Cardiomyopathies: Sorting Through the Differences

An Overview

Class Code: FF518A
Cardiomyopathy

- Heterogeneous group of diseases of the myocardium
- Associated with mechanical and/or electrical dysfunction
- Usually (but not invariable) exhibit inappropriate ventricular hypertrophy or dilation
- Due to a variety of causes


European Society of Cardiology

“Cardiomyopathy is a myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to cause the observed myocardial abnormality” (p. 271).

(Elliot et al., 2008)
AHA Classification of Cardiomyopathy

- Primary (genetic)
- Mixed (genetic and nongenetic)
- Acquired
- Secondary
Primary (Genetic) Cardiomyopathy

- Previously idiopathic
- Processes confined to diseases of the heart muscle
- Hypertrophic cardiomyopathy
- Arrhythmogenic right ventricular cardiomyopathy / dysplasia
- Left ventricle noncompaction
- Conduction system disease
- Ion channelopathies
  - Long-QT syndrome
  - Brugada syndrome
  - Catecholaminergic polymorphic ventricular tachycardia
  - Short-QT syndrome
  - Idiopathic ventricular fibrillation

Mixed Cardiomyopathy

- Genetic and non-genetic
- Dilated cardiomyopathy
- Primary restrictive non-hypertrophied cardiomyopathy
Acquired Cardiomyopathy

- Myocarditis
  - Inflammatory cardiomyopathy
- Stress cardiomyopathy
  - “Tako-Tsubo”
- Peripartum cardiomyopathy
- Alcoholic dilated cardiomyopathy

Secondary Cardiomyopathy

- Infiltrative disorders
- Storage disease
- Toxicity
- Endomyocardial disorders
- Inflammatory disorders
- Neuromuscular/neurological disorders
- Nutritional deficiencies
- Autoimmune/collagen disorders
- Electrolyte imbalances
- Consequences of cancer therapy
Recommendation for Less Complex Classification

Morphological and functional phenotypes
- Dilated cardiomyopathy
- Restrictive cardiomyopathy
- Hypertrophic cardiomyopathy
- Arrhythmogenic right ventricular cardiomyopathy
- Unclassified

Elimination of Some Former Processes

- Valvular heart disease
- Systemic hypertension
- Congenital heart disease
- Atherosclerotic coronary artery disease producing ischemic myocardial damage secondary to impairment in coronary flow (ischemic cardiomyopathy)

(Elliott et al., 2008; Maron et al., 2006).
Our Focus Today

- Dilated cardiomyopathy (DCM)
- Restrictive cardiomyopathy (RCM)
- Hypertrophic cardiomyopathy (HCM)
- Arrhythmogenic right ventricular cardiomyopathy (ARVC)
- Tako-Tsubo cardiomyopathy (TCM)

Systolic Dysfunction (HFrEF)
Diastolic Dysfunction (HFpEF)
HFrEF - Systolic Dysfunction

- Impaired wall motion and ejection
- Dilated chamber
- 50% of HF Population
- **Hallmark**: Decreased LV Ejection Fraction $\leq 40\%$
- Coronary artery disease is cause in 2/3 of patients
- Remainder – other causes of LV dysfunction

Cardiomyopathy not synonymous with HF

HFrpEF - Diastolic Dysfunction

- Filling impairment
- Normal chamber size
- 50% of patients with HF have preserved LV function
- Normal EF or elevated
- Caused by
  - Hypertension
  - **Restrictive myopathy (C)**
  - Ischemic heart disease
  - **Hypertrophy (D)**
  - Valve disease
  - Idiopathic

Primarily a disease of elderly women with HTN
HFpEF - Diastolic Dysfunction

- Diagnosis is made when rate of ventricular filling is slow
- Elevated left ventricular filling pressures when volume and contractility are normal

In practice: the diagnosis is made when a patient has typical signs and symptoms of heart failure and has a normal or elevated ejection fraction with no valve disease.

Evaluation and Treatment Strategies

- Identification
- Echocardiogram is the most common tool for diagnosis
- Cardiac MR
- MRI
- Endomyocardial Biopsy
- Treat the cause
- Treat the dysfunction
  - HFrEF vs HFpEF
  - Mortality Benefit
  - Symptom Relief
**Stages, Phenotypes and Treatment of HF**

**STAGE A**
At high risk for HF but without structural heart disease or symptoms of HF
- Patients with:
  - HTN
  - Atherosclerotic disease
  - DM
  - obesity
  - Metabolic syndrome
  - With family history of cardiomyopathy

**STAGE B**
Structural heart disease but without signs or symptoms of HF
- Patients with:
  - Previous MI
  - LV remodeling including LVH and low EF
  - Asymptomatic valvular disease

**STAGE C**
Structural heart disease with prior or current symptoms of HF
- Patients with:
  - Known structural heart disease and HF signs and symptoms
  - Refractory symptoms of HF at rest despite GDMT

