Prevalence and Clinical Correlates of Right Ventricular Dysfunction in Patients With Hypertrophic Cardiomyopathy

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Hypertrophic cardiomyopathy (HC) is a disease that mainly affects the left ventricle (LV), however recent studies have suggested that it can also be associated with right ventricular (RV) dysfunction. The objective of this study was to determine the prevalence of RV dysfunction in patients with HC and its relation with LV function and outcome. A total of 324 consecutive patients with HC who received care at Stanford Hospital from 1999 to 2012 were included in the study. A group of 99 prospectively recruited age- and gender-matched healthy volunteers were used as controls. RV function was quantified using the RV fractional area change, tricuspid annular plane systolic excursion (TAPSE), and RV myocardial performance index (RVMPI). Compared with the controls, the patients with HC had a higher RVMPI (0.51 ± 0.18 vs 0.25 ± 0.06, p <0.001) and lower TAPSE (20 ± 3 vs 24 ± 4, p <0.001). RV dysfunction based on an RVMPI >0.4 and TAPSE <16 mm was found in 71% and 11% of the HC and control groups, respectively. Worst LV function and greater pulmonary pressures were independent correlates of RV dysfunction. At an average follow-up of 3.7 ± 2.3 years, 17 patients had died and 4 had undergone heart transplantation. LV ejection fraction <50% and TAPSE <16 mm were independent correlates of outcome (hazard ratio 3.98, 95% confidence interval 1.22 to 13.04, p = 0.02; and hazard ratio 3.66, 95% confidence interval 1.38 to 9.69, p = 0.009, respectively). In conclusion, RV dysfunction based on the RVMPI is common in patients with HC and more frequently observed in patients with LV dysfunction and pulmonary hypertension. RV dysfunction based on the TAPSE was independently associated with an increased likelihood of death or transplantation. **(Am J Cardiol 2014;113:361−367)**

Hypertrophic cardiomyopathy (HC) is a genetic disease associated with left ventricular (LV) hypertrophy,[1] which commonly involves the left ventricular septum. Recent studies have shown that cardiac hypertrophy is not limited to the left ventricle. In a magnetic resonance study by Maron et al,[2] the right ventricular (RV) wall thickness was increased in patients with HC compared with controls. To date, however, only limited data are available on the prevalence and clinical correlates of RV dysfunction in patients with HC. In the present study, we sought to determine the prevalence of RV dysfunction in a large series of patients with HC, using 3 echocardiographic indexes: the tricuspid annular plane systolic excursion (TAPSE), RV fractional area change, and RV myocardial performance index (RVMPI). We also sought to determine which factors were associated with a higher incidence of RV dysfunction. Finally, our last objective was to determine in an exploratory analysis whether RV systolic dysfunction was independently associated with an increased likelihood of death or transplantation.

Methods

From January 1999 to January 2012, 434 consecutive patients with HC were enrolled in the Stanford Inherited Cardiomyopathy Registry. In the present study, we retrospectively analyzed 324 cases in which the calculation of the MPI of both ventricles was feasible. The patients were enrolled in the study at their first echocardiogram. We excluded from our analysis technically difficult studies, examinations that did not allow an assessment of the MPI, patients in atrial fibrillation, and those in paced rhythm (because an MPI evaluation in these 2 situations has been considered to be unreliable). The diagnosis of HC was determined by the presence of significant LV hypertrophy (end-diastolic wall thickness ≥15 mm at M-mode or 2-dimensional echocardiography) in the absence of other etiologies, according to international criteria, or a wall thickness of 13 to 15 mm, in presence of abnormal electrocardiographic findings, or a familial history of inherited cardiomyopathy.[1] Patients with LV systolic dysfunction at enrollment were included in the present study if they had a clear documented history of HC and preserved LV ejection fraction on previous echocardiographic examinations

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See page 366 for disclosure information.

