Cardiomyopathy: How Can One Name Mean So Many Things

ARRHYTHMOGENIC
HYPERPLASTIC
RESTRICTIVE
DILATED
TAKO-TSABO

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NTI Session: 198

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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

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

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
  - Dilated Cardiomyopathy
  - Restrictive Cardiomyopathy
  - Hypertrophic Cardiomyopathy
  - Arrhythmogenic Cardiomyopathy
  - Stress Induced Cardiomyopathy (Tako-Tsabo)
Cardiac Diastole (Atrial & Ventricular): Early Passive Ventricular Filling
Atrial Systole & Ventricular Diastole: Late Active Ventricular Filling

Atrial Kick

Beginning Ventricular Systole: Isovolumic Contraction
### Ventricular Systole: Ejection

![Diagram of ventricular systole]

- **Passive Ventricular Filling**
  - S3
- **Active Ventricular Filling**
  - Atrial Kick – S4
- **Valves Open**
  - Mitral
  - Tricuspid
  - Don't open well
    - Stenosis
- **Valves Closed**
  - Aortic
  - Pulmonic
  - Don't close well
    - Regurgitation

### Heart Sounds – the Basis for the sounds

<table>
<thead>
<tr>
<th>Diastole</th>
<th>Systole</th>
</tr>
</thead>
<tbody>
<tr>
<td>Passive Ventricular Filling</td>
<td>Isovolumic contraction</td>
</tr>
<tr>
<td>Active Ventricular Filling</td>
<td>Ejection of LV Contents</td>
</tr>
<tr>
<td>Atrial Kick – S4</td>
<td>Valves Open:</td>
</tr>
<tr>
<td>Mitral</td>
<td>Aortic</td>
</tr>
<tr>
<td>Tricuspid</td>
<td>Pulmonic</td>
</tr>
<tr>
<td>Don't open well</td>
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</tr>
<tr>
<td>Stenosis</td>
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</tr>
<tr>
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<td>Valves Closed</td>
</tr>
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</tr>
<tr>
<td>Regurgitation</td>
<td>Regurgitation</td>
</tr>
</tbody>
</table>
Basic Heart Sounds

\( S_1 \)

Closure of the Mitral (M\(_1\)) valve and the Tricuspid (T\(_1\)) valve

Beginning of Ventricular Systole and Atrial Diastole

Location: Mitral area

Intensity: Directly related to force of contraction

Duration: Short

Quality: Dull

Pitch: High

---

Basic Heart Sounds

\( S_2 \)

Closure of Aortic (A\(_2\)) and Pulmonic (P\(_2\)) Valve

End of Ventricular Systole

Location: Pulmonic area

Intensity: Directly related to closing pressure in the aorta and pulmonary artery

Duration: Shorter than \( S_1 \)

Quality: Booming

Pitch: High
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

Patient position: left lateral decubitus position
Location:
• Left-sided S3 – mitral area.
• Right-sided S3 – tricuspid area.
Intensity
• Left-sided heard best during expiration.
• Right-sided heard best during inspiration.
Duration: short.
Quality: dull, thud like.
Pitch: low.
May be normal in children, young adults (up to 35-40) and in the 3rd trimester of pregnancy.
Diastolic Filling Sounds
$S_4$ - 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
$S_4$ - Atrial Gallop

Patient position: left lateral decubitus position.

Location
- Left-sided $S_4$ – mitral area.
- Right-sided $S_4$ – tricuspid area.

Intensity
- Left-sided louder on expiration.
- Right-sided louder on inspiration.

