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 – 1. Coxsackie B 2. HIV	Systemic Diseases: 1. SLE 2. Sarcoidosis 3. Vasculitides(Wegener's) 4. Celiac disease
Bacterial – 1. Corynebacterium diphtheriae	Neoplastic infiltration
Protozoan – 1. Trypanosoma cruzi (Chagas disease)	Drugs & toxins: 1. Ethanol 2. Cocaine 3. Radiation 4. Chemotherapeutic agents - Doxorubicin
Spirochete 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

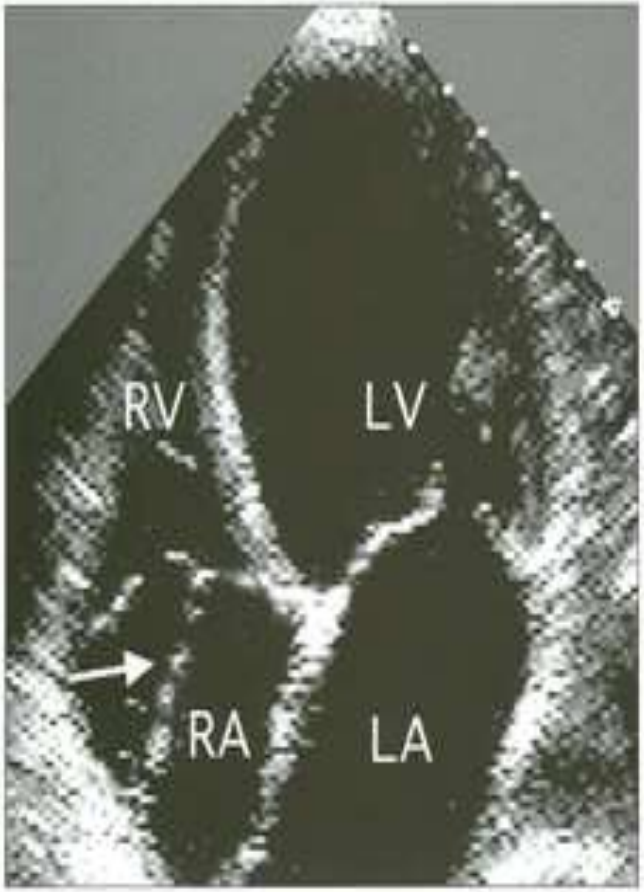
- Familial is inherited in autosomal dominant pattern (90% of cases)
- To date, more than 22 DCM susceptibility genes & 4 more chromosome loci have been implicated
- Penetrance of DCM is highly variable
- No clinically available genetic test

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



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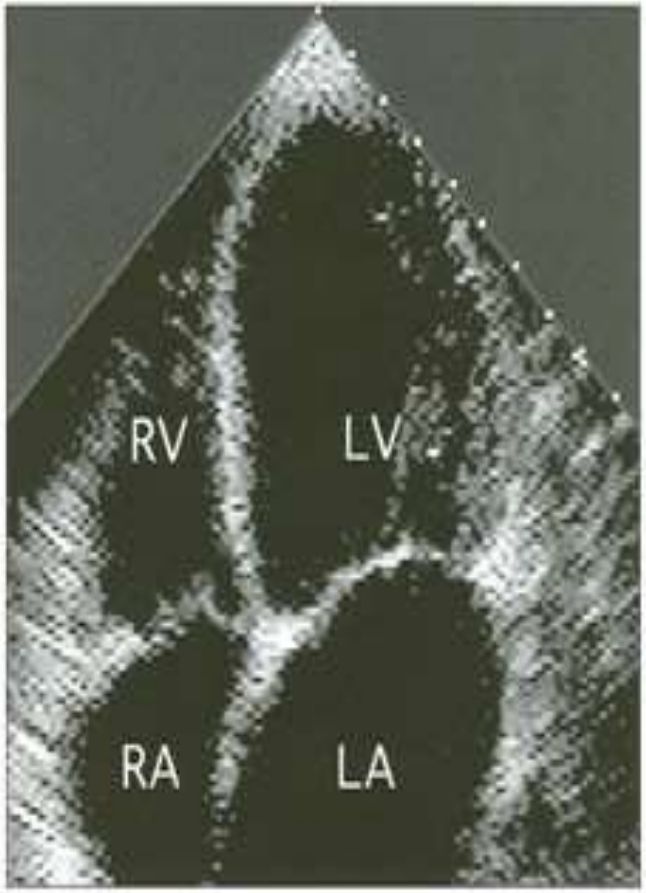

