ABSTRACT

Intraoperative transesophageal echocardiography (TEE) for pediatric congenital heart disease has become a standard of care in most institutions in the USA and across Europe. Advances in technology have facilitated the safe use of this tool in patients as small as 2.5 kg and have proved invaluable in guiding surgical repair of congenital cardiac lesions as well as guiding anesthesia management for cardiac and noncardiac procedures. This article describes a systematic approach to image acquisition in children with congenital heart disease (CHD). Specific congenital heart lesions are also described with an emphasis on intraoperative TEE data acquisition and analysis.

Keywords: Transesophageal, Echocardiography, Congenital heart disease.

How to cite this article: Navaratham M, Punn R, Tacy TA. Imaging Congenital Heart Disease in the Perioperative Period using Transesophageal Echocardiography. J Perioper Echocardiogr 2014;2(1):10-28.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

The routine use of transesophageal echocardiography in congenital heart surgery has been shown to be safe, improve surgical results,¹ and be costeffective.² For these reasons, intraoperative transesophageal echocardiography is an invaluable addition to the surgery for congenital heart disease. Technological advancements, particularly the use of small probe sizes, have significantly improved patient safety and success of cardiac surgery in infants and children.³⁻¹¹ Previous publications have reviewed TEE imaging in the child with CHD, and are excellent resources of information.¹²⁻¹⁵ The focus of this article is a review of some of the more commonly encountered forms on congenital heart disease, with a description of the most useful views by transesophageal imaging for each.

A Systematic Approach to Data Acquisition

The transesophageal echocardiogram examination in children with CHD should be based on a systematic, organized and easily reproducible approach. This provides more efficient use of physician and patient time, avoidance of inadvertent omission of important views, and facilitates study review.¹⁶ The American Society of Echocardiography (ASE)¹⁷ and Society of Cardiac Anesthesiologists (SCA)¹⁸ have established guidelines for performing intraoperative multi-plane TEE examination in adults which consists of a series of 20 cross-sectional views of the heart and great vessels.

We review herein a systematic approach to acquiring essential TEE information in children with CHD, which is based on the ASE guidelines. It is our practice to perform a complete TEE in the operating room preoperatively, as the window provided by TEE may be superior to the transthoracic window.

STANDARD TRANSESOPHAGEAL VIEWS

Midesophageal Views

A complete TEE exam can begin with the images taken first with the probe in the midesophageal area, which provides proximity, and hence best image resolution to the cardiac structures of interest. Technological advancements, particularly the use of small probe sizes, have significantly improved patient safety and success of cardiac surgery in infants and children.³⁻¹¹ Previous publications have reviewed TEE imaging in the child with CHD, and are excellent resources of information.¹²⁻¹⁵ The focus of this article is a review of some of the more commonly encountered forms on congenital heart disease, with a description of the most useful views by transesophageal imaging for each.

The routine use of transesophageal echocardiography in congenital heart surgery has been shown to be safe, improve surgical results,¹ and be costeffective.² For these reasons, intraoperative transesophageal echocardiography is an invaluable addition to the surgery for congenital heart disease. Technological advancements, particularly the use of small probe sizes, have significantly improved patient safety and success of cardiac surgery in infants and children.³⁻¹¹ Previous publications have reviewed TEE imaging in the child with CHD, and are excellent resources of information.¹²⁻¹⁵ The focus of this article is a review of some of the more commonly encountered forms on congenital heart disease, with a description of the most useful views by transesophageal imaging for each.

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Midesophageal Views

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Four-Chamber View (0-30°)

This is one of the most high yield TEE views. With the probe in a neutral position in the midesophagus the image is acquired with a multiplane angle ranging from 0 to 30°. Correct image display is achieved when both atrioventricular valves are seen (Fig. 1). This cross-sectional view is most useful for the initial 2D imaging of chamber size and morphology as well as the assessment of atrioventricular valve function.

To image the LV and LV inflow, the probe should be retroflexed to correct for LV apex foreshortening. This view will illustrate the LV from inflow to apex and can be a good

![Image of a medical chart with a patient's information]

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http://journals.sagepub.com/doi/full/10.5005/jp-journals-10034-1015
initial demonstration of LV function. Further retroflexion, (or advancing the probe lower in the esophagus) will also demonstrate the coronary sinus. An enlarged coronary sinus is suggestive of a persistent left superior vena cava (SVC). Mitral and tricuspid valve morphology and function can be assessed by 2D imaging, color flow Doppler and inflow spectral Doppler (PW and CW). This view is also useful for demonstrating intracardiac air following separation from cardiopulmonary bypass-air will rise to the highest site within the LV, along the interventricular septum.

Pulling up the probe and slight flexion from the mid-esophageal position will reveal the LVOT (Fig. 2). LVOT color Doppler can be recorded from this view, looking for either LV outflow tract obstruction (LVOTO), or aortic insufficiency. Usually the angle of insonation is poor for Doppler interrogation of flow velocities. Pulling the probe up higher in the esophagus to get just above the level of the LVOT followed by ante flexing and rotating counterclockwise slightly (i.e. to the left of the LVOT) will illustrate the main pulmonary artery (MPA), conduit and branch pulmonary arteries. This view provides an excellent angle for Doppler interrogation of the MPA, and often provides the clearest view of any valve in the pulmonary position (Fig. 3).

