**Hypertrophic Cardiomyopathy – an emergency medicine review - Grand Rounds Summary**

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* Hypertrophic Cardiomyopathy (ie. HCM) refers to the genetic condition resulting in abnormal LV thickening or cardiac muscle hypertrophy without chamber dilation as well as the absence of other identifiable cause.
* Classified by obstructive and non-obstructive physiology
* Common condition, with an incidence of 1:500
* Inherited in an autosomal dominant fashion
* Phenotypic expression excessive myosin and binding protein crossbridging, leading to asymmetric muscle hypertrophy
* Pathophysiology can be distilled down to myocardial cell disarray and fibrosis, abnormal coronary vasculature and ischemia, and dynamic LV outflow tract obstruction (LVOTO)
* LV obstruction is due to both septal hypertrophy encroaching into LVOT as well as a phenomenon known as systolic anterior motion of the mitral valve (SAM)
* SAM is exacerbated with increases in the LVOT gradient
* Classically presents as exertional chest pain, shortness of breath, palpitations and presyncope/syncope. Can also present with sudden death.
* Echo is the key to diagnosis. Can also diagnose dynamic obstruction and observe SAM. This is most apparent clinically with the M-Mode function on POCUS.
* Outpatient management includes BB, CBB and disopyramide. There is a small role for ventricular pacing. For patients with refractory symptoms, surgical myomectomies and alcohol ablations are considered. All patients are risk stratified for ICD placement.
* Hemodynamic goals: Reduce rate and contractility, maintain sinus rhythm, maintain afterload, increase/maintain preload
* Be careful with procedural sedation. In patients who require sedation or anesthesia, pick agents with appropriate doses to maintain hemodynamic goals. This is most important in HCM patients with obstructive physiology.
* If a patient with HCM presents in shock and/or pulmonary edema, look for signs of obstruction on ultrasound (ie. SAM). If observed, BB such as Esmolol and afterload improving agents such as phenylephrine should be go-to agents. Maintain sinus rhythm. Improve preload. Avoid PPV.
* In HCM patients who present in VT storm, reduce the sympathetic drive with sedation/anesthesia/analgesia while maintaining hemodynamic goals. Reduce LVOT obstruction with esmolol and phenylephrine, in addition to amiodarone. Avoid epinephrine, even in arrest. Stellate ganglion blocks work well.