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REVIEW ARTICLE

Transesophageal Echocardiography for Tetralogy of Fallot Repair: What a Perioperative Physician need to know?

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ABSTRACT

Transesophageal echocardiography (TEE) is now an integral part of intraoperative management of TOF patients undergoing intracardiac repair. With the availability of micro TEE probes, intraoperative TEE care can now be provided to even the smallest of patients. It provides live images of the anatomical and pathophysiological state of the heart and allows perioperative physicians to modify surgical and medical treatment perioperatively.

During pre-bypass period, TEE confirms preoperative diagnosis and can provide additional information which might be missed on transthoracic echocardiography (TTE). It also helps in modifying intraoperative surgical plan if new findings are detected intraoperatively. In addition, real time information on volume status and inotropy helps in management of hemodynamics and preventing hypercyanotic spells in pre-bypass period.

Adequacy of surgical repair can be assessed in immediate post-bypass period and any residual defect can be corrected before patient leaves the operating room. Post repair information on anatomical and pathophysiologic status helps guiding management in intensive care unit.

Keywords: Perioperative, Transesophageal echocardiography, Tetralogy of fallot, Intracardiac repair.

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INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease constituting around 10% of all congenital heart disease.[1,2] The diagnosis of TOF includes a constellation of anatomical abnormalities including a nonrestrictive ventricular septum defect (VSD), aortic override, right ventricle outflow tract obstruction (RVOTO) and right ventricle hypertrophy (RVH). Total surgical correction under cardiopulmonary bypass support is now the treatment of choice.

Intraoperative transesophageal echocardiography (TEE) is now recommended for all the patients undergoing intracardiac repair (ICR) for TOF.[3] With the availability of micro TEE probes, intraoperative TEE care can now be provided to even the smallest of patients. Many studies have shown the beneficial effects of intraoperative TEE care in pediatric cardiac surgeries in general and in TOF as well.[4-7] TEE provides live images of the anatomical and pathophysiological state of the heart and allows perioperative physicians to modify surgical and/or medical treatment perioperatively. During the pre-bypass period, TEE overcomes some of limitations of TTE, such as like suboptimal windows, or poor patient cooperation, and confirms the preoperative diagnosis. TEE may provide additional information which might be missed on TTE. In addition, real time information on volume status and cardiac function helps in management of hemodynamics and preventing hypercyanotic spells during the pre-bypass period. A pre-bypass examination also serves as a baseline for comparison with post-repair assessment.

A detailed description of TEE assessment of TOF patients is discussed below. For uniformity of nomenclature used while describing TEE views, latest guidelines for TEE evaluation as described by American Society of Echocardiography are being used.[8]

Ventricle Septal Defect

Ventricle septal defect in TOF patients is perimembranous in most and juxtaarterial or doubly committed in some of the TOF patients. There may be additional muscular VSDs in a small subset of patients. The VSD is usually large and unrestrictive. The direction of blood flow across the VSD depends on the degree of RVOT obstruction. The pre-bypass TEE helps in detecting the size, number and location of VSDs.

A perimembranous VSD is typically seen at probe angle of 0°, in midesophageal 5 chamber (ME5Ch) view

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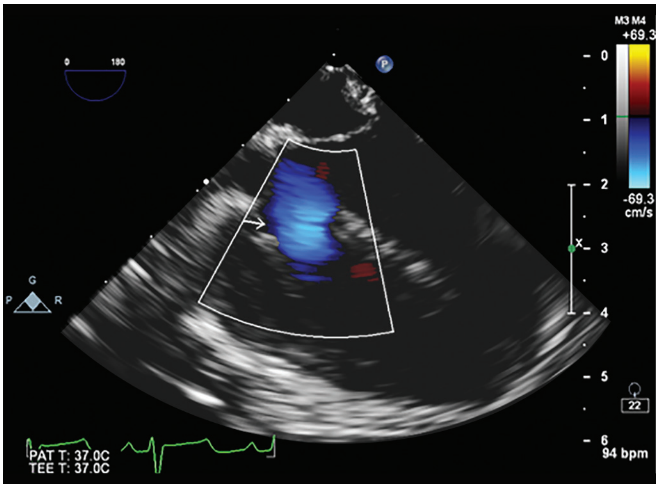


Fig. 1: ME5Ch view showing large perimembranous VSD (arrow) with right to left shunting in a patient with TOF (Videos 1A and B).

