Cardiomyopathy: Sorting Through the Differences to Understand the Clinical Implications

ARRHYTHMOGENIC  DILATED
RESTRICTIVE  HYPERTROPHIC
TAKO-TSUBO

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Class Code: M18oM6oo
Mastery is not something that strikes in an instant, like a thunderbolt, but a gathering power that moves steadily through time, like weather.

- John Champlain Gardner Jr. (1933-1982)

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

Maron, B.J. et al Contemporary Definitions and Classification of the Cardiomyopathies: An American Heart Association Scientific Statement From the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention Circulation, Apr 2006; 113: 1807 - 1816.
“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).

(Elliott et al., 2008)
Primary (Genetic) Cardiomyopathy

- 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

(Elliott et al., 2008)
Elimination of Some Former Processes (Once classified as secondary CM)

- 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) (Picture B)
- Restrictive cardiomyopathy (RCM) (Picture C)
- Hypertrophic cardiomyopathy (HCM) (Picture D)
- Arrhythmogenic right ventricular cardiomyopathy (ARVC)
- Tako-Tsubo cardiomyopathy (TCM)
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
Activation of SNS

First Responder
- Decreased CO → ↓ BP → activates baroreceptors and vasomotor regulatory centers in medulla

Increase circulating catecholamines
- Stimulates alpha and beta receptors
  - Increase HR
  - Peripheral vasoconstriction
  - Contractility

Positive effect: ↑ CO and BP
Negative effect: ↑ O2 demand → ischemia, arrhythmias, sudden death

Chronic Stimulation of SNS

Norepinephrine (circulating catecholamine) is Cardiotoxic
- Decreases heart’s ability to respond to sympathetic stimulation
- Down regulation of B1 receptor sites (less sensitive)
- Contributes to decreased exercise tolerance
- Can also lead to ventricular remodeling

Be aware of your patient’s heart rate response to activity.
Activation of RAAS

Kidney’s response to decreased perfusion due to decreasing CO
Concentrations of angiotensin II, and aldosterone rise as end result
• Potent vasoconstriction
• Sodium/water absorption increases

Result
• Increased preload and increased afterload
• Increased myocardial oxygen demand

Angiotensinogen → Renin → Angiotensin I → Angiotensin Converting Enzyme (ACE) → Angiotensin II → Vasoconstriction → Increased preload and afterload

Angiotensinogen → Renin → Angiotensin I → Angiotensin Converting Enzyme (ACE) → Angiotensin II → Aldosterone → Sodium and water retention and potassium loss
### Harmful Result of RAAS Activation

Enhanced preload increases end-diastolic volume dilating the LV
LV becomes overstretched
LV changes size and shape (ventricular remodeling)
Contractility decreases
Congestive symptoms develop

### 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

Definition of Heart Failure

<table>
<thead>
<tr>
<th>Classification</th>
<th>Ejection Fraction</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. Heart Failure with Reduced Ejection Fraction (HF(r)EF)</td>
<td>( \leq 40% )</td>
<td>Also referred to as systolic HF.</td>
</tr>
<tr>
<td>II. Heart Failure with Preserved Ejection Fraction (HF(p)EF)</td>
<td>( \geq 50% )</td>
<td>Also referred to as diastolic HF.</td>
</tr>
<tr>
<td>III. HF(p)EF, Borderline</td>
<td>41% - 49%</td>
<td>These patients fall into a borderline or intermediate group.</td>
</tr>
<tr>
<td>IV. HF(p)EF Improved</td>
<td>&gt;40%</td>
<td>Subset of patients with HF(p)EF previously had HF(r)EF.</td>
</tr>
</tbody>
</table>
The Good Guys: Natriuretic Peptides

