Pericardial Diseases

David Stultz, MD, FACC
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www.drstultz.com
Goals of Conference

- Identify signs and symptoms of pericardial diseases
- Discuss workup and treatment strategies for pericarditis
- List common causes of pericarditis and pericardial effusion
The Pericardium

- **Outer fibrous layer**
- **Inner lining (serous)**
  - Visceral pericardium (epicardium)
  - Parietal pericardium lines outer fibrous layer
- **Pericardial space is in between visceral and parietal pericardium**
  - Normally 15-50mL of fluid
Case #1

- 32 year old Male
- 1 week ago started with sore throat, myalgias
- Developed chest pain (pressure)
  - Left substernal
  - Radiating to neck
  - Improves when sitting up
- No significant medical, family, social history
- No medications
- Physical Exam unremarkable
Laboratory Data

- WBC 12.8
- Renal panel normal
- LDL 113
- Troponin I 4.0
- CPK 240, CK-MB 21.8 (index 9.1%)
What is the most likely diagnosis?

1. Acute myocardial infarction
2. Acute pericarditis
3. Acute myopericarditis
4. Constrictive pericarditis
5. Pericardial tamponade
What would you do next?

1. Cardiac Catheterization
2. Echocardiography

Or go straight to treatment without imaging:

3. High dose NSAIDS
4. High dose NSAIDS + Colchicine
5. Prednisone
Now What Would You Do For Treatment?

1. Indomethacin 25mg po q6-8h
2. Ibuprofen 800mg po q6h
3. Aspirin 650-800mg po q6-8h
4. Indomethacin + colchicine 0.6mg daily
5. Ibuprofen + colchicine 0.6mg daily
6. Aspirin + colchicine 0.6mg daily
7. Prednisone 60mg po daily with taper
What is the Most Common Cause of Acute Pericarditis?

1. Idiopathic/Viral
2. Bacterial
3. Malignancy
4. Uremia
5. Acute Myocardial Infarction
6. Autoimmune disease
Acute Pericarditis

- Inflammation of Pericardium
- Symptoms include sharp chest pain
- Often improved with upright position

Common Causes of Acute Pericarditis

- **Idiopathic (85-90%)**
- **Infectious**
  - Viral (1-2%)
  - Bacterial (1-2%)
  - Tuberculous (4%)
- **Neoplastic disease (7%)**
- **Uremia**
  - Before dialysis (5%)
  - After initiation of dialysis (13%)
- **Systemic autoimmune disease (3-5%)**

- **As a complication of**
  - Acute myocardial infarction (5-20%)
  - Myocarditis (30%)

Rare Causes of Acute Pericarditis

- After cardiotomy or thoracic surgery
- Aortic dissection
- Chest wall trauma
- Chest wall irradiation
- Adverse drug reaction
- Rare Infectious causes
  - Fungal
  - Parasites

Diagnostic Criteria for Pericarditis

Two of Four criteria

- Characteristic chest pain
- Pericardial friction rub
- Suggestive EKG changes
- New or worsening pericardial effusion

Pericarditis Chest Pain

- Sudden onset
- Retrosternal
- Pleuritic/Sharp
- Worse with inspiration
- Improved when sitting up or leaning forward
- Chest pain can radiate

Pericarditis Friction Rub

- Caused by rubbing of inflamed visceral and parietal pericardium
- Variable over time
- Present in 85% of patients with pericarditis at some point
- High pitched scratch or squeak at left lower sternal border
- Classically 3 phases
  - Atrial systole,
  - Ventricular systole
  - Rapid ventricular filling during early diastole
- May be only biphasic or monophasic

http://depts.washington.edu/physdx/heart/tech5.html
Pericarditis EKG Changes

- Diffuse ST segment elevation and PR segment depression
- **Stage 1**
  - Hours to days
  - ST elevation and PR depression
  - Possible PR segment elevation in aVR
- **Stage 2**
  - Normalization of ST and PR segments
- **Stage 3**
  - Diffuse T wave inversions
- **Stage 4**
  - EKG normalizes (or T wave inversions persist)

