A 19-year-old woman with mild exercise intolerance was referred for follow-up examination of a known ventricular septal defect (VSD; Figure 1A). By continuous-wave Doppler, a gradient across the tricuspid valve of 60 mm Hg was recorded. The parasternal short-axis view revealed an anomalous muscle bundle with insertion at the interventricular septum and the right ventricular (RV) free wall (Figure 1B). RV obstruction was suggested by a turbulent Doppler color flow velocity pattern (Figure 1C). However, it was impossible to locate the origin of the pressure gradient exactly using pulsed-wave Doppler, and the patient was referred for cardiac catheterization to rule out pulmonary hypertension. Right heart catheterization revealed normal pulmonary artery pressure (16/6 mm Hg) and a normal pressure in the RV outflow tract (16/4 mm Hg). The oximetric determined pulmonic-to-systemic shunt ratio was 1.3, confirming a hemodynamically nonsignificant small left-to-right shunt. On advancing the catheter into the RV apex, a pressure of 70/5 mm Hg was recorded (Figure 2). Placement of the catheter tip in the VSD was excluded by oxymetry. Thus, the diagnosis of a double-chambered right ventricle was confirmed.

Double-chambered right ventricle is a rare congenital heart disorder involving 2 different RV pressure compartments that is often associated with malalignment VSD. Usually, the obstruction is caused by an anomalous muscle bundle crossing the RV from the interventricular septum to the RV free wall. We emphasize that an increased tricuspid regurgitant gradient, as measured by continuous-wave Doppler, may be caused by a double-chambered right ventricle and not by pulmonary hypertension.

Figure 1. Echocardiographic findings (parasternal short-axis view) in a patient with double-chambered right ventricle and malalignment VSD. A, Malalignment VSD with shunt into the RV (small arrow). B, Anomalous RV muscle bundle (large arrow). C, Turbulent Doppler color flow velocity pattern, suggesting RV obstruction. LV indicates left ventricle; PV, pulmonic valve; and TV, tricuspid valve.
Figure 2. RV pressure tracings in a patient with double-chambered right ventricle. A, High pressure values in the RV apex. B, Normal pressure values in the RV outflow tract.
Double-Chambered Right Ventricle
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