

# Role of Comprehensive Perioperative Transesophageal Echocardiography in Ebstein's Anomaly

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## ABSTRACT

Ebstein's anomaly (EA) is a complex congenital anomaly with a broad morphological spectrum. Although typically characterized by apical displacement of the septal tricuspid leaflet (STL)  $\geq 8$  mm/m<sup>2</sup> body surface area (BSA), diagnostic categorization is confusing in cases with minor downward displacement of the leaflets. A comprehensive transesophageal echocardiography (TEE) has an integral role in the surgical decision-making and perioperative management of EA. Preoperative TEE aims to provide the morphological diagnosis, coexistent cardiac lesions most commonly, atrial septal defect (ASD), degree of atrialization of the right ventricle (RV), size of the functional RV, biventricular function, quantification of the resultant tricuspid regurgitation (TR), and the amenability of the leaflets to surgical repair. Postoperatively, TEE examination can elucidate valuable information about the competency of tricuspid apparatus, RV dysfunction, and any residual shunting across ASD. Three-dimensional echocardiography has been especially employed to study the leaflet morphology and coaptation defects. The TEE is also indispensable in guiding the management of critically sick neonates with EA. The Great Ormond Street Echocardiography (GOSE) score predicts mortality in neonatal EA and has been proposed for risk stratification. The TEE is also helpful in identifying the cases which are not suitable for a biventricular repair and the ones which may require a valve replacement.

**Keywords:** Ebstein's anomaly, Perioperative, Transesophageal echocardiography.

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## INTRODUCTION

Ebstein's anomaly is characterized by a spectrum of morphological features involving the tricuspid valve (TV) and the RV.<sup>1</sup> The leaflets and tensile apparatus of the atrioventricular valves are formed by a process of delamination of the ventricular layers. The process

of delamination is incomplete in EA with a resultant apical displacement of septal or posterior TV leaflets. In the most typical form, septal leaflet is the most nondelaminated with the anterior leaflet being the most delaminated leaflet. Usually, the anterior tricuspid leaflet (ATL) becomes enlarged to appear "sail" like. The tricuspid leaflets tend to be distorted, thickened, and dysplastic. The abnormal leaflets are often tethered due to the shortening of the chordae.

The functional tricuspid annulus is displaced downward in a spiral manner below the true tricuspid annulus resulting in valvular incompetence of varying severity. Thereby, the RV gets divided into two parts. The proximal part becomes atrialized with the remaining distal portion being the functional part. The atrialized part becomes dilated, thinned, and dyskinetic in long-standing disease, limiting its contractility with increased propensity for atrial arrhythmias. The functional portion of RV is smaller than the anatomic RV and myopathic with a decline in the number of myocardial fibers.<sup>2</sup>

Considering the morphological variability and the coexistent anomalies in EA, the spectrum or magnitude of disease and age of the patient dictate the definitive or palliative surgical management. The present article aims to review the role of TEE in surgical decision-making and perioperative management in patients with EA.

## PREOPERATIVE INFORMATION ON TEE

### Morphological Diagnosis

Echocardiography provides the definitive diagnosis and aids in the anatomical evaluation in EA. The characteristic echocardiographic finding is the apical displacement of the septal leaflet at the crux of the heart, computed as the displacement index  $\geq 8$  mm/m<sup>2</sup> BSA<sup>3</sup> (Fig. 1 and Video 1). In various other forms of congenital TR with minor apical displacement of the TV leaflets, it is confusing to characterize the TV anomaly as EA. It is crucial to identify such Ebsteinoid lesions, as the management strategies are different.

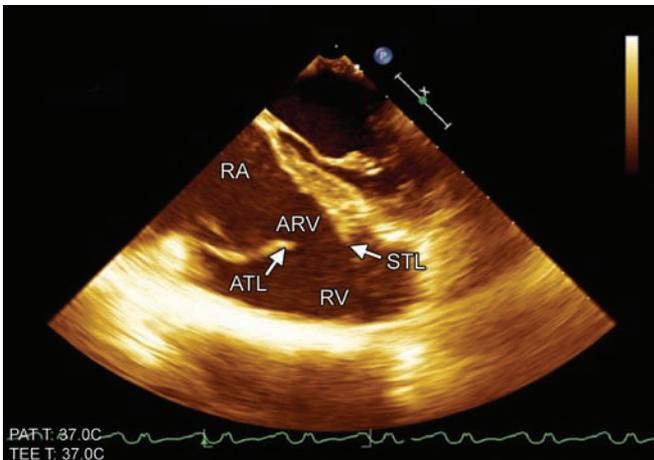
### TV Leaflet Anatomy and Mobility

The morphology and the degree of delamination of the various TV leaflets need to be meticulously evaluated in

