

Hypertrophic nonobstructive cardiomyopathy as a cause of severe restrictive physiology

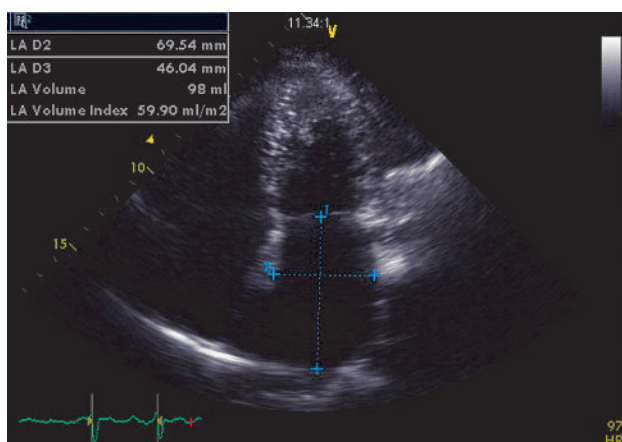
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A 66-year-old woman with a family history of hypertrophic cardiomyopathy (HCM) presented with severe-progressive exertional dyspnoea. She had recently complained of palpitations corresponding to atrial fibrillation on the ECG. A transthoracic echocardiogram (TTE) showed nonobstructive, slightly asymmetrical left ventricular hypertrophy (septum and posterior wall thickness respectively measured at 20 and 15 mm). Left ventricular (LV) cavity and ejection fraction (70%) were normal, but both atria were enlarged (fig. 1A). Diastolic function assessment was suggestive of restrictive physiology (fig. 1B). The patient did not improve despite successful cardioversion and intensive medical therapy with maximally-tolerated doses of beta-blockers and verapamil. She was then referred to the catheterisation laboratory to undergo left and right heart catheterisation and coronary angiography after aggressive forced diuresis with intravenous diuretics. The coronary angiogram did not show significant coronary artery disease (narrowing <30%). Right heart, pulmonary artery (PA), left atrial (via trans-septal approach) and left heart catheterisations were performed

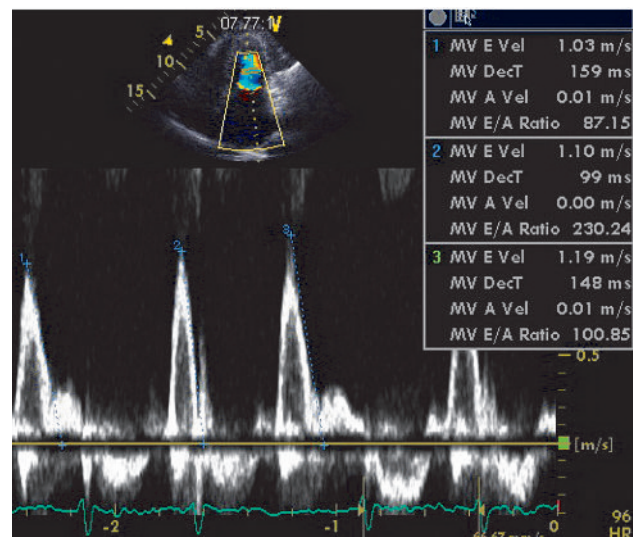
Figure 1

A: Four-chamber view showing a severely dilated left atrium.
B: Deceleration time <150 m/sec (in atrial fibrillation), suggestive of a restrictive physiology.



a

No conflict of interest to disclose.



b

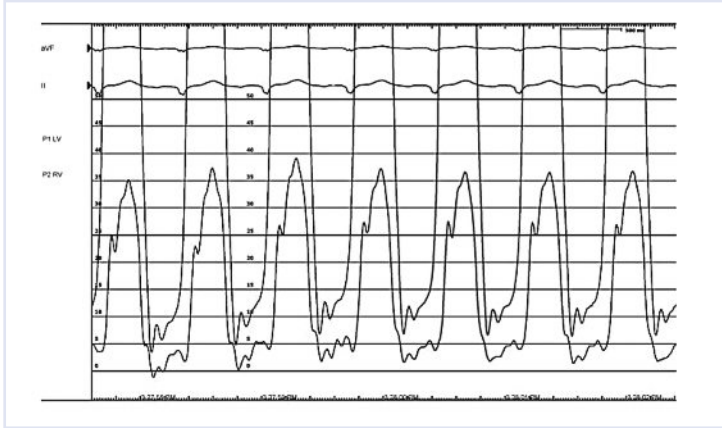
and revealed signs of restrictive physiology without intraventricular dynamic obstruction (fig. 2, table 1). However, the PA pressure was only mildly elevated and the ventricular diastolic pressure showed a subtle dip-plateau pattern (square root sign).

