

Myocarditis and Cardiomyopathy



Myocarditis

- ◆ Myocarditis is an inflammation of the myocardium, the thick muscular layer making up the major portion of your heart.
- ◆ Often follows URI
- ◆ May present with chest pain (either pleuritic or non-specific) or signs of heart failure
- ◆ ECG may show sinus tachycardia, nonspecific repolarization abnormalities, and intraventricular conduction abnormalities
- ◆ Echocardiography documents cardiomegaly & contractile dysfunction
- ◆ Myocardial biopsy, although not sensitive, may reveal characteristic inflammatory pattern (ex. Giant Cell)

Myocarditis basics

- ◆ Wide spectrum of clinical consequences
 - Mild & self-limited with few symptoms or severe with progression to CHF & dilated CM
 - Very localized or diffuse
 - Clinical involvement can be limited to the heart or be part of widespread systemic disorder

Epidemiology

- ◆ No accurate estimate of incidence as many cases are mild & brief and diagnosis is not made.
- ◆ No reliable gold-standard noninvasive test to confirm diagnosis
- ◆ Have a high clinical suspicion, if we don't think of it, we won't dx it

Infectious	Noninfectious
Viruses – <ol style="list-style-type: none"> 1. Coxsackie B 2. HIV 	Systemic Diseases: <ol style="list-style-type: none"> 1. SLE 2. Sarcoidosis 3. Vasculitides(Wegener's) 4. Celiac disease
Bacterial – <ol style="list-style-type: none"> 1. Corynebacterium diphtheriae 	Neoplastic infiltration
Protozoan – <ol style="list-style-type: none"> 1. Trypanosoma cruzi (Chagas disease) 	Drugs & toxins: <ol style="list-style-type: none"> 1. Ethanol 2. Cocaine 3. Radiation 4. Chemotherapeutic agents - Doxorubicin
Spirochete <ol style="list-style-type: none"> 1. Borrelia burgdorferi 	

Signs & Symptoms

- ◆ Days to weeks after onset of acute febrile illness or with heart failure without any known antecedent symptoms; highly variable
- ◆ Onset of heart failure may be abrupt and fulminant or gradual.
- ◆ May mimic acute MI with ST elevation, positive cardiac markers, regional wall motion abnormalities

ECG & CXR

- ◆ ECG - nonspecific ST-T changes and conduction delays are common
 - Ventricular ectopy may be only clinical finding
- ◆ CXR - cardiomegaly is frequent, may have evidence for pulmonary venous hypertension & pulmonary edema

Diagnostics

- ◆ Wbc's often elevated
- ◆ ESR increased
- ◆ Troponins elevated in 1/3
- ◆ CK-MB elevated in 10%
- ◆ Echocardiogram helps evaluate cardiac function & exclude other causes
- ◆ Cardiac MRI improving in ability to see abnormalities in myocardium

Endomyocardial Bx

- ◆ Pathologic exam may reveal lymphocytic inflammatory response with necrosis, but this is not sensitive b/c of the patchy areas of distribution.
- ◆ “Dallas” criteria for histopathologic dx
- ◆ May see “Giant cells”

Treatment & Prognosis

- ◆ Fulminant myocarditis pt may present with cardiogenic shock
 - Ventricles are thickened, usually not dilated
- ◆ Subacute pt have dilated cardiomyopathy
- ◆ Chronic pt may have mildly dilated LV and more of a restrictive cardiomyopathy

Treatment

- ◆ Antibiotics if specific agent is identified
- ◆ Standard HF therapy
- ◆ Arrhythmia suppression
- ◆ Limited exercise role during recovery
- ◆ IVIG and steroids are controversial
- ◆ Fulminant myocarditis need aggressive short term support from intra-aortic balloon pumps &/or LVAD

Cardiomyopathy

- ◆ Group of entities that affect the myocardium primarily and are NOT associated with another major cause of heart disease (like coronary artery disease, valvular heart disease)
- ◆ Controversial classifications

