Cardiomyopathies & Heart Failure

Cardiomyopathy – Definition

Cardio | Myopathy
---|---
| → Heart

- **Myo**– Greek μυο
  - muscle
- **Pathos** – pathy Greek
  - suffering
- Muscle disease in which the muscle fibers do not function resulting in Muscular weakness

Cardiomyopathy vs Heart Failure

| Cardiomyopathy | Heart Failure |
---|---|
| → Primary disorder of cardiac muscle causing abnormal myocardial performance
  - Associated with mechanical and/or electrical dysfunction
  - Usually exhibits ventricular hypertrophy or dilation
  - Often leads to progressive heart failure |
| → Complex clinical syndrome
  - Develops from any cardiac disorder that impairs the ability of the ventricle to fill or eject adequately
  - Pathologic state in which the heart is unable to pump enough oxygenate blood to meet the metabolic needs of the body |

Test Blue print

| CMC | CSC |
---|---|
| Cardiac Patient Care Problems (476) | Complications of Heart Surgery & interventions |
| - Acute Coronary Syndrome
  - Dysrhythmia |
| - Heart Failure |
| - Other Cardiac Issues
  - Cardiomyopathies
  - Pulmonary Hypertension
  - Vascular Issues |
  - Right sided failure
  - Pulmonary Hypertension
  - Vasopressors/vasodilators
  - Ventricular Assist Devices |

Cardiomyopathy leads to Heart Failure

- In the past, left ventricular dysfunction and cardiomyopathy were sometimes thought of as equivalent to HF
- But rather they are structural or functional reasons for the development of HF

Cardiomyopathies

- Primary disorder of cardiac muscle causing abnormal myocardial performance
Cardiomyopathy

- How can one name mean so many different things?
- One name – Many causes
- Work in a group and write down as many cardiomyopathies that you can think of.

Cardiomyopathies

- Hypertrophic
- Stressed Induced (Takotsubo)
- Restrictive
- Dilated
  - Idiopathic
  - Valvular
  - Genetic
  - Ischemic
  - Hypertensive
  - Myocarditis (Infection/Inflammatory)
  - Chemotherapy induced
  - Periparum syndrome related to toxemia
  - Cardiotoxic induced (effects of drugs or alcohol)

Cardiomyopathies

- Hypertrophic
- Dilated (ischemic and nonischemic)
- Stressed Induced (Takotsubo)
- Restrictive *

* Not on CMC test blue print

Hypertrophic Cardiomyopathy (HCM)

- Myocardial hypertrophy without the presence of associated hemodynamic stress (no 7 in afterload)
- Hypertrophy of the heart muscle including the septum and ventricular free wall
- Previously called IHSS – idiopathic hypertrophic subaortic stenosis
- Leading cause of death in athletics < 35 y/o

Hypertrophic Obstructive Cardiomyopathy (HOCM)

- Subgroup of patients with HCM develop obstruction
- Once obstruction occurs it is called: Hypertrophic obstructive cardiomyopathy (HOCM)
Hypertrophic Cardiomyopathy (HCM)

**Pathophysiology**
1. Hypertrophy of heart muscle including septum and ventricular free wall.
2. Rigid, noncompliant ventricles do not stretch.
3. Causes diastolic dysfunction.
4. \( \downarrow \) preload and cardiac output.
5. Left atrial dilatation from inability to empty LA.
6. Mitral regurgitation occurs from papillary muscles and mitral valve pulled out of alignment.

Hypertrophy of LV, septum and ventricular wall, LA enlargement, MR.

**Hypertrophic Obstructive Cardiomyopathy (HOCM)**

**Pathophysiology**
1. With severe hypertrophy, left ventricular outflow tract becomes obstructed — especially with \( \uparrow \) contractility from \( \uparrow \) catecholamines (exercise).
2. Decrease in blood flow to coronary arteries (angina) and brain (syncope).
3. May result in sudden cardiac death.

LV outflow tract obstructed – syncope, sudden death.

**Hypertrophic Cardiomyopathy (HCM)**

**Causes**
- Probably genetic.
- May occur as early as the 1st year of life.
- Develops most commonly during adolescence.
- Hypertrophy manifests after age 20.
- Diagnosis is usually made by age 25.
- Persons with normal echo and EKG after 25 y/o are unlikely to develop HCM.

Clinical Presentation
- Sudden cardiac death often the first presentation.
- Often diagnosed incidentally as may be asymptomatic.
- Dyspnea on exertion – relieves with rest.
- Syncope on exertion or rest.
- Palpitations.
- Jugular venous palpitation.
- Associated with prominent "a" wave secondary to \( \downarrow \) RV compliance.
- Heart Sounds.
- Harsh systolic murmur LSB.
- Murmur increases with movement.
- S4 from LVH.
- EKG.
- Repolarization abnormalities.
- Atrial enlargement (large p waves).
- Pathological Q waves – inferior leads.

Sudden cardiac death often the first presentation.