- Cardiac hormones secreted by myocytes
- **Atrial natriuretic peptide (ANP)**
  - Produced in atria
- **Brain natriuretic peptide (BNP)**
  - Produced in ventricles in response to increased ventricular pressure/stretching
  - Stronger release than ANP
- Promote vasodilatation (preload/afterload reduction)
- Reduce sodium/water retention (diuretic response)
- Reduce production/action of vasoconstrictor peptides
- Plasma concentrations elevated in patients in fluid overload
- Neseritide (Natrecor) is the synthetic form of BNP

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
Heart Failure Symptoms

- Exercise intolerance (hallmark)
  - Ability to perform ADLs
  - Fatigue
  - Dyspnea
- Paroxysmal nocturnal dyspnea
- Frequent night urination with less during the day
- Peripheral edema/weight change
- Chest pain
- GI problems
- Confusion/altered mental status

Physical Exam Findings

- General Appearance (resting dyspnea, cyanosis, cachexia)
- Weight gain
- BP/HR
  - Include orthostatic pressures
- JVD
- Hepatojugular reflux
- Edema
- Displaced apical impulse
- S3/S4
- Lung sounds
- Murmur of Mitral Regurgitation
Mitral Regurgitation

**Timing:** Holosystolic

**Location:** Mitral area

**Radiation:** To the left axilla

**Configuration:** Plateau

**Pitch:** High

**Quality:** Blowing, harsh or musical
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 - S₃

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>
</table>
| - Chamber size  
- Wall thickness/shape  
- Eccentric hypertrophy  
- Usually thin  
- Clot formation  
- Ejection Fraction  
- Normal 55-65%  
- Mild Dysfunction 41-55%  
- Moderate Dysfunction 26-40%  
- Severe Dysfunction <26% | - Not needed to diagnose DCM  
- Used to assist with defining cause of CM | - Enlarged silhouette  
- Congestion  
- Pleural Effusion | - Atrial Fibrillation  
- Left Bundle Branch Block  
- Large QRS Complexes  
- Hypertrophy  
- Abnormal P waves |
Treatment Strategies
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
  - Hypertrophic disease
  - DM
  - Obesity
  - Metabolic syndrome
  - Using cardiotoxins
  - 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

**STAGE D**
Refractory HF

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

**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, palpitation 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>
<tr>
<td>C</td>
<td>II Symptomatic with moderate exertion</td>
</tr>
<tr>
<td>D</td>
<td>IV Symptomatic at rest</td>
</tr>
</tbody>
</table>

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

**THERAPY**
- **Goals**
  - Control symptoms
  - Improve HRQOL
  - Prevent hospitalization
  - Prevent mortality
- **Strategies**
  - Diuretics to relieve symptoms of congestion
  - Follow guidelines driven indications for comorbidities, e.g., HTN, HF, CAD, DM
  - Revascularization or valvular surgery as appropriate

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

**THERAPY**
- **Goals**
  - Control symptoms
  - Improve HRQOL
  - Prevent hospitalization
  - Prevent mortality
- **Strategies**
  - Diuretics to relieve symptoms of congestion
  - Follow guidelines driven indications for comorbidities, e.g., HTN, HF, CAD, DM
  - Revascularization or valvular surgery as appropriate

**STAGE C**
Structural heart disease with prior or current symptoms of heart failure
- Patients with:
  - Known structural heart disease and HF signs and symptoms

**THERAPY**
- **Goals**
  - Control symptoms
  - Patient education
  - Prevent hospitalization
  - Establish patient’s end-of-life goals
- **Options**
  - Advanced care measures
  - Heart transplant
  - Chronic kidney disease
  - Temporary or permanent MCS
  - Palliative care surgery or drug
  - Palliative care and hospice
  - ICD deactivation

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

**THERAPY**
- **Goals**
  - Control symptoms
  - Improve HRQOL
  - Reduce hospital readmissions
  - Establish patient’s end-of-life goals
- **Options**
  - Advanced care measures
  - Heart transplant
  - Chronic kidney disease
  - Temporary or permanent MCS
  - Palliative care surgery or drug
  - Palliative care and hospice
  - ICD deactivation
**STAGE A**
At high risk for HF but without structural heart disease or symptoms of HF

