Mid-ventricular paradoxical dynamic obstruction in a right-sided secondary hypertrophic cardiomyopathy

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Isolated right ventricular (RV) hypertrophy is a common diagnosis at echocardiography. Mid-ventricular obstructive involvement represents however a really unusual finding. Currently available studies on right-sided hypertrophic cardiomyopathy, regardless of whether they are associated with RV outflow tract obstruction, are usually limited to single cases. Besides, there is still no agreement about the clinical relevance and management of RV hypertrophy.

In the present study the authors describe the echocardiographic findings of a patient presenting with RV hypertrophy in whom a rare coexistence of both fixed RV outflow tract and intraventricular dynamic obstruction was observed. Some functional effects of verapamil treatment are also discussed.

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Introduction

Right ventricular outflow tract (RVOT) obstruction has been described as being consequent to pulmonary valve disease or, less frequently, as a localized dynamic process in patients with right-sided hypertrophic cardiomyopathy (HCM)[1-4].

Various degrees of right ventricular (RV) hypertrophy have been demonstrated to be associated with left ventricular HCM. However, in the absence of the latter feature, RV HCM as well as the presence of secondary dynamic obstruction are considered as being rather infrequent[5-12].

In this report we present a case in which a non-valvular RVOT obstruction induced RV hypertrophy and an unusual intraventricular dynamic (paradoxical) obstructive involvement.

Case report

A 53-year-old Caucasian man was admitted to the Cardiology Unit of the Messina University Hospital due to the recent onset of atypical chest pain, dyspnea and tachycardia.

The patient's cardiac risk factors included a family history of myocardial infarction (his father) and hypertension (his mother). His medical history included mild hypertension lasting 3 years and he was on a sodium-restricted diet. He did not have any other risk factors.

On admission, physical examination revealed a healthy-appearing male with a blood pressure of 150/85 mmHg, a heart rate of 65 b/min and a breathing rate of 20 cycles/min. A grade 3/6 Levine systolic murmur irradiating to the apex but not to the carotid arteries could be heard at the upper sternal borders.

Standard ECG showed normal sinus rhythm, incomplete right bundle branch block and widespread unspecific ST-T segment abnormalities. Left pulmonary artery branch enlargement was suspected at chest X-ray. All serum tests, including D-peptides and fibrin degradation products, were normal. A small hypoperfusive apical area in the left lung was disclosed at ^{99m}technetium-macroaggregated albumin scintigraphy. Chest computed tomographic scan confirmed the isolated left pulmonary artery aneurysm (Θ = 55 mm), but excluded any acute parenchymal perfusion defect.

Transthoracic echocardiography, performed as recommended by the American Society of Echocardiography criteria[13], revealed a normal cardiac chamber size and a preserved ventricular systolic function (Table I). The interventricular septum, right lateral wall and moderator band were found to be thickened and a diagnosis of RV hy-

Table I. Main Doppler echocardiographic data measured at the time of admission of the patient.

LV diastolic diameter (M-mode measurement) (mm)	48
IVS diastolic thickness (mm)	11
Posterior wall diastolic thickness (mm)	8
LV mass (g)	185
LV mass index (g/m^2)	149
LV diastolic volume (SR method, apical 4-chamber) (ml)	75
LV systolic volume (SR method, apical 4-chamber) (ml)	27
LV ejection fraction (%)	64
RV diastolic volume (SR method, apical 4-chamber) (ml)	22
RV systolic volume (SR method, apical 4-chamber) (ml)	9
RV ejection fraction (%)	59
RV anterior wall (mm)	9
RV lateral wall (mm)	10
RV moderator band (mm)	10
Mitral valve E/A ratio	0.78
Mitral E-wave deceleration time (ms)	196
LA systolic area (apical 4-chamber) (mm^2)	1400
RA systolic area (apical 4-chamber) (mm^2)	1200
LV outflow tract peak gradient (mmHg)	7.3
RV outflow tract peak gradient (mmHg)	68.2
Tricuspid valve E/A ratio	0.67
Tricuspid E-wave acceleration time (ms)	65
Tricuspid E-wave acceleration rate (cm/s^2)	1210
Tricuspid E-wave deceleration time (ms)	180
Tricuspid valve regurgitation peak velocity (cm/s)	250
Systolic pulmonary artery pressure (mmHg)	30

IVS = interventricular septum; LA = left atrial; LV = left ventricular; RA = right atrial; RV = right ventricular; SR = Simpson rule (single-plane).

pertrophy was made on the basis of the criteria of McKenna et al.[14]. The pulmonary root diameter was 30 mm and the valve leaflets' excursion only mildly reduced.