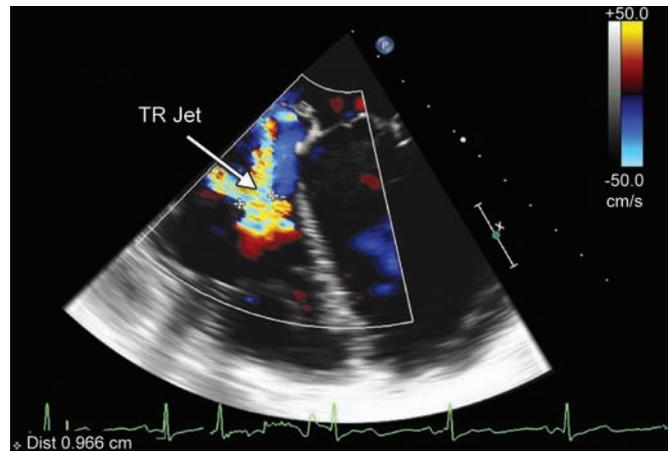
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**Fig. 1:** Two-dimensional mid-esophageal four-chamber view at 0° showing ATL, apically displaced STL, and a large atrialized portion of RV (ARV)



**Fig. 2:** Two-dimensional mid-esophageal four-chamber view at an angle of 0° with color Doppler across TV showing a regurgitant jet, with a vena contracta of 0.95 cm

the preoperative TEE. Midesophageal (ME) views like four-chamber view, RV inflow–outflow view, modified bicaval view, and transgastric RV basal short axis views are the most useful for this purpose.<sup>4</sup> The presence of leaflet fenestrations or tethering (significant adherence of the edge or body of the leaflet to underlying endocardium) may be evident in certain cases. The mobility of the leading edges of the leaflets can be studied in the ME four-chamber view.

### TV Annulus Assessment

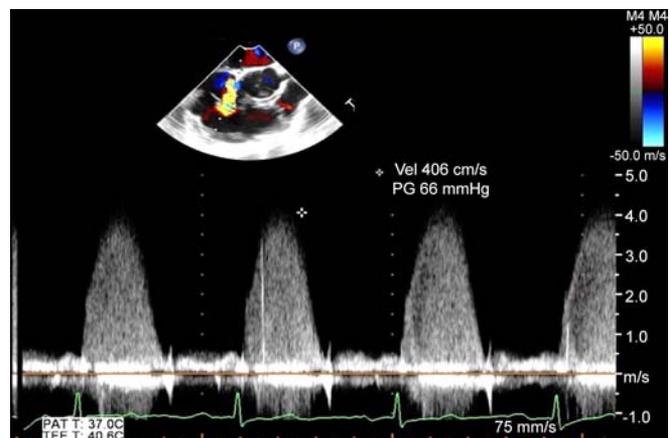
The quantification of the TV annulus indexed to the BSA of the patient is a valuable preoperative information, especially in the setting of TR. This is assessed in the ME four-chamber view and transgastric RV inflow–outflow view.

### The Degree of Atrialization of the RV

The fraction of the RV which becomes atrialized should be quantified on TEE, along with the assessment of the size and functioning of the RV distal to the functional annulus.

### Doppler Interrogation

Unrepaired EA is accompanied by moderate-to-severe TR wherein color flow mapping may fail to characterize the regurgitation jet completely owing to the relatively low-pressure difference between RV and right atrium (RA). Thus, the degree of TR is better demonstrated with continuous wave Doppler interrogation of low-velocity triangular Doppler trace across the TV (Figs 2 and 3 and Video 2) in the ME RV inflow–outflow view and modified bicaval view. Doppler interrogation also helps in identifying any evidence of obstruction in the right ventricular outflow tract (RVOT) and in assessment of



**Fig. 3:** Two-dimensional ME right ventricular inflow–outflow view at 62° showing continuous wave Doppler assessment across TV regurgitant jet, with a peak gradient of 66 mmHg

direction of shunting across interatrial septum (IAS) in an associated ASD.

