Acyanotic Congenital Heart Disease and Transesophageal Echocardiography

Abstract
The spectrum of congenital heart disease (CHD) seen in the adult varies widely. Malformations range from mild anomalies requiring no intervention to extremely complex pathologies characterized by the presence of multiple coexistent defects. Echocardiography represents the primary noninvasive imaging modality in the assessment of these lesions. The transesophageal approach expands the applications of echocardiography by allowing the acquisition of anatomic and functional information that may not be obtainable by transthoracic imaging.

Keywords: Acyanotic congenital heart disease, atrial septal defect, transesophageal echocardiography

Introduction
Transesophageal echocardiography (TEE) is an essential diagnostic and monitoring device in pediatric cardiac surgery for confirming preoperative diagnoses, demonstrating previously unappreciated anatomic details, formulating surgical plans, evaluating immediate surgical results, detecting residual lesions which are significant, and guiding surgical revisions.[1‑5]

Atrial Septal Defect
When larger than 5 mm, a patent foramen ovale is considered a secundum atrial septal defect (ASD). Primum ASD is at the inferior aspect of the interatrial septum and is associated with a cleft mitral valve (MV). Sinus venosus ASD is often associated with partial anomalous pulmonary venous return. The least common ASD is an unroofed coronary sinus, and it is often associated with left superior vena cava. The right ventricle (RV) versus left ventricle (LV) compliance, rather than the size of ASD, determines the amount of shunting. RV volume load occurs. High pulmonary artery (PA) pressures may occur with advancement in age. Volume overload creates a paradoxically moving interatrial septum. As with ventricular septal defect (VSD), definitive treatment is indicated when pulmonary blood flow (PBF): systemic blood flow (Qp:Qs) is >1.5:1. ASD can be closed primarily or with pericardial/Dacron patch. Transcatheter closure is another option.[6,7]

Secundum atrial septal defect
Defect is seen in the foramen ovale area in mid esophageal four-chamber view (ME 4C V) - Figure 1a, mid-esophageal bicaval view (ME bicaval V) Figure 1b, mid-esophageal aortic valve short axis view (ME AV SAX V), mid-esophageal right ventricular inflow-outflow view (ME RV inflow-outflow V), and transgastric (TG) bicaval[8] equivalent views.

TEE guidance is used for transcatheter closure. MV and tricuspid valves (TV) as well as total septal length is assessed in ME 4C V. MV must be examined for prolapse. When a large device is deployed, atrioventricular valve (AVV) closure may be affected. Diameter of ASD, septal rims and total septal length are measured using TEE. Baseline view of valve function before device deployment must be acquired to enable comparison with postdeployment image. Catheter course from inferior vena cava (IVC) to right atrium (RA) and then to left atrium (LA) via defect is seen in ME AV SAX V. The superior and inferior septal rims are seen in the bicaval view.

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Primum atrial septal defect

Defect is seen using ME 4C V [Figure 1c] in the posterior and inferior part of interatrial septum, above the AVVs. Both these valves originate at the same level. Primum ASD is frequently associated with partial or complete atrio-VSD. It is also commonly associated with cleft MV. Using color flow Doppler (CFD) and spectral Doppler Mitral regurgitation if present must be diagnosed and graded.

It is necessary to look for left ventricular outflow tract (LVOT) obstruction using two-dimensional (2D) TEE, CFD, and spectral Doppler in modified ME 5C V, mid esophageal aortic valve long axis view (ME AV LAX V), mid esophageal long axis view, and deep TG long axis view (deep TG LAX V). LVOT obstruction may be present due to:

- Aberrant chords inserting into interventricular septum
- discrete membrane in LVOT
- “goose neck deformity” or long tunnel type obstruction of LVOT.

Sinus venosus atrial septal defect

The defect is to be looked for in ME 4C, ME bicaval, and deep TG bicaval equivalent views. Superior defect (more common) is located near superior vena cava (SVC) above foramen ovale [Figure 1d]. The inferior defect (rare) is situated below foramen ovale, in inferior part of interatrial septum, near IVC [Figure 1e]. These ASDs are associated with partial anomalous pulmonary venous drainage. CFD and pulse wave Doppler (PWD) are used to assess drainage of all pulmonary veins. ME 4C V and ME bicaval V with rightward rotation is used to look for right pulmonary veins. ME 4C and ME 2C views are used to look for left pulmonary veins. By advancing and withdrawing the probe the SVC and IVC needs to be searched for high normal velocity flow or aliasing, suggestive of anomalous pulmonary venous drainage of upper and lower pulmonary veins.