Cardiomyopathy

- ◆ *Dilated (congestive) cardiomyopathy* is a group of heart muscle disorders in which the ventricles enlarge but are not able to pump enough blood for the body's needs, resulting in heart failure. (Example - CAD, myocarditis, EtOH, HIV)
- ◆ *Hypertrophic cardiomyopathy* includes a group of heart disorders in which the walls of the ventricles thicken (hypertrophy) and become stiff, even though the workload of the heart is not increased. (Example – congenital HOCM, or acquired)
- ◆ *Restrictive (infiltrative) cardiomyopathy* includes a group of heart disorders in which the walls of the ventricles become stiff, but not necessarily thickened, and resist normal filling with blood between heartbeats. (Example – radiation, amyloidosis)

New Categories

- ◆ Tako-Tsubo - “broken heart syndrome” transient cardiomyopathy secondary to high catecholamine surge
- ◆ Noncompaction - embryologic defect resulting in massive trabeculation of LV
- ◆ Arrhythmogenic right ventricular - most common cause of SCD in young Italians

Primary dilated CM

- ◆ 36.5 per 100,000 persons in US
- ◆ mortality rate 25% at 1 yr
- ◆ 50% at 5 years
- ◆ Most pts are asymptomatic for years
- ◆ 80% of gene carriers younger than 20 are asymptomatic
- ◆ Careful family member screening about 30% of DCM are familial

Primary DCM

- ◆ LV dilation and systolic function <50%
- ◆ Blacks > whites
- ◆ Men > women
- ◆ Chronic EtOH and unrecognized myocarditis are frequent causes
- ◆ Chronic tachycardia may also cause
- ◆ RV may be primarily involved - unusual CM with adipose displacing myocardial cells

Clinical Findings

- ◆ Usually symptoms of HF develop slowly
- ◆ Initial presentation may be severe left or biventricular failure
- ◆ May be recognized b/c of asymptomatic cardiomegaly or ECG abnormalities (including arrhythmias)

Molecular Basis of DCM

- ◆ Familial is inherited in autosomal dominant pattern (90% of cases)
- ◆ To date, more than 22 DCM susceptibility genes & 4 more chromosome loci have been implicated
- ◆ Penetrance of DCM is highly variable
- ◆ No clinically available genetic test

