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# A 59-Year-Old Woman with Shortness of Breath: Hypertonic Cardiomyopathy Masquerading as Critical Aortic Stenosis in a Patient with Bicuspid Aortic Valve

Avaneesh Jakkoju, MD, Mehnaz Rahman, MD, Murtuza Ali, MD,  
Fred A. Lopez, MD

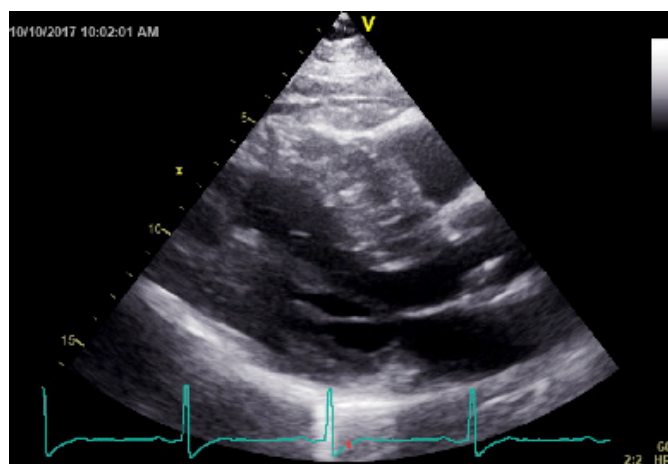
**Hypertrophic cardiomyopathy (HCM) with varied genotypes and phenotypes is the most common genetic cardiovascular disease.<sup>1</sup> Described 50 years ago, at which time it was thought to be fatal and without any treatment options, our understanding and management of HCM has significantly improved. A significant number of patients with HCM are often undiagnosed until later in life, as it often does not have any clinical manifestations.<sup>2</sup> Left ventricular hypertrophy (LVH) often poses a diagnostic challenge as to whether it is from primary genetic abnormality versus secondary to systemic hypertension or aortic stenosis. We present a case of HCM, which was diagnostically challenging because of a coexisting bicuspid aortic valve (BAV).**

## CASE PRESENTATION

A 59-year-old woman with past medical history of hypertension presented to her primary care physician's office with a three-month history of progressive dyspnea on exertion, effort intolerance and paroxysms of near syncope. Work up included an echocardiogram that showed a BAV with severe LVH, and near cavity obliteration with increased velocity across left ventricular outflow tract (LVOT) and aortic valve (AV). Peak velocities across the LVOT and AV were 5 m/sec (normal: <2 m/sec) with a peak gradient of 100 mmHg (normal: <16 mmHg) and mean gradient of 60 mmHg (normal: <5 mmHg). The patient was referred to the cardiology clinic with suspected severe aortic stenosis.

Upon initial presentation to the clinic, she continued to complain of shortness of breath with minimal exertion, fatigue, effort intolerance, and near syncope. Physical examination was significant for a blood pressure of 90/75 mmHg. Cardiac auscultation revealed an early peaking systolic murmur without respiratory variation, and a preserved S2 heart sound. A repeat transthoracic echocardiogram (TTE) showed significant LVH with septal thickness of 3.03 cm (normal: 0.6–1.0 cm) (Figure 1) causing dynamic outflow obstruction, bicuspid aortic valve (Figure 2) with fused non-coronary and left coronary cusps and good leaflet motion. Using the continuity equation, her aortic

valve area was calculated as 1.8 cm<sup>2</sup> (normal: 2.5–4.5 cm<sup>2</sup>). With Valsalva maneuver the LVOT velocity increased from 1.5 m/sec to 4 m/sec (Figure 3). A diagnosis of hypertrophic cardiomyopathy (HCM) with intracavitary dynamic obstruction and concomitant bicuspid aortic valve without aortic stenosis was made.



**Figure 1. Parasternal long axis view on TTE showing asymmetric septal hypertrophy**

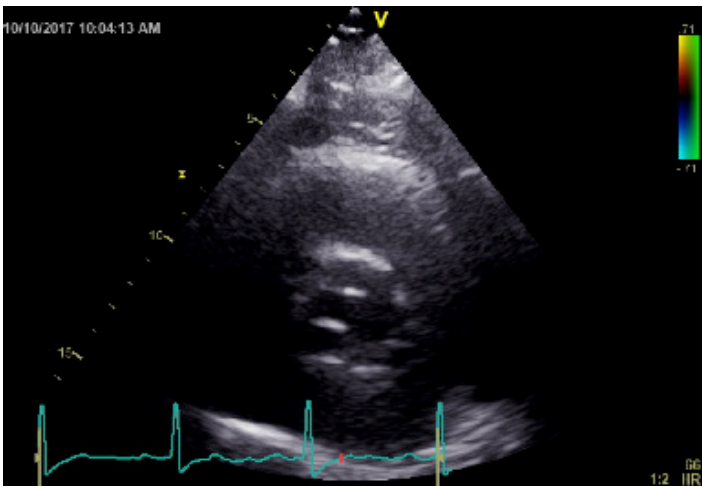


Figure 2. Parasternal short axis view on the echocardiogram shows bicuspid aortic valve

