

Paradoxical systolic and diastolic flow abnormalities in hypertrophic cardiomyopathy with mid-cavity systolic obstruction

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Abstract

Systolic obstruction of the left ventricular outflow tract, either at rest or during provocation, is a frequent finding in patients with hypertrophic cardiomyopathy. Mid-cavity obstruction is uncommon, and intraventricular diastolic pressure gradients in association with either mid-cavity or apical hypertrophy are reported only rarely. We describe a patient with hypertrophic cardiomyopathy with evidence of systolic mid-cavity obstruction, and with complex diastolic paradoxical flow abnormalities within the left ventricular cavity detected by colour and pulsed-wave Doppler. Contrast echocardiography confirmed the presence of diastolic and systolic obstruction at the mid-ventricular level, with evidence of an apical cavity sequestered from the main body of the left ventricle during systolic mid-cavity obliteration. (Cardiol J 2011; 18, 3: 314–317)

Key words: hypertrophic cardiomyopathy, mid-cavity obstruction, left ventricular hypertrophy, Doppler echocardiography

Case report

A 70 year-old woman presented with an abnormal electrocardiogram which had been detected during pre-operative assessment for cholecystectomy. She had no significant past medical history, and blood pressure was normal. She gave a history of exertional chest pain and breathlessness. Coronary angiography demonstrated no significant coronary artery disease. However, left ventricular (LV) angiography showed a spade-like appearance of the LV cavity suggestive of possible apical hypertrophic cardiomyopathy. She was then referred for detailed transthoracic echocardiography.

Standard transthoracic images confirmed severe LV hypertrophy predominantly affecting the septum, and the anterior and lateral walls as well as the papillary muscle. End-diastolic septal thick-

ness was 2.1 cm. Left ventricular systolic function was normal. Contrast echocardiography (Sonovue, Bracco Imaging) confirmed hypertrophic changes predominantly affecting the mid-cavity, with systolic mid-cavity obliteration, and relative sparing of the apex which became sequestered from the remainder of the cavity during systole. The mid-cavity region of the LV appeared narrowed even during diastole (Fig. 1).

Mild diastolic impairment was evidenced by reversal of the E:A waves on both trans-mitral Doppler flow and mitral annular tissue Doppler velocity profiles, producing a borderline E:E' ratio of 10 (Fig. 2). Systolic anterior motion of the mitral valve was not present, and there was no significant mitral regurgitation. Detailed Doppler examination confirmed no evidence of LV outflow obstruction. However, colour flow Doppler confirmed turbulent

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Received: 20.01.2010

Accepted: 11.05.2010

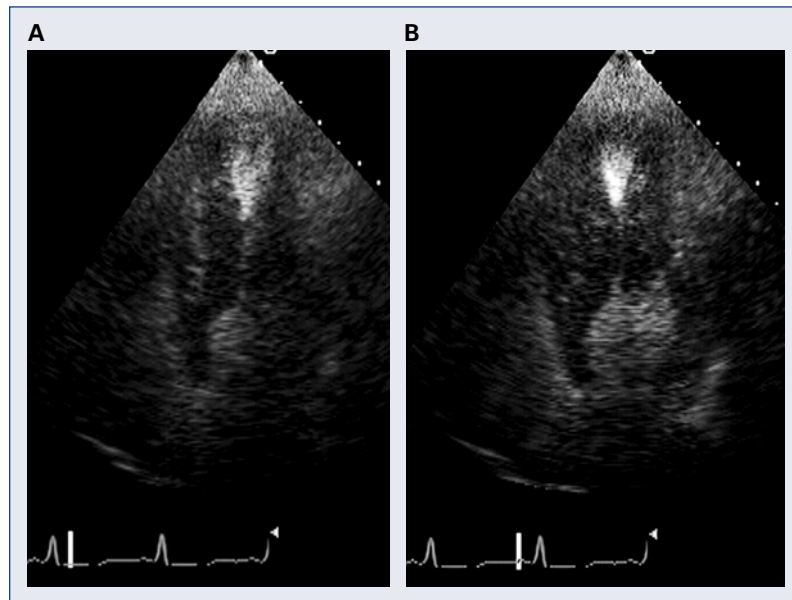


Figure 1. Contrast enhanced left ventricular cavity opacification, demonstrating hypertrophic changes at the mid-cavity level of the left ventricle, causing obliteration of the mid-cavity during systole (A), and narrowing of the mid-cavity during diastole (B).

