Hypertrophic Obstructive Cardiomyopathy

HOCM (a.k.a. IHSS) is an autosomal dominant genetic disorder of cardiac sarcomeres of which a subset of patients (~25%) have a dynamic obstruction to left ventricular outflow at rest, while other patients have dynamic outflow obstruction only during exertion and some have no appreciable outflow obstruction (despite being genetically susceptible)

ANESTHETIC CONSIDERATIONS:

- Cardiac dysfunction
  - Dynamic LV outflow obstruction
  - Mitral regurgitation
  - Hypertrophy with diastolic dysfunction and ischemia
  - Arrhythmias / sudden death
  - CPR ineffective??
- Medications
  - Beta-blockers or CCB
  - Anti-arrhythmics
  - Anti-coagulants
  - Diuretics
- Pacemakers or AICD

ANESTHETIC GOALS:

- Overall, goal is to minimize systolic pressure gradient between LV and aorta
- Avoid hypotension (phenylephrine)
  - Profound hypotension in setting of **hypovolemia**, decreased preload / afterload or increased contractility → always keep full, full, full (first treatment for hypotension!!) and keep afterload up, up, up (second treatment for hypotension is pure vasoconstrictor), finally consider decreasing contractility (volatile good for this)
- Contractility: avoid increasing (beta blockers, CCB, halothane etc.)
- Rate: avoid tachycardia
- Rhythm: strict NSR
- Afterload: Maintain
- Preload: FULL
- MVO_2: monitor for ischemia

HISTORY

- Many patients may have minor or no symptoms (many individuals diagnosed during family screening)
- Resting pressure gradient between LV outflow tract and aorta present in only ~25% of patients whereas other patients only have a gradient with exertion
- LVOT Obstruction
  - Dyspnea on exertion (90%)
  - Pre-syncope or syncope
  - Postural light-headedness
- Hypertrophy
  - Chest pain (25-30%)
    - Majority have normal angiogram
    - Commonly precipitated or worsened by heavy meals
  - Diastolic dysfunction
- Arrhythmias
  - Supraventricular Arrhythmias (A-fib, A-flutter)
    - Can proceed to ventricular arrhythmias
  - Ventricular Arrhythmias
    - Ventricular premature beats
    - Non-sustained VT
      - Increased risk for SCD (8.6% per year vs. 1% per year)
- Sudden Cardiac Death
  - Risk factors
    - Syncope and / or family history of SCD
    - LV wall thickness > 30 mm
    - Non-sustained ventricular tachycardia
    - Abnormal blood pressure response with exercise

PHYSICAL

- **GENERAL**
  - May be normal
- **CVS**
  - LV outflow obstruction
    - Harsh crescendo-decrescendo systolic murmur
    - May also reflect MR
    - Worsened with decreased SVR, pre-load:
      - Squatting to upright posture
      - Valsalva maneuver
    - Improved with increased pre-load, increased SVR
      - Handgrips

        - Raising legs
    - May have evidence of diastolic dysfunction
        - S3 or S4
- **RESP**
    - Crackles / wheeze due to pulmonary congestion

INVESTIGATIONS

- **Labs**
    -
- **Imaging**
    - **EKG**
        - Normal EKG is uncommon (only 7%)
        - LVH with strain
        - Prominent abnormal Q waves in inferior and lateral leads
        - P-wave abnormalities
        - LAD
    - **Echocardiography**
        - Hypertrophy:
            - Variable patterns: septal hypertrophy, apical hypertrophy, LV free wall, RV hypertrophy
            - DDx: HTN, Aortic Stenosis, Athlete's Heart, Fabry's disease
        - Resting LVOT obstruction present in only ~25% of patients
        - Mitral regurgitation
            - Obstructive HOCM
                - Almost all patients
                - Secondary to systolic anterior movement of mitral valve (SAM)
            - Non-obstructive HCM
                - 20-30% of patients and normally mild
        - Diastolic Dysfunction
            - Present in most patients
            - A risk factor for death or sustained VT
    - **MIBI**
        - Often abnormal in patients with HOCM:
            - Asymmetric distribution of hypertrophy
            - Microvascular disease
    - **Cardiac MRI**
        - Useful if suboptimal Echo or hypertrophy localized to apex
    - **Catheterization**
        - Assessment of severity of LV obstruction requires estimation of three hemodynamic factors
            - Cardiac output
            - LV filling pressure or left atrial pressure
            - LV outflow tract pressure gradient
        - Performed if Echo inadequate or obstruction complex