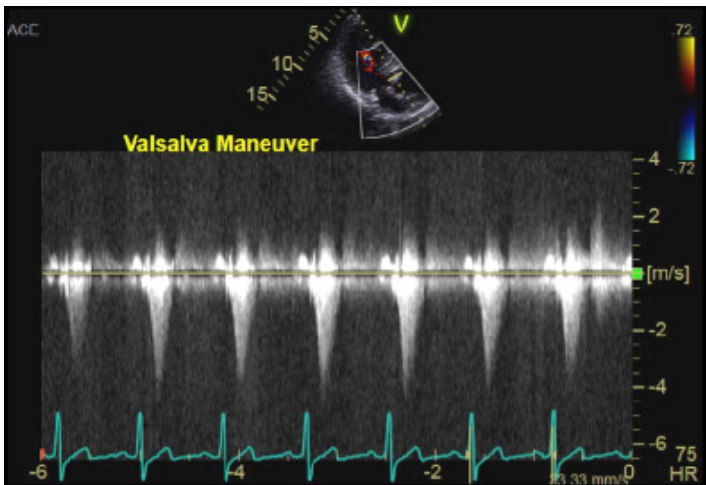


Figure 3. Increase in gradient across LVOT with Valsalva maneuver

## DISCUSSION

Hypertrophic cardiomyopathy (HCM) is inherited in an autosomal dominant pattern with equal preponderance for men and women. However, women are more often undiagnosed at an early stage and often present later in life with advanced heart failure symptoms.<sup>3</sup> The disease is characterized by a thickened left ventricle in the absence of other precipitating conditions such as systemic hypertension and aortic stenosis. A significant number of patients have normal life expectancy and do not manifest any signs or symptoms. About 10-15% patients develop progressive heart failure symptoms NYHA class III or higher.<sup>3</sup> The incidence of progressive heart failure is dependent on coexisting atrial fibrillation, the degree of diastolic dysfunction, and dynamic left ventricle outflow obstruction. HCM is the most common cause of sudden cardiac death in young patients and can cause symptoms of heart failure at any age.<sup>4, 5, 6</sup> Timely diagnosis and

appropriate management is crucial to the management of these patients.

A transthoracic echo is a very useful initial diagnostic modality. 2D echocardiographic imaging can demonstrate a thick sigmoid-shaped septum with hyperdynamic LV and a small cavity often described as "banana shaped." Septal hypertrophy is defined as asymmetric when the septal thickness is  $\geq 1.6$  times the thickness of the posterior wall.<sup>7,8</sup> Elevated flow velocity across the LV outflow tract that peaks in the late systole is typically noted. Most patients with HCM have a long anterior mitral valve leaflet which due to flow drag forces and suctioning effect (Venturi effect) moves anteriorly in systole increasing the obstruction to LV outflow.<sup>8</sup> This effect can also cause a posteriorly-directed mitral regurgitation jet. Continuous wave (CW) Doppler measurements can show the gradient of obstruction across the LVOT. These values have shown good correlation with invasive measurements.<sup>9</sup> CW Doppler measures peak blood acceleration along its cursor line.<sup>8</sup> However the left ventricular outflow tract and aortic valve are very close to each other and when measured with CW Doppler these gradients can falsely be attributed to AV gradients. Pulse Wave (PW) Doppler measures peak blood acceleration at a particular point where the cursor is placed. Given the high intra-cardiac velocities, PW Doppler can be used to sequentially interrogate from the LV apex down to the LVOT in order to confirm the anatomical level of obstruction.<sup>8</sup> Outflow gradient across the LVOT is dynamic in patients with HCM. A change in loading conditions can alter the degree of obstruction. In our patient, a Valsalva maneuver increased the velocity across LVOT from 1.5 m/sec-4 m/sec, increasing the gradient from 9 mmHg-64 mmHg.

Diagnosing HCM may often be challenging and it is important to differentiate it from the physiologic form of LVH. If an echocardiogram fails to establish a diagnosis of HCM and if clinical suspicion for HCM is high, Cardiac Magnetic Resonance (CMR) imaging may help to establish the diagnosis. Increased ventricular mass index and asymmetric septal hypertrophy can be seen on CMR.<sup>10</sup> An end diastolic wall thickness to LV cavity volumeratio of less than 0.15 mm/ml/m<sup>2</sup> suggests the physiologic form of LVH.<sup>11</sup> Late gadolinium enhancement on CMR has been shown to be associated with microvascular dysfunction, sudden cardiac death and progressive LV dilatation.<sup>10</sup>

## CONCLUSION

The prevalence of BAV in patients with HCM is around 0.9%, which is similar to the general population.<sup>12</sup> Although the concurrent presentation is low, when present, BAV may be a confounding factor that can lead to unwarranted aortic valve replacement surgery. Thorough diagnostic evaluation and an understanding of their hemodynamic interplay is paramount in guiding therapy.

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