**STAGE B**
Structural heart disease with symptoms of HF
- Known structural heart disease and HF signs and symptoms
- With family history of cardiomyopathy
- Development of symptoms of HF

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

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

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**THERAPY**

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

**Strategies**
- Identification of comorbidities

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

**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

**In selected patients**
- CRT
- ICD
- Revascularization or valvular surgery as appropriate

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

- ACE Inhibitors: LVEF ≤ 40%
- Betablockers: LVEF ≤ 40%
- Aldosterone Antagonists:
  - LVEF ≤ 35 NYHA II-IV
- Digoxin
  - LVEF ≤ 40% Persistent symptoms on ACE I, BB, and Diuretic
- Hydralazine/Isosorbide Dinitrate
- African American with Class C NYHA II-IV
- In addition to standard therapy
- Diuretics
- Cardiac Resynchronization Therapy
- Cardiovertor Defibrillator

Cardiac Resynchronization Therapy
Cardiac Resynchronization Therapy (CRT)

- Treatment modality for heart failure not just pacing
- Treatment modality in conjunction with drug therapy

Goals:
- Improve hemodynamics by restoring synchrony of ventricular contraction
- Improve quality of life
- Decrease mortality and morbidity

Cardiac Resynchronization Therapy

Goal: Force biventricular pacing
Goal: Ventricular Pacing 90% of time or greater

Causes of Loss of Bi V pacing:
- Long AV Delays
- Prolonged PVARP
- ST with 1 degree AV Block
- Lead dislodgement
Normal Ventricular Depolarization

Ventricular Depolarization with LBBB
Indications for CRT Therapy

Patient with cardiomyopathy on GDMT for >3 mo or on GDMT and >40 d after MI, or with implantation of pacing or defibrillation device for special indications

LVEF < 35%

Evaluate general health status

Comorbidities and/or frailty limit survival with good functional capacity to <1 y

Continue GDMT without implanted device

Acceptable noncardiac health

Evaluate NYHA clinical status

NYHA class I
- LVEF ≤ 30%
- QRS ≥ 150 ms
- LBBB pattern
- Sinus rhythm
- Ischemic cardiomyopathy
- QRS ≤ 150 ms
- Non-LBBB pattern
- Sinus rhythm

NYHA class II
- LVEF ≤ 35%
- QRS 120-149 ms
- LBBB pattern
- Sinus rhythm
- NYHA class III & Ambulatory class IV
- LVEF ≤ 35%
- QRS 150 ms
- Non-LBBB pattern
- Sinus rhythm
- Anticipated to require frequent ventricular pacing (>40%)
- Atrial fibrillation, if ventricular pacing is required and rate control will result in near 100% ventricular pacing with CRT

Special CRT Indications

NYHA class III & Ambulatory class IV
- LVEF ≤ 35%
- QRS ≥ 150 ms
- Non-LBBB pattern
- Sinus rhythm

Colors correspond to the class of recommendations in the ACCF/AHA Table 1.

Benefit for NYHA class I and II patients has only been shown in CRT-D trials, and while patients may not experience immediate symptomatic benefit, late remodeling may be avoided along with long-term HF consequences. There are no trials that support CRT-pacing (without ICD) in NYHA class I and II patients. Thus, it is anticipated these patients would receive CRT-D unless clinical reasons or personal wishes make CRT-pacing more appropriate. In patients who are NYHA class III and ambulatory class IV, CRT-D may be chosen but clinical reasons and personal wishes may make CRT-pacing appropriate to improve symptoms and quality of life when an ICD is not expected to produce meaningful benefit in survival.