**STAGE D**
Refractory HF
- Patients with:
  - Marked HF symptoms at rest
  - Recurrent hospitalizations despite GDMT

**THERAPY**

**Goals**
- Control symptoms
- Improve HRQOL
- Prevent hospitalization
- Prevent mortality

**Strategies**
- Identification of comorbidities
- Treatment
  - Drugs
    - ACEI or ARB as appropriate
    - Beta blockers as appropriate
    - In selected patients:
      - ICD
      - Revascularization or valvular surgery as appropriate

**Options**
- Cardiovertor Defibrillator
- ACE Inhibitors
- Betablockers
- Aldosterone Antagonists
- Digoxin
- Diuretics
- Cardiac Resynchronization Therapy
- Cardiovertor Defibrillator

**HFrEF vs HFP EF**

**HFrEF**
- Focus on allowing for diastolic filling time
  - ACE Inhibitors
  - Betablockers
  - Aldosterone Antagonists
  - Digoxin
  - Diuretics
  - Cardiac Resynchronization Therapy
  - Cardiovertor Defibrillator

**HFP EF**
- Focus on slow heart rate
  - Maintain sinus rhythm
  - Cautious alterations in volume
  - Manage blood pressure
  - Cardiovertor Defibrillator
Cardiomyopathies: Sorting Through the Differences

Compare and Contrast Cardiomyopathies

Class Code: FF518B

Dilated Cardiomyopathy
Dilated Cardiomyopathy

Most common form of cardiomyopathy
• Idiopathic
• Genetic disorders
• Viral / Bacterial Infection
• Hyperthyroidism
• Chemotherapy
• Peripartum Syndrome Related to Toxicity
• Cardiotoxic Effects of Drugs or alcohol

Changes in Dilated Cardiomyopathy

- Ventricular Dilatation
- Decreased Ventricular Contractility
- Decreased Ejection of Ventricular Contents
- Increased Ventricular Pressure / Volume
- Increased Atrial Pressure / Volume
- Atrial Dilatation
- Atrial Overload
- Increased Pulmonary Pressure / Volume
- Fluid Accumulates in Pulmonary Capillary Bed
- Symptoms

- Activation of Neuro-hormonal Responses
- Vasoconstriction / Fluid Retention

Dilated Mitral Valve Annulus
Mitral Regurgitation
Systolic Dysfunction

- Impaired Contractility
- Decreased LV Ejection Fraction $\leq 40\%$
- Eccentric Hypertrophy
- Elongated myocytes
- Volume overload

Clinical Syndrome Resulting in Clinical Manifestations

Dyspnea and fatigue
  - May limit exercise tolerance

Fluid Overload
  - May lead to pulmonary congestion and peripheral edema

Impaired functional capacity and quality of life
Dilated Cardiomyopathy
Presentation

- Displace apical impulse
- S3 or S4
  - Left lateral position
  - Bell of Stethoscope
- Mitral regurgitation
  - Blowing/scratchy
  - Systolic
- Rales, if in failure
- Diminished breath sounds with effusions
- JVD
- HJR
- Edema

Diastolic Filling Sounds
S3 - Ventricular Gallop

Early diastolic filling sound
Caused by increased pressure and resistance to filling.
Most frequently associated with systolic dysfunction
Associated with:
- Fluid overload state
- Right or left ventricular failure
- Ischemia
- Aortic regurgitation
- Mitral regurgitation
Diastolic Filling Sounds - $S_3$

Patient position: left lateral decubitus position
Location: Mitral area.
Intensity: Heard best during expiration.
Duration: short.
Quality: dull, thud like.
Pitch: low. (Bell of Stethoscope)
May be normal in children, young adults (up to 35-40) and in the 3rd trimester of pregnancy.

Dilated Cardiomyopathy Diagnosis

<table>
<thead>
<tr>
<th>Echo</th>
<th>Cath</th>
<th>Chest X-Ray</th>
<th>ECG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chamber size</td>
<td>Not needed to diagnose DCM</td>
<td>Enlarged silhouette</td>
<td>Atrial Fibrillation</td>
</tr>
<tr>
<td>Wall thickness/shape</td>
<td></td>
<td>Congestion</td>
<td>Left Bundle Branch Block</td>
</tr>
<tr>
<td>Eccentric hypertrophy</td>
<td></td>
<td>Pleural Effusion</td>
<td>Large QRS Complexes</td>
</tr>
<tr>
<td>Usually thin</td>
<td></td>
<td></td>
<td>Hypertrophy</td>
</tr>
<tr>
<td>Clot formation</td>
<td></td>
<td></td>
<td>Abnormal P waves</td>
</tr>
<tr>
<td>Ejection Fraction</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal 55-65%</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Mild Dysfunction 42-55%</td>
<td></td>
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</tr>
<tr>
<td>Moderate Dysfunction 26-40%</td>
<td></td>
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<tr>
<td>Severe Dysfunction &lt;26%</td>
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</tbody>
</table>
Patient with LVEF of 10%, dilated left ventricle and left atrium.