OPTIMIZATION

- Need to assess degree of medical optimization from:
    - LVOT obstruction (want to reduce LVOT gradient)
        - Preoperative sedation to prevent anxiety induced sympathetic stimulation
        - Replace any preoperatively **fluid** deficit
        - Is the patient adequately **beta-blocked or CCB**?
        - Is there **any surgical procedure** that could be performed?
            - LV myectomy
                - Patients with obstructive HCM who are severely symptomatic NYHA class III or IV or have recurrent syncope despite pharmacologic therapy are candidates for LV myectomy (i.e. > 50 mmHg at rest)
                - Can eliminate or significantly decrease LVOT obstruction
            - MVR, mitral valve plication
        - Non-surgical approach
            - Alcohol ablation of septal perforators
    - Hypertrophy
        - Ischemia
            - Is an adequate etiology of ischemia been sought and can this be further optimized?
        - Diastolic dysfunction
            - Does the patient require diuretics if significant SOBOE or obvious clinical CHF?
            - Careful with decreased preload causing increased LVOT obstruction
    - Arrhythmias
        - Does this patient require medical management (amiodarone) or pacemaker / AICD prior to procedure?
        - Current 2002 ACC / AHA / NASPE guidelines suggest pacemaker only for HCM patients with sinus node dysfunction or AV block
        - DDD pacing was previously used as alternative to myectomy to improve symptoms – not supported by outcome data (Uptodate)
    - Anticoagulation

        - Patients with paroxysmal or chronic atrial fibrillation are at high risk for thromboembolism - long term anticoagulation
        - May have to reverse, hold or convert to heparin for surgery

ANESTHETIC OPTIONS

- Either general anesthesia or regional anesthesia OK if hemodynamic goals can be met
- Some feel that neuraxial anesthesia relatively contra-indicated due to unfavorable alteration in physiology (i.e. decreased preload, decreased afterload, increased HR)

ANESTHETIC SETUP

- **Drugs**
    - Alpha agonists (e.g. phenylephrine) preferred to maintain SVR
    - Beta-blockers or CCB
    - Anti-arrhythmics (e.g. amiodarone, lidocaine) for immediate treatment of arrhythmias
- **Equipment**
    - Patients should have full hemodynamic monitoring to ensure anesthetic goals are met
        - Arterial line
        - CVP
        - 5 lead EKG
    - Have immediate availability for cardioversion or pacing
        - Pacer pads
        - Consider pacing PAC in high risk patients

MANAGEMENT OF ANESTHESIA

- **Induction**
    - Ensure patient has deep plane of anesthesia prior to airway manipulation
- **Maintenance**
    - Volatiles well tolerated due to myocardial depressant properties although be wary of arrhythmogenic potential
- **Emergence**
    - Avoid sympathetic activation

DISPOSITION & MONITORING

- Good pain control with PCA or epidural (watch for hypotension) to avoid SNS activation and tachycardia
- Major surgery with potential for pain, fluid shifts should be admitted to a monitored High dependency unit
- Intermediate and minor procedures may be discharged to the ward or home

