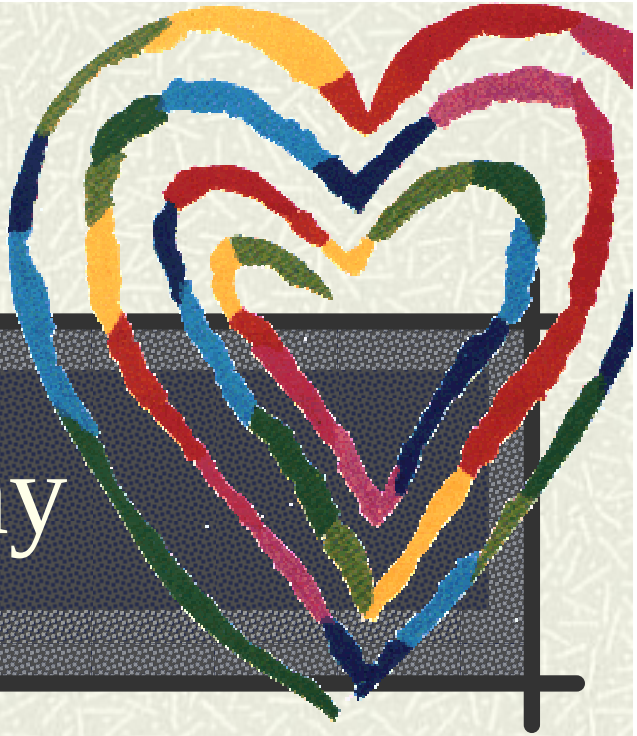
# Cardiomyopathy



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