### Stages, Phenotypes and Treatment of HF

#### STAGE A
At high risk for HF but without structural heart disease or symptoms of HF

- Patients with:
  - HTN
  - Atherosclerotic disease
  - DM
  - Obesity
  - Metabolic syndrome or patients
  - Using cardiotoxins
  - With family history of cardiomyopathy

#### STAGE B
Structural heart disease but without signs or symptoms of HF

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- Patients with:
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#### STAGE D
Refractory HF

- Patients with:
  - Marked HF symptoms at rest
  - Recurrent hospitalizations despite GDMT

### Therapy

**Goals**
- Control symptoms
- Improve HRQOL
- Prevent hospitalization
- Prevent mortality

**Strategies**
- Identification of comorbidities
- Treatment
  - Diuresis to relieve symptoms of congestion
  - Follow guideline driven indications for comorbidities, e.g., HTN, AF, CAD, DM
  - Revascularization or valvular surgery as appropriate

**Goals**
- Control symptoms
- Patient education
- Prevent hospitalization
- Prevent mortality

**Drugs for routine use**
- Diuretics for fluid retention
- ACEI or ARB
- Beta blockers
- Aldosterone antagonists

**Drugs for use in selected patients**
- Hydralazine/isosorbide dinitrate
- ACEI and ARB
- Digoxin
- CRT
- ICD
- Revascularization or valvular surgery as appropriate

**Drugs**
- ACEI or ARB as appropriate
- Beta blockers as appropriate
- ICD
- Revascularization or valvular surgery as appropriate

**Options**
- Advanced care measures
- Heart transplant
- Chronic inotropes
- Temporary or permanent MCS
- Experimental surgery or drugs
- Palliative care and hospice
- ICD deactivation

**Heart Healthy Lifestyle**
- Prevent vascular, coronary disease
- Prevent LV structural abnormalities

**Options**
- ACEI or ARB in appropriate patients for vascular disease or DM
- Statins as appropriate

**GDMT**
- Heart failure
e
- Patients with:
  - Marked HF symptoms at rest
  - Recurrent hospitalizations despite GDMT
- Previous MI
- LV remodeling including LVH and low EF
- Asymptomatic valvular disease

**Patients with**:
- HTN
- Atherosclerotic disease
- DM
- Obesity
- Metabolic syndrome

**Options**
- Using cardiotoxins
- With family history of cardiomyopathy

**Development of symptoms of HF**
- Structural heart disease
- Patients with:
  - Previous MI
  - LV remodeling including LVH and low EF
  - Asymptomatic valvular disease

**HFpEF**
- Refractory symptoms of HF at rest, despite GDMT

**HFrEF**
- Refractory symptoms of HF with prior or current symptoms of HF
- Patients with:
  - Known structural heart disease and HF signs and symptoms

**Heart Failure**
- Refractory symptoms of HF at rest, despite GDMT
- Marked HF symptoms at rest
- Recurrent hospitalizations despite GDMT

**Heart Healthy Lifestyle**
- Prevent vascular, coronary disease
- Prevent LV structural abnormalities
### Classification of Heart Failure
New York Heart Association

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
</table>
| 1     | - No limitation of physical activity  
       |   - Physical activity does not cause fatigue, palpitation or shortness of breath |
| 2     | - Slight limitation of physical activity  
       |   - Comfortable at rest, but physical activity results in fatigue, palpitations or shortness of breath |
| 3-A   | - Limitation of physical activity  
       |   - Comfortable at rest, but ordinary activity causes fatigue, palpitations or shortness of breath |
| 3-B   | - Significant limitation of physical activity  
       |   - Comfortable at rest, but minimal activity causes fatigue, palpitations or shortness of breath |
| 4     | - Unable to carry on any physical activity without discomfort  
       |   - Symptoms of heart failure at rest |

### Stages/Classification of Heart Failure

<table>
<thead>
<tr>
<th>ACC-AHA Stage</th>
<th>NYHA Functional Classification</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>None</td>
</tr>
<tr>
<td>B</td>
<td>I  Asymptomatic</td>
</tr>
</tbody>
</table>
| C             | II  Symptomatic with moderate exertion  
       |   III Symptomatic with minimal exertion  
       |   IV Symptomatic at rest |
| D             | Refractory heart failure requiring specialized interventions |
Dilated Cardiomyopathy
Acute Treatment

**Reduce Preload**
- Diuretics
- Venous Vasodilators
- Low Dose NTG
- Neseritide

**Reduce Afterload**
- Arterial Vasodilators
- High Dose NTG
- Neseritide
- Nitroprusside
- Intra Aortic Balloon Pump

**Increase Contractility**
- Decrease afterload
- Positive Inotropes
- Dobutamine
- Milronone

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Stages, Phenotypes and Treatment of HF