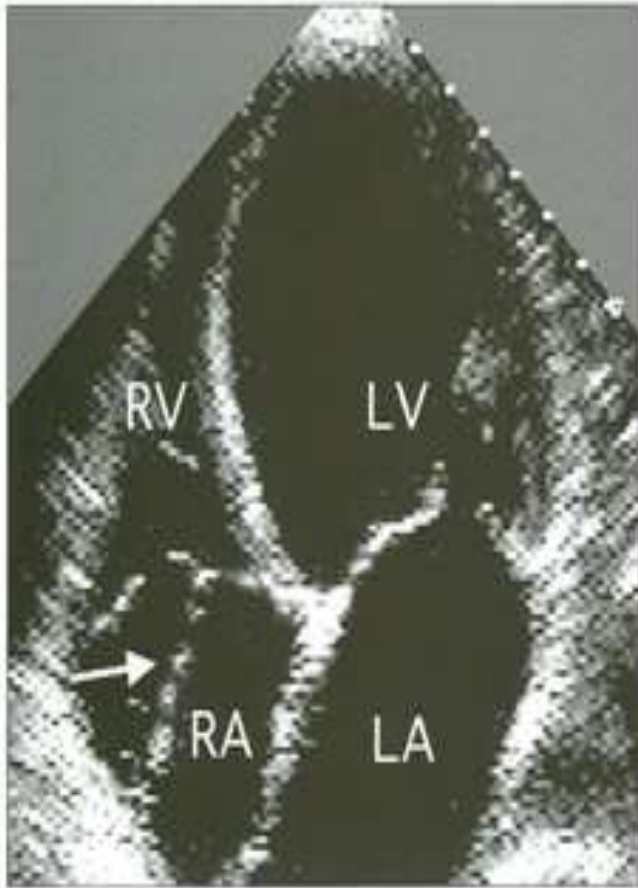
Physical Exam

◆ Physical exam

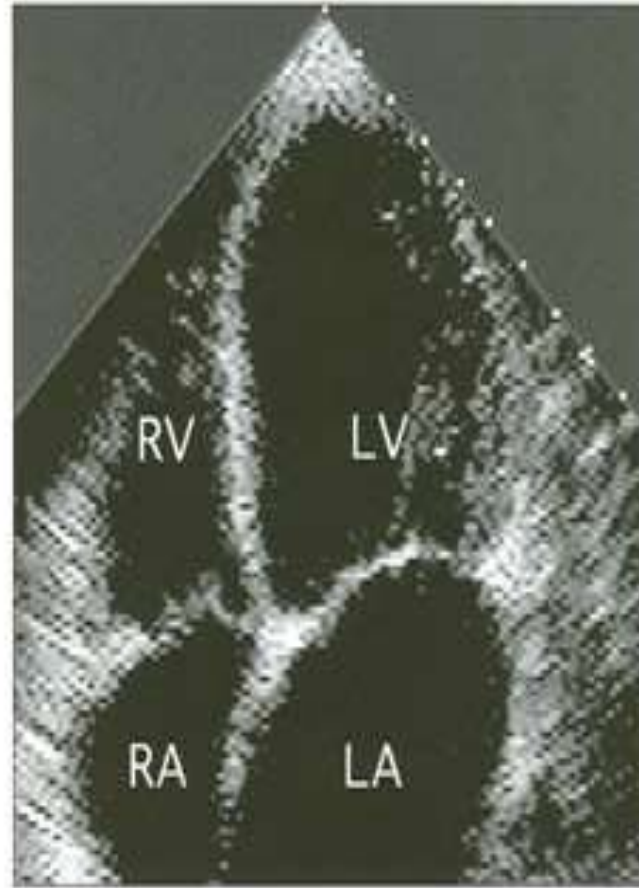
- Rales
- elevated JVP
- cardiomegaly
- S3 gallop
- Murmur of functional mitral or tricuspid regurgitation
- Peripheral edema
- Ascites

Diagnostics

- ◆ **ECG** - ST-T changes, conduction abnormalities, ventricular ectopy, LBBB
- ◆ **CXR** - Enlarged heart, pulmonary congestion
- ◆ **ECHO** - LV dilation & dysfunction
- ◆ **Catheterization** - LV dilation & dysfunction, high diastolic pressures, low cardiac output



a



b

Treatment of DCM

◆ Standard HF therapy

- ACE inhibitor
- B-blocker
- Diuretic
- Aldosterone antagonist
- Digoxin is 2nd line agent
- CCB should generally be avoided
- Na⁺ restriction helpful