Laboratory Studies in Pericarditis

- **Nonspecific elevation of inflammatory markers**
  - Erythrocyte sedimentation rate
  - C-reactive protein
  - White blood cell count

- **Viral titers and cultures not useful**

- **ANA, Rheumatoid factor useful only if other autoimmune findings are present**

- **Elevated troponin**
  - Mild increase when present
  - Usually patent coronary arteries at catheterization
  - Usually resolve in 1-2 weeks
  - Prognosis is good

Myopericarditis

- Predominantly pericardial involvement with associated myocardial inflammation.
- Diagnosed after pericarditis diagnosed and evidence of myocardial involvement
  - Abnormal cardiac enzymes
  - New onset of global or regional left ventricular dysfunction
- Endomyocardial biopsy not needed
- Approximately 17% of patients ultimately diagnosed with pericarditis undergo heart catheterization

Imaging Studies

- Chest X-ray – Usually not helpful unless there is a significant pericardial effusion
- Echocardiography – Indicated for hemodynamic compromise
- Computed Tomography – useful to measure pericardial thickness (usually 1-2mm) and pericardial effusion
- Cardiac MRI – Delayed gadolinium enhancement shows inflammation of pericarditis

Cardiac MRI of Pericarditis
Delayed Gadolinium Enhancement

Initial Evaluation

- **History**
  - Any suggestion of malignancy or autoimmune disease

- **Physical Examination**
  - Friction rub
  - Signs of Tamponade
    - Pulsus Paradoxicus
    - Kussmaul’s sign
    - Beck’s Triad

Initial Evaluation

- EKG
- Chest X-ray
- CBC
- Erythrocyte Sedimentation Rate and/or C-Reactive Program

Selected Evaluations

- Echocardiogram for hemodynamic compromise
- Suspected autoimmune disease
  - Antinuclear Antibody titers
  - Rheumatoid Factor
- Suspected infectious disease
  - Tuberculin skin testing
  - Human Immunodeficiency Virus
  - Blood Cultures
- Malignancy workup

Hospitalization

Any high risk features
- Fever
- Leukocytosis
- Large pericardial effusion (>20 mm)
- Cardiac tamponade
- Acute trauma
- Immunosuppressed state
- Anticoagulated patient
- Failure of NSAID treatment
- Abnormal troponin
- Recurrent pericarditis.

Outpatient “Day Hospital” Management

- No high risk features
- 254 out of 300 cases low risk
  - Treated with Aspirin 800mg q6-8h x 2-3 weeks with tapering
  - Baseline echocardiogram
  - Clinical and echo follow-up periodically over 1 year
- Mean follow-up of 38 months
  - 43 (16.9%) cases of relapses
  - 4 (1.6%) cases of constrictive pericarditis
  - No cases of cardiac tamponade
- Failure to respond to Aspirin after 7-10 days predicted higher rates of complication

Treatment of Pericarditis

- **NSAIDS**
  - High dose Aspirin or ibuprofen
    - Aspirin 800mg q6-8h x 7-10 days then taper off over 2-3 weeks
    - GI prophylaxis recommended
  - Indomethacin not recommended in patients with coronary disease

- **Colchicine**
  - Use in conjunction with aspirin for 4-6 weeks
  - Caution with severe renal insufficiency, hepatobiliary dysfunction, gastrointestinal motility disorders

- **Corticosteroids**
  - Reserved for patients failing initial therapy with NSAID+colchicine
  - Increased risk of relapsing pericarditis
  - Consider using in
    - Autoimmune disease
    - Connective tissue disorder
    - Uremic pericarditis
  - Prednisone 1mg/kg/day, taper after 2-4 weeks of therapy
COPE Trial

120 Patients with first episode pericarditis

Conventional
- ASA 800mg q6-8h for 7-10 days with tapering over 3-4 weeks

Experimental
- ASA + Colchicine 1-2mg day 1 then 0.5-1mg daily for 3 months

Addition of Colchicine beneficial
- Lower rate of recurrence at 18 months (11% vs. 33%)
- Better 72 hour symptom resolution (12% vs. 37%)