### Biventricular Function, the Motion and Position of the Interventricular Septum

Right ventricular function assessment can be challenging in EA considering the morphologically variable substrates. Tricuspid annular plane systolic excursion is usually high or above normal in EA and can falsely overestimate RV function. Thereby, right ventricular fractional area change (RV-FAC) and Tei index are recommended for quantifying RV function.<sup>5,6</sup> Left ventricular systolic function and ejection fraction can be calculated using conventional parameters. Any evidence of regional wall motion abnormalities and paradoxical septal motion should be noted, as this may affect the surgical decision-making.

### Associated Cardiac Lesions

An ASD or a patent foramen ovale (PFO) is the most consistently observed cardiac lesion on TEE in EA.<sup>7</sup>

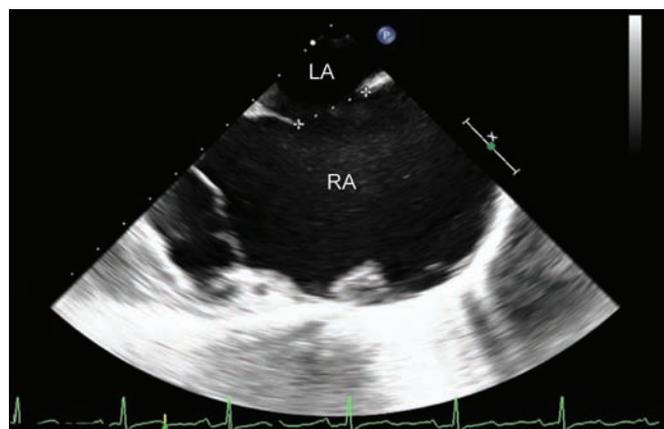
Interatrial septum should be thoroughly inspected in the bicaval TEE view (Fig. 4). Pulmonary atresia or stenosis, patent ductus arteriosus, tetralogy of Fallot, ventricular septal defect, transposition of great arteries, or congenital mitral stenosis may coexist in conjunction with EA.

### Disease Severity Classification

Carpentier et al.<sup>8</sup> proposed a severity classification system based on the size of the functional RV and mobility of the ATL (Table 1).<sup>1</sup>

### Role in Surgical Decision-making and Guiding the TV Repair

Assessment of the feasibility of a surgical TV repair is one of the initial and most relevant steps in planning the management of EA. The most widely practiced repair technique is the cone reconstruction wherein surgical delamination and mobilization of leaflet tissue are performed, followed by the anchoring of rotated reconstructed leaflet tissue at the true tricuspid annulus.



**Fig. 4:** Two-dimensional mid-esophageal bicaval view at an angle of 119° showing an ASD of about 2.05 cm in a case of EA. LA: Left atrium

**Table 1:** Severity classification of EA

Type A (minimal disease)	A large, mobile anterior leaflet Only a small part of RV is atrialized with an adequate volume of true RV
Type B (intermediate disease)	A mobile anterior leaflet but with abnormal chordae A large atrialized component of RV with a reduced size of true RV
Type C (severe disease)	The anterior leaflet movement is severely restricted and may result in RVOT obstruction Large atrialized RV with a small true RV
Type D (tricuspid sac lesion)	The anterior leaflet is immobile The RV is almost completely atrialized, except for a small infundibular component

The repair technique results in a cone-shaped TV with the RV apex forming the vertex of the cone. For this repair technique, a large-sized, mobile, adequately delaminated anterior leaflet with a free leading edge is a prerequisite. Table 2 summarizes the characteristics of the TV leaflets on TEE which favor a successful repair.<sup>9</sup>

### EMERGING ROLE OF THREE-DIMENSIONAL (3D) ECHOCARDIOGRAPHY

Real-time 3D echocardiography has revolutionized the perioperative management of congenital cardiac anomalies. The technique of multiplanar reconstruction (MPR) further helps in understanding the cardiac morphology (Table 3). Recent literature describes the correlation of the 3D-TEE findings with intraoperative surgical findings in EA, thereby, dictating the surgical management<sup>10,11</sup> (Figs 5 and 6 and Video 3).