Aggressive treatment with diuretics typically decreases filling pressures (preload) acutely, as confirmed by the low absolute LV and RV end-diastolic pressures, and affects recognition of the typical restrictive pressure tracings. We therefore performed an intravenous fluid challenge with 500 ml of NaCl 0.9% over approximately 10 minutes. The fluid challenge provoked an increase in systolic RV pressure from 36 mm Hg to 57 mm Hg (fig. 3), confirming a significant preload increase (probably more representative of this patient's usual clinical condition). The typical "M or W" pattern of restriction became more obvious on the RA pressure

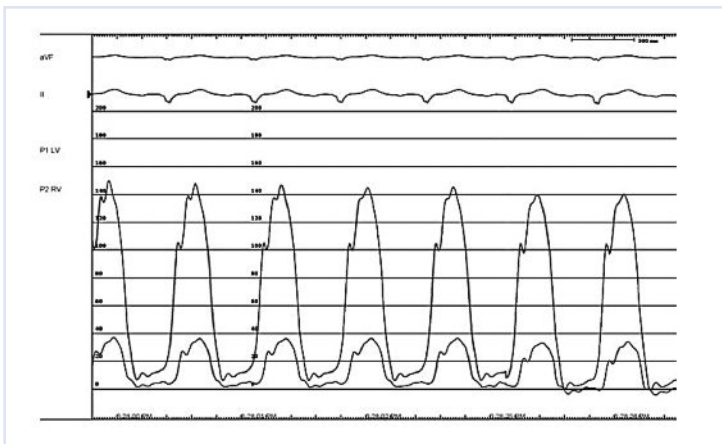
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Figure 2A

RV and LV pressure tracing (50 mm Hg scale). RV systolic pressure / RV end-diastolic pressure is greater than 3 (36/4). LV end-diastolic pressure (13 mm Hg) is greater than RV end-diastolic pressure (4 mm Hg) by more than 5 mm Hg at rest. Systolic RV pressure usually exceeds 50 mm Hg. In this case, a value of 36 mm Hg was measured after aggressive forced diuresis (and before fluid challenge). The ventricular diastolic pressure showed subtle dip-plateau pattern (square root sign).

**Figure 2B**

RV and LV pressure tracing (200 mm Hg scale).



tracing (prominent x and y descent) (fig. 4), as did the ventricular dip-plateau pattern (square root sign). These findings were interpreted as suggestive of a severe restrictive physiology (in addition to the concomitant PA pressure value >50 mm Hg, elevated LV end-diastolic pressure and ventricular diastolic pressures differential >5 mm Hg). This patient did not undergo endomyocardial biopsy or MRI in view of the documented familial history of HCM, absence of clinical findings suggestive of other diagnoses, and typical echocardiographic findings. We did however perform genetic testing for HCM.

HCM is typically classified in the subgroup of non-infiltrative restrictive cardiomyopathy [1]. Most patients with HCM exhibit significant abnormalities of diastolic function at rest and under stress, even in the absence of an intraventricular pressure gradient [2]. These abnormalities of global diastolic filling are largely independent of the extent and distribution of myocardial hypertrophy [3]. HCM may cause abnormal distensibility of the ventricle due to fibrosis or cellular disorganisation. Nonobstructive HCM often presents with the clinical manifestation of restrictive cardiomyopathy, and is also associated with atrial fibrillation.

Finally, we believe it is crucial to determine whether the baseline filling pressures are artificially low, as is often the case following aggressive diuresis, since this situation may mask the typical haemodynamic patterns of restriction. This case, with demonstrative haemodynamic tracings for restrictive cardiomyopathy, clearly shows the effect of fluid challenge, which is often underused in catheterisation laboratories, in a physiology of this kind.