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**Therapy**
- Goals:
  - Control symptoms
  - Improve HRQOL
  - Prevent hospitalization
  - Prevent mortality

**Options**
- Advanced care measures
- Heart transplant
- Chronic therapies
- Temporary or permanent ICD
- Palliative care and hospice
- ICD deactivation

**THERAPY**
- Goals:
  - Control symptoms
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Heart Failure

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**Options**
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- Heart transplant
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- Palliative care and hospice
- ICD deactivation
Dilated Cardiomyopathy
Outcomes

- 50% mortality 5 years after diagnosis
- Progressive Heart Failure
- Sudden Death – 40%
- Embolic Stroke

Restrictive Cardiomyopathy
Restrictive Cardiomyopathy

- Rigidity of myocardial wall
- Not secondary to untreated hypertension, aortic stenosis or hypertrophy seen with HCM
- Results in decreased ability of chamber walls to expand during ventricular diastole
- Diastolic dysfunction
- Least common form of Cardiomyopathy
  - 5% of all primary heart muscle diseases (Goswami & Reddy, 2003)

Restrictive Cardiomyopathy

Primary Causes
Endomyocardial Diseases
- Eosinophilic Endomyocardial Fibrosis
- Endocardial Fibrosis
- Cardiac Transplant
- Anthracycline Toxicity
- Idiopathic
- Loeffler’s Endocarditis

Secondary Causes
Infiltrative disorders
- Amyloidosis
  - 90% of RCM in North America
- Sarcoidosis
- Radiation carditis
Storage Diseases
- Hemochromatosis
- Glycogen storage disease
- Fabry’s Disease
Amyloidosis

- Abnormal protein that infiltrates healthy tissue
- Most common form in the US is immunoglobulin light chain amyloidosis – AL amyloidosis
- Hematologic malignancy that results in deposits of protein fibrils (amyloid) in tissue resulting in organ dysfunction
- Heart becomes rubbery, with thick – not dilated – ventricular walls
  - Including septum
  - Ventricular chambers become smaller

Amyloidosis

- Incidence 8.0 per 1 million people
- More often men then women
- 60% of those with amyloidosis have involvement of the myocardium
- High mortality with median survival of 13.2 months
  - 4 months if heart failure is present
Amyloidosis

- Endomyocardial Fibrosis
  - Common in Africa
  - Fibrosis of ventricular endocardium and subendocardium
  - Extends to both ventricles
  - 50% mortality at 2 years
- Loeffler Endocarditis
  - Idiopathic eosinophilic syndrome
  - Eosinophilic infiltration results in myocardial fibrosis
  - Common in Africa, Asia, and South America

Idiopathic RCM

- Diagnosis of exclusion
- More often women than men
- All other sources of diastolic dysfunction have been ruled out.
- Rule out:
  - Hypertension for more than 5 years
  - Previous ischemic heart disease
  - Native valvular disease
  - Previous chest radiation
  - Connective tissue disorders, amyloidosis, hemochromatoisis, eosinophilic syndrome, alcoholism or intake of cardiotoxid drugs
Symptoms of Heart Failure

Fluid Accumulates in Pulmonary Capillary Bed

Atrium dilates due to increased volume and pressure

Increased volume and pressure in pulmonary symptoms

Ventricular chamber has limited ability to expand during filling

Decreased volume available for next ejection

Decreased stroke volume and cardiac output

Physiologic Changes in Restrictive Cardiomyopathy

Primary Diastolic Dysfunction

• Filling Impairment
• Rate of ventricular filling is slow
• Elevated left ventricular filling pressure when volume and contractility are normal
• Pressure overload
• Often elevated left ventricular ejection fraction
Restrictive Cardiomyopathy

- Fatigue, weakness
- Decrease in activity intolerance
- Hypotension – narrow pulse pressure
- Syncope
- Palpitations with arrhythmias
- Pale/ cool
- Peripheral pulses decreased
- Right sided failure

S4
- Left Lateral Position
- Bell of Stethoscope
- Murmur of Mitral Regurgitation
- Systolic Murmur
- 5th ICS MCL
- Mitral insufficiency
- Dilation of atrium
- Papillary muscle dysfunction
- Fibrosis of leaflets

Diastolic Filling Sounds

S4 - Atrial Gallop

Late diastolic filling sound
Caused by atrial contraction and the propulsion of blood into a noncompliant (stiff) ventricle.
Most frequently associated with diastolic dysfunction

Associated with:
- Fluid overload state
- Systemic hypertension
- Restrictive cardiomyopathy
- Ischemia
- Aortic stenosis
- Hypertrophic cardiomyopathy

May be normal in athletes
Diastolic Filling Sounds
S₄ - Atrial Gallop

Patient position: left lateral decubitus position.

Location
- Left-sided S₄ – mitral area.
- Intensity: louder on expiration.

