Pulmonary Valvular Stenosis

in L-Transposition of the Great Arteries with Ventricular Septal Defect

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A 39-year-old man presented with a fever of 102 °F. He had a history of viridans group streptococci (VGS) bacteremia, levo-transposition of the great vessels (also known as congenitally corrected transposition), large perimembranous ventricular septal defect (VSD), and valvular pulmonary stenosis. Prior cardiac catheterization had shown his circulation to be well balanced, with a ratio of pulmonary blood flow to systemic blood flow of 1.4:1. Examination revealed nothing unusual except for a harsh systolic ejection murmur along the left sternal border. No emboli or immunologic phenomena were seen. The patient's white blood cell count and cell count differential were normal, but his erythrocyte sedimentation rate and C-reactive protein level were elevated. During his initial 48 hours after presentation, 8 of 8 blood cultures grew VGS.

We performed transesophageal echocardiography (TEE) to look for valvular vegetations and found neither intracardiac vegetations nor other evidence of endocarditis. Depicted are atrial–ventricular discordance (Fig. 1); ventriculo–arterial discordance (Figs. 2 and 3); short-axis view of the semilunar valves (Figs. 4 and 5); 3-dimensional

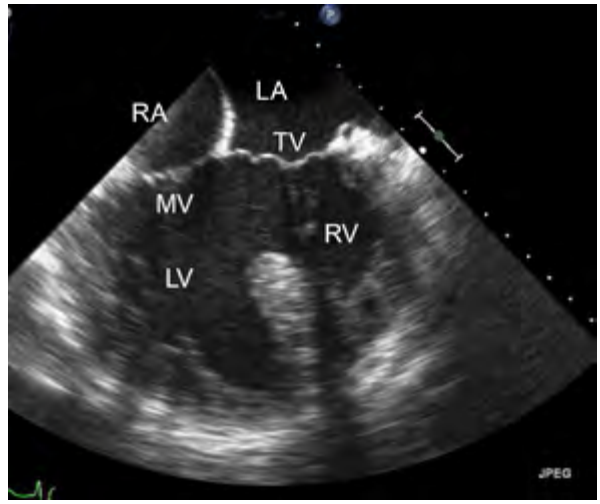

Fig. 1 *Transesophageal echocardiogram (0° midesophageal view) depicts a 4-chamber view. Note the usual arrangement of the atria, with atrioventricular discordance and the large inlet ventricular septal defect.*

LA = left atrium; LV = left ventricle; MV = mitral valve; RA = right atrium; RV = right ventricle; TV = tricuspid valve

Supplemental motion image is available for Figure 1.

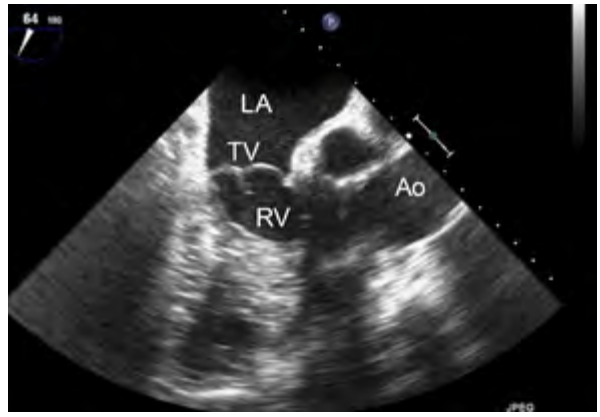

Fig. 2 *Transesophageal echocardiogram (60° midesophageal view) depicts atrioventricular and ventriculoarterial discordance between the left atrium, right ventricle, and aorta.*

Ao = aorta; LA = left atrium; RV = right ventricle; TV = tricuspid valve

Supplemental motion image is available for Figure 2.

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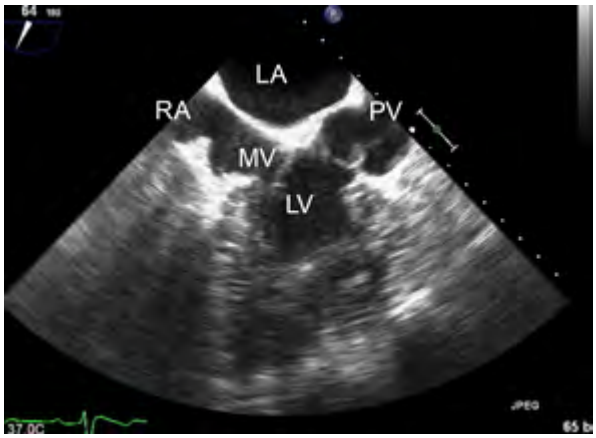

Fig. 3 *Transesophageal echocardiogram (60° midesophageal view) depicts atrioventricular and ventriculoarterial discordance between the right atrium, left ventricle, and pulmonary artery.*

LA = left atrium; LV = left ventricle; MV = mitral valve; PV = pulmonary valve; RA = right atrium

Supplemental motion image is available for Figure 3.

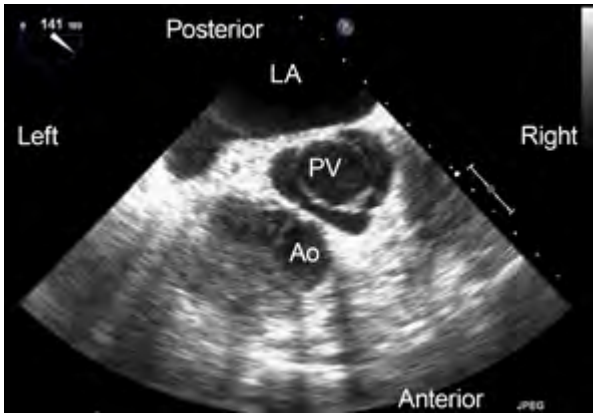

Fig. 4 *Transesophageal echocardiogram (140° midesophageal view) depicts short-axis view across the semilunar valves caudally. The aorta is anterior and leftward (L-transposition) to the pulmonary valve.*

Ao = aorta; LA = left atrium; PV = pulmonary valve

Supplemental motion image is available for Figure 4.