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Treatment of DCM

- 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

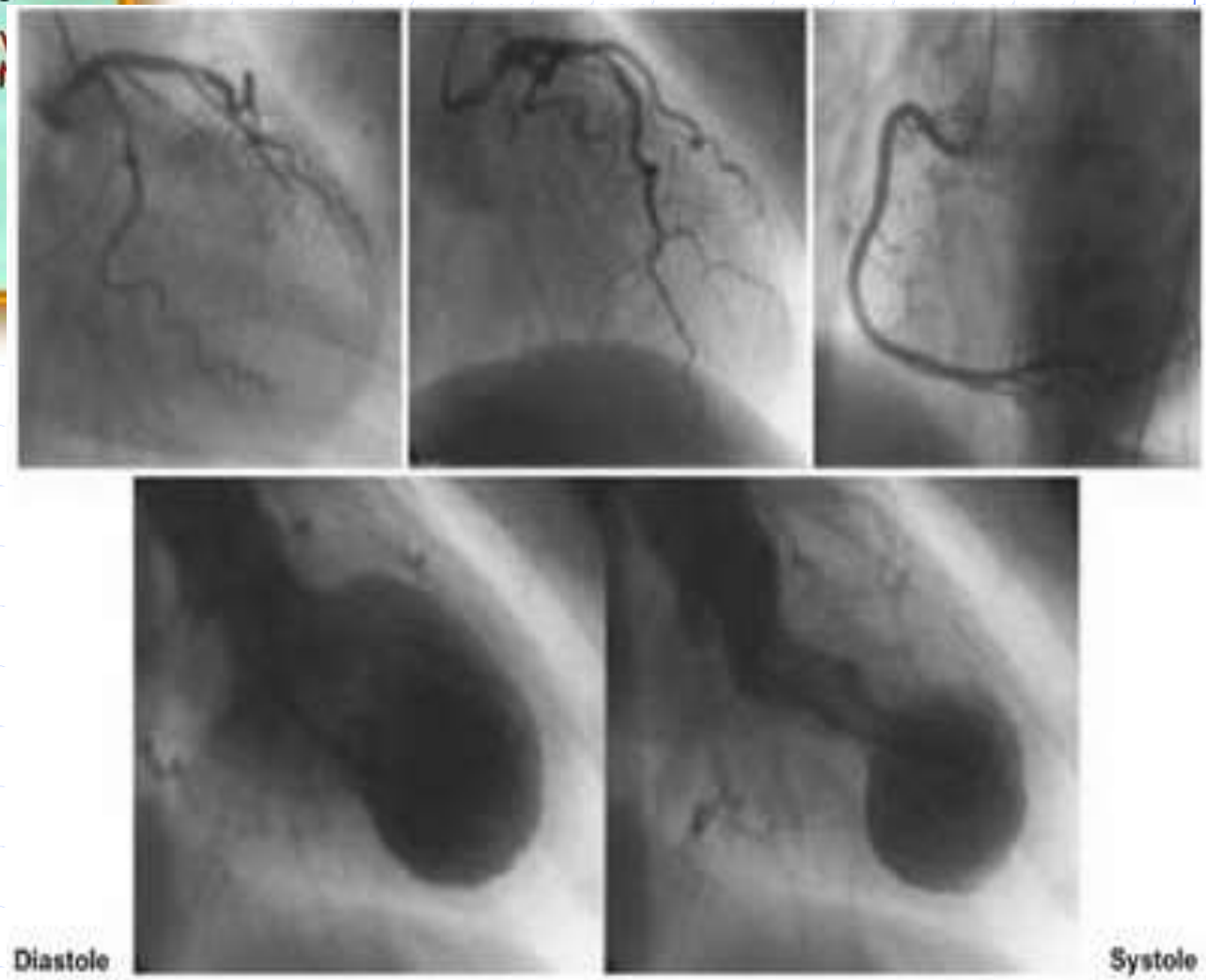
Family Screening

- 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


JAPANESE
FISHERMEN
ABOVE
(octopus trap
in japanese)
Tako-tsubo
Calm, happy
octopus