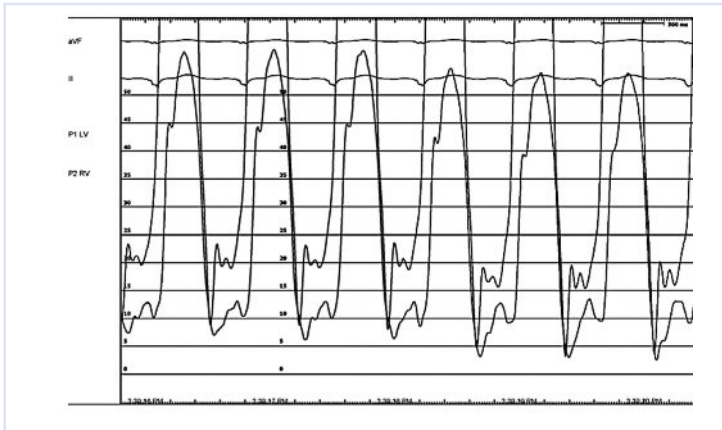
Table 1

Haemodynamic criteria for restrictive cardiomyopathy and constrictive pericarditis [4].

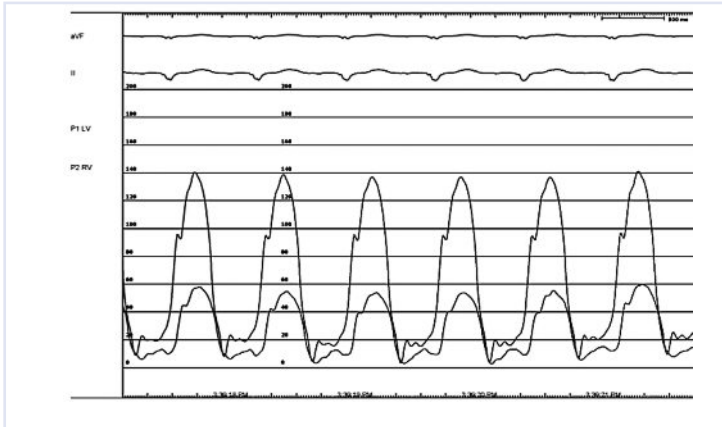
Restrictive cardiomyopathy	Constrictive pericarditis
RVDP plateau <1/3 RVSP peak	RVDP plateau >1/3 RVSP peak
RVSP/RVDP >3	RVSP/RVDP <3
LVEDP >RVEDP (>5 mm Hg at rest) ↑ by fluid challenge, exercise and Valsalva manoeuvre	LVEDP = RVEDP No effect of these manoeuvres
Normal respiratory variation in left-right pressures (no ventricular interdependence)	Exaggerated respiratory variation in left-right pressures (ventricular interdependence)
	Increase in systolic area index (ratio of RV area to LV area in inspiration versus expiration) ⁵
RVSP typically >50 mm Hg	RVSP moderately elevated (typically <40 mm Hg)
Prominent y descent in venous pressure: variable	Prominent y descent in venous pressure
LV: left ventricle, RV: right ventricle, RVDP: right ventricular diastolic pressure, RVSP: right ventricular systolic pressure, RVEDP: right ventricular end-diastolic pressure, LVEDP: left ventricular end-diastolic pressure	

Figure 3A

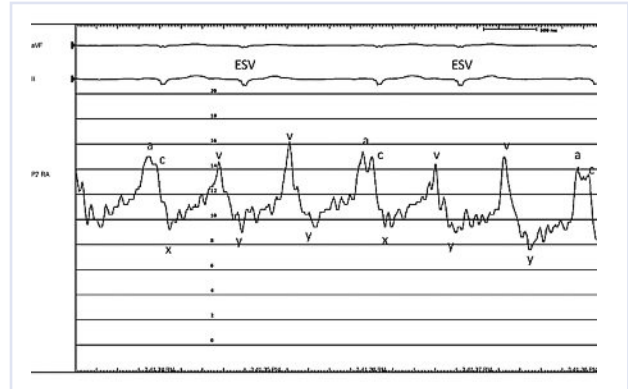
RV and LV pressure tracing (50 mm Hg scale) after fluid challenge. Systolic RV pressure increased from 36 mm Hg to 57 mm Hg. RV systolic pressure / RV end-diastolic pressure is greater than 3 (57/12).

**Figure 3B**

RV and LV pressure tracing (200 mm Hg scale) after fluid challenge.

**Figure 4**

Right atrial (RA) pressure after fluid challenge. The mean RA pressure is elevated (11 mm Hg). The atrial pressure tracing shows a prominent y descent followed by a rapid rise and plateau. The X descent is also rapid. This combination results in the characteristic M waveform in the atrial pressure tracing, also found in the context of constrictive pericarditis.



References

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