Routine re-evaluation of pacing burden is important in the treatment of HF. If HF worsens assess CRT function.
Implantable Cardioverter Defibrillators

Heart Rhythm Society
Sudden Cardiac Death Primary Prevention Protocols

Ejection Fraction ≤
- 35% for Non-Ischemic Cardiomyopathy
- 40% for Ischemic Cardiomyopathy

Any Cardiomyopathy
- Not on Optimal Medical Therapy

Post-MI or Ischemic Cardiomyopathy
- With Permanent Left Ventricular Assist Device
- Post-MI Without Permanent Left Ventricular Assist Device
- Post-MI Without Permanent Left Ventricular Assist Device and EF ≥ 36–40%
- ESD (≥ 180 days) or EF < 36%
- EF ≤ 29%
- Hypertrophic Cardiomyopathy
- Marfan Syndrome
- Other Cardiomyopathies

Discharge Home, Continue Optimisation of Medical Therapy
Consider Consultation with Heart Rhythm Specialist/Consider Wearable Cardioverter Defibrillator

Reassess EF at 3 Months

Reassess EF if EF ≤ 30%

EF = 36–40%
Monitoring
- EF < 30%
- EF ≥ 40%

Refer for Consultation with Heart Rhythm Specialist

Learn more at www.HRSonline.org
ICD Device

**Pulse Generator**
- Single chamber, dual chamber, or biventricular pacing
- Back up pacing
- Antitachycardia pacing
- Implanted subcutaneously – same as pacemaker

**Defibrillator lead**
- Detects arrhythmias
- Delivers therapy
- Defibrillator lead capable of pacing and defibrillating
- Placed in right ventricle

ICD Functions

**ATP-Anti tachycardia Pacing**
- Tiered Antiarrhythmic Therapies
ICD Functions

- **Cardioversion Shock**
  - Delivers shocks from 0.1 to 30 joules synchronized on the R wave

- **Defibrillating Shock**
  - Delivers high energy (20-34 joules) unsynchronized shock for VF
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

Dilated Cardiomyopathy
Outcomes

- 50% mortality 5 years after diagnosis
- Progressive Heart Failure
- Sudden Death – 40%
- Embolic Stroke
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 with normal systolic function
• 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
- Loffler’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

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**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 8.9 per 1 million people
- More often men than 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
### Other Types of Amyloidosis

<table>
<thead>
<tr>
<th><strong>TTR Amyloidosis</strong></th>
<th><strong>SSA Amyloidosis</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Amyloid Transthyretin</td>
<td>Variant of transthyretin</td>
</tr>
<tr>
<td>Autosomal dominant inherited trait</td>
<td>Primarily a disease of men &gt; 70</td>
</tr>
<tr>
<td>Produces a genetic mutation of transthyretin (serum protein carrier of thyroxine and retinol)</td>
<td>Impacts most major organ systems</td>
</tr>
<tr>
<td>Protein produced in the liver</td>
<td>Has a much better survival</td>
</tr>
<tr>
<td>Infiltrates nervous system</td>
<td>Median survival reported at 75 months</td>
</tr>
<tr>
<td>Severe peripheral &amp; autonomic neuropathy, renal failure &amp; blindness.</td>
<td>Incidence is relatively low</td>
</tr>
<tr>
<td>Impact on the heart varies from patient to patient</td>
<td>Less frequent than AL Amyloidosis.</td>
</tr>
<tr>
<td>Does result in heart failure when the heart is involved</td>
<td>Newer imaging techniques recognizing this more often and is now thought to be more common than previously reported</td>
</tr>
<tr>
<td>Occurs less frequently than the other types</td>
<td></td>
</tr>
<tr>
<td>May be successfully treated with liver transplantation.</td>
<td></td>
</tr>
</tbody>
</table>

### Other Primary Causes of RCM

- **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
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 cardiotoxic drugs

Physiologic Changes in Restrictive Cardiomyopathy

Ventricular chamber has limited ability to expand during filling

- Decreased volume available for next ejection
- Decreased stroke volume and cardiac output

- Atrium dilates due to increased volume and pressure
- Increased volume and pressure in pulmonary symptoms

- Fluid Accumulates in Pulmonary Capillary Bed

Symptoms of Heart Failure
HFpEF - 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.
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
Patient position: left lateral decubitus position.