Prognosis

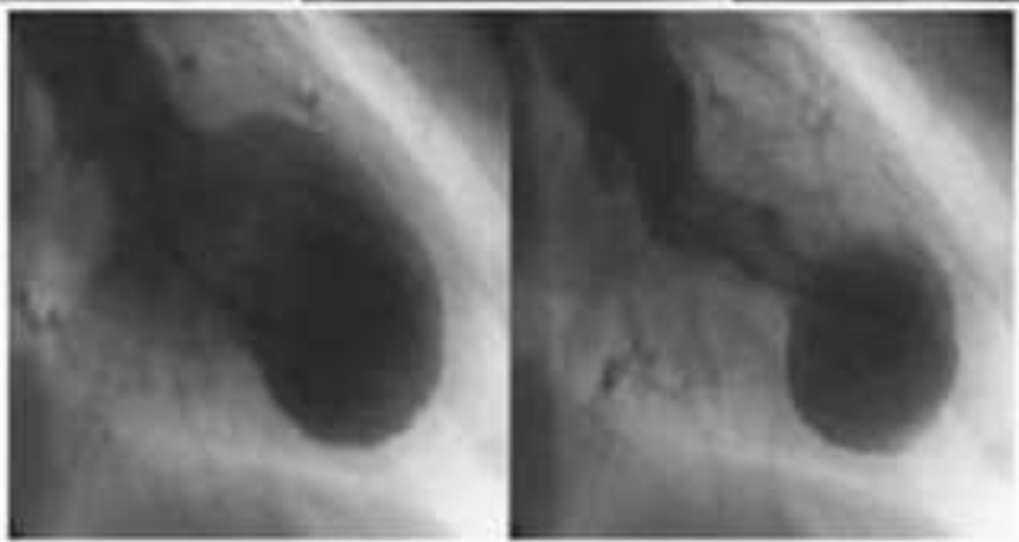
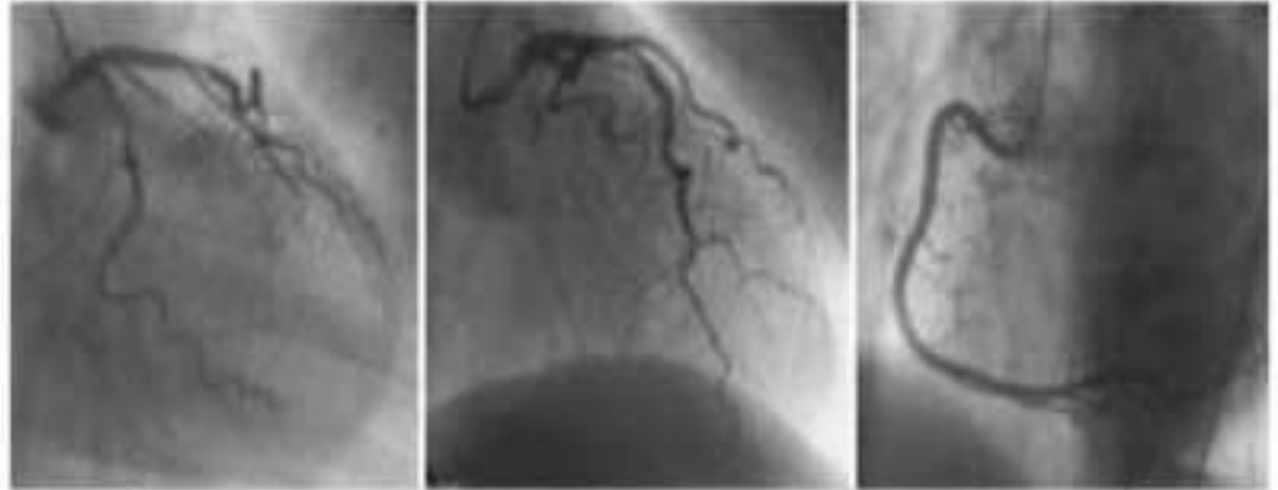
- ◆ DCM without HF is variable outcome
 - ranges from stable to rapid decline
 - Once HF is manifest, outcome similar to other types of HF, annual mortality 15%
 - Arterial & pulmonary emboli more common in DCM than ischemic - consider anticoagulation

Family Screening

- ◆ Take thorough family hx
- ◆ Dx of familial based on dx of DCM in 2 or more close relatives
- ◆ All 1st degree relatives of index case should receive screening with ECG and echocardiogram
- ◆ After initial - screen, repeat every 3-5 years

Tako-Tsubo

- ◆ LV apical ballooning after a high catecholamine stress which results in LV shape similar to octopus pot (takotsubo pot which is Japanese octopus trap)
- ◆ Has been described following stressful event like hypoglycemia, earthquakes, following surgery, after emotional stress



