflow tract (*LVOT*) demonstrates that there is no residual VSD by color Doppler interrogation after tetralogy of Fallot repair. In addition, flow is laminar into the left ventricular outflow tract. *Ao* aorta, *LA* left atrium, *RV* right ventricle

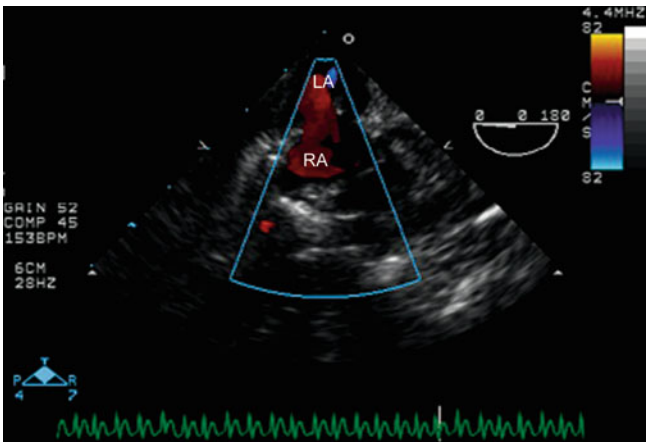
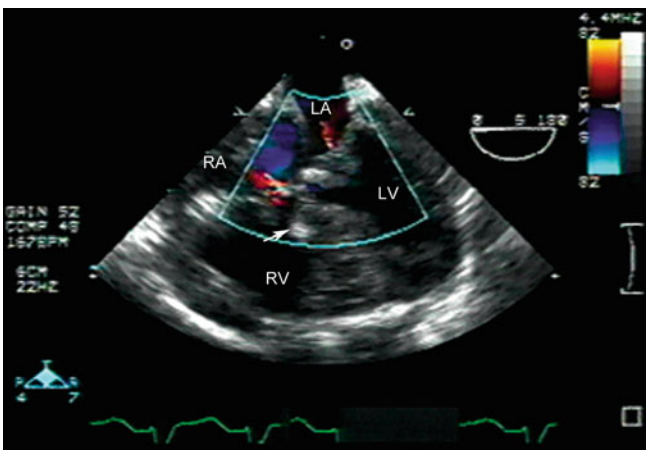


Fig. 12.13 Mid esophageal four chamber view, with probe withdrawn superiorly to the level of the atrial septum and rotated clockwise toward the patient's right side. Color Doppler interrogation demonstrates a residual right to left shunt (*red*) through an atrial communication that was intentionally left patent during surgery. This type of shunting typically occurs in the early postoperative period after tetralogy of Fallot repair. *LA* left atrium, *RA* right atrium



surgically but in some instances, a transcatheter approach (notably using the Melody valve) can be employed (Chap. 16). In addition, tricuspid valve replacement might be indicated in cases of severe tricuspid regurgitation (Fig. 12.15, Video 12.12). RV systolic pressure estimate can be performed if tricuspid regurgitation is present. Aortic root dilation is a common finding in adults with TOF and can be associated with the development of aortic regurgitation over time. Patients with TOF can also develop important arrhythmias such as ventricular tachycardia. In some cases, TEE can help distinguish the type of arrhythmia.

Double Outlet Right Ventricle

Anatomy

Double outlet right ventricle (DORV) is not a specific congenital malformation, but rather, an abnormal ventriculo-

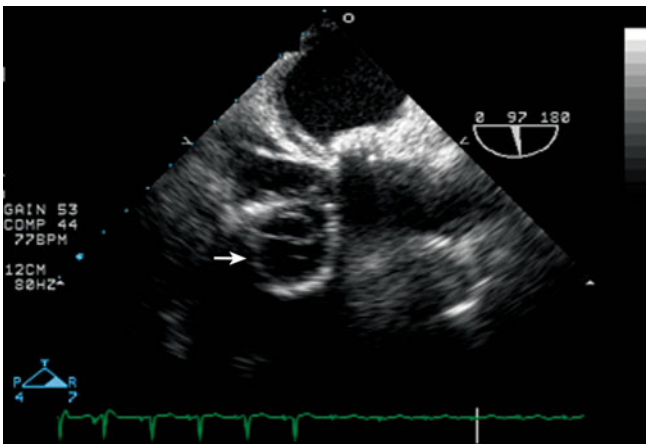


Fig. 12.15 Severe tricuspid regurgitation prompted tricuspid valve replacement in this adult long after tetralogy of Fallot repair; as seen here in a modified mid esophageal right ventricular inflow-outflow view (multiplane angle 97°), the prosthetic valve appears in cross section as indicated by the *arrow*

Fig. 12.14 Mid esophageal four chamber view in a patient who has undergone tetralogy of Fallot and atrioventricular canal repair. A large ventricular septal defect patch is seen (*arrow*). Minimal residual atrioventricular valve regurgitation is seen across the right and left atrioventricular valves. *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

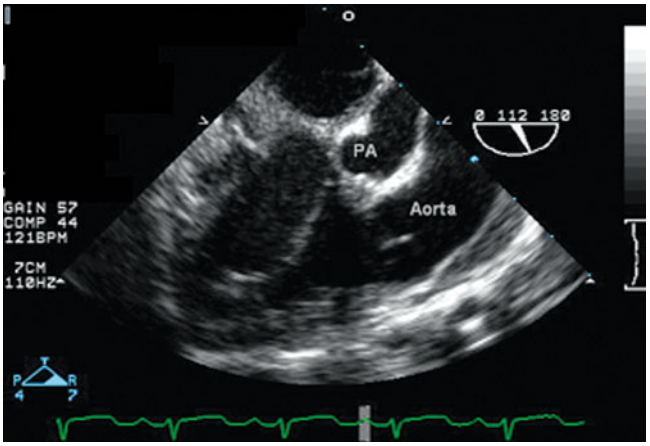


Fig. 12.16 Modified mid esophageal right ventricular inflow-outflow view (multiplane angle 110°) in a patient with double outlet right ventricle with posterior malalignment of the conal septum and subaortic ventricular septal defect, demonstrating that the aorta is anterior to the smaller pulmonary artery (*PA*). Both great vessels arise from the anterior right ventricle. The conal septum is seen between the aorta and PA

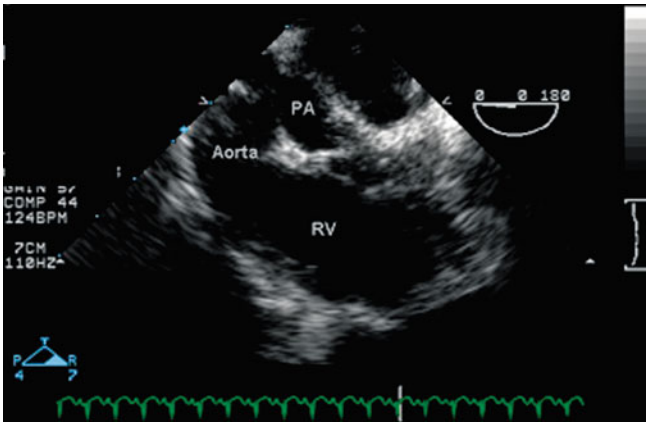


Fig. 12.17 Mid esophageal four chamber view with the probe withdrawn towards the base of the heart in the same patient displayed in Fig. 12.16 with double outlet right ventricle, posterior malalignment of the conal septum with subaortic ventricular septal defect; in this image, the aorta and pulmonary artery (*PA*) come into view, both arising from the right ventricle (*RV*). The PA is smaller than the aorta because of the conal septal malalignment

arterial alignment whereby both great arteries arise from the morphologic RV (Figs. 12.16 and 12.17, Video 12.13). By most criteria, both great vessels must sit more than 50 % over the RV [27]. Typically, there is *bilateral conus*, with myocardium separating both great vessels from the AV valves. However, other conal anatomy can occur in DORV [28]. A VSD is present in almost all cases and usually acts as the "outlet" from the left ventricle (LV). There are rare reports of DORV with an intact ventricular septum [29, 30]. In DORV, the ventricular loop may be "D" (normal) or "L" (inverted) and this will affect the relationship of the great arteries to the ventricle and to each other.

The anatomic and physiologic spectrum of DORV is wide. Associated anomalies are common and include ventricular hypoplasia, superior/inferior ventricles, mitral valve anomalies (including mitral atresia, straddling or parachute mitral valve), common AV canal (in heterotaxy syndrome) and outflow obstruction [28, 31, 32].

The pathophysiology can be similar to TOF, transposition of the great arteries, or a large VSD. This physiology is determined by the location of the great vessels in relationship to the VSD and the degree of outflow obstruction. Of critical importance is the type and location of the VSD: it often determines the type of surgical repair as well as, in some instances, the viability for two-ventricle versus a single ventricle surgical strategy. By far, the most common type of VSD in DORV is the malalignment type, however any type of VSD (such as AV canal, mid muscular, or apical muscular) can occur [33]. The VSD is also classified as one of four types, based upon its relationship to the great vessels: subaortic, subpulmonary, noncommitted, and doubly committed. In those patients with DORV with malalignment type VSD, the most common location of the VSD is subaortic. This is seen on echocardiography as muscle between the mitral and aortic valves. If there is no pulmonary stenosis, the physiology is similar to a large unrestrictive VSD because the blood flow streams from the LV into the aorta. If there is pulmonary stenosis, the physiology is similar to TOF. However, the distinction between DORV and TOF is of utmost importance because those patients with TOF-like DORV have complete subaortic conus that can result in subaortic obstruction after surgery [34]. Subaortic stenosis is extremely rare in TOF.

DORV with subpulmonary VSD is classically termed a "Taussig-Bing" anomaly. In this defect, the great vessels are spatially oriented side-by-side, bilateral conus is present, and the aorta is to the right of the pulmonary artery. The blood from the LV streams into the pulmonary artery so that the physiology is similar to transposition of the great arteries (see below). The subaortic conus is usually small, thus arch obstruction is a frequent association. Some cases of DORV with subpulmonary VSD are not classic Taussig-Bing anomalies but rather fall within the spectrum of that anatomic description. The distinction is critically important when determining surgical strategy.

More uncommon locations of the VSD in DORV include non-committed or doubly-committed subtypes. In DORV with non-committed VSD, the VSD is usually a muscular or a canal (inlet) type VSD. Since it is far from both outflow tracts, baffling procedures may pose a significant challenge. Often, AV valve tissue is obstructing the potential pathway from the LV to the aorta. Moreover, straddling tricuspid valve can occur in association with a canal type VSD. When a muscular VSD is present, there is a tendency for the defect

to get smaller over time because it is surrounded by muscle; therefore in general, muscular VSDs are not considered reliable pathways to a great artery. In the rare doubly-committed type, both great vessels sit directly over the VSD.

Other variations of DORV exist, most commonly including those with mitral atresia or hypoplasia associated with left ventricular hypoplasia. When there is significant ventricular hypoplasia, the single ventricle surgical strategy is usually indicated. DORV is often seen in patients with superior/inferior ventricles with criss-cross AV valves. In this rare anomaly, the RV is situated superior to the LV and it is typically hypoplastic. The VSD is usually a canal (inlet) type.

Genetic syndromes can be seen with DORV but there is not one particular association. Transforming growth factor (TGF)-beta 2 knockout mice have been observed to develop DORV in the majority of cases [35]. Heterotaxy syndrome often has DORV as one of its cardiac components.

TEE Evaluation

There are pertinent findings seen with DORV to keep in mind when performing a TEE:

- Location and size of the VSD.
- Relationship of great vessels to the VSD.
- Presence of additional VSDs.
- Restrictive VSD due to accessory AV valve tissue.
- Anatomy and size of AV valves.
- Presence and severity of outflow tract obstruction (and to which great vessel).
- Coronary anomalies, particularly if the arterial switch operation is being considered.

One of the most important aspects of the pre-operative evaluation of the patient with DORV is assessment of location and size of the VSD and its relationship to the great vessels (Fig. 12.18, Video 12.14). This, along with the other echocardiographic goals mentioned above, can often help in the determination of the optimal surgical strategy. TEE is an excellent tool to assess for AV valve abnormalities that might preclude a biventricular repair. For example, a straddling AV valve over a VSD usually obstructs the pathway from the LV to a great artery. TEE can also assess whether a VSD is restrictive and the mechanism of the restriction (i.e. AV valve tissue, conal muscle) [36]. Moreover, important anatomic features to help determine a surgical strategy include prominence of the conal septum, presence of tricuspid valve attachments to conal septum, and the distance between the pulmonary and tricuspid valves [37].

The standard ME 4 Ch view may not show evidence of pathology other than RV hypertrophy. However, probe withdrawal and anteflexion will demonstrate that both great vessels arise mostly or completely from the anatomic RV (Fig. 12.16 and 12.17). The anatomy of the AV valves can be seen in this view, as well as in a sweep of the multiplane angle from 0 to 90° that incorporates both the ME RV In-Out

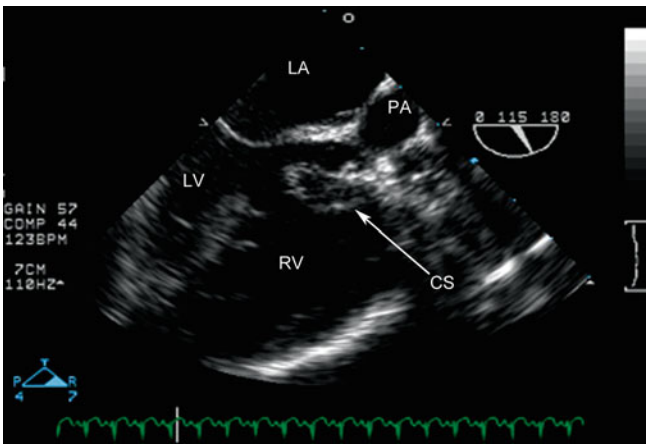


Fig. 12.18 Mid esophageal aortic valve long axis view in the same patient as Figs. 12.16 and 12.17 with double outlet right ventricle, posterior malalignment of the conal septum (*CS*), and subpulmonary ventricular septal defect, highlighting the posterior deviation of the conal septum that results in severe subpulmonary stenosis. The conal septum is also hypertrophied. *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *RV* right ventricle

and ME 2 Ch views. Alignment of the great vessels is dependent on their position; therefore no single TEE view will be adequate for all patients with DORV. The advantage of the multiplane probe is that the best view of the outflow tracts can be determined as the study is performed. The DTG LAX and DTG Sagittal views are also useful to visualize the great arteries and their relationship to the VSD (Fig. 12.19, Video 12.15). Additional VSDs can be viewed in the ME 4 Ch and ME RV In-Out views, with variation of multiplane angle between 0 and 90° and axial probe rotation to perform a complete sweep of the ventricular septum. The transgastric short axis views (TG Basal SAX, TG Mid SAX) are also useful to evaluate for additional VSDs. Other associated anomalies can be demonstrated using TEE, including AV canal defect (as is common in heterotaxy syndrome), as well as juxtaposition of the atrial appendages, which can be seen from a modified ME 4 Ch view as well as ME AV SAX view (Fig. 12.20, Video 12.16). Systemic venous connections can be evaluated with the LE Situs SAX and lower esophageal IVC long axis (LE IVC LAX) views, as well as the ME Bicaval view (Chap. 4). Pulmonary venous connections can be evaluated using the modified ME 4 Ch and ME Bicaval/ME 2Ch views, as described in Chaps. 4 and 6.

