TEE Evaluation of Tetralogy of Fallot
and Double Outlet Right Ventricle

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Echocardiography is the diagnostic modality of choice for the initial evaluation and serial assessment in most types of pediatric heart disease. The morphologic diagnosis of tetralogy of Fallot (TOF) and double outlet right ventricle (DORV) is established in the majority of cases using this imaging approach alone. The primary goal of the initial echocardiographic evaluation is to outline the anatomic abnormalities in detail, evaluate associated pathology, and perform a physiologic assessment. Rarely, additional studies are necessary for diagnostic purposes or in the initial management of affected patients with typical anatomy. In those with tetralogy variants (pulmonary atresia with ventricular septal defects and aortopulmonary collaterals, absent pulmonary valve syndrome), additional imaging studies may be required pre and post operatively to fully delineate the morphologic abnormalities.

The segmental morphologic analysis is the preferred approach in the echocardiographic assessment of patients with congenital heart disease. In those with TOF typical findings include levocardia (cardiac apex and mass to the left) and normal arrangement of the abdominal viscera and atria (visceroatrial situs solitus). The atrioventricular and ventriculoarterial connections are usually concordant. Although it is well recognized that TOF represents a spectrum of disease and in fact specific types of DORV may be considered by some to be part of this spectrum, in TOF the anatomic anomalies are all the result of a basic malformation, namely an underdeveloped pulmonary infundibulum.

Double outlet right ventricle refers to the ventriculoarterial connection where both great arteries are nearly or completely aligned with respect to the right ventricle. This is thought to be the result of abnormal conal development leading to pathologic great artery relationships.
Various criteria have been proposed to characterize this pathology leading to significant controversy. These include the 50% rule, the absence of fibrous discontinuity between the aortic valve and adjacent atrioventricular valve, and the morphology of the conus septum. The anatomic and physiology diversity of DORV relates to the location of the ventricular septal defect and great artery relationships.

The primary applications of transesophageal echocardiography (TEE) in patients with TOF or DORV include those related to the intraoperative setting, either during initial surgical correction or when reoperation is needed to address sequelae or less likely, residual or recurrent pathology. The main goals of the examination are the confirmation of the preoperative diagnoses, baseline assessment, and identification of hemodynamically significant defects immediately following the surgical intervention that may necessitate revision. Additional benefits of TEE include: evaluation of associated defects, assessment of atrioventricular and semilunar valve competence, monitoring of ventricular preload and function, determining the adequacy of cardiac deairing, and guidance of the surgical revision is necessary. When TEE is contraindicated epicardial imaging may be considered.

The use of TEE in patients with TOF or DORV for diagnostic purposes alone is not as frequent. On occasion it is applied to define anatomic details not obtainable by other imaging modalities or to assess potential residual/recurrent lesions, sequelae or related acquired pathology. This may be the case in those with suboptimal transthoracic windows such as postoperative patients or adults with a history of these defects. Imaging via the transesophageal approach in these patients may also be of benefit in the cardiac catheterization laboratory during catheter-based interventions for monitoring of the procedure, management of potential complications, minimizing the use of fluoroscopy as well as the need for angiography.

The section that follows highlights the anatomic and hemodynamic alterations in the typical patient with TOF and how TEE can be applied to this assessment. As the focus of the session is TOF, only selected features of DORV are included.
Anatomic/Hemodynamic Abnormalities and Transesophageal Echocardiographic Evaluation

Pulmonary Outflow Tract Obstruction

The small size of the subpulmonary infundibulum, an essential feature, plays a critical role in this lesion. The characteristic finding is anterosuperior displacement of the conal or infundibular septum with respect to the crest of the muscular ventricular septum. This encroaches the right ventricular outflow tract and contributes to the stenosis. Pulmonary valve stenosis is a frequent but not universal feature, resulting from the obstructive infundibulum. Stenosis or hypoplasia of the main and branch pulmonary arteries may be present. Obstructive gradients can occur at any or all of these levels.