Pericardiocentesis for Pericarditis

- Suspected etiology
  - Purulent
  - Tuberculous
  - Malignancy

- Persistent symptomatic pericardial effusion

Relapsing pericarditis

- Usual first recurrence within 18 months
- Incessant
  - Pericarditis returns within 6 weeks of treatment discontinuation
- Intermittent
- Usually responds to steroids
- Usual causes
  - Autoimmune
  - Viral or other infection
  - Post-pericardial/Post-myocardial injury syndromes
- Consider pericardiectomy in extreme cases

Case #1

- Diagnosis: Myopericarditis
- Treated with ibuprofen 800mg po q8h x 5 days then 400mg po q12h x 5 days
- Started on carvedilol 3.125mg po q12h
- Discharged after overnight hospitalization
- Doing well at 3 month follow-up
53 year old male

Increasing chest pressure and neck pain
- Almost constant
- Worse with activity or deep breath
- Better when sitting up

Fevers, chills, nausea, and vomiting 3 months ago

Past medical history
- Hypertension
- Hyperlipidemia

No pertinent medications, family or social history
Case #2 EKG
CAT Scan of the Chest
Cardiac Tamponade

- Increased fluid in pericardial space
- Increases intracardiac pressures
- Impairs normal cardiac filling
- Exaggerated by respirations
  - Inspiration decreases right ventricular pressure but increases left ventricular pressure
- May be acute, subacute, or chronic
  - Cardiac procedures are the most common acute cause!

Diagnosis

- Symptoms may include chest discomfort, shortness of breath
- Pulsus paradoxus
  - Decrease in systolic blood pressure of >10mmHg with inspiration
- Jugular venous distension
  - Normal x descent (atrial diastole) with blunted y descent (atrial systole)

Echocardiography in Tamponade

- Right atrial collapse (late diastole)
- Right ventricular collapse (early diastole)
- Respiratory variation of transvalvular inflow (i.e. the echo pulsus paradoxus)
  - Tricuspid >40%
  - Mitral >25%
Right Atrial Collapse
Tricuspid Valve Inflow with Respiratory Variation
Mitral Valve Inflow with Respiratory Variation
Treatment of Tamponade

- **IV fluids can be a temporizing measure**
- **Ultimately need to evacuate fluid**
  - Percutaneous pericardiocentesis
  - Surgical or percutaneous balloon pericardiotomy
- **Analyze fluid (as clinically indicated) for**
  - Gram stain
  - Bacterial cultures
  - Acid-fast bacilli and culture
  - Cytology
- **In setting of aortic dissection, pericardiocentesis should NOT be done!**

Pericardial effusion without tamponade

- 1/3 of patients with large (>20mm) pericardial effusion develop tamponade
  - Consider pericardiocentesis if effusion persists more than 1 month

- Regular clinical and echocardiographic follow-up recommended

- Consider thoracic duct obstruction with chylopericardium if persistent

- Consider hypothyroidism

Pericardial Hematoma

- Blood clot in pericardial space
- Causes tamponade physiology
- Diagnosed by transthoracic or transesophageal echocardiogram

Etiology

- Iatrogenic
  - post cardiac surgery or other procedure
- Aortic dissection
- Trauma
Case #2 Treatment

- Thyroid studies normal
- Rheumatoid Factor, ANA negative
- Erythrocyte Sedimentation rate 83
- Pericardiocentesis performed
  - Cytology negative
- Treated with NSAIDS
- Diagnosis: idiopathic/viral pericarditis
What is the Most Common Cause of non-Iatrogenic Cardiac Tamponade?