Lastly, relaxing the probe and advancing into the esophagus to return to the retrocardiac position and a four-chamber view and then rotation counterclockwise (i.e. tilting to the left) will demonstrate left pulmonary vein (LPV) flow into the LA with color Doppler (Figs 4A and B). Pulsed wave (PW) Doppler interrogation of this signal is useful in evaluating pulmonary vein flow: a large flow reversal with atrial contraction in the pulmonary veins may indicate LA pressure elevation, or junctional ectopic rhythm (Fig. 5), while a large flow reversal with ventricular contraction in the pulmonary veins indicates moderate to severe mitral regurgitation. A more chaotic signal may indicate other atrial rhythm problems. Imaging of the atrial and ventricular septum may be suboptimal in this view, as the imaging beam is parallel to these structures, which often results in image drop-out.
Fig. 5: An abnormal pulmonary venous Doppler patterns may indicate LA pressure elevation, or junctional ectopic rhythm, as seen in this image.

Fig. 6: A short-axis view of the aortic valve at 30 to 50° beam angle, midesophageal position (LA: left atrium; Ao: aorta; RV: right ventricle).

Fig. 7: At 60 to 80° in the midesophageal position, the tricuspid valve anatomy can be assessed, as well as any tricuspid regurgitation (RV: right ventricle; TR jet: tricuspid regurgitant jet).

Fig. 8: At 60 to 80° in the midesophageal position, the right ventricular anatomy can be assessed from tricuspid valve to pulmonary valve (RA: right atrium; RV: right ventricle; MPA: main pulmonary artery).

Fig. 9: At 60 to 80° in the midesophageal position, the interatrial septum may be viewed with slight counter clockwise rotation, to assess for any secundum atrial septal defects. (LA: left atrium; RA: right atrium).

Fig. 10: With rightward rotation toward the right axilla, the right pulmonary veins can be seen in long-axis as they approach the left atrium (RPVs: right pulmonary veins).
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Fig. 11: At 90°, with rightward angulation, the bicaval view can be displayed (SVC: superior vena cava; IVC: inferior vena cava)

Fig. 12: At 90°, leftward rotation will lead to a view of the left atrium, LV inflow, left atrial appendage and anterior and posterior mitral valve leaflets (LA: left atrium; LV: left ventricle; LAA: left atrial appendage)

Fig. 13: By further rotation the probe leftward from the left ventricular inflow and LA appendage view at 90° imaging, the descending aorta can be viewed in long-axis, and PW Doppler can be used to evaluate the pattern of flow. Here, holodiastolic retrograde flow is seen in the patient with a large patent ductus arteriosus (DAO: descending aorta)

Fig. 14: At 120° in the midesophageal position, the left atrium, mitral valve, left ventricle, LVOT, aortic valve and proximal ascending aorta in addition to portions of the IVS and right ventricle are seen (LA: left atrium; LV: left ventricle; AAo: ascending aorta)

Fig. 15: At 120° with leftward rotation of the probe from the LVOT view, one will see the left main, left anterior descending, and circumflex coronary arteries, with good alignment for Doppler PW interrogation of flow. (LMCA: left main coronary artery; Cx: circumflex coronary artery; LAD: left anterior descending coronary artery)

Fig. 16: At 120° with leftward rotation of the probe from the view of the left atrium, one will see the left pulmonary veins as they approach the left atrium
Aortic Valve Short Axis View (30-50°)

From the midesophageal five-chamber view (i.e. the LVOT axial view described above) changing the multiplane angle to approximately 30° will illustrate a cross-sectional view of the aortic valve that allows for assessment of valve cusp and commissure morphology. Color flow Doppler applied in this view can assess valve stenosis or regurgitation, as well as assess the etiology of the regurgitation (Fig. 6). The coronary origins can usually be viewed from this angle, although Doppler interrogation often not useful due to poor angle of incidence.

RV Inflow-Outflow View (60-80°)

From the aortic valve short axis view, rotating to a multiplane angle of 60 to 80° will reveal the right ventricular inflow-outflow tract view. First, view the tricuspid valve anatomy, and assess any tricuspid regurgitation (TR) using Doppler interrogation (Fig. 7). The right ventricular anatomy can be assessed from TV to PV (Fig. 8), in addition to qualitative assessment of RV function. This view may allow for assessment of PV diameter and valve motion, although the anterior location of the pulmonary valve may provide less clear imaging of this structure in the larger patient- in these instances the view at 0 to 30° is often useful.

Once the RV inflow, ventricle, and outflows have been assessed, the interatrial septum may be viewed with slight counterclockwise rotation, to assess for any secundum atrial septal defects (Fig. 9).

Pulling up further in the esophagus and rotating clockwise toward the right axilla, just past the SVC, with color Doppler on will allow for visualization of the right pulmonary veins as they enter the LA. Recall that the pulmonary veins are located inferior to the pulmonary arteries, and position the transducer accordingly. This view allows for visualization of all three RPVs as the approach the LA - they will be seen as red joining together at the top of the image, and are well aligned for Doppler interrogation (Fig. 10).

Midesophageal Aortic Valve Long Axis View (120°)

This view is obtained at the midesophageal level with an angle ranging from 120 to 140°. This view illustrates the left atrium, mitral valve, left ventricle, LVOT, aortic valve and proximal ascending aorta in addition to portions of the IVS and right ventricle (Fig. 14). This view is useful in the assessment of mitral and aortic valve pathologies, LVOT obstruction, VSDs in addition to assessment of septal thickness, systolic anterior motion (SAM) and mitral regurgitation (MR) in hypertrophic cardiomyopathy (HCM).