TEE:

- Anatomy and Suggested Two Dimensional Views:
  - RVOT: ME RV inflow-outflow, ME Asc Ao SAX, Deep TG (RVOT), additional RVOT/PA views

Hypoplasia of the pulmonary valve annulus is often present and may have implications regarding surgical repair (need for transannular patch). The size of the valve annulus, main and proximal branch pulmonary arteries can be assessed in these views, in addition to details regarding valve morphology (number of cusps, doming, thickening, dysplasia).

In the postoperative patient with TOF or related anatomic variant, native pulmonary or conduit regurgitation represents a frequent indication for reoperation. The views noted above are helpful in characterizing the pulmonary outflow tract in this setting (morphology, dilation, aneurysm) and assessing candidacy for percutaneous pulmonary valve implantation if an option.
Outflow tract obstruction occurs in the majority of patients with DORV (over 70%). This most commonly affects those with a subaortic VSD.

- Color Doppler:
  
  - determine nature, location, and severity of the obstruction
  
  - in the postop patient: assess for residual obstruction, degree of pulmonary/conduit regurgitation (patency of left ventricular outflow post DORV repair)

- Spectral Doppler (obtained in views that allow for optimal alignment of the Doppler cursor to the direction of flow):
  
  - evaluate severity of right ventricular outflow tract obstruction
    
    (estimation of gradient(s) derived from peak velocity)

_Ventricular Septal Defect (VSD)_

The interventricular communication is generally unrestrictive in TOF. The classic defect is an anterior malalignment-type outlet type (large conoventricular or subaortic defect). Other anatomic defects may be present in the ventricular septum (perimembranous, atrioventricular septal defect, inlet, muscular, doubly committed subarterial). The direction of shunting across the defect is related to the right ventricular outflow obstruction. The shunt direction is left to right if minimal outflow obstruction if present and predominantly right to left during hypercyanotic episodes. In most cases the shunt is bidirectional and the ventricular pressures are equal.

The location of the VSD is variable in DORV accounting for the physiologic diversity of this defect. A subaortic location occurs in most patients, however a subpulmonary location, doubly committed or noncommitted (remote) location of the VSD may alternatively be the case.
TEE:

- Anatomy and Suggested Two Dimensional Views:
  - VSD (location, size): ME 4 Ch, ME Ao SAX, ME LAX, deep TG (LVOT)

- Color Doppler:
  - assess direction of flow across VSD
  - evaluate additional shunts (ventricular, atrial and great artery levels)
  - evaluate for residual ventricular level shunting

In some cases, closure of the large conoventricular defect in TOF may unmask smaller ventricular septal defects. This should be evaluated in the post bypass examination.

- Spectral Doppler:
  - determine peak velocity of VSD jet (usually low due to unrestrictive nature of defect)

If residual defect, determine degree of restriction based on jet velocity.

- Contrast Echocardiography:
  - may assist in the evaluation of residual intracardiac shunts

Aortic Override:

In TOF the aortic root characteristically overrides the ventricular septum/defect. This is manifested as a biventricular connection of the leaflets of the aortic valve. In DORV the spatial relations of the arterial trunks ranges from normal to side-by-side to malposed. As in the case of the location of the VSD, influencing the physiology and surgical approach.
TEE:
- Anatomy and Suggested Two Dimensional Views:
  - Aortic relationship to the ventricular septum: ME LAX, deep TG (LVOT)
  
The degree of aortic override has been used as criteria for distinguishing between TOF and double outlet right ventricle (less than and more than 50% override respectively). However, it is important to recognize that the degree of override may vary according to the angle of two-dimensional interrogation, thus it is helpful to also consider additional criteria that would support one or the other diagnosis.

  Aortic root dilation is a frequent finding, which may be associated with aortic regurgitation.

*Right Ventricular Hypertrophy*

The compensatory response to the pressure load on the right ventricle results in increased wall thickness. This, as well the size of the ventricular cavities and the biventricular systolic function can be readily assessed qualitatively by TEE

TEE:
- Suggested Views:
  - LV/RV: ME 4 Ch, TG Mid SAX, ME RV inflow-outflow, TG RV inflow
REFERENCES