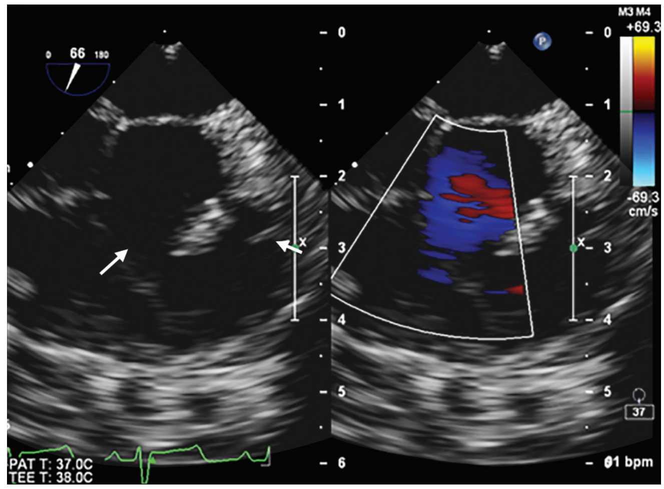


Fig. 2: MERV inflow outflow view showing typical perimembranous VSD (arrow) at 7 o'clock position in a patient with TOF (Video 2)

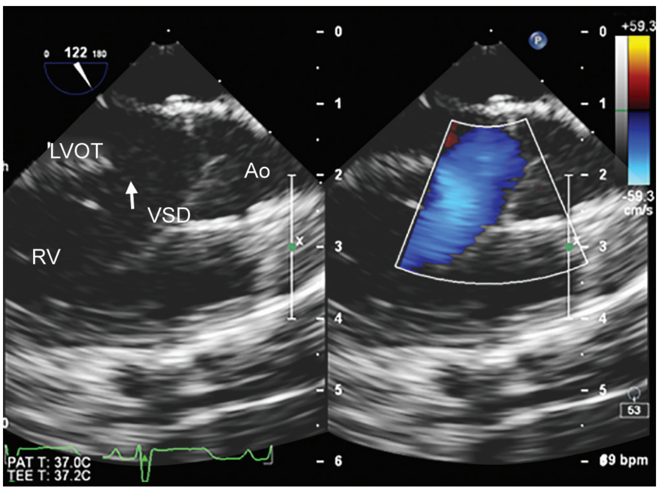


Fig. 3: ME aortic valve LAX view showing VSD (arrow) and aortic over ride in a patient with TOF. Note that aortic valve is overriding the interventricular septum and appears to be arising from both left and right ventricles (Videos 3A and B)

(Fig. 1) and disappears as the probe is moved inferiorly to the midesophageal 4 chamber (ME4Ch) view (also at about 0°). When the imaging angle is adjusted to 40 to 70°, the midesophageal RV inflow outflow (MERV inflow outflow) view can be seen. This provides imaging equivalent to the TTE parasternal aortic short axis view, however the image display is inverted upside down. Thus, a typical perimembranous VSD will be seen at 7 o' clock position (Fig. 2) while a subpulmonic VSD will be located at 5 O' clock position in MEAoSAX view. A perimembranous VSD can also be seen in midesophageal aortic valve long axis (MEAV LAX) view (angle 90-120°) and deep transgastric 5 chamber (DeepTG5Ch) view (angle 0-20°).

Additional muscular VSDs can be seen in ME4Ch view, ME5Ch view, MidEsophageal long axis (ME LAX) view and trans gastric mid papillary short axis (TGSAX) views. However, when a large and unrestrictive VSD is present as in TOF, flow through small muscular VSDs may be undetectable by color flow imaging until after the large VSD has been closed. This is simply a limitation of imaging that is inherent to the perioprative changes occurring during TOF repair.

Aortic Override

Both VSD and aortic override develop as a result of malalignment between the trabecular and outlet ventricular septae. Since the outlet, or conal septum is anteriorly malaligned, the aortic root arises partially from the right ventricle. If the aortic override exceeds 50%, and is associated with discontinuity of the aortic and mitral fibrous rings due to the presence of a subaortic conus, the cardiac condition is double outlet right ventricle (DORV). ME AVLAX (Fig. 3), ME5Ch view and TGLAX views are most commonly used to assess the degree of aortic override. Aortic root dilatation and aortic regurgitation can be associated in TOF patients and is assessed by ME AVSAX, MEAVLAX and TGLAX views.