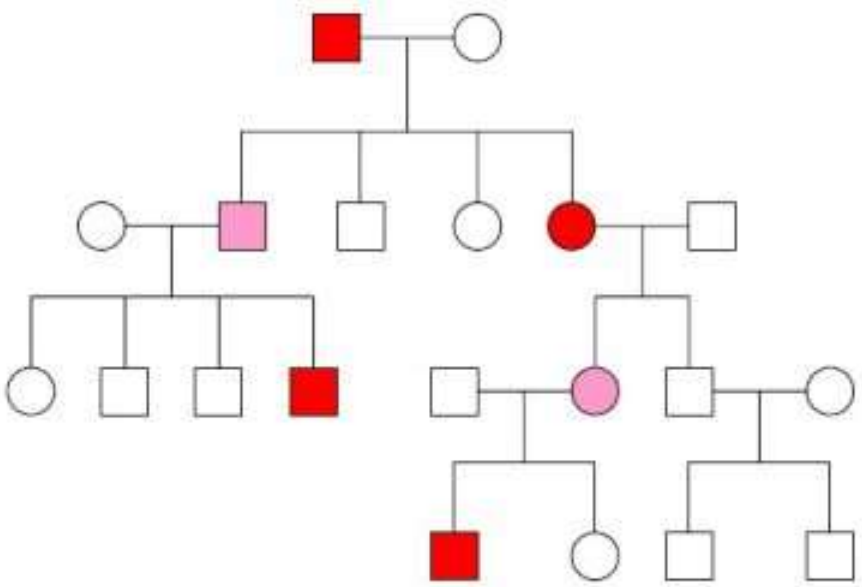

Diastole
Systole

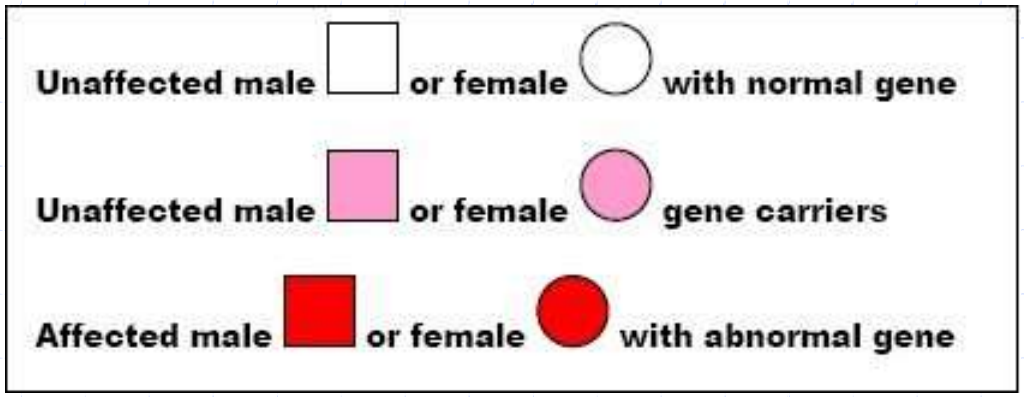
Tako-Tsubo

- 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

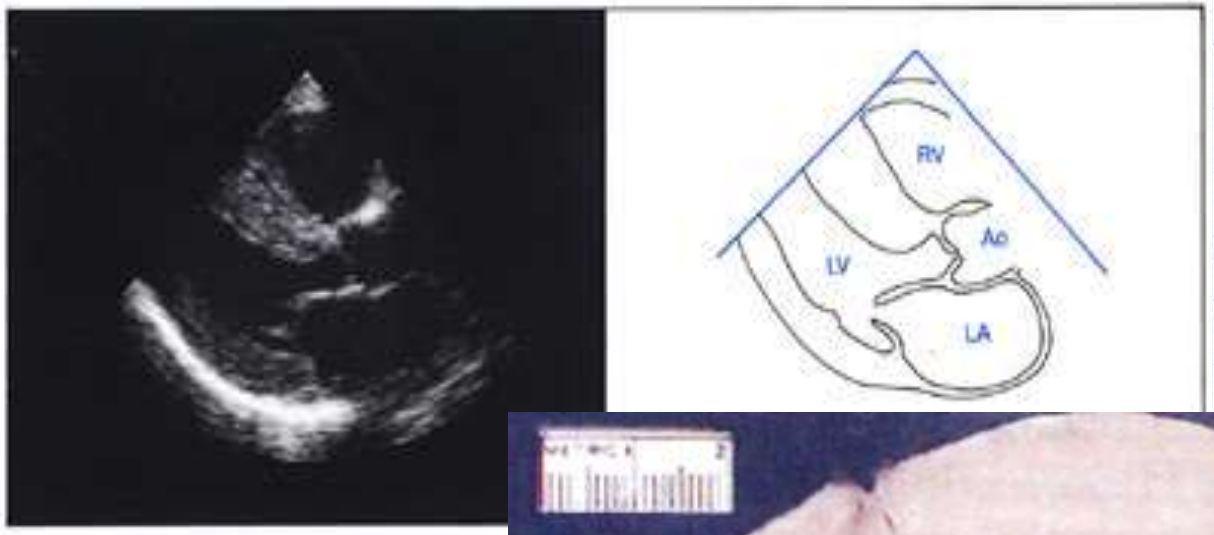
- Most frequent symptoms are dyspnea, chest pain, syncope (often post-exertion)
- Ventricular arrhythmias are common and SCD may occur, especially in athletes after extreme exertion