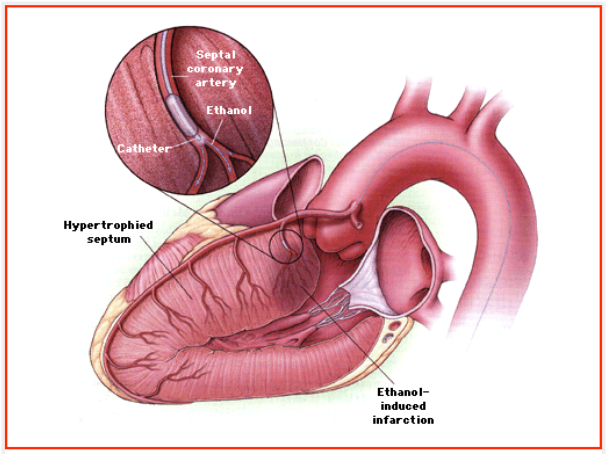
COMPLICATIONS

- Acute LVOT obstruction / SAM
    - Always consider LVOT obstruction as primary cause of hypotension
    - Should be treated with aggressive volume, vasopressors (e.g. phenylephrine) and beta-blocker (esmolol) or CCB (verapamil)
    - Temporary dual chamber pacing (can decrease LVOT obstruction by 50%, but no improvement in functional capacity)
- Ischemia
    - Once again, ensure that hemodynamic goals are optimized
    - Patients likely require increased SVR to perfuse hypertrophic myocardium
    - Important to treat precipitating factors quickly as this can lead to arrhythmias or worsening LVOT obstruction
- Arrhythmia
    - Poorly tolerated
    - Treat immediately with volume (often due to hypovolemia) and cardioversion

PATHOPHYSIOLOGY

- HOCM (a.k.a. IHSS) is an autosomal dominant genetic disorder of cardiac sarcomeres of which a subset of patients (~25%) have a dynamic obstruction to left ventricular outflow
- Many patients may have minor or no symptoms (many individuals diagnosed during family screening)
- Variability in age of onset
    - Severity and progression of symptoms varies markedly with genetic abnormality
- Resting pressure gradient between LV outflow tract and aorta present in only ~25% of patients whereas other patients only have a gradient with exertion
- No predictable correlation between the degree of obstruction and symptoms:
    - Some patients with severe LVOT obstruction remain asymptomatic
    - Sudden death or cardiac arrest may be the initial presentation in some patients
- **LVOT Obstruction**
    - Dyspnea on exertion (90%)
    - Pre-syncope or syncope
    - Postural light-headedness
- **Hypertrophy**
    - Chest pain (25-30%)
        - Majority have normal angiogram
        - Commonly precipitated or worsened by heavy meals
    - Diastolic dysfunction
- **Arrhythmias**
    - Supraventricular Arrhythmias (A-fib, A-flutter)
        - Common (up to 25%)
        - May be associated with increased mortality
        - May be poorly tolerated d/t presence of physiologic changes:

        - Diastolic dysfunction
        - Increased HR
    - Can proceed to ventricular arrhythmias
- **Ventricular Arrhythmias**
    - Ventricular premature beats
        - Common in patients with HOCM
        - No evidence that frequent VPB are at increased risk for sustained ventricular arrhythmias
    - Non-sustained VT
        - 15-25% of patients
        - Increased risk for sudden cardiac death (8.6% per year vs. 1% per year)
- **Sudden Cardiac Death**
    - Risk factors
        - Syncope and / or family history of sudden cardiac death
        - LV wall thickness > 30 mm
        - Non-sustained ventricular tachycardia
        - Abnormal blood pressure response with exercise
    - AICD
        - Secondary prevention in survivors of cardiac arrest or sustained VT
        - Primary prevention in high risk patients
- **Management of HOCM**
    - LVOT obstruction
        - Pharmacotherapy
            - Beta-blockers
            - Calcium channel blockers
            - Disopyramide
                - antiarrhythmic with negative inotropic properties
        - Surgical Therapy
            - LV myectomy +/- MVR
            - Septal ablation with EtOH
    - Hypertrophy
        - Diuretics in patients with diastolic dysfunction
    - Arrhythmias
        - Amiodarone
        - Anticoagulants
        - Pacemaker
        - AICD



Ethanol ablation in HCM Representation of ethanol ablation via a septal coronary artery in hypertrophic cardiomyopathy (HCM). A section of hypertrophied left ventricle is shown, while the inset demonstrates a balloon occluding a septal coronary artery and ethanol-induced infarction. (Reproduced with permission from: Braunwald, E. Hypertrophic cardiomyopathy--the benefits of a multidisciplinary approach. N Engl J Med 2002; 347:1306. Copyright © 2002 Massachusetts Medical Society.)

REFERENCES

- Uptodate
- Roizen, Essence of Anesthesia