Slight leftward rotation of the probe while focusing on the left coronary cusp will display the LMCA, LAD and CX, with good alignment for Doppler PW interrogation of flow (Fig. 15). The left pulmonary veins can also be seen if one returns to the LV inflow/outflow view, pulls the probe up slightly in the esophagus and rotates counterclockwise (i.e. to the left) with color flow Doppler on. If the long axis of the descending aorta is seen, the rotation is too far. The left upper and lower pulmonary veins will be seen joining together at the top of the image, as they join the LA (Fig. 16). PW Doppler of the PVs can be used for further interrogation.
Fig. 17: At 0 to 20°, transgastric short axis view, a short axis view of the LV is seen (LV: left ventricle; RV: right ventricle)

Fig. 18: At 0 to 20°, transgastric short axis view, with some slight rightward angulation from the LV short axis view, a short axis view of the mitral valve is seen (MV: mitral valve)

Fig. 19: The right ventricular outflow tract is imaged by further advancement of the probe into the stomach further and rightward rotation while maintaining flexion (RVOT: right ventricular outflow tract; LV: left ventricle)

Fig. 20: Transgastric imaging at 90° provides a sagittal plane through the LVOT (LVOT: left ventricular outflow tract; RV: right ventricle)

Fig. 21: A secundum atrial septal defect is imaged at 0°, a catheter is seen crossing the defect (ASD: atrial septal defect)

Fig. 22: During transcatheter device closure of an atrial septal defect, the defect and the catheter course can be followed at beam angle of 30 to 50° (LA: left atrium; RA: right atrium)
Transgastric Views

Imaging the heart with the probe in the stomach will provide additional information to that acquired from midesophageal imaging. These views are, in the authors experience, a bit more variable, since the probe position within the stomach, and the degree of flexion obtained is less consistent between patients than in retrocardiac imaging.

Transgastric Short-axis View (0-20°)

This cross-sectional view is obtained by advancing the probe into the stomach followed by flexion, and provides a short axis view of the mitral valve and LV that is useful for the quick assessment of LV filling and global and segmental LV function (Fig. 17). In every postoperative patient separating from bypass, a moment should be taken to specifically use this view to make an assessment about ventricular function.

Pulling up a little higher while maintaining probe flexion will demonstrate mitral valve anatomy (Fig. 18). Assessment of the mitral valve with CD in this view may provide information about the morphology and site of the origin of any mitral regurgitant jet. If the probe is advanced into the stomach further and rotated rightward while maintaining flexion, one will image the right ventricular outflow tract (Fig. 19). At times, the RVOT is imaged by simply adjusting the angle of the imaging beam until the RVOT is seen, and the resultant angle may be somewhat variable.

Transgastric Sagittal View (90°)

This view can be complementary to the deep transgastric short axis view and is obtained with the probe deep in the stomach, rotating to the left, and flexing with a multi-plane angle of 90° (Fig. 20). Data acquired from this view is helpful is assessing the anatomy of the LVOT, aortic valve and ventricular septum and provides an excellent alignment for spectral Doppler interrogation of the LVOT.

TRANSESOPHAGEAL IMAGING OF SPECIFIC CONGENITAL LESIONS

Atrial Septal Defects

Defects of the interatrial septum are the third most common type of congenital heart defect with an estimated incidence of 56 per 100,000 live births, however this incidence appears to be increasing with improved echo detection - recent estimates are thought to approach 100 per 100,000 live births.

There are four main morphological types of atrial septal defects (ASD), and the goals of imaging should be to discern which type this patient has, as well as assess for known associated findings.

Secundum ASDs

After patent foramen ovale, secundum ASDs are the most common cause of an atrial level shunt and are centrally located in the region of the fossa ovalis. The defect is surrounded by atrial septal tissue, and the defect itself is not confluent with any of the venous structures entering the atria.

Echocardiography: Transcatheter closure of a secundum atrial septal defect is often performed with guidance using transesophageal imaging. At 0°, the total septal length may be assessed, as well as the mitral and tricuspid valves. This is especially valuable when a large device may be deployed, which may affect AV valve closure in some cases. Thus, a baseline view of valve function/regurgitation prior to device deployment is prudent for the purpose of providing valve function comparison to post deployment images. In addition, confirming normal pulmonary venous return to the LA can be done at 0°. The atrial septum and the defect are often well imaged at 30 to 50° (Fig. 21). This angulation also allows for the delineation of the catheter course from IVC to LA via the defect (Fig. 22). The 90° view images the superior and inferior septal rims when imaged in the bicaval plane (Fig. 23).

Primum ASDs

Primum ASDs are located in the inferior portion of the interatrial septum extending to the junction of the atrioventricular valves. The floor if the primum ASD is the atrioventricular valves. Premium ASDs are frequently associated with partial or complete atrioventricular septal defects (AVSD). A partial AVSD is comprised of a primum ASD, cleft mitral valve, and may have LVOT obstruction if chordal attachments to the interventricular septum obstruct the LV outflow. Complete AVSD include a primum atrial and inlet ventricular septal defect, and can have a single large atrioventricular valve separating right and left atria from their respective ventricles.