Duration: Short

Quality: Thud like

Pitch: Low

Diagnosing Restrictive Cardiomyopathy

Rule Out Other Causes of Diastolic Differentiate from Constrictive Pericarditis
## Clinical Features of Constrictive Pericarditis and Restrictive Cardiomyopathy

<table>
<thead>
<tr>
<th>Clinical Features</th>
<th>Constrictive Pericarditis</th>
<th>Restrictive Cardiomyopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>History</td>
<td>Prior history of pericarditis or condition that causes pericardial disease</td>
<td>History of systemic disease (eg. Amyloidosis, hemochromatosis)</td>
</tr>
<tr>
<td>Heart Sounds</td>
<td>Pericardial knock, high frequency sound</td>
<td>Presence of loud diastolic filling sound, low frequency sound</td>
</tr>
<tr>
<td>Murmurs</td>
<td>No murmurs</td>
<td>Murmurs of mitral and tricuspid regurgitation</td>
</tr>
<tr>
<td>Cardiac Pressures</td>
<td>Left side filling pressures (PCWP) and right side filling pressures (CVP) are elevated and equal.</td>
<td>Left side filling pressures are generally &gt; right sided filling pressures by 5 mmHg or more.</td>
</tr>
<tr>
<td>Chest X-ray</td>
<td>Pericardial calcification is visible.</td>
<td>Atrial dilation with normal ventricular size</td>
</tr>
<tr>
<td>CT Scan / MRI</td>
<td>Pericardial thickening is visible.</td>
<td>No pericardial thickening; myocardial thickening can be present with amyloid infiltrates.</td>
</tr>
<tr>
<td>Echocardiogram</td>
<td>Normal sized ventricles and atria; pericardial thickening; pericardial effusion may be noted.</td>
<td>Biatrrial dilation with normal ventricles. Normal systolic function. Speckled texture of myocardium if amyloid infiltrate is present.</td>
</tr>
</tbody>
</table>

## Diagnosing Restrictive Cardiomyopathy

### Echo
- Chamber size
- Enlarged L Atrium
- Wall thickness
- Increased in infiltrative disorders
- Ejection Fraction – Normal or high
- Valve functioning
- Speckled appearance on myocardium with amyloidosis

### ECG
- Low QRS voltage
- No-specific ST-T wave changes
- P wave abnormalities
- Arrhythmias
- High incidence of atrial fibrillation
- Conduction abnormalities
- Blocks can develop

### Chest X-Ray
- Dilated atrium
- Congestion if in HF
- Calcified pericardium can be seen in constrictive pericarditis

### Cardiac Catheterization
- Full cath not necessary
- Hemodynamic measurements valuable
- Elevated LVEDP
- Elevated PAOP
- Elevated RA Pressures
- Elevated pulmonary pressures
Figure 10.2: Classic ECG of patient with cardiac amyloidosis. Note the low voltage QRS amplitude in the limb leads and V4-V6. A conduction abnormality with left anterior hemiblock is present. Pseudo infarct pattern is present in the precordial leads with slight ST elevation V1-V3 and a QS pattern in the same leads.

Figure 10.3: A: Normal P wave in Lead II and Lead V1. B: Right atrial hypertrophy in Lead II and Lead V1. C: Left Atrial Hypertrophy in Lead II and V1.
Additional diagnosis

- Cardiac Magnet Resonance (CMR)
- Differentiating Constrictive Pericarditis from RCM
- Greater accuracy of wall thickness than echocardiogram
Restrictive Cardiomyopathy Diagnosis

Endomyocardial Biopsy
• Septal wall of RV
• Multiple sites
• Essential for diagnosis of RCM

Restrictive Cardiomyopathy Treatment

<table>
<thead>
<tr>
<th>Reduce Diastolic Dysfunction</th>
<th>Treat Rhythm</th>
<th>Conduction Abnormalities</th>
<th>Ventricular Arrhythmias</th>
</tr>
</thead>
<tbody>
<tr>
<td>• No direct medications</td>
<td>• AF Control</td>
<td>• May require pacemaker</td>
<td>• Based on hemodynamic response</td>
</tr>
<tr>
<td>• Treat affects of restriction</td>
<td>• Loss of atrial kick</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Careful control of volume</td>
<td>• Decreased filling</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Diuretics - Fluid overload</td>
<td>• Digoxin cautiously in amyloidosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Decrease afterload - Arterial Vasodilators</td>
<td>• Calcium channel blockers detrimental in amyloidosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Careful with venous vasodilators</td>
<td>• Beta blocker OK</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Restrictive Cardiomyopathy Treatment

