Cardiomyopathy: Sorting Through the Differences

ARRHYTHMOGENIC
HYPERTROPHIC
DILATED
RESTRICTIVE
TAKO-TSABO

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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.
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 (postpartum) 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

Functional Classification of Cardiomyopathy

- Pathological situation occurring regardless of cause
- Provides a discussion based on patient presentation and related pathology
- Cause often unknown
- Describes the ventricular changes that occur
  - Restrictive Cardiomyopathy
  - Hypertrophic Cardiomyopathy
  - Dilated Cardiomyopathy
  - Arrhythmogenic Cardiomyopathy
  - Stress Induced Cardiomyopathy (Tako-Tsabo)
Dilated Cardiomyopathy

- Most common form of cardiomyopathy
- Causes
  - Idiopathic
  - Ischemic
  - Genetic disorders
  - Hypertension
  - Viral / Bacterial Infection
  - Hyperthyroidism
  - Valvular Heart Disease
  - Chemotherapy
  - Peripartum Syndrome Related to Toxicity
  - Cardiotoxic Effects of Drugs or alcohol
Symptoms

Fluid Accumulates in Pulmonary Capillary Bed

Increased Pulmonary Pressure / Volume

Atrial Overload

Increased Pulmonary Pressure / Volume

Fluid Accumulates in Pulmonary Capillary Bed

Atrial Dilatation

Atrial Overload

Increased Atrial Pressure / Volume

Mitral Regurgitation

Dilated Mitral Valve Annulus

changes in Dilated Cardiomyopathy

Ventricular Dilatation

Decreased Ventricular Contractility

Decreased Ejection of Ventricular Contents

Increased Ventricular Pressure / Volume

Activation of Neurohormonal Responses

Vasoconstriction / Fluid Retention

Systolic Dysfunction

• Impaired Contractility
• Decreased LV Ejection Fraction < 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
**Dilated Cardiomyopathy Diagnosis**

<table>
<thead>
<tr>
<th><strong>Echo</strong></th>
<th><strong>Cath</strong></th>
<th><strong>Chest X-Ray</strong></th>
<th><strong>ECG</strong></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>
<td></td>
</tr>
<tr>
<td>Mild Dysfunction 41-55%</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Moderate Dysfunction 26-40%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe Dysfunction &lt;26%</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Stages of Heart Failure ACC / AHA**

<table>
<thead>
<tr>
<th><strong>Stage A</strong></th>
<th><strong>Stage B</strong></th>
<th><strong>Stage C</strong></th>
<th><strong>Stage D</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>At high risk for HF but without structural heart disease or symptoms of HF.</td>
<td>Structural heart disease but without signs or symptoms of Heart Failure</td>
<td>Structural heart disease with prior or current symptoms of HF.</td>
<td>Structural heart disease with prior or current symptoms of HF.</td>
</tr>
<tr>
<td>HPTN</td>
<td>Previous MI</td>
<td>Know structural disease and SOB, fatigue, reduced exercise tolerance.</td>
<td>Know structural disease and SOB, fatigue, reduced exercise tolerance.</td>
</tr>
<tr>
<td>CAD</td>
<td>LV Remodeling including LVH and low EF</td>
<td></td>
<td></td>
</tr>
<tr>
<td>DM</td>
<td>A symptomatic valvular disease</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Obesity</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Metabolic syndrome</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Family HX CM</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
# Utilizing Stages as a Guide to Therapy

<table>
<thead>
<tr>
<th>Stage A</th>
<th>Stage B</th>
<th>Stage C</th>
<th>Stage D</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Treat HTN</td>
<td>• All measures as stage A</td>
<td>• All measures under stage A</td>
<td>• All measures under stage A, B and C</td>
</tr>
<tr>
<td>• Treat DM</td>
<td>• ACE-I in select patients</td>
<td>• Dietary salt restriction</td>
<td>• Mechanical assist</td>
</tr>
<tr>
<td>• Smoking Cessation</td>
<td>• Beta Blockers in select patients</td>
<td>• Diuretics</td>
<td>• Transplantation</td>
</tr>
<tr>
<td>• Treat Lipids</td>
<td>• Implanted defibrillators</td>
<td>• Digitalis</td>
<td>• Palliative Care</td>
</tr>
<tr>
<td>• Regular Exercise</td>
<td></td>
<td>• Aldosterone Antagonists</td>
<td>• Hospice</td>
</tr>
<tr>
<td>• DC Alcohol / Drug Use</td>
<td></td>
<td>• Cardiac Resynchronization Therapy - CRT</td>
<td></td>
</tr>
<tr>
<td>• ACE-I in select patients</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Dilated Cardiomyopathy

**Acute Treatment**

<table>
<thead>
<tr>
<th>Reduce Preload</th>
<th>Reduce Afterload</th>
<th>Increase Contractility</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Diuretics</td>
<td>• Arterial Vasodilators</td>
<td>• Decrease afterload</td>
</tr>
<tr>
<td>• Venous Vasodilators</td>
<td>• High Dose NTG</td>
<td>• Positive Inotropes</td>
</tr>
<tr>
<td>• Low Dose NTG</td>
<td>• Neseritide</td>
<td>• Dobutamine</td>
</tr>
<tr>
<td>• Neseritide</td>
<td>• Nitroprusside</td>
<td>• Milronone</td>
</tr>
<tr>
<td></td>
<td>• Intra Aortic Balloon Pump</td>
<td></td>
</tr>
</tbody>
</table>
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
- 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
- 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
### Physiologic Changes in Restrictive Cardiomyopathy