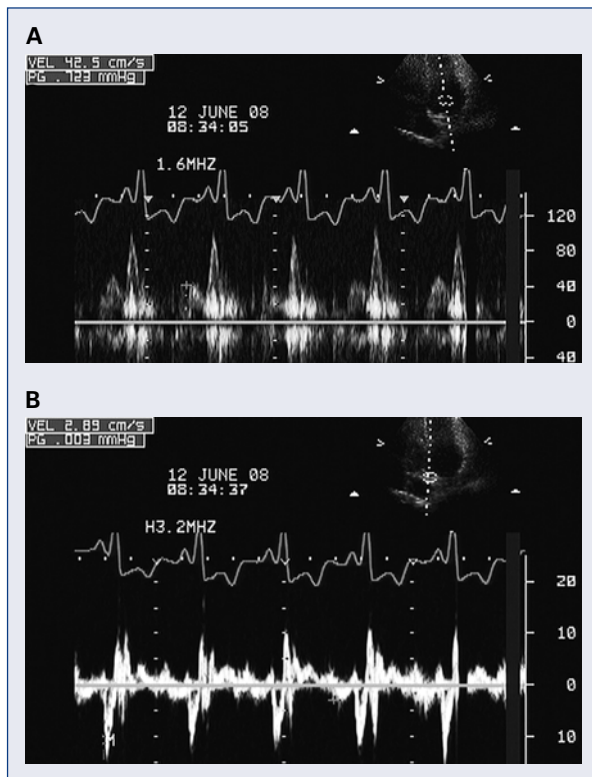


Figure 2. Pulsed wave Doppler trace of mitral valve inflow velocities (A) demonstrating reversal of normal E:A velocity ratio. Tissue Doppler trace of the septal mitral annular velocity (B), also showing reversal of the normal E:A ratio. The calculated E:E' ratio was 10.

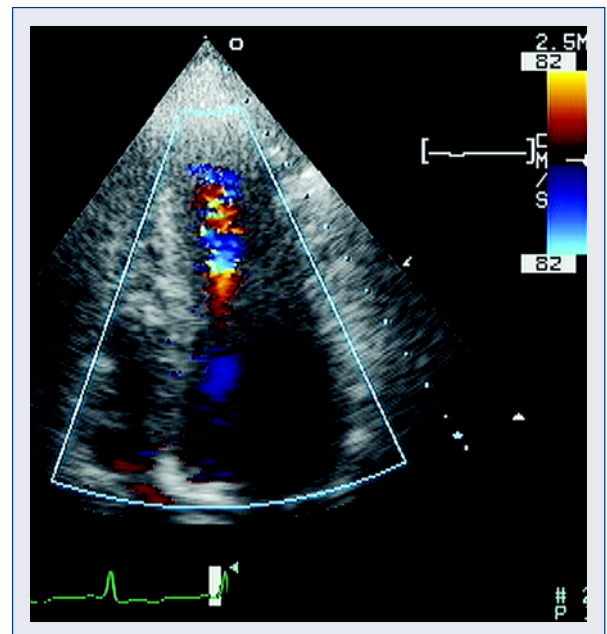


Figure 3. Colour flow Doppler showing evidence of high velocity turbulent flow within the mid-cavity of the left ventricle during diastole.

mid-cavity systolic and diastolic flow (Fig. 3). Pulsed-wave and continuous wave Doppler analysis detected complex abnormalities of systolic and diastolic flow within the LV cavity between the base and apex of the heart. Systolic flow from the apex

