Adult Congenital Heart Disease

18

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Abbreviations

ASD	Atrial septal defect
CFD	Color flow Doppler
CHD	Congenital heart disease
IAS	Interatrial septum
IJ	Internal jugular
LAX	Long-axis
LVOT	Left ventricular outflow tract
ME	Midesophageal
PA	Pulmonary artery
PASP	Pulmonary artery systolic pressure
PDA	Patent ductus arteriosus
PFO	Patent foramen ovale
RV	Right ventricle
RVOT	Right ventricular outflow tract
SAX	Short-axis
SVC	Superior vena cava
TEE	Transesophageal echocardiography
ToF	Tetralogy of Fallot
TTE	Transthoracic echocardiography
VSD	Ventricular septal defect

Supplementary Information The online version of this chapter (https://doi.org/10.1007/978-3-030-84349-6_18) contains supplementary material, which is available to authorized users.

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Introduction

With the advancement of technology, transesophageal echocardiography (TEE) has quickly become invaluable in pediatric congenital heart surgery, both as a diagnostic tool and to assess repair [1]. With more congenital heart disease (CHD) patients now surviving longer, it is not difficult to imagine that these patients will ultimately need further anesthesia for noncardiac and cardiac procedures [2]. Echocardiography has remained the cornerstone for diagnosing and managing these conditions. The basic perioperative transesophageal echocardiography consensus statement suggests that the echocardiographer should be familiar with, and able to recognize, simple congenital heart disease lesions. Complex lesions are beyond the scope of a basic echocardiographer, and if suspected, consultation with an advanced echocardiographer or a switch to another diagnostic technique is warranted. Perioperatively, TEE provides useful information about the real time monitoring of ventricular filling, myocardial performance, and identification of intracardiac shunting, in addition to optimization of hemodynamic management strategies. A brief outline of the major congenital cardiac lesions and their echocardiographic correlates is provided here. There is also an introduction to the applicability of transthoracic echocardiography (TTE) to these lesions as well. The bicuspid aortic valve, which is the most common congenital

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heart disease in adulthood, is discussed separately in Chap. 9.

Atrial Septal Defect (ASD) (Highlight Box 18.1)

Highlight Box 18.1

Atrial septal defects	
2D	• Echogenic defect of tissue in interatrial septum ◦ Secundum – ME four-chamber or bicaval views ◦ Primum – ME four-chamber view ◦ Sinus venosus – ME bicaval view ◦ Unroofed coronary sinus – difficult to visualize on 2D • Associated findings ◦ Cleft anterior mitral leaflet (primum) ◦ Anomalous pulmonary venous return (sinus venosus) ◦ Atrial enlargement ◦ Ventricular dilation
CFD	• Interatrial flow – note directionality ◦ May or may not be turbulent (dependent on ASD size)
Spectral	• Calculate pulmonary to systemic flow ratio (Qp/Qs)

ME midesophageal, *ASD* atrial septal defect

Atrial septal defects (ASDs) account for 7–8% of all congenital heart disease and, thus, are relatively common, either in combination with other lesions or by themselves. The location and size, which are related to its embryonic origin, often determine the magnitude of the hemodynamic effects. Due to the proximity of the left atrium to the probe, TEE results in excellent imaging of the inter-atrial septum (IAS) and is superior to TTE in this respect.

A patent foramen ovale (PFO), present in up to 27% of the population, can cause an intracardiac shunt if right atrial pressure exceeds the left atrial pressure [3]. Although a PFO represents a possible communication between the atria, it is technically not considered an ASD, as there is no actual defect or tissue missing. A PFO may be more easily identified with the help of color flow Doppler (CFD). While the flow through a PFO may be small, lowering the aliasing velocity on CFD scale may help to identify the lower flow (Fig. 18.1 and Video 18.1). An agitated saline study with identification of "microbubbles" moving across the foramen during a Valsalva maneuver can also help identify a PFO. The use of a Valsalva maneuver is important as the release of the Valsalva maneuver temporarily increases right atrial pressure in comparison to left atrial pressure, thereby creating right-to-left flow, allowing visualization of the agitated saline crossing the interatrial septum [3] (Fig. 18.2 and Video 18.2).

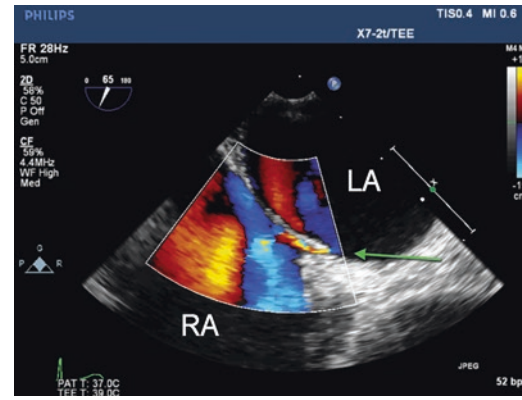

Fig. 18.1 Midesophageal view of the interatrial septum with color flow Doppler. The *green arrow* indicates a left-to-right shunt across a patent foramen ovale (PFO). *LA* left atrium, *RA* right atrium