**Normal Echocardiogram**

**HOCM**

<table>
<thead>
<tr>
<th>Diastole</th>
<th>Systole</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image" alt="Diastole Image" /></td>
<td><img src="image" alt="Systole Image" /></td>
</tr>
</tbody>
</table>
Hypertrophic Cardiomyopathy (HCM)

Clinical Management

- Symptom relief
- Prevention of sudden cardiac death
- Beta blockers for chest pain and dyspnea with exertion in HOCM
- Disopyramide (Norpace and Rythmodan) - reduces obstruction by ↓ inotropic action
- Verapamil - used only for mild obstruction
- Annual kick more essential than normal
- If symptoms persist
  - Ventricular Septal myectomy - removal of muscle from septum.
  - Percutaneous septal alcohol ablation - causes controlled septal MI
- ICD
  - History of cardiac arrest or sustained ventricular dysrhythmias
  - Multiple clinical risk factors
  - Counseling & genetic testing
  - Restrict from intense competitive sports
  - SBE prophylaxis for HOCM

Symptom relief & prevention of cardiac arrest

Medications

- Beta blockers
  - 1st choice
  - Increase exercise tolerance
  - ↓ heart rate
  - Improves LV relaxation
  - Control of arrhythmias
- Disopyramide (Norpace and Rythmodan)
  - Negative inotrope (↓ contractility)
  - Used with BB to treat LV outflow tract obstruction
  - ↓ SAM
  - Assists with HR control
  - Monitor QT - may cause arrhythmias
  - Class I antiarrhythmic
- Calcium Channel Blockers
  - Verapamil or diltiazem
  - Used only for mild obstruction
  - Use if BB ineffective
  - ↓ LV wall tension
  - Negative inotrope
  - ↓ LHR
- Antiarrhythmic medications
  - Treat A fib and/or vent arrhythmias
  - Amiodarone or sotalol

Disopyramide may cause uncomfortable anticholinergic side effects and may enhance the hypoglycemic effect of gliclazide, insulin, and metformin. SA

Case study

- 25 y/o female presents to ED complaining of significant chest pain that was continuous for several hours.
- What diagnostic test do you want?

Should you go to Cath lab?
More History

- History of palpitations, dyspnea on mild exertion and recurrent syncope.
- Three episodes of syncope in last week
- Hx of seizures
- Systolic murmur III/IV
- Meds:
  - ASA
  - Metoprolol Tartrate
  - Phenytoin
  - Valium, prn
  - Zolpidem

- EF 67%
- Troponin 0.5
- What diagnosis do you think?

Diagnosis is Hypertrophic Obstructive Cardiomyopathy.

Septal Myectomy
Dilated Cardiomyopathy

- Enlarged, dilated cardiac chamber
- Can affect one or all four chambers
- As chamber enlarges, its ability to contract becomes impaired, resulting in systolic dysfunction
- Most common cause of HF

Dilated Cardiomyopathy (DCM)

Clinical Presentation
- Hallmark signs
  - Weakness
  - Fatigue
  - Decreased activity tolerance
  - Biventricular hypertrophy
  - Bialtral enlargement
  - Decreased EF
  - S3, Systolic murmur

Symptoms develop slowly; patient adjusts ADLs without realizing it.

Dilated Cardiomyopathy

Causes
- Idiopathic
- Valvular
- Genetic
- Ischemic
- Hypertensive
- Myocarditis (Infection/Inflammatory)
- Chemotherapy
- Periparum syndrome related to toxemia
- Cardiotoxic effects of drugs or alcohol

Primary vs Secondary Dilated Cardiomyopathy

Primary
- Idiopathic Dilated Cardiomyopathy (IDC)

Secondary
- Ischemic Dilated Cardiomyopathy
- Hypertensive Dilated Cardiomyopathy
- Valvular Dilated Cardiomyopathy
- Anthracycline Dilated Cardiomyopathy
- Peripartum Dilated Cardiomyopathy
- Alcoholic Dilated Cardiomyopathy

Dilated Cardiomyopathy

Clinical Management
- Five year survival = 50%
- Eliminate the cause
- Medical treatment same as for HF
- Symptom relief
  - Preload reduction
  - Afterload reduction
  - Increased contractility

Eliminate the cause and symptom relief
### Idiopathic Dilated Cardiomyopathy (IDC)

<table>
<thead>
<tr>
<th>Causes</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>› 50% of IDC is familial</td>
<td>› ACE inhibitors</td>
</tr>
<tr>
<td>› Suspected when other causes are excluded</td>
<td>› Beta blockers</td>
</tr>
<tr>
<td>◦ CAD</td>
<td>› Anticoagulation is required due to the risk of thromboembolism</td>
</tr>
<tr>
<td>◦ Thyroid disease</td>
<td>› Improvement of LV function is often better in IDC than in patients with</td>
</tr>
<tr>
<td>◦ Valvular abnormalities</td>
<td>IDC (idiopathic)</td>
</tr>
<tr>
<td>◦ Infiltrative causes</td>
<td>Improvement of LV function is often better in IDC than in patients with</td>
</tr>
<tr>
<td>◦ Hypertension</td>
<td>IDC (idiopathic)</td>
</tr>
<tr>
<td>◦ Alcohol</td>
<td>›ampoacal and improves wall stress but not depressed LVP</td>
</tr>
<tr>
<td></td>
<td>› ACE inhibitors</td>
</tr>
<tr>
<td></td>
<td>› Beta blockers</td>
</tr>
<tr>
<td></td>
<td>› Anticoagulation is required due to the risk of thromboembolism</td>
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<tr>
<td></td>
<td>› Improvement of LV function is often better in IDC than in patients with</td>
</tr>
<tr>
<td></td>
<td>IDC (idiopathic)</td>
</tr>
<tr>
<td></td>
<td>› Amiodarone to prevent dysrhythmias</td>
</tr>
<tr>
<td></td>
<td>› Monitor electrolytes</td>
</tr>
<tr>
<td></td>
<td>› Prognosis is worse for IDM than nonischemic cardiomyopathy.</td>
</tr>
</tbody>
</table>

