Cardiomyopathy: The Good, the Bad....and the Insurable?

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Objectives

- Overview of common cardiomyopathies
  - Dilated
  - Hypertrophic

- Distinguish factors that places a case into favorable vs. less favorable risk category

- High-level review of rare cardiomyopathies
  - Takotsubo
  - Arrhythmogenic RV dysplasia
Defining Cardiomyopathies

AHA 2006: “A heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilatation and are due to a variety of causes that frequently are genetic.”
Cardiomyopathies: WHO

- Dilated (DCM, Congestive)
- Hypertrophic (IHSS, HCM, ASH)
- Rare (unclassified)
  - Takotsubo Cardiomyopathy
  - Arrhythmogenic Right Ventricular Dysplasia (ARVD)
Case Study - What’s the risk?

30 Y/O Male nonsmoker

- Medical Questionnaire: 2011 Heart issues caused by the flu.

Medical Records:
- 3/11 presented to ER for SOB w/CP. Had recent upper respiratory infection. Echocardiogram: LV mildly dilated. LV function was mild-moderately impaired w/EF estimated around 40%. Mild biatrial enlargement. Started on ramipril (Altace)

  - 10/11: repeat echo: EF 48%, normal biatrial size. Mild LV dysfunction and dilation, mild MR/TR

  - 11/12 echo: EF 50%, mild MR/TR, LV normal size with LV function lower limits of normal

  - 12/13 echo: essentially unchanged with EF 50%
Dilated Cardiomyopathy (DCM)
Dilated Cardiomyopathy (DCM)

- Key features
  - Dilated (enlarged) ventricle
  - Systolic dysfunction
    - Reduced ejection fraction (EF)
    - Wall motion abnormalities/global hypokinesis
- Heart muscle weakens leading to heart failure
DCM Etiology

• **Primary:**
  - Idiopathic - unknown etiology
  - Genetic (20-~50% familial link)

• **Secondary: (acquired)**
  - Tachycardia- induced (SVT)
  - Peri-partum
  - Ischemic
  - Inflammatory (infectious and non-infectious)
  - Toxic (chronic ETOH, chemotherapy agents)
DCM Epidemiology/Prevalence

- Prevalence: 36 per 100,000 of the general population
- Ages 20 – 60
  - Male:female (3:1)
- Common cause of heart failure
- Most common indication for individuals referred for cardiac transplantation
DCM Prognosis/ Mortality Risk

- Progressive disorder: Prognosis depends on etiology
- Early diagnosis, asymptomatic cases, and those with reversed LV remodeling show a good prognosis
- Poor once heart failure has occurred
- Risk of sudden death is consistent with LVEF (<40%)
- 20% mortality at 5 years from symptom onset
DCM: Signs and Symptoms

Symptoms of heart failure

- Dyspnea (rest, exertion)
- Edema, abdominal pain
- Fatigue and weakness
  - Low cardiac output
- Hypotension, tachycardia

EKG

- ST/T changes, poor R wave progression, Q waves, CLBBB

Arrhythmias

- Atrial fibrillation, complex PVCs, sudden cardiac death (SCD)
DCM Diagnostic Studies

• 2D-doppler echocardiogram (gold standard)
  – systolic dysfunction w/low EF (< 40 - 45%)
  – *global* hypokinesis, diffuse LV wall motion abnormalities, global dysfunction
  – increased LV internal diameter in diastole (LVIDd >5.8 cm)

• NT-pro BNP

• Chest x-ray

• Cardiac catheterization
  – age > 40, ischemia history, high risk profile
DCM Management

• Medical Therapy
  – ACE inhibitors (lisinopril, ramipril)
  – Angiotensin II receptor blockers - ARBS (losartan, olmesartan)
  – Beta blockers (Coreg)
  – Digoxin, diuretics
  – Antiarrhythmics
  – Anticoagulation
    • A-fib
    • EF < 30%,
    • previous embolic event

• Other treatments - severe disease
  – Implantable defibrillator (ICD)
  – Biventricular pacemaker
  – Cardiac resynchronization therapy (CRT)
DCM Management (cont.)

Uninsurable:

- **Ventricular Assist Device (VAD)**
- **Cardiac Transplantation**
  - DCM the most common reason
  - Survival at one year is ~88%
  - 5 years ~72%
# Underwriting Tips for DCM