Duration: Short
Quality: Thud like
Pitch: Low
Murmurs

High blood flow through a normal or abnormal valve

Forward flow through a narrowed or irregular orifice into a dilated chamber or vessel

Backward or regurgitant flow through an incompetent valve

Heart Sounds:
the Basis for the sounds

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<td>• Don't close well</td>
<td></td>
</tr>
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<td>• Regurgitation</td>
<td></td>
</tr>
</tbody>
</table>
Systolic vs Diastolic Dysfunction

Systolic Dysfunction

Impaired wall motion and ejection
Dilated chamber
2/3 of Heart Failure Population
Hallmark: Decreased LV Ejection Fraction < 40%
Ischemia is cause in 2/3 of patients
Multiple other causes including
  - Mitral Regurgitation
  - Dilated Cardiomyopathy
  - Untreated diastolic dysfunction
Diastolic Dysfunction

Filling impairment
Normal chamber size
20 to 40% of patients with HF have preserved LV function
Normal EF or elevated

Caused by
- Hypertension
- Restrictive myopathy
- Ischemic heart disease
- Ventricular hypertrophy
- Valve disease
- Idiopathic

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

Cardiac Output

Heart Rate X Stroke Volume

Preload Afterload Contractility

Same four components also determine myocardial oxygen demand

Preload

The ventricle is preloaded for ejection.

It’s about stretch!
**Preload**

End-diastolic stretch on myocardial muscles fibers

Determined by:
- Volume of blood filling the ventricle at end of diastole
- Greater the volume the greater the stretch (muscle fiber length)
  - Greater the stretch the greater the contraction
  - Greater the contraction the greater cardiac output
  
  TO A POINT

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**Non Invasive Assessment of Preload**

<table>
<thead>
<tr>
<th>Right Ventricular Preload</th>
<th>Left Ventricular Preload</th>
</tr>
</thead>
<tbody>
<tr>
<td>JVD</td>
<td>Orthopnea / PND/Dyspnea</td>
</tr>
<tr>
<td>Hepatojugular reflux</td>
<td>CXR report of vascular / interstitial edema</td>
</tr>
<tr>
<td>Peripheral edema *</td>
<td>Rales/crackles</td>
</tr>
<tr>
<td>Weight*</td>
<td>- Consider role of lymph drainage</td>
</tr>
<tr>
<td></td>
<td>S3 Gallop</td>
</tr>
<tr>
<td></td>
<td>Frothy sputum</td>
</tr>
<tr>
<td></td>
<td>Hypoxemia from decreased diffusion of oxygen</td>
</tr>
<tr>
<td></td>
<td>Pre-renal AKI (BUN/creatinine ratio &gt; 20:1)</td>
</tr>
<tr>
<td></td>
<td>Weight*</td>
</tr>
</tbody>
</table>
### Factors Influencing Preload

<table>
<thead>
<tr>
<th>Body Position</th>
<th>Circulating blood volume</th>
</tr>
</thead>
<tbody>
<tr>
<td>Venous Tone</td>
<td>Hypervolemia</td>
</tr>
<tr>
<td>Intrathoracic pressure</td>
<td>Hypovolemia</td>
</tr>
<tr>
<td>Intrapericardial pressure</td>
<td>Third spacing</td>
</tr>
<tr>
<td>Dysrhythmias</td>
<td>Distribution of blood volume</td>
</tr>
<tr>
<td>Atrial Kick</td>
<td>Sepsis</td>
</tr>
<tr>
<td>LV Function</td>
<td>Anaphylaxis</td>
</tr>
<tr>
<td></td>
<td>Venous vasodilators</td>
</tr>
</tbody>
</table>

**Afterload**

*After the ventricle is loaded, it must work to eject the contents! It’s about pressure!"*
Afterload

Pressure ventricle needs to overcome to eject blood volume

Left ventricle:
- Systemic vascular resistance (SVR)
- Other components
  - Valve compliance
  - Viscosity of blood
  - Arterial wall compliance
  - Aortic compliance

Right ventricle:
- Pulmonary vascular resistance

BP and Afterload

Blood pressure does not equal afterload

\[
\text{Blood Pressure (MAP)} = \text{Cardiac Output} \times \text{Systemic Vascular Resistance (Afterload)}
\]
Low BP could be due to:

- Low CO
  - HR too slow or too fast
  - Preload too low or too high
  - Contractility low
- Low SVR
  - Vasodilation due to sepsis, anaphylaxis, altered neurological function, drugs