### Ischemic Dilated Cardiomyopathy (IDM)

<table>
<thead>
<tr>
<th>Causes</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>› Most common type of dilated cardiomyopathy</td>
<td>› ACE inhibitors</td>
</tr>
<tr>
<td>› Occurs when CAD or ischemic heart disease causes remodeling of the</td>
<td>› Beta blockers</td>
</tr>
<tr>
<td>LV with an associated reduction of EF</td>
<td>› Anticoagulation is required due to the risk of thromboembolism</td>
</tr>
<tr>
<td>› Remodeling is the compensatory response of the ventricles to improve</td>
<td>› ICD</td>
</tr>
<tr>
<td>its function</td>
<td>› Amiodarone to prevent dysrhythmias</td>
</tr>
<tr>
<td>◦ Harms ventricular muscle</td>
<td>› Monitor electrolytes</td>
</tr>
<tr>
<td>◦ Worsens stroke volume</td>
<td>› Prognosis is worse for IDM than nonischemic cardiomyopathy.</td>
</tr>
<tr>
<td>◦ Develops ventricular dilation</td>
<td></td>
</tr>
<tr>
<td>◦ Decreases EF</td>
<td></td>
</tr>
</tbody>
</table>

### Hypertensive Dilated Cardiomyopathy

<table>
<thead>
<tr>
<th>Causes</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>› Diagnosed when systolic function remains depressed despite adequate</td>
<td>› Same as IDM (ischemic)</td>
</tr>
<tr>
<td>treatment of hypertension</td>
<td>› Afterload reduction is the most important goal</td>
</tr>
<tr>
<td>› Myocardial systolic function is depressed out of proportion to the</td>
<td>› Antihypertensive vasodilators</td>
</tr>
<tr>
<td>increase in wall stress</td>
<td>› Amiodipine (Norvasc, besylate, mesylate or maleate)</td>
</tr>
<tr>
<td>› Prognosis is influenced by other comorbidities</td>
<td>› Alpha–blocking agents</td>
</tr>
<tr>
<td></td>
<td>› Alfuzosin (Uroxatral)</td>
</tr>
<tr>
<td></td>
<td>› Doxazosin (Cardura)</td>
</tr>
<tr>
<td></td>
<td>› Prazosin (Minipress)</td>
</tr>
<tr>
<td></td>
<td>› Terazosin</td>
</tr>
<tr>
<td></td>
<td>› Tamsulosin (Flomax)</td>
</tr>
<tr>
<td>Afterload reduction – Alpha–blocking agents</td>
<td></td>
</tr>
</tbody>
</table>

### Valvular Dilated Cardiomyopathy

<table>
<thead>
<tr>
<th>Causes</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>› Myocardial systolic function is depressed out of proportion to the</td>
<td>› Valve replacement or repair</td>
</tr>
<tr>
<td>increase in wall stress secondary to valvular abnormalities</td>
<td>› improves wall stress but not depressed LVP</td>
</tr>
<tr>
<td>› Most caused by left sided valves</td>
<td>› ACEI &amp; BB</td>
</tr>
<tr>
<td>◦ MR &amp; AR</td>
<td>› Aggressive afterload reduction</td>
</tr>
<tr>
<td>◦ AS less common cause</td>
<td>› Hydralazine</td>
</tr>
<tr>
<td></td>
<td>› Nitrates</td>
</tr>
<tr>
<td></td>
<td>› nitroglycerin, isosorbide dinitrate, isosorbide mononitrate.</td>
</tr>
<tr>
<td></td>
<td>› With AR– calcium channel blockers</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Peripartum Dilated Cardiomyopathy

<table>
<thead>
<tr>
<th>Causes</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>› Occurs when myocardial systolic dysfunction occurs during the last</td>
<td>› Treatment is aggressive &amp; consistent with IDC (idiopathic)</td>
</tr>
<tr>
<td>trimester of pregnancy or within 6 months of childbearing.</td>
<td>› 50% will recover completely</td>
</tr>
<tr>
<td>› Outcomes are better with peripartum than with other dilated</td>
<td>› Small minority will need transplant</td>
</tr>
<tr>
<td>cardiomyopathies</td>
<td></td>
</tr>
</tbody>
</table>

### Case Study

7/23/2016
37 y/o African American presents to ED with Shortness of Breath

- BP 152/102
- HR 100
- RR 28
- T 98.9 oral
- SpO2 88% room air
- Loose cough
- Coarse rhonchi and scattered wheezes
- 2+ pitting edema

PMH

- Asthma
- Pancreatitis
- Diabetes, type II (diet controlled)
- Smokes 4 cigarettes/day x 15 years
- Cocaine use in the past
- Sister and daughter → sickle cell anemia

Labs

- WBC 8.2
- Hgb 10.2
- HCT 32.7
- Glucose 79
- Potassium 3.2
- Creatinine 1.2
- Troponin 0.06

Symptoms

- C/o not breathing “normally” with increased effort and shortness of breath
- Moderate SOB at rest
- Decreased exercise capacity with exertional SOB
- Started two weeks ago and getting worse
- Albuterol treatment taken 1 hour ago at home

Admission EKG (37 y/o)

Admission CXR

A. Pneumonia
B. Pulmonary Edema
C. Cardiomyopathy with Pulmonary Edema
D. All of the above
More history
- C-section two weeks ago for failure to progress
  - 4th child
- SOB and wheezing have become progressively worse since delivery
- Low grade fever past few days
- Productive cough

Admission Diagnosis
- Bilateral pneumonia vs CHF
- ? Peripartum cardiomyopathy
- Treatments
  - Albuterol/Atrovent nebs
  - Prednisone 50 mg po
  - Methylprednisolone 125 mg IV
  - Lasix 60 mg IV
  - Any concerns??
  - Ceftriazone 1 gm IV
  - Azithromycin 500 mg IV
  - Echocardiogram

