A Case of a Thick Heart: Hypertrophic Obstructive Cardiomyopathy

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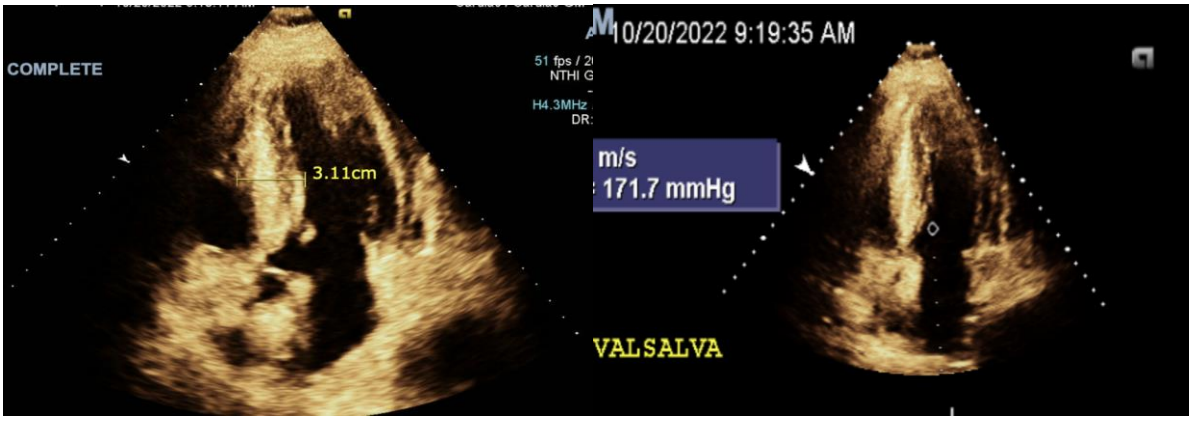
Background:

Hypertrophic cardiomyopathy is a genetic disorder characterized by left ventricular hypertrophy unexplained by secondary causes and a nondilated left ventricle with preserved or increased ejection fraction. Prevalence has been estimated at 0.16% to 0.29% in the general adult population. In the United States alone, there are fewer than 100 deaths per year due to HOCM at a rate of 1:220,000 athletes. We report newly diagnosed Hypertrophic Obstructive Cardiomyopathy.

Case Presentation:

40-year-old gentleman with past medical history of CVA with residual left sided weakness, polysubstance abuse presented to the emergency department with the chief complaint of shortness of breath and waxing and waning substernal chest pain of 2 days duration. He also reported associated orthopnea and paroxysmal nocturnal dyspnea. Physical examination revealed Grade 2/6 systolic ejection murmur at the left sternal border which improved with squatting of the patient. Family history of sudden cardiac death. UDS was positive for Cocaine. Bedside POCUS was done, Hypertrophic Obstructive Cardiomyopathy was suspected. 2D echocardiogram showed Left Ventricular EF 65%, Eccentric LVH with septal thickness of 3.1 cm and post wall thickness of 1.9 cm. There was a peak Left ventricular outflow tract gradient of 117 mmHg, which increased to 171 mmHg with Valsalva maneuver. Vasodilators and preload reducing antihypertensives were avoided given the diagnosis of HOCM. We attempted to control the patient's high blood pressure using Verapamil. Betablockers were avoided due to recent use of cocaine to avoid profound hypertension. After careful consideration, we introduced Diltiazem as opposed to Verapamil, since his blood pressure remained in 200s despite using Verapamil 120 mg every 8 hours. ICD placement was recommended by Electrophysiologist due to high risk of sudden cardiac death. The patient left against medical advice on the third day of hospitalization.



Conclusion:

The first line medical treatment for symptomatic HOCM is beta blockers or cardiac selective calcium channel blockers may be used. Mavacamten is a new FDA approved cardiac myosin inhibitor indicated for the treatment of adults with symptomatic NYHA class II-III HOCM to improve functional capacity and symptoms.