More on Vascular Tone

Increased vascular tone is usually associated with compensation for low Stroke Volume
- Acute Cardiogenic shock
- Hypovolemic shock

Decreased vascular tone is usually due to abnormally pathological
- Sepsis
- Anaphylaxis
- Altered neurological control
### Non Invasive Assessment of Afterload

<table>
<thead>
<tr>
<th>Right Ventricular Afterload</th>
<th>Left Ventricular Afterload</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>NOTE</strong>: Any hypoxemia, positive pressure ventilation, and PEEP increase the workload of the right ventricle.</td>
<td><strong>Diastolic BP</strong> is closest noninvasive measurement. Evaluate the pulse pressure</td>
</tr>
</tbody>
</table>

### Use of Pulse Pressure & Heart Rate

<table>
<thead>
<tr>
<th>PP &lt; 35 with tachycardia (in absence of beta blocker)</th>
<th>PP &gt; 35 with tachycardia</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Early sign of inadequate blood volume</td>
<td>• Early sign of oxygenation failure</td>
</tr>
<tr>
<td></td>
<td>• Delivery cannot meet demand</td>
</tr>
</tbody>
</table>
### Causes of Increased LV Afterload

- Arterial vasoconstrictors
- Hypertension
- Aortic valve stenosis
- Increased blood viscosity
- Hypothermia
- Compensatory vasoconstriction from hypotension in shock

### Causes of Decreased LV Afterload

- Arterial vasodilators
- Hyperthermia
- Vasogenic shock states (sepsis and anaphylactic) where the body cannot compensate with vasoconstriction
- Chronic Aortic Regurgitation
  - hyperdynamic cardiac output therefore lowering systemic vascular resistance

### Increased Right Sided Afterload

#### Pulmonary hypertension
- mPAP > 25 mmHg or > 30 mmHg with exercise
- PVR > 250 dynes/sec/cm\(^5\)

#### Causes
- Hypoxemia
- Acidosis
- Inflammation
- Hypothermia
- Excess sympathetic stimulation
- Pulmonary endothelial dysfunction
  - Impaired nitric oxide and prostacyclin (PGI\(_2\)) release
- Primary pulmonary hypertension
Contractility

Ability of myocardium to contract independent of preload or afterload
• Velocity and extent of myocardial fiber shortening
• Inotropic state

Contractility

Related to degree of myocardial fiber stretch (preload) and wall tension (afterload).
Influences myocardial oxygen consumption

↑ contractility
⇒ ↑ myocardial workload
⇒ ↑ myocardial oxygen consumption
Important Points about Contractility

No accurate way to measure contractility

\textbf{Noninvasive Assessment: Ejection Fraction}

Low cardiac output does not necessarily mean diminished contractility (i.e. hypovolemia)

Correct preload and afterload problems first in a patient with a low ejection fraction.

Increasing contractility with medications will also increase myocardial oxygen demand.

Factors Altering Contractility

\textbf{Decreased contractility}
- Excessive preload or afterload
- Drugs – negative inotropes
- Myocardial damage
- Ischemia
- Cardiomyopathy
- Hypothyroidism
- Changes in ionic environment: hypoxia, acidosis or electrolyte imbalance

\textbf{Increased contractility}
- Drugs
- Positive inotropes
- Hyperthyroidism
- Adrenal Medulla Tumor
**Heart Rate**

Mathematically, heart rate increases cardiac output.

Physiological limit where increased heart rate will decrease cardiac output due to decreased filling time (decreased preload).

Consider as first line strategy to increase cardiac output when temporary pacemaker in place.