Echo
- LV mildly dilated
- EF 35%
- Mild to moderate aortic regurgitation
- Moderate mitral regurgitation
- RV mildly enlarged

Discharged 2 days later
- Community acquired pneumonia
- Asthma exacerbation
- Peripartum cardiomyopathy
- Anemia secondary to postpartum state
- Lisinopril
- Metoprolol tartrate
- Prednisone
- Flovent Diskus
- Albuterol
- Augmentin
- Ferrous Sulfate

CXR three weeks later

Peripartum Cardiomyopathy (PPCM)
Also called pregnancy associated cardiomyopathy
Peripartum Cardiomyopathy (PPCM)

5th leading cause of mortality during the pregnancy period


Peripartum Cardiomyopathy (PPCM)

- Incidence per live births
  - 1:4350 USA – 10 years ago
  - 1:2399 USA – 2011
  - ↑ maternal age, ↑ multifetal pregnancy, ↑ recognition PPCM
  - 1:1000 South Africa
  - 1:300 Haiti
  - 1:100 Nigeria
- Cause – Unknown
- Usually occurs with first or second pregnancy


Pregnancy

- High output state
- 30% decrease in systemic vascular resistance
- 30–40% increase in cardiac output by 2nd and 3rd trimester
- Changes may not resolve completely until 12 weeks postpartum

Source: Garg, J et al. Peripartum Cardiomyopathy Cardiology in Review 2019

PPCM Risk Factors

- Advancing maternal age > 30 years
- Extreme age (very young or advanced age)
- African descent
- Multi-fetal gestation
- History of preeclampsia, eclampsia, postpartum hypertension
- Long term (> 4 weeks) use of beta adrenergic agonists (terbutaline) for preterm labor suppression
  - Tocolytic agents used > 4 weeks have higher incidence of pulmonary edema (terbutaline, salbutamol, ritodrine, and magnesium sulfate)
- Maternal cocaine abuse


PPCM signs and symptoms

- Similar to other forms of systolic HF
  - Dyspnea – most common
  - Tachycardia
  - Early sign
  - Cough
  - Orthopnea
  - Paroxysmal nocturnal dyspnea (PND)
  - Pedal edema
  - Nonspecific fatigue
  - Hemoptysis
- High clinical suspicion
  - Elevated jugular venous pressure
  - Displaced apical impulse
  - S3
  - Murmurs from tricuspid or mitral regurgitation

Source: Tsang, W Cardiomyopathy in the last month or the first five months after pregnancy

1. Cardiomyopathy in the last month or the first five months after pregnancy
2. Absence of another identifiable cause of HF
3. EF < 45%
   - LV may or may not be dilated

PPCM symptoms

- Often missed or delayed
- Similar signs and symptoms of normal pregnancy

Comparison of Symptoms

<table>
<thead>
<tr>
<th>Preeclampsia</th>
<th>Similar Symptoms</th>
<th>PPCM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Edema</td>
<td>Weight gain</td>
<td>Dyssynea</td>
</tr>
<tr>
<td>Proteinuria</td>
<td>Tachycardia</td>
<td>Cough</td>
</tr>
<tr>
<td>Headache</td>
<td>Adventitious breath sounds</td>
<td>Chest pain</td>
</tr>
<tr>
<td>Blurred vision</td>
<td>Hypertension</td>
<td>Palpitations</td>
</tr>
<tr>
<td>Decreased renal perfusion</td>
<td>Fatigue</td>
<td>Third heart sound S3</td>
</tr>
<tr>
<td>Hyperactive DTRs</td>
<td></td>
<td>Jugular venous distension</td>
</tr>
<tr>
<td>Impaired hepatic function</td>
<td></td>
<td>Tachypnea</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Murmur</td>
</tr>
</tbody>
</table>

Early Recognition is Key!

- For patients who develop
  - Dyspnea
  - Increasing blood pressure
  - Increased edema or edema that doesn’t decrease
  - Chest pain
  - Tachycardia
  - Nonproductive cough
- Detailed, serial, frequent nursing assessments with accurate documentation
- Comprehensive and thorough communication with provider
- BNP, Echo
- Cardiology consult

PPCM Diagnosis

Three Clinical criteria
1. Development of HF end of pregnancy or first 5 months post delivery
2. Absence of other identifiable cause of HF
3. EF < 45%

PPCM Prognosis

- Related to the severity of cardiac dysfunction and return of ventricular function
- Outcomes are better with peripartum than with other dilated cardiomyopathies
- 50% will recover completely
- Small minority will need transplant

Outcome with a cardiomyopathy is related to the etiology

In a study of 1339 patients with a cardiomyopathy of various etiologies, the adjusted Kaplan-Meier estimates of survival is related to the underlying cause of cardiomyopathy; only ischaemic cardiomyopathy and cardiomyopathy due to causes for which survival was significantly different from that in patients with ischaemic cardiomyopathy are shown. The best estimate is in those with a peripartum cardiomyopathy and the worst outcome is in those with an infiltrative cardiomyopathy or that due to HCM infection.

Source: Tsang, W. Anticipatory Cardiomyopathy, Etiology and Prognosis. Retrieved Feb 9, 2011 from UpToDate
Causes

- Thiamine deficiencies
- Diagnosed when there is a history of sustained and heavy alcohol consumption and other causes of dilated cardiomyopathy are excluded.
- Toxic effects of alcohol are thought to cause the nonspecific changes in the myocardium.
- Thiamine deficiencies can compromise cardiac function.

Treatment

- Alcohol abstinence
- Same as for IDC (idiopathic)
- Prognosis is somewhat better than for IDC depending on the degree of myocardial impairment and alcohol abstinence.