1. Idiopathic/Viral
2. Malignancy
3. Uremia
4. Acute Myocardial Infarction
5. Autoimmune disease
6. Trauma
7. Hypothyroidism
Case #3

- 50 year old female
- Increasing dyspnea and orthopnea over several days
- Bilateral lower extremity edema and night sweats for two days
- Dry cough and low-grade fever (101°F) 10 days ago
- Recent chest pains
  - Thoracentesis for bilateral pleural effusions
- Recently diagnosed atrial fibrillation & atrial flutter on admission
- Past Medical History
  - Hypothyroidism
  - Hodgkin lymphoma (radiation & chemotherapy)
  - Basal cell carcinoma
- Medications
  - Levothyroxine 112 mcg/day, diltiazem 30 mg q12h, propafenone 150mg q12h, warfarin
Case #3

- WBC 13.6, Hgb 12.3, Platelets 384
- Na 131, K+ 3.9, creatinine 1.0
- TSH 25.4, free T4 1.0
- Cholesterol 131, trigs 54, LDL 89, HDL 31
- BNP 833
- Erythrocyte sedimentation rate 120
- C Reactive protein 115
- INR 4
CT Scan of the Chest
What is the Most Likely Diagnosis?

1. Hypothyroidism
2. Acute pericarditis
3. Pericardial tamponade
4. Restrictive cardiomyopathy
5. Constrictive pericarditis
6. Ischemic cardiomyopathy
Constrictive Pericarditis

- Symptoms of heart failure with preserved ejection fraction
- Due to thickening of pericardium
  - Impairs diastolic filling
- Etiology in developed countries
  - Idiopathic
  - Cardiac surgery
  - Pericarditis
  - Mediastinal radiation therapy
- Tuberculosis is major cause in developing countries

Constrictive Pericarditis vs. Restrictive Cardiomyopathy

- Restrictive Cardiomyopathy is RARE
  - Amyloidosis
  - Sarcoidosis
  - Hypereosinophilic syndromes
  - Endomyocardial fibrosis
  - Chemotherapy or Radiation

Gross Specimens
Constrictive pericarditis vs. Restrictive Cardiomyopathy

Diagnosis of Constrictive Pericarditis

- **History**

- **Physical examination**
  - Jugular venous distention
  - Kussmaul’s sign (rise in JVD with inspiration)
  - Pericardial knock

- **EKG** – Nonspecific
Chest X-ray in Constrictive Pericarditis

Constrictive pericarditis vs. Restrictive Cardiomyopathy

- In Constrictive Pericarditis there is ventricular interdependence accentuated by respiration
  - As inspiration occurs, RV filling improves at the expense of LV filling
  - RV pressure increases as LV pressure decreases
- Echocardiographic criteria based on this phenomena
- Can be measured invasively

Hemodynamics in a Nutshell

Constrictive pericarditis vs. Restrictive Cardiomyopathy

Treatment of Constrictive Pericarditis

- If transient due to acute inflammation, medical therapy
- If chronic, pericardiectomy is considered
  - Surgical mortality approaches 6%
  - Must be a complete pericardiectomy

Case #3 Treatment

- Started on furosemide
- Levothyroxine dose increased
- Referred to tertiary care center for pericardiectomy
Effusive-Constrictive Pericarditis

- Both pericardial effusion and constrictive pericarditis
- Constrictive hemodynamics may persist after pericardiocentesis
- Initial treatment as pericarditis
- May be transient, resolve in 2-3 months
- May require pericardiectomy if symptoms persist

Congenital Absence of Pericardium

- Usually partial absence of left pericardium
- Male predominance
- Associated with
  - Atrial septal defect
  - Bicuspid aortic valve
  - Bronchogenic cysts
- Usually asymptomatic
- May require surgical closure of partial defect if symptomatic

Chest X-ray of Partially Absent Pericardium

http://www.bcm.edu/radiology/cases/pediatric/text/7d-desc.htm
Pericardial Cyst

- Benign
- Incidental mass on CT or Chest X-ray
- Usually located at right costophrenic angle
- Echocardiography, cardiac CT, or cardiac MRI can differentiate
  - Malignancy
  - Diaphragmatic hernia
  - Cardiac chamber enlargement
- No treatment necessary if asymptomatic

Conclusions

- **Acute pericarditis**
  - Most often viral
  - Treat with high dose NSAIDS + colchicine

- **Pericardial tamponade**
  - Often caused by malignancy
  - Volume support until pericardiocentesis

- **Constrictive pericarditis**
  - Suspect with diastolic heart failure symptoms, thickened pericardium, and history of pericarditis or radiation exposure