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**Dilated Cardiomyopathy**

[Images of a heart and athletic balls]
Most common form of cardiomyopathy
- Idiopathic
- Ischemic
- Genetic disorders
- Hypertension
- Viral / Bacterial Infection
- Hyperthyroidism
- Valvular Heart Disease
- 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 < 40%
- Eccentric Hypertrophy
- Elongated myocytes
- Volume overload

Pathophysiology

- Complex process involving continually emerging symptoms and deterioration
- Myocardial dysfunction initially results from any number of triggers
- Compensatory mechanisms help, then harm
Pathophysiology
The Real Culprit = Neurohormonal Response

Three significant events occur
1. Sympathetic Nervous System (SNS) stimulation
2. Activation of the Renin-Angiotensin-Aldosterone System (RAAS)
3. Ventricular Remodeling

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
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 = enhanced preload and afterload
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

HF as Progressive Disorder

Initial injury or stress on myocardium
Change in geometry of left ventricle
- Dilates
- Hypertrophies
- Becomes more spherical
Decreases mechanical performance of LV and increases regurgitation through mitral valve
These effects sustain and enhance the remodeling process
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
  - Left lateral position
  - Bell of Stethoscope
- Mitral regurgitation
  - Blowing/scratchy
  - Systolic
- Rales, if in failure
- Diminished breath sounds with effusions
- JVD
- HJR
- Edema
BNP Lab Test

Useful to differentiate “SOB”
High negative predictive value (<100 pg/ml we know it is not HF)
Must be used with assessment
May be elevated for other reasons

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>
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<td></td>
<td></td>
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<tr>
<td>Mild Dysfunction</td>
<td></td>
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</tr>
<tr>
<td>41-55%</td>
<td></td>
<td></td>
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<tr>
<td>Moderate Dysfunction</td>
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<td></td>
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<tr>
<td>26-40%</td>
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<td></td>
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<tr>
<td>Severe Dysfunction</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>&lt;26%</td>
<td></td>
<td></td>
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</tbody>
</table>
## Stages of Heart Failure

**ACC / AHA**

<table>
<thead>
<tr>
<th>Stage A</th>
<th>Stage B</th>
<th>Stage C</th>
<th>Stage D</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>
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</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>• Aymptomatic 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>
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## Utilizing Stages as a Guide to Therapy

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<th>Stage B</th>
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<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>• Daily Weight</td>
<td>• Transplantation</td>
</tr>
<tr>
<td>• Treat Lipids</td>
<td>• Implantable defibrillators</td>
<td>• Diuretics</td>
<td>• Palliative Care</td>
</tr>
<tr>
<td>• Regular Exercise</td>
<td></td>
<td>• Digitalis</td>
<td></td>
</tr>
<tr>
<td>• DC Alcohol / Drug Use</td>
<td></td>
<td>• Aldosterone Antagonists</td>
<td>• Hospice</td>
</tr>
<tr>
<td>• ACE-I in select patients</td>
<td></td>
<td>• Cardiac Resynchronization Therapy -CRT</td>
<td></td>
</tr>
</tbody>
</table>
Medication – Ace Inhibitors

- Definitive evidence of mortality/morbidity reduction
- Inhibits conversion of angiotensin I to II
- Interferes with ventricular remodeling
- Slows disease progression
- Used for mortality benefit not symptom relief
- May need to adjust diuretic dose and monitor K+
- Renal function and ACE-I
- Cough / angioedema
- Use of ARB or hydralazine/isordil
- ACE Inhibitor suffix: “pril”
  - Captopril, enalapril, lisonopril, perindopril, ramipril, trandolapril

Medications
Angiotensin Receptor Blockers

- ACE Inhibitors remain the first choice for inhibition of RAAS
- ARB’s are a reasonable alternative to ACE Inhibitor if intolerant to ACE Inhibitor due to cough or angioedema
- Directly block angiotensin II
- Combination of ACE I and ARB
- Reasonable alternative to ACE I as 1st line therapy for patients with mild / moderate HF & reduced LVEF, especially if already take ARB for other reason (HTN)
- ARB Suffix: “sartan”
Medication – Beta Blockers

- Decrease mortality/hospitalization
- Even better in combination with ACE Inhibitor
- Enhances overall well being
- Slows disease progression
- Inhibits ventricular remodeling and apoptosis
- Inhibits adverse effects of SNS
- Decrease myocardial oxygen consumption
- When to initiate?
- Not all beta blockers are equal
  - Metoprolol, bisoprolol, carvedilol
- Beta blocker suffix: “olol”