Stressed Induced Cardiomyopathy

- Broken Heart Syndrome
- Takotsubo Cardiomyopathy
- A specific syndrome of stress-related reversible cardiomyopathy
- Mimics acute myocardial infarction without obstructive disease

Case Study

- 69 y/o female comes to ED with c/o of severe chest discomfort
- PMH: mild HTN and hyperlipidemia
- B/P 173/89, HR 91, RR 21
- SpO₂ 98% on 2 l/np

EKG on admission

- Rural hospital with no cath lab
- NTG 0.4 mg SL x 3 in 30 minutes
- ASA 81 mg po
- Metoprolol 25 mg po
- Retavase
More history....

- A few hours earlier in the same ED, her husband came in full arrest and was not able to be resuscitated

Labs on admission

- CK = 156
- CKMB = 10.7 ↑
- Myoglobin = 298 ↑
- Troponin I = 2.91 ↑
- BNP = 35

Cardiac Cath findings

- Markedly depressed LV function with ejection fraction = 5 – 10%
- Severe hypokinesis to akinesis of the distal 2/3 anterolateral, apical, and inferior walls.
- The basal segments contract vigorously giving it very Japanese amphora shape suggestive of Takotsubo cardiomyopathy

Management

- Transferred to CVICU
- No IABP due to hemodynamically stable and recent Retavase
- Diagnosis: Broken Heart Syndrome or Takatsubo cardiomyopathy

No relief of symptoms... Repeat EKG No improvement

Transported via helicopter to hospital with cardiac cath

Normal coronary anatomy - No CAD
Discharged the next day so she could attend her husband’s funeral

- Discharge medications
- Aldactone 25 mg every day
- Alprazolam 0.5 mg prn
- Altace 2.5 mg every day
- ASA 81 mg every day
- Coreg 6.35 mg every 12 hours
- Coumadin 5 mg po every day
- Lasix 20 mg every other day
- Lipitor 40 mg po at hs

6 weeks later

- EF 60%
- Patient doing well

Stressed Induced Cardiomyopathy

- Broken Heart Syndrome
- Takotsubo Cardiomyopathy

- A specific syndrome of stress-related reversible cardiomyopathy
- Mimics acute myocardial infarction without obstructive disease

Precipitating factors

Marked psychosocial or physical stress

Original name given “Takotsubo Cardiomyopathy”

- Takotsubo is the narrow-necked bulging container used by Japanese fisherman to trap octopus
- The shape of the takotsubo pot resembles the distorted ballooning ventricle.
- Also called: Transient Left Ventricular Apical Ballooning
Etiology

- Unclear etiology
- 1 – 2% of patients who have S/S AMI have apical ballooning (Japan & USA)
- 6–9 times more common in women
- 6% of women with AMI have apical ballooning
- Most often in postmenopausal women

Takotsubo Cardiomyopathy

Clinical Presentation

- Chest pain
- ST segment changes
- Cardiac biomarkers
  - Only moderately elevated
  - Do not follow the typical rise-fall-pattern seen with AMI

12 Lead EKG Variable findings

- ST segment elevation or depression usually in the precordial leads (V2 – V5)
- Reciprocal changes in the inferior leads may not occur
- Q waves usually do not develop or Q waves V3 – V6
- Deeply inverted T waves are common in the recovery period
- Markedly prolonged QT interval

Pathophysiology

- Marked systolic ballooning of the ventricular apex
- Hypercontractility of the base of the heart
- Now thought to be related to stunning of the myocardium related to excessive catecholamines
- Since preceded by increased psychosocial or physical stress suggest an association with ↑ SNS activity
- Catecholamines have a toxic effect on the myocardium

Catecholamine levels reported to be 7 – 34 times as high as the normal 2 – 3 elevation in classic AMI patients

A 12-lead electrocardiogram showing ST-segment elevations and T-wave inversions in the right precordial leads, which is a typical pattern observed in Takotsubo cardiomyopathy

Takotsubo Cardiomyopathy

Diagnosis

Cardiac Cath/Echo

- Systolic ballooning of the ventricle, akinetic or dyskinetic left ventricle
- Ejection fraction markedly decreased in the acute phase – as low as 14 – 40%
- No significant coronary artery disease to account for the marked left ventricular dysfunction

Nuclear stress testing

- Evidence of reversible myocardial injury

Low EF – no CAD – precipitating stressor

Normal LV on Echo

- Systole
- Diastole
Left ventriculogram in systole (3a) and diastole (3b) to illustrate the ballooning.

65-year-old female admitted to a local ED due to chest pain in the retrosternal region associated with worsening dyspnea. Referral of the symptoms, the patient reported a significant stress episode following a stressful quarrel with her husband.

Takotsubo Cardiomyopathy

**Diagnosis**

- Immediately difficult to differentiate between STEMI caused by thrombosis
- Suspect Takotsubo Cardiomyopathy when obstructive CAD is not present to explain the LV dysfunction
- Confirmation of diagnosis: typical octopus morphology of LV
- Stressor considered supportive evidence
- Complete resolution of LV dysfunction weeks after the event

Low EF - no CAD - precipitating stressor – octopus morphology

**Clinical Management**

- Management of cardiogenic shock
  - Vasopressors
  - Pacemaker
  - Intraaortic balloon pump (IABP)
  - Support until LV recovers
- Supportive management
  - Arrhythmias → antiarrhythmic drugs
  - Diuretics → pulmonary congestion
  - β Blockers, vasodilators, ACEI, vasococtrioters, IABP → left sided HF
  - Short term anticoagulant → prevent LV thrombi