### POSTREPAIR TEE

#### Adequacy of Surgical Repair and Competency of the TV Apparatus

The postoperative TEE examination should focus on the assessment of the surgical TV repair (Fig. 7). Any residual regurgitation (Fig. 8 and Video 4) or a resultant TV stenosis postrepair should be quantified and discussed with the surgical team.

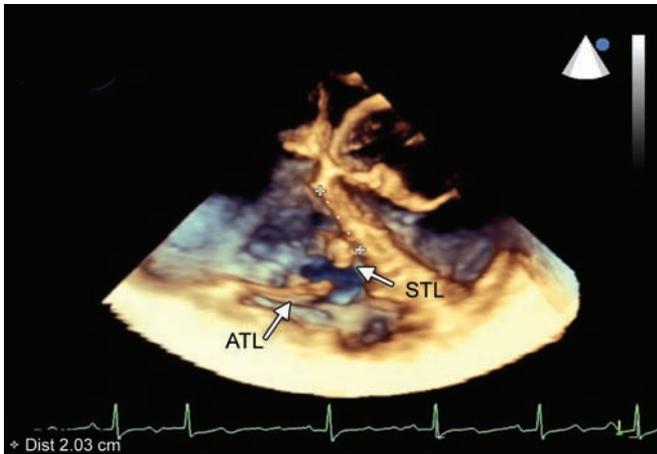
- Quantifying the degree of RV dysfunction: Varying extents of RV dysfunction may be present in the postoperative period, which remains a major issue of concern. TEE-based RV function assessment with RV-FAC and Tei index can guide the choice of inotropes and vasoactive agents postoperatively.

**Table 2:** Transesophageal echocardiography findings conducive to TV repair

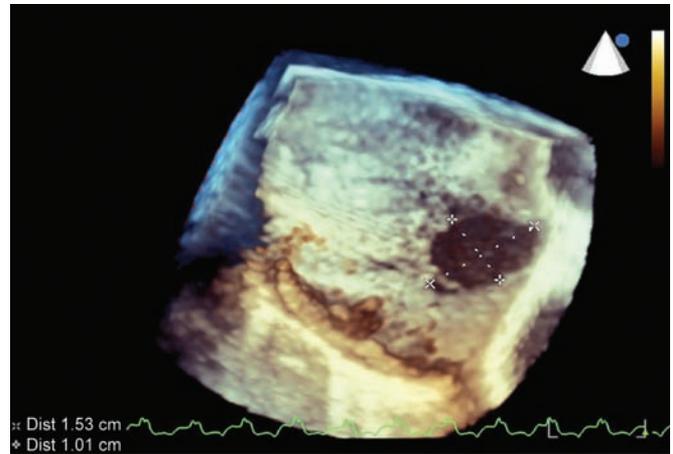
1.	Large, mobile anterior leaflet
2.	A free leading edge of the anterior leaflet
3.	Few free wall attachments with at least 50% delamination of the anterior leaflet
4.	Absence of any leaflet tethering or leaflet tissue in the RVOT
5.	Any amount of delaminated septal leaflet tissue (especially helpful in cone type repair)

**Table 3:** Various features of EA best evaluated with 3D TEE

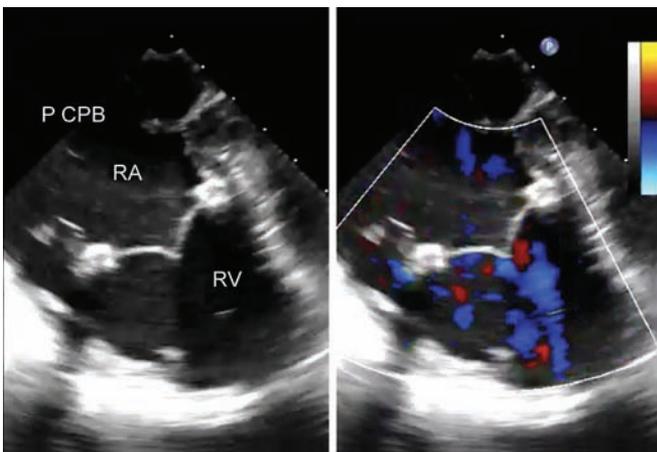
1.	Apical displacement of the TV leaflets
2.	Rotational anomaly of the TV (difficult to appreciate on two-dimensional TEE)
3.	TV dysplasia
4.	Leaflet morphology with degree of delamination
5.	Leaflet coaptation defects
6.	Coexisting anomalies of the RVOT, pulmonary valve, and tensile apparatus