Color Doppler can be used to assess for preoperative AV and semilunar valve regurgitation as well as restriction of flow across the VSD. Color interrogation can also be useful to assess inflow into each ventricle, particularly when straddling or overriding AV valves are suspected. Pulsed and continuous wave Doppler assessment of outflow tract obstruction is dependent on the angle of interrogation. The DTG LAX and DTG Sagittal views can be very helpful for this purpose. LV systolic pressure estimate (as measured by

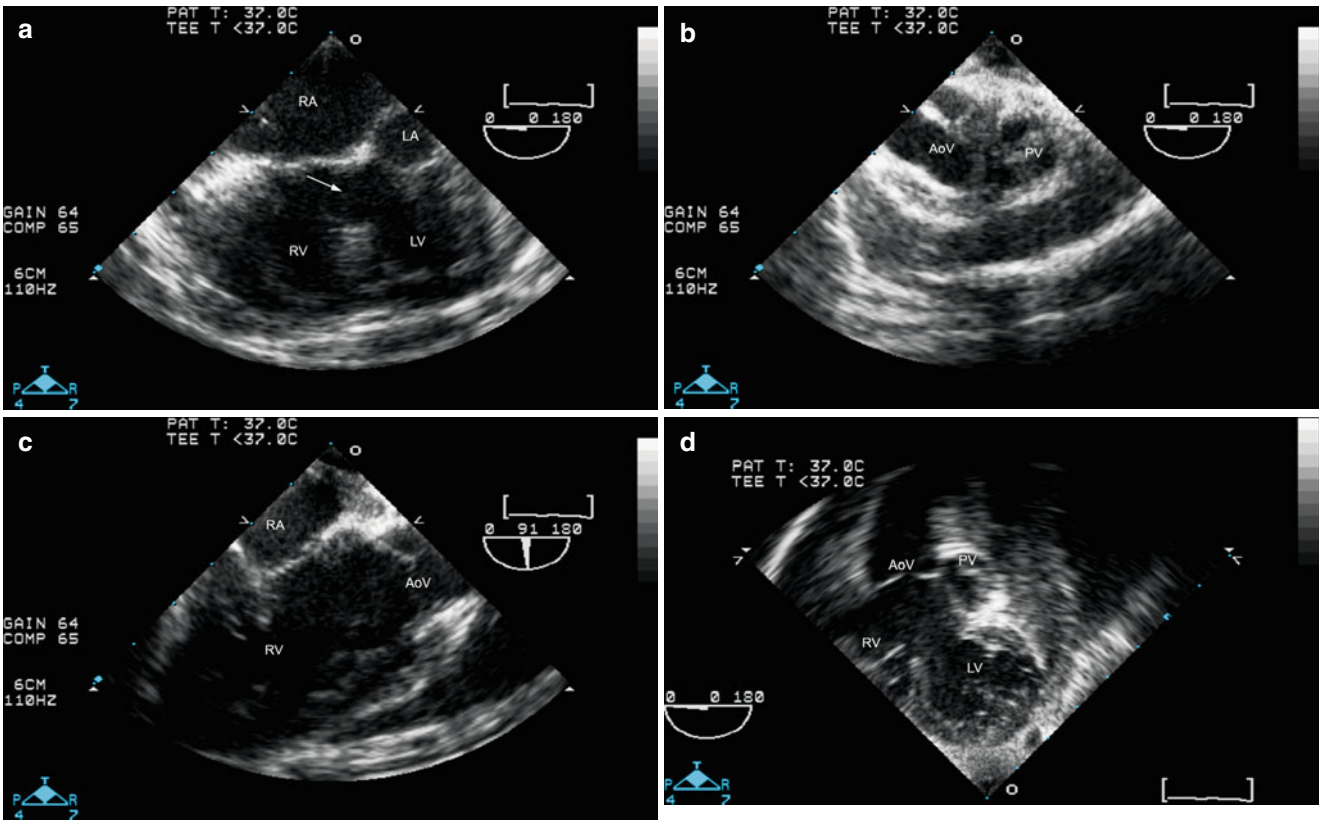


Fig. 12.19 Double outlet right ventricle with pulmonary outflow tract obstruction. Mid esophageal four chamber view (**a**) shows a large ventricular septal defect (*arrow*). With probe withdrawal and anteflexion, the semilunar valves are visualized (**b**). However the pathway from left ventricle (*LV*) to aortic valve (*AoV*) is not clearly shown. Rotation of multiplane angle to about 90° (**c**) shows right atrium (*RA*) and right ventricle (*RV*) as well as AoV, but it is still unclear whether the pathway from LV to AoV is unobstructed. The deep transgastric long axis view (**d**) shows that this pathway is unobstructed. This patient successfully underwent a patch closure of the ventricular septal defect and relief of pulmonary outflow tract stenosis.

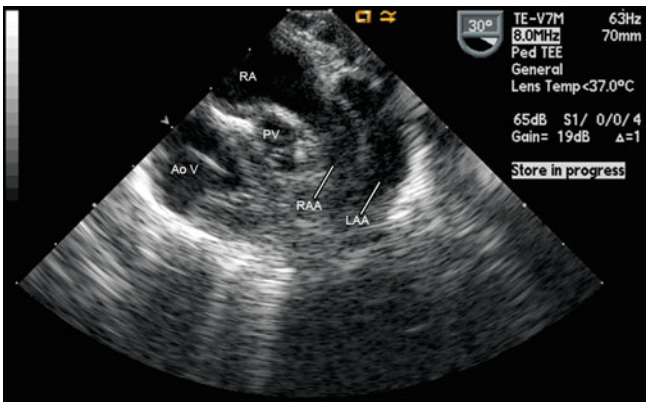


Fig. 12.20 Left juxtaposition of the atrial appendages in a patient with double outlet right ventricle and pulmonary stenosis. This image was obtained from a mid esophageal aortic valve short axis view. Note how the right atrial appendage (*RAA*) crosses posterior to the great arteries to lie just anterior to the left atrial appendage (*LAA*). *AoV* aortic valve, *RA* right atrium, *PV* pulmonary valve

mitral regurgitation jet) is important if a restrictive VSD is suspected. The LV may be at suprasystemic pressure in these instances. This method of assessment, performed in the ME 4 Ch and ME 2 Ch views, is particularly important when the VSD jet is not at an appropriate angle for interrogation.

Surgical Considerations

When both ventricles and both AV valves are well developed, the goal of surgical repair is to utilize the VSD to baffle the LV to one of the great arteries. If baffled to the aorta, a "physiologic repair" is achieved. If baffled to the pulmonary artery, an arterial switch operation is required. In cases where the VSD is remote from the great arteries, septation might not be possible. In addition, presence of a straddling tricuspid valve over a canal type VSD or straddling mitral valve over a malalignment VSD may also preclude septation into two ventricles. In these cases, staged surgical palliation culminating in the Fontan operation is usually performed [38].

For those undergoing a biventricular repair, selection of the appropriate operation depends upon an assessment of great artery relationship in relation to the ventricles and VSD, whether outflow tract obstruction is present and if so, which outflow tract is obstructed and to what degree. Based upon these assessments, a determination can be made as to

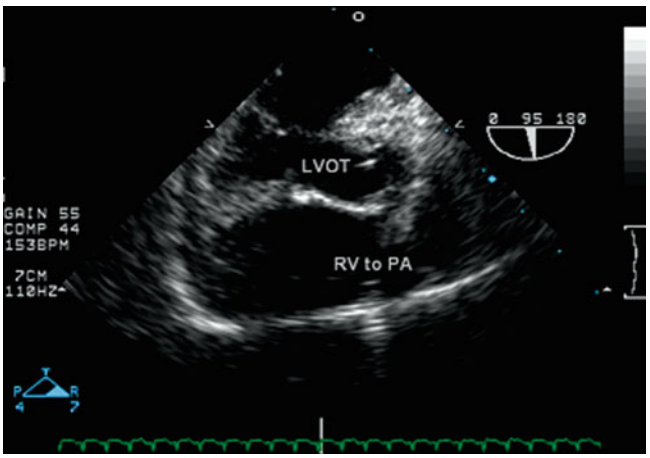


Fig. 12.21 Mid esophageal aortic valve long axis view in the patient from Figs. 12.16, 12.17, and 12.18 with double outlet right ventricle, posterior malalignment of the conal septum with subpulmonary ventricular septal defect after the Nikaidoh procedure (aortic translocation). The translocation of the aorta has placed the aortic valve closer to the left ventricle, improving left ventricle to aortic valve alignment and reducing the possibility of subaortic obstruction once the ventricular septal defect has been closed by a patch. This image demonstrates the "physiologic" repair achieved by baffling the ventricular septal defect to the aorta in its new position and the right ventricle to pulmonary artery conduit. *LVOT* left ventricular outflow tract, *PA* pulmonary artery, *RV* right ventricle

the most suitable operation to effect a complete biventricular repair. The most common operations include:

- Patch closure of VSD (baffle to the aorta).
- Rastelli type procedure: Baffle closure of VSD to aorta, with repair of obstructed RV outflow tract (outflow patch or RV to pulmonary artery conduit).
- Arterial switch operation with closure of VSD to neo-aorta.
- Nikaidoh operation with translocation of the aorta into the pulmonary position over the LV, VSD closure and placement of an RV to pulmonary artery conduit [39, 40]. This operation is designed to bring the aorta and aortic valve closer to the LV, thus reducing the risk of postoperative LV outflow tract obstruction (Fig. 12.21, Video 12.17).

Postoperative assessment is dependent upon the surgical intervention. In those patients with a subaortic VSD, the two most important issues for intraoperative TEE are assessment for a residual VSD and determination of possible LV outflow tract obstruction from the baffle to the aorta. Subaortic conus as well as VSD patch position may result in significant subaortic obstruction. The best view to assess the LV outflow tract is variable, depending on the relationship of the aorta to the LV. Though the ME AV LAX view with a multiplane angle of 120° is best for a normal LV outflow tract, this may not be the case in DORV, thus trying different TEE views, probe rotations, and multiplane angles, will often be required to obtain the best image. Doppler interrogation of the outflow tracts can also be challenging. A variety of different TEE views will often be necessary to obtain the optimal Doppler angle of interrogation; these usually include a combination of mid esophageal, transgastric, and (when available) deep transgastric views. Severity of obstruction may also have to be determined by other methods such as smallest diameter. In some cases, direct pressure measurements in the LV and the aorta might be required.

Assessment for residual ventricular communications is very important in patients undergoing repair of DORV. Intramural type defects can occur in this lesion as well (See section "Tetralogy of Fallot"). In patients with Taussig-Bing anatomy that undergo the arterial switch operation, intraoperative TEE assesses adequacy of the neo-aortic and neo-pulmonic anastomoses (see section "Transposition of the Great Arteries") as well as the adequacy of the repair of associated defects.

Truncus Arteriosus

Anatomy

Truncus arteriosus communis, also known more simply as truncus arteriosus (TA) or common arterial trunk, is characterized by the following anatomic features: a solitary great vessel arises from the heart, giving rise to the aorta, at least one pulmonary artery, and at least one coronary artery. Almost always, there is a large VSD in which the septal band (septomarginal trabeculation) forms the "floor" of the VSD, and the truncal root overrides the crest of the ventricular septum so that the solitary truncal valve forms the "roof" of the VSD [41]. The morphology of the VSD in TA is virtually identical in to that of TOF [42], however in TA the infundibular septum is absent. TA must be distinguished from TOF/PA. In TOF/PA a remnant of the main pulmonary artery and valve can often be seen. In the most extreme form of TOF/PA, the pulmonary circulation may be supplied directly from the aorta. Thus, the distinction between TOF/PA and TA is sometimes difficult. In TA, the solitary semilunar valve, the truncal valve, is often morphologically abnormal. Though a tricuspid truncal valve is the most common anatomic subtype (69 %), other variations can occur including (in order of frequency), quadricuspid (22 %), bicuspid (9 %), and rarely valves with five leaflets (0.3 %) [43]. Because of these frequent abnormalities in valvar anatomy, truncal valve stenosis and/or regurgitation are common, and if clinically significant, they may need to be addressed surgically. Approximately 25 % of patients with TA have a right aortic arch. Other associated cardiac anomalies include aberrant origin of a subclavian artery from the descending aorta, persistent left superior vena cava to coronary sinus connection, coronary anomalies, and an atrial communication in the

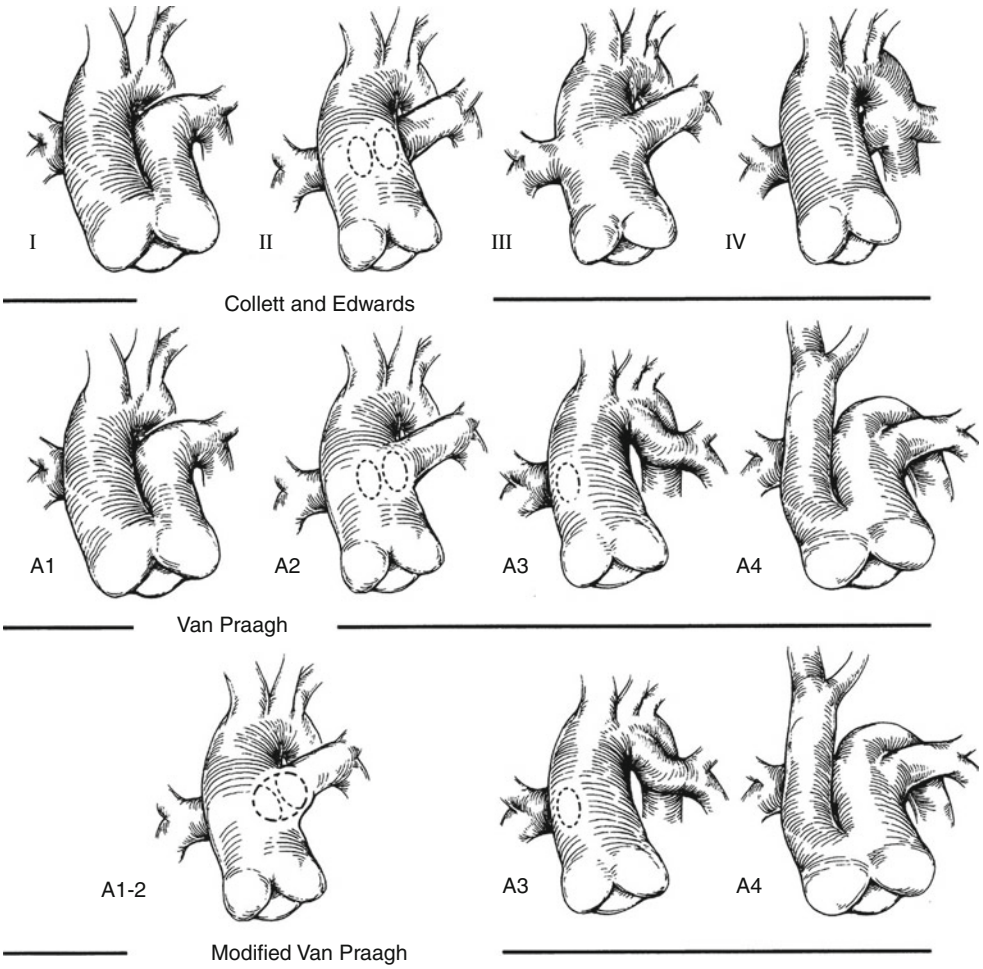


Fig. 12.22 The two common classification systems for truncus arteriosus: Collett and Edwards (*first row*) and Van Praagh (*second row*). A modified Van Praagh classification system is shown on the *third row*. See text for details (Reprinted from: Jacobs [46] with permission from Elsevier)

form of a patent foramen ovale or secundum atrial septal defect. There have been rare reports of TA with single ventricle. In the absence of aortic arch obstruction a ductus is rarely seen in this lesion.

Two classification schemes have been proposed for this malformation: one described by Collett and Edwards [44], the other by Van Praagh [42, 45]. Both are based principally upon the manner in which the pulmonary arteries arise. The classifications share similarities although there are also some important differences (Fig. 12.22).

The Collett and Edwards classification is listed as follows:

Type 1: There is partial absence of the aorto-pulmonary artery septum, and a short main pulmonary artery segment (of variable length) that arises from the common trunk and gives rise to the branch pulmonary arteries. This is the most common type, comprising 48–68 % of all TA [43].

Type 2: There is total absence of the aorto-pulmonary septum and the pulmonary arteries arise directly from the posterior aspect of the common trunk. This type comprises 29–48 % of all TA [43].

Type 3: The branch pulmonary arteries arise separately from the common trunk and are remote from each other. This type comprises 6–10 % of all TA [43].

Type 4: The pulmonary circulation derives from the collateral arteries arising from the descending aorta. This type is now considered a form of TOF/PA with aortopulmonary collaterals, and is not generally used as a classification for TA.

The Van Praagh classification is listed below:

Type A1: There is partial absence of the aorto-pulmonary artery septum: there is a main pulmonary artery segment (of variable length) that arises from the common trunk and gives rise to the branch pulmonary arteries.

Type A2: There is total absence of the aorto-pulmonary septum and the pulmonary arteries arise directly from the posterior aspect of the common trunk.

Type A3: One of the pulmonary arteries arises directly from the common trunk and one arises directly from a collateral supply (usually the ductus arteriosus). This is also known as truncus with absent pulmonary artery.

Type A4: Truncus arteriosus in association with interruption of the aortic arch. The common trunk gives rise to the

ascending aorta and a main pulmonary artery, from which the ductus arteriosus and the branch pulmonary arteries arise. The transverse arch is absent, thus there is an interrupted aortic arch. The ductus arteriosus provides blood to the descending aorta, thus this type of TA is a ductal-dependent lesion. The interruption is usually type B (between the left carotid and the left subclavian arteries). This type occurs in 13 % of cases.

