Management of Cardiomyopathy

• 3rd most common form of heart disease in U.S.
• 2nd most common cause of adolescent sudden death (IHSS or HOCM)
• directly affects cardiac structure and impairs myocardial function

Management of Cardiomyopathy

• Four types
  – Dilated Cardiomyopathy (DCM)
  – Hypertrophied Cardiomyopathy (HCM)
  – Restrictive Cardiomyopathy
  – Dysrhythmic right ventricular Cardiomyopathy
Management of Cardiomyopathy

Dilated Cardiomyopathy
- dilation and compensatory hypertrophy of myocardium
- depressed systolic function and pump failure with low cardiac output
- 80% of DCM cases are idiopathic
- most common age of diagnosis 20-50yrs
- African Americans and males have 2.5x increased risk

Dilated Cardiomyopathy
- Etiology
  - Ischemic
  - Valvular
  - Hypertensive
  - Inflammatory (Infectious/Noninfectious – peripartum)
  - Toxic (alcohol)
  - Metabolic (diabetes)
  - Nutritional (thiamine, selenium)
**Dilated Cardiomyopathy**

**Clinical Presentation**

- signs and symptoms of CHF
  - dyspnea on exertion
  - orthopnea
  - paroxysmal nocturnal dyspnea (PND)
- chest pain can occur due to low coronary vascular reserve

**Dilated Cardiomyopathy**

**Clinical Presentation**

- mural thrombi formation can occur
- adventitious heart sounds
  - holosystolic regurgitant murmur
  - gallop ($S_3$)
- other
  - dependent edema
  - bibasilar rales

**Dilated Cardiomyopathy**

**Diagnosis**

- CXR
  - enlarged heart
  - biventricular enlargement
  - pulmonary vascular congestion (cephalization)
Dilated Cardiomyopathy

Diagnosis

- ECG
  - LVH - poor R wave progression
  - Left atrial enlargement – Q waves
  - Atrial fibrillation

- Echocardiography – confirms diagnosis
  - Ventricular enlargement
  - Increased systolic and diastolic volumes
  - Decreased EF

Dilated Cardiomyopathy

Differential

- Acute MI
- Restrictive Pericarditis
- Acute valvular disruption
- Sepsis
- Any other condition that results in low cardiac output state
Dilated Cardiomyopathy

- Newly diagnosed or symptomatic DCM-admit
- IV lasix and digoxin-improve symptoms
- ACE-inhibitors and β-blockers-improve survival
- Amiodarone- for complex ventricular ectopy
- Anticoagulation can be considered

Peripartum Cardiomyopathy

- Peripartum cardiomyopathy (PPCM) is defined as the onset of acute heart failure without demonstrable cause in the last trimester of pregnancy or within the first 5 months after delivery
- form of Dilated Cardiomyopathy
- left ventricular systolic dysfunction
- results in signs and symptoms of heart failure
- often unrecognized, as symptoms of normal pregnancy commonly mimic those of mild heart failure
Peripartum Cardiomyopathy

1. Development of Cardiac failure in the last month of pregnancy or within 5 month after delivery
2. Absence of an identifiable cause for the cardiac failure
3. Absence of recognizable heart disease prior to the last month of pregnancy
4. Left ventricular systolic dysfunction demonstrated by classic echocardiographic criteria such as depressed shortening fraction or ejection fraction

Peripartum Cardiomyopathy

Etiology still unknown
- nutritional deficiencies
- small vessel coronary artery abnormality
- hormonal effects
- toxemia
- maternal immunologic response to fetal antigen
- myocarditis

Peripartum Cardiomyopathy

• Predisposing Factors
  – maternal age greater than 30 years
  – multiparous or eclamptic patients
  – twinning
  – racial origin (black)
  – hypertension
  – nutritional deficiencies
Symptoms of worsening cardiac failure like:
- dyspnea on exertion
- fatigue
- ankle edema
- embolic phenomena
- atypical chest pain
- hemoptysis

Many of above symptoms may occur even in normal pregnancy and can be mistaken for a diseased state.

**Postpartum Cardiomyopathy**

- Signs
  - evidence of a raised CVP
  - tachycardia
  - cardiomegaly with a gallop rhythm (S₃)
  - mitral regurgitation
  - pulmonary crackles and
  - peripheral edema

**Postpartum Cardiomyopathy**

The prognosis
- 50-60% patients show complete or near complete recovery within the first 6 months postpartum.
- in others, either continued clinical deterioration leading to early death or persistent left ventricular dysfunction and chronic heart failure results.
- an initial high risk period with mortality of 25-50% in the first 3 months postpartum.
- patients with persistent cardiomegaly at 6 months have a reported mortality of 85% at 5 years.
Postpartum Cardiomyopathy
Management
Vigorous treatment of acute heart failure
  – oxygen
  – diuretics
  – digoxin
  – vasodilators
• Use of ACE inhibitors in early pregnancy should be avoided as it has teratogenic effects on fetus

Postpartum Cardiomyopathy
Anticoagulant therapy is recommended because of high incidence of thromboembolic events in PPCM
  ▪ patient on oral anticoagulants require change to parenteral anticoagulants with short half life
  ▪ dose adjusted according to the PTT which may be discontinued before delivery
  ▪ after delivery Warfarin may be used