No fibrinolysis — Supportive management

Takotsubo Cardiomyopathy

**Prognosis**

- Left ventricular function improves rapidly
- Often within 7 – 30 days
- EKG changes may be slower to resolve
- Generally favorable prognosis
- Mortality of 0 – 8%

LV function recovers

Takotsubo Cardiomyopathy

**Restrictive Cardiomyopathy (RC)**

- Restrictive filling and reduced diastolic volume of either or both ventricles
- Least common cardiomyopathy (5%)
Restrictive Cardiomyopathy (RC)

Pathophysiology

1. Disease process causes noncompliant ventricles
2. Noncompliant ventricles resist ventricular filling
3. $↓$ diastolic filling $⇒$ $↓$ blood volume $⇒$ $↓$ SV
4. Results in $↑$ blood volume and pressure in atria
5. Blood backs up into the lungs (left sided failure) and then in the venous circulation (right sided failure)
6. Disease process affects both ventricles
7. Ventricle size usually normal or slightly decreased
8. Systolic function is not usually affected

Heart becomes noncompliant and cannot stretch and fill

Characteristics

- Syndrome preceded by an initiating cardiovascular event (MI, hypertension, etc)
- On the cardiac continuum HF is an end event – represents the most severe manifestation of cardiovascular disease

Physiologic Changes in Dilated Cardiomyopathy

Heart Failure

Characteristics

1. Arises from alterations in systolic and diastolic dysfunction
   - Systolic Dysfunction
   - Diastolic Dysfunction
2. Systolic and Diastolic Dysfunction are progressive syndromes that develop over the course of many years
3. Heart Failure preferred term
   - Not all HF patients (especially those with diastolic dysfunction) exhibit symptoms of congestion

HF is a progressive syndrome $⇒$ develops over many years
EF 5 – 10%

Diastole

Systole

Clinical Progression of Heart Failure

HF progression

~50% mortality within 5 Years of Diagnosis

Systolic vs Diastolic HF

Diastolic Dysfunction

- Heart failure symptoms with EF ≥ 40%
- Hypertrophic CMP, HTN, Ischemia, Age
- Imbalance in volume/pressure relationship

Systolic Dysfunction

- Depressed contractility EF ≤ 40%
- CAD, Valvular disease, Ischemic and Idiopathic CMP

Types of heart failure
Diastolic Dysfunction (HFpEF)

Pathophysiology

1. Ventricular muscle thickens (concentric hypertrophy)
2. Ventricular cavity size may remain normal or become smaller
3. Noncompliant ventricle unable to relax, impairing filling
4. To ↑ filling, left atrial pressure ↑; leading to pulmonary congestion

1. EF normal in diastolic dysfunction

Diastolic Dysfunction (HFrEF)

Pathophysiology

1. LV wall thins and the cavity dilates (eccentric hypertrophy)
2. Thin, dilated ventricle unable to contract effectively
3. EF decreases
4. Leads to ↓ CO, ↑ LVEDV, ↑ preload → pulmonary congestion
5. Dilated cardiomyopathy common cause of systolic HF
6. Cardiomyopathy and systolic dysfunction should NOT be used interchangeably

1. EF < 40% systolic dysfunction

Diastolic Dysfunction

Clinical Presentation

- Symptomatic with exertion and ↑ HR
  - Faster HR ↓ filling time & ↓ CO
  - Exercise → ↑ catecholamines → ↑ HR → worsens diastolic function
  - Flash pulmonary edema can develop during periods of ischemia

Elevated heart rate worsens diastolic function

Diastolic Dysfunction

Diagnosis

- Three conditions required:
  1. Signs and symptoms of HF
  2. Normal or only slightly decreased EF
  3. Increased diastolic filling pressures and abnormal relaxation of LF
- Diagnosis made in patients presenting with the clinical syndrome of HF with no evidence of systolic dysfunction

1. Commonly associated with chronic hypertension or ischemic heart disease
New York Heart Association (NYHA) Classification of Heart Failure

<table>
<thead>
<tr>
<th>Class</th>
<th>Patient Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I (Mild)</td>
<td>No limitation of physical activity. Ordinary physical activity does not cause undue fatigue, rapid/irregular heartbeat (palpitation) or shortness of breath (dyspnea).</td>
</tr>
<tr>
<td>Class II (Mild)</td>
<td>Slight limitation of physical activity. Comfortable at rest, but ordinary physical activity results in fatigue, rapid/irregular heartbeat (palpitation) or shortness of breath (dyspnea).</td>
</tr>
<tr>
<td>Class III (Moderate)</td>
<td>Marked limitation of physical activity. Comfortable at rest, but less than ordinary activity causes fatigue, rapid/irregular heartbeat (palpitation) or shortness of breath (dyspnea).</td>
</tr>
<tr>
<td>Class IV (Severe)</td>
<td>Unable to carry out any physical activity without discomfort. Symptoms of fatigue, rapid/irregular heartbeat (palpitation) or shortness of breath (dyspnea) are present at rest. If any physical activity is undertaken, discomfort increases.</td>
</tr>
</tbody>
</table>

Causes

- Usually results from prolonged LV failure
- Right ventricular MI or inferior wall MI
- Primary pulmonary hypertension
- Acute or chronic lung disease
- Chronic severe tricuspid regurgitation
Your patient has an S 3

Neurohormonal Responses in HF
1. Activation of Sympathetic Nervous System (SNS)
2. Renin–Angiotensin–Aldosterone System (RAAS) Kicks in...