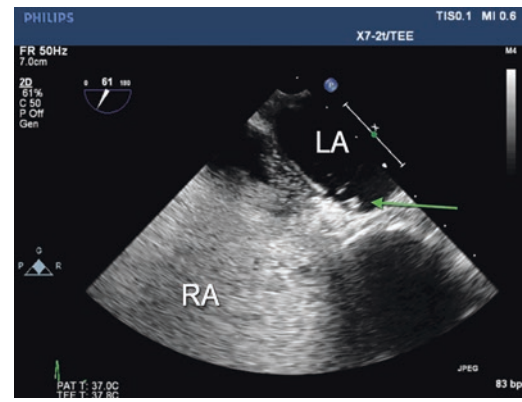

Fig. 18.2 Midesophageal view of the interatrial septum during agitated saline injection. The *green arrow* indicates transseptal flow of agitated saline from the right atrium (RA) to left atrium (LA)

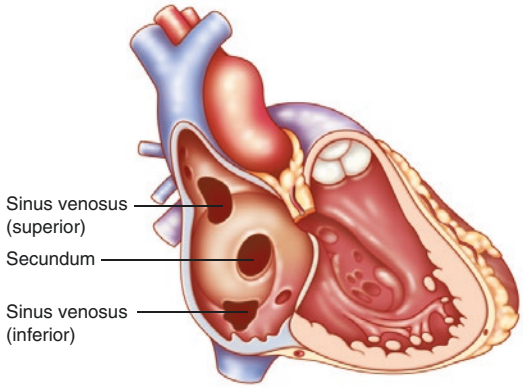

Fig. 18.3 Diagram of interatrial septum from the perspective of the right atrium and right ventricle

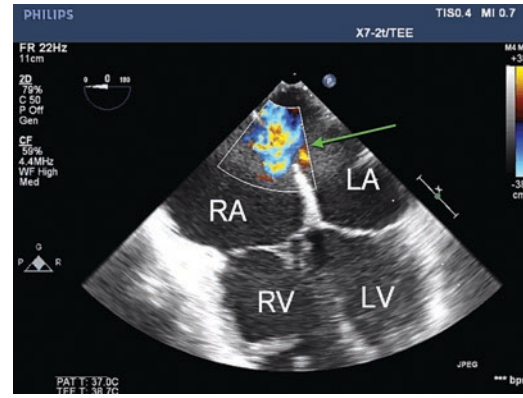

Fig. 18.4 Midesophageal four-chamber view demonstrating an ostium secundum ASD (*green arrow*) with left-to-right flow noted on color flow Doppler. *RA* right atrium, *LA* left atrium, *RV* right ventricle, *LV* left ventricle

The defects or gaps truly classified as ASDs involve some degree of absent tissue, allowing the potential for various degrees of intracardiac shunting. The ASDs are classified based upon their location which relates to the defect during embryologic development. The subdivided defects are described below (Fig. 18.3).

Ostium Secundum ASD

This is the most common ASD (approximately 70%) and generally occurs in the area contained in the limbus of the fossa ovalis [4]. During embryologic development, the septum primum grows toward the atrioventricular canal. An ostium develops centrally (termed the ostium secundum) which allows oxygenated blood in utero to cross the interatrial septum. Subsequently a septum secundum develops to cover this ostium yet still allows flow through as the foramen ovale. After birth with the increase in left atrial pressure from increased pulmonary blood flow, the foramen ovale is functionally pushed closed. Fusion of the two septae finalizes the process, leaving a fossa ovalis. A defective closure of the ostium secundum leads to the ostium secundum ASD (Fig. 18.4 and Video 18.3). It may be circular in shape or may be a series of fenestrations associated with an aneurysmal interatrial septum.

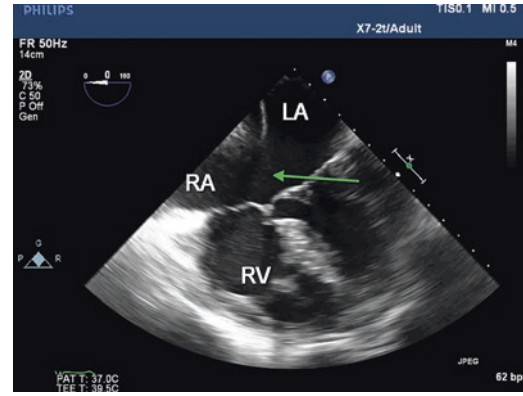

Fig. 18.5 Midesophageal four-chamber view demonstrating an ostium primum ASD (*green arrow*). *RA* right atrium, *LA* left atrium, *RV* right ventricle

Ostium Primum ASD

These defects are the second most common ASD (approximately 20%) and occur in the inferior and anterior portion of the IAS, near the atrioventricular valves. This defect generally represents the smallest degree of an atrioventricular canal defect. During embryologic development, the septum primum develops in the direction of the atrioventricular valves leaving the ostium primum to be covered by the septum secundum. Failure of this closure leaves the ostium primum atrial septal defect (Fig. 18.5 and Video 18.4). As