For obtaining the modified deep TG bicaval view,[8] the standard TG short axis view is first obtained. The probe flexion is then undone. In the neutral position, the probe is advanced 2–3 cm and rotated in the clockwise direction by 90°. The probe is then antiflexed and the multiplanar angle switched to 90°–110°. The SVC is seen in the center of the sector scan. Using this view, the sinus venousus ASD can be visualized [Figure 2]. In this view, SVC-RA junction can be aligned with spectral Doppler to get flow gradients following surgical repair of sinus venosus ASD.

Coronary sinus atrial septal defect

The defect may be anywhere along the course of the coronary sinus. It is looked for using ME 4C [in area of coronary sinus in AV groove – Figure 1f], modified ME bicaval view near entrance of IVC, and ME 2 CV.

Contrast saline solution should be injected into left hand to determine whether left SVC (LSVC) is present. Immediate opacification of LA occurs before RA if LSVC and coronary sinus ASD are present. Bubbles first enter RA (via innominate and right SVC) when there is coronary sinus ASD without LSVC.

In all types of atrial septal defect

- The interatrial septum must be examined for multiple defects and the size of ASD must be measured in multiple planes. CFD and PWD are used to identify the size and direction of flow
- The pulmonary veins must be examined to find out where they drain. Using PWD velocity of flow in pulmonary veins can be assessed. Although the velocity of flow in pulmonary veins may be high due to high PBF, peak and mean velocities are not significantly elevated
- It is necessary to look for RA and RV enlargement and to assess biventricular function
- MV and TV annulus must be measured. Prolapse or cleft in MV needs to be looked for. CFD and spectral Doppler is used to grade any Mital or/and TR. LSVC is
searched for between left upper pulmonary vein (LUPV) and left atrial appendage (LAA) in ME 2C V and ME 4C V.

- The pulmonary valve (PV) must be examined (ME ascending aortic SAX and TG RV basal view) for leaflets doming, commissural fusion, and restricted motion. From TG basal SAX view, turn the probe clockwise to see TG RV basal view. CFD and continuous wave Doppler (CWD) are used to look for Pulmonary stenosis. Velocities of up to 2.5 m/s at the PV may occur due to ASD flow.

**Transesophageal echocardiography assessment after surgery/device closure**

- ASD closure is confirmed preferably with bubble contrast study.
- MV needs to be assessed and any MR quantified.
- Following sinus venous ASD repair, proper drainage of pulmonary veins into LA and absence of narrowing of pulmonary veins, SVC, SVC baffle, and SVC-RA appendage anastomosis (post-warden repair) must be confirmed. If superior vena cava-right atrial appendage anastomosis is narrow, increase PWD gradients are observed at this site in TG RV inflow view and TG RV inflow-outflow view. Usually, SVC-RAA gradient of more than 5 mmHg is unacceptable. Tacy[9] have mentioned that loss of biphasic pattern in SVC is more indicative of obstruction rather than absolute value of gradient as it varies with the loading conditions.
- The absence of pulmonary vein obstruction must be confirmed after device closure of ASD. If the pulsed wave Doppler gradient is less than 2 mmHg, pulmonary venous obstruction is ruled out.

**Ventricular Septal Defect**

VSD is the most common form of congenital heart disease. VSDs are classified as perimembranous (the most common), inlet, trabecular, and infundibular (supracristal, subpulmonic, and doubly committed subarterial). Biventricular volume load occurs in restrictive VSD. RV volume and pressure overload occur in unrestrictive VSD. Aortic regurgitation (AR) may be present in subarterial VSD. Patients with multiple VSDs and severe congestive heart failure (CHF) may have a PA band placed to limit PBF before definitive repair. Surgery consists of over-sew or patch closure of VSD using cardiopulmonary bypass. After bypass, a residual VSD more than 0.5 cm indicates a need to return to bypass for additional patching.[21]

**Perimembranous ventricular septal defect**

This VSD is seen in ME RV inflow-outflow [Figure 3a], ME AV SAX V, and modified ME 5C V (obtained by withdrawing and flexing the probe from the ME 4C V). The defect is adjacent to septal leaflet of TV. The leaflet tissue may completely or partially close the defect. In the ME AV LAX V, the defect is seen adjacent to right coronary cusp of aortic valve which may be prolapsed into the defect due to venturi effect from VSD flow.