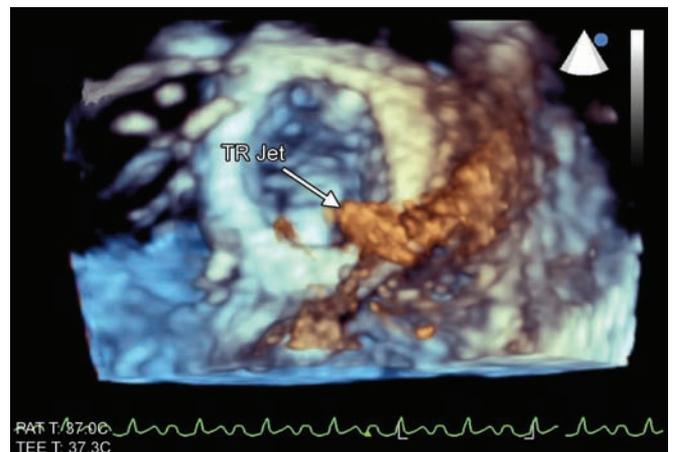
**Fig. 5:** Three-dimensional image of mid-esophageal four-chamber view at 0° showing a characteristic large sail-like ATL and a displaced septal leaflet with a displacement index of 1.35 cm/m<sup>2</sup> (2.03 cm/BSA of 1.5 m<sup>2</sup>)



**Fig. 6:** Three-dimensional view of TV showing lack of coaptation zone measuring 1.53 × 1.10 cm



**Fig. 7:** Two-dimensional mid-esophageal four-chamber view at 0° with color compared post-TV repair in EA showing adequate coaptation of leaflets with no regurgitation



**Fig. 8:** Three-dimensional image of post-TV repair in EA showing residual regurgitant jet

- Residual shunting across IAS or PFO: Any degree of blood flow across the IAS should be meticulously evaluated in order to rule out a residual shunt.

**TEE-BASED RISK STRATIFICATION**

The Celermajer et al.<sup>12</sup> score or the Great Ormond Street Score is an echocardiographic-based scoring system which is useful to prognosticate neonates with EA. Higher the grade, poorer the prognosis.

This four-chamber-based echocardiographic scoring is calculated as:

$$\frac{\text{Area of RA} + \text{area of atrialized RV}}{\text{Area of the functional RV} + \text{left atrium} + \text{left ventricle}}$$

The estimated mortality rate with a GOSE score of <0.5 is almost negligible in the background of an adequate surgical correction, whereas a score of 0.5 to 0.99 is associated with 10% mortality. A score of 1 to 1.5 accounts for a mortality rate of 44 to 100% and a score

more than 1.5 has a poor survival rate with an almost 100% mortality (Table 4).<sup>11</sup>

**TEE IN SPECIAL SITUATIONS**

**Critically Ill Neonates**

In the early neonatal period, the physiological elevation in pulmonary vascular resistance can exaggerate the TR associated with EA, resulting in an increased right to left shunting. They are also at elevated risk of right-sided heart failure and low cardiac output states.

An echocardiographic evaluation in the cases of neonatal EA helps to rule out any associated RVOT

**Table 4:** GOSE scoring system

Grade	GOSE ratio
1	<0.5
2	0.5–0.99
3	1–1.49
4	≥1.5



obstruction secondary to anatomic or functional pulmonary atresia which may be evident as a failure of anterograde pulmonary flow.<sup>13</sup> A trial of prostaglandin infusion may be attempted in neonates with functional pulmonary atresia. The subsequent failure of prostaglandin weaning after stabilization may necessitate a simple systemic to pulmonary artery shunt. As discussed, computing a composite Celermajer Score is useful in predicting mortality in these subset of patients to decide upon further management.