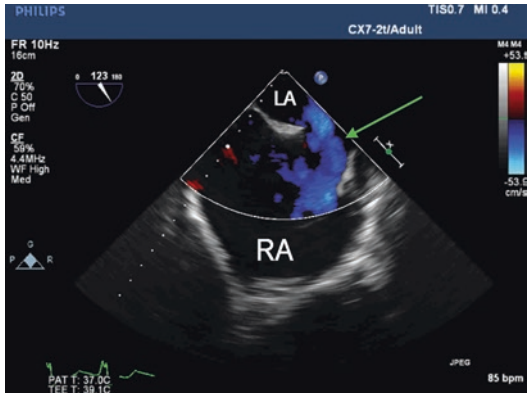

Fig. 18.6 Midesophageal bicaval view in a patient with a superior sinus venosus ASD and a grossly dilated right atrium (RA). The *green arrow* indicates left-to-right flow from the left atrium (LA) to the RA near the superior vena cava and RA junction

the endocardial cushion is also involved in the development of the atrioventricular valves, ostium primum defects can be associated with a cleft in the anterior mitral leaflet.

Sinus Venosus ASD

This defect represents an atrial communication adjacent to the attachment of either the superior or inferior vena cava, and results in the respective vena cava over-riding the defect. Sinus venosus defects account for about 8% of all ASDs [4]. Embryologically, the vena cavae are derived from the sinus venosus. Abnormal resorption of the sinus venosus leads to a defect between the cavae and the left atrium (Fig. 18.6 and Video 18.5). The defect is often associated with partial anomalous pulmonary venous return (i.e., anomalous right upper pulmonary vein draining into the left atrium).

Coronary Sinus ASD (Unroofed Coronary Sinus)

These rare defects (< 1%) result from a partial or complete defect in the separation between the LA and the coronary sinus, resulting in "unroofing" and causing communication between the right and the left atria.

Transesophageal Echocardiographic Examination for ASDs

The midesophageal (ME) four-chamber view can interrogate the majority of the interatrial septum, though probe withdrawal and insertion may be required for superiorly and inferiorly located ostium secundum defects as well as sinus venosus defects. Advancing the probe to the AV groove allows detection of ostium primum defects, evaluation of the coronary sinus, and Doppler evaluation of atrioventricular valves. Rotation of the probe to right and left is recommended to thoroughly interrogate the area. The ME aortic valve short-axis (SAX) or ME right ventricular inflow-outflow views can also be used to evaluate the septum, TV, and PV. In addition, these views can be used to quantify tricuspid regurgitation, estimate pulmonary artery systolic pressure (PASP) and right ventricular (RV) function, and search for abnormalities of venous return. The ME bicaval view provides a good cross-sectional display of the septum (superior to inferior), aligns the Doppler beam perpendicular to the septum, and is also an excellent view for agitated saline studies. This view is not particularly suited for detection of ostium primum defects but can be modified by clockwise or counterclockwise rotation and multiplane manipulation to detect and evaluate all other ASDs. The transgastric midpapillary SAX views can be used to detect flattening of the interventricular septum and help diagnose RV pressure or volume overload (see Chap. 10). While beyond the scope of this textbook, TEE, especially 3D TEE, can be invaluable during device closure of ASDs by identifying the site and size of the defect, evaluating adequacy of the tissue ring around the defect (generally 5 mm) to hold the device, and following the deployment of the device in real time.

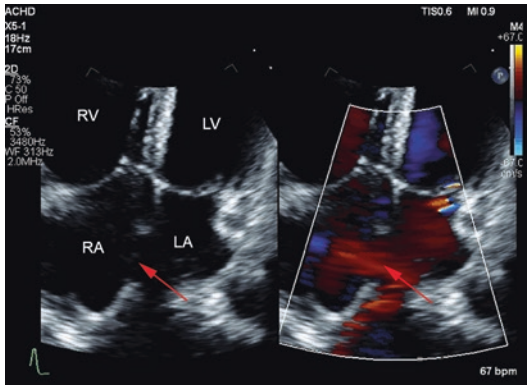

Fig. 18.7 Apical four-chamber view with a zoomed perspective of the interatrial septum with color compare feature (two-dimensional image on left; color flow Doppler applied on the right). The *red arrow* indicates a secundum ASD on both images. *RA* right atrium, *LA* left atrium, *RV* right ventricle, *LV* left ventricle

Transthoracic Echocardiographic Examination for ASDs

Factors that alert the clinician to the possibility of an ASD (during a TTE exam) include a hypermobile interatrial septum, abrupt septal irregularity, right atrial and ventricular volume overload, and pulmonary artery dilatation. A subcostal window, if it can be obtained, is ideal for examination of the IAS since the septum is mostly perpendicular to the Echo beam, and any existing shunt should be parallel to it. The apical four-chamber view, although satisfactory, suffers from the flow being perpendicular to the beam, limiting the sensitivity of Doppler evaluation (Fig. 18.7 and Video 18.6). Out of the subtypes, TTE is the most sensitive in detecting primum ASDs (100%) and the least sensitive when it comes to sinus venosus ASDs (44%) [5]. This is probably a reflection of distance from the probe and the variability in anatomy. TEE is considered superior to diagnose and quantify ASDs and is needed when TTE is either indeterminate or technically limited.