Right Ventricular Outflow Tract, Pulmonary Valve and Pulmonary Arteries

The right ventricular outflow tract obstruction in TOF patients is typically infundibular but may involve the pulmonary valve, main and branch pulmonary arteries. The pulmonary valve annulus may be small and in extreme cases atretic in a separate subset of patients termed as TOF with pulmonary atresia. The pulmonary blood flow in TOF with pulmonary atresia patients depends on the presence of a patent ductus arteriosus (PDA) or major aorto-pulmonary collateral arteries (MAPCAs). Of special interest is another subset of patients of TOF with absent pulmonary valve occurring in approximately 3% of patients with TOF. In these patients, there is often annulus hypoplasia associated

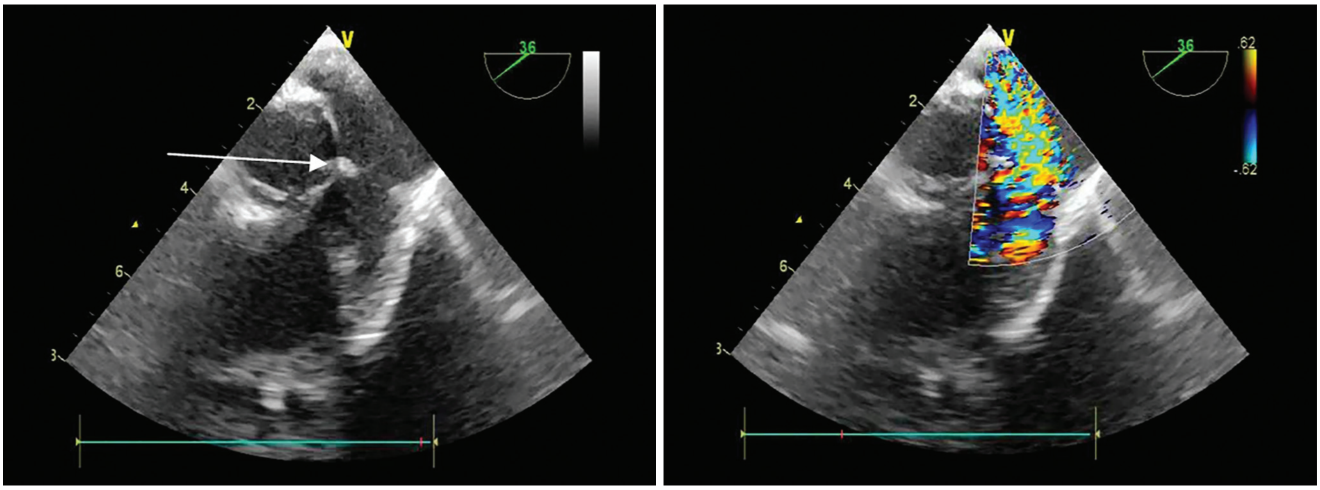


Fig. 4: Mid esophageal RV inflow outflow view in a patient of TOF with absent pulmonary valve. Note absent (rudimentary) pulmonary valve and aneurismal pulmonary artery (Videos 4A and B)

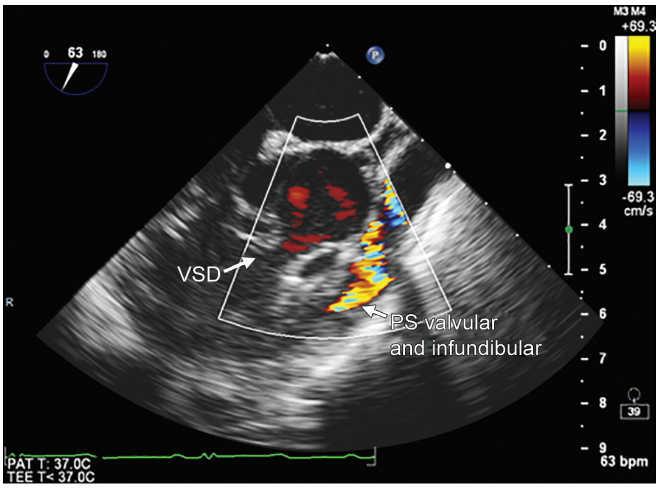


Fig. 5: MERV inflow outflow view showing VSD and PS involving infundibulum and pulmonary valve (Video 5)

with a rudimentary and malformed pulmonary valve, which results combined pulmonary stenosis (PS) and regurgitation leading to massive aneurysmal dilatation of pulmonary arteries (Fig. 4). In the most severe subset of these patients, the large and pulsatile pulmonary arteries compress the airways, and can lead to significant airway compromise postnatally. The RV may also be enlarged and demonstrates paradoxical septal motion.