Of note, the original Van Praagh classification provided a subclassification of each type: **A** (with a VSD) and **B** (no VSD). Thus a Type 1 TA could be divided into Type A1 and Type B1, depending upon the presence/absence of a VSD [42]. However given the fact that a VSD is almost always present, the Type B subclassifications are rarely seen. The reader will also note that the first two types of TA for both classification schemes are essentially identical; these are by far the common types of TA encountered. Another important point is that many TA patients are classified as "Type 1" when, in reality, the two pulmonary artery orifices are located so close together that no main pulmonary artery segment is present. This type is often referred to as "truncus arteriosus Type 1–2" or "Type 1½" (Fig. 12.22) [46].

Genetic syndromes are quite common in association with TA. In fact, 40 % of patients have 22q11 deletion syndrome; this association is particularly high in those with a right aortic arch [47]. Other chromosomal defects such as isochromosome 8q have been implicated as well [48].

TEE Evaluation

The pertinent aspects of cardiac anatomy and function that should be evaluated in a patient with TA includes the following:

- Assessment of the VSD and evaluation for potential additional VSDs.
- Evaluation of truncal valve anatomy and function.
- Determination of the origin of both pulmonary arteries from the common trunk as well as their anatomy.
- Evaluation of AV valve function.
- Evaluation of ventricular systolic function.
- Comparison of ascending aorta to main pulmonary artery size. In Type 1 TA (Type A1 of Van Praagh), the ascending aorta is generally much larger than the main pulmonary artery segment. However in TA patients with an interrupted aortic arch (Type A4 of Van Praagh), the ascending aortic diameter will usually be smaller than that of the main pulmonary artery [42, 45, 49].
- Assessment of other possible cardiac abnormalities, including atrial septal defects, systemic/pulmonary venous anomalies, and possible coronary artery anomalies.

TEE can be very helpful in the pre-operative evaluation of the patient with TA. A careful TEE study can be used to evaluate all of the elements listed above. The ME 4 Ch view provides for evaluation of AV valve function and global assessment of ventricular performance. Withdrawal of the probe with anteflexion will demonstrate the truncal valve. Forward rotation of the multiplane angle to 30–45° will often reveal the truncal valve anatomy *en face* (Fig. 12.23, Video 12.18). Similar to all conotruncal defects, complementary TEE views (modified ME 4 Ch, ME Bicaval, LE Situs SAX and LE IVC LAX) establish the systemic venous connections and confirm the presence of an atrial communication, if present. The VSD can be visualized in orthogonal planes in the ME 4 Ch view (using probe anteflexion) or the ME AV LAX view (Fig. 12.24, Video 12.19). In Type 1 or 2 TA, the mid ME Asc Ao SAX and mid esophageal ascending aortic long axis (ME Asc Ao LAX) views, will often demonstrate the pulmonary arteries originating from the posterior aspect of the trunk (Fig. 12.24, Video 12.19). It can be difficult to see the entire length of the branch pulmonary arteries by TEE, but significant portions can be visualized using the aforementioned TEE views as well as the upper esophageal views (UE PA LAX, UE Ao Arch SAX).

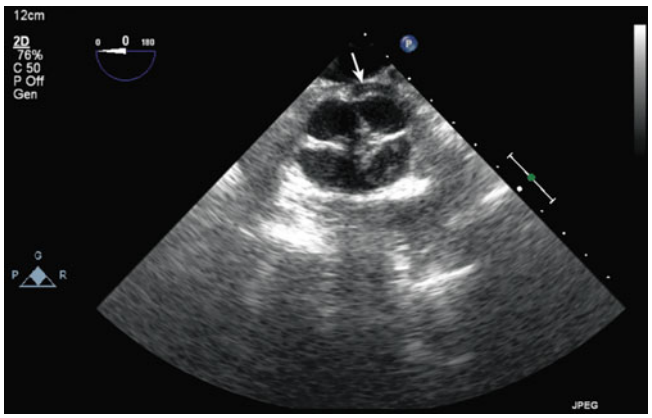


Fig. 12.23 Truncal valve seen *en face* from a modified mid esophageal aortic valve short axis view. This shows a quadricuspid truncal valve with thickened edges and a central area of noncoaptation. Note the left main coronary artery arising from the rightward, posterior cusp (*arrow*)

It is important to consider that TEE probe placement can be difficult in small infants with TA especially when there is associated 22q11 deletion syndrome (Chap. 3).

Surgical Considerations

Surgery for TA is usually performed in the neonatal period because most patients become symptomatic with congestive symptoms and pulmonary vascular obstructive disease can develop early. The type of surgical approach is dictated by the truncal anatomy. For the common forms of TA (Types 1 and 2) surgical repair consists of closure of the VSD to the truncal valve, establishing continuity between the LV and truncal valve, removal of the pulmonary arteries from the common trunk, and in most cases, placement of a conduit between the RV and the pulmonary arteries. In TA Type A4

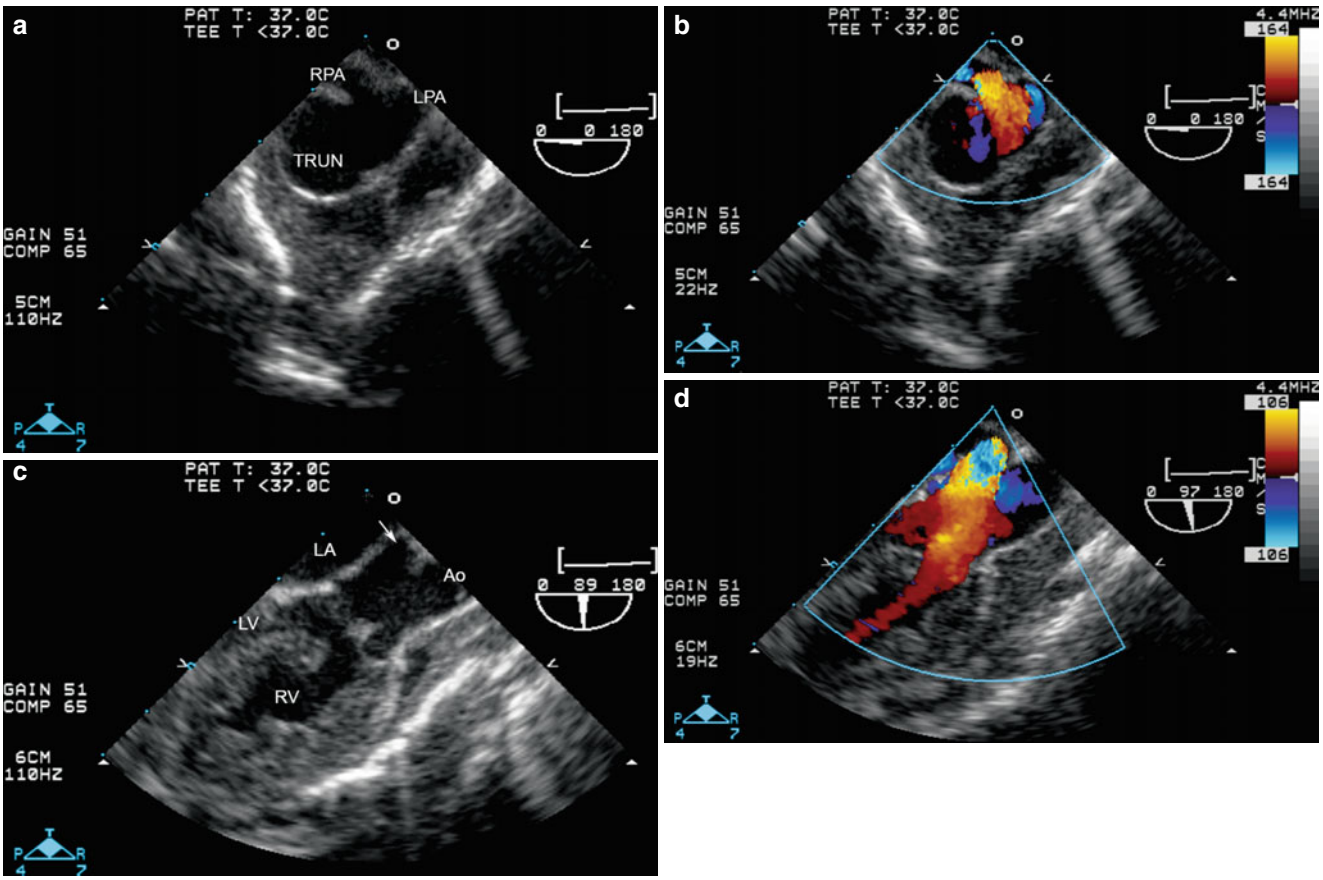


Fig. 12.24 Truncus arteriosus Type "1½" as seen from the mid esophageal ascending aortic short axis (**a**, **b**) and mid esophageal aortic valve long axis (**c**, **d**) views. From the short axis view, the right pulmonary artery (*RPA*) and left pulmonary artery (*LPA*) origins are seen immediately adjacent to each other, arising from the posterior aspect of the trunk (*TRUN*). From the long axis view this posterior origin of the pulmonary arteries is also seen (*arrow*), and the truncal valve shown to override the ventricular septal defect. *Ao* ascending aorta, *LA* left atrium, *LV* left ventricle, *RV* right ventricle

(of Van Praagh), the interrupted arch is addressed in addition to VSD closure and RV to pulmonary artery conduit, as noted above. For all TA repairs, if the truncal valve manifests significant stenosis and/or regurgitation, surgical intervention might be necessary.

Postoperative TEE helps determine adequacy of the repair. The goals of this imaging are to assure that there is unobstructed flow from the LV to the truncal root, no significant residual VSD and unobstructed flow from the RV to the branch pulmonary arteries. Evaluation of the interatrial septum should be undertaken for residual shunting. It should be noted that in some cases, an intentional small interatrial communication remains, allowing for potential right to left shunting and the maintenance of cardiac output during the immediate postoperative period. In addition, the postbypass examination is of benefit in the evaluation of truncal valve function (estimation of residual severity of obstruction and/or regurgitation) in cases where concomitant valve interventions were performed.

The ME 4 Ch view sweeping from inferior to superior demonstrates that the VSD patch is in the correct position and color flow Doppler can be used to determine whether there is residual shunting (Fig. 12.25, Video 12.20). The intramural type VSD previously described can also occur with TA repair [23]. Assessment for a residual VSD should always occur in more than one view. The ME RV In-Out view is quite useful to demonstrate a peri-patch residual leak. In addition, the RV to pulmonary artery pathway is best seen in this view with a counterclockwise turn of the probe toward the left side of the patient. Doppler interrogation of this pathway can also be performed to assess for outflow tract obstruction. The LV to truncal valve pathway can be seen well in the ME AV LAX view along with assessment of truncal stenosis and regurgitation (Fig. 12.26, Video 12.21) [50]. When available, the deep transgastric views (DTG LAX, DTG Sagittal) can provide visualization of the LV outflow tract and proximal RV to pulmonary artery conduit, as well as excellent angles of insonation for spectral

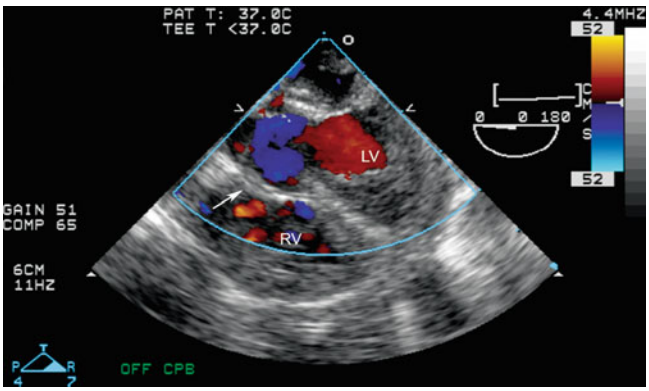


Fig. 12.25 Postop truncus arteriosus repair, seen from mid esophageal four chamber view with probe anteflexion. The ventricular septal defect patch is shown by the *arrow*; no residual defect is present by color flow imaging. *LV* left ventricle, *RV* right ventricle,

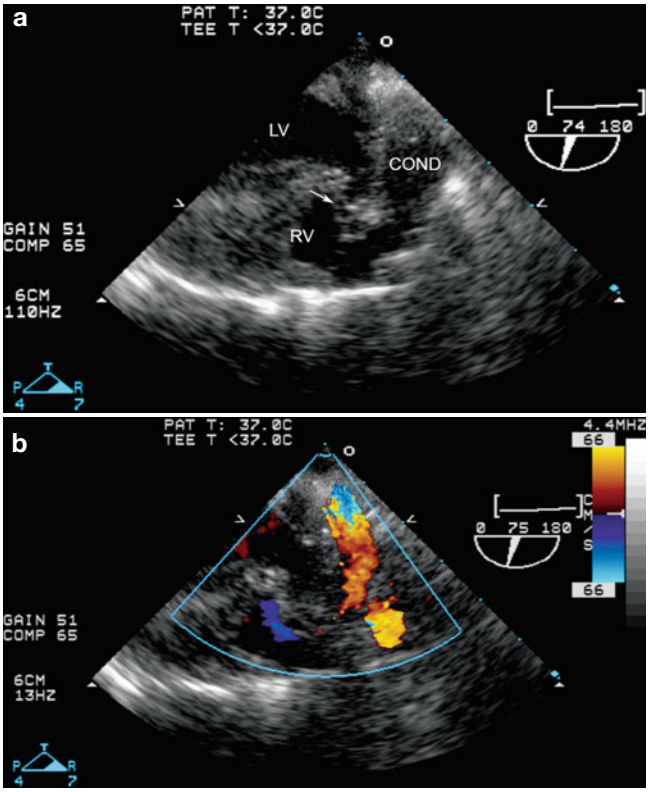


Fig. 12.26 Postop truncus arteriosus repair, seen from a modified mid esophageal aortic valve long axis view, multiplane angle about 75°, with imaging (**a**) and color flow Doppler (**b**). The ventricular septal defect patch (*arrow*) is seen as well as the origin of the conduit (*COND*) from the right ventricle (*RV*) to the pulmonary artery. *LV* left ventricle

Doppler assessment. The ME 4 Ch and ME Bicaval views can be used to assess the atrial septum, and the ME 4 Ch, ME RV In-Out, and ME 2 Ch views can facilitate the evaluation AV valve function.

Transposition of the Great Arteries

Anatomy

Transposition of the great arteries, also known as D-transposition of the great arteries, or complete or physiologically uncorrected transposition of the great arteries, is defined as an abnormal ventriculoarterial alignment whereby the aorta arises from the RV and the pulmonary artery from the LV. This results in ventriculoarterial *discordance*. However, there remains atrioventricular *concordance* (right atrium connected with RV, left atrium with LV). This anatomy produces a clinical picture of parallel circulations, in which deoxygenated systemic venous blood returns predominantly back to the aorta, and oxygenated pulmonary venous blood predominantly back to the pulmonary arteries. Thus these patients are usually cyanotic, sometimes profoundly so. Transposition of the great arteries is generally seen in combination with visceroatrial situs solitus and a D-ventricular loop (the 'normal' ventricular loop for situs solitus), hence the widely used abbreviation D-TGA, which will also be used for the remainder of this chapter. In its most common form, subaortic conus is present with absence of subpulmonary conus; thus there is mitral valve to pulmonary valve fibrous continuity. However, the conal anatomy may vary: in a series of 117 patients with D-TGA and VSD, subaortic conus was present in almost 90 % of patients, while bilateral conus, subpulmonary conus only and bilaterally deficient conus (extremely rare) were present in the remaining 10 % of patients [51]. In most patients with D-TGA, the aorta is spatially oriented anterior and rightward to the pulmonary artery [52]. In some cases the great vessels are side by side. In more unusual cases, the aorta is leftward or posterior to the pulmonary artery. Also, this entity occurs rarely in the setting of visceroatrial situs inversus [53]. For the purposes of this chapter, we will address the most common anatomic manifestation of D-TGA. Those familiar with the nomenclature of Van Praagh will recognize this as transposition of the great arteries with cardiac segments {S,D,D}: situs solitus of the atria, D-ventricular loop and D-position of the great arteries (with an anterior and rightward aorta) [54].