Myocarditis
• Inflammation of myocardium
• Can be result of systemic disorder or infectious agent
  ▪ Viral-Coxsackie B, echovirus, influenza, parainfluenza, Epstein-Bar, and HIV
  ▪ Bacterial-C. Diphtheria, N. meningitidis, M. pneumonia, and β-hemolytic strep
  ▪ Frequently coexistent with pericarditis
Myocarditis
Clinical Feature
• Fever, tachycardia out of proportion to fever, myalgias, headache, rigors
• Chest pain due to coexisting pericarditis
• Pericardial friction rub
• Severe cases may have CHF symptoms

Myocarditis
Diagnosis and Differential
• EKG - nonspecific changes, aV block, prolonged QRS duration, or ST elevation (with pericarditis)
• CXR - Normal
• Cardiac Enzymes - may be elevated
• Differential - ischemia or infarct, valvular disease, and sepsis

Myocarditis
ED Care and Disposition
• Supportive care
• Blood cultures
• Antibiotics for bacterial cause
• Watch for signs of progressive heart failure
Management of Cardiomyopathy

- **Alcoholic Dilated Cardiomyopathy**
  - Abnormal compliance-impaired diastolic relaxation and filling-output usually normal
  - 50% are hereditary
  - Prevalence 1 in 500, Mortality 1%
  - Mortality 4-6% in childhood/adolescence

Hypertrophic Cardiomyopathy

- Asymmetric LVH and/or RVH-primarily involves septum-usually without dilation
- Abnormal compliance-impaired diastolic relaxation and filling-output usually normal
- 50% are hereditary
- Prevalence 1 in 500, Mortality 1%
- Mortality 4-6% in childhood/adolescence

Hypertrophic Cardiomyopathy

Clinical Features

- Symptom severity progresses with age
- Dyspnea on exertion-most common initial or presenting symptom
- Angina-like chest pain, palpitations and syncope may also be present
Hypertrophic Cardiomyopathy

Exam
- Fourth heart sound (S₄)
- Hyperdynamic apical pulse
- Precordial lift
- Systolic ejection murmur at apex or lower left sternal border
- Murmur increased with valsala maneuver

Hypertrophic Cardiomyopathy

Diagnosis
- EKG - LVH - 30%
- Left atrial enlargement - 25-50%
- Large septal Q waves - 25%

Hypertrophic Cardiomyopathy

Diagnosis
- CXR - usually normal
  - Mild cardiomegaly

- Usually normal
  - Mild cardiomegaly
Hypertrophic Cardiomyopathy

Diagnosis
• Echocardiography
  – study of choice
  – demonstrates disproportionate septal hypertrophy

• Pts who c/o exercise intolerance or CP with typical HCM murmur- needs echo- refer to cardiology
• β-blocker- treatment of choice for HCM with CP
• Discourage vigorous exercise
• Admit HCM with syncopal episode

Treatment
Symptomatic Management
■ β-blockers
  – ↓ MVO₂
  – ↓ gradient (exercise)
  – arrhythmias
■ Calcium Channel blockers
■ Anti-arrhythmics
  – Atrial Fibrillation
    ■ Amiodarone
    ■ Disopyramide
  – AICD for sudden death
■ Antibiotic prophylaxis for endocarditis
No therapy has been shown to improve mortality
Treatment (Surgical)

For severe symptoms with large outflow gradient (>50mmHg)
*Does not prevent Sudden Cardiac Death*

- **Myomyectomy**
  - removal of small portion of upper IV septum
  - +/- mitral valve replacement
  - 5 year symptomatic benefit in ~ 70% of patients

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Treatment (Surgical)

- **Dual Camber (DDD pacemaker) pacing**
  - decreases LVOT gradient (by ~25%)
  - randomized trials have shown little long-term benefit
  - possible favorable morphologic changes

- **ETOH septal ablation**

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Restrictive Cardiomyopathy

- One of least common cardiomyopathies
- Ventricular volume and wall thickness is normal
- Decreased volume of both ventricles
- Mostly idiopathic - sometimes familial
Restrictive Cardiomyopathy

• Systemic disorders
  – amyloidosis
  – sarcoidosis
  – hemochromatosis
  – scleroderma
  – carcinoid

Restrictive Cardiomyopathy
Clinical Features

• Symptoms of CHF
  – dyspnea
  – orthopnea
  – pedal edema
  – chest pain (rare)

Restrictive Cardiomyopathy
Clinical Features

• Exam – may have
  – S₃ or S₄ gallop
  – rales
  – JVD
  – Kussmaul’s sign (JVD with inspiration)
  – hepatomegaly
  – pedal edema
  – ascites
Restrictive Cardiomyopathy

Diagnosis

- CXR - signs of CHF without cardiomegaly

Restrictive Cardiomyopathy

Diagnosis

- EKG - nonspecific changes most likely
- Conduction disturbances
- low-voltage QRS complexes are common with amyloidosis or sarcoidosis

Restrictive Cardiomyopathy

Differential Diagnosis

- Constrictive pericarditis
- Diastolic left ventricular dysfunction (due to ischemic or hypertensive heart disease)
- Need to differentiate restrictive cardiomyopathy from constrictive pericarditis (surgical treatment)
Restrictive Cardiomyopathy

Treatment
- Symptom directed
  - Diuretics
  - ACE inhibitors
- Corticosteroids for sarcoidosis
- Chelation therapy for hemochromatosis
- Admission based on severity of symptoms and availability of prompt follow up

Dysrhythmogenic Right Ventricular Cardiomyopathy (DRVC)

- Most rare form of cardiomyopathy
- Progressive replacement of RV myocardium with fibrofatty tissue
- Typical presentation of sudden death in young or middle aged pt
- Exam usually normal
- EKG - RBBB may be present
- Echo - necessary for diagnosis