### One and a Half Ventricle/Single Ventricle Pathway

Demonstration of a poor RV function on echocardiography, as evidenced by a reduced Tei index or RV-FAC, precludes a successful biventricular repair. The subset of patients developing right heart dysfunction post-cardiopulmonary bypass would benefit from bidirectional cavo-pulmonary shunting (BDCPS).<sup>14,15</sup> Mild-to-moderate TV stenosis resulting from an aggressive repair, evident on Doppler as a gradient more than 6 to 7 mm Hg, may constitute another indication for BDCPS. Ideally, this shunting should be performed only if the pulmonary artery pressures are low as evident by a left ventricular end-diastolic pressure less than 15 mm Hg, transpulmonary gradient less than 10 mm Hg, and the mean pulmonary arterial pressure less than 18 to 20 mm Hg.<sup>16</sup>

A univentricular strategy or Starne's<sup>17-20</sup> approach is contemplated in neonates with an anatomic obstruction in the RV outflow due to the abnormal placement of the dysmorphic anterior leaflet encroaching the RVOT, rendering a successful repair unfeasible.

### TV Replacement

Echocardiographic assessment (EA) of anterior leaflet morphology with regard to the size, mobility, free leading edge, and degree of delamination may help to assess the feasibility of TV repair. Displacement of the anterior leaflet into the RV, extreme thickening, and dense attachment of the leaflet edge to the ventricular wall may preclude repair.<sup>21</sup> TV replacement is adopted as the last resort in background of a massively enlarged RV with dilated annulus, keeping the age of the patient in consideration.

### Plication of the Atrialized Ventricle

During plication of the atrialized portion of the RV, accidental injury to the right coronary artery and its branches may occur.<sup>22</sup> This may manifest as new onset regional wall motion abnormalities detected in the postoperative TEE examination.

### EVIDENCE

Quinonez et al.<sup>14</sup> reviewed their experience with one and a half ventricle repair in EA. The authors elaborated the role of BDCPS in EA as a preplanned procedure, an intraoperative rescue procedure in difficult CPB weaning or as an alternate to cardiac transplantation in a select group. Out of the 169 patients with EA who were taken up for surgical intervention, 14 patients underwent BDCPS. Nine were performed as planned procedures owing to the preoperative RV dysfunction and five BDCPS were done as a salvage manoeuvre in background of severe hemodynamic instability post-CPB. Three patients who were initially planned for cardiac transplantation owing to severe left ventricular dysfunction underwent BDCPS, which allowed a conventional repair in them. Thus, echocardiographic assessment of ventricular function can help in identifying patients who can be benefited with a BDCPS.

Tang et al.<sup>23</sup> retrospectively analyzed the intraoperative two-dimensional (2D) TEE data of 164 patients of EA in comparison with the operative findings. The analysis revealed high consistency rates of intraoperative 2D TEE data with the surgical findings to the extent of 93.2 and 96.1% for septal and posterior leaflets respectively. However, the TEE findings with regard to the anterior leaflet corroborated in only about 40.1% cases. The preoperative color Doppler demonstrated a severe degree of TR in 150 cases and moderate TR in the rest of the cases. Post-repair TEE revealed a moderate degree of TR persisting in 58 cases which indicated a revision valvuloplasty to address the regurgitation.

Planinc et al.<sup>24</sup> reported a case of EA which was diagnosed by 2D transthoracic echocardiography. Though they were able to elicit information regarding the morphology and function of the RV with 2D echocardiography, the TV apparatus morphology, and the true origin of septal and posterior leaflets with the coaptation zone was not well appreciated. The MPR mode and 3D reconstruction modalities were utilized to identify a complete agenesis of the posterior leaflet which accounted for the large zone of lack of coaptation and resulting TR; 3D analysis also revealed a complete absence of posterior leaflet subvalvular apparatus and posterior leaflet delamination. Thus, to get additional information regarding the morphology and orientation of TV apparatus, a 3D echocardiographic examination is mandatory.

van Noord et al.<sup>25</sup> described the application of perioperative 3D TEE during a TV repair for EA. The 3D TEE findings regarding morphology of TV, RVOT, and pulmonary valve were found to be consistent with intraoperative findings. The maintainability problem

report (MPR) mode analysis revealed a bicuspid pulmonary valve, which could not be detected during the preoperative 2D echocardiography. Additional information on the underlying morphology available in 3D imaging can be useful in the surgical conduct of the lesions harboring anatomical variations.