Echocardiography: At 0°, the defect is often well seen, and in the case of AVSDs, the inferior position of the plane of the AV valve is noted (Fig. 24). The mitral valve imaging may be performed initially at 0°. Often, when a cleft is present, the regurgitant jet is noted to be located more medially than at the coaptation points (Fig. 25). In addition the anterior leaflet may appear ‘split’, which is actually an image of the two separate leaflets overlapping within the ultrasound beam. This defect is not amenable to transcatheter closure.

Superior Sinus Venosus ASDs

Superior sinus venosus ASDs are defects in the wall between the right upper pulmonary veins, the posterior SVC and
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Fig. 23: The secundum atrial septal defect is viewed at the 90° bicaval view (LA: left atrium; RA: right atrium; SVC: superior vena cava; ASD: atrial septal defect)

Fig. 24: A primum ASD is seen at 0°, as well as both AV valves at the same level (LA: left atrium; RA: right atrium; LV: left ventricle, RV: right ventricle)

Fig. 25: The regurgitant jet through a cleft mitral valve is seen at 0° (LA: left atrium; RA: right atrium; LV: left ventricle; RV: right ventricle)

Fig. 26: At 0°, when the probe pulled cranially while viewing the SVC, an anomalous right pulmonary vein is seen entering it (RUPV: right upper pulmonary vein; SVC: superior vena cava)

Figs 27A and B: (A) In the sagittal plane of the left atrium and left ventricle, the normal coronary sinus (CS) septum is seen, (B) the absence of septum is evident in this patient with a CS septal defect

Fig. 28: In this image, which is the same orientation as demonstrated in Fig. 14, contrast was injected into the left arm. Opacification was seen first in the coronary sinus (CS), then a brief flash of contrast was seen entering the left atrium (LA), establishing the diagnosis of CS septal defect. This contrast was then washed back into the CS and into the right atrium, so that overall there was scant contrast seen in the left ventricle
the high atrial septum. Typically, the SVC appears to sit over the defect. The relatively uncommon inferior types are found near the inferior vena caval-atrial junction. Sinus venous defects are often associated with partial anomalous pulmonary venous drainage especially the right-sided veins.23 PAPVC is more common in patient with sinus venous ASDs—occurring in up to 85% of cases, compared to a 10 to 15% incidence PAPVC in patients with secundum ASDs.24,25 Often, it is the presence of the associated lesion which leads to detection of the anomalous PVC. TEE has been shown to be superior to TTE in the diagnosis of sinus venous defects.26,27

**Echocardiography:** At 0°, the defect is often not well seen, as the plane of this defect is perpendicular to the imaging plane. However, at 90° the defect is well seen, as well as the appearance of SVC “override” over the defect. Anomalous right pulmonary veins draining to the SVC are often best visualized by imaging the SVC/RA junction at 0° (which transects the SVC in the horizontal plane), and withdrawing as you observe the transition from high RA to the round shape of the SVC. If one sees a change in the round shape to a teardrop appearance, then you are likely seeing the point of entry of an anomalous RPV into the SVC (Fig. 26).28 Similarly, anomalous venous connections to the free wall of the RA can be assessed by advancing the probe within the esophagus.

**Coronary Sinus Septal Defects**

Coronary sinus septal defects are rare and are the result of a communication between the left atrium and coronary sinus. They are defined on echocardiography as an enlarged coronary sinus with a deficient roof and are frequently associated with a persistent left SVC.

**Echocardiography:** In the parasternal long axis view, a dilated coronary sinus may present the first clue that an abnormality is present. The coronary sinus ostium may be dilated, and may be confused with a primum ASD—the mouth of the coronary sinus may be distinguished from a primum ASD on the basis of a normal-appearing mitral valve, and the coronary sinus orifice’s location posterior to the MV annulus. This is observed as a defect that becomes more visible when the probe is advanced from the usual atrial mitral valve, atrial septal junction to the coronary sinus view. In the sagittal plane, in the two-chamber view, the dilated coronary sinus is seen, as is the septum separating the coronary sinus from the left atrium. A defect in this septum is clearly demonstrated using this plane of imaging (Figs 27A and B).

Contrast echocardiography is helpful— injection into a left arm vein will result in sequential opacification of the LSVC, coronary sinus, left atrium (Fig. 28), and finally the right atrium via the coronary sinus orifice;29 although this sequence is variable as it depends on regional pressure gradients. For example, if the shunt direction is predominantly left to right (that is, from the left atrium, to the coronary sinus via the defect, to the right atrium), injection into the left arm will stream predominantly into the right atrium, with negative contrast seen from the left atrium into the coronary sinus.

**Ventricular Septal Defects**

Isolated ventricular septal defects (VSD) are the most common congenital cardiac lesion identified in infancy.21 While some—particularly those located in the perimembranous or muscular region—close spontaneously, 15 to 20% of patients with isolated VSDs require surgery. VSDs are classified into 4 main groups according the location of the defect in the ventricular septum.

**Perimembranous VSDs**

Perimembranous VSDs are subaortic (outlet) defects, located between the tricuspid and aortic valves. They may be associated with ventricular septal aneurysms or tricuspid tissue which may restrict the flow of blood across the defect. Secondary lesions may occur in perimembranous defects, and a thorough investigation for each of these should be part of any preoperative transesophageal echocardiogram.