<table>
<thead>
<tr>
<th>Ventricular chamber has limited ability to expand during filling</th>
</tr>
</thead>
<tbody>
<tr>
<td>Decreased volume available for next ejection</td>
</tr>
<tr>
<td>Decreased stroke volume and cardiac output</td>
</tr>
<tr>
<td>Atrium dilates due to increased volume and pressure</td>
</tr>
<tr>
<td>Increased volume and pressure in pulmonary symptoms</td>
</tr>
<tr>
<td>Fluid Accumulates in Pulmonary Capillary Bed</td>
</tr>
<tr>
<td>Symptoms of Heart Failure</td>
</tr>
</tbody>
</table>

### 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
- Syncope
- Palpitations with arrhythmias
- Pale/cool
- Peripheral pulses decreased

- 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

Diagnosing Restrictive Cardiomyopathy

- Rule Out Other Causes of Diastolic Dysfunction
  - Aortic Stenosis
  - Hypertrophic Cardiomyopathy
  - Hypertensive Cardiovascular Disease
- Differentiate from Constrictive Pericarditis
## Differentiation of RCM from Constrictive Pericarditis

<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 (e.g., Amyloidosis, Hemochromatosis)</td>
</tr>
<tr>
<td>Heart Sounds</td>
<td>Pericardial knock, high frequency sound</td>
<td>Presence of loud diastolic filling sound S3, low frequency sound</td>
</tr>
<tr>
<td>Murmurs</td>
<td>No murmurs</td>
<td>Murmurs of mitral and tricuspid insufficiency Arrhythmias</td>
</tr>
<tr>
<td>Heart Pressures</td>
<td>L &amp; R filling pressures up and equal (Elevated JVP)</td>
<td>L sided filling pressures &gt; R sided filling pressures</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
- Conduction abnormalities

### 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
### 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>
</tr>
</thead>
<tbody>
<tr>
<td>- No direct medications</td>
</tr>
<tr>
<td>- Treat affect of restriction</td>
</tr>
<tr>
<td>- Careful control of volume</td>
</tr>
<tr>
<td>- Diuretics - Fluid overload</td>
</tr>
<tr>
<td>- Decrease afterload - Arterial Vasodilators</td>
</tr>
<tr>
<td>- Careful with venous vasodilators</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Treat Rhythm</th>
</tr>
</thead>
<tbody>
<tr>
<td>- AF Control</td>
</tr>
<tr>
<td>- Loss of atrial kick</td>
</tr>
<tr>
<td>- Decreased filling</td>
</tr>
<tr>
<td>- Digoxin cautiously in amyloidosis</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Conduction Abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>- May require pacemaker</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Ventricular Arrhythmias</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Based on hemodynamic response</td>
</tr>
</tbody>
</table>

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**www.cardionursing.com**
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 diseases</td>
</tr>
<tr>
<td>• High risk with TR and MR</td>
<td></td>
<td></td>
<td>• Disease will affect new organs</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
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

![Normal Muscle Structure](image1) ![Myocardial Disarray](image2)

Hypertrophic Cardiomyopathy

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

![Heart with Hypertrophic Cardiomyopathy](image3)
Hypertrophic Cardiomyopathy

- May involve entire septum or only a portion of septum

Physiologic Changes with Hypertrophic Cardiomyopathy

- Ventricular chamber size decreases as enlarging walls close in on chamber
- Stiff walls resist filling (diastolic dysfunction)
- Compensation for decreased filling -> hyperdynamic systolic dysfunction
- Passive filling from the atria is slowed
- EF increases to 70-80%
- Atrial kick more essential than normal
- Atrial dilatation due to increase in pressure and volume
- Transferred to pulmonary system
- Mitral Regurgitation
- Symptoms of HF may develop
• 30-50% of HCM patients have obstruction
• 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

**HOCM**
**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 $\rightarrow$ $\downarrow$ left ventricular filling
  - Decreased left ventricular filling $\rightarrow$ $\uparrow$ 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 usually 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>

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)

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

### Anti arrhythmic Therapy
- Atrial Fibrillation
- Most common arrhythmia
- Poorly tolerated
- Anticoagulation
- Amiodarone or sotalol
- 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

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

Normal life span

Once diagnosed – routine follow up every 12 -18 months

SCD primary cause of shortened life span

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

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
## Arrhythmogenic Cardiomyopathy

### 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></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>
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</tr>
</tbody>
</table>
Arrhythmogenic Cardiomyopathy
Treatment

- No cure
- Goal: Manage arrhythmias
- Antiarrhythmics
- 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

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

Tako Tsubo Cardiomyopathy Diagnosis

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

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

THANK YOU!!!

Nurses Make a Difference