Activation of Sympathetic Nervous System (SNS)
- Low Cardiac Output
- Hypoventilation
- Hypovolemia
- Decreased Renal perfusion
- Afferent Arteriole (baroreceptors)
- Release Renin (a messenger)
- Go to Liver to stimulate Angiotensin I production
- Angiotensin I goes to the Lung
- Angiotensin Converting Enzyme (ACE) located in the pulmonary vascular membrane
- Converts Angiotensin I to Angiotensin II
- Angiotensin II
- Growth Factor
- Potential Vasconstrictor
- Increases B/P
- Increases SVR
- Converted to Aldosterone
- Secreted by Adrenal Cortex
- Increased Na + absorption
- Excretes K + for Na +

Renin–Angiotensin–Aldosterone System (RAAS)
- Low Cardiac Output
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SNS Activation
- Cardiac Output
- Activates SNS
- Compensates by releasing neurohormones (Norepinephrine, Aldosterone, Vasopression)
- Sodium & Water Retention
- Tachycardia
- Myocardial Oxygen Time
- Diastolic Filling Time
- Vasoconstriction
- Atrial Filling
NT proBNP (N-Terminal Pro-B)
- Elevates with Heart Failure
- Not interchangeable with BNP
- proBNP is measured before it is metabolized into BNP (Hence proBNP)
- 5:1 ratio to BNP
- proBNP divided by 5 gives a number similar to the BNP values
  - For example: proBNP of 500/5 = 100
  - proBNP < 450, probably not Heart Failure
  - If age over 50, need to look at specific ranges
    - 50 – 75 years proBNP > 900pg/mL consistent with HF
    - > 75 years proBNP > 1800pg/mL consistent with HF

Heart Failure

Pharmacological Treatment
- Three primary goals of medical treatment
  1. Reduce preload
  2. Reduce afterload
  3. Increase contractility

Paradigm for Outpatient Management of Heart Failure

<table>
<thead>
<tr>
<th>Stabilize patient</th>
<th>Stabilize the disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diuretic</td>
<td>ACE inhibition</td>
</tr>
<tr>
<td></td>
<td>α-blockade</td>
</tr>
<tr>
<td></td>
<td>Aldosterone inhibitors</td>
</tr>
<tr>
<td></td>
<td>Treat residual symptoms</td>
</tr>
<tr>
<td></td>
<td>Digoxin</td>
</tr>
</tbody>
</table>

Reduce Preload
- Diuretics
- Venous Vasodilators
  - ACE (Angiotensin-converting enzyme) Inhibitor
    - Blocks the RAAS effect of reabsorption of sodium and water and thus decreases volume overload
  - Aldosterone antagonists
    - Spironolactone & eplerenone
    - Added to increase diuretic effect if symptoms at rest
  - Nitrates
    - Dilates veins, allowing more blood to remain in the vascular system and sending less to the heart

Reduce Afterload
- Arterial Vasodilators
  - ACE (Angiotensin-converting enzyme) Inhibitor
    - Blocks the RAAS effect of vasoconstriction
    - Enhances the action of kinins, which promotes a positive vasodilatory effect
  - Angiotensin Receptor Blockers
    - Directly blocks angiotensin II – results vasodilatory effect
    - Used if patient cannot tolerate ACEI due to cough or angioedema
  - Hydralazine and Nitrate Combination
  - Hydralazine – arterial vasodilator
    - Recommended in African Americans with systolic dysfunction
    - Oral nitrates are venous vasodilators (preload reduction)
  - Milrinone (Primacor) Phosphodiesterase inhibitor
    - Causes vasodilation to preload and afterload

Beta Blockers
- Blocks the neurohormonal response of chronic SNS stimulation
- Slows heart rate for better diastolic ventricular filling
- Not initiated when fluid overload or in a decompensated state
- Initiate after fluid status optimized (no longer needing IV diuretics or IV vasodilators)
- Reduces arrhythmias
- Slows disease progression
- Carvedilol (Coreg), Metoprolol (Lopressor), and Bisoprolol (Zebeta)
Increase Contractility

- Increase contractility by first ↓ afterload
- Digoxin
  - Oral medicine of choice to assist with contractility
  - Enhances inotropy of cardiac muscle
  - Reduces activation of SNS and RAAS
  - Most effective in patients with low EF
- Dobutamine
  - ↑ contractility by stimulating beta receptors
  - Milrinone (Primacor) Phosphodiesterase inhibitor
    - Increases calcium ion uptake.
    - Has positive inotropic effect

Medications to Avoid in HF

- NSAIDS (non-steroidal anti-inflammatory drugs)
  - ↑ risk of fluid retention and renal failure
  - May diminish the efficacy of diuretics and ACE inhibitors
- Most antiarrhythmics
  - Poorly tolerated due to proarrhythmic & cardiodepressant effects
  - Amiodarone – does not adversely effect HF survival
- Calcium channel blockers
  - Amiodipine is okay
  - Avandia (Rosiglitazone Maleate)
    - Can cause fluid retention and exacerbate HF

Cardiac Resynchronization Therapy

Patient Indications

CRT device:
- Moderate to severe HF (NYHA Class III/IV) patients
- Symptomatic despite optimal, medical therapy
- QRS ≥ 130 msec
- LVEF ≤ 35%

CRT plus ICD:
- Same as above with ICD indication

CRT Systems

- Atrial lead in right atrial appendage (same as conventional pacing)
- Right–ventricular lead at the RV apex or RV outflow tract
- Left-ventricular lead is passed through the coronary sinus and into one of the vessels on the outside of the heart