Medication - Diuretics

- Goal = decrease congestive symptoms
- Added for fluid overload – Stage C
- Loop diuretics preferred
  - Thiazide ok with overloaded HTN patients
- Keep K >4 and Mg >2
- Consider adding NA restriction as well
Medication - Digoxin

Stage C
- Added in patients with persistent symptoms already on ACE Inhibitor, Beta-blocker and diuretic
- Positive inotropic effect – weak effect
- Enzyme inhibition in noncardiac tissues – reduces sympathetic flow
- Improved symptoms, exercise tolerance and quality of life
- No reduction in mortality
- Beta-blocker better for rate control
- Low dose: 0.125mg daily
- No need for loading dose

Medications
Aldosterone Antagonists

- Stage C and D
- New York Heart Association Class IV, systolic dysfunction
  - EF <35%
- Already on ACE I, Beta Blocker, Diuretics, Digoxin
- Reduce death and rehospitalizations
- Spironolactone (RALES Trial)
  - Survival benefit in NYHA Functional Class 3 or 4 HF
- Epleranone
- High risk for HYPERkalemia
  - Do not initiate in patients with elevated creatinine (M>2.5 mg/dL; W>2.0 mg/dL) or elevated potassium (>5.0 mEq/L)
Hydralazine & Isosorbide Dinitrate

- Combination of fixed dose of Hydralazine & Isosorbide Dinitrate to a standard medical regimen for HF, including ACEIs and beta blockers, is recommended in order to improve outcomes for patients self-described as African Americans, with NYHA functional class II of IV HF. (Class IA)

Medication - Other

Warfarin recommended if HF and a fib

Antiarrhythmics
- ICD preferred if had VF or unstable VT
- Amiodarone for symptomatic ventricular arrhythmias and/or not ICD candidate

Avoid:
- NSAIDS
- Avandia
Non Pharmacologic Treatment Strategies

Stages to Guide Therapy

<table>
<thead>
<tr>
<th>Stage A</th>
<th>Stage B</th>
<th>Stage C</th>
<th>Stage D</th>
</tr>
</thead>
</table>
| Class IA-C)  
• Treat HTN  
• Treat DM  
• Smoking Cessation  
• Treat Lipids  
• Regular Exercise  
• DC Alcohol / Drug Use  
• Treat thyroid disorders  
• ACE-I in select patients | • All measures as stage A  
• ACE-I in select patients  
• Beta Blockers in select patients  
• Implantable defibrillators | • All measures under stage A  
• Dietary salt restriction  
• Daily Weight  
• Medications for routine use:  
  - Diuretics  
  - ACE - I  
  - Beta-Blockers  
  - Digitalis  
  - Aldosterone Antagonists  
  - Hydralazine/Isordil (African Americans)  
• Exercise training  
• Devices in select patients  
• Resynchronization Therapy  
• Implantable defibrillators | • All measures under A, B and C  
• Mechanical assist  
• Transplantation  
• Palliative Care  
• Hospice |
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
Indications for CRT

- Stage C
- NYHA Class III or ambulatory Class IV
- EF ≤ 35%
- Demonstrates cardiac dysynchrony
  - QRS duration ≥ 120 ms
- Optimal medical therapy
  - ACE Inhibitor
  - Beta Blocker
  - Diuretic

Normal Ventricular Depolarization

- Mitral valve closed to prevent regurgitation
- Septum moves leftward and functions as part of LV to eject blood
- Papillary muscles contract with LV
Ventricular Depolarization with LBBB

CRT

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
Automatic Implantable Cardioverter Defibrillators

Indications for ICD

• LVEF ≤35% due to prior MI ≥ 40 days old and NYHA functional Class II or III.
• Nonischemic DCM, LVEF ≤35% and NYHA functional Class II or III.
• LV dysfunction due to prior MI ≥ 40 days old, LVEF ≤30%, and are in NYHA functional Class I.
• Nonsustained VT due to prior MI, LVEF ≤40%, and inducible VF or sustained VT at electrophysiological
Implantable Cardiovertor Defibrillator - Indications

