Isolated Right Ventricular Apical Hypoplasia Without Atrial Septal Defect

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A 25-year-old woman was admitted due to an abnormal electrocardiogram (Supplementary Figure A). Transthoracic ultrasound cardiogram showed hypoplasia from the middle portion to the apex in the right ventricle (RV) and enlargement in the right atrium (Figure A,B; Supplementary Movie). Cardiac magnetic resonance imaging (MRI) showed the absence of the trabecular portion of the RV and the presence of normally developed tricuspid and pulmonary valves (Figure C,D). Cardiac catheterization showed no evidence of intercardiac shunt. The most striking finding on the pressure curves was a wave in the pulmonary artery and RV, which indicated that atrial kick was prominent (Figure F–H). Blood flow analysis of the pulmonary artery trunk derived from phase-contrast MRI showed an abnormal blood flow during diastole that could be attributed to the transmission of the flow by the right atrial kick (Figure I).

Disclosures
The authors declare no conflicts of interest.

Supplementary Files

Supplementary Movie. Color Doppler imaging of a transthoracic ultrasound cardiogram.
Please find supplementary file(s):

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Figure. Transthoracic ultrasound cardiogram in (A) diastole and (B) systole. (C,D) Cardiac magnetic resonance imaging in (C) diastole and (D) systole. (E) Schematic diagram of the 4 cavities. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle. (F–I) Upper curves, electrocardiogram. (F–H) Lower curves, intracardiac pressure on right heart catheterization (Y axis, mmHg). (F) High a waves (white arrows) and low ventricular v waves (black arrows) were observed in the RA. (G) Systolic pressure was decreasing in the RV (18 mmHg, arrowheads), and a right atrial a wave (white arrows) was transmitted to the RV. (H) A right atrial a wave (white arrows) was also recorded in the pulmonary artery (PA). (I) Abnormal blood flow of the PA trunk during diastole derived from phase-contrast magnetic resonance imaging (white arrow).