<table>
<thead>
<tr>
<th>Treat for Thromboembolic Complications</th>
<th>Treat Underlying Disease Process</th>
<th>Valve Replacement</th>
<th>Cardiac Transplant</th>
</tr>
</thead>
<tbody>
<tr>
<td>Highest risk in endocardial fibrosis</td>
<td>No cure for Amyloidosis</td>
<td>May provide symptomatic relieve</td>
<td>Beneficial in idiopathic / familial</td>
</tr>
<tr>
<td>High risk with enlarged atrium</td>
<td>Steroids and chemo helpful in slowing progression of disease process</td>
<td>High mortality</td>
<td>Need heart and liver with hemochromatosis</td>
</tr>
<tr>
<td>High risk with AF</td>
<td>Chelation for hemochromatosis</td>
<td></td>
<td>Limited usefulness in infiltrative</td>
</tr>
<tr>
<td>High risk with TR and MR</td>
<td></td>
<td></td>
<td>Amyloid patients transplanted follow with 6-12 months of chemotherapy</td>
</tr>
</tbody>
</table>

Restrictive Cardiomyopathy Outcomes

Poorest mortality of all cardiomyopathies

90% mortality rate at 10 years (Kavinsky & Parrillo, 2000).

Amyloid Heart

80% mortality at 2 years
Linking Knowledge to Practice

- Medications usually used in the treatment of atrial fibrillation may not be well tolerated in RCM. Careful assessment of the response to routine medications is important in this population and should not be taken lightly.
- Anything that would normally cause the heart rate to increase, including activity, decreased blood pressure, fever, shivering, and low blood volume, results in a further decrease in stroke volume in patients with RCM.
- Patients with RCM should be closely monitored for signs of decreased CO that may result from over diuresis. Signs include hypotension, especially orthostatic hypotension, lethargy, increased heart rate, and increased blood urea nitrogen levels.
- To differentiate cardiac ascites from non-cardiac ascites, utilize the assessment of JVD. JVD will be present with cardiac ascites and not with non-cardiac ascites.
- Amyloidosis can be a devastating diagnosis requiring a great deal of support for the patient and family.

Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy

1 of every 500 (Maron et al, 2003)
Primary genetic cardiomyopathy
Effects men and women equally
Hypertrophy of myocardial muscle mass in the absence of increased ventricular afterload
Associated with decreased ventricular filling (diastolic dysfunction) and decreased cardiac output
Most common cause of sudden death in young adults
Cause unknown
- 50% transmitted genetically

Hypertrophic Cardiomyopathy

- Disarray of cardiac myofibrils with hypertrophy of myocytes
- Cells take on a variety of shapes
- Myocardial scarring and fibrosis occurs
Hypertrophic Cardiomyopathy

- Usually only effects Left Ventricle
- Changes may be symmetrical
- Asymmetrical septal hypertrophy is more common

Hypertrophic Cardiomyopathy

May involve entire septum or only a portion of septum
Symptoms of HF may develop
Transferred to pulmonary system
Atrial dilatation due to increase in pressure and volume
Stiff walls resist filling (diastolic dysfunction)
Passive filling from the atria is slowed
Atrial kick more essential than normal
Ventricular chamber size decreases as enlarging walls close in on chamber
Compensation for decreased filling -> hyperdynamic systolic dysfunction
EF increases to 70-80%
Mitral Regurgitation
Transferred to pulmonary system
Symptoms of HF may develop

Physiologic Changes with Hypertrophic Cardiomyopathy

OBSTRUCTIVE Hypertrophic Cardiomyopathy

• 35% of HCM patients have obstruction at rest
• 35% additionally have obstruction with provocation
• Obstruction of outflow tract
• Septal wall enlarges into ventricular cavity
• Anterior leaflet of mitral valve drawn towards the septum during ejection
• Early closure of aortic valve, decreased ejection time, decreased cardiac output
Hypertrophic Cardiomyopathy Presentation

- Many asymptomatic for years
- Incidence of sudden death often first presentation
  - Or identified during screening of relative of patient with HCM
- Symptoms related to severity of diastolic dysfunction
- Heart failure
  - Dyspnea #1 sign
- Syncope / palpitations with activity
- Chest pain
- Supraventricular arrhythmias
- Development of mitral regurgitation
Hypertrophic Cardiomyopathy
Presentation

• Bisferiens Carotid Pulse (HOCM)
  • Brisk initial upstroke
  • Collapse of pulse then secondary rise
  • Must differentiate from AS – delayed upstroke
• PMI forceful and brisk
• S₄
• MR murmur
• Systolic murmur with obstructive disease process
  • Differentiating between HOCM and Aortic Stenosis

Subvalvular Left Ventricular Outflow Obstruction Systolic Murmur

• Timing: Mid systolic
• Location: best heard along left sternal boarder
• Radiation: usually does not radiate
• Configuration: crescendo-decrescendo
• Intensity: grade 3/6 to 4/6
• Pitch: medium
• Quality: harsh or rough
Subvalvular Left Ventricular Outflow Obstruction Systolic Murmur

HOCM murmur louder during Valsalva’s maneuver

Decreases venous return to the heart
- Decreased preload → ↓ left ventricular filling
- Decreased left ventricular filling → ↑ obstruction