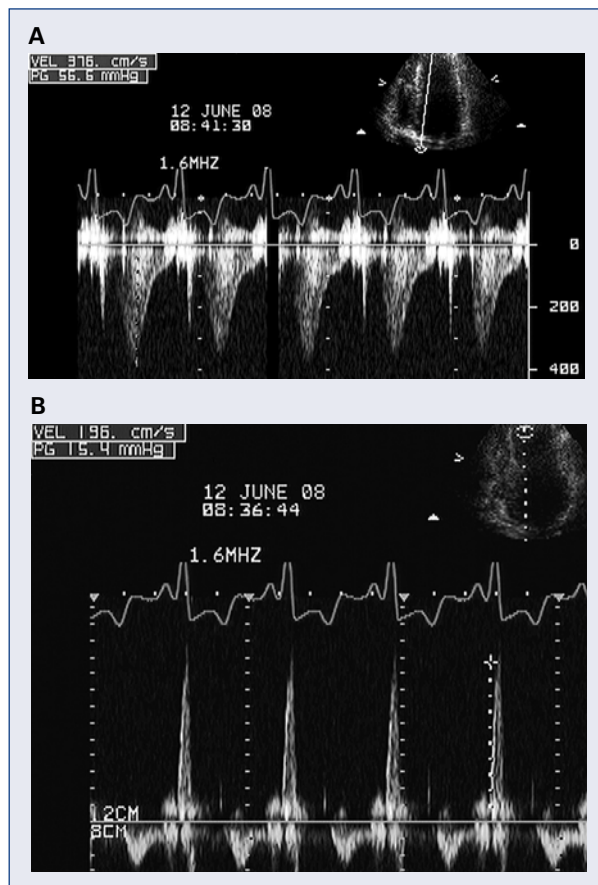


Figure 4A, B. Continuous wave Doppler tracing from the left ventricular cavity demonstrating high velocity early diastolic flow from the apex towards the base of the heart (A). Pulsed wave Doppler from the mid-cavity level showing filling of the apical component of the left ventricular cavity in association with the QRS complex.

was represented by a brief, aborted forward wave. In early diastole, high velocity flow (peak velocity 3.7 m/s) was detected from the apex towards the base of the heart. Using the modified Bernoulli equation, this flow generated a pressure gradient within the cavity from apex to base of 57 mm Hg (Fig. 4A). In late diastole and early systole, a brief phase of high velocity flow (peak velocity 1.9 m/s) was detected towards the cardiac apex (Fig. 4B). This flow appeared to represent filling of the apical cavity through the narrowed mid-cavity region, either via a delayed transmission of the atrial component of ventricular filling (a delay of 100 ms was measured compared to the transmitral A wave), or by retrograde flow into the apical cavity during isovolumic contraction.

The patient was started on beta-blocker therapy, which was subsequently switched to verapamil due to adverse effects. Her symptoms improved

significantly on medical therapy, although echocardiography was unchanged, with a persistent intracavity early diastolic velocity of 3.7 m/s. No further treatment is planned at present.

Discussion

Left ventricular outflow tract obstruction occurs with a reported incidence of approximately 25% in patients with hypertrophic cardiomyopathy at rest, rising to as high as 70% following provocation with exercise [1]. Mid-cavity systolic obstruction is much more uncommon. Intra-cavity diastolic flow abnormalities have been reported on only a few occasions, typically in association with either apical or mid-cavity hypertrophic cardiomyopathy.

This phenomenon was first described in detail in three patients with either mid-cavity or apical hypertrophy, and a muscular ‘tunnel’ connecting the main body of the left ventricle to a hypokinetic apical cavity [2]. Brief early systolic flow from the apical chamber towards the base of the heart was noted, which terminated during systole as the muscular tunnel closes. Early in diastole, paradoxical flow was found to occur from the apex towards the base. This flow is simultaneous with, and in the opposite direction to, mitral inflow. Filling of the apical chamber occurred only late in diastole. These findings were attributed to high pressure in the apical cavity during systole as the mid-cavity obstruction to apical outflow progresses, and persistence of this high apical pressure in early diastole due to delayed apical relaxation.

Similar findings have been confirmed subsequently [3–7], with other reports detailing variations of this process. Complete reversal of normal systolic and diastolic flow patterns has been described [6], with outflow from the apical cavity occurring only in diastole, and inflow only in early systole. This functional separation of the apical region from the remainder of the cavity is attributed to thinning and akinesia of the apical region, rendering the apical cavity passive with no contribution to pressure generation or flow. It is hypothesised that long-standing systolic and diastolic obstruction to flow from the apical chamber may lead to increased wall stress at the apex, resulting in ischemia, and eventual myocardial necrosis and infarction. Evidence of apical aneurysm formation has been reported in association with a fixed, non-perfused defect on myocardial perfusion scintigraphy [7]. There is also evidence that this group of patients is at higher risk of systemic embolism, ventricular

arrhythmias, and defects on myocardial perfusion scintigraphy than those with evidence of mid-cavity obstruction but no paradoxical blood flow [8].