D-TGA occurs in three principal anatomic manifestations: (a) with an intact ventricular septum; (b) with a VSD; (c) with a VSD and LV outflow tract obstruction. D-TGA with an intact ventricular septum and no other cardiac abnormalities (other than a patent foramen ovale or patent ductus arteriosus) is the most frequent anatomic manifestation of D-TGA, occurring in approximately 50 % of all patients. Of the remaining 50 %, the most commonly associated cardiac defect is a VSD, occurring in 40–45 % of all patients. Any type of VSD is possible in D-TGA; the most common type is the perimembranous/conoventricular defect (33 %), but other types are possible including muscular (27 %), malalign-

ment (30 %), AV canal (5 %), and conal septal hypoplasia (5 %) [55]. When there is malalignment of the conal septum in D-TGA, two anatomic subtypes are seen: (1) anterior malalignment results in subaortic narrowing in association with distal arch obstruction, (2) posterior malalignment results in LV outflow tract obstruction consisting of subpulmonic obstruction and often, valvar pulmonary stenosis. The combination of VSD and LV outflow tract obstruction is seen in about 10–12 % of D-TGA patients; isolated LV outflow tract obstruction (without a VSD) occurs in only about 5 % of all patients [55, 56].

Coronary anatomy in D-TGA is variable. The so called "usual" coronary anatomy occurs in approximately two thirds of cases: the right coronary artery (RCA) arises from the right-sided sinus of Valsalva and the left main coronary artery (LCA) (anterior descending coronary and circumflex artery) arises from the left-sided sinus of Valsalva. In approximately 20 % of cases, the circumflex artery comes off the RCA and courses posterior to the pulmonary artery. Rarer anatomical subtypes include single RCA, single LCA, or inverted coronaries. In some patients, a portion of the coronary artery may take an intramural course in the wall of the aorta. This anatomy may complicate surgical repair [57].

Other cardiac anomalies that can be seen with D-TGA include aortic arch abnormalities such as coarctation (seen in about 5 % of all patients), as well as tricuspid and mitral valve abnormalities which have been reported in 20–30 % of autopsy series, though most of these AV abnormalities are functionally insignificant [55]. However AV abnormalities must be evaluated carefully, particularly when a VSD is present: overriding or straddling AV valve tissue can present problems from a cardiac surgery standpoint. Left juxtaposition of the atrial appendages (discussed above) occurs in about 2–5 % of D-TGA patients; when present, it often heralds more significant pathology such as dextrocardia, RV hypoplasia, and tricuspid valve stenosis or atresia [56].

Genetic causes of D-TGA are poorly defined and likely multifactorial. Environmental factors such as maternal diabetes or fetal exposure to retinoic acids have been implicated. In contrast to other conotruncal defects, isolated D-TGA is not usually linked to genetic syndromes such as 22q11.2 deletion syndrome [58].

TEE Evaluation

There are pertinent associated cardiac lesions seen with D-TGA that need to be kept in mind when performing a TEE:

- Size, location, and shunting pattern across the atrial communication
- Size and location of VSD(s), if present, and shunting pattern across the defect(s). It is important to remember that, in D-TGA/VSD and no pulmonary outflow obstruction, VSD shunting pattern is generally from the RV to LV, which is the reverse of that expected with a VSD and normally related (nontransposed) great arteries.
- Anatomy and size of AV valves.
- Presence and severity of outflow tract obstruction.
- Semilunar valve anatomy.
- Coronary anomalies, particularly if the arterial switch operation is being considered.

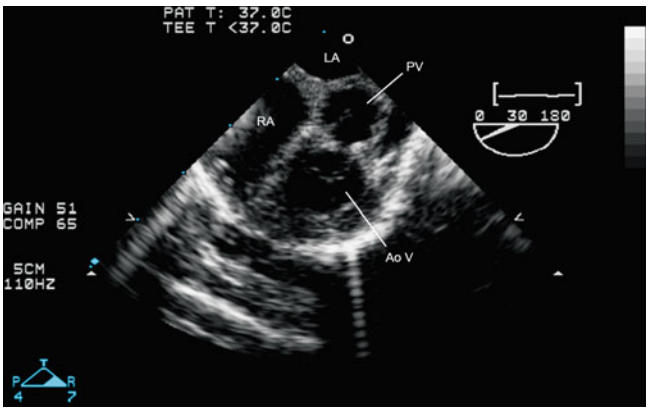


Fig. 12.27 D-transposition of the great arteries. Using a mid esophageal aortic valve short axis view, both semilunar valves are seen *en face*, with the aortic valve (*AoV*) anterior and rightward to the pulmonary valve (*PV*). *LA* left atrium, *RA* right atrium

In D-TGA with an intact ventricular septum, a pre-operative TEE is rarely needed for diagnostic assessment. However, it can still be very useful for confirmation of the pre-operative findings, for the assessment of AV and semilunar valve function, for evaluation of ventricular performance prior to surgical intervention, and as a reference for comparison with the post-operative examination. The ME 4 Ch sweep provides a good global view of these aspects of D-TGA evaluation. In some cases, the coronary anatomy can be well-delineated by TEE. The ME AV SAX view with multiplane angle 30–45° can be used to determine semilunar valve anatomy (Fig. 12.27, Video 12.22). Because both great vessels are parallel, rather than the normal criss-cross orientation, both semilunar valves can generally be seen in the same plane. The coronary artery origins can sometimes be seen from this view (Fig. 12.28, Video 12.23). Using a modified ME 4 Ch view with slight probe withdrawal and anteflexion, the outflow tracts, semilunar valves and great arteries can be shown (Fig. 12.29, Video 12.24); these structures can also be displayed in an orthogonal manner using the ME RV In-Out, ME 2 Ch, ME LAX, and ME AV LAX views (Fig. 12.30, Video 12.25). A very abnormal pulmonary valve and/or narrowed pulmonary outflow tract may preclude the ability to perform an arterial switch procedure (Fig. 12.31, Video 12.26). In such cases, a Rastelli procedure (discussed below) might be necessary; the pre-operative TEE can be used to help determine the pathway from the LV to the aorta and to find the best view to evaluate this after surgery.

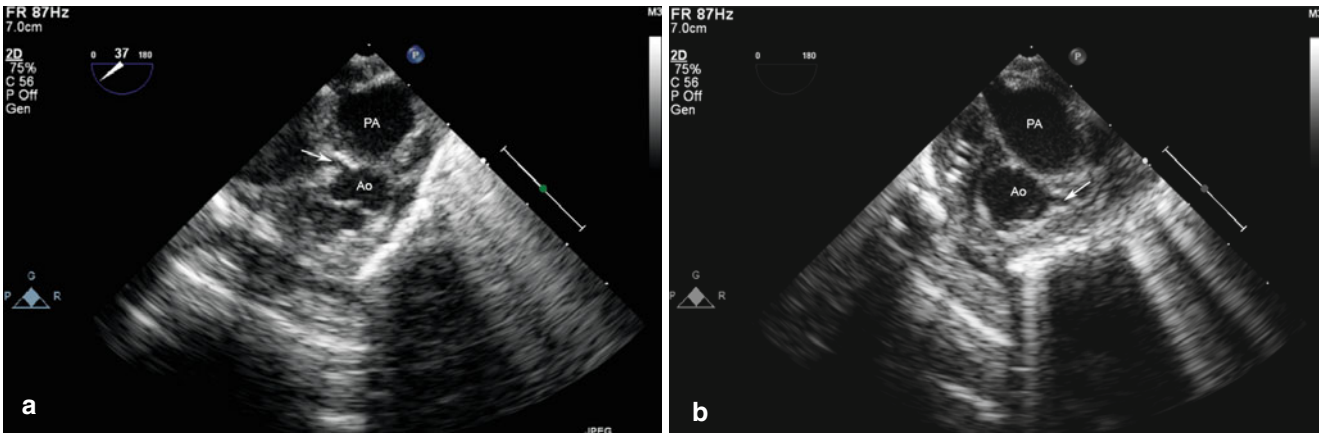


Fig. 12.28 Origins of the right (**a**) and left (**b**) coronary arteries (*arrows*) from the opposite aortic valve sinuses facing the pulmonary artery in transposition of the great arteries, as seen from the mid esophageal aortic valve short axis view. This coronary pattern is the most common type seen in this cardiac defect. *Ao* aorta, *PA* pulmonary artery

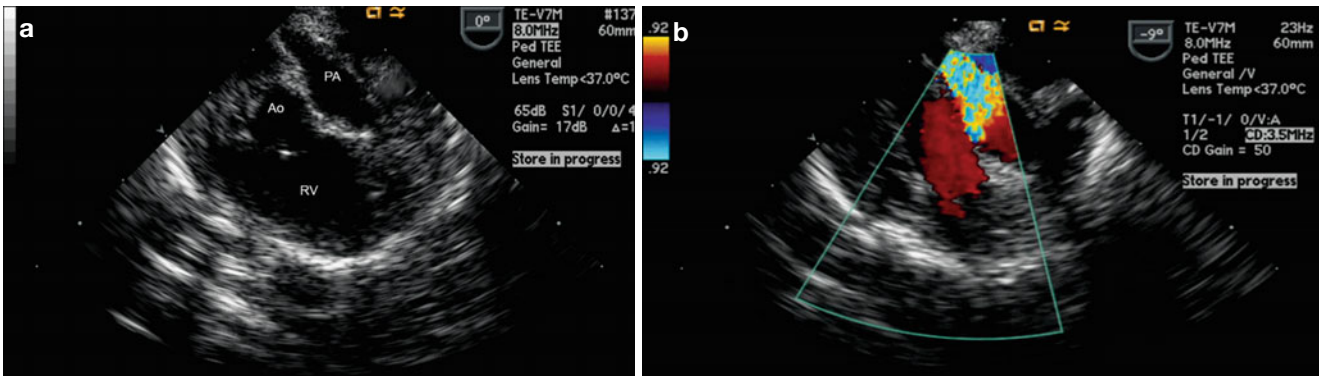


Fig. 12.29 Transposition of the great arteries, as viewed from a mid esophageal view. With anteflexion and slight probe withdrawal from the mid esophageal four chamber view, both great arteries are seen in parallel, with the aorta (*Ao*) arising from the right ventricle (*RV*), and the pulmonary artery (*PA*) from the left ventricle (**a**). Color flow Doppler shows continuous flow in the PA from ductal shunting (**b**)

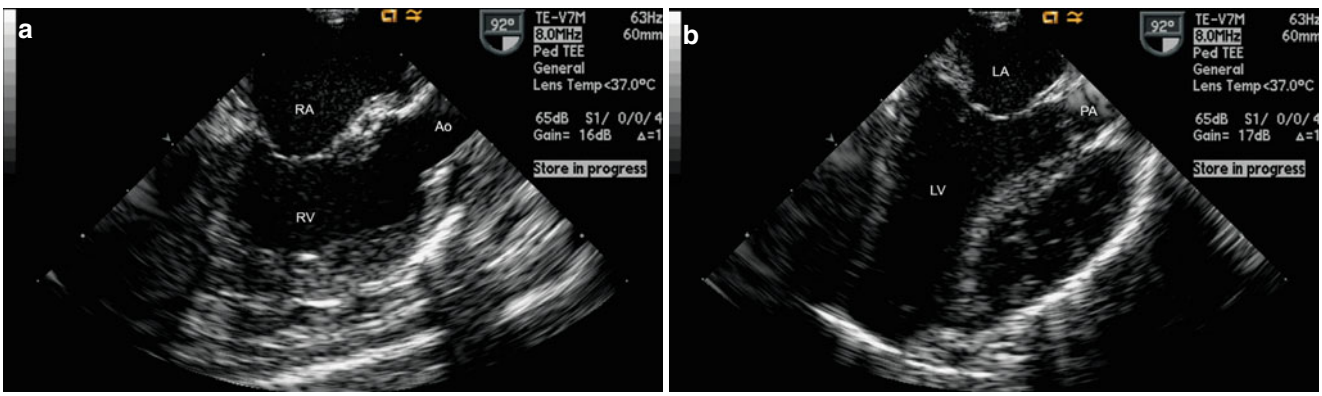


Fig. 12.30 Transposition of the great arteries, as viewed from a mid esophageal right ventricular inflow-outflow/long axis view, multiplane angle about 90°. The aorta arises from the right ventricle (**a**), and the pulmonary artery from the left ventricle. (**b**) Both great arteries have the same alignment, and therefore take a parallel course. *Ao* aorta, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle

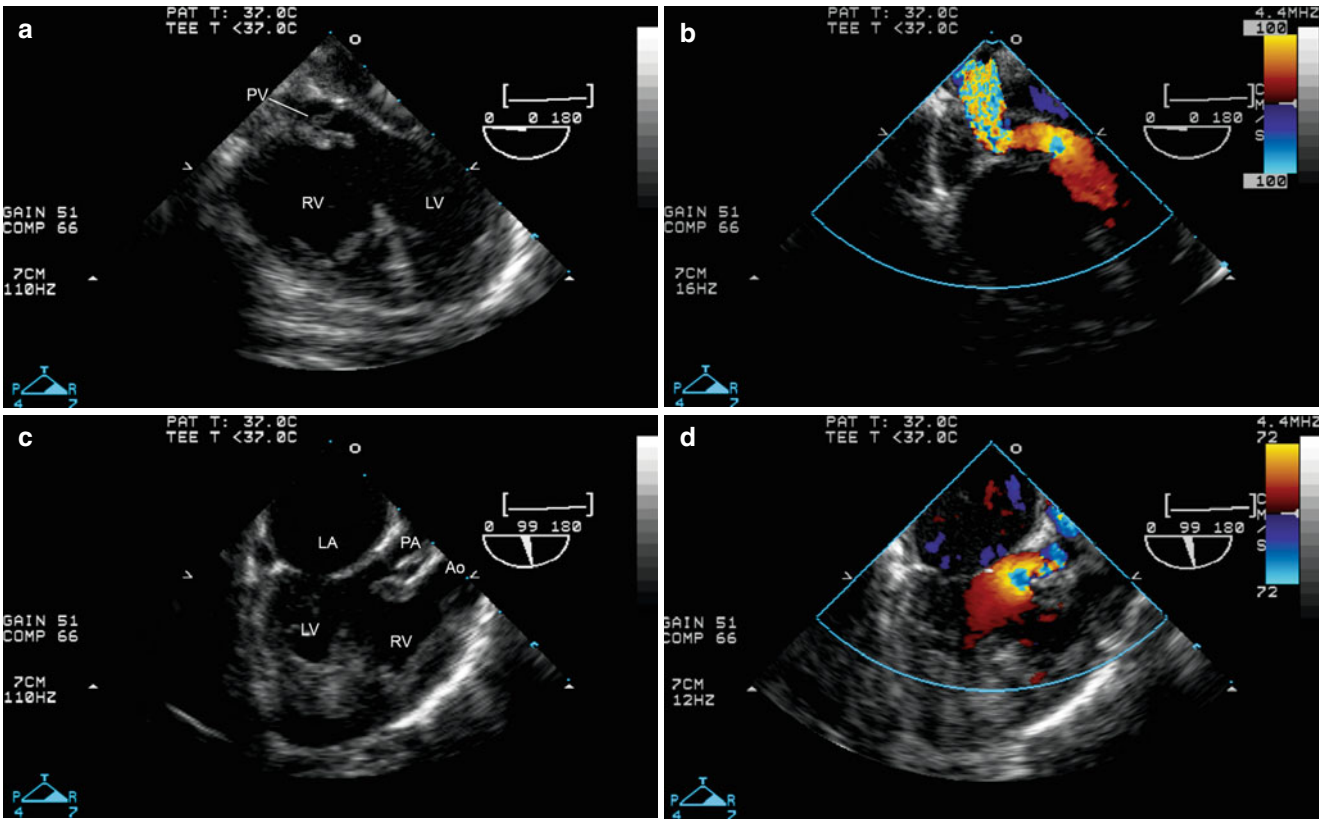


Fig. 12.31 D-transposition of the great arteries, with a large ventricular septal defect and pulmonary outflow obstruction, as seen from a modified mid esophageal four chamber view (**a**, **b**) and mid esophageal long axis view (**c**, **d**). Posteriorly malaligned infundibular septum produces prominent subpulmonary narrowing, and turbulence is seen by color flow Doppler across the pulmonary outflow tract. *Ao* aorta, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *PV* pulmonary valve, *RV* right ventricle

Surgical Considerations

Surgical intervention for D-TGA has progressed from the atrial switch operation to the arterial switch operation over the course of the last 25 years. The atrial switch procedure (variations include the Mustard and the Senning procedures) is now performed in very rare circumstances (late presentation, significantly abnormal pulmonary valve, isolated ventricular inversion, and in those with congenitally corrected TGA, see below). In this procedure, superior and inferior vena caval venous return are baffled directly to the mitral valve, while the pulmonary venous blood is channeled into the tricuspid valve. This procedure has been abandoned as the favored surgical option for D-TGA because of many long-term complications including: RV failure (the RV must act as the systemic ventricle), tricuspid regurgitation, as well as sinus node dysfunction and significant atrial arrhythmias from the extensive atrial suture line [59].