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

Duration: Short

Quality: Thud like

Pitch: Low

Diastolic Filling Sounds

$S_4$ - Atrial Gallop

"Believe Me"

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><strong>History</strong></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><strong>Heart Sounds</strong></td>
<td>Pericardial knock, high frequency sound</td>
<td>Presence of loud diastolic filling sound, low frequency sound</td>
</tr>
<tr>
<td><strong>Murmurs</strong></td>
<td>No murmurs</td>
<td>Murmurs of mitral and tricuspid regurgitation</td>
</tr>
<tr>
<td><strong>Cardiac Pressures</strong></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><strong>Chest X-ray</strong></td>
<td>Pericardial calcification is visible.</td>
<td>Atrial dilation with normal ventricular size</td>
</tr>
<tr>
<td><strong>CT Scan / MRI</strong></td>
<td>Pericardial thickening is visible.</td>
<td>No pericardial thickening; myocardial thickening can be present with amyloid infiltrates.</td>
</tr>
<tr>
<td><strong>Echocardiogram</strong></td>
<td>Normal sized ventricles and atria; pericardial thickening; pericardial effusion may be noted.</td>
<td>Biatrial dilation with normal ventricles. Normal systolic function. Speckled texture of myocardium if amyloid infiltrate is present.</td>
</tr>
</tbody>
</table>

### Diagnosing Restrictive Cardiomyopathy

<table>
<thead>
<tr>
<th>Echo</th>
<th>ECG</th>
<th>Chest X-Ray</th>
<th>Cardiac Catheterization</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Chamber size</td>
<td>• Low QRS voltage</td>
<td>• Dilated atrium</td>
<td>• Full cath not necessary</td>
</tr>
<tr>
<td>• Enlarged L Atrium</td>
<td>• No-specific ST-T wave changes</td>
<td>• Congestion if in HF</td>
<td>• Hemodynamic measurements valuable</td>
</tr>
<tr>
<td>• Wall thickness</td>
<td>• P wave abnormalities</td>
<td>• Calcified pericardium can be seen in constrictive pericarditis</td>
<td>• Elevated LVEDP</td>
</tr>
<tr>
<td>• Increased in infiltrative disorders</td>
<td>• Arrhythmias</td>
<td>• Cardiac MR</td>
<td>• Elevated PAOP</td>
</tr>
<tr>
<td>• Ejection Fraction – Normal or high</td>
<td>• High incidence of atrial fibrillation</td>
<td></td>
<td>• Elevated RA Pressures</td>
</tr>
<tr>
<td>• Valve functioning</td>
<td>• Conduction abnormalities</td>
<td>• Greater accuracy for measuring wall thickness.</td>
<td>• Elevated pulmonary pressures</td>
</tr>
<tr>
<td>• Speckled appearance on myocardium with amyloidosis</td>
<td>• Blocks can develop</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
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
- Gold standard
- Septal wall of RV
- Multiple sites
- Essential for diagnosis of RCM
- Low yield procedure in early stages of disease
- High yield procedure when amyloid changes noted on ECHO
## 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 affect of restriction</td>
<td>• Loss of atrial kick</td>
<td>• If concern for consistent RV pacing</td>
<td>• Most often have conduction abnormalities</td>
</tr>
<tr>
<td>• HR control</td>
<td>• Decreased filling</td>
<td>consider Cardiac Resynchronization therapy</td>
<td>• Not increased risk for ventricular arrhythmias</td>
</tr>
<tr>
<td>• Careful control of volume</td>
<td>• Digoxin cautiously in amyloidosis</td>
<td>• Calcium channel blockers detrimental in amyloidosis</td>
<td></td>
</tr>
<tr>
<td>• Decrease afterload - Arterial Vasodilators</td>
<td>• Binds to amyloid deposits</td>
<td>• Reports of clinical deterioration with CCBs</td>
<td></td>
</tr>
<tr>
<td>• Assist in stoke volume</td>
<td>• Susceptible to toxicity</td>
<td>• Beta blocker OK</td>
<td></td>
</tr>
<tr>
<td>• Careful with venous vasodilators</td>
<td>• Calcium channel blockers detrimental in amyloidosis</td>
<td>• Amiodarone OK</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Decrease afterload - Arterial Vasodilators</td>
<td>• Anticoagulate</td>
<td></td>
</tr>
</tbody>
</table>