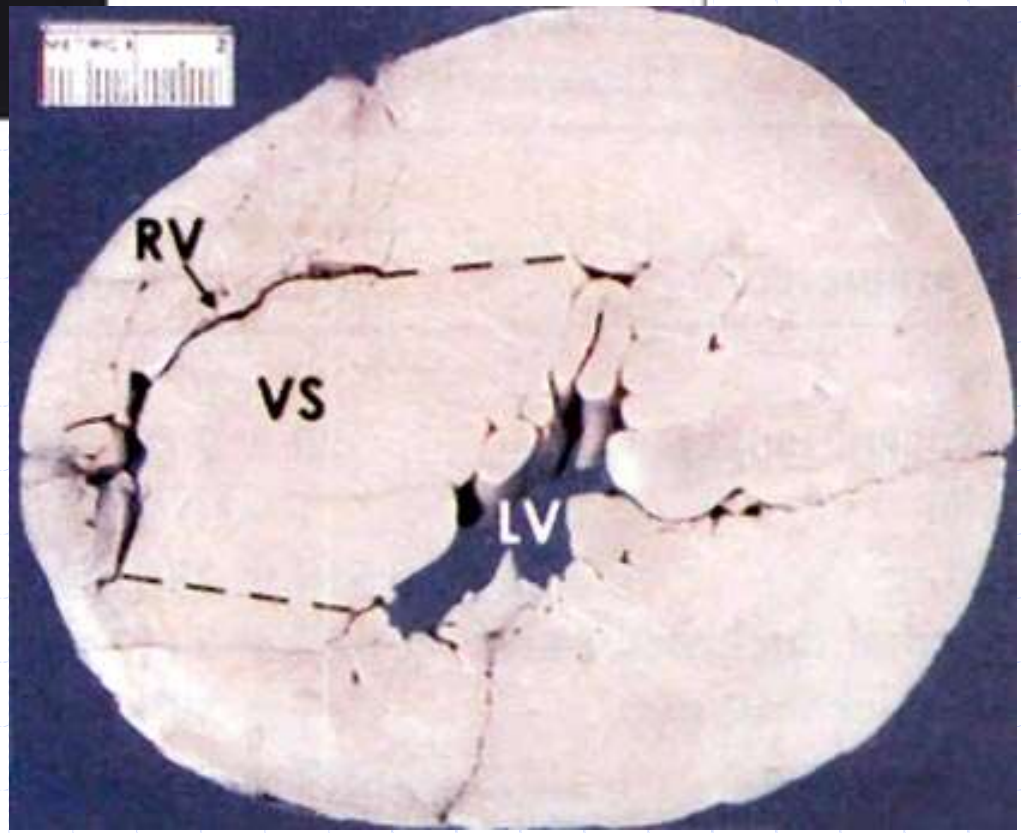
Physical Exam of HCM

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

Diagnostic Studies HCM

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


RV
Ao
LV
LA


RV
VS
LV

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