The arterial switch operation is now the preferred corrective surgical repair in those patients with isolated D-TGA or in those with a VSD but without significant narrowing of the pulmonary outflow tract. It is also the suitable surgical intervention for patients with the Taussig-Bing-type of DORV where there is a subpulmonary VSD (see section "Double Outlet Right Ventricle"). In this procedure, both great arteries are transected above their respective sinuses and the vessels are "switched", restoring the normal ventricular-arterial connection. The pulmonary artery is brought in front of the aorta by means of the "Lecompte maneuver" (both the right and left pulmonary artery lie anterior to the aorta) and the coronary arteries are translocated to the neo-aorta [60]. Associated defects are addressed at the time of surgery, included intracardiac communications, if present [61].

In cases of D-TGA with posterior malalignment VSD and LV outflow tract stenosis (subpulmonary/pulmonary valve stenosis), an arterial switch operation is not feasible. For these cases, other surgical options may be considered including the Rastelli operation. In this procedure an intracardiac tunnel is created to allow ejection of left ventricular blood through the VSD into the rightward aorta, while at the same time eliminating ventricular level shunting, and a conduit is placed from the RV to the pulmonary artery [61].

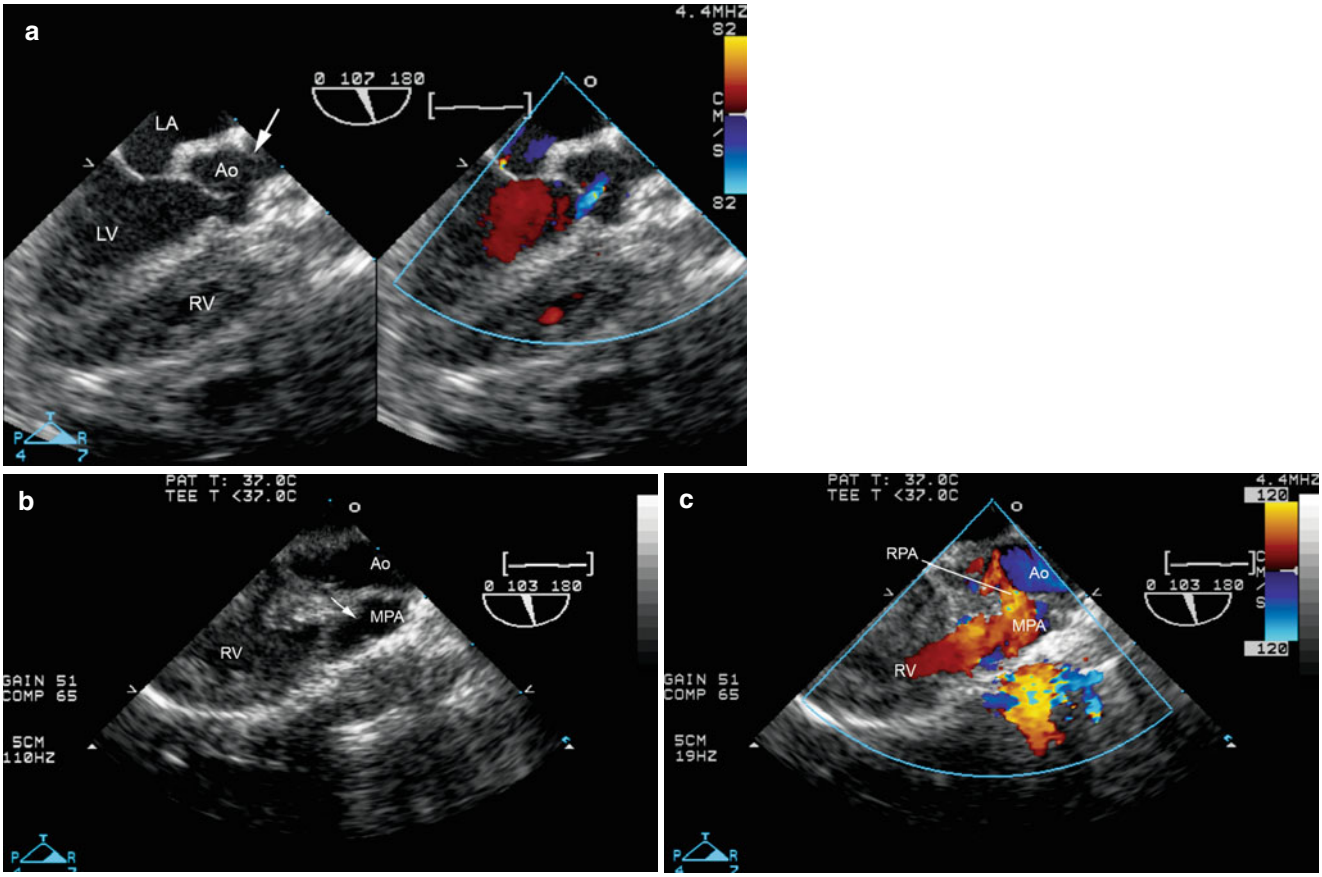


Fig. 12.32 Postoperative study following arterial switch operation. (**a**) From the mid esophageal long axis view, the figure shows the neo-aortic valve with a small amount of regurgitation. The aortic anastomosis is shown by the *arrow*. (**b**) From the mid esophageal ascending aorta long axis view, the figure shows the pulmonary artery anastomosis (*arrow*); the branch pulmonary arteries arise just above (superior to) the anastomosis. In (**c**) the right pulmonary artery is seen. *Ao* aorta, *LA* left atrium, *LV* left ventricle, *MPA* main pulmonary artery, *RPA* right pulmonary artery, *RV* right ventricle

TEE Evaluation Following Cardiac Surgery

Arterial Switch Operation

Postoperative TEE for the arterial switch operation has several goals including: (1) assuring patent right and left ventricular outflow tracts after transection and "switch" of the great arteries, (2) determination of regional and global systolic ventricular function after transfer of the coronary arteries, (3) assessment for residual intracardiac shunts (if present pre-operatively), (4) assessment of semilunar (particularly neo-aortic valve) competency. The ME 4 Ch view provides a good cross section for global assessment of ventricular performance. The TG Basal SAX and transgastric two chamber (TG 2 Ch) views provide excellent images to determine if there are regional wall motion abnormalities consistent with coronary obstruction during the transfer to the neo-aorta. The ME RV In-Out and ME AV LAX views demonstrate the right and left ventricular outflow tracts (Fig. 12.32, Video 12.27). The supravalvar regions are seen as the probe is withdrawn to the upper esophagus; this is important because the suture lines can cause obstruction or semilunar valve regurgitation might be present. Often the branch pulmonary arteries are difficult to image because they are the most anterior structures, however, withdrawal to the UE PA LAX view can visualize the branch pulmonary arteries anterior to, and straddling the ascending aorta (Fig. 12.33, Video 12.28).

Color Doppler can be used to assess for AV valve regurgitation, which can occur in the presence of ventricular dysfunction. Coronary ischemia can also result in papillary muscle dysfunction leading to mitral regurgitation. If tricuspid regurgitation is present, an RV systolic pressure estimate can be performed in the ME 4 Ch view to determine the pulmonary artery pressure (assuming that the branch pulmonary arteries are unobstructed). Pulsed and continuous wave Doppler are used to assess the severity of outflow tract obstruction, if this is present. The deep transgastric views (DTG LAX, DTG Sagittal), when available, are particularly helpful in this regard. Neo-aortic and neo-pulmonary valve function should be determined, because valve distortion can occur with the procedure. With the color flow at low-scale,

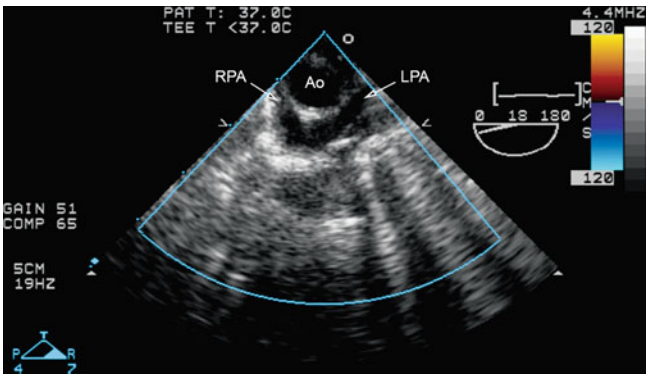


Fig. 12.33 Postoperative study following arterial switch operation, showing the branch pulmonary arteries from the upper esophageal pulmonary artery long axis view. A Lecompte maneuver was performed so the main and branch pulmonary arteries are situated anterior to the aorta (*Ao*). *LPA* left pulmonary artery, *RPA* right pulmonary artery

coronary blood flow in diastole can also be seen in some cases.

In the longer term, TEE provides information on potential sequelae of the arterial switch operation: supravalvar pulmonary stenosis, supravalvar aortic stenosis, dilation of the neoaortic root, and neoaortic regurgitation. At midterm follow-up, trivial to mild neoaortic regurgitation develops in the majority of patients. A smaller percentage of patients develop more significant regurgitation. Importantly, the neoaortic root frequently exhibits progressive dilation that is out of proportion to somatic growth [62]. The extent of this dilation into adulthood and the risk for dissection or rupture remain to be seen.

Rastelli Procedure

The postoperative TEE for the Rastelli procedure differs in its objectives from the arterial switch operation. Goals for this study include (1) surveillance for residual VSD, (2) assurance of an unobstructed pathway from the LV to the aorta, (3) assessment of the function of the RV to pulmonary artery conduit. Multiple sweeps are used to assess for a residual VSD. It is important to sweep from the most posterior aspect of the heart to the great vessels to assure that a residual VSD is not missed. This should be performed in the ME 4 Ch, ME RV In-Out, and ME AV SAX views. Intramural VSDs (discussed under TOF) can occur with this procedure as well. In the ME RV In-Out view, the conduit can be assessed in the most anterior region. Withdrawal of the probe to the ME Asc Ao LAX and UE Ao Arch SAX will demonstrate the most distal portion of the conduit as it is connected to the branch pulmonary arteries. Rotating the multiplane angle to 0–10°, using the ME AV SAX and UE PA LAX views, the right pulmonary artery is usually seen well. The left pulmonary artery can be difficult to visualize with TEE, but it can be seen in some patients using the UE PA LAX view and leftward probe rotation. The ME AV LAX view is an important view after a Rastelli procedure to assure that there is an unobstructed pathway from the LV to the aorta. If the angle of interrogation is appropriate, pulsed and continuous wave Doppler can also be performed in this view. The angle to image the pathway may need to be altered depending on the relationship of the aorta to the LV. As with the other outflow tract abnormalities mentioned above, DTG LAX and DTG Sagittal views can provide important additional information of the LV to aorta pathway, as well as excellent angles for spectral Doppler interrogation. Alternatively, the transgastric long axis (TG LAX) view, multiplane angle 90°-120°, is also excellent to evaluate this pathway and also allows for spectral Doppler evaluation. This view can be used in older patients and/or when the DTG views are not available (see Chap. 4 for a detailed description of the TG LAX view).

Long-term assessment of the Rastelli operation primarily involves evaluation of conduit patency and regurgitation. Almost all patients who undergo the Rastelli operation during infancy or childhood require reoperation for conduit failure later in life [63]. Subaortic obstruction can develop late after repair as well.

Atrial Switch Operation

Prior to the 1980s, surgical management for D-TGA was achieved using a procedure that rerouted the systemic venous return to the LV, and pulmonary venous return to the RV. These procedures were known as the atrial switch operation or atrial baffle procedure, and venous flow redirection was accomplished by the use of two principal surgical variations: the Mustard and Senning procedures [56, 61]. TEE is a very important tool to assess the long-term outcome after the atrial switch. Many regions of concern are difficult to visualize using TTE, particularly the venous baffles. Surveillance by TEE in these patients provides superior visualization, particularly in older patients with marginal TTE imaging [64]. In addition, TEE is frequently used in patients with atrial flutter or fibrillation prior to cardioversion to assure that there is no thrombus present. Other long-term complications after the atrial switch that can be diagnosed using TEE include: residual intracardiac shunts, baffle leaks, pulmonary or systemic venous pathway obstruction, tricuspid and/or mitral valve regurgitation, thrombi/vegetations on pacing leads, right and/or left ventricular outflow tract obstruction, and semilunar valve insufficiency [59]. Contrast injection into the venous circulation during the TEE can help discern baffle leaks. TEE guidance can be used for device closure of such leaks (Figs. 12.34 and 12.35, Videos 12.29 and 12.30). The TG Basal SAX will give an excellent view of overall ventricular performance. It is important to keep in mind that the ventricular septum will be shifted toward the LV because of the systemic RV pressure. The pulmonary venous baffle can best be viewed in ME 4 Ch view (Fig. 12.36, Video 12.31) and with the probe rotated counterclockwise to the left the entrance of

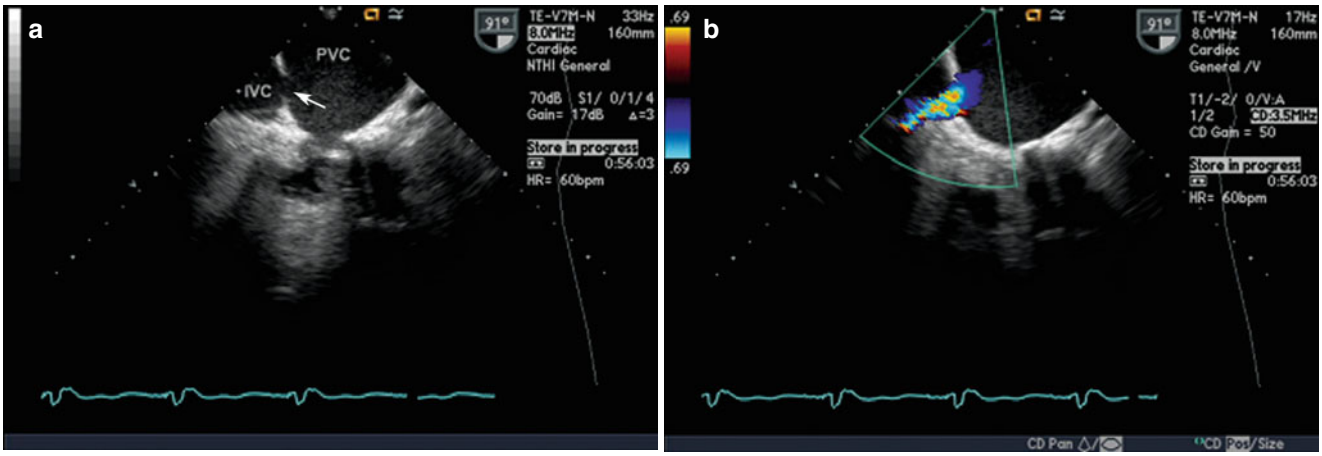


Fig. 12.34 Modified mid esophageal bicaval view with the probe advanced toward the liver in an adult who has undergone an atrial switch operation for D-transposition of the great arteries. (**a**) A communication (*baffle leak*) between the inferior vena cava (*IVC*) and pulmonary venous channel (*PVC*) is seen by two-dimensional imaging (*arrow*). (**b**) The color flow demonstrates flow from the pulmonary venous channel to the IVC (*left* to *right shunt*)

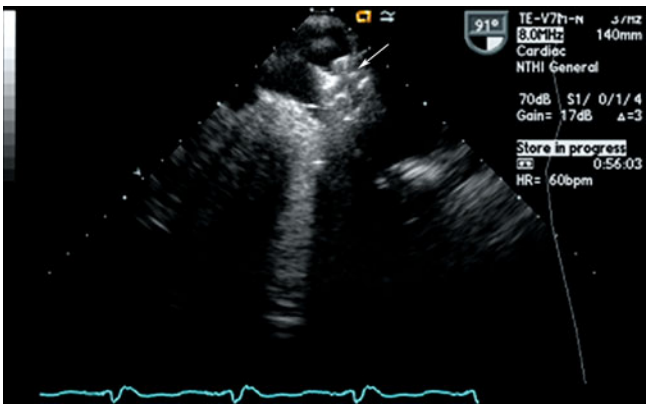


Fig. 12.35 Modified mid esophageal bicaval view with the probe advanced toward the liver in the same patient as Fig. 12.34 with the atrial switch and baffle leak. This image is obtained during transcatheter device deployment. The image demonstrates an Amplatzer device (*arrow*) just prior to its release

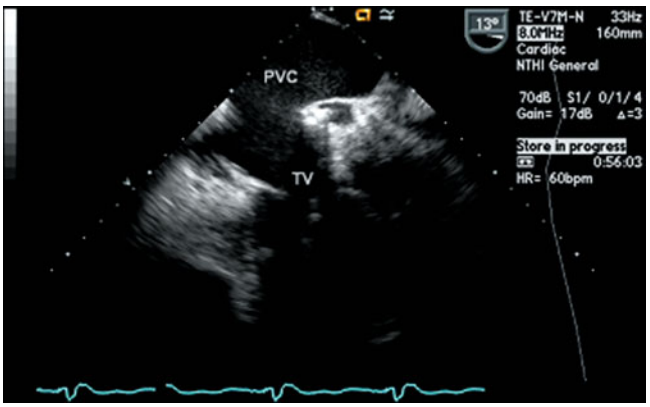


Fig. 12.36 Mid esophageal four chamber view of a patient after atrial switch operation demonstrating the pulmonary venous channel (*PVC*) as it makes its way to the tricuspid valve (*TV*). In this patient, the pathway is unobstructed

the left pulmonary veins can be seen. In the same plane, tilting the probe clockwise, toward the right, the entrance of the right pulmonary veins can be seen. The LE Situs SAX allows identification of the IVC within the liver. Withdrawal of the TEE probe shows the IVC entering into the inferior limb of the intra-atrial baffle and subsequent counterclockwise rotation connects the baffle to the mitral valve (ME 4 Ch view). Turning the multiplane angle to 90°, the superior limb of the systemic venous pathway is best seen in ME Bicaval view (Fig. 12.37, Video 12.32). With rotation of the probe toward the left, the baffle can be seen as it leads to the mitral valve. Obstruction is more common in the superior limb of the baffle because of the remnant of septum secundum along the pathway (Figs. 12.38 and 12.39, Video 12.33). Some patients have pacemaker wires in the superior limb that further contribute to narrowing along this venous pathway (Figs. 12.37, 12.38, and 12.39, Videos 12.32 and 12.33).