Ventricular Septal Defect (VSD) (Highlight Box 18.2)

Highlight Box 18.2

Ventricular septal defects	
2D	• Echogenic defect of tissue in interventricular septum ◦ ME or apical four-chamber views (muscular and inlet) ◦ ME AV SAX or PSAX basal level view (perimembranous and outlet) • Associated findings ◦ Atrial enlargement ◦ Ventricular dilation
CFD	• Presence of interventricular flow ◦ May or may not be turbulent (dependent on VSD size)
Spectral	• Calculate pulmonary to systemic flow ratio (Qp/Qs) • Estimate pulmonary arterial systolic pressure (TR jet)

ME midesophageal, *AV* aortic valve, *SAX* short-axis, *PSAX* parasternal short-axis, *VSD* ventricular septal defect

Ventricular septal defects (VSDs) are present in approximately 10% of all adults with congenital heart disease and can occur in isolation or associated with other disorders. The ventricular septum can be divided into four components, each with its distinct morphology: membranous, inlet, trabecular (muscular), and outlet [6] (Fig. 18.8). VSDs follow similar nomenclature but can span more than one segment. Spontaneous closure is more likely for VSDs of the membranous or muscular type.

The most common of the four subtypes is the perimembranous VSD, occurring in 75–80% of all VSDs. This defect is found in the membranous portion of the septum beneath the tricuspid valve and allows a connection to the left ventricular outflow tract immediately beneath the aortic valve [6]. It is best seen in the ME right ventricular

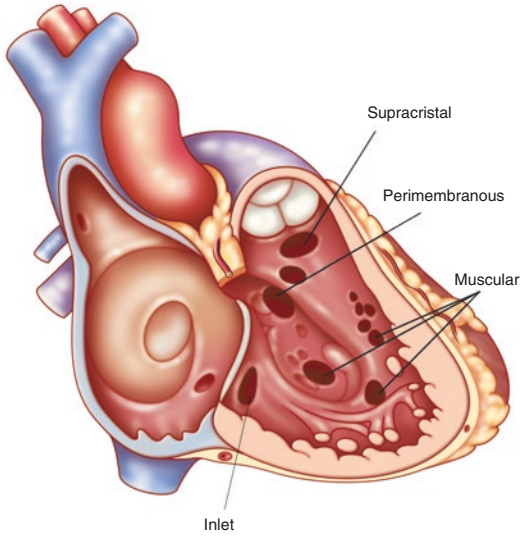

Fig. 18.8 Diagram of the interventricular septum from the perspective of the right atrium and right ventricle

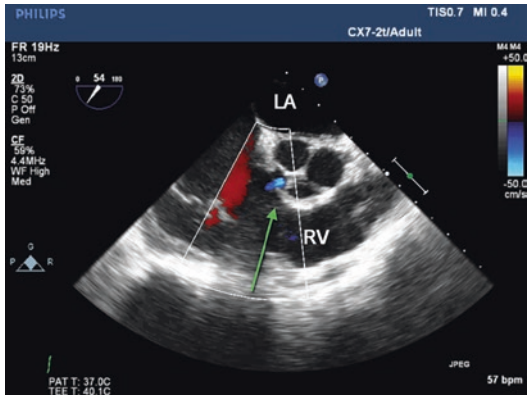

Fig. 18.9 Midesophageal right ventricular inflow-outflow view in a patient with a perimembranous ventricular septal defect (VSD) indicated by the *green arrow*. *LA* left atrium, *RV* right ventricle

inflow-outflow or ME aortic valve SAX views (Fig. 18.9 and Video 18.7). Associated aneurysm of the membranous septum or accessory tricuspid tissue may be visualized. Perimembranous defects that occur high in the left ventricular outflow tract (LVOT) can result in aortic regurgitation due to cusp herniation through the defect (most commonly the right coronary cusp).

Inlet VSDs, also part of the endocardial cushion defect spectrum, are located in the posterior portion

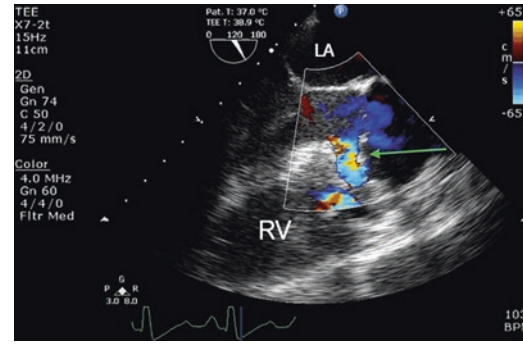

Fig. 18.10 Midesophageal aortic valve long-axis view in a patient with an outlet VSD (*green arrow*). *LA* left atrium, *RV* right ventricle

of the interventricular septum immediately below the mitral and tricuspid valves [6]. Echocardiographically, these two valves tend to be located at the same level; however the normal insertion of the tricuspid valve is typically a few millimeters inferiorly. These defects are large and generally do not close spontaneously. Multiple configurations of the atrioventricular valves can occur, the details of which are outside the scope of this text. Endocardial cushion defects represent defects in the separation of the right and the left heart chambers and can have complete absence of the septae, one common atrioventricular valve, and an ostium primum ASD, among other abnormalities.