In the RVOT a partially occluding thin membrane was identified a few millimeters below the pulmonary valve (Fig. 1). Color-Doppler aliasing across the left side of the bundle was shown and the post-stenotic jet (maximum peak velocity at continuous wave Doppler 413 cm/s; Fig. 2A) appeared to be directed precisely towards the enlarged pulmonary artery branch. No relevant mitral or tricuspid valve regurgitation was found. Moreover, a paradoxical (from the apex to the tricuspid valve) systolic jet, determining a systolic gradient of 44.4 mmHg (Fig. 2C), was observed within the RV chamber (Fig. 3).

The diastolic RV filling pattern was as follows: E/A ratio inversion (E-wave peak velocity 70 cm/s, A-wave peak velocity 100 cm/s), rapid E-wave acceleration time (mean value over 5 cycles 65 ms), E-wave acceleration rate (mean value 1210 cm/s^2), and normal deceleration time (mean value 180 ms) (Fig. 2E). Moreover, tissue Doppler imaging revealed that the RV myocardial relaxation time was prolonged (140 ms).

On these grounds, a diagnosis of secondary RV hypertrophy associated with unusual mid-ventricular obstruction was made, and the patient was prescribed verapamil 240 mg daily.

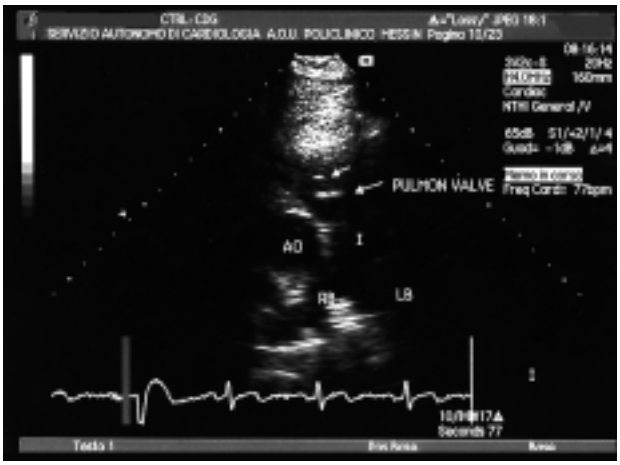

Figure 1. *Standard echocardiographic short-axis view showing the presence of an abnormal bundle* (short arrow) *just below the pulmonary valve leaflets* (long arrow). *Note the asymmetry of the pulmonary artery branches (left > right). Two-dimensional image settings: 4 MHz transducer frequency in harmonic imaging; total gain 65 dB; gray scale – 1dB. AO = transverse aortic root; LB = left pulmonary artery branch; RB = right pulmonary artery branch.*

At 10 weeks of follow-up the patient's clinical conditions as well as the RV diastolic function (E/A ratio ≥ 1; E-wave acceleration time 90 ms; E-wave acceleration rate 845 cm/s^2) improved; the mid-ventricular obstructive gradient decreased to 22 mmHg, but the RV relaxation time and the RVOT gradient were only slightly improved by treatment (Fig. 2).

Discussion

To date, the real clinical impact of isolated RV hypertrophy has not been well established. In daily clinical practice. In fact, the association of mild RV concentric hypertrophy with left-sided HCM is not an unusual finding[1-3,5].

In the present report we describe a case of secondary RV HCM, the clinical interest of which grounds on the coexistence of fixed RVOT stenosis and uncommon intraventricular dynamic (paradoxical) obstruction. To our knowledge, this is the first study showing this type of RV dynamic obstruction.

The pathophysiological mechanism leading to obstruction likely consisted of an abnormal systolic interaction between the moderator band and the hypertrophic RV lateral wall (Fig. 3, bottom panel). In such circumstances, given the scant information in the literature, we are still too inexperienced to manage such a clinical picture.

Paradoxically, it could also be hypothesized that the obstruction provided a sort of "protective" mechanism by limiting the transmission of the systolic pressure into the RV apical region.