Transesophageal echocardiography provides anatomical detail that may help refine the surgical plan, specifically regarding the type of repair-whether valve sparing surgery, use of transannular patch or RV to pulmonary artery conduit will be technique employed. MERV inflow-outflow view (Fig. 5), transgastric RV basal (TG RV basal) view (Figs 6A and B) and TG RVinflow outflow views (Fig. 7) can clearly show RVOT, pulmonary valve and main pulmonary artery. ME ascending aortic short axis view (Figs 8A and B) is commonly used to interrogate main and branch pulmonary arteries, as well as ME5Ch view pulled further out and rotated leftward to view the MPA (or RV to PA conduit) at 0°. This view is particularly useful for Doppler interrogation of flow beyond the pulmonary valve. Also, the presence of PDA or patent BT shunt can be seen in this view or the ME ascending aortic short axis view by using color and spectral Doppler imaging. However, TEE assessment of left pulmonary artery (LPA) can be challenging because of interposition of left bronchus between esophagus posteriorly and LPA anteriorly.

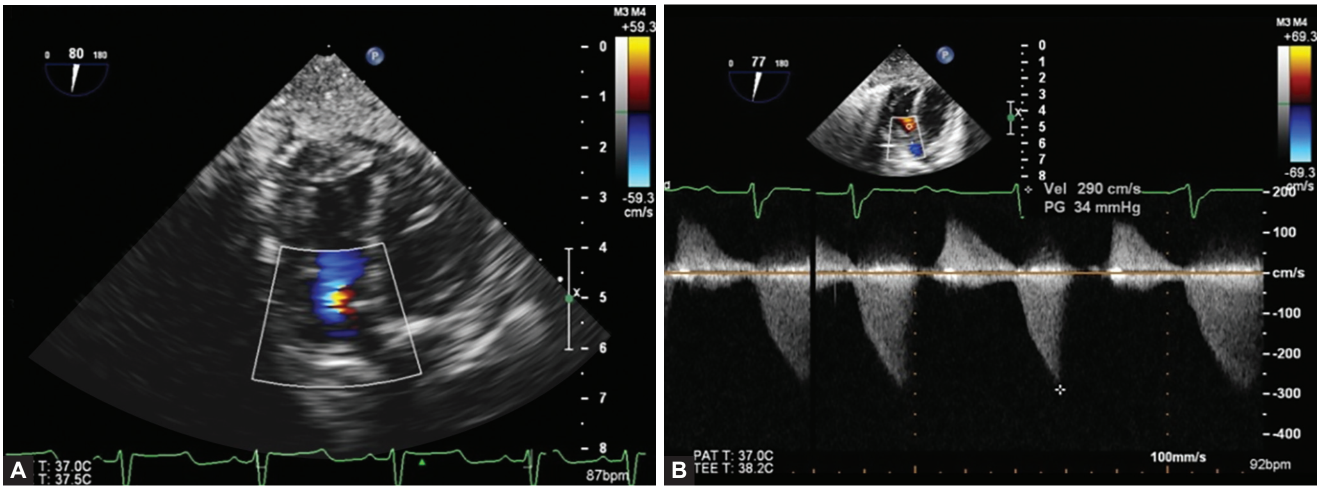
Application of continuous wave Doppler (CWD) across the RVOT shows characteristic 'dagger shaped' signal suggestive of dynamic obstruction due to infundibular hypertrophy (Fig. 6B). However, there may be a double envelope formation on CWD spectrum indicating both infundibular and valvular PS. The use of color and pulsed wave Doppler (PWD) helps in localizing the site of obstruction. A high ME5Chview, ME Asc Ao SAX, TG RV basal and TGRV inflow outflow views are best for Doppler alignment and estimation of peak pressure gradients.

Right Ventricle Hypertrophy

Right ventricle hypertrophy in TOF patients occurs as a compensatory response to increased RV after load due to outflow obstruction. RV hypertrophy can be appreciated in ME4Ch, MERV inflow-outflow views and TG mid papillary SAX views (Fig. 9).

Coronary Artery Anomalies

Coronary artery anomalies can be seen in approximately 10% of cases of TOF.[9,10] Most common is aberrant left anterior descending (LAD) artery originating from right coronary artery (RCA) that crosses over the RVOT anteriorly and can be the cause for suboptimal RVOT



Figs 6A and B: (A) Transgastric RV basal view showing narrowed RVOT (arrow) in a patient with TOF, (B) Application of continuous wave Doppler across RVOT shows characteristic dagger shaped signal typical of infundibular stenosis (Video 6)