Muscular defects, approximately 5–20% of all VSDs, occur centrally or apically in the trabecular portion and can have multiple openings ("Swiss cheese" appearance). Apical VSDs may occur after myocardial infarctions [7]. Color flow Doppler is invaluable to detect multiple defects in the muscular septum.

Outlet VSDs are also known by several terms: supracristal, infundibular, doubly committed, or subarterial VSDs. Irrespective of the nomenclature used, they occur in the region just below the aortic and pulmonic valves and can have associated aortic insufficiency (related to the herniation of the right coronary cusp). Interrogation of the outflow tracts side by side, done as a modification of the ME aortic valve long-axis (LAX) view or the ME right ventricular inflow-outflow view, is used to detect these defects (Fig. 18.10 and Video 18.8).

Transesophageal Echocardiographic Examination of VSDs

The complexity of the interventricular septum requires multiple views as well as rotation and use of the multiplane angle at nonstandard imaging planes. The ME four-chamber, ME aortic valve short-axis and long-axis, ME RV inflow-outflow, and deep transgastric long-axis views are recommended for a focused interrogation. Apart from number, size, location, and nature of the defect(s), other pertinent findings to look for include additional congenital lesions, aortic valve abnormalities, signs of RV pressure and volume overload, and functional consequences of the VSD. Doppler interrogation can quantify the nature and magnitude of the intracardiac shunt, estimate valvular regurgitation, and estimate PASP (see Chap. 4).

High velocity through the defect as evidenced by Doppler interrogation is indicative of a restrictive shunt, whereas low, non-turbulent flow denotes a nonrestrictive defect. Generally, a nonrestrictive defect indicates a more severe lesion [8, 9]. The ratio of pulmonary-to-systemic blood flow (Qp/Qs) should be measured since it has diagnostic and therapeutic implications. A high Qp/Qs indicates that there is a significant left-to-right shunt, which may eventually lead to pulmonary overcirculation and Eisenmenger's syndrome. A low Qp/Qs (< 1) is indicative of a right-to-left shunt.

Transthoracic Echocardiographic Examination for VSDs

Most patients with VSDs are investigated initially with a TTE for a murmur or other clinical indications. The ventricular septum is closer to the transthoracic probe, favoring this mode of imaging. Atrioventricular valves that lie on the same level or unusual LVOT shape can provide a clue to the presence of a VSD. Certain syndromes, such as trisomy 21 (Down syndrome, associated with an inlet VSD), CHARGE and Noonan syndromes, or VACTERL syndrome, are associated with VSDs as well. Abnormal flow

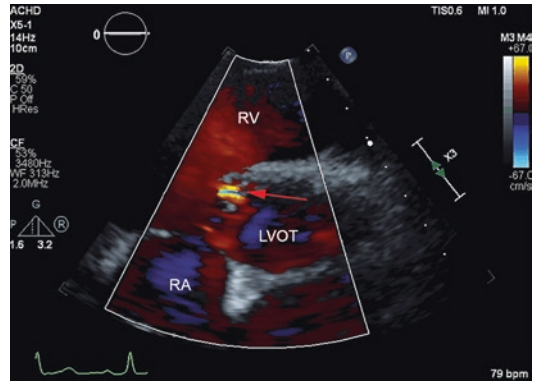

Fig. 18.11 Parasternal short-axis view at a basal level with color flow Doppler demonstrating a perimembranous VSD (*red arrow*). *RV* right ventricle, *RA* right atrium, *LVOT* left ventricular outflow tract

detection with CFD is the first step to detection of a VSD. The parasternal long-axis view (and small variations thereof) is very helpful to detect perimembranous and outlet VSDs. The parasternal short-axis view with CFD box applied over the conal septum helps both in detection and identification (Fig. 18.11 and Video 18.9). Inlet VSDs are best visualized in the apical four-chamber view with CFD interrogation of the septum in the region of the atrioventricular valves. Muscular VSDs can be best detected in the apical four-chamber view but requires an off-axis view and sweeping loops since their trajectory can be convoluted (Fig. 18.12 and Video 18.10). Small jets of flow within the trabecula (especially in the RV) can occur that need to be distinguished from true flow across the septum. Restriction across the defect (usually a gradient exceeding 20–30 mm Hg), the direction of flow during the cardiac cycle, effect on other structures (dilation of chambers), and presence of pulmonary hypertension are all important parts of a thorough examination [10].

