

42. Hypertrophic Cardiomyopathy

1. Acquired (most common)
 - a. Hypertension
 - b. Aortic Stenosis
2. Familial HCM
 - a. Mutations in seven different genes found in ~50% of cases
 - b. Most common association: mutations in β -cardiac myosin H chain gene
 - c. Cardiac troponin T, troponin I, and alpha-tropomyosin genes mutations also reported
 - d. In young athletes with sudden death, HCM most common cardiac anomaly on autopsy
3. Apical Hypertrophic Cardiomyopathy
 - a. Yamaguchi's Syndrome - hypertrophy localized to ventricular apex
 - b. Deeply inverted T waves in lateral leads on ECG
 - c. Natural history includes atypical chest pain symptoms, low risk MI
4. HOCM Hypertrophic Obstructive Cardiomyopathy, Bifed Pulse, louder murmur with Valsalva!

Brockenbrough Effect:

Post-PVC contraction results in an increase in LVOT gradient (worsening of obstruction) and therefore no increase in stroke volume and no change in arterial pulse pressure.

Post PAC Augmentation of outflow gradient and murmur

Notice: The "Bifed" pulse

Treatment

- a. Maintain low heart rate to allow time for filling
- b. Drugs with anti-inotropic activity help relax muscle
- c. Reversal of ischemia, which itself can worsen ability of muscle to relax
- d. TRANSCORONARY ABLATION OF SEPTAL HYPERTROPHY (TASH) w/Alcohol.
- e. Implantable cardioverter-defibrillator (ICD) terminate ventricular arrhythmias and are likely have a role in therapy for cases of familial HCM