Role of transesophageal echocardiography for intraoperative decision-making in double-chamber right ventricle with ventricular septal defect and absent pulmonary valve

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Absent pulmonary valve is a variant of tetralogy of Fallot, characterized by small or rudimentary, stenotic or regurgitant pulmonary valve with dilated pulmonary artery. It is uncommonly associated with double-chamber right ventricle. We hereby report a case with preoperative diagnosis of ventricular septal defect with pulmonary stenosis based on routine transthoracic echocardiography. Intraoperatively, transesophageal echocardiography showed a rare combination of double-chamber right ventricle in association with ventricular septal defect and absent pulmonary valve, which was confirmed during surgery.

Keywords:
absent pulmonary valve, double-chamber right ventricle, transesophageal echocardiography, ventricular septal defect

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Introduction

Double-chambered right ventricle (DCRV) accounts for only 0.5–2% of all congenital heart diseases. It is a rare disorder, most commonly associated with a membranous-type ventricular septal defect (VSD) [1]. The other coexisting lesions include subaortic stenosis, pulmonary valve stenosis, double-outlet right ventricle (RV), tetralogy of Fallot (TOF), anomalous pulmonary venous drainage, dextro transposition of the great arteries or corrected transposition of the great arteries, pulmonary atresia with intact ventricular septum, and Ebstein anomaly [2]. The association of absent pulmonary valve with DCRV has been described only once in the literature [3].

Case report

A female patient, aged 25 years old, weighing 45 kg, presented with mild dyspnea on exertion. Transthoracic echocardiography (TTE) and cardiac catheterization diagnosed a cyanotic congenital heart disease characterized by malaligned subaortic VSD with no aortic override, valvular and infundibular pulmonary stenosis (PS), bilateral confluent pulmonary arteries, normal biventricular function, normal coronaries, and no aortopulmonary collaterals. The initial surgical plan was the repair of ventricular septal defect and infundibular resection/pulmonary valvotomy.

After uneventful induction of anesthesia and endotracheal intubation, transesophageal echocardiographic (TEE) examination revealed a subaortic VSD, anomalous muscle band in RV suggestive of type 1 DCRV, and rudimentary pulmonary valve (Figs 1 and 2 and Videos 1 and 2). Pulmonary artery was dilated and aneurysmal, as seen in absent pulmonary valve variants of TOF in contrast to hypoplastic pulmonary artery seen in classical TOF. DCRV showed a peak gradient of 27 mmHg across the intraventricular muscular bundle. Color Doppler showed trivial regurgitation across the pulmonary annulus and right to left shunt across the VSD. Intraoperative surgical findings confirmed large subaortic VSD, with no aortic override, and thickened intraventricular muscle band in the RV with rudimentary tissue at the pulmonary annulus. Corrective surgery was performed that included VSD closure using Dacron patch, RV muscle band resection, bicuspid pulmonary valve reconstruction with thin polytetrafluroethylene membrane (0.1 mm) using the Graham Nunn's technique [4], and right ventricular outflow tract (RVOT) reconstruction with transannular polytetrafluroethylene patch (Fig. 3 and Video 3).

Discussion

DCRV is characterized by the presence of an anomalous muscle bundle, which divides the RV into two s

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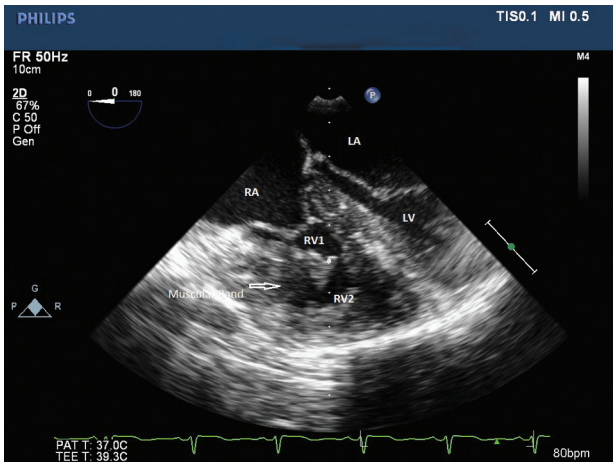
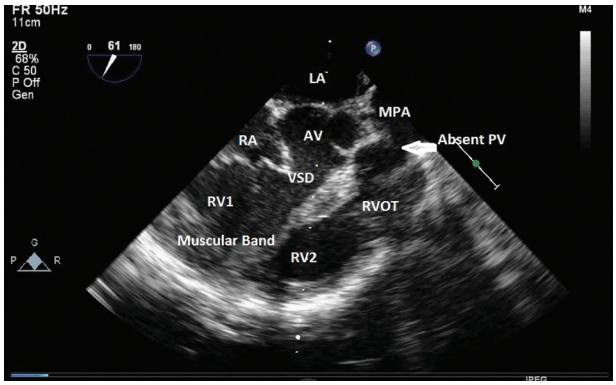
Figure 1