Persistent Left Superior Vena Cava (SVC)

Approximately 0.5% of the population has a persistent left SVC, which drains into the coronary sinus 90% of the time. It is uncommonly

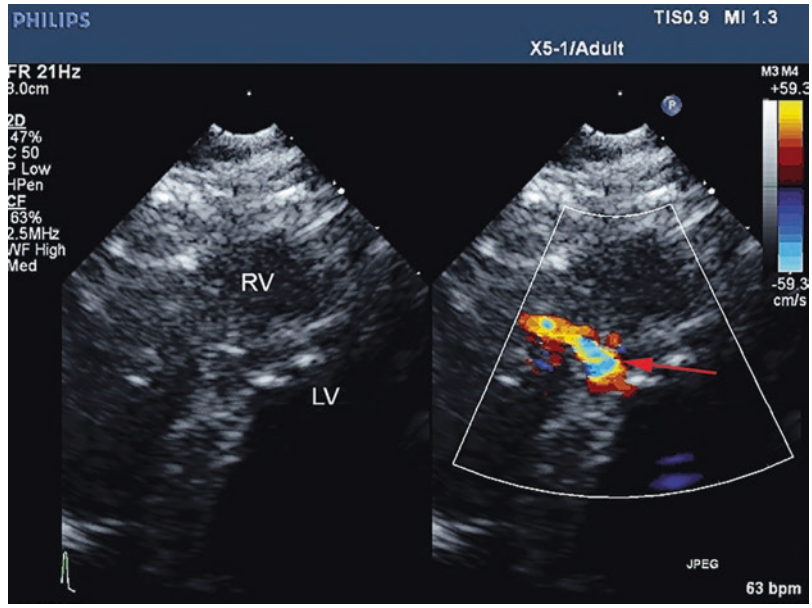

Fig. 18.12 Apical four-chamber view with an RV focus, zoomed perspective on interventricular septum with color compare feature (two-dimensional image on left; color flow Doppler applied on the right). Color flow Doppler demonstrates a muscular VSD (*red arrow*). *RV* right ventricle, *LV* left ventricle

associated with an absent right SVC. During embryologic development, there are two superior vena cavae. Normally the left-sided SVC regresses with blood from the internal jugular (IJ) and left subclavian returning to the heart via the innominate vein. In the setting of a persistent left SVC, the left IJ and subclavian typically return blood flow to the heart via the left SVC into the coronary sinus. In its presence, central venous cannulae and pacemakers can take an abnormal orientation. In cardiac surgery, retrograde cardioplegia can prove ineffective and venous cannulation strategies may need to be adjusted.

The coronary sinus can be imaged by pushing the probe in from a ME four-chamber view, a modified bicaval view, or in the posterior atrioventricular groove on the ME two-chamber view (Fig. 18.13 and Video 18.11). Dilatation of the coronary sinus (> 10 mm in diameter) should arouse suspicion for a persistent left SVC. Other causes such as elevated right atrial pressures from heart failure, atresia or stenosis of the ostium, or a coronary artery fistula to the coronary sinus are alternative causes for coronary sinus dilatation and should be evaluated as well. The suspicion can then be confirmed with agitated saline injection into the left upper extremity and resultant

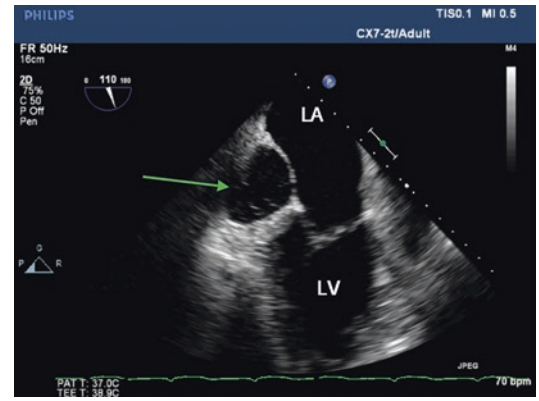

Fig. 18.13 Midesophageal two-chamber view with the dilated coronary sinus (*green arrow*) in cross section to the posterior aspect of the top of the left ventricle in a patient with a persistent left superior vena cava. *LA* left atrium, *LV* left ventricle

coronary sinus opacification. A large coronary sinus and its drainage into the right atrium have been mistaken for an ASD in some patients, and this makes thorough imaging essential [11]. The principles of identification and characterization for this condition remain similar with TTE and require agitated saline injection into the left arm resulting in opacification of the coronary sinus prior to the right atrium. Visualization of a dilated coronary sinus is done in the parasternal long-

axis view (where it lies adjacent to the posterior mitral annulus) [12].

Patent Ductus Arteriosus/ Aorticopulmonary Window

The ductus arteriosus is a vascular communication between the proximal descending aorta and the main or the left pulmonary artery in its roof. It closes spontaneously after birth but can persist to adulthood in rare cases causing a left-to-right shunt. It is usually a co-incidental finding picked up due to a murmur that leads to echocardiography [13]. Echocardiography is helpful not only to diagnose the lesion but also to evaluate the shunt magnitude and volume load, estimate pulmonary artery pressures, and identify associated cardiac pathology. Some patients may present with endarteritis (endocarditis of the ductus), which is responsible for almost half of the deaths in adult patients with a patent ductus arteriosus (PDA) [14]. The pulmonary side of the ductus is more commonly the site of infection. The patent ductus can be closed either surgically or with a transcatheter device with excellent results. Upper esophageal views have been used to visualize a PDA, with nonstandard orientation of the multiplane angles. Since the connection between the aortic isthmus and the main pulmonary artery (PA) hides anterior to the left mainstem bronchus, it is often difficult to visualize with TEE. The demonstration of flow abnormality in the PA using color flow Doppler is excellent supportive evidence, but by itself not diagnostic of a PDA. Proper parallel alignment of the Doppler beam with the flow is even more difficult. It is important to note the FiO_2 during the shunt calculation since hyperoxia leads to reduction in PA pressures and an increase in the shunt [14]. Using a transthoracic approach, the PDA is imaged from the parasternal short-axis view at the base of the heart, suprasternal long-axis view, and high left parasternal long-axis views. Detailed description of the techniques is outside the scope of this text and can be found elsewhere [15]. The aorticopulmonary window represents a more proximal communication between the ascending aorta and the PA and can be easier to visualize with TEE. Hemodynamic consequences tend to be similar to a PDA.

