Cardiomyopathy

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Objectives

- Discuss the pathophysiology and etiologies of dilated, hypertrophic, and restrictive cardiomyopathy
- Review current medical and nursing management of patients with cardiomyopathy
- Discuss future trends in management
Cardiomyopathy

- Irreversible primary, progressive disease of the heart muscle
- Damage to the myocardial cells
- Progressive deterioration

Predominant affect is on the myocardium
Cardiomyopathy

“A diverse group of conditions whose final, common pathway is myocardial dysfunction”

Let’s talk!

WHO/ISFC classification: Based on pathophysiological features

Primary & Secondary: CM & Specific CM
Etiology

1. Cardiovascular disease (remodeling)
2. Infectious
3. Toxins
4. Systemic connective tissue diseases
5. Infiltrative and proliferative diseases
6. Nutritional deficiencies
7. Idiopathic
Classification

- Arrhythmogenic Right Ventricular CM
- Dilated
- Hypertrophic
- Restrictive
- Unclassified CM
Arrhythmogenic Right Ventricular Cardiomyopathy

Heart muscle disease characterized by replacement of the muscle by fibrous scar and fatty tissue. RV tends to be most affected.
Arrhythmogenic Right Ventricular Cardiomyopathy

- Symptoms early teens to second decade
- Etiology unknown (familial). Incidence: 1:3000-10,000
- Arrhythmia most prominent
- CHF, tricuspid regurgitation
- Symptoms
  - Palpitations
  - Syncope
  - Fatigue
  - Heart failure
  - Ventricular arrhythmias
Arrhythmogenic Right Ventricular Cardiomyopathy

- Antiarrhythmics – Amiodarone/Sotolol
- ACE
- Anticoagulation
- Digoxin
- Diuretics

Arrhythmia most prominent
CHF, tricuspid regurgitation, embolus
Arrhythmogenic Right Ventricular Cardiomyopathy

- Antiarrhythmics – Amiodarone/Sotolol
- ACE
- Anticoagulation
- Digoxin
- Diuretics

- Cardioversion
- Ablation
- Pacemaker
- ICD
- Surgery

Arrhythmia most prominent
CHF, tricuspid regurgitation, embolus
Dilated Cardiomyopathy (DCM)

EF less than 40% in the presence of increased LV dimensions
Dilated Cardiomyopathy (DCM)

- Cardiac enlargement
- Hypertrophy?
- Impaired systolic function of either or both ventricles

Dilation and impaired contraction

Apoptosis - necrosis - fibrosis

Laplace’s law
Dilated Cardiomyopathy (DCM)

- Most prominent CM
- Incidence – 36 cases/100,000 per year (Diagnostic criteria are lacking)
- Males and Africans
- Middle age
- IDCM - accounts for 25% of all heart failure cases
### DCM: Etiologies

- **Primary - Idiopathic**

- **Secondary:**
  - Electrolyte abnormalities
  - Endocrine abnormalities
  - Hypertension*
  - Infectious causes
  - Infiltrative diseases
  - Ischemia*
  - Neuromuscular diseases
  - Nutritional abnormalities
  - Rheumatologic diseases
  - Tachyarrhythmias
  - Toxins
  - Valvular heart disease*

*WHO classified as specific cardiomyopathies
Alcoholic Cardiomyopathy (DCM)

- CHF, HTN, CVA, arrhythmia, sudden death
- Major cause of secondary, nonischemic CM
  - 1/3 of all cases
- Three mechanisms
  - Direct toxic effect of alcohol/metabolites
  - Nutritional (thiamine)
  - Alcohol additives (cobalt)
- Men 30-55 years of age >10 year consumption
DCM: Clinical Presentation

- Fatigue/weakness
- Weight loss
- Dyspnea on exertion
- Peripheral edema
- BP
- Pulsus alternans
- Pulsatile jugular veins
- Apical displacement
- S3 / S4
- Murmurs
DCM: Clinical Presentation

- Orthopnea
- PND
- Chest pain
- Abdominal pain
- Emboli
- Dysrhythmias
- Syncope
- Sudden death
DCM: Diagnostic Tests

- CXR - enlargement
- EKG - tachyarrhythmias, Q waves, R-wave
- Echocardiography - diffuse global dysfunction. (MV?)
- Catheterization
Dilated Cardiomyopathy (DCM)
DCM: Management

- Sodium restriction
- Vasodilators (arterial/venous)
- ACE, ARB
- Beta-Blockers
- Cardioversion
- Pacemakers
- Diuretics
- Anticoagulation
- Antiarrhythmics (amiodarone)
- Heart transplant

Adrenergic and renin-angiotensin systems
Hypertrophic Cardiomyopathy

Stiffness of the LV with resultant impaired ventricular filling
Hypertrophic Cardiomyopathy

- Myocardial mass
- Ventricular cavities
- LV over RV
- Atria
- Heterogeneous
Hypertrophic Cardiomyopathy

Disproportionate thickening of the intraventricular septum. Greater hypertrophy of the ventricular septum than of the ventricular chambers.
Hypertrophic Cardiomyopathy

- Excessive thickening of the heart muscle.
- Myocardial disarray - normal alignment of muscle cells is absent
- Abnormalities of collagen deposition and altered contractile proteins in the myocytes (whole structure changes)
- Fibrosis – visible scar
- Myocardial ischemia - abnormal intramural coronary arteries
Hypertrophic Cardiomyopathy

**Asymmetric septal hypertrophy without obstruction**
- Asymmetric septal hypertrophy (ASH)
- Mitral valve in normal position
- Cavity reduced in size

**Symmetric hypertrophy**
- Symmetric or concentric hypertrophy

**Asymmetric septal hypertrophy with obstruction**
- Blood leaks back through mitral valve = mitral regurgitation
- Mitral valve presses against septum causing obstruction to blood flow
- Systolic anterior motion of the mitral valve (SAM)

**Apical hypertrophy**
- Apical hypertrophy
- Small cavity remains
Hypertrophic Cardiomyopathy

- Rare genetic disease
  - IHSS - Idiopathic hypertrophic subaortic stenosis
  - Asymmetric septal hypertrophy
  - Muscular subaortic stenosis

- Aortic stenosis
- Hypertension
- HCM vs. physiological hypertrophy
- HCM in the elderly
Hypertrophic Cardiomyopathy

1. Hyperdynamic state – septal thickening
2. Diastolic dysfunction – thickened muscle usually contracts well but doesn’t relax. Higher pressures result to allow expansion for the inflow of blood.
3. Possible outflow obstruction (~25%): MV involvement
4. Myocardial ischemia

Systolic dysfunction (pump) vs. Diastolic dysfunction (fill)
Need to differentiate systolic and diastolic dysfunction
Hypertrophic Cardiomyopathy

Clinical Manifestations

- Mild to asymptomatic – screenings
  - Sudden death

- Dyspnea – most common from diastolic dysfunction
Hypertrophic Cardiomyopathy

Clinical Manifestations:
- Angina, fatigue, syncope, dysrhythmias (more common)
- Palpitations, PND, CHF, dizziness (less common)
Clinical Manifestations

- S4 / S3 may be heard with outflow obstructions as well as a systolic murmur
- Ventricular arrhythmias – ¾ of patients
- SVT – ¼ to ½ of patients. Less tolerated
- EP testing – limited predictive value
- CXR – normal to cardiomegaly
- Squat position
Hypertrophic Cardiomyopathy

**Echocardiogram – screening and diagnosis**

- Cardinal sign is LV hypertrophy of septum and anterolateral free wall
- Variability in hypertrophy
- Dilated left atrium
- Normal to near-normal EF
- Septum at least 1.3 to 1.5 times the thickness of the posterior wall (15 mm). Average finding is 20 mm
- Outflow tract obstructions; MV / pressure gradient changes
- Diastolic dysfunction
R-wave in AVL >11mm;
R wave height in Lead I plus the S wave depth in Lead III > 25 mm
*S wave depth in V1 plus the height in V5 that exceeds 35 mm
Hypertrophic Cardiomyopathy

Morbidity / Mortality

- Mortality – 1%-3% per year
- Some remain stable or improve. Clinical deterioration is slow
- Sudden death – higher in children/adolescents
- Patients with gradients are more likely to deteriorate
- Atrial fibrillation – may lead to increase symptoms (LA dilation)
- LV dilation and dysfunction (DCM) occurs in 5-10%. Wall thinning and scar formation
Hypertrophic Cardiomyopathy

- **No symptoms** - No treatment
- **Mild symptoms** - Drug treatment
- **Moderate / severe symptoms**
  - Non-obstructive – Beta blockers, Calcium antagonists (Diuretics)
  - Obstructive: Drug treatment, alcohol ablation, myectomy, pacemaker
Restrictive Cardiomyopathy

Heart with Hypertrophic Cardiomyopathy

Heart with Restrictive Cardiomyopathy

Growth and arrangement of muscle fibers are abnormal. Heart walls thicken, especially in the left ventricle.

Ventricle walls stiffen and lose flexibility.
Restrictive Cardiomyopathy

- Myocardium becomes rigid, noncompliant
- Diastolic dysfunction
- Ventricular filling
- Systolic function preserved
- Resembles constrictive pericarditis
- Prevalence: <5% of CM in western world
Restrictive Cardiomyopathy

- **Idiopathic**
- **Noninfiltrative:**
  - Scleroderma
- **Infiltrative:**
  - Amyloidosis, Sarcoidosis
- **Storage Disease:**
  - Hemochromatosis
- **Endomyocardial:**
  - Metastatic cancers
  - Radiation
Restrictive Cardiomyopathy

Clinical heart failure – right failure prominent
- JVD
- S3, S4, or both
- Elevation in CVP
- Peripheral edema, liver enlargement, ascites
- Exercise intolerance
- Weakness
- Dyspnea

- AV block
- Symptomatic bradycardia
- Atrial fibrillation
Restrictive Cardiomyopathy

- **CXR**
- **CT / MRI**
- **Echocardiogram** – dilated atria, increased early LV filling velocity, decreased atrial filling velocity, and decreased isovolumetric relaxation time
- **Endomyocardial biopsy**
Restrictive Cardiomyopathy

- No satisfactory medical therapy (treat secondary causes)
- Drug therapy must be used with caution:
  - Diuretics for extremely high filling pressures
  - Vasodilators may decrease filling pressure
  - Calcium channel blockers to improve diastolic compliance
  - Digitalis and other inotropic agents are not indicated
Summary

Dilated Cardiomyopathy

Hypertrophic Cardiomyopathy

Restrictive Cardiomyopathy