Congenitally Corrected Transposition of the Great Arteries

Anatomy

Congenitally corrected transposition of the great arteries (CCTGA), also known as physiologically corrected or L-transposition of the great arteries, is a cardiac defect in which there is both atrioventricular (AV) *and* ventriculoarterial discordance. The AV discordance results from ventricular inversion, in which the looping is the opposite of normal: in situs solitus, the RV ventricle assumes an L-loop (left hand) configuration, and in situs inversus, a D-loop (right hand). Thus, the right atrium empties into the morphologic LV, and the left atrium into the morphologic RV. In addition, the great arteries are transposed so that there is ventriculoarterial

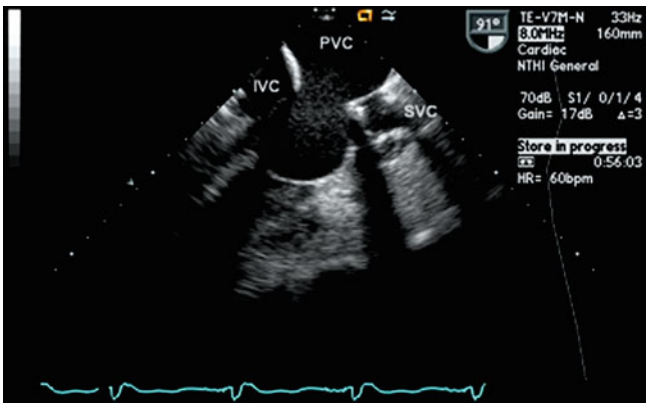


Fig. 12.37 Mid esophageal bicaval view with probe turned clockwise toward the patient's right side. This image is from a patient who has superior systemic venous limb obstruction after atrial switch operation. In this view, both inferior (*IVC*) and superior (*SVC*) limbs of the systemic venous channel are seen with the pulmonary venous channel (*PVC*) coursing in between. Pacing wires (echobright structures) are seen in the superior limb

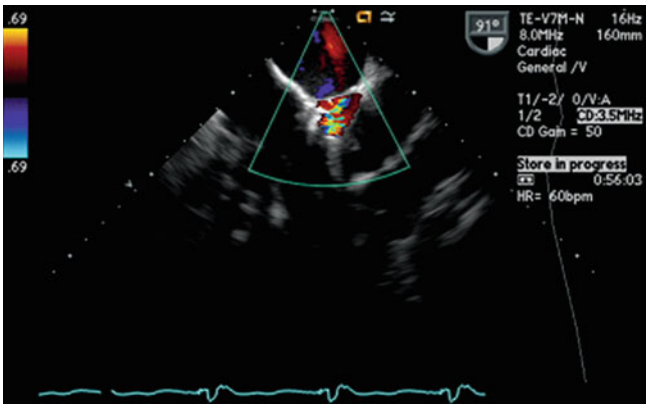


Fig. 12.38 Modified mid esophageal right ventricular inflow-outflow view using color Doppler in the same patient as Fig. 12.37 after atrial switch operation with superior limb obstruction. Aliasing of the color flow is seen in the superior limb of the channel

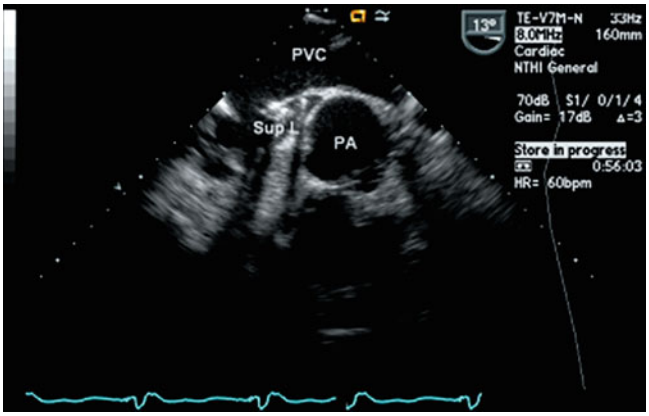


Fig. 12.39 Mid esophageal four chamber view in the same patient as Figs. 12.37 and 12.38 who has superior limb obstruction after atrial switch operation. This image demonstrates significant narrowing of the superior limb of the systemic venous channel as it crosses posterior to the pulmonary artery (*PA*). Transvenous pacing wires are seen (echobright structures) within the superior limb of the systemic venous channel (*Sup L*), which likely contribute to the obstruction. *PVC* pulmonary venous channel

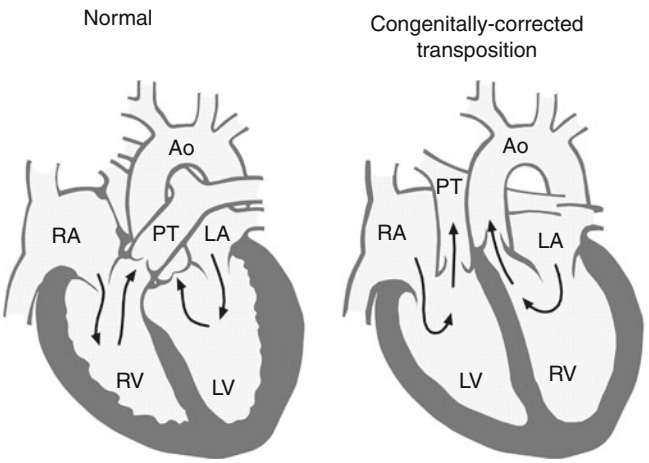


Fig. 12.40 Side by side schematic drawings of a normal heart and a heart with congenitally corrected transposition of the great arteries (*CCTGA*). The left diagram (*normal heart*) demonstrates normal atrioventricular (*AV*) and ventriculoarterial (*VA*) relationships; the right diagram (*CCTGA*) demonstrates both AV and VA discordance. In CCTGA, the normal great arterial relationships are lost; the aorta is situated anterior and leftward to the pulmonary trunk (*PT*), and the great arteries are oriented parallel to each other. *Ao* indicates aorta, *LA* morphological left atrium, *LV* morphological LV, *RA* morphological right atrium, *RV* morphological RV (From Warnes [65]; used with permission)

discordance: similar to D-TGA, the aorta arises from the morphologic RV, and pulmonary artery from the morphologic LV, and there is generally subaortic conus present with absence of subpulmonary conus, and mitral valve to pulmonary valve fibrous continuity. With situs solitus, great artery spatial positioning is such that the aorta is situated leftward and anterior to the pulmonary artery, and both great arteries are oriented parallel to each other (Fig. 12.40). Because of the "double discordance", CCTGA patients are physiologically corrected (systemic venous blood flows to the pulmonary artery, pulmonary venous blood to the aorta) and generally have normal systemic arterial oxygen saturation, unlike D-TGA. Most cases of CCTGA are seen with situs solitus, although CCTGA can definitely be seen with situs inversus. For the purposes of this chapter, we will address only the more common form of CCTGA with visceroatrial situs solitus. Again, those familiar with the nomenclature of Van Praagh will recognize this as transposition of the great arteries with cardiac segments {S,L,L}: situs solitus of the atria, L-ventricular loop, and L-position of the great arteries (aorta anterior and leftward to the pulmonary artery) [54].

While CCTGA can occur without other cardiac defects, this is usually not the case. Most cases of CCTGA have associated defects, which include the following:

- A VSD is present in approximately 80 % of patients [66]. It is usually a perimembranous defect but can occur in any

portion of the ventricular septum. The VSD can be variable in size and physiologic significance; when moderate to large, there can be hemodynamically significant left to right shunting and pulmonary overcirculation. With perimembranous VSDs, it is not unusual to have AV valve tissue and/or chordae (from either AV valve) attached to the crest of the ventricular septum, and sometimes confluent with tissue covering the VSD. This may have surgical implications.

- Left ventricular (pulmonary) outflow tract obstruction is seen in 40–50 % of patients [66]. It can be valvar, subvalvar, or both. Subvalvar pulmonary obstruction is usually the result of fibrous tissue originating from the membranous septum and attaching to the mitral valve, creating a fibrous collar. Subpulmonary stenosis can also be caused by an excrescence (or "windsock") of tissue protruding from the membranous septum into the pulmonary outflow tract [67, 68].
- Abnormalities of the systemic AV (tricuspid) valve have been described in 90 % of patients evaluated by autopsy [66]; these usually manifest as an Ebstein-type malformation of the valve, characterized by inferiorly displaced leaflets with impaired mobility. The valve anatomy differs from that of a true right sided Ebstein anomaly: there is little to no atrialized portion of the RV, adherence of the septal/posterior leaflets is limited, and the anterior leaflet is not sail-like [65, 69, 70]. Other tricuspid valve abnormalities, such as deficient leaflet tissue and thickened valve leaflets, as well as abnormal papillary musculature, can be seen in CCTGA [71]. The abnormal tricuspid valve presents with a variable degree of valvar regurgitation. In some patients, despite a very abnormal appearance to the valve, the degree of regurgitation can be surprisingly mild.
- Other abnormalities seen in association with CCTGA include subaortic stenosis and atrial septal defects. Aortic insufficiency can develop and progress over time [72]. The coronary arteries are the reverse of normal: the anterior descending and circumflex arise from the right facing sinus of Valsalva, the "right" coronary artery from the left facing sinus of Valvalva. Patients with CCTGA are also at increased risk for spontaneous development of complete heart block, and this risk increases with time at the rate of approximately 2 % per year [65].

The clinical presentation of CCTGA is variable, depending upon whether associated cardiac lesions are present, and the severity of these lesions. If other cardiac defects are present, such as a moderate VSD or significant AV valve regurgitation, patients can present in childhood with signs/symptoms of congestive heart failure. If significant pulmonary outflow tract obstruction is present, patients can present with cyanosis. When CCTGA occurs without any associated cardiac lesions, patients can be asymptomatic for decades [73]. However in many of these patients, systemic AV valve regurgitation can develop with time and patients can eventually become symptomatic. Of concern is that, by the fourth and fifth decades of life, a significant proportion of patients with or without associated cardiac lesions will have developed congestive heart failure and systemic ventricular (RV) dysfunction. This complication is particularly seen in those patients with important tricuspid (systemic AV valve) regurgitation [72, 74]. Thus CCTGA is one of the cardiac defects commonly encountered by specialists who treat adults with congenital heart disease (Chap. 18).

The genetic basis of CCTGA is still unknown [75]. There appears to be an increased incidence among families [76, 77], with a recurrence risk given between 2.6 and 5.2 % [78].

TEE Evaluation

Evaluation of the CCTGA patient should begin with a careful segmental analysis. In order to establish the diagnosis of atrioventricular and ventriculoarterial discordance, it is first necessary to identify the morphology of the three principal segments (atria, ventricles, great arteries), and then determine the manner in which these individual segments connect to each other (Chap. 4). This analysis can be further complicated by the high incidence of cardiac malposition in CCTGA patients (mesocardia in 15 %, dextrocardia in 30 %), despite the fact that most of these patients will have visceroatrial *situs solitus*. Thus the methods outlined in Chap. 4 for segmental evaluation hold an even greater importance in the assessment of these patients.

There are particular anatomic features of CCTGA that should be evaluated by TEE. These include the following:

- Anatomy of the AV valves, particularly the systemic (tricuspid) AV valve.
- Ventricular septum, and if a VSD is present, its size and location. In many CCTGA patients the ventricular septum is oriented in a straight sagittal (antero-posterior) plane, which differs from the angled orientation of the septum in normal patients.
- Evaluation of the pulmonary outflow tract, particularly in regard to subpulmonary and pulmonary valve obstruction.
- Relationship of the AV valves to the VSD (if present) and any pulmonary outflow tract obstruction (fibrous tissue, excrescences, etc.)
- Aortic valve anatomy.
- Atrial septal defect.
- Systemic and pulmonary venous return.
- Morphology and size of the ventricles.

Atrial situs can be evaluated using the techniques outlined in Chap. 4. The LE Situs SAX and LE IVC LAX views are useful for localizing the entrance of the IVC and coronary sinus, two markers that identify the morphologic right atrium. This evaluation also becomes important in identifying the location of the inferior systemic venous pathway when a double switch operation (discussed below) is planned. The ME 4 Ch view and its modifications (clockwise/counterclockwise rotation), is the most useful TEE view for evalua-

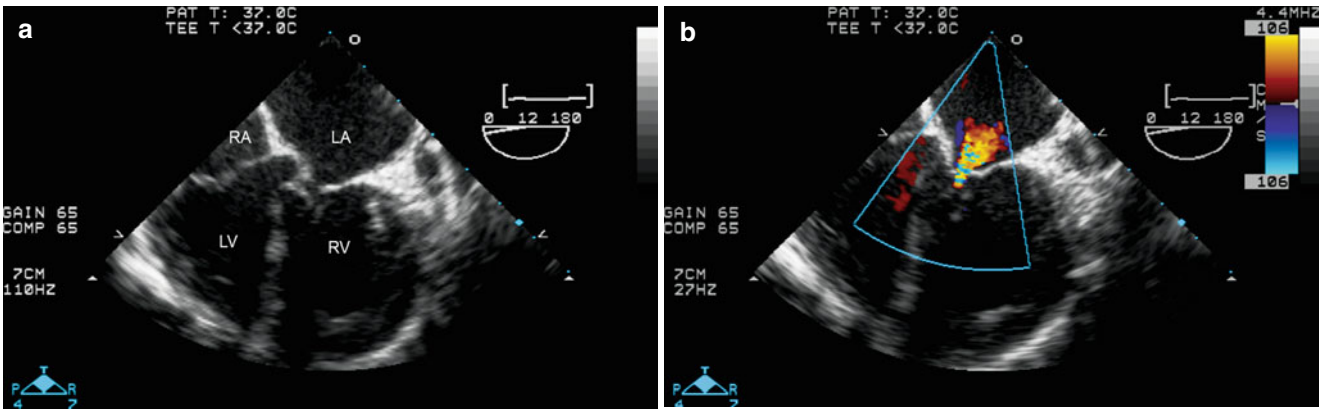


Fig. 12.41 Congenitally corrected transposition of the great arteries. Using a mid esophageal four chamber view (**a**), marked inferior displacement of the septal leaflet of the left sided tricuspid valve is seen, compared with the right sided mitral valve. Incomplete coaptation of this valve results in a mild to moderate degree of tricuspid insufficiency, as noted in (**b**). *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

tion of CCTGA, as a number of important structures can be visualized: (1) the atrial appendages and systemic/pulmonary venous return; (2) the atrial septum; (3) the AV valves and the offset between tricuspid and mitral valves (reverse of normal); (4) the inlet, membranous, and trabecular portions of the ventricular septum; (5) the pulmonary outflow tract and, with probe withdrawal and slight anteflexion, the pulmonary valve. Thus the ME 4 Ch view allows evaluation of the defects most commonly seen in CCTGA [79]. The degree of inferior tricuspid valve displacement is very well shown with the ME 4 Ch, and color flow Doppler will provide an excellent profile of the degree of AV valve insufficiency (if any) (Fig. 12.41, Video 12.34). Further views of the left sided tricuspid valve can be provided with rotation of the multiplane angle between 0° and 90°, along with slight counterclockwise rotation, to obtain the equivalent of the ME 2 Ch and ME LAX views (Fig. 12.42, Video 12.35). The ventricular septum will also be well seen using the ME 4 Ch view. Again, given the straight sagittal orientation of the ventricular septum in many CCTGA patients, the ME 4 Ch, with varying amounts of retroflexion and anteflexion, provides the best sweep of this structure. This view also provides excellent depiction of perimembranous VSDs, along with detail regarding AV valve chordal attachments to the crest of ventricular septum and/or straddling across the septum (Fig. 12.43, Video 12.36). Additional views of the ventricular septum and ventricular morphology can be provided by the TG Basal and TG Mid SAX views; the ventricular inversion will be apparent with the two LV papillary muscles on the patient's right, and the trabeculated RV on the patient's left (Fig. 12.44, Video 12.37). In the transgastric position, additional views of the tricuspid valve can be obtained by rotating the multiplane angle to 90° and slight counterclockwise rotation to obtain the TG 2 Ch view.