Secondary Prevention (IA)
• Symptoms of HF
• History of cardiac arrest, VF, or hemodynamically destabilizing VT

Primary Prevention (IA)
• Non-ischemic dilated myopathy or ischemic heart disease > 40 days post-MI or 90 days post intervention
• EF ≤ 35%
• NYHA class II or III in optimal medical therapy
• Not recommended in Stage D

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 Termination Therapies

ATP-Antitachycardia Pacing
- Painless
- “Slow” VT’s
- Burst
- Ramp
- Decremental Scanning

Cardioversion Shock
Defibrillating Shock

Other Device Features

Brady Pacing

Atrial Diagnostics
- Differentiates between SVT and VT

Stored Electrograms
- Store arrhythmia event

Noninvasive EPS
- EP study through implantable leads
Refractory End-Stage HF

Stage D
- Meticulous fluid control
- Referral for cardiac transplant
- Discuss Prognosis: Survival and Function
- LV Assist Therapy
- No Chronic Intermittent Inotropic Support in refractory end stage HF
- No left ventriculectomy
- Compassionate End-of Life Care
- Advanced Directives, Hospice / Palliative Care Referrals
- ICD Deactivation

Acute Decompensated Heart Failure

<table>
<thead>
<tr>
<th>Warm and Dry</th>
<th>Warm and Wet</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal Perfusion</td>
<td>Normal Perfusion</td>
</tr>
<tr>
<td>No Congestion</td>
<td>Congestion</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Cold and Dry</th>
<th>Cold and Wet</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low Perfusion</td>
<td>Low Perfusion</td>
</tr>
<tr>
<td>No Congestion</td>
<td>Congestion</td>
</tr>
</tbody>
</table>

Assessment of Skin and Lungs
Fluid Overload vs. Hypoperfusion

**Hypoperfusion**
- Narrow pulse pressure
- Resting tachycardia
- Cool Skin
- Altered mentation
- Decreased urine output
- Increased BUN/Creatinine
- Cheyne Stokes Respirations

**Fluid Overload**
- Weight gain
- Peripheral edema
- Jugular venous distention
- SOB
- Crackles

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>Nesentide</td>
<td>Dobutamine</td>
</tr>
<tr>
<td>Nesentide</td>
<td>Nitroprusside</td>
<td>Milronone</td>
</tr>
<tr>
<td>Intra Aortic Balloon Pump</td>
<td></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

- 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

### 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

<table>
<thead>
<tr>
<th><strong>Echo</strong></th>
<th><strong>ECG</strong></th>
<th><strong>Chest X-Ray</strong></th>
<th><strong>Cardiac Catheterization</strong></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></td>
<td>• Elevated PAOP</td>
</tr>
<tr>
<td>• Ejection Fraction – Normal or high</td>
<td>• Conduction abnormalities</td>
<td></td>
<td>• Elevated RA Pressures</td>
</tr>
<tr>
<td>• Valve functioning</td>
<td></td>
<td></td>
<td>• Elevated pulmonary pressures</td>
</tr>
<tr>
<td>• Speckled appearance on myocardium with amyloidosis</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### 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

---

**Normal Patient**

**Patient with Amyloid Deposits in Heart**
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 affect 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></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Careful with venous vasodilators</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Restrictive Cardiomyopathy Treatment

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Underlying Disease Process</th>
<th>Valve Replacement</th>
<th>Cardiac Transplant</th>
</tr>
</thead>
<tbody>
<tr>
<td>Treat for Thromboembolic Complications</td>
<td>No cure for Amyloidosis</td>
<td>May provide symptomatic relief</td>
<td>Beneficial in idiopathic / familial</td>
</tr>
<tr>
<td>• Highest risk in endocardial fibrosis</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 enlarged atrium</td>
<td>• Chelation for hemochromatosis</td>
<td></td>
<td>• Limited usefulness in infiltrative diseases</td>
</tr>
<tr>
<td>• High risk with AF</td>
<td></td>
<td></td>
<td>• Disease will affect new organs</td>
</tr>
<tr>
<td>• High risk with TR and MR</td>
<td></td>
<td></td>
<td></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

