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 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
Nils Kucher, Christian Seiler, Yves Allemann and Franz Robert Eberli

Circulation. 2001;103:e105-e106
doi: 10.1161/01.CIR.103.21.e105
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2001 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/103/21/e105