**Inlet ventricular septal defect**

Inlet VSD is located inferiorly in the ventricular septum close to AVVs [Figure 3b]. It is seen in ME 4C and in TG basal SAX views. It is often associated with atroventricular canal defect (AVCD). The AVV must be examined for override. More than 50% of the TV may open into LV. It may also be associated with LVOT obstruction. The LVOT must be examined for membranes, chords, and tunnel like obstruction (use ME LAX V, TG LAX V, and deep TG LAX V). CFD and CWD is used to look for stenosis/regurgitation of AVVs.

**Outlet ventricular septal defect**

It is observed adjacent to PV in ME RV inflow-outflow view. In the ME AV LAX V with outward rotation [Figure 3c], it is seen adjacent to the right coronary cusp. This view is used to look for AR (CFD) and prolapse of right coronary cusp.

**Muscular ventricular septal defect**

Muscular VSDs are looked for using 2D and CFD in ME 4C [Figure 3d], ME LAX V, ME AV LAX V, TG mid SAX V, and deep TG LAX V. The entire interventricular septum must be searched for multiple defects. Muscular VSD may be anterior, mid-ventricular, posterior, and apical, depending on their location in the muscular septum. It has a rim totally made up of muscle.

**In all types of ventricular septal defect**

- Search for multiple VSDs should be done and size of each VSD assessed (small 1–2 mm: moderate 3–5 mm; large >5 mm).
- It is necessary to look for left atrial enlargement, left ventricular enlargement, right atrial enlargement.
• Measure annulus of TV if dilated, and assess biventricular function
• Direction of blood flow must be assessed using CFD. Flow is left to right in uncomplicated VSD. However, in early part of diastole, a short reversal of flow is usual
• CWD is used to measure RV pressure which is $= \text{pulmonary artery pressure (PAP)} = \text{systolic blood pressure (BP)} - 4 \times \text{velocity across VSD}$
• The presence of any AR is assessed using CFD, CWD/PWD.

**Transesophageal echocardiography assessment after surgical repair**

• Closure of VSD (and any co-existing ASD) must be confirmed and diligent search must be done to look for residual or additional VSDs. Residual defects <3 mm are detected by TEE but are hemodynamically insignificant and usually do not require immediate reoperation.
• RV pressure or PA pressure is measured using VSD flow or tricuspid regurgitation (TR) jet velocity if present.
• Search is done for AR, aortic cusp perforation or prolapse, TR.
• Biventricular function needs to be assessed (can be affected by injury to coronary artery, ventriculotomy).

**In atrial septal defect and ventricular septal defect**

• Pulmonary hypertension, RV hypertrophy, Right to Left shunt, and systolic flattening of interventricular septum points toward the possibility of Eisenmenger syndrome.
• TR is searched for (using CFD) and if it is present, is present, continuous wave is used to measure right ventricular pressure = PAP = 4 multiplied by square of TR velocity + right atrial pressure.
• Qp: Qs (ratio of which is measured before and after surgery) using velocity time integral at LVOT and right ventricular outflow tract (RVOT) as well diameter of LVOT and RVOT.

**Atrioventricular Canal Defect**

Partial AV canal defect consists of four components: (1) primum ASD, (2) inlet VSD with restrictive or no ventricular shunting, (3) cleft anterior mitral leaflet, (4) widened anteroseptal tricuspid commissure. Complete AV canal defect consists of a large septal defect with interatrial and interventricular components and a common atrio-ventricular (AV) valve that connects both atria to both ventricles. In balanced AV canal defect, LV and RV contribute fair parts to the total work of the heart. In unbalanced AV canal defect, there is single ventricle physiology. PA banding may be chosen over complete repair on rare occasions when the infant is small. The goal is to reduce pressure distal to the band to one-half that of systemic BP, when inspired oxygen concentration is 21%. After banding, saturations of 78% to 90% should be acceptable on an inspired oxygen concentration of 50%. Palliation allows the child to grow and pulmonary edema to resolve before complete repair is attempted. Complete repair includes repair of valve clefts, closure of ASD and VSD, and creation of two AV valves.

1. Interatrial septum is examined for Primum ASD (ME 4C) and interventricular septum for inlet VSD (ME 4C, TG Mid SAX). Partial AVCD [Figure 4a] consists of Primum ASD and cleft MV. Complete AVCD [Figure 4b] consists of primum ASD, inlet VSD, and common AVV. Size of defect and direction of flow is assessed using CFD and PWD.