Any factor that decreases venous return to the heart increases the murmur in HOCM
- Squatting increases venous return
- Standing decreases venous return

Aortic stenosis murmur becomes quieter during Valsalva’s maneuver

Hypertrophic Cardiomyopathy Diagnosis

<table>
<thead>
<tr>
<th>ECHO</th>
<th>ECG</th>
<th>Cardiac Cath</th>
</tr>
</thead>
</table>
| • Wall thickness  
• LV size  
• Hyperdynamic LV function  
• Atrial size  
• MV leaflets  
• LV outflow obstruction | • LV hypertrophy  
• Deep symmetrical T wave inversions  
• P wave abnormalities  
• Arrhythmias | • Not very helpful  
• Do not often find CAD with HCM |
Hypertrophic Cardiomyopathy

Treatment

• Goals
  • Relief of symptoms
  • Preventing complications
  • Preventing or reducing risk of sudden death
  • No evidence to support treatment of non-symptomatic patients

Beta Blockers
  • 1st choice (with or without HOCM)
  • Symptomatic benefit / improved exercise tolerance
  • Decreases HR
  • Improves LV relaxation
  • Helps control arrhythmias

Calcium Channel Blockers
  • If Beta Blocker not effective
  • Decrease LV wall tension
  • Decreases HR
  • Diltiazem or Verapamil (no nifedipine D/T vasodilatation)
Hypertrophic Cardiomyopathy
Treatment

Disopyramide
- Negative inotrope
- Class I antiarrhythmic
- Use with BB to treat LV outflow track obstruction
- Assists in HR control
- May cause ventricular

Anti arrhythmic Therapy
- Atrial Fibrillation
- Most common arrhythmia
- Poorly tolerated
- Anticoagulation
- Amiodarone or sotolol
- Obstructive or non-obstructive OK
- Ventricular or atrial arrhythmias

Other Medications
- Diuretics
- With caution
- ACE Inhibitors and NTG
- Avoided in HOCM
- Positive Inotropes
- Strictly avoid any medication that increases contractility in HOCM

Pregnancy
- Not restricted in non-obstructive disease

Endocarditis Prophylaxis
- NO LONGER INDICATED (was previously indicated in obstructive disease only)

Non-Obstructive Disease Treatment
- More difficult to treat if no symptoms
- Ultimately evolves into dilated cardiomyopathy
Surgical Myectomy

- Marked outflow obstruction
- On maximum medical therapy
- NYHA Class III or IV
- MV Replacement or repair at same time (increases operative mortality)
- Improvement noted immediately and last 20-30 years
- Survival Rates 80% at 10 years
- May need pacemaker (2%)

Percutaneous Alcohol Septal Ablation

- Symptomatic with full therapy
- NYHA Class III or IV
- Not appropriate if MVR needed
- Cath Lab Procedure
- Catheter in septal perforator
- Ethyl alcohol injected
- Myocardial infarction occurs
- Enlarged septum eventually shrinks
- May need pacemaker (20%)
Risk for Sudden Death

- One or more 1st degree relative with an episode of SCD
- Left ventricular wall thickness greater than 35 mm
- Prolonged or repetitive non-sustained ventricular tachycardia on Holter monitor
- Hypotensive BP response to exercise
- Syncope or near syncope

Family Evaluation

- Screen 1st degree relatives
- Genetic testing best if available
- Screenings
  - Annually from age 12 - 18 then every 5 years
  - Not necessary in relatives < 12 unless a particularly high risk family profile or a desire to play intense competitive sports.
- Screenings include:
  - Physical exam
  - 12 lead ECG
  - ECHO
Normal life span

Once diagnosed – routine follow up every 12 -18 months

SCD primary cause of shortened life span

Outcomes

Linking Knowledge to Practice

- Volume balance in patients with HCM is critical. Because the ventricular chamber is no longer able to expand during filling, the ventricle must fill fully in order to produce adequate stroke volume.

- The development of atrial fibrillation results in a loss of atrial kick and subsequent loss of filling, especially when heart rates are high. Atrial fibrillation can be very poorly tolerated (heart failure, hypotension) in those with significant hypertrophy. Assess the patient carefully for signs of decompensation and anticipate cardioversion if the rhythm is poorly tolerated.

- Beta blockers can cause fatigue, impotence, and sleep disturbances, especially with initial dosing. These symptoms can cause patients to stop taking the beta blocker. Inform the patient that the symptoms generally ease, especially the fatigue, over time as the patient’s body adjusts to the medication. Patients should be aware of these effects and encouraged to continue the medication as the body adjusts to the changes.

- Implantation of an ICD is a very emotional process, and patients should be provided with emotional support. Many facilities that place ICDs are aware of an ICD support group that can be helpful to patients dealing with the emotions associated with sudden death and implantation of this device.