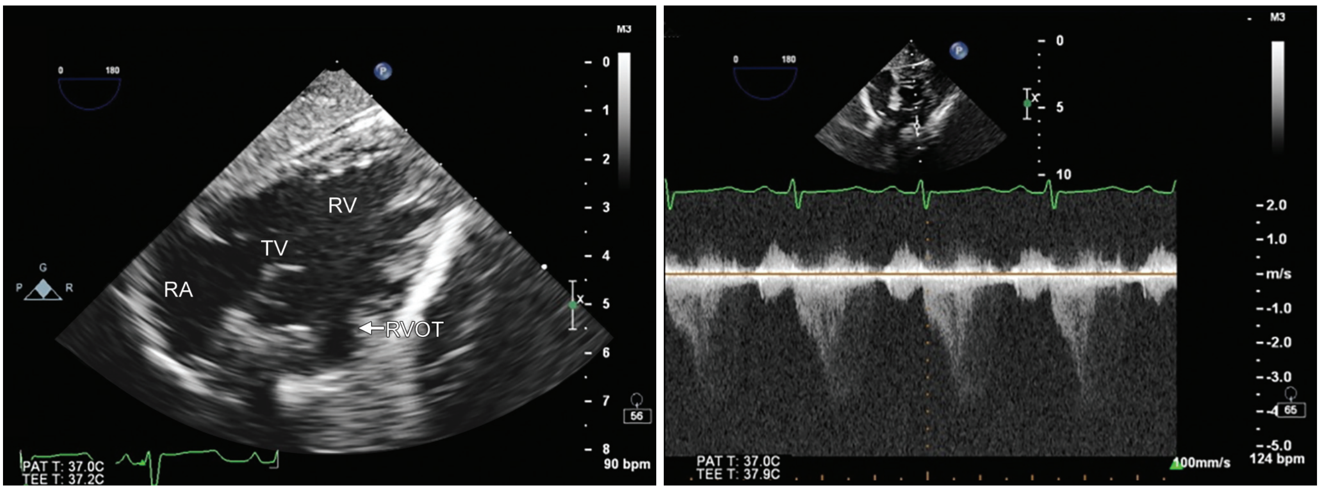
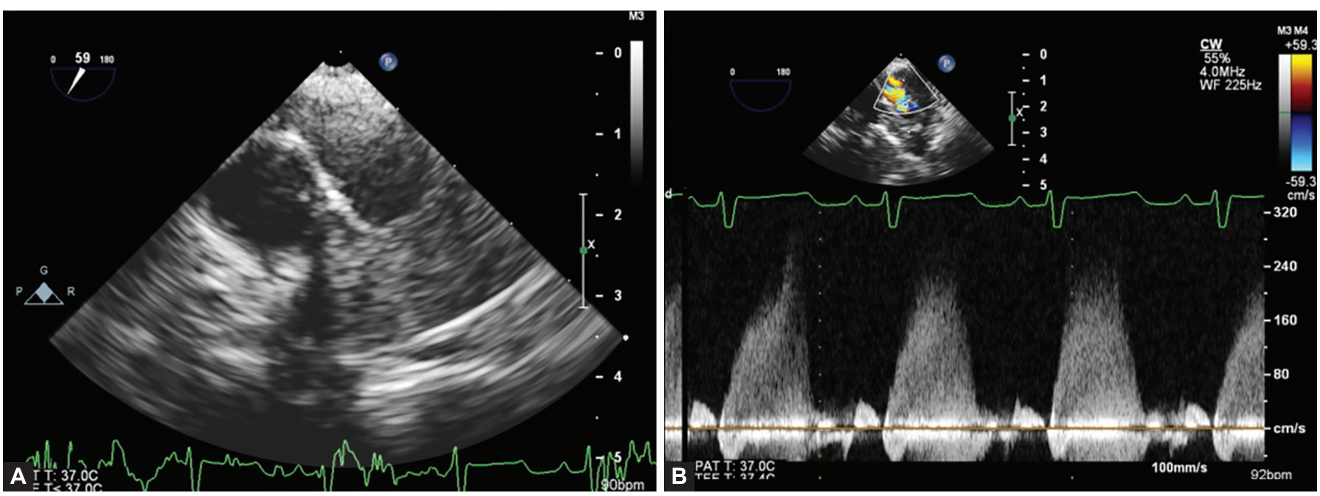


Fig. 7: Transgastric RV inflow outflow view showing narrowed RVOT in a patient with TOF (Video 7)



Figs 8A and B: (A) ME ascending aortic SAX view showing main and branch pulmonary arteries, (B) application of continuous wave Doppler for estimation of pressure gradient across main pulmonary artery (Video 8)

resection and residual gradient. Although, TEE is not the modality of choice to detect an abnormal coronary artery crossing RVOT, an aberrant coronary artery can be picked up by TEE in some cases. MEAVSAX view can show an aberrant LAD crossing RVOT (Fig. 10). Since dual LADs may be present in TOF, the visualization of a LAD in the usual position does not preclude presence of an LAD off the right coronary cusp in these patients.