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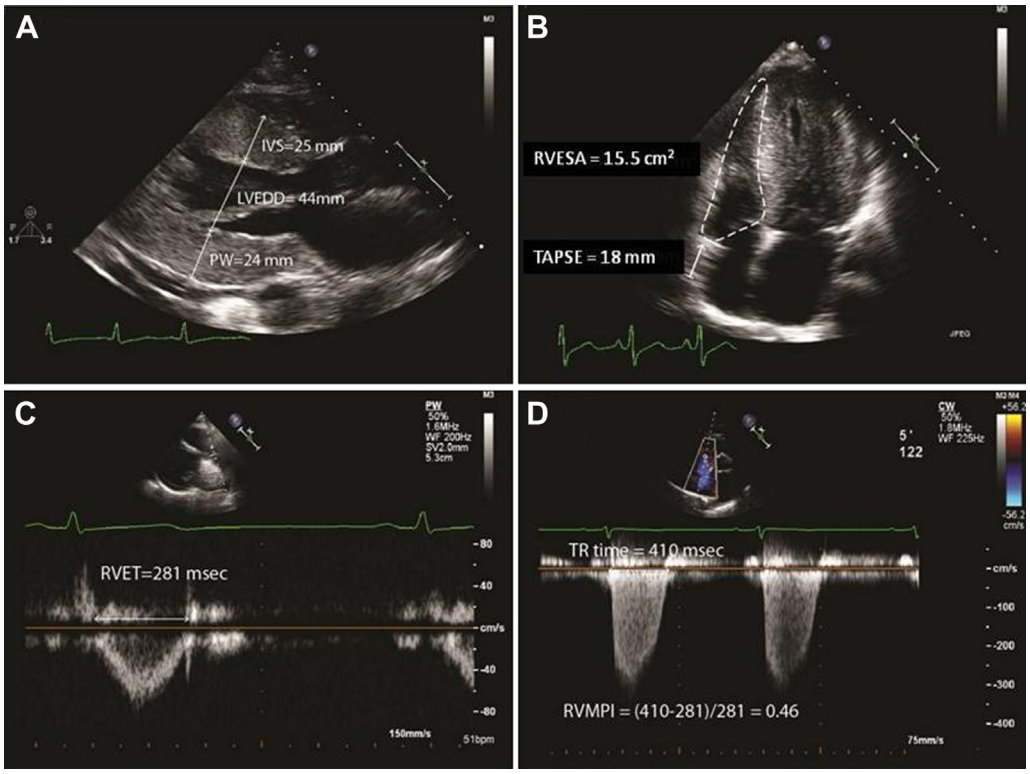

Figure 1. Echocardiographic evaluation. *(A)* Parasternal long-axis view and 2-dimensional measurement of LV end-diastolic diameter and walls. *(B)* Apical 4-chamber view and assessment of RV end-systolic area (RVESA) and TAPSE. *(C)* Doppler analysis of flow at the RV outflow tract with assessment of RV ejection time (RVET). *(D)* Continuous wave Doppler analysis of tricuspid regurgitation (TR) time and RVMPI.

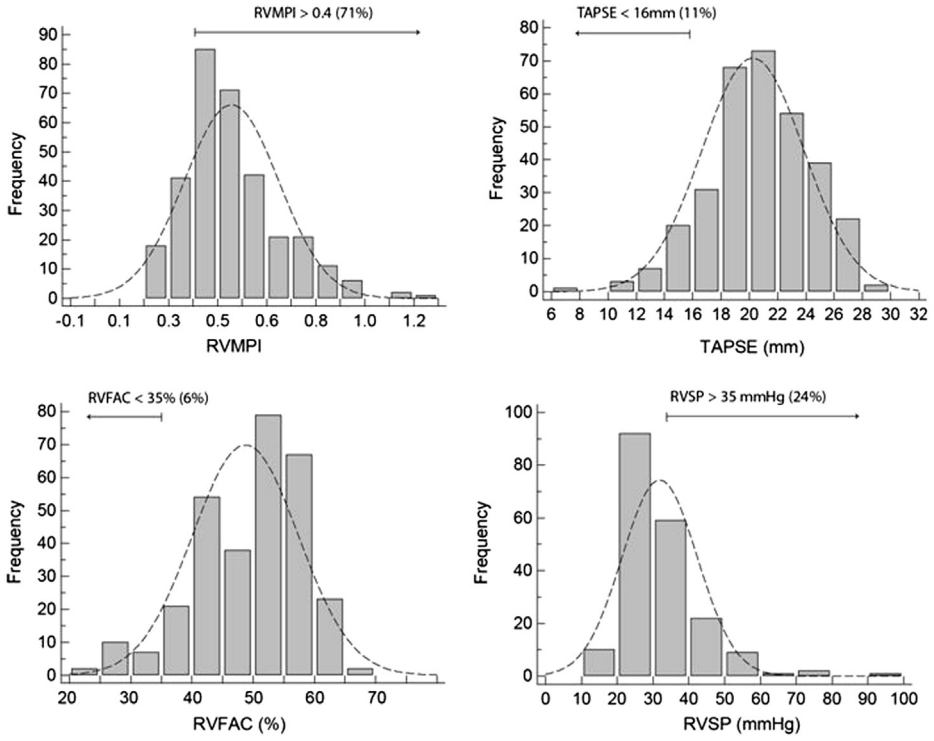

Figure 2. RV function and RV systolic pressure in HC and percentage of abnormal findings. RVFAC = RV fractional area change; RVSP = RV systolic pressure.