A double chambered right ventricle (DCRV), also known as an anomalous muscle bundle within the right ventricle, may develop in the setting of a perimembranous VSD.30 Subaortic stenosis has been noted in a subset of patients with perimembranous defects,31 and is thought to be related to altered shear stress forces on the endothelial surface of the septum.32 Perimembranous VSDs can also be associated with distortion of an adjacent aortic valve cups due to herniation into the defect. This is thought to be due to a suction on the valve from a Venturi effect, caused by flow acceleration from the LV into the RV via the defect.33 In the setting of a restrictive and otherwise hemodynamically insignificant intracardiac shunt, progressive aortic regurgitation may in itself be an indication for surgical intervention.

**Echocardiography:** In the 0° view, when viewing the AV valves, the perimembranous VSD is often not seen. The VSD may be observed in the 0° view when the probe is drawn upward within the esophagus to view the LVOT and its entry to the aorta. Preoperatively, an assessment of the AV valves at 0°, particularly the tricuspid valve, is helpful to provide a comparison for postoperative tricuspid valve function. A VSD patch may distort the tricuspid valve or interfere with septal leaflet function, and having images of the valve, its motion, and regurgitation is critical to the assessment of any
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**Fig. 29:** In the midesophageal position at 30 to 50°, the septum between the tricuspid valve (TV) and aorta is seen well, and any defects in this region is well demonstrated (RV: right ventricle; VSD: ventricular septal defect)

**Fig. 30:** A right ventricular anomalous muscle bundle is seen in any view of the RVOT, here demonstrated at 30 to 50°

**Fig. 31:** From a transgastric view at 0°, muscular ventricular septal defects can be noted in both 2D imaging (left) or with color Doppler imaging (right) (LV: left ventricle; RV: right ventricle; VSD: ventricular septal defect)

**Fig. 32:** An inlet VSD is seen in the 0° plane, at the level of the AV valve opening, with the AV valves at same level (LA: left atrium; RA: right atrium; LV: left ventricle; RV: right ventricle; VSD: ventricular septal defect)

**Fig. 33:** At 30 to 50°, the RV inflow to outflow is seen, and the supracristal defect can be found in this view immediately below the pulmonary valve. The subpulmonic position is seen by 2D imaging, and disturbed flow into the pulmonary valve is seen with color Doppler imaging (RA: right atrium; RV: right ventricle; VSD: ventricular septal defect; MPA: main pulmonary artery)

**Fig. 34:** This diagram depicts the abnormal connections in complete transposition of the great arteries (LA: left atrium; RA: right atrium; LV: left ventricle; RV: right ventricle; Ao: aorta; PA: pulmonary arteries)
postoperative valve regurgitation. At 30 to 50°, the defect between the tricuspid valve and aorta is seen well (Fig. 29). This view is also useful for assessing with 2D imaging the RV myocardium, looking for any anomalous muscle bundles that may signal a double chambered RV (Fig. 30). Imaging at 90° with slight angulation rightward may also provide views of the VSD, and is a particularly useful view of the LVOT and aortic valve in the assessment for subaortic obstruction and aortic valve distortion/aortic regurgitation. In the setting of VSD repair with subaortic stenosis resection, perioperative evaluation of the LVOT with both color and continuous wave Doppler is useful to assess the degree of stenosis.

Muscular VSDs

Muscular VSDs account for 20% of VSDs and occur in the muscular, or trabecular portion of the ventricular septum. Apical muscular VSDs can be particularly challenging to identify by either transthoracic or transesophageal imaging. When imaging, sometimes the first clue to the presence of an apical muscular VSD is a sense of disconnect between adjacent septal segments—once this clue is pursued with focused imaging, the defect can be better appreciated. Any time a muscular VSD is identified, one must suspect multiple muscular VSDs, and image with purpose to either identify or rule this possibility out.

Echocardiography: If the ventricular pressures are identical- for example, if there is a large outlet VSD—identification of additional muscular VSDs can be quite difficult. This is because the sentinel finding of a high velocity color jet across the septum will be lacking in the setting of high RV pressures. In addition, muscular VSDs may be unrestrictive, yet challenging to identify due to septal overlap in some imaging planes. The muscular septum is best interrogated in a series of sweeps, first with 2D imaging at the highest resolution available, followed by color Doppler imaging. At 0°, the ‘sweep’ takes form in advancement of the probe within the esophagus. The transgastric views are useful for apical imaging, and transgastric imaging of the interventricular septum at 0 to 20°, both at neutral, with leftward, and then rightward angulation (Fig. 31), and additional imaging at 90 to 120° will interrogate much of the trabecular septum.