Transesophageal Echocardiographic Evaluation of Tetralogy of Fallot (Highlight Box 18.3)

Highlight Box 18.3

Tetralogy of Fallot	
2D	• Pulmonic stenosis ◦ Narrowed RVOT in ME RV inflow-outflow or PSAX view • Right ventricular hypertrophy ◦ Measure in ME or apical four-chamber view • Overriding aorta ◦ Observed in ME LAX or PLAX view • Ventricular septal defect ◦ Typical perimembranous VSD in ME AV SAX or PSAX view
CFD	• Presence of interventricular flow ◦ May or may not be turbulent (dependent on VSD size) • Turbulence in RVOT/pulmonary artery
Spectral	• Calculate pulmonary to systemic flow ratio (Qp/Qs) • Estimate pulmonary arterial systolic pressure (TR jet)

RVOT right ventricular outflow tract, *ME* mid-esophageal, *RV* right ventricular, *PSAX* parasternal short-axis, *LAX* long-axis, *PLAX* parasternal long-axis view, *VSD* ventricular septal defect, *TR* tricuspid regurgitation

Classically, tetralogy of Fallot (ToF) patients manifest a VSD, pulmonic stenosis, an overriding aorta, and RV hypertrophy. Addition of an ASD makes it a pentalogy (present in about a third of cases). Multiple other congenital cardiac lesions can accompany a ToF, such as right aortic arch, systemic venous abnormalities, and LVOT obstruction, among others. Most patients require surgery early in life due to cyanosis from right-to-left shunt; thus patients that survive into adulthood generally have little RV obstruction. The goals of echocardiography should include (apart

from confirming the diagnosis) quantification of the RV obstruction, VSD shunt magnitude, and direction and detection of associated anomalies.

The large, perimembranous VSD is best seen in the ME aortic valve LAX or SAX views. The defect is located between the right and the noncoronary cusps of the aortic valve. This generally permits shunt interrogation by Doppler (either color or spectral). The entire septum should be carefully interrogated to rule out additional defects. The ME aortic valve LAX view can also demonstrate the aortic override, which can be variable in presentation. In the post-repair adult, the repair of the VSD using a patch still permits identification of the override (Fig. 18.14 and Video 18.12). Involvement of the aortic valve, either as part of the primary lesion or during repair, needs to be carefully evaluated.

Manipulation of the multiplane angle from either the ME aortic valve LAX or ME right ventricular inflow-outflow view can be used to interrogate the RV outflow tract (RVOT). Obstruction is suggested (using color flow Doppler) by the presence of aliasing and turbulence in the RVOT. Transgastric views can be used to examine the RVOT as well. These views can help detect abnormalities of the pulmonic valve, if present. RV wall thickness can be measured in the ME right ventricular inflow-outflow to quantify the hypertrophy. Interrogation of the RV outflow and the pulmonary artery with 2D, color flow Doppler, and spectral Doppler is helpful after repair since stenosis can persist and even worsen as the patient continues to grow into adulthood.

After repair, left and right ventricular outflow obstruction and valvular regurgitation remain the focus. Assessment of RV systolic pressure, size, and function is vital to the exam. Tricuspid and pulmonic regurgitation is not uncommon, and their quantification can be useful for future comparison.

Transthoracic Echocardiographic Examination of Tetralogy of Fallot

Noninvasive fetal echocardiography can detect ToF as early as at 14 weeks of gestation. In early childhood, TTE is invaluable in the diagnosis and follow-up of ToF. The subcostal views (long- and short-axis) are utilized to evaluate the atrial septum and connections in the heart at the level of the valves. They can also help in assessing the VSD, RVOT obstruction, and potential ASDs. An apical four-chamber view can visualize both the atrioventricular valves and the VSD. The parasternal long-axis view is the best to identify the type of VSD and the degree of the aortic override (Fig. 18.15 and Video 18.13). The aorta overrides the VSD by less than 50% of the aortic diameter

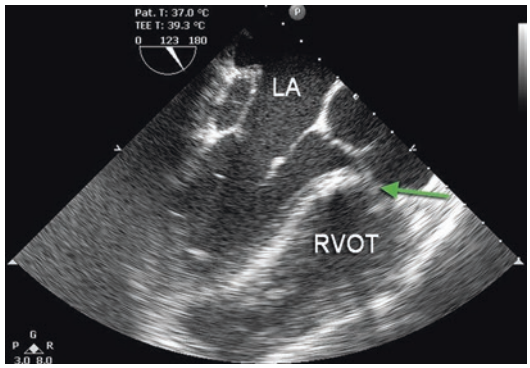

Fig. 18.14 Midesophageal long-axis view in a patient with tetralogy of Fallot after VSD repair. Note the overriding aorta (*green arrow*) that remains and the evidence of right ventricular hypertrophy. Incidentally, there is a dilated coronary sinus resulting from a persistent left-sided superior vena cava. *LA* left atrium, *RVOT* right ventricular outflow tract