The pulmonary outflow tract can be evaluated with several views. It can be visualized with the ME 4 Ch view and

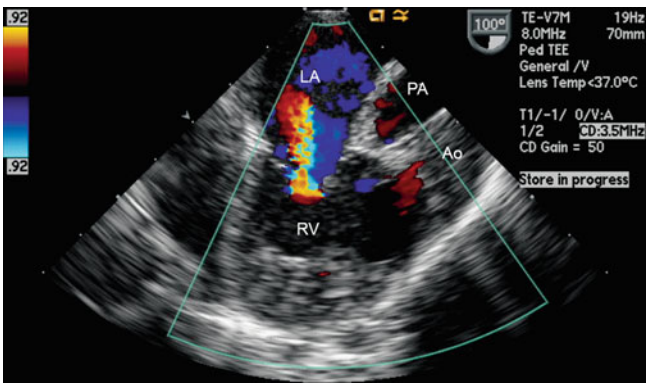


Fig. 12.42 Congenitally corrected transposition of the great arteries. Using the mid esophageal long axis view, the left sided tricuspid valve is seen, with significant regurgitation. *Ao* aorta, *LA* left atrium, *PA* pulmonary artery, *RV* right ventricle

slight probe withdrawal, as well as a small amount of probe anteflexion. However, the best views of the outflow tract will be obtained by a small amount of clockwise rotation as well as rotation of the multiplane angle to 90°–110°. This sagittally oriented view more fully displays the long axis of the pulmonary outflow tract, and provides anatomic detail regarding subpulmonary stenosis (if present) [67, 80, 81], including the distance between area of stenosis and the pulmonary valve (Fig. 12.45, Video 12.38). This view also provides excellent color flow Doppler visualization for evaluation of outflow tract stenosis, and fair to good angles for spectral Doppler quantification. Supplementary cross-sections from the transgastric (TG LAX) and deep transgastric views (DTG LAX, DTG Sagittal) offer additional angles for Doppler interrogation of the pulmonary outflow tract (Fig. 12.46, Video 12.39).

Like D-TGA, the great arteries of CCTGA arise in parallel fashion from the ventricles. From the ME 4 Ch view, a sweep using both probe withdrawal and anteflexion will

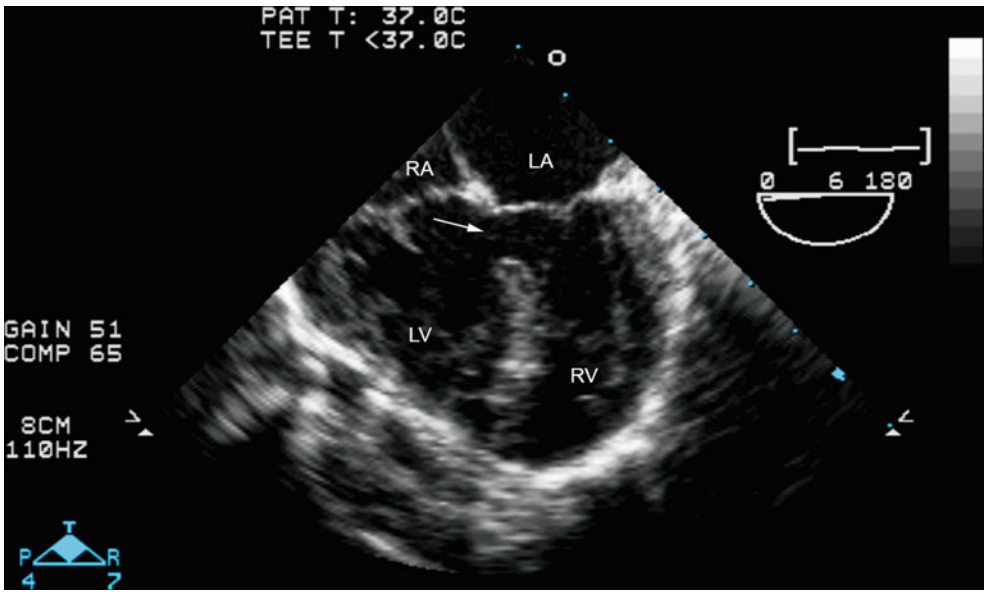


Fig. 12.43 Mid esophageal four chamber view shows a large perimembranous ventricular septal defect (*arrow*) in a patient with congenitally corrected transposition of the great arteries. Note the left sided tricuspid valve chordal attachment to ventricular septum, as well as the presence of a moderator band in the left sided right ventricle (*RV*). *LA* left atrium, *LV* left ventricle, *RA* right atrium

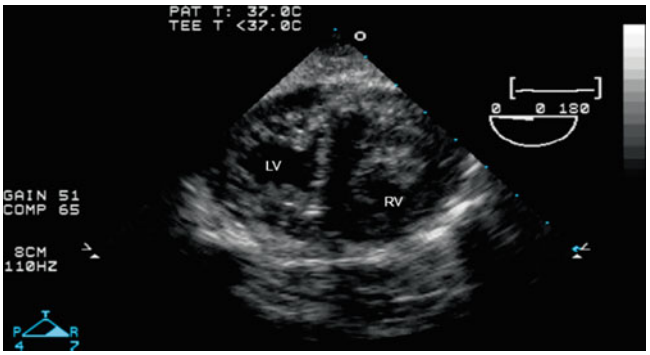


Fig. 12.44 Congenitally corrected transposition of the great arteries. Transgastric mid short axis view shows the inverted ventricles in cross section. Note the smooth walled septal surface in the left ventricle (*LV*), and the prominent moderator band in the right ventricle (*RV*)

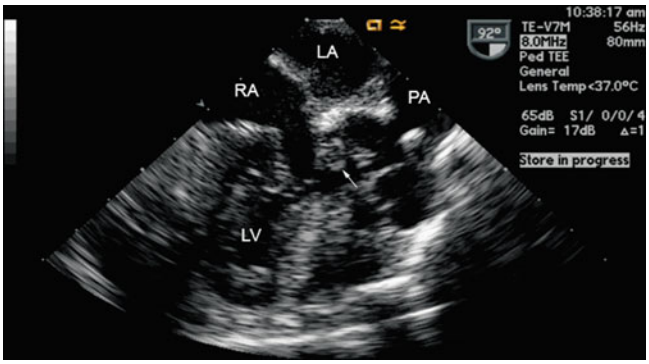


Fig. 12.45 Congenitally corrected transposition of the great arteries, as seen from the mid esophageal window in an approximately 90° sagittal plane. There is a "windsock" aneurysm of tissue (*arrow*) originating from the anterior leaflet of mitral valve and protruding into the pulmonary outflow tract, just below the pulmonary valve. In addition fibrous tissue from the mitral valve extends across the outflow tract and attaches to the ventricular septum. Neither of these was obstructive. A pulmonary artery band is seen in the main pulmonary artery, with flow acceleration noted across it. *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium

display both semilunar valves arising from their respective ventricles, and (because of the parallel great artery arrangement) an *en face* view of both semilunar valves in the same plane (Fig. 12.47, Video 12.40). Most commonly, the aortic valve will be seen anterior and leftward to the pulmonary valve. At this point, if the multiplane angle is rotated between 80° and 100° (a view approximating the ME AV LAX view), both semilunar valves and great arteries will be seen in parallel, with slight clockwise/counterclockwise rotation bringing each great artery into view [80]. These views are useful to measure semilunar valve annular diameters and also to observe the motion of the valves, including evaluation by color flow Doppler (Fig. 12.48, Video 12.41). When the aortic valve is visualized, the inflow and outflow portions of the RV can be seen, along with subaortic conus and tricuspid/aortic fibrous discontinuity. The deep transgastric views (DTG LAX, DTG Sagittal) also provide additional visualization of the intracardiac anatomy, including the semilunar valves. Given the higher incidence of cardiac malposition and sagittal orientation of the ventricular septum, the DTG LAX views generally provide more useful information regarding the intracardiac anatomy and ventricular outflow tracts (Figs. 12.46 and 12.49, Videos 12.39 and 12.42). For CCTGA, it should be noted that the more sagitally oriented views (ME LAX, ME AV LAX, DTG Sagittal, sometimes ME RV In-Out) can be confusing, because the ventriculoarterial discordant connections, along with the parallel alignment of the great arteries from the ventricles, very much resembles that of D-TGA (as described earlier). Without proper left-right orientation, it can be difficult to discern visceroatrial situs,

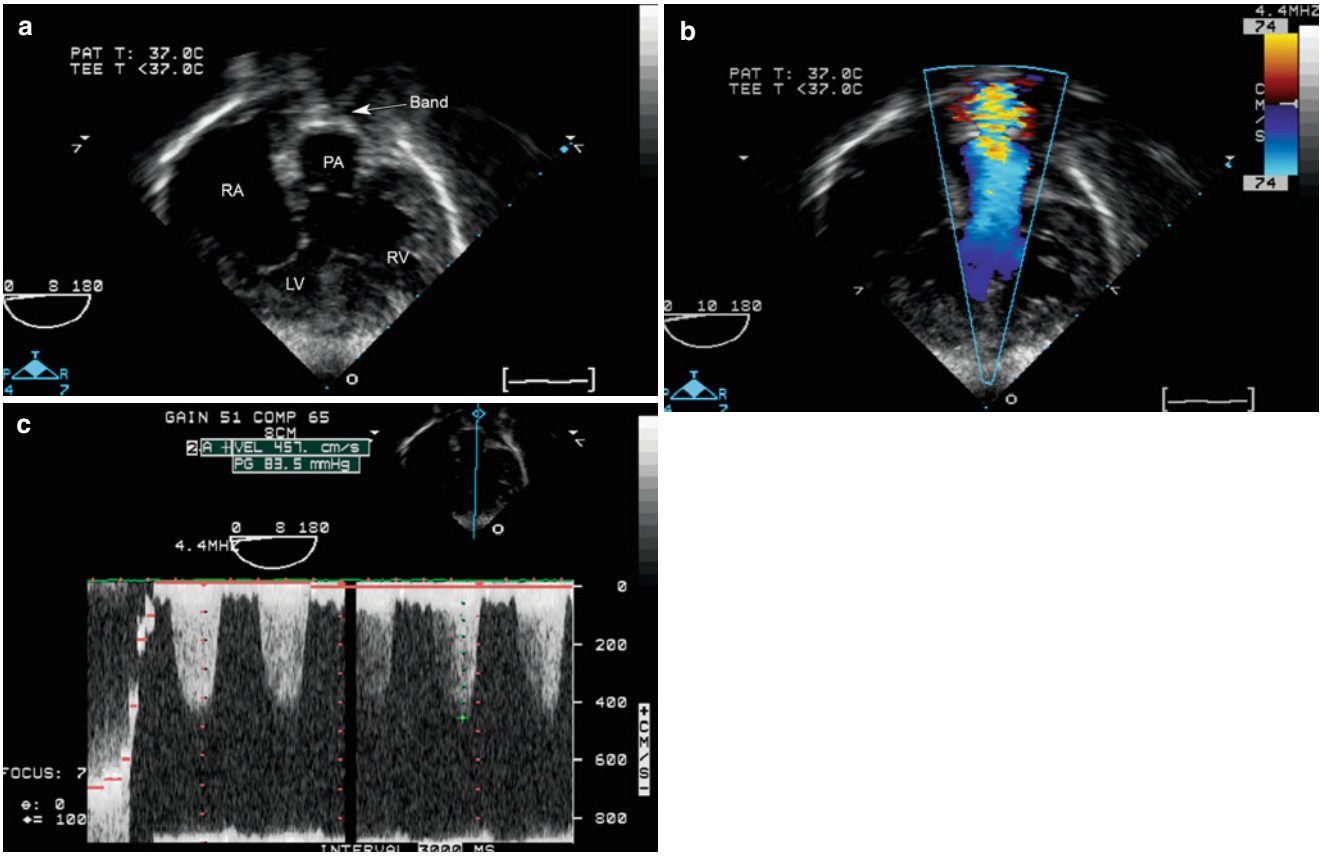


Fig. 12.46 Deep transgastric long axis view in a patient with congenitally corrected transposition of the great arteries and ventricular septal defect, in whom a pulmonary artery band (*Band*) was placed. (**a**) Shows the band well above the pulmonary valve. (**b**) Shows significant aliasing across the band, and (**c**) Displays the corresponding spectral Doppler tracing, with a peak gradient of 84 mmHg. Note the excellent angle for Doppler interrogation. *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle

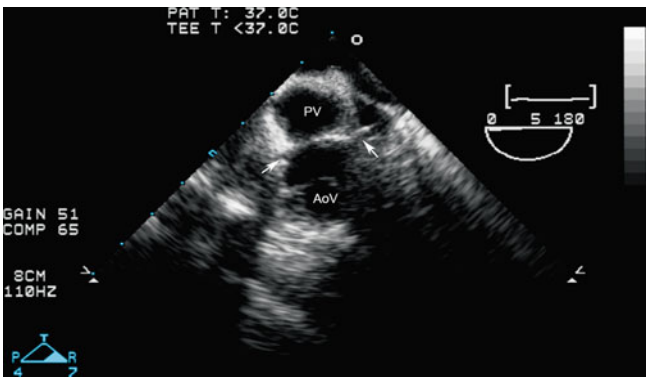


Fig. 12.47 Mid esophageal aortic valve short axis view in a patient with congenitally corrected transposition of the great arteries. Note the position of the aortic valve (*AoV*) anterior and slightly to the left of the pulmonary valve (*PV*). Coronary arteries (*arrows*) are seen to arise from the right and left facing sinuses of Valsalva

AV concordance/discordance, or relationship of the great arteries to each other. Thus in CCTGA (as well as D-TGA), the more sagittal views should always be used in conjunction with those that display left-right orientation, such as ME 4 Ch, ME AV SAX, DTG LAX, etc. Alternatively, the examiner can perform a careful right/left or left/right sweep, but meticulous attention and notation must be made as to the direction of the sweep, and the structures visualized at each point (Video 12.43).

Returning to 0° in the mid esophagus, further probe withdrawal to the upper esophageal windows (UE Ao Arch LAX, UE PA LAX) will display the branch pulmonary arteries and ascending aorta, as well as the aortic arch, and descending aorta. A ductus arteriosus, if patent, can also be seen from these views. These same structures can also be evaluated from a more sagittal perspective by turning the multiplane angle to approximately 90° (producing the UE Ao Arch SAX view), and rotating the TEE probe clockwise/counterclockwise to visualize the different vascular structures.