Diastole

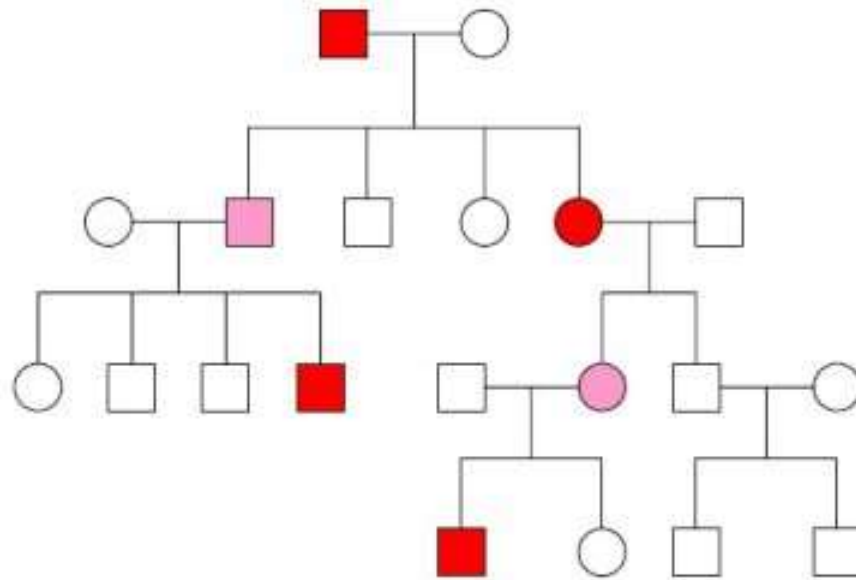
Systole



Tako-Tsubo



- ◆ Presents as acute anterior MI with chest pain or SOB
- ◆ Usually in post-menopausal women
- ◆ Cardiac catheterization reveals clean coronary arteries
- ◆ Prognosis is good unless there is serious complication (like MR, ventricular rupture, v-tachycardia)



Arrhythmogenic right ventricular

- ◆ Primary heart muscle disease with progressive degeneration & fibrous fatty replacement or total or partial RV
- ◆ Major cause of SCD in young, especially in some regions of Italy, still exists in US - consider in young pt with syncope, palpitations, and aborted SCD
- ◆ Familial in 50% of cases



Unaffected male  **or female**  **with normal gene**

Unaffected male  **or female**  **gene carriers**

Affected male  **or female**  **with abnormal gene**

Noncompaction

- ◆ Congenital disorder with hypertrophied LV with deep trabeculations
- ◆ Decreased systolic fxn
- ◆ Can be isolated or occur with other congenital heart diseases
- ◆ Facial abnormalities and neurologic problems also occur in high proportion of pts with LVNC
- ◆ Some genetic links, screen 1st degree relatives

Hypertrophic CM

- ◆ LV outflow tract is often narrowed during systole between bulging septum & anteriorly displaced mitral valve which causes a dynamic obstruction
- ◆ Can be congenital (HOCM) or acquired (Idiopathic hypertrophic subaortic stenosis)
- ◆ obstruction worsened by things that increase myocardial contractility including sympathetic stimulation, digoxin, PVC or things that decrease filling like Valsalva, peripheral vasodilators

Hypertrophic CM

- ◆ Amount of obstruction is preload & afterload dependent and can vary from day to day
- ◆ Consequence of this hypertrophy is elevated diastolic pressures rather than systolic dysfunction
- ◆ LV > RV
- ◆ Atria are often significantly enlarged

Hypertrophic CM

- ◆ HOCM is inherited as autosomal dominant trait and is caused by mutations in a number of genes
- ◆ Most of these genes code for myosin heavy chains or proteins that regulate calcium binding
- ◆ Prognosis is related to specific gene mutation

