**Cardiomyopathy**

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**Disclosures**

No Conflicts

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**Cardiomyopathy**

WHO Classification

Anatomy & physiology of the LV

1. Dilated
   - Enlarged
   - Systolic dysfunction
2. Hypertrophic
   - Thickened
   - Diastolic dysfunction
3. Restrictive
   - Diastolic dysfunction
4. Arrhythmogenic RV dysplasia
   - Fibrofatty replacement
5. Unclassified
   - Fibroelastosis
   - LV noncompaction

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Dilated Cardiomyopathy

Dilation and impaired contraction of ventricles:
- Reduced systolic function with or without heart failure
- Characterized by myocyte damage
- Multiple etiologies with similar resultant pathophysiology

Many of the cases are idiopathic
- Incidence of idiopathic dilated CM 5-8/100,000
- Incidence likely higher due to mild, asymptomatic cases
- 3X more prevalent among males and African-Americans

DCM: Etiology

- Ischemic
- Valvular (aortic and mitral valve disease)
- Hypertensive (end stage)
- Familial
- Idiopathic
- Infectious
  - Viral: Cox B, CMV, HSV, HCV
  - Rickettsial: Lyme Disease
  - Parasitic: Chagas’ Disease, Toxoplasmosis
- Non-infectious
  - Collagen/Vascular Disease (SLE, RA)
  - Peripartum
- Toxic
- Alcohol, Anthracyclines (adriamycin), Cocaine
- Metabolic
  - Endocrine: thyroid, pheochromocytoma, DM, acromegaly
  - Nutritional
    - Thiamine, selenium, carnitine
    - Neuromuscular (Duchene’s Muscular Dystrophy–x-linked)
Ischemic Cardiomyopathy

Patients with history of MI or revascularization (CABG or PCI)
Patients with >75% stenosis of the left main or proximal LAD
Patients with >75% stenosis of two or more epicardial vessels

Prognosis depends on Etiology

40 year old female presents with "crushing chest pain"
Definition of myocarditis

Inflammation of the heart muscle secondary to injury

- Ischemic damage
- Mechanical trauma
- Genetic cardiomyopathies
- Exposure to discrete external antigens
  - Viruses, bacteria, parasites, toxins, drugs
- Internal triggers
  - Autoimmune activation against self antigens
**Incidence**

Difficult to ascertain, depends on criteria used
- Estimated 8 to 10 per 100,000
- Unselected autopsy series as high as 1 to 4 per 100
- Young adults with sudden cardiac death, estimated 8.6%
- Idiopathic dilated cardiomyopathy patients only → 10-40% are secondary to myocarditis

**Population**

Bimodal age distribution
- Young children and teenagers: acute presentation
  - Exuberant response to initial exposure of antigen
- Older adults: Subtle and insidious symptoms of dilated cardiomyopathy and heart failure
  - Mature immune system with greater tolerance

**Clinical presentation**

Wide-ranging clinical presentation contributes to difficult diagnosis and classification
- Asymptomatic ECG or echocardiographic abnormalities
- Cardiac dysfunction, arrhythmias, heart failure and hemodynamic collapse
**Acute Myocarditis Presentation**

- Fatigue 82%
- Dyspnea on exertion 81%
- Arrhythmias 55%
- Palpitations 49%
- Chest pain at rest 26%

**Acute Myocarditis Presentation**

Acute ischemic syndrome type symptoms

- Elevated troponin
- ST-segment elevation on ECG
- Segmental wall motion abnormalities on echocardiography

Viral prodrome symptoms 20-80%

- Fever
- Chills
- Myalgias
- Constitutional symptoms

**Fulminant Myocarditis Presentation**

- Abrupt onset within 2 weeks of a viral illness
- Hemodynamic compromise
- Hypotension requiring pressors and mechanical support
- Echocardiogram reveals diffuse global hypofunction
- Thickening of the ventricular wall probably due to myocardial edema from myocardial inflammation and cytokine release
Endomyocardial biopsy in fulminant myocarditis

- Typical and diffuse myocarditis in each histologic section

Pathogenesis
Diagnostics: Expanded Criteria for Diagnosis of Myocarditis

Category I: Clinical Symptoms
- Clinical heart failure
- Fever
- Viral prodrome
- Fatigue
- Dyspnea on exertion
- Chest pain
- Palpitations
- Pre-syncope or syncope

Category II: Evidence of Cardiac Structural or Functional Perturbation in the absence of Regional Coronary Ischemia

Echocardiography evidence
- Regional wall motion abnormalities
- Cardiac dilation
- Regional cardiac hypertrophy
Troponin release
- High sensitivity (>0.1 ng/mL)
Normal coronary angiography or
Absence of reversible ischemia by coronary distribution on perfusion scan

Category III: Cardiac Magnetic Resonance Imaging

Increased myocardial T2 signal on inversion recovery sequence
Delayed contrast enhancement after gadolinium-DTPA infusion
Category IV: Myocardial biopsy – Pathologic or Molecular Analysis

- Pathology findings compatible with Dallas criteria
- Presence of viral genome of polymerase chain reaction or in situ hybridization
  - 80-100% specificity when performed from myocardial biopsy

Treatments/Therapeutic Approaches

- Supportive Therapy
- Immunosuppression
- Interferon
- Intravenous Immune Globulin
- Immune Adsorption Therapy
- Hemodynamic Support
- Vaccination

Supportive Therapy

- First-line therapy
- Only a small proportion of patient require hemodynamic support
- Treat this group same as for clinical heart failure
  - Diuretics
  - IV Vasodilators: Nitroglycerin, Nesiritide
  - ACEi, ARB, B-blockers when stable
  - Anti-inflammatory properties
Hemodynamic Support

Patients with fulminant myocarditis and cardiogenic shock may require
- Intra-aortic balloon pump
- Ventricular assist devices
- Extracorporeal membrane oxygenation (ECMO)
Prognosis

Several studies have looked at clinical variables that predict adverse outcomes (death and transplantation):
- Syncope
- Bundle branch block
- EF <40%

Other factors:
- NYHA Class III or IV
- PCWP <15mmHg
- Immunopathologic evidence of myocardial inflammation
- Failure to use B-blockers
- BiV failure
- Giant cell or viral genome on biopsy

DCM: toxic

Alcoholic cardiomyopathy
Chronic use

Mechanism?:
- Myocyte cell death and fibrosis
- Directly inhibits:
  - mitochondrial oxidative phosphorylation
  - Fatty acid oxidation

Risk vs intake

[Graph showing Relative Risk of Mortality]
Treatment
Guideline directed medical therapy
Reversible with abstinence

Questions