2. AVVs are examined for MV cleft, and leaflet prolapse, and straddling using ME 4C V, ME LAX V, TG basal SAX V, TG LAX V. ME 4C and TG mid-SAX views are used to assess whether the defect is balanced. If the LV is small (unbalanced AVCD), single ventricle repair may be required. Size and function of ventricles are assessed. When insertions of straddling cords are into the opposite ventricle, it may not be possible to obtain 2 competent AVVs (necessitating single ventricle repair) Any stenosis or regurgitation of AVVs should be graded. Any LVOTO should be identified using CFD (ME LAX) and the gradient measured (TG LAX V, deep TG LAX V).

3. It is necessary to look for LSVC. If LSVC draining into coronary sinus (CS) is present, the atrial patch should not be placed in such a way that the coronary sinus drains into LA.

4. RV pressure or PA pressure is measured using TR or VSD jet.

5. Postoperative TEE:
   1. Closure of ASD and VSD is confirmed and AVVs are examined for any residual stenosis/regurgitation/prolapse/flail.
   2. Biventricular function is assessed, and PA pressure is measured if there is TR.

**Persistent Ductus Arteriosus**

Patients are usually asymptomatic if the ductus is small. A large shunt may predispose to lower respiratory tract...
infection, atelectasis, and CHF. Left-to-right shunting and volume overload of the pulmonary circulation occur. Pulmonary edema, hypotension, and acidosis may occur. If left untreated pulmonary overcirculation may cause irreversible pulmonary vascular occlusive disease (PVOD), which leads to right-to-left shunting (Eisenmenger’s syndrome).

Definitive treatment: Since persistent ductus arteriosus (PDA) is a risk for endocarditis, paradoxical emboli, and CHF, the existence of PDA is an indication for closure. However, the presence of severe PVOD is a contraindication for PDA interruption. PDA may be ligated or divided via left thoracotomy. Video-assisted thoracoscopic surgery is another option. Transcatheter closure of PDA is very popular.

ME ascending aorta, SAX view [Figure 5a] is used to image PDA. Although it is difficult to visualize PDA by 2D TEE, ductal flow can be seen in this view (using CFD) as continuous high velocity turbulent aliased flow in the main pulmonary artery (MPA). CWD at this site shows flow throughout the cardiac cycle, and it may be possible to delineate the PDA and its attachment to PA using CFD. The same views obtained after surgery should show the absence of turbulence in the MPA.

In postsurgical period, any persistent shunt is looked for, and biventricular function is assessed.

Aorto-pulmonary Window

In this anomaly, both aortic as well as PV apparatus are well defined and there is a communication between the ascending aorta and the pulmonary trunk and/or the right PA. Patients with pulmonary over-circulation should be managed to maintain pulmonary vascular tone. Surgical snaring of the PA can assist in increasing diastolic BP and coronary perfusion. Perioperative pulmonary hypertension frequently requires hyperventilation, 100% oxygen, systemic alkalinization, deep sedation, and paralysis.

The defect can be imaged in mid esophageal ascending aorta short axis view [Figure 5b].

Coarctation of Aorta

Coarctation of the aorta most commonly is characterized by a narrowing of the aortic lumen opposite the opening to the ductus arteriosus and just distally to the opening of the left subclavian artery. As many as 85% of patients with coarctation will have a bicuspid aortic valve. The diagnostic feature of coarctation of the aorta is systolic and mean BP differences between the upper extremities and lower extremities. The manifestations can be mild or severe depending on the severity of obstruction. In severe obstruction, the PDA can be the only source of systemic supply and LV dysfunction is common. If the infant is <1 month of age, intravenous prostaglandin is utilized to open the closed ductus arteriosus. The aortic obstruction may precipitate CHF and cardiogenic shock in the neonate. Such patients require inotropic agents and diuretics. Metabolic acidosis should be corrected to improve left ventricular function. Unrelieved coarctation results in upper extremity hypertension. The most common sequelae of coarctation of the aorta are systolic hypertension, left ventricular hypertrophy (LVH), development of collateral vessels, recurrent or residual coarctation, aneurysm formation, aortic dissection, intracranial hemorrhage secondary to intracranial aneurysm formation, and sudden death. Resection of narrowing is the surgical treatment, which is usually done through thoracotomy. Left thoracotomy is done in case of left aortic arch. The surgical approaches to coarctation repair include resection of the coarcted area with direct end-to-end anastomosis, subclavian patch aortoplasty, or aortoplasty with homograft/synthetic graft material. After coarctation repair, hypertension is treated with esmolol or nitroprusside. Catheter-based therapy is more commonly used in older patients.