### Treat for Thromboembolic Disease Process
- Highest risk in endocardial fibrosis
- High risk with enlarged atrium
- High risk with AF
- High risk with TR and MR

### Treat Underlying Disease Process
- No cure for Amyloidosis
- Steroids and chemo helpful in slowing progression of disease process
- Chelation for hemochromatosis

### Valve Replacement
- May provide symptomatic relieve
- High mortality

### Cardiac Transplant
- Beneficial in idiopathic / familial
- Need heart and liver with hemochromatosis
- Limited usefulness in infiltrative disorder
- Amyloid patients transplanted follow with 6-12 months of chemotherapy
Restrictive Cardiomyopathy Outcomes

Poorest mortality of all cardiomyopathies
90% mortality rate at 10 years (Kavinsky & Parrillo, 2000).

Amyloid Heart
• 80% mortality at 2 years
• SSA: Median survival 60 months
• AL: 5.4 months
• Idiopathic:
  • 64% 5 year survival
  • 37% 5 year survival

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

- 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
Atrial kick more essential than normal
Passive filling from the atria is slowed
Stiff walls resist filling (diastolic dysfunction)
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

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>
<tbody>
<tr>
<td>Wall thickness</td>
<td>LV hypertrophy</td>
<td>Not very helpful</td>
</tr>
<tr>
<td>LV size</td>
<td>Deep symmetrical T wave inversions</td>
<td>Do not often find CAD with HCM</td>
</tr>
<tr>
<td>Hyperdynamic LV function</td>
<td>P wave abnormalities</td>
<td></td>
</tr>
<tr>
<td>Atrial size</td>
<td>Arrhythmias</td>
<td></td>
</tr>
<tr>
<td>MV leaflets</td>
<td></td>
<td></td>
</tr>
<tr>
<td>LV outflow obstruction</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

---
Advances in technology has increased use of CMR in:
- Diagnosing HCM
- Identifying the distribution and extent of the hypertrophy
- Utilized when echocardiographic evidence of HCM is questionable or non-diagnostic and there is a high suspicion of HCM.
- 6% of patients with suspected HCM identified with CMR when the echocardiogram was inconclusive (Gersh et al., 2011).
- Apical hypertrophy more readily seen with CMR than with ECHO.
- May be used in risk stratification for SCD.
  - Wall thickness measurements - more precise on CMR
  - Can be helpful in the decision making process when surgical intervention is being considered.
  - Able to specifically define the extent and location of the septal hypertrophy.
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**

**PREVENTION**

**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 All family members should be educated on how to perform CPR.
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 effects 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>• Noticeable structural and functional changes</td>
<td>• Right ventricular dilation</td>
<td>• Disease spreads to left ventricle</td>
</tr>
<tr>
<td>• Often asymptomatic</td>
<td>• Palpitations, pre-syncope, syncope, arrhythmias</td>
<td>• Decreased contractility</td>
<td>• Signs of biventricular failure</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Signs of right sided heart 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>
<tbody>
<tr>
<td>• T Wave inversion in leads V1-V6</td>
<td>• RV enlargement and dysfunction</td>
<td>Unreliable</td>
<td>• Detect fatty infiltrate</td>
</tr>
<tr>
<td>• Epsilon wave</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• VT with LBBB pattern</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Conduction delays through right bundle</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