Hypertrophic CM

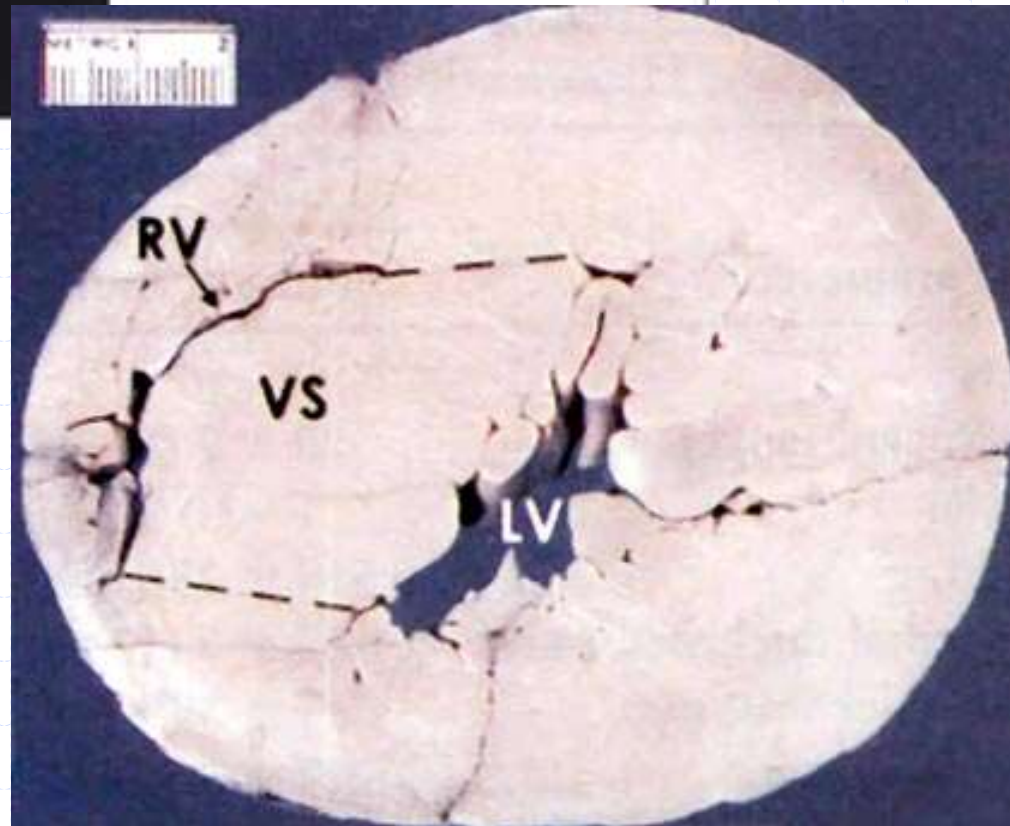
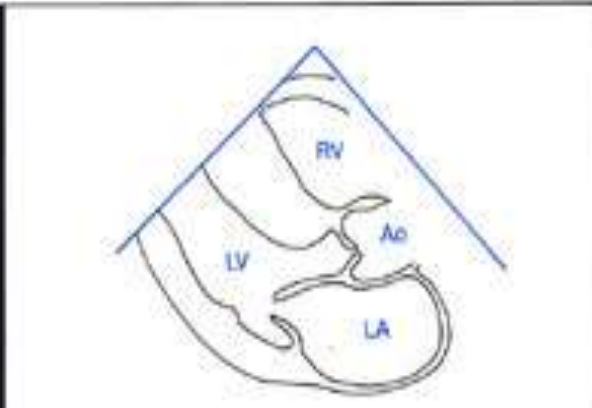
- ◆ Most frequent symptoms are dyspnea, chest pain, syncope (often post-exertion)
- ◆ Ventricular arrhythmias are common and SCD may occur, especially in athletes after extreme exertion

Physical Exam of HCM

- ◆ Bisferiens carotid pulse
- ◆ triple apical impulse
- ◆ loud S4
- ◆ Loud systolic murmur left sternal border that increases with Valsalva and decreases with squatting
- ◆ These maneuvers distinguish HCM from aortic stenosis

Diagnostic Studies HCM

- ◆ **ECG** - Left ventricular hypertrophy
- ◆ **CXR** - usually normal, if ascending aorta is dilated probably aortic stenosis
- ◆ **ECHO** - diagnostic, reveals asymmetric LVH, small & hypercontractile LV, thick septum
- ◆ **Catheterization** - confirms dx



Treatment of HCM

- ◆ Beta blockers are initial drugs of choice
- ◆ Slower heart rate allows more time for diastolic filling
- ◆ CCB have also been effective
- ◆ Endocarditis prophylaxis is indicated
- ◆ Atrial fibrillation usually worsens symptoms and should be aggressively treated
- ◆ Consider dual chamber pacing
- ◆ Can have surgical myomectomy or alcohol ablation

Evaluate for ICD

- ◆ Those at highest risk for SCD
 - prior hx of cardiac arrest
 - spontaneous ventricular tachycardia on ambulatory monitoring
 - family hx of HCM related SCD in relatives <45
 - Severe LVH
 - unexplained syncope, especially in young pt with exertion
 - abnormal BP response to exercise (hypotension)
 - Increased delayed enhancement on MRI

Restrictive CM

- ◆ Idiopathic or systemic myocardial dx characterized by impaired diastolic filling with preserved contractile fxn.
- ◆ Usually caused by infiltration or fibrosis
- ◆ Amyloidosis is most common
- ◆ Right heart failure dominates
- ◆ Pulmonary HTN present