## Favorable And Unfavorable Features of DCM

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<thead>
<tr>
<th></th>
<th>Favorable</th>
<th>Unfavorable</th>
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<tbody>
<tr>
<td><strong>Types of DCM</strong></td>
<td><strong>Reversible</strong>&lt;br&gt;• Peripartum&lt;br&gt;• Tachycardia-induced</td>
<td>Variable outcomes&lt;br&gt;• Idiopathic&lt;br&gt;• Genetic&lt;br&gt;• Viral</td>
</tr>
<tr>
<td><strong>Echocardiogram findings</strong></td>
<td>• EF of ≥ 50 %&lt;br&gt;• LVID(d) &lt; 5.8 cm&lt;br&gt;• Normal LV wall motion</td>
<td>• Severe if &lt; 40%&lt;br&gt;• LVIDd of 5.8 cm or greater&lt;br&gt;• Global hypokinesis</td>
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<tr>
<td><strong>Signs and Symptoms</strong></td>
<td>No signs or symptoms or well controlled on treatment</td>
<td>Signs and symptoms of heart failure (dyspnea, edema, exercise intolerance, chest pain)</td>
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<tr>
<td><strong>Treatment</strong></td>
<td>• No current treatment&lt;br&gt;• Treatment limited to ACE inhibitors, ARBS, and beta-blockers</td>
<td>• Diuretics&lt;br&gt;• Antiarrhythmics&lt;br&gt;• Anticoagulant (warfarin)&lt;br&gt;• Surgery</td>
</tr>
<tr>
<td><strong>Family history of sudden cardiac death (SCD)</strong></td>
<td>None</td>
<td>+FH of sudden cardiac death (SCD) related to CM in parent or sibling</td>
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Case Study - What’s the risk?

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- Medical Questionnaire: 2009 Heart issues caused by the flu

Medical Records
- 3/11 presented to ER for SOB w/CP. Had recent upper respiratory infection. Echocardiogram: LV mildly dilated. LV function was mildly-moderately impaired w/EF estimated around 40%. Mild biatrial enlargement. Started on ramipril (Altace)
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Cardiologist’s diagnosis: Stable non-ischemic cardiomyopathy. Remains asymptomatic and continue ramipril

Favorable? Unfavorable?
Case Study- What's the risk?

38 Y/O Male nonsmoker

- Medical Questionnaire: Mild Cardiomyopathy

- Medical Records:
  - 7/12 resting echo:
    - Mild to moderate asymmetrical hypertrophy (ASH) with septal thickness of 1.4 to 1.5 cm
    - Diastolic relaxation abnormality
    - Systolic anterior motion (SAM) of mitral valve & no significant LVOT gradient
  
  - 8/13 resting echo:
    - Mild to moderate ASH with septal thickness of 1.3-1.5
    - Diastolic dysfunction
    - Systolic anterior motion (SAM) of mitral valve without an increase in LV outflow tract gradient. Trace MR
    - No significant change from 7/12 echo

  - GXT: No symptoms with 13.5 mets achieved - Physiologic B/P and HR response with exercise, no arrhythmias

  - Meds: Toprol XL- 25 mg a day
HYPERTROPHIC CARDIOMYOPATHY (HCM)
Asymmetrical Septal Hypertrophy (ASH)

LV outflow tract (LVOT) obstruction, especially with LVOT gradient > 30 mm Hg at rest or provoked
HCM Epidemiology/Etiology

• Prevalence: 1 in 500 of the general population
• Male = Females
• Etiology: known genetic risk
  – 60-70% familial (autosomal dominant)
  – 30-40% sporadic mutations
  – Variable expression
• Mutations cause abnormal heart muscle cells
  – Cell enlargement and myocardial disarray
  – Substrate for EKG changes and arrhythmias

[Images of normal and disarray muscle structures]
HCM Prognosis and Mortality Risk

- Annual mortality rate:
  - Adults – 1-2 %
  - Children/Adolescents – 2- 6%

- It is the most common cause of sudden cardiac death (SCD) in young people in US
  - 36% of deaths in athletes <35 years

- Sustained V- Tach and V- Fib are the most likely mechanism of syncope and SCD
HCM Risk Factors for Poor Outcomes

- Syncope history (no attributable cause)
- Family history of SCD in 1st degree relative
- Early age of diagnosis (< 14 years)
- Non-sustained ventricular tachycardia
- Severe hypertrophy of ≥3.0cm
HCM: Sign and Symptoms

- Asymptomatic: echo results
- Symptomatic:
  - dyspnea (95%)
  - chest pain (75%)
  - syncope (near syncope)
  - fatigue
  - palpitations
Diagnostic Studies:

EKG findings:
- Abnormal in 75-90 % of cases
- LVH, strain pattern
- Abnormal ST/T’s, giant T wave inversions in lateral precordial leads
- Bundle branch blocks
- Atrial Fib and Ventricular arrhythmias
HCM Resting Echocardiogram

gold standard

- LV hypertrophy of septum and posterior wall
  - IVSd /LVPWd greater > 1.4 cm
- Asymmetrical Septal Hypertrophy (ASH) at least 1.4 times the thickness of posterior wall may be present
- Diastolic dysfunction (abnormal relaxation)
- Systolic anterior motion of mitral valve (SAM) may be present
- Outflow tract obstruction (LVOT) may be present
  - > 30 mm Hg at rest or provoked with exercise
HCM Management

- Beta-blockers: slow HR and allow LV relaxation and better filling
- Calcium Channel-blockers
- Antiarrhythmic drugs – low dose amiodarone
- Prophylactic ICD
- Septal Myectomy
- Percutaneous Alcohol Septal Ablation
- Cardiac Transplant
HCM Management (cont.)