Schwitz et al.<sup>26</sup> reported a 41-year-old woman, a diagnosed case of EA with severe TR and Celemajer index of 0.55. Considering the dysplastic nature of the TV, a bioprosthetic valve replacement was performed with no significant gradient across the valve. Five years later, the patient became symptomatic. Transthoracic echocardiography revealed fusion and restricted motion of the TV leaflets, resulting in TV stenosis with a mean gradient of 10 mm Hg, and a mild TR. Following the refusal of the patient for undergoing a redo cardiac surgery, a percutaneous transfemoral TV implantation was planned with the aid of 3D TEE and cardiac computed tomography with regard to the feasibility and appropriate valve size. A 29 mm bioprosthetic valve was implanted in a hybrid catheterization lab set-up under real-time TEE guidance. Periprocedural TEE demonstrated a reduction of the transvalvular mean gradient from 7 to 1 mm Hg.

Cardiac anomalies associated with TR and predominant right-sided chamber enlargement can mimic EA resulting in a wrong diagnosis and management. Ammash et al.<sup>27</sup> retrospectively analyzed a subset of 22 patients with Ebstein's like lesions with similar clinical picture. All of them had right atrial and ventricular enlargement and 82% had associated RV dysfunction. But the usual features of EA like septal leaflet displacement  $\geq 8$  mm/m<sup>2</sup> and large redundant anterior leaflet were absent on echocardiography. In two peculiar cases, TEE examination further substantiated the transthoracic echocardiographic findings. The echocardiographic observations were confirmed in the 15 patients who underwent surgery. The TR in the studied subset was secondary to multiple etiologies like TV dysplasia, valve prolapse, trauma, endocarditis, RV dysplasia, and annular dilatation secondary to pulmonary valve regurgitation. Recognition of the mimics of EA has got important surgical implications.

## CONCLUSION

Ebstein's anomaly is a complex congenital anomaly with several morphological characteristics of varying severity. Intraoperative decision-making with regard to tailoring the surgical procedure to the patient's morphology is an important predictor of outcome. A comprehensive TEE examination helps in assessing the feasibility and adequacy of repair in EA and detecting postrepair issues that can complicate the postoperative course of the cohort of these

patients. The role of TEE in surgical decision-making continues to evolve with the advent of echocardiographic advancements, such as 3D TEE and MPR.

## REFERENCES

1. Attenhofer Jost CH, Connolly HM, Dearani JA, Edwards WD, Danielson GK. Ebstein's anomaly. *Circulation* 2007 Jan;115(2): 277-285.
2. Anderson KR, Lie JT. The right ventricular myocardium in Ebstein's anomaly: a morphometric histopathologic study. *Mayo Clin Proc* 1979 Mar;54(3):181-184.
3. Seward JB. Ebstein's anomaly: ultrasound imaging and hemodynamic evaluation. *Echocardiography* 1993 Nov;10(6): 641-664.
4. Huttin O, Voilliot D, Mandry D, Venner C, Juillière Y, Selton-Suty C. All you need to know about the tricuspid valve: tricuspid valve imaging and tricuspid regurgitation analysis. *Arch Cardiovasc Dis* 2016 Jan;109(1):67-80.
5. Gatzoulis, MA.; Webb, GD.; Daubeney, PE. Diagnosis and management of adult congenital heart disease. 3rd ed. Philadelphia (PA): Elsevier; 2018. p. 66.
6. Eidem BW, Tei C, O'Leary PW, Cetta F, Seward JB. Nongeometric quantitative assessment of right and left ventricular function: myocardial performance index in normal children and patients with Ebstein anomaly. *J Am Soc Echocardiogr* 1998 Sep;11(9):849 -856.
7. Watson H. Natural history of Ebstein's anomaly of the tricuspid valve in childhood and adolescence: an international cooperative study of 505 cases. *Br Heart J* 1974 May;36(5):417-427.
8. Carpentier A, Chauvaud S, Mace L, Relland J, Mihaileanu S, Marino JP, Abry B, Guibourt P. A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. *J Thorac Cardiovasc Surg* 1988 Jul;96(1):92-101.
9. Dearani JA, Bacha E, da Silva JP. Cone reconstruction of the tricuspid valve for Ebstein's anomaly: anatomic repair. *Oper Tech Thorac Cardiovasc Surg* 2008 Summer;13(2):109-125.
10. Bharucha T, Anderson RH, Lim ZS, Vettukattil JJ. Multiplane review of three-dimensional echocardiography gives new insights into the morphology of Ebstein's malformation. *Cardiol Young* 2010 Feb;20(1):49-53.
11. Vettukattil JJ, Bharucha T, Anderson RH. Defining Ebstein's malformation using three dimensional echocardiography. *Interact Cardiovasc Thorac Surg* 2007 Dec;6(6):685-690.
12. Celermajer DS, Cullen S, Sullivan ID, Spiegelhalter DJ, Wyse RK, Deanfield JE. Outcome in neonates with Ebstein's anomaly. *J Am Coll Cardiol* 1992 Apr;19(5):1041-1046.
13. Jaquiss RD, Imamura M. Management of Ebstein's anomaly and pure tricuspid insufficiency in the neonate. *Semin Thorac Cardiovasc Surg* 2007 Autumn;19(3):258-263.
14. Quinonez LG, Dearani JA, Puga FJ, O'Leary PW, Driscoll DJ, Connolly HM, Danielson GK. Results of the 1.5-ventricle repair for Ebstein anomaly and the failing right ventricle. *J Thorac Cardiovasc Surg* 2007 May;133(5):1303-1310.
15. Raju VK, Dearani JA, Burkhart HM, Grogan M, Phillips SD, Ammash N, Pike RP, Johnson JN, O'Leary PW. Right ventricular unloading for heart failure related to Ebstein malformation. *Ann Thorac Surg* 2014 Jul;98(1):167-174.
16. Dearani JA, O'Leary PW, Danielson GK. Surgical treatment of Ebstein's malformation: state of the art in 2006. *Cardiol Young* 2006 Sep;16(Suppl 3):12-20.