Nonpharmacological Treatments of HF

- Diet
- Daily weights
- Exercise
- Teach back
Heart Failure
Call 911 for:
- Unexplained Chest Pain
- Unexplained Shortness of Breath
- Weight that goes up 3 pounds in 1 day or 5 pounds in 1 week
- New swelling in feet, ankles, hands, abdomen
- Cough that does not go away
- Increased shortness of breath especially with rest or when laying down
- Less energy than usual
- Unexplained dizziness, confusion or anxiety
You are doing well when:
- Weight is stable
- Able to do normal activities
- No change in symptoms

Cardiogenic Shock
- Caused by massive insult to LV
  - usually MI
  - May be complication post cardiac surgery
- Results in profound LV dysfunction

Clinical Presentation
- Systolic BP < 90
- Decreased sensorium
- Cool, pale, moist skin
- Peripheral cyanosis
- Decreased urine output
- Tachycardia
- Weak, thready pulse
- Tachypnea
- Hypoxia
- S3, S4
- Distended neck veins if right sided failure
- Crackles or adventitious lung sounds if pulmonary edema

Caused by massive insult to LV

Know Normal Values!

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Normal Values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac Output (CO)</td>
<td>4 – 8 l/min</td>
</tr>
<tr>
<td>Cardiac Index (CI)</td>
<td>2.5 – 4.2 l/min/m²</td>
</tr>
<tr>
<td>Right atrial pressure (CVP)</td>
<td>0 – 8 mmHg</td>
</tr>
<tr>
<td>Pulmonary artery pressure (PAS/PAD)</td>
<td>15 – 30/6 – 12 mmHg</td>
</tr>
<tr>
<td>Pulmonary artery occlusive pressure</td>
<td>4 – 12 mmHg</td>
</tr>
<tr>
<td>Systemic vascular resistance (SVR)</td>
<td>770 – 1500 dyne/sec/cm²</td>
</tr>
<tr>
<td>Pulmonary vascular resistance (PVR)</td>
<td>20 – 120 dyne/sec/cm²</td>
</tr>
<tr>
<td>Stroke Volume (SV)</td>
<td>60 – 130 ml/beat</td>
</tr>
<tr>
<td>Stroke Volume Index (SVI)</td>
<td>30 – 65 ml/beat/m²</td>
</tr>
<tr>
<td>Arterial oxygenation saturation</td>
<td>95 – 100 %</td>
</tr>
<tr>
<td>Venous oxygenation saturation</td>
<td>60 – 80 %</td>
</tr>
</tbody>
</table>

Source: Modified from Critical Care Nursing: Principles of Practice, 11th ed.

Draw arrows to indicate if the hemodynamic parameters would be increased, decreased or normal.
Cardiogenic Shock

Management

- Maximize oxygen to tissues
- Increase myocardial oxygen supply
- Decrease myocardial oxygen demand
- Improve contractility with supportive measures
- Mechanical support with IABP
- Left ventricular assist devices
### LVAD ~ Left Ventricular Assist Device

- A mechanical circulatory support (MCS) pump
  - Supports the damaged heart
  - Reestablished normal hemodynamics
  - Restores blood flow to vital organs
- Used in decompensating advanced heart failure patients who fail to improve or stabilize with optimal medical therapy.

### Indications for Use

- **Bridge to Transplant (BTT)**
  - Non-reversible left heart failure
  - Imminent risk of death
  - Candidate for cardiac transplantation
- **Destination Therapy (DT)**
  - NYHA Class IIIb or IV heart failure
  - Optimal medical therapy 45 of last 60 days
  - Not candidate for heart transplantation
- **Bridge to Recovery (BTR)**
  - Reversible cardiac insults (AMI, PP cardiomyopathy, myocarditis)
- **Bridge to Candidacy (BTC)**
  - Not currently listed for transplant
  - No absolute or permanent contraindication to transplant

### VAD

- **No pulse**
- **Anticoagulation needed**
- **Battery backup and Emergency plan**
Assessment
“Back to the Basics”

- Good peripheral circulation
  - Warm and pink legs and fingers
- Brisk capillary refill (< 5 seconds)
- Appropriate mentation
  - Follows commands
  - Able to talk to you and carry on normal conversation
  - Does not fall asleep
- Adequate urine output


Emergency Management

- Arrhythmias are major cause of emergencies
  - If LVAD is running, blood is still circulating
  - OK to defibrillate or cardiovert or external pace
  - Do not stop the pump!
  - Do not place pads over implanted pump
- Do NOT do chest compressions.
  - Compressions will displace cannulas
  - Call implant center before doing compressions
  - If LVAD is running, blood is still circulating
- Dehydration can cause lethal arrhythmias.
  - Need immediate bolus

Vital Signs

- **B/P** Take manually with doppler.
  - The pressure you hear is the MAP
  - MAP should be 70 – 80
  - MAP > 90, call
  - With automatic BP you may get a systolic & diastolic pressure with a very narrow pulse pressure
- **Pulse** – you may feel one radially, but it’s not accurate. Need to Dopple.
- **Telemetry** – LVAD does not affect rhythm
- **Pulse ox** – likely accurate

What’s New? LifeVest

- The LifeVest is worn outside the body rather than implanted in the chest.
- This device continuously monitors the patient’s heart with dry, non-adhesive sensing electrodes to detect life-threatening abnormal heart rhythms.
- If a life-threatening rhythm is detected, the device alerts the patient prior to delivering a treatment shock, and thus allows a conscious patient to delay the treatment shock.
- If the patient becomes unconscious, the device releases a Blue™ gel over the therapy electrodes and delivers an electrical shock to restore normal rhythm.

Source: http://lifewayz.com

SmitCo Personal Defibrillator
https://www.youtube.com/watch?v=TwP55lrg8Z8