(3D) imaging of the domed unicommissural pulmonic valve (Fig. 6); and a peak gradient of 80 mmHg, which suggests protected pulmonary arterial pressures (Fig. 7).

In Figure 1, the atrioventricular (AV) valves appear to be in the same plane, which raises the possibility of a partial AV canal with a large inlet VSD—rather than the usual apical displacement of the tricuspid valve (not shown) seen in structurally normal hearts and in congenitally corrected transposition. Also note that the AV valves follow their respective ventricles (Figs. 1–3), not their supplying atria. In comparing planar (Figs. 4 and 5) with 3D imaging (Fig. 6), note that the "bicommissural" valve is better defined with 3D imaging as unicommissural.

Comment

Only 1% of L-transposition cases are uncomplicated: 90% have systemic tricuspid valve abnormalities, 70% have VSDs, and 40% have pulmonary stenosis. Arrhythmias include a 2%-per-year risk of complete AV block because of abnormal position of the AV node and accessory pathways (for example, pre-excitation or Wolff-Parkinson-White syndrome) associated with Ebsteinoid tricuspid valves.[1] Severe systemic tricuspid regurgitation is a class I indication for repair and early referral; in a study by Mongeon and colleagues, patients in whom the preoperative systemic ventricular ejection fraction was >0.40 had better outcomes.[2] Our bacteremic patient had normal biventricular function

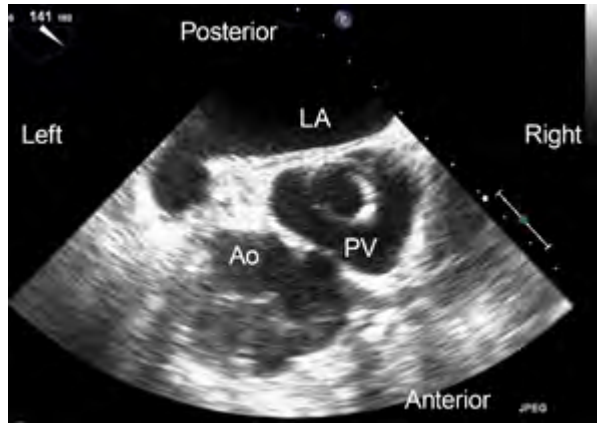

Fig. 5 *Transesophageal echocardiogram (140° midesophageal view) depicts short-axis view across the semilunar valves cranially. The pulmonary valve appears to be bicommissural, stenotic, thickened, and domed.*

Ao = aorta; LA = left atrium; PV = pulmonary valve

Supplemental motion image is available for Figure 5.

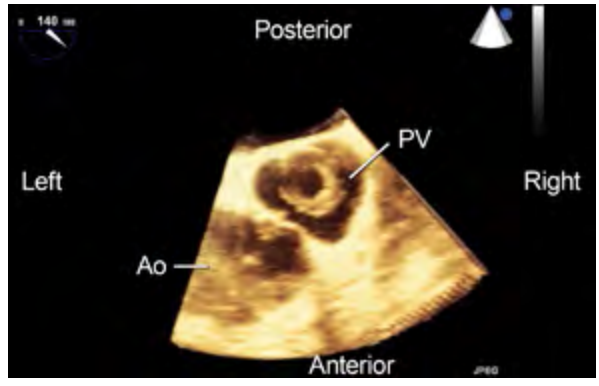

Fig. 6 *Three-dimensional transesophageal echocardiogram (140° midesophageal view) depicts short-axis view and level of pulmonary stenosis. The pulmonary valve is clearly unicommissural.*

Ao = aorta; PV = pulmonary valve

Supplemental motion image is available for Figure 6.

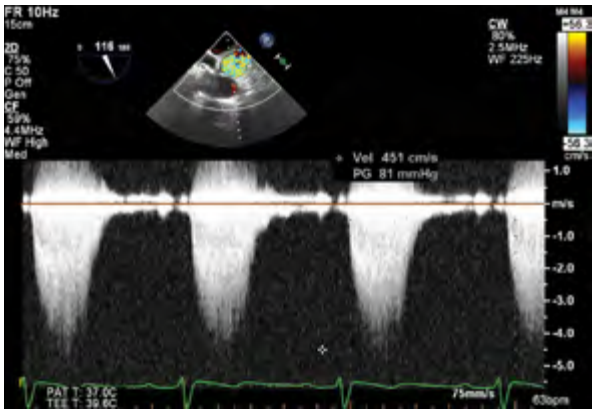

Fig. 7 *Continuous-wave Doppler echocardiogram across the pulmonary valve depicts an 80-mmHg gradient (4.5 m/s peak velocity) in the setting of systemic right ventricular and protected pulmonary arterial pressures.*

and remained in sinus rhythm with only trivial tricuspid regurgitation. However, closure of his VSD would be indicated should he develop confirmed infective endocarditis.[1] As in structurally normal hearts, 3D TEE can be useful in congenital heart disease when endocarditis is suspected. In addition, it can confirm complex anatomic arrangements and define associated defects.

References

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2. Mongeon FP, Connolly HM, Dearani JA, Li Z, Warnes CA. Congenitally corrected transposition of the great arteries ventricular function at the time of systemic atrioventricular valve replacement predicts long-term ventricular function. J Am Coll Cardiol 2011;57(20):2008-17.