The narrow coarctation segment (including posterior shelf inside aortic lumen) and poststenotic dilation of descending thoracic aorta distal to the coarctation site can be seen in upper esophageal aortic arch SAX and descending thoracic aortic LAX view [Figure 6a]. CFD shows aliased flow. CWD measures gradient of >2.5 m/s. In descending aortic SAX view, collateral vessels which brings blood flow to descending aorta may be visualized (using CFD). PWD interrogation of these collateral vessels show flow signals in systole and diastole.

Using ME 4-CH, ME 2-CH, AV LAX and TG SAX, mitral and aortic valvular morphology and function are assessed, any subvalvular and supravalvular aortic obstruction is searched for, and LV mass and function are assessed.

Residual gradient, re-coarctation, and aortic aneurysm formation is searched for during postsurgical TEE examination.

Aortic stenosis

Aortic stenosis may be valvular, subvalvular, or supravalvular. Aortic valve stenosis causes progressive
LVH with associated sequelae of decreased ventricular function and myocardial ischemia. These factors place the patient at increased risk for sudden death. Congenitally bicuspid aortic valve orifice produces varying degrees of obstruction, which results in stenosis with a systolic ejection murmur. Supravalvular aortic stenosis may be complicated by involvement of the coronary artery ostia in the stenotic segment. Coronary ischemia may occur, especially with low systemic vascular resistance when the ostia are distal to the stenotic segment.

Surgical treatment of critical aortic stenosis consists of complete repair with aortic valvotomy. The Ross procedure is an innovative technique that utilizes the patient’s own PV for aortic valve replacement. A cryopreserved, valved pulmonary homograft is inserted in the position of the original pulmonary root. Transcatheter dilation of aortic stenosis and aortic valve replacement are other treatment options.

Precardiopulmonary bypass transesophageal echocardiography

ME AV SAX V is used to see morphology of AV. Deep TG LAX V is used to assess aortic stenosis (measure peak gradient across obstruction), any associated AR, and to measure the aortic root size. Location and severity of obstruction (subvalvar, valvar, and supravalvar) must be evaluated. To look for LVH and to assess LV function, ME 4C V is used. Supravalvular aortic stenosis is seen in ME AV LAX V [Figure 6b].

Postcardiopulmonary bypass transesophageal echocardiography

After surgery, TEE is used to look for any residual outflow obstruction or aortic insufficiency, any new VSD, or mitral regurgitation. Ventricular function (segmental and global) is also assessed. Function of right ventricular homograft is assessed after Ross procedure.

Pulmonic Stenosis

Pulmonary stenosis may be subvalvular, valvular, or supravalvular. Stenosis of PA may occur near the bifurcation and may involve more peripheral branches. PS may be an isolated lesion or may be seen in association with complex congenital lesions, rubella syndrome, or Williams’ syndrome.

Intraoperative TEE should be used for patients with PS and right ventricular or biventricular dysfunction. In these patients, loading conditions and ventricular function monitoring are required.

For valuation of RVOT and PV, ME RV inflow-outflow view is used. ME ascending aortic SAX view is used to evaluate the MPA and right PA. For measurement of gradients across the PV, TG RV basal view, TG RV inflow-outflow view, and upper esophageal aortic arch SAX views [Figure 6c] can be used. Anatomy of PV, the location and severity of obstruction (subvalvar, valvar, and supravalvar), and size of PAs must be determined. It is also necessary to evaluate for intracardiac shunts, to evaluate for ventricular hypertrophy, and to assess ventricular function.

Postsurgical evaluation includes assessment of RV size and function as well as any residual pulmonary outflow tract obstruction or pulmonary regurgitation.

Cor-triatriatum

Cor-triatriatum is diagnosed if the LAA is inferior to a stenotic ring in the LA. If the opening of the ring is severely restrictive, the patient develops symptoms similar to those of severe mitral stenosis.

ME 4C, mid-esophageal commissural [Figure 6d] and ME 2C views enable visualization of a fibromuscular membrane which courses between the atrial septum and the junction of the LUPV and the LAA. The atrial accessory chamber receives the LUPV. 2D TEE shows the opening in the membrane. Using PWD, the degree of narrowing can be assessed.

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