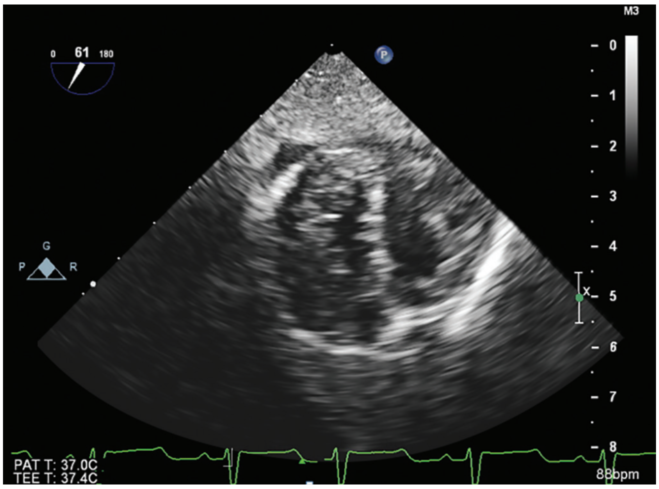


Fig. 9: Transgastric mid papillary SAX view showing hypertrophied RV (Video 9)

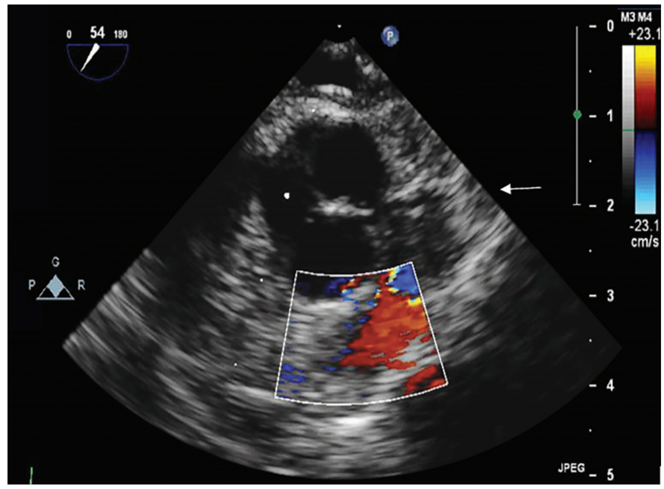
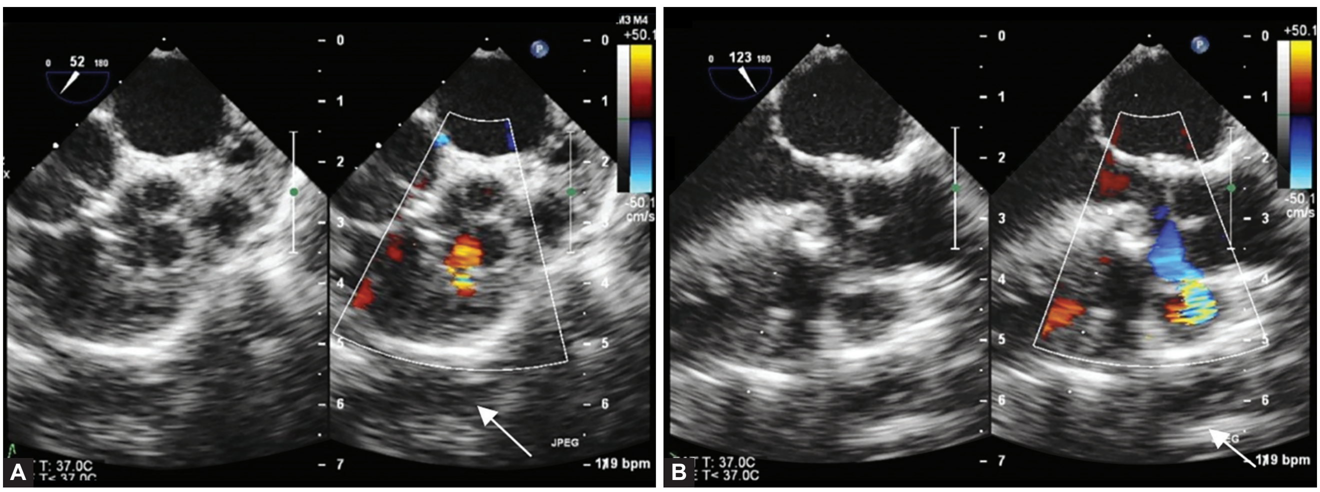