Inlet VSDs

Inlet VSDs are located posterior and inferior to the septal leaflet of the tricuspid valve are termed inlet VSDs, or VSDs of the AV canal type. Echocardiographically, these defects are recognized by seeing the AV valves at the same level without the normal offset and inferior tricuspid valve position. Inlet VSDs may be associated with a straddling tricuspid valve. An associated primum ASD indicates that this is not an isolated inlet VSD, but is part of the spectrum of an AVSD.
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Figs 38A to F: Typical patterns of coronary artery anatomy variations in d-TGA, as viewed from an esophageal window: (A) Usual pattern: left main from the left facing sinus, RCA from the right facing sinus (B) Cx from RCA: LAD from the left facing sinus, RCA and Cx from the right facing sinus, (C) Single RCA: RCA and left main from the right facing sinus, (D) Single LCA: Left main and RCA from the left facing sinus; (E) Inverted Origins: Left main from the right facing sinus and RCA from the left facing sinus, (F) Inverted RCA and Cx: LAD and RCA from the left facing sinus and the LCx from the right facing sinus, RCA: right coronary artery; LCA: left coronary artery; LAD: left anterior descending artery; Cx: circumflex artery; Ao: aorta; PA: pulmonary artery

Echocardiography: The inlet defect is best viewed in the 0° view, when viewing the AV valves, after the probe is advanced within the esophagus to view the region just posterior and inferior to the atroventricular valves. Special focus on the tricuspid valve, with assessment for tricuspid valve straddle, is indicated in the setting of an inlet VSD and especially so when there is some degree of RV hypoplasia. In the transgastric 0° plane, at the level of the mitral valve opening, the defect may also be visible with some rightward angulation. If the defect is clearly seen between the AV valves, it is likely that it is an inlet VSD, or an inlet VSD with trabecular extension (Fig. 32).

Doubly Committed Subarterial VSDs
Doubly committed subarterial VSDs are found in the outlet septum immediately below the aortic and pulmonary valves. There is fibrous continuity between the pulmonary and aortic valves, which are consequently at the same level due to lack of interposed septum. They can also be referred to as
supracristal VSDs. Due to the lack of support to the aortic valve from below, as well as the close proximity of the aortic valve leaflets to the VSD shunt, these defects have a higher incidence of aortic coronary cusp prolapse with resultant aortic regurgitation.35

Echocardiography: Since the doubly committed subarterial defect is located underneath both the aortic and pulmonary valves, it is located in the leftward outlet septum. At 30 to 50°, the RV inflow to outflow is seen, and the defect can be found in this view immediately below the pulmonary valve (Fig. 33).

D-Transposition of the Great Vessels

Complete transposition of the great vessels (d-TGA) is a congenital heart condition where great vessel arises off of the inappropriate ventricle and often results in severe cyanosis secondary to inadequate mixing between the systemic and pulmonary circulations (Fig. 34). Intercirculatory mixing is necessary for survival and can occur at the level of the atrial septum, ventricular septum, or patent ductus arteriosus.

d-TGA requires surgical intervention for long term survival given a 52% mortality by 1 month of age and a 89% mortality by 1 year without therapy.36 Overall, this cardiac condition is rare, with an overall incidence of 0.27 per 1000 births and it ranks 7 (9.1%) among other forms of congenital heart disease.37,38

There are two basic categories of d-TGA: simple and complex. Simple d-TGA has no other associated cardiac lesions other than an atrial communication and patent ductus arteriosus.

Echocardiography: From the standard 0°, four chambers can be identified with a right-sided right ventricle and a left-sided left ventricle. A patent foramen ovale or secundum atrial septal defect will be present and will likely shunt left to right by color Doppler (Fig. 35). The systemic and pulmonary venous return will be normal since venous abnormalities are not usually associated with this condition. With flexion or withdrawing the TEE probe cranially, the great vessels can be identified parallel to each other; the aorta will arise off of the right ventricle while the main pulmonary artery will arise off of the left ventricle (Fig. 36). Frequently the pulmonary artery bifurcation can be visualized to establish the imaging beam to 110 to 130° to view the posterior main pulmonary artery and the anterior aorta (Fig. 39). Transgastric imaging will avail the same image from 50 to 80° (Fig. 40). Transgastric views of the LV short axis should be used to get a baseline assessment of function/wall motion abnormalities. This can be compared to the postoperative imaging to detect any changes.

Complex d-TGA

Complex d-TGA may be associated with a ventricular septal defect, left ventricular outflow tract (subpulmonary) obstruction, or the more rare findings of juxtaposed atrial appendages, or situs inversus totalis.

Echocardiography: From the standard 0° image, muscular ventricular septal defects can be noted. Whenever one ventricular septal defect is determined, additional defects must be excluded. Color Doppler can be helpful to determine the presence of a defect and direction of shunting; however, since frequently there is a large patent ductus arteriosus, the flow will be of low velocity and can be difficult to assess. Flexing, or withdrawing the probe cranially can be useful to detect perimembranous ventricular septal defects. Imaging at 30 to 50° with some leftward rotation is useful to detect subpulmonary obstruction, whereas 90 to 120° is more suited to image left ventricular outflow tract obstruction (Fig. 41).

Tetralogy of Fallot

Tetralogy of Fallot (TOF) results from anterior deviation of the conal septum, resulting in an anterior malalignment ventricular septal defect, aortic override, and varying degrees of pulmonary stenosis. Right ventricular hypertrophy is the fourth feature, hence tetralogy, and is secondary to the high pressure exerted upon it. Tetralogy of Fallot is a spectrum related to the degree of right ventricular outflow tract obstruction- the severity of the obstruction leads to different physiology at manifestation. If there is very mild pulmonary stenosis, TOF is similar to ventricular septal defect physiology, whereas severe pulmonary stenosis or atresia results cyanosis, since the ventricular septal defect shunt is from the right to left ventricle. TOF is one of the most common forms of cyanotic congenital heart disease; however, the overall prevalence is only 0.49 in
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**Fig. 41:** As in any form of LVOT obstruction, subpulmonary obstruction in d-TGA can be seen well from the 120° transgastric view. (LVOTO: left ventricular outflow tract obstruction; Ao: aorta; PA: main pulmonary artery)