Amyloidosis

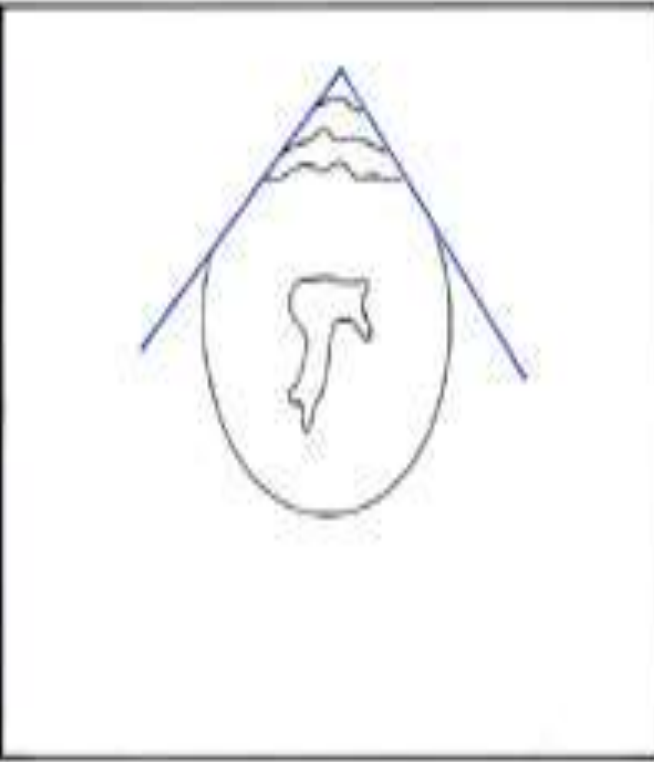
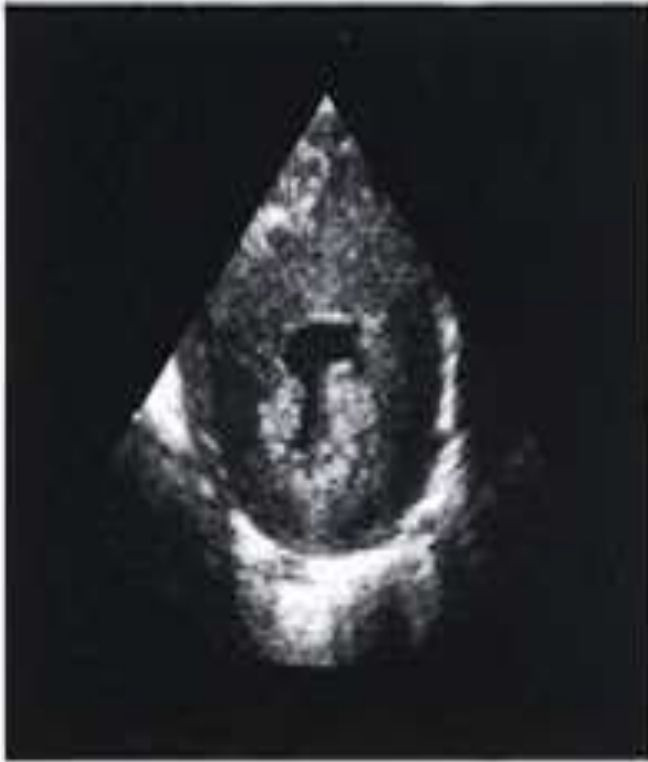
- ◆ Deposition of various proteins within myocardium
- ◆ Primary is from immunoglobulin light chains (often as consequence of multiple myeloma)
- ◆ Heart may be only involved organ

Clinical Findings of RCM

- ◆ Dyspnea & fatigue from pulmonary venous congestion, edema, ascites from RHF common presenting symptoms
- ◆ Must distinguish from constrictive pericarditis.
- ◆ Key feature is ventricular interaction is accentuated with respiration in constrictive, that is absent in restrictive

Diagnostic Studies for RCM

- ◆ **ECG** - low voltage
- ◆ **ECHO** - ventricular hypertrophy, small thickened LV with bright myocardium
- ◆ Abdominal fat pad easiest place to bx for diagnosis



Treatment & Prognosis RCM

- ◆ Little useful therapy exists
- ◆ Diuretics can help
- ◆ Digoxin may worsen
- ◆ May qualify for transplant
- ◆ Overall prognosis very poor