In our patient, we noted very brief aborted early systolic flow, suggesting that the mid-cavity muscular tunnel is obliterated very early in systole. Paradoxical flow from the apex towards the base begins in early diastole, occurring almost throughout diastole, with a peak pressure gradient from apex to base of 57 mm Hg. Filling of the apical cavity occurs consequent with the QRS complex, consistent with either a delayed transmission of the mitral A wave into the apical cavity, or more likely with systolic filling during isovolumetric contraction. These findings suggest that mid-cavity obstruction is persistent throughout the cardiac cycle, and this is confirmed by the two-dimensional appearance of the mid-cavity when delineated using intravenous contrast in our case.

Following treatment with beta-blockade, our patient's symptoms improved significantly, although there is very little data to guide treatment in this situation. Barbier and Bartorelli [6] reported improvement in the mitral inflow Doppler characteristics, but not in the mid-cavity Doppler parameters following 12 months' treatment with verapamil in a single case. Yamamoto et al. [9] demonstrated reduction in the velocity of both the paradoxical jet and the peak systolic velocity at the mid-cavity narrowing in a single patient following administration of propranolol.

Conclusions

Mid-cavity obstruction is a relatively uncommon finding in patients with hypertrophic cardiomyopathy, and abnormalities of diastolic flow with paradoxically directed jets appear to be rare. Identifying these abnormalities is important, as they may be a cause of both symptoms and potentially longer term complications. Echocardiography in this group

of patients requires a thorough assessment of intracavity hemodynamics using colour flow Doppler, as well as pulsed-wave and continuous-wave Doppler, in conjunction with 2D appearances, augmented by contrast opacification if images are suboptimal.

Acknowledgements

The authors do not report any conflict of interest regarding this work.

References

1. Maron MS, Olivitto I, Zenovich AG et al. Hypertrophic cardiomyopathy is predominantly a disease of left ventricular outflow tract obstruction. *Circulation*, 2006; 114: 2232–2239.
2. Zoghbi WA, Haichin RN, Quinones MA. Mid-cavity obstruction in apical hypertrophy: Doppler evidence of diastolic intraventricular gradient with higher apical pressure. *Am Heart J*, 1988; 116: 1469–1474.
3. Keller H, Kageneck VV, Buss J, Stegaru B. Diastolic intraventricular gradient in hypertrophic cardiomyopathy with apical hypertrophy. *Eur Heart J*, 1993; 14: 425–427.
4. Ioannides MA, Konis P, Georgiou G, Nicolaidis E. Diastolic gradient in hypertrophic cardiomyopathy of the apical type. *Eur J Echocardiography*, 2004; 5: 79–81.
5. Mixon TA, Giebel DW. Abnormal diastolic flow demonstrated by color M-mode echocardiography in hypertrophic cardiomyopathy with mid-ventricular cavity obliteration. *Echocardiography*, 2004; 21: 49–53.
6. Barbier P, Bartorelli A. Doppler evidence of abnormal intracavity systolic and diastolic flow in hypertrophic cardiomyopathy with midventricular obstruction. *Am Heart J*, 1993; 126: 483–487.
7. Shen AYJ, Gupta N, Kapoor G. Severe intraventricular diastolic gradient due to hypertrophic cardiomyopathy and systolic left ventricular midcavitary obstruction. *Echocardiography*, 2005; 22: 47–48.
8. Nakamura T, Matsubara K, Furukawa K et al. Diastolic paradoxical jet flow in patients with hypertrophic cardiomyopathy: Evidence of concealed apical asynergy with cavity obliteration. *J Am Coll Cardiol*, 1992; 19: 516–524.
9. Yamamoto K, Masuyama T, Nishikawa N, Tanouchi J, Hori M. Variability of diastolic paradoxical jet flow in a patient with hypertrophic cardiomyopathy with midventricular obstruction: Dissociation between systolic and diastolic intracavitary flow dynamics. *J Am Soc Echocardiogr*, 1993; 6: 628–630.