Besides, the increased mean right atrial pressure was thought to be a possible consequence of the RV

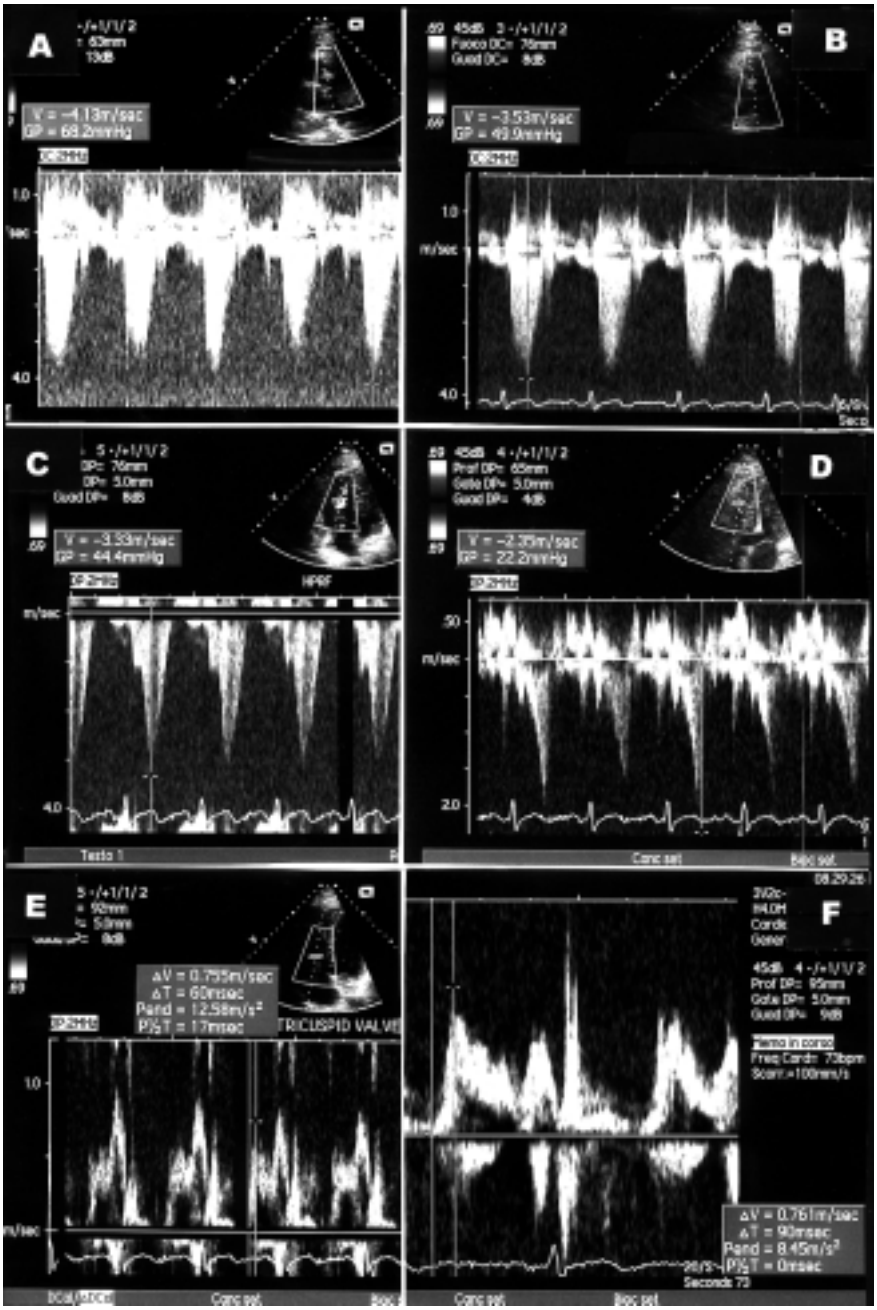

Figure 2. *Flow velocity measurements at Doppler echocardiography before* (right panels) *and after* (left panels) *treatment.* A *and* B: *continuous wave Doppler assessment of the pulmonary peak gradient (at baseline 68.2 mmHg; at follow-up 49.9 mmHg).* C *and* D: *intraventricular obstructive dynamic gradient measured at high pulse repetition frequency pulsed wave Doppler (at baseline 44.4 mmHg; at follow-up 22.2 mmHg).* E *and* F: *right ventricular filling pattern at pulsed wave Doppler sampling; one out of the five values of the E-wave deceleration rate is displayed (at baseline 1258 cm/s^2; at follow-up 845 cm/s^2). Echo-Doppler settings: 4 MHz transducer frequency in harmonic imaging; Doppler gain 13 dB* (A)*, 8 dB* (B, C, E)*, 4 dB* (D) *and 9 dB* (F)*; Doppler sampling depth 76 mm* (C)*, 65 mm* (D)*, 92 mm* (E) *and 95 mm* (F)*; Doppler spectrum recording at 50 mm/s, with the exception of* panel F *(100 mm/s). GP = peak gradient; Pend = E-wave deceleration rate.*

outflow obstruction and diastolic dysfunction. As reported in recent studies[15,16], this observation was also supported by the prolonged myocardial relaxation time at tissue Doppler imaging and by the E-wave acceleration rate. The favorable clinical and functional response to verapamil treatment probably increased the chances of an improved treatment of both the systolic and diastolic RV impairment, as already demonstrated in patients with left-sided HCM[17].