Fig. 10: ME aortic valve SAX view in a patient with aberrant LAD arising from RCA and crossing RVOT. Note color flow in the region of RVOT which was originating from RCA in the patient



Figs 11A and B: Residual VSD post TOF repair: (A) MERV inflow outflow view showing leak across the VSD patch (arrow), (B) ME aortic valve LAX view showing residual VSD (arrow) in same patient after TOF repair (Videos 11A and B)

TEE Post Intracardiac Repair

Weaning from CPB

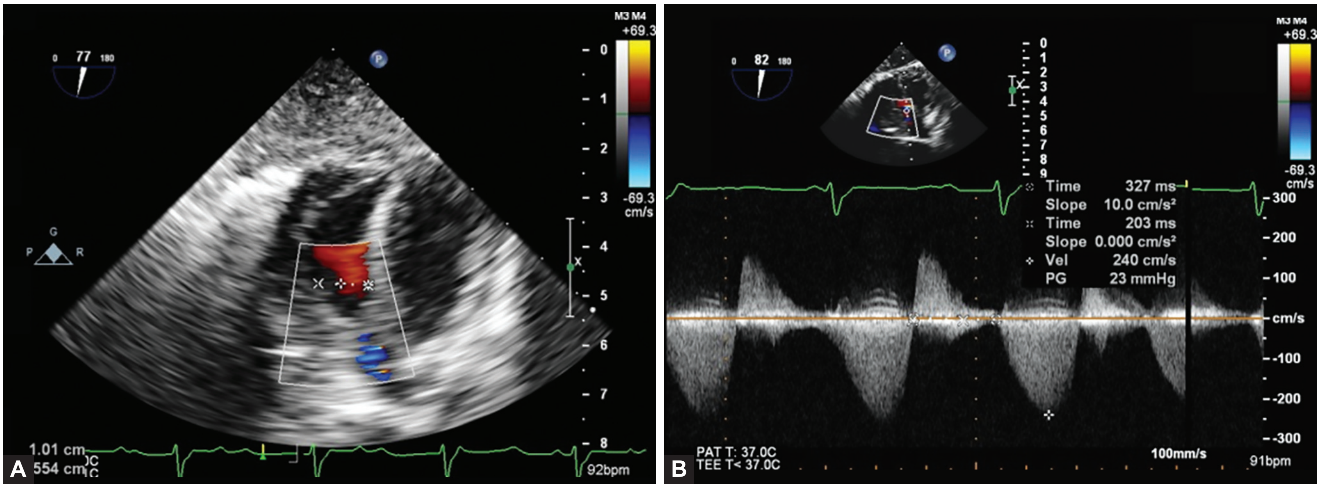
Transesophageal echocardiography helps in adequate deairing of heart before weaning off CPB. Also, it provides vital information on the volume status and biventricular contractility. TG mid SAX view is used for assessing ventricular filling and contractility.

Residual VSD

Residual VSD post TOF correction can be due to patch leak or unaddressed muscular VSD. TEE views used for assessing VSD in pre-bypass period can be used for detecting residual VSD (Figs 11A and B). TEE can guide the surgeon to the site and magnitude of the residual VSD. A VSD dimension <3 mm is usually not hemodynamically significant while a defect >5 mm should be repaired.Similarly, hemodynamic calculations can be done and a ratio of pulmonary to systemic blood flow (Qp:Qs) more than 1.5 should be repaired. An alternative method is to look for step up in oxygen saturation from right atrium to pulmonary artery; a step up > 7% is usually clinically significant.

Residual RVOT Obstruction

Residual RVOT obstruction can be at the level of the infundibulum, pulmonary valve, main or branch pulmonary arteries. By the time imaging is being performed, the surgeon has often determined whether they feel a thrill in the RVOT, and often questioning the surgeons about their findings can be helful in the imaging process. RVOT obstruction can be dynamic (hypertrophied muscle bundles) or fixed (i.e. small pulmonary annulus, small sized main and branch pulmonary arteries). TEE images as described for pre- bypass assessment of RVOT obstruction can be used for post-bypass assessment as well. TEE not only helps in grading the degree of residual obstruction but also guides surgeons in further resection by exactly showing the site of obstruction. Application