- All family members should be educated on how to perform CPR correctly.
Arrhythmogenic Cardiomyopathy

- Inherited muscle disorder
- Often referred to as Arrhythmogenic Right-Ventricular Dysplasia (ARVD)
- Manifest as an arrhythmia, heart failure, or sudden death
- Genetic characteristics include autosomal dominance inheritance (most common)
- Most frequently affects the right ventricle
- More often than thought also affects left ventricle
- More often males than females
Arrhythmogenic Cardiomyopathy

- Cardiomyocyte replaced with fibro fatty tissue
- Initially patchy infiltration
- Progressive loss of muscle leads to thinning of the ventricular wall, dilation and pump dysfunction
- Thinnest portions of the right ventricle affected first
- Triangle of dysplasia: Inflow, outflow, apical regions of RV
## Disease Progression

<table>
<thead>
<tr>
<th>Early / Concealed phase</th>
<th>Overt Phase</th>
<th>Impaired contractility and right-sided failure</th>
<th>Bi-ventricular failure</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Subtle structural changes</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Often asymptomatic</td>
<td>• Noticeable structural and functional changes</td>
<td>• Right ventricular dilation</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Palpitations, pre-syncope, syncope, arrhythmias</td>
<td>• Decreased contractility</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Signs of right sided heart failure</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Disease spreads to left ventricle</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Signs of biventricular failure</td>
<td></td>
</tr>
</tbody>
</table>

## Arrhythmogenic Cardiomyopathy Presentation

- Palpitations
- Presyncope
- Syncope
- Often episode of sudden cardiac death is first presentation
- Signs of heart failure are late sign
## Diagnosis

<table>
<thead>
<tr>
<th>ECG</th>
<th>Echo</th>
<th>Endomyocardial Biopsy</th>
<th>MRI / CT</th>
</tr>
</thead>
</table>
| • T Wave inversion in leads V1-V6  
• Epsilon wave  
• VT with LBBB pattern  
• Conduction delays through right bundle | • RV enlargement and dysfunction | | • Detect fatty infiltrate |

### Figure 10.16: Prolonged upstroke of the S wave.

### Figure 10.17: Epsilon waves in a patient with ARVC.
Arrhythmogenic Cardiomyopathy
Treatment

- No cure
- Goal: Manage arrhythmias
- Antiarrhythmics: Amiodarone and beta blockers
- Implantable Cardiovertor Defibrillator
- Radiofrequency catheter ablation if unsuccessful in treating VT with antiarrhythmics
- Refrain from competitive / intense sports
- Screening of family members
  - 1st and 2nd degree relatives
ICD Implantation

- Recommended for the prevention of SCD in patients with ARVC and documented sustain VT or VF who are optimal medical therapy and have a reasonable expectation of good survival for more than one year (Class IB recommendation).

- ICD implantation can be effective for the prevention of SCD in patients with ARVC with extensive disease, including those with LV involvement, 1 or more affected family member with SCD, or undiagnosed syncope when VT or VF has not been excluded as the cause of syncope, who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year (Class IIa recommendation). (Zipes et al., 2006).

Outcomes

Progressive disease

- Long term prognosis continues to be evaluated
Tako-Tsubo Cardiomyopathy

- Transient left ventricular apical ballooning
- Abrupt onset of ballooning or dilatation of left ventricle
- Post menopausal women
- Occurs after psychosocial or physical stressors
- Also referred to as Stress Cardiomyopathy
- Cause unknown
  - Related to excessive catecholamines
## Tako-Tsubo Cardiomyopathy

Chest Pain mimicking acute MI  
ST-segment changes similar to anterior MI  
Elevated cardiac biomarkers  
Dyspnea  
Hypotension  
Signs of left ventricular failure

## Diagnosis

<table>
<thead>
<tr>
<th>ECG</th>
<th>Cardiac Biomarkers</th>
<th>Cardiac Cath</th>
<th>Echo</th>
</tr>
</thead>
</table>
| ST elevation mimicking AMI  
Prolonged QT interval | Mildly elevated  
Do not follow same rise and fall as AMI | No significant coronary artery disease  
Visualize ballooning of LV | LV  
Dysfunction with decreased ejection fraction  
Visualize ballooning of LV |
Treatment

- Goals: Similar to patients with Acute MI
- Treat Left Ventricular Failure
  - Cardiogenic Shock
  - IABP
  - Arrhythmias
  - Hypotension
  - Avoid inotropes
  - Cardiac Rehabilitation
  - Stress Reduction
Cardiomyopathy: Sorting Through the Differences

- Heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction
- Usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilation
  **THINK FUNCTIONAL CARDIOMYOPATHY**
- Pathological situation occurring regardless of cause
- Provides a discussion based on patient presentation and related pathology
- Describes the ventricular changes that occur
BE THE BEST THAT YOU CAN BE EVERY DAY. YOUR PATIENTS ARE COUNTING ON IT!