Surgical Considerations

The first important determination in a patient with CCTGA is whether a biventricular repair is feasible. If biventricular surgery can be performed, several options are available. The remainder of this section discusses biventricular repair of CCTGA; the surgical approach to the single ventricle is discussed in Chap. 10.

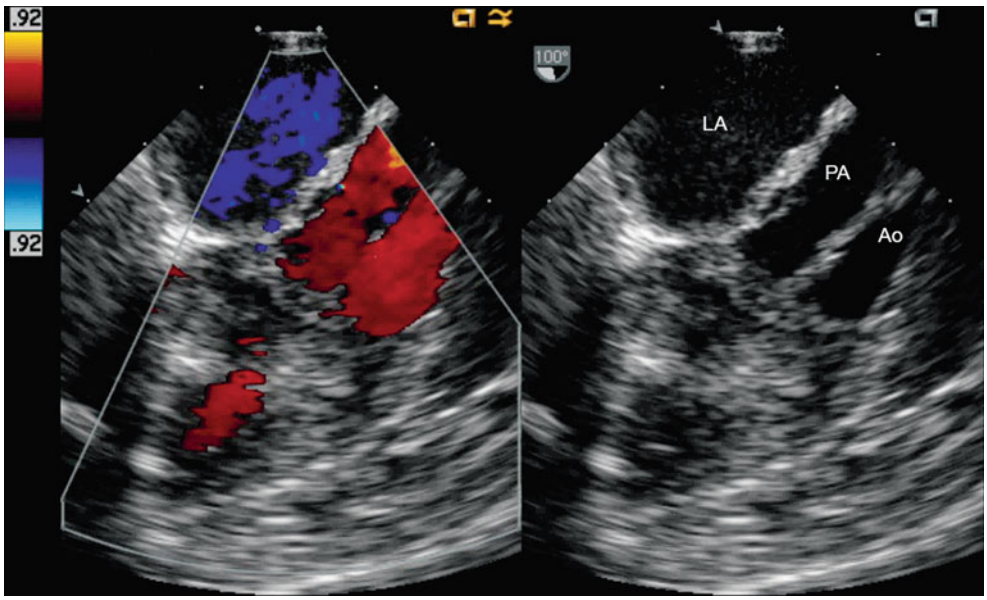


Fig. 12.48 Mid esophageal long axis view in congenitally corrected transposition of the great arteries, showing the parallel course of both aorta (*Ao*) and pulmonary artery (*PA*) as they arise from the heart. In this view, the ventriculoarterial discordance, and parallel arrangement of the great arteries, is very similar to that seen in D-transposition of the great arteries. Thus complementary views and sweeps are necessary to determine visceroatrial situs and atrioventricular connections. *LA* left atrium

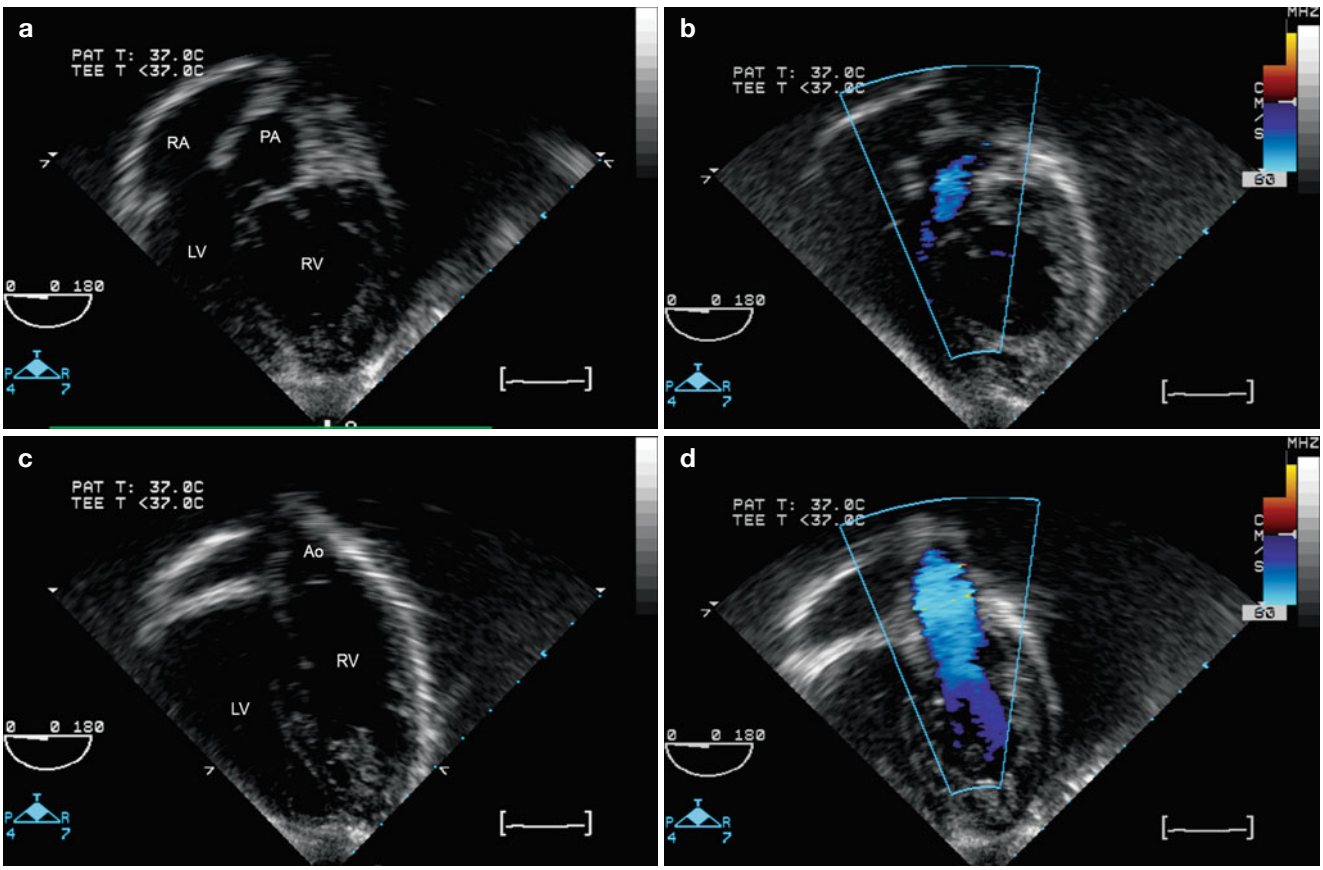


Fig. 12.49 Deep transgastric long axis view in a patient with congenitally corrected transposition of the great arteries. (**a**, **b**) show the right atrium (*RA*), left ventricle (*LV*), and pulmonary artery (*PA*), with ventricular septum bowing toward the LV. Unobstructed flow is seen across the pulmonary outflow tract. In (**c**) the probe has been withdrawn slightly to point the tip more anteriorly, visualizing the anterior aorta (*Ao*) arising from the right ventricle (*RV*). Unobstructed flow is seen into the aorta (**d**)

Biventricular surgery for CCTGA can be divided into two major classes, depending upon which morphologic ventricle assumes the role of the systemic ventricle.

Physiologic Repair

In this type of repair, also referred to as 'classic repair for CCTGA', the RV remains the systemic ventricle. Surgery is directed toward addressing important associated cardiac defects, including closure of atrial and ventricular septal defects, and tricuspid valve repair or replacement if significant tricuspid regurgitation is present. Pulmonary outflow obstruction (if present) is addressed either by direct relief of the subpulmonary obstruction, or via alternate pulmonary ventricle (LV) to pulmonary artery connection using either a conduit or direct pulmonary artery to ventricle anastomosis (*réparation à l'étage ventriculaire*, or REV procedure) [82]. In these types of surgeries, postoperative TEE is used to assess the adequacy of the repairs, using the ME 4 Ch view and its modifications to focus upon tricuspid valve anatomy/function, pulmonary outflow tract, and the atrial/ventricular septa. If an LV to pulmonary artery conduit has been placed, it can be visualized using mid and upper esophageal views with rotation of the multiplane angle between 80° and 110°, the imaging plane depending upon the exact placement of the conduit (which generally lies to the right of the aorta). The REV procedure can also be visualized in a similar manner.

Anatomic Repair

This type of repair has gained popularity over recent years, as preferences for surgical repair of CCTGA have been influenced by recent studies demonstrating a high incidence of deterioration of systemic AV valve and morphologic RV function in adulthood [72, 74, 83], and also suboptimal outcomes with physiologic repair [84, 85]. The anatomic repair for CCTGA is designed to redirect blood flow in such a manner that the LV becomes the systemic ventricle [86]. There are two principal forms of anatomic repair. In the first, both an atrial switch (Senning, Mustard) and arterial switch operation are performed at the same time. This surgery, also known as a "double switch", re-routes pulmonary venous blood through the LV to the aorta, and systemic venous blood through the RV to the pulmonary arteries (Fig. 12.50). All other cardiac lesions (e.g. atrial and ventricular septal defects) are addressed at the same time. In those patients with an intact ventricular septum or a pressure-restrictive VSD, LV "retraining" is necessary due to the fact that the LV, adapted to low pulmonary artery pressures, lacks sufficient muscle mass to maintain cardiac output against higher systemic arterial afterload. Thus a preparatory pulmonary arterial band is placed and after a sufficient period of time (months to years in most cases), the double switch is performed [87]. The second type of anatomic repair is utilized when there is pulmonary outflow obstruction (precluding an arterial switch), as well as an adequate size VSD. In these cases an atrial switch procedure is performed, along with

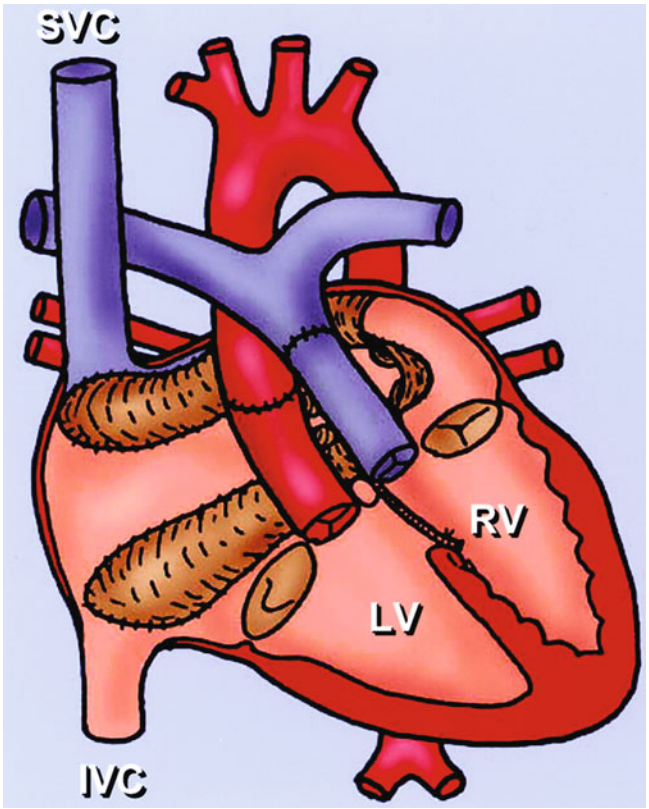


Fig. 12.50 Double-switch operation for congenitally corrected transposition of the great arteries, using the Mustard atrial baffle technique and arterial switch procedure. Venous blood from the superior and inferior vena cava (*SVC*, *IVC*) is directed to the right ventricle (*RV*) and then to the pulmonary trunk, and pulmonary venous blood is directed to the left ventricle (*LV*) and then to the aorta. The ventricular septal defect has been closed with a patch (From Warnes [65]; used with permission)

VSD closure to the aorta and an RV to pulmonary connection (usually a conduit). No coronary artery transfer is performed. This second type of operation is often referred to as an "atrial switch-Rastelli" and does not require a preparatory pulmonary artery band. In some institutions, for both the "double switch" as well as "atrial switch-Rastelli", a bidirectional cavopulmonary anastomosis (Glenn procedure) is performed along with an intra-atrial baffle redirecting IVC blood return to the morphologic RV (hemi-Mustard). This procedure, performed to partially unload the RV [82, 88, 89], this is procedure is akin to the "one and a half" ventricle strategy used for patients with pulmonary atresia/intact ventricular septum (hypoplastic right heart) or severe forms of Ebstein anomaly [90–92].

It should be noted that, in those cases with significant pulmonary outflow obstruction and intact ventricular septum or a small or remote VSD, neither anatomic repair is feasible. In these cases a physiologic repair must be performed, with or without the need for a "one and a half" ventricle modification.

The postoperative TEE evaluation of the anatomic correction of CCTGA is essentially the same as that described for the repairs described earlier in this chapter. Specifically, TEE

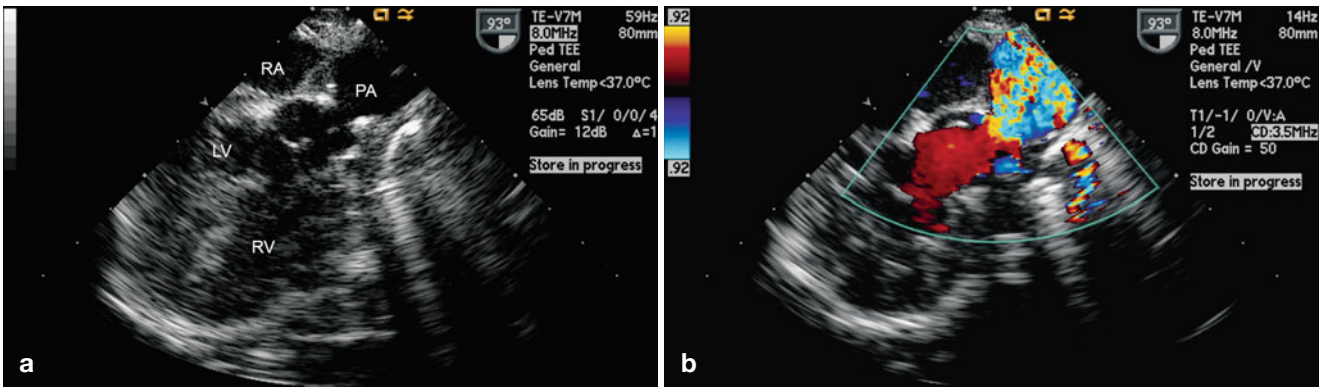


Fig. 12.51 Same patient as Fig. 12.44. The pulmonary artery band is clearly seen well above the pulmonary valve (**a**), and color flow Doppler demonstrates significant aliasing across the band (**b**). *LV* left ventricle, *PA* pulmonary artery, *RA* right atrium, *RV* right ventricle

evaluation of the atrial switch, arterial switch, and Rastelli repairs should be performed in a similar manner, with TEE views and multiplane angle adjustments made to accommodate the alterations in anatomy seen with CCTGA. It is important for the TEE examiner to have a complete understanding of the surgery that has been performed. For those patients who have undergone a preparatory pulmonary artery band, postoperative TEE can be used to evaluate LV size and function, pulmonary valve function, band placement, and the gradient across the pulmonary artery band. A combination of deep transgastric, mid and upper esophageal views can be used to assess the gradient across the band (Figs. 12.46 and 12.51, Videos 12.39 and 12.44). In some of these cases, tricuspid regurgitation significantly lessens after band placement, as the ventricular septum shifts toward the RV and decreases tricuspid annular diameter [89, 93]. This change in tricuspid regurgitation can be compared between pre and postoperative TEE studies. The altered septal geometry may also impact the systolic function of the systemic RV.

It remains to be seen whether the anatomic repair for CCTGA will eventually result in improved long-term outcomes as compared to the natural history of unoperated patients, and to those patients who have undergone physiologic repairs. The short to intermediate term results are promising, but long-term data are still lacking [93]. Specifically, there is the potential for arrhythmias and sinus node dysfunction from the atrial switch, and late LV dysfunction, neo-aortic root dilation, and neo-aortic valve insufficiency from the arterial switch [94]. Close long-term follow-up will be required to determine the actual incidence and severity of these potential complications.

Summary

The majority of conotruncal defects require surgical intervention. TEE evaluation of these malformations can be performed prior to surgery, during intra-operative repair and in the assessment of the long-term outcome of surgery for these defects. The flexibility of the multi-plane probe allows for unusual angles of interrogation when standard views are not adequate. Outcomes for conotruncal malformations have been improved by our ability to assess repairs in the operating room and to perform surveillance long after repair particularly when TTE windows are inadequate. Three-dimensional TEE is a new application that has the potential to help determine the most appropriate surgical pathway for patients with complex anatomy.

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