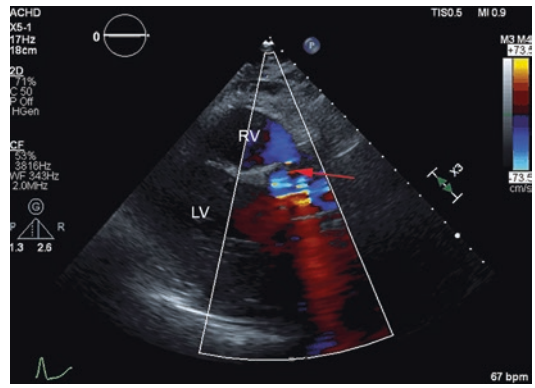

Fig. 18.15 Parasternal long-axis view in a patient with tetralogy of Fallot. The *red arrow* indicates the overriding aorta and a perimembranous VSD detected on color flow Doppler. *RV* right ventricle, *LV* left ventricle

in ToF, whereas the override is more than 50% in double-outlet right ventricle. The parasternal short-axis view can provide information regarding the coronary arterial configuration, VSD margins, and the RVOT anatomy. The suprasternal view is vital to assess the right- or left-sidedness of the aortic arch, as well as visualizing the branching from the aortic arch [16].

Conclusion

With more congenital heart disease patients now surviving longer, these patients will increasingly require clinical management related to or unrelated to their heart. Echocardiography has remained the cornerstone of diagnosing and managing these conditions. The basic echocardiographer should be familiar with and able to recognize simple congenital heart disease. Complex lesions should involve consultation with an advanced echocardiographer. Perioperative TEE or point-of-care TTE can provide useful information about the real-time monitoring of ventricular filling, myocardial performance, and identification of intra-cardiac shunting, in addition to optimization of hemodynamic management strategies.

Questions

1. Which of the following views provides the best visualization to detect shunt across a patent foramen ovale?

   A. Apical four-chamber view
   B. Transgastric midpapillary short-axis view
   C. Midesophageal bicaval view
   D. Midesophageal AV long-axis view

2. Which of the following is the most common kind of ventricular septal defect?

   A. Perimembranous
   B. Inlet
   C. Outlet
   D. Muscular

3. Which of the following congenital conditions in adults are amenable to device closure?

   A. Sinus venosus ASD
   B. Patent ductus arteriosus
   C. Complete AV cushion defect
   D. Situs inversus

4. Which of the following is a reason to use a Valsalva maneuver during a bubble study to detect a patent foramen ovale?

   A. To increase the turbulence of the blood across a shunt
   B. To induce a right-to-left shunt
   C. To enable positioning of the echo probe closer to the interatrial septum
   D. To reduce transit time of the echo contrast to reach the atrium

5. A 25-year-old woman is diagnosed with a ventricular septal defect, moderate tricuspid regurgitation, and no other valvular abnormalities. The peak velocity across the VSD is measured at 4 m/s. Her right atrial pressure (or CVP) is 4 mmHg, and her systolic BP is 110/70 mmHg. What is the peak systolic pulmonary pressure?

   A. 46 mmHg
   B. 50 mmHg
   C. 64 mmHg
   D. 94 mmHg

6. Which of the following is most true regarding restrictive VSDs?

   A. Compared to a nonrestrictive defect, they are easier to detect using 2D imaging.
   B. Reduction in the Nyquist limit using color flow Doppler aids in detection.

C. Restrictive VSDs are generally less severe than nonrestrictive defects.
D. Restrictive VSDs exhibit non-turbulent flow.

7. In the midesophageal bicaval view, following release of a Valsalva maneuver, which of the following would be expected regarding flow through a patent foramen ovale?

A. Flow should be toward the probe and the right side of the display.
B. Flow does not occur due to closure of the flap between the septum secundum and septum primum.
C. Flow is more difficult to detect with the color scale reduced to 30 cm/s.
D. Flow can only be detected with bubble contrast.

8. Which of the following conditions does not result in the dilation of the coronary sinus?

A. Unroofed coronary sinus
B. Persistent left SVC
C. Severe right ventricular pressure or volume overload
D. Presence of a Eustachian valve

9. Which of the following is most true regarding detection of VSDs with echocardiography?

A. Transthoracic views are not sensitive for small VSDs.
B. A low Qp/Qs indicates that there is a right-to-left shunt.
C. Bubble studies are generally not used for detection due to the risk for air emboli.
D. Flow is usually right to left.

10. Which of the following describes an ostium primum ASD?

A. It is the most common type of ASD.
B. It results from failure of fusion between the septum primum and the septum secundum.
C. It is the result of incomplete fusion between the septum primum and the endocardial cushion.
D. It is a communication between the ostium primum and the ostium secundum.

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