17. Reemtsen BL, Fagan BT, Wells WJ, Starnes VA. Current surgical therapy for Ebstein anomaly in neonates. *J Thorac Cardiovasc Surg* 2006 Dec;132(6):1285-1290.
18. Reemtsen BL, Polimenakos AC, Fagan BT, Wells WJ, Starnes VA. Fate of the right ventricle after fenestrated right ventricular exclusion for severe neonatal Ebstein anomaly. *J Thorac Cardiovasc Surg* 2007 Dec;134(6):1406-1410, discussion 1410-1412.
19. Reemtsen BL, Starnes VA. Fenestrated right ventricular exclusion (Starnes' procedure) for severe neonatal Ebstein's anomaly. *Oper Tech Thorac Cardiovasc Surg* 2008 Summer; 13(2):91-100.
20. Starnes VA, Pitlick PT, Bernstein D, Griffin ML, Choy M, Shumway NE. Ebstein's anomaly appearing in the neonate. A new surgical approach. *J Thorac Cardiovasc Surg* 1991 Jun;101(6):1082-1087.
21. Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Oliver WC Jr. Operative treatment of Ebstein's anomaly. *J Thorac Cardiovasc Surg* 1992 Nov;104(5):1195-1202.
22. Brown ML, Dearani JA, Danielson GK, Cetta F, Connolly HM, Warnes CA, Li Z, Hodge DO, Driscoll DJ. Functional status after operation for Ebstein anomaly: the Mayo Clinic experience. *J Am Coll Cardiol* 2008 Aug;52(6):460-466.
23. Tang XJ, Bao M, Zhao H, Wang LY, Wu QY. Intraoperative transesophageal echocardiography in the operation of Ebstein's anomaly: a retrospective study. *Chin Med J (Engl)* 2017 Jul;130(3):1540-1543.
24. Planinc I, Baricevic Z, Reskovic Luksic V, Lovric D, Veceric S, Hanezevacki JS. Three dimensional echocardiography in classification of Ebsteins anomaly: a case report on agenesis of the tricuspid valve posterior leaflet. *Cardiologia Croatica* 2013 May/Jun;8(5-6):201-202.
25. van Noord PT, Scohy TV, McGhie J, Bogers AJ. Three-dimensional transesophageal echocardiography in Ebstein's anomaly. *Interact Cardiovasc Thorac Surg* 2010 May;10(5):836-837.
26. Schwitz F, Wenaweser P, Kadner A, Wustmann K, Windecker S, Schwerzmann M. A valve in valve approach for Ebstein's anomaly. *Cardiovasc Med* 2015 Apr;18(4):139-141.
27. Ammash NM, Warnes CA, Connolly HM, Danielson GK, Seward JB. Mimics of Ebstein's anomaly. *Am Heart J* 1997 Sep;134(3):508-513.