![ECG Image](image1.png)

![Echo Image](image2.png)

![MRI Image](image3.png)
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 Cardioverter 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 Ila recommendation). (Zipes et al., 2006).
Heart Transplant

- Represents less than 1% of the heart transplant population
- Viable option for those with refractory or terminal heart failure
- Disease process does not rule out heart transplantation
- Outcomes after heart transplant demonstrate a survival similar to HCM, DCM, & others while significantly better than restrictive & ischemic disorders

Outcomes

Progressive disease

Long term prognosis continues to be evaluated
Population is small
Not in the literature 30 years ago
Median survival – 60 years.
SCD primary cause of mortality
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

Tako Tsubo Cardiomyopathy 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 |
# Modified Proposed Mayo Clinic Criteria for Apical Ballooning Syndrome

<table>
<thead>
<tr>
<th>1.</th>
<th>Transient hypokinesis, akinesis, or dyskinesis of the left ventricular mid segments with or without apical involvement; the regional wall motion abnormalities extend beyond a single epicardial vascular distribution; a stressful trigger is often, but not always present.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Absence of obstructive coronary disease or angiographic evidence of acute plaque rupture.</td>
</tr>
<tr>
<td>1.</td>
<td>New electrocardiographic abnormalities (either ST-segment elevation and/or T-wave inversion) or modest elevation in cardiac troponin.</td>
</tr>
<tr>
<td>1.</td>
<td>Absence of: Pheochromocytoma or Myocarditis</td>
</tr>
</tbody>
</table>

*From Prasad, Lerman, & Rihal, 2009*
Treatment

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

Outcomes

- Recovery is good in this population
- Improvement of LV dysfunction occurs rapidly with much improvement seen within the hospital stay
- Full resolution of LV dysfunction seen in 1-3 months
- Most common complications associated with TCM include:
  - Heart failure
  - Mitral regurgitation
  - Cardiogenic shock
  - In hospital mortality rates are low - reported at 1-2%
  - Deaths result from:
    - Cardiogenic shock
    - Malignant arrhythmias
    - Free wall rupture
    - Systemic embolization
- Post-discharge mortality nearly 13% at 7 years with over 52% dying from cancer and others from other non-cardiac related causes.
- Incidence of recurrence is low - between 2-10%
- Most repeat events being stimulated by an event similar to the initial event.
Linking Knowledge to Practice

✓ Monitor for the development of hypotension. Hypotension with a new systolic murmur may represent LVOT obstruction.

✓ Monitor QT intervals inpatients that develop deep symmetrical T wave inversions. Risk for TdP increases with recent conversion of atrial fibrillation to sinus rhythm, administration of QT prolonging agents, hypokalemia, hypocalcemia, severe hypomagnesemia, and bradycardia.

✓ When assessing QT intervals remember that the QTc (QT corrected for the patient’s heart rate) should be used. (Chapter 5). A QTc > 0.50 sec (500 msec) places the patient at high risk for TdP.

✓ Emotional support is an important aspect of care in this population. Caring practice extends beyond the patient to the family and loved ones. The family can be supported in many ways. The top identified needs of families of critically ill patients include a need for information, a need for proximity to the patient, a need for assurance that the best possible care is being given, a need for support, and a need for comfort (Pryzby, 2005).

Cardiomyopathy: Sorting Through the Differences

- Heterogeneous group of diseases of the myocardium associated with mechanical and / or electrical dysfunction
- Usually (but not invariable) 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
My Vision Statement

Impact every patient and family on their journey and provide safe passage by meeting them where they are, connecting with them in a meaningful way, and delivering care with wisdom and intention.

- Cindy
BE THE BEST THAT YOU CAN BE EVERY DAY. YOUR PATIENTS ARE COUNTING ON IT!

Handouts are available on the NTI Network today and will be available next week at www.cardionursing.com