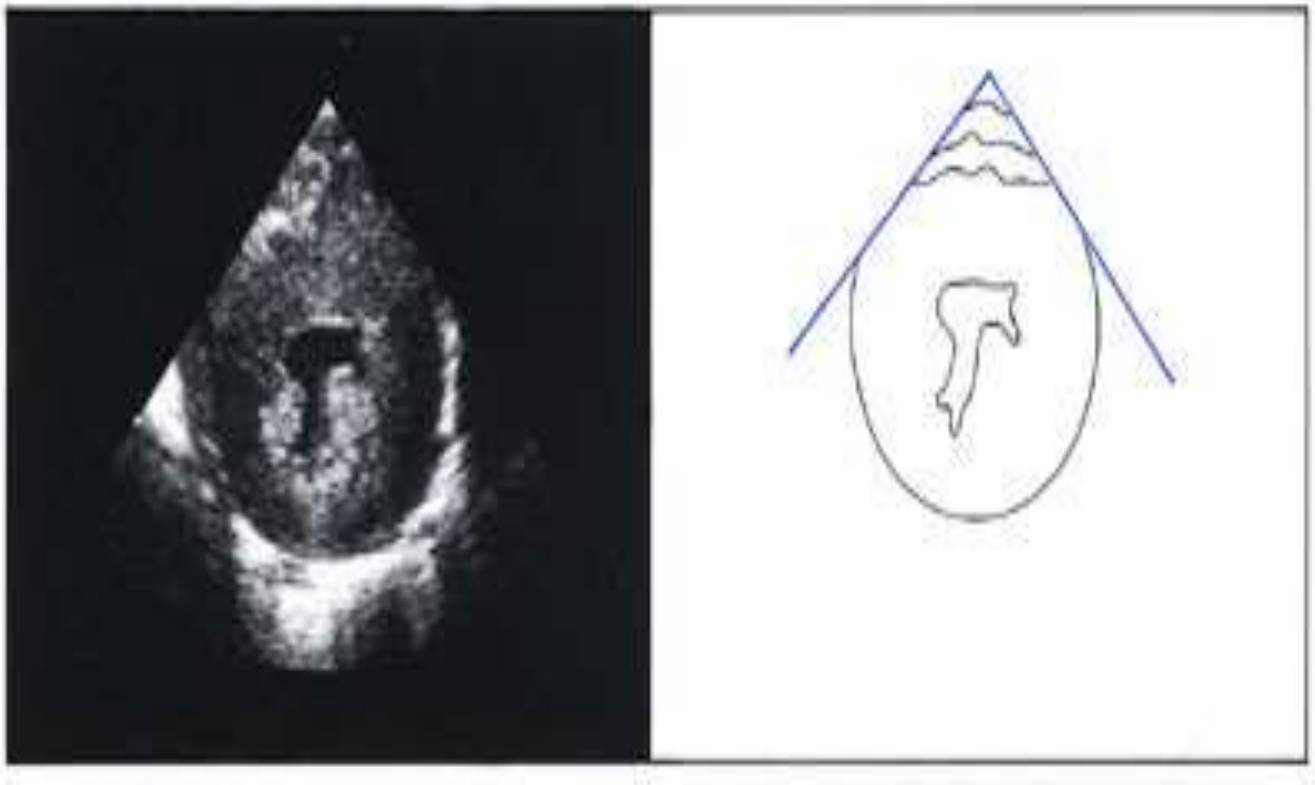
- Deposition of various proteins within myocardium
- Primary is from immunoglobulin light chains (often as consequence of multiple myeloma)
- Heart may be only involved organ

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