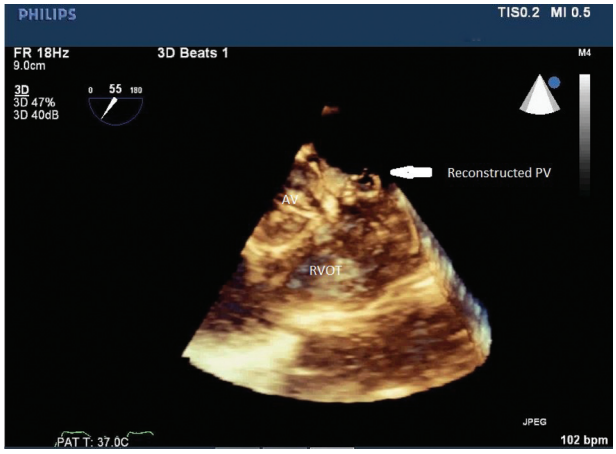
Two-dimensional mid-esophageal four-chamber view at a sector angle of 0° showing the thickened intraventricular muscle band in the right ventricle. LA, left atrium; LV, left ventricle; RA, right atrium; RV1, proximal right ventricle; RV2, distal right ventricle.

Figure 2



Two-dimensional mid-esophageal right ventricular inflow–outflow view at a sector angle of 61° showing ventricular septal defect and rudimentary pulmonary valve. AV, aortic valve; LA, left ventricle; MPA, main pulmonary artery; PV, pulmonary valve; RA, right atrium; RV1, proximal right ventricle; RV2, distal right ventricle; RVOT, right ventricular outflow tract; VSD, ventricular septal defect.

Figure 3



Three-dimensional mid-esophageal right ventricular inflow–outflow view at a sector angle of 55° showing reconstructed RVOT and PV. AV, aortic valve; PV, pulmonary valve; RVOT, right ventricular outflow tract.

eparate chambers, namely the proximal high-pressure and the distal low-pressure chambers. The location of the muscle bundle can be high (horizontal), close to the pulmonary valve, or low (oblique), close to the apex. Galiuto *et al.* [5] divided DCRV into two distinct types of intracavitary obstruction. Type I DCRV is characterized by the presence of anomalous muscle bundle crossing the RV cavity causing intraventricular obstruction. Type 2 DCRV is characterized by the absence of anomalous muscle bundle. There is marked parietal and septal muscle hypertrophy leading to intraventricular obstruction. The intraventricular pressure gradient of type 1 DCRV is greater than that of type 2 DCRV. Ventricular septal defect is more commonly associated with type 2 DCRV [5].

Chevers [6] first described congenital absence of the pulmonary valve leaflets in 1847. The earliest report of the association of absent pulmonary valve and VSD was by Royer and Wilson [7]. Absent pulmonary valve with VSD and DCRV was first reported in 2014 [3].

Absent pulmonary valve is a rare congenital anomaly, characterized by dysplastic or absent pulmonary valve tissue and severe pulmonary regurgitation. Rarely isolated, it is usually associated with TOF [8] with VSD and obstructive subvalvular pulmonary ring, but not typically with DCRV [3]. Absent pulmonary valve with aneurysmally dilated pulmonary arteries may present with life-threatening respiratory obstruction and cardiac failure in a neonate. It can remain asymptomatic in a few patients for many years [8].

Diagnosis of DCRV can often be missed on routine adult TTE, as the RV outflow tract is not always a routine part of the adult echocardiographic examination and can be difficult to visualize [9]. When evaluating the patient, cardiologists should also image the entire right heart complex [9]. RVOT is best visualized in subcostal view than the parasternal and apical views [10]. TEE is much more useful than TTE for delineating anatomy and estimating the pressure gradient [1]. Routine preoperative TEE is not required in all cases of VSD and PS. However, in adults with poor echocardiographic window, TEE should be performed.The preoperative TTE of this patient did not diagnose DCRV. Erroneous PS was diagnosed instead of DCRV. Probably on TTE examination, by using continuous-wave Doppler, the

pressure gradients across the anomalous muscle bundle between inflow and the outflow portion of RV were erroneously interpreted as gradients across the pulmonary valve. To prevent this error, the site of obstruction could be located by detecting the origin of high-velocity flow using color Doppler and appearance of the mosaic pattern.

In our case, diagnosis was missed on cardiac catheterization as well. DCRV can be confused with infundibular PS on angiography due to the mal-placement of the catheter tip for measuring gradient. The positioning of the catheter tip can thus be misleading. In our setup, diagnosis is made on TTE, and cardiac catheterization is performed to rule out additional VSD or aortopulmonary collaterals in cases of VSD and PS. Performing an additional lateral view for RV angiogram would have clearly delineated DCRV. However, concerns about contrast load and the need for additional angiogram apart from left ventricle, aortic root, and descending aorta do not permit two RV angiograms. RV angiogram is done in anterioposterior view generally to profile pulmonary arteries and its bifurcation. In most of the cases, anterioposterior view is sufficient to delineate the site of RVOT obstruction, but was not in this particular case. If lateral view was performed, it could have helped in making the diagnosis.

In this case, intraoperative TEE had a better visualization of the RV and RVOT leading to a diagnosis of DCRV with VSD and absent pulmonary valve, which was subsequently confirmed during surgery.

Conclusion

DCRV with VSD and absent pulmonary valve is a rare combination. RVOT examination by TTE should be routinely performed in adults. This case further emphasizes the role of intraoperative TEE in diagnosis and change in the surgical management plan during cardiac surgeries.

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Conflicts of interest

There are no conflicts of interest.

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