**Fig. 42:** The aortic override of the VSD is seen at 0° when the probe is withdrawn to view the LVOT (Ao: aorta; VSD: ventricular septal defect)

**Fig. 43:** Some excessive RV muscle bundles are restricting the VSD in this view

**Fig. 44:** With the imaging bear at 60 to 90° for an RVOT view, the degree of infundibular obstruction and pulmonary hypoplasia can be assessed

**Fig. 45:** In the 120° view of the LVOT in TOF, the VSD, anterior displacement of the aorta and conal septum are seen, and aortic override of the VSD are seen (Ao: aorta; LV: left ventricle; RV: right ventricle)

**Fig. 46:** From a transgastric window (here, at 0°), the RVOT obstruction from the anteriorly deviated conal septum is seen
1000 live births. The associated defects with TOF include: coronary artery anomalies, right-sided aortic arch with a retroesophageal (aberrant) subclavian artery, major aortopulmonary collaterals, main and branch pulmonary artery stenosis, secundum atrial septal defect, additional muscular ventricular septal defects, persistent left superior vena cava, anomalous pulmonary venous return, aortic abnormalities and atrioventricular canal.

**Echocardiography:** From the standard 0° view, four chambers will be identified and the conotruncal defect of TOF is difficult to appreciate. A patent foramen ovale or secundum atrial septal defect must be detected from this view. Given the possibility of venous anomalous, one must confirm appropriate drainage of the pulmonary veins into the left atrium with clockwise and counterclockwise rotation of the TEE probe. Of note, the cardiac mass may be more horizontal than normal due to the TOF defect rendering the assessment of tricuspid or mitral Doppler difficult. Additionally, a dilated coronary sinus may be noted and indicate a persistent left superior vena cava. The entire ventricular septum should be scanned for muscular ventricular septal defects which can be difficult since the right and left ventricular pressures should be equal, due to the large anterior malalignment VSD of TOF. With anteflexion and superior withdrawal of the TEE probe, this anterior malalignment ventricular septal defect will be noted with an overriding aorta (Fig. 42). At this level, it is important to establish if there is right to left or left to right ventricular shunting and determine if there is any restriction to flow. The vast majority of TOF ventricular septal defects are large and unrestrictive; however, there are cases of restrictive ventricular septal defects which are usually caused by either the tricuspid valve tissue of muscle bundles (Fig. 43). In these cases, the right ventricular pressure will be suprasystemic. Withdrawing the probe toward the base of the heart, the aortic valve will be visualized. Given the association of coronary artery anomalies, the short axis of the aortic valve should be used to track each coronary artery carefully. The most important coronary anomaly is finding a large coronary artery crossing the infundibulum such as the left anterior descending coronary artery. With a transannular patch repair, this type of vessel can be transected. When the probe is rotated to 60 to 90° for an RVOT view, the degree of infundibular obstruction and pulmonary hypoplasia can be assessed (Fig. 44). The pulmonary valve at its hinge points should be measured in systole to establish the degree of hypoplasia compared to the normal population for which there are normal values (http://parameterz.blogspot.com/).

At 100 to 120° views, the ventricular septal defect of TOF can be more readily visualized and the direction of shunting can be established (Fig. 45). With further cranial withdrawal of the TEE probe, one can determine the size of the aortic root including the aortic annulus, sinuses, sinotubular junction and ascending aorta. These aortic dimensions can be enlarged in TOF. The degree of aortic insufficiency should also be established with color Doppler. Transgastric imaging is also very important in patients with TOF. The 0° transgastric view of the ventricular septum can be used to determine the presence of additional ventricular septal defects by sweeping in a superior-inferior direction. Views of the RVOT—at both 0° and in the orthogonal plane at 90°—are very useful in TOF to establish the degree of subpulmonary obstruction (Fig. 46).

**Double Outlet Right Ventricle**

Double outlet right ventricle (DORV) is a less common form of congenital heart disease, occurring in about 12.7 per 1000 live births. This can be a very challenging form of CHD with much anatomic variability. What is common throughout the types of DORV are that both great arteries arise completely or predominantly from the right ventricle, and the posterior great vessel is separated from the mitral valve and ventricular mass by a segment of muscular tissue, or conus septum, which is considered by many to be the key distinguishing feature of DORV. Since both great vessels arise from atop a conus, the semilunar valves are near or at the same level. A ventricular septal defect is almost always present, and in many instances, the posterior great vessel may override the ventricular septal defect. What is variable in DORV are the (1) relationship between the VSD and the great arteries, (2) position of the great arteries in relation to each other and (3) presence of additional malformations. Each of these features are used in the classification of DORV.

**DORV with Subaortic VSD**

DORV with subaortic VSD occurs in about 50% of all DORV. In this subtype, the VSD may be in continuity with the tricuspid valve, and the aortic valve or the subaortic conus is superior margin of VSD. The great vessel arrangement is often close to normal—the aortic position is rightward, and either side by side or slightly posterior to the pulmonary valve. The resultant physiology is similar to a simple VSD physiology, with a significant left to right shunt, and repair consists often of only VSD closure. This type of DORV may be associated with either subvalvular pulmonary stenosis—when this is present, the physiology more closely resembles the physiology of TOF, depending on the degree of PS.