Hypertrophic Cardiomyopathy

- Usually only effects Left Ventricle
- Changes may be symmetrical
- Asymmetrical septal hypertrophy is more common
May involve entire septum or only a portion of septum

Physiologic Changes with Hypertrophic Cardiomyopathy

- Compensation for decreased filling -> hyperdynamic systolic dysfunction
- EF increases to 70-80%
- Stiff walls resist filling (diastolic dysfunction)
- Passive filling from the atria is slowed
- Atrial kick more essential than normal
- Atrial dilatation due to increase in pressure and volume
- Transferred to pulmonary system
- Symptoms of HF may develop
OBSTRUCTIVE Hypertrophic Cardiomyopathy

- 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
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>

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
### Hypertrophic Cardiomyopathy Treatment

<table>
<thead>
<tr>
<th>Beta Blockers</th>
<th>Calcium Channel Blockers</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st choice (with or without HOCM)</td>
<td>If Beta Blocker not effective</td>
</tr>
<tr>
<td>Symptomatic benefit / improved exercise tolerance</td>
<td>Decrease LV wall tension</td>
</tr>
<tr>
<td>Decreases HR</td>
<td>Decreases HR</td>
</tr>
<tr>
<td>Improves LV relaxation</td>
<td>Diltiazem or Verapamil (no nifedipine D/T vasodilatation)</td>
</tr>
<tr>
<td>Helps control arrhythmias</td>
<td></td>
</tr>
</tbody>
</table>

### Hypertrophic Cardiomyopathy Treatment

<table>
<thead>
<tr>
<th>Disopyramide</th>
<th>Antiarrhythmic Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Negative inotrope</td>
<td>Atrial Fibrillation</td>
</tr>
<tr>
<td>Class I antiarrhythmic</td>
<td>Most common arrhythmia</td>
</tr>
<tr>
<td>Use with BB to treat LV outflow track obstruction</td>
<td>Poorly tolerated</td>
</tr>
<tr>
<td>Assists in HR control</td>
<td>Anticoagulation</td>
</tr>
<tr>
<td>May cause ventricular arrhythmias – monitor QT</td>
<td>Amiodarone or sotold</td>
</tr>
<tr>
<td></td>
<td>Obstructive or non-obstructive OK</td>
</tr>
<tr>
<td></td>
<td>Ventricular or atrial arrhythmias</td>
</tr>
</tbody>
</table>
## Hypertrophic Cardiomyopathy Treatment

### Other Medications

<table>
<thead>
<tr>
<th>Diuretics</th>
<th>ACE Inhibitors and NTG</th>
</tr>
</thead>
<tbody>
<tr>
<td>• With caution</td>
<td>• Avoided in HOCM</td>
</tr>
</tbody>
</table>

### Positive Inotropes

| • Strictly avoid any medication that increases contractility in HOCM |

### Pregnancy

- Not restricted in non-obstructive disease

## Hypertrophic Cardiomyopathy Treatment

### 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

SCD primary cause of shortened life span.

Arrhythmogenic Cardiomyopathy

Outcomes

Arrhythmogenic Cardiomyopathy

Outcomes

Arrhythmogenic Cardiomyopathy

Outcomes

Arrhythmogenic Cardiomyopathy

Outcomes
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
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>
<td></td>
</tr>
</tbody>
</table>

V3
Arrhythmogenic Cardiomyopathy

Treatment

- No cure
- Goal: Manage arrhythmias
- Antiarrhythmics
- 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

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>Echo</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>
</tr>
</tbody>
</table>
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 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
BE THE BEST THAT YOU CAN BE EVERY DAY. YOUR PATIENTS ARE COUNTING ON IT!

Nurses Make a Difference

Slides will be available next week at www.cardionursing.com