Figs 12A and B: Pulmonary regurgitation post TOF repair: (A) Transgastric RV basal view showing PR jet (arrow), (B) Calculation of PR index using continuous wave Doppler in same view. PR index is ratio of duration of PR signal by total diastole duration. PR index in this case is 203 msec/327 msec = 0.62 , hence severe PR (Videos 12A and B)

of CW Doppler across the site of obstruction is used for estimation of pressure gradient, while color Doppler and PW Doppler are used for locating the site of obstruction. CW Doppler estimation can be accurately used if angle of alignment between blood flow and Doppler beam is less than 20°. A peak velocity > 3.0 m/s across the suspected site of obstruction is considered significant and requires further resection. In case of dynamic obstruction, estimation of pressure gradients should be done after correction of hypovolemia and minimising inotropic support as hypovolemia and increased use of inotropes can lead to overestimation of pressure gradients. For these reasons, it is prudent to request direct pressure measurement of the RV when a substantial obstruction is suspected by imaging. An RV systolic pressure that exceeds 2/3 of the systemic blood pressure is often considered unacceptable, and cause for surgical revision of the RVOT.

Pulmonary Regurgitation

The degree of Pulmonary Regurgitation (PR) post TOF correction depends on type of RVOT reconstruction. Pulmonary regurgitation can be severe in patients with transannular patch reconstruction of RVOT. Significant PR eventually leads to RV volume overload and RV dysfunction. The severity of PR is graded by the size of regurgitant jet relative to RVOT diameter (mild <1/3rd, moderate 1/3rd to 2/3rd and severe >2/3rd).[11-13] The presence of diastolic flow reversal in branch pulmonary arteries is suggestive of severe PR. The PR index is the ratio of PR signal duration in spectral Doppler trace to total duration of diastole (Fig. 12). As PR severity increases, early equilibration of pressure across RV and PA occurs and thus PR index decreases. A value of PR index <0.77 is correlated with severe PR as derived by cardiac magnetic resonance.[14]

Right Ventricle Function Assessment

Right ventricle dysfunction is an inevitable sequelae of TOF repair, however, its severity varies among the patients. Residual RVOT obstruction, pulmonary regurgitation, ventriculotomy (incision on RV/RVOT) and inadequate myocardial protection of the hypertrophied RV can all contribute to right ventricular dysfunction. A hypertrophied RV is also prone to develop diastolic dysfunction post TOF repair. Echocardiographic assessment of RV function parameters have been less well studied and even less studied while using TEE. Because of typical crescent shape of RV, global RV function assessment by 2D echo in one single image/plane cannot be done accurately. Hence, multiple images assessing RV in different planes should be used.

Right-ventricle fractional area change (FAC) is defined as [(end-diastolic area – end-systolic area)/end-diastolic area] × 100. ME4Ch view can be used to estimate RVFAC and a value < 35% suggests RV systolic dysfunction.[15,16] Although, RVFAC can be easily estimated by tracing endocardial borders of RV in systole and diastole, studies have shown weak correlation with cardiac magnetic resonance derived RV ejection fraction.[17]

Longitudinal RV function as assessed by tricuspid annulus peak systolic excursion (TAPSE) has been shown to correlate well with global RV function in patients with normal RV function. In a recent study, TAPSE as derived by TTE was not associated with cardiac magnetic resonance derived RV ejection fraction and exercise tolerance in patients post TOF correction.[18] Transgastric RV inflow outflow view is used for TAPSE estimation by TEE. TAPSE estimation by TEE may be further limited by inadequate angle between motion of tricuspid annulus and M-mode in TEE views.

Tissue Doppler imaging (TDI) derived lateral tricuspid annulus peak systolic velocity (S′) can also be used for RV systolic assessment. RV myocardial performance index (MPI or Tei index) can also be calculated.

Right ventricle diastolic performance assessment post TOF repair involves a combination of parameters like trans-tricuspid inflow velocities, hepatic venous flow reversal, IVC diameter variation with respiration and presence of antegrade flow in MPA during late diastole.[19] The forward diastolic flow in pulmonary artery occurs due to premature opening of pulmonary valve during atrial systole, when right ventricle end diastolic pressure exceeds pulmonary arterial end diastolic pressure, thus suggesting severe RV diastolic dysfunction where RV acts as a passive conduit between right atrium and pulmonary artery.

Position of interventricle septum also provides information regarding RV volume or pressure oiverload state. Flattening of interventricle septum during systole suggests pressure overloaded RV while during diastole suggests volume overloaded RV.

SUMMARY

Transesophageal echocardiography is now an integral part of intraoperative management of TOF patients undergoing intracardiac repair. TEE by providing real time information on anatomy, pathophysiology and hemodynamics in these patients helps in clinical management. TEE also helps in modifying intraoperative surgical plan if new findings are detected intraoperatively. Any residual lesion detected post repair can be taken care of inside the operating room and thus reduces the incidence of resurgery. Post repair information on anatomical and pathophysiologic status helps guiding management in intensive care unit.

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