**Echocardiography:** At 0°, the view at the level of the AV valves is relatively normal, until the probe is drawn upward in the esophagus to view the LVOT. At this point, the VSD
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Fig. 47: The RVOT anatomy is well seen at 30 to 50° as well as in the transgastric view of the RVOT (PA: main pulmonary artery)

Fig. 48: In Taussig-Bing form of DORV, when the probe is withdrawn to demonstrate the ventriculoarterial connections, the posterior position of the pulmonary artery, and the commitment of the VSD to the pulmonary artery, are seen (Ao: aorta, PA: main pulmonary artery)

Fig. 49: Transgastric imaging at 0 to 30° demonstrates the great vessels, the conal septum guarding the subaortic region, and the subpulmonary position of the VSD (Ao: aorta; PA: main pulmonary artery)

Fig. 50: Transgastric imaging at 90° demonstrates the subpulmonary position of the VSD (VSD: ventricular septal defect, PA: main pulmonary artery)

Fig. 51: The Doppler pattern in the descending aorta can be assessed for evidence of residual arch obstruction. A normal upstroke is demonstrated here, whereas a blunted or delayed upstroke may indicate obstruction

Fig. 52: An large inlet VSD is seen in this 90° midesophageal window, as well as a large, posteriorly positioned pulmonary valve, in this patient with DORV and a remote VSD (LA: left atrium; RA: right atrium; PA: main pulmonary artery; VSD: ventricular septal defect)
is seen, along with the overriding aortic valve. This can appear virtually identical to tetralogy of Fallot, however in the 120° view, the separation between aortic valve and mitral valve can be observed. The RVOT is interrogated at 30 to 50°, as well as in the transgastric view of the RVOT (Fig. 47). Often, anterior displacement of the outlet septum is seen as the etiology of the RVOT obstruction.

**DORV with a Subpulmonary VSD**

DORV with a subpulmonary VSD occurs in about 25% of all DORV. Is is also known as Taussig Bing malformation. In this anatomic variant, the aortic valve is rightward relative to the pulmonary valve, and is either slightly anterior to or beside the pulmonary valve. The VSD is located beneath the pulmonary valve, and the conal septum ‘shields’ the aorta somewhat from the VSD flow. Since most of the VSD flow from the LV is directed toward the PA, and most of the flow from the RV is directed toward the aorta, these patients can have transposition physiology. In addition, this lesion is associated with a 50% incidence of aortic arch hypoplasia and/or interruption. Surgical repair usually consists of an arterial switch operation, VSD closure and possibly arch repair/reconstruction.

**Echocardiography:** At 0°, the view at the level of the AV valves is relatively normal, until the probe is drawn upward in the esophagus to view the LVOT (Fig. 48). At this point, the VSD is seen, along with the posterior semilunar valve, which then bifurcates into the branch pulmonary arteries, identifying this valve as a pulmonary valve. At 30 to 50°, the anterior location of the aorta may be seen, and the size of the aortic valve annulus may be noted to be smaller than the more posterior pulmonary valve. Transgastric imaging at 0 to 30° may demonstrate both great vessels and the relative sizes of their valve annuli (Fig. 49), while in the sagittal plane (90°) may demonstrate the subpulmonary positioning of the VSD (Fig. 50). When arch hypoplasia or interruption is present and arch repair is planned as part of the complete repair, then imaging and PW Doppler of the descending aorta pre and postoperatively is recommended to assess, especially in the postoperative period- for any delay in systolic upstroke which could indicate residual arch obstruction (Fig. 51). This image is achieved by rotating the entire TEE probe posteriorly to view the descending aorta, playing it in it cross section in the center of the imaging plane, and then rotating the transducer beam from 0° to 90° to view the descending aorta in its long axis. With slight flexion, a mild angulation of the aorta is seen, which can then undergo Doppler interrogation. The angle in incidence will be high—but it is the pattern of the Doppler signal, and not its actual amplitude—that is going to be assessed.

**DORV with a Noncommitted VSD**

In this anatomic variant of DORV, the VSD is most commonly located in either the inlet or the muscular portion of the interventricular septum. Approximately 15% of DORV are of this type. The VSD is termed ‘noncommitted’ or ‘remote’ because of the distance between it and the semilunar valves. Surgical repair may be possible by routing the defect to a semilunar valve via a baffle, or may not be possible, leaving single ventricle palliation as the only surgical option.

**Echocardiography:** The imaging is mainly dependent upon the location of the VSD. In the case of an inlet VSD, the defect may be seen at 0°, yet its relationship to the great vessels is better appreciated at a sagittal or orthogonal plane, around 90° (Fig. 52). The transgastric views may be particularly helpful for demonstrating the relationship between defect and semilunar valves, and the plane of imaging where this is best displayed may be dependent on the individual anatomy.

**SUMMARY**

This document reviews a suggested comprehensive TEE examination, and which views may be particularly useful in specific types of congenital heart disease. In general, a complete study is recommended in the immediate preoperative period in the operating room, to thoroughly evaluate the anatomy before surgical intervention commences. Postoperatively, a more directed study is more practical, and is directed to evaluating both the surgical procedure as well as to reevaluate ventricular function.

In congenital heart disease, there are not only spectacular variations in anatomy, as well as unexpected findings. Taking a systemic, similar, organized approach to imaging these patients is essential to an accurate imaging evaluation.

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