Cardiomyopathy

- Disease of Heart Muscle
- Multiple etiologies from intrinsic vs extrinsic factors
- 3 primary patterns
  - Dilated
  - Hypertrophic
  - Restrictive

WHO Classification

- A. Functional Classification (intrinsic to myocardium)
  - 1. Dilated Cardiomyopathy
  - 2. Hypertrophic cardiomyopathy
  - 3. Restrictive Cardiomyopathy
  - 4. RV Dysplasia
  - 5. Unclassified (Obliterative)
- B. Specific Cardiomyopathies (secondary to external diseases)

Functional Classification of Cardiomyopathies

- I Cardiac Dilatation
- II Cardiac Hypertrophy
  - With Obstruction
  - Without Obstruction
- Cardiac Restriction

Graph showing pressure vs volume for normal and CHF conditions.
Specific Cardiomyopathies

- Ischemic
- Valvular
- Hypertensive
- Inflammatory (Idiopathic, Autoimmune, Infectious)
- Metabolic (Endocrine, Amyloid)
- General system Disease (Connective Tissue Disorders)
- Muscular Dystrophies
- Neuromuscular Disorders
- Sensitivity and Toxic Reactions
- Peripartum
### Etiology

<table>
<thead>
<tr>
<th>Category</th>
<th>Conditions</th>
<th>Comment</th>
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<tbody>
<tr>
<td>Genetic</td>
<td>Fabry, Kearns-Sayre Syndrome,</td>
<td>Right Ventricular Dysplasia present with</td>
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<tr>
<td></td>
<td>Right Ventricular Dysplasia</td>
<td>ventricular arrhythmias.</td>
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<tr>
<td>Endocrine</td>
<td>Hypothyroidism, Hyperthyroidism,</td>
<td>Pheochromocytoma, Acromegaly, Diabetes.</td>
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<tr>
<td>Metabolic</td>
<td>Hypocalcemia, Hyperphosphatemia,</td>
<td>Uremia</td>
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<tr>
<td></td>
<td>Uremia</td>
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</tbody>
</table>

### Clinical Presentation

- Dyspnea (Asthma, unrelenting URI)
- Fatigue
- Arrhythmias (Syncope, Palpitations, Dizziness)
- Chest pain
- Edema
- Febrile illness with SOB

### Diagnosis

- Physical Exam: JVD, S3, Rales, Hepatomegaly, edema,
- Labs: Elevated BNP, low serum Na,
- ECHO: key diagnostic tool to help determine etiology of CHF-myocardial disease, valvular, pericardial
- EKG: BBB, acute or old MI, arrhythmia
- I. heart cath/Endomyocardial Biopsy

### Symptomatic HF - Low EF (Stage C)

- Volume Regulation
  - Na restriction
  - Loop diuretic to renal threshold
- BHD dosing
- Drugs: Spironolactone (monitor for 
  - ACEI/ARB & titrate dose
  - β blocker & titrate dose

- Neurohormonal Blockade
  - Biventricular Pacing +/- ICD
  - Drugs: Devices:
  - NYHA Class III/IV
  - Consider investigational drugs

- Surgery:
  - Consider Transplant
  - Consider LVAD

- Non transplant candidate?
Diagnoses made by Endomyocardial Bx

1. Myocarditis
   - Giant Cell
   - CMV
   - Toxo
   - Chagas
   - Rheumatic
   - Lyme
2. Infiltrative
   - Amyloid
   - Sarcoid
   - Hemochromatosis
   - Carcinoid
   - Hyperesinophilic
   - Tumors
3. Toxins
   - Doxorubicin
   - Chloroquine
   - Radiation Injury
4. Genetic
   - Fabry
   - Kearns-Sayre Syndrome
   - RV Dysplasia

Potentially Reversible Dilated Cardiomyopathies

- Ischemic with viable myocardium
- Uncorrected Valvular Disease
- Hypersensitivity
- Inflammatory
  - CMV
  - Toxo
  - Lyme
- Toxic
  - Alcohol
  - Cocaine
  - Cobalt
- Endocrine
  - Hypothyroidism
  - Phaeochromocytoma
- Metabolic
  - HypoCa, HypoP
  - Uremia
  - Carnitine
- Nutritional
  - Selenium, Thiamine
- Infiltrative
  - Hemochromatosis
  - Sarcoidosis

Case #1: Dilated Cardiomyopathy

- Ischemic with viable myocardium
- Uncorrected Valvular Disease
- Hypersensitivity
- Inflammatory
  - CMV
  - Toxo
  - Lyme
- Toxic
  - Alcohol
  - Cocaine
  - Cobalt
- Endocrine
  - Hypothyroidism
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The patient was a 53 year old diabetic, hypertensive black male who was diagnosed with a dilated cardiomyopathy in 1998. Coronary artery catheterization revealed normal coronary vessels with an ejection fraction of 39%. Treatment with enalopril and furosemide was initiated. The patient did well until 10/00, when he developed increasing shortness of breath and was hospitalized for decompensated heart failure. He was treated with aggressive diuresis and optimization of vasodilator therapy including beta blockade with carvediol. In 11/00, patient developed ventricular tachycardia and required AICD implantation and treatment with amiodarone for recurrent VT. He again presented 1/31/03 with decreasing exertional tolerance, increasing abdominal girth, peripheral edema, and nightly PND. The patient had been compliant with his medical regimen and diet; he denied fever, palpitations, dizziness, blood loss.