- **Life style changes:**
  - Avoid extreme activity and competitive sports (i.e. basketball, weight lifting)

- **Family screening:**
  - EKG
  - Resting echo
  - Genetic testing
## Favorable And Unfavorable Features of HCM

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| **Echocardiogram findings** | • IVS(d) or LVID (d) ≤1.4 cm  
• No or mild diastolic dysfunction  
• No outflow tract obstruction | • IVS(d) or LVID (d) >2.0 cm  
• Moderate or severe diastolic function  
• LV outflow tract gradient ≥ 30 mmHg at rest or provoked |
| **EKG findings**          | • Normal                                                                  | • Increased QRS voltage  
• Diffuse T waves changes  
• Atrial fibrillation  
• Ventricular arrhythmia (VT) |
| **Symptoms**              | • Asymptomatic                                                            | • Dyspnea  
• Chest pain  
• Palpitations |
| **Treatment**             | • Treatment limited to beta-blockers or calcium channel blockers         | • Diuretics  
• Antiarrhythmics/ anticoagulant (warfarin)  
• ICD  
• Surgery |
| **Risk factors for sudden cardiac death (SCD)** | • None                                                                    | • Syncope  
• Nonsustained ventricular tachycardia history  
• Family history of sudden cardiac death in parent and sibling |
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Favorable? Unfavorable?
Case Study - What’s the risk?

49 Y/O Male

- MHQ: 10 years ago diagnosed with Apical HCM

- Medical Records:
  - Deeply inverted T waves prompted the initial CV evaluation
  - Numerous diagnostic studies performed:
    - Multiple negative perfusion stress tests
    - EBCT calcium score of zero
    - Cardiac MRI demonstrated focal asymmetric apical hypertrophy with no LVOT obstruction – subsequent MRI 3 yrs ago -> no progression
    - Current resting echo shows hypertrophy of apical wall with basal septal measurement of 1.4 cm and diastolic dysfunction

- Meds: atorvastatin

Favorable? Unfavorable?
Apical Hypertrophic Cardiomyopathy
## Athletic Heart: Is it really?

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<th>HCM</th>
<th>Athletic Heart</th>
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<tbody>
<tr>
<td>• Echo</td>
<td>• Occurs in <em>elite</em> athletes</td>
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<tr>
<td>• Can be <em>asymmetric</em></td>
<td>• <em>Concentric &amp; regresses</em></td>
</tr>
<tr>
<td>• Wall thickness: &gt; 1.5 cm</td>
<td>• &lt; 1.4 cm</td>
</tr>
<tr>
<td>• LVIDd normal or small: &lt; 4.5 cm</td>
<td>• &lt; 6.3 cm</td>
</tr>
<tr>
<td>• <em>Diastolic dysfunction</em></td>
<td>• <em>Normal systolic and diastolic function</em></td>
</tr>
<tr>
<td>• No change or regression</td>
<td>• Regression of echo finding</td>
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Questions???
Takotsubo Cardiomyopathy
(Stress –Induced Cardiomyopathy)

- Ventriculogram and/or echocardiography show akinesis or dyskinesia of apical and mid segments of left ventricle
- Average EF of 20 – 49%, wall motion abnormalities
Takotsubo Cardiomyopathy is *Unique*

- **Women:** 90% of cases
- **Postmenopausal:** 90% (mean age: 68.5)
- **Triggered by an extreme emotional or physical stressor**
- **Treatment**
  - Treat LV dysfunction with acute MI protocol
- **Prognosis**
  - 95% complete recovery within 4-8 weeks
  - 3% recurrence
  - 1% mortality
Arrhythmogenic Right Ventricular Dysplasia (ARVD)

- Replacement of myocardium muscle by fibro-fatty scars.
- Right ventricle the most affected.
ARVD Epidemiology/Etiology

- Prevalence: 1 in 5000
- 3:1 men: women
- Etiology:
  - Genetic
  - Possible viral infection/sporadic mutation
- Arrhythmia the most prominent symptom.
  - 50% of individuals present with ventricular arrhythmias
  - Syncope or SCD: Up to 80%
Diagnostic Studies

- **Echocardiogram**
  - RV dilatation, regional wall motion abnormalities

- **Cardiac MRI** *(gold standard)*
  - fibro-fatty infiltration of RV myocardium

- **Right Heart Catheterization** *
  - Myocardial biopsy
  - Sensitivity low (< 20% - 67%) due to segmental nature of disease
ARVD Management

• Antiarrhythmic drugs
• ACE inhibitors
• Catheter ablation for ventricular arrhythmias
• ICD
• Heart Transplantation
• Screen family members