In our opinion, the peculiar morphology of the bundle disclosed in the RVOT, determining the high-velocity jet towards the left pulmonary artery branch, should be considered the main determinant of its isolated enlargement. On the other hand, both bundle and pulmonary artery aneurysm might also simply coexist on the basis of a common gene disorder, although there are no decisive data on the pathognomonic pathways[5,18].

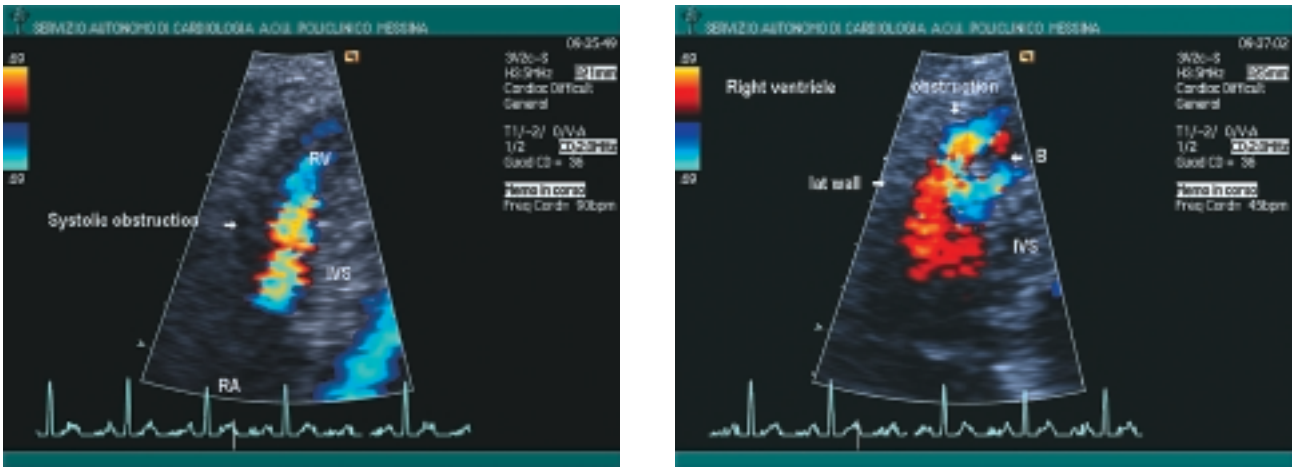

Figure 3. *Echocardiographic 4-chamber apical view in the left extralateral supine decubitus. Panels showing the presence of color Doppler aliasing into the right ventricular chamber. Magnified imaging* (right panel) *demonstrates how the obstruction was related to an abnormal interaction between the moderator band and the lateral wall. Color Doppler imaging settings: 3.5 MHz transducer frequency in harmonic imaging; color Doppler gain 36 dB; velocity range ± 69 cm/s; flow persistence 1/4. IVS = interventricular septum; RA = right atrium; RV = right ventricle.*

Notwithstanding this latter consideration, a major limitation in the present report is the lack of a gene study with histological validation. However, the aim of this study was essentially to examine the functional aspects of this rare cardiac disease.

Since left-sided HCM is considered among the major causes of sudden death in youths, many efforts have been made to identify the earliest markers for stratifying high-risk populations[1,2,14,19-21]. On the contrary, no precise indications on RV HCM are currently available for clinical practice. In such patients medical and surgical strategies have been attempted with variable success in the past years but their appropriateness has not been univocally proven[5-12]. Beta-blockers, for instance, were demonstrated to improve symptoms and exercise capacity[22]. In the present study, the clinical and functional efficacy of verapamil was demonstrated.

Being hypertrophy the final common pathway of various sarcomeric alterations, further considerations must be made bearing in mind the profound phenotypic heterogeneity existing in patients with HCM. Even in the presence of RV hypertrophy, only a larger experience with a proper follow-up will contribute to improve our knowledge about this rare cardiac disease, and hopefully clarify some of the numerous still unanswered clinical questions.

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