His chronic medical regimen included aldactone 25 mg daily, accupril 20 mg daily, amiodarone 200 mg BID, carvediol 3.125 mg BID, furosemide 160 mg daily, coumadin and glyburide.

Physical exam was notable for: BP of 78/48 mm Hg, pulse 60 bpm, and respirations 20/min; JVD at 15 cm, bibasilar rales, S3 and II/VI holosystolic murmur, hepatomegaly, and +3 pitting pre-tibial edema.

Laboratory data analysis showed a hemoglobin of 10.8 gm/dl, hematocrit 34%, BUN 36, Creatinine 1.8 mg/dl. EKG: NSR LVH.

CXR: massive cardiomegaly, pulmonary venous redistribution, Kerley B lines, blunting of the costophrenic angles.

Dilated cardiomyopathy continued

Hospital Course:
He was treated acutely with Milrinone and intravenous diuretics. Right heart catheterization revealed a right atrial pressure of 20, PA 30 /17, PCW 19 mm Hg, cardiac output of 1.36 L/min with a pulmonary artery saturation of 36%. Echocardiogram demonstrated 4 chamber enlargement with a left ventricular ejection fraction <20%. He diuresed approximately 20 lbs. Peak VO2 was 10.6 ml/kg/min. He was listed for cardiac transplantation. He was discharged on an increased dose of accupril 20 mg BID and furosemide 160 mg BID. Aldactone was added to the regimen.

Dilated cardiomyopathy continued

Dilated cardiomyopathy continued
Case #2: Myocarditis

JL is a 31 year old man who presented complaining of malaise, shortness of breath, paroxysmal nocturnal dyspnea, orthopnea, nausea and vomiting. He denied any chest pain, syncope, fever or chills. Until the day of admission he was in his usual state of very good health, and exercised daily, up to 100 miles (160 kilometers) per day.

On physical examination he was found to be in pulmonary edema with a normal size heart.

Left ventricular ejection fraction was 20% by transthoracic echocardiogram.

There was no family history of heart disease, no history of drug or alcohol abuse.

PHYSICAL EXAMINATION:
- Temp 101.4
- Heart rate 135
- Blood pressure 91/63
- Respiratory rate 32
- Weight 170 pounds.

On physical examination he appeared pale,
had jugular venous distention,
had bilateral crackles on auscultation of his lungs.

His heart was tachycardic with a regular rhythm and S1-S2 and S3 gallop heart sounds.

He had no peripheral edema.

His abdomen was soft, nontender.

Neurologic exam was grossly intact and he was alert, awake and oriented times three.

A left cardiac catheterization performed showed clean coronary arteries.

An endomyocardial biopsy was done.

Case #3: Restrictive Cardiomyopathy
Well developed female in no acute distress

Skin: multiple eccymoses

HEENT: macroglossia, peri-orbital erythema

Neck: JVD 8cm

Lungs: decreased breath sounds on R about 1/3 way up

Heart: PMI 5th ICS, MCL, S1, S2, S3

Abd: Bowel sounds normoactive, nontender 12cm liver

Ext: 1+ edema

Labs:

WBC 8.5  H/H 11/33  Plt 235

Na 135  BUN 45  Cr 2.2

24 hour urine 427 g/day of protein

SPEP: Small monoclonal spike

EKG: NSR, low voltage, poor R wave progression

Restrictive Cardiomyopathy continued

ECHO:

Left ventricle is moderately hypertrophied with moderately reduced ejection fraction estimated at 30%. Right ventricle is moderately hypokinetic. Mild mitral regurgitation is seen. Trace aortic regurgitation is seen. Left atrium is moderately dilated. Right ventricular systolic pressure is estimated at 45mmHg.

Cardiac Cath: Normal coronary arteries; Left ventriculogram: moderately reduced LV function with an ejection fraction of 30%, trace mitral regurgitation. Abnormal right heart hemodynamics with an right atrial pressure 8, right ventricular systolic pressure 39/8, pulmonary artery pressure 40/21 with mean of 26, pulmonary capillary wedge of 21 mm Hg, PA sat 52% left ventricular diastolic pressure is 22. There was no equalization of right and left ventricular end diastolic pressures. Cardiac Output by Thermal Dilution was 2.29 L/min, Cardiac Index 1.25

Fat Pad Biopsy: negative for amyloid.

An endomyocardial biopsy showed diffuse interstitial, perimyocytic, and endocardial infiltrates of amyloid with focal vascular involvement.

Restrictive Cardiomyopathy continued

Amyloid

EKG in Amyloid Pt

Note: low voltage, Poor R wave progression

Hemodynamic Tracing in Amyloid
Classification of Amyloidosis

Case #4: Hypertrophic Cardiomyopathy

Amyloid

Case #2: Hypertrophic Cardiomyopathy

Case #4: Hypertrophic Cardiomyopathy
Hemodynamic Classification of Hypertrophic Cardiomyopathy

- Obstructive
  - Subaortic
  - midventricular
- Non-obstructive
  - Normal or supranormal LV fn
  - Impaired systolic function (end stage)
Genetic Mutations in HCM

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Pedigrees
HCM

- Autosomal Dominant Disease that affects males and females equally
- 50% of the offspring of affected individuals will be at risk for inheriting the gene and developing disease
- In any one family, all members have the same mutation
- Onset of clinical symptoms is delayed until adolescence or early adulthood
- Clinical features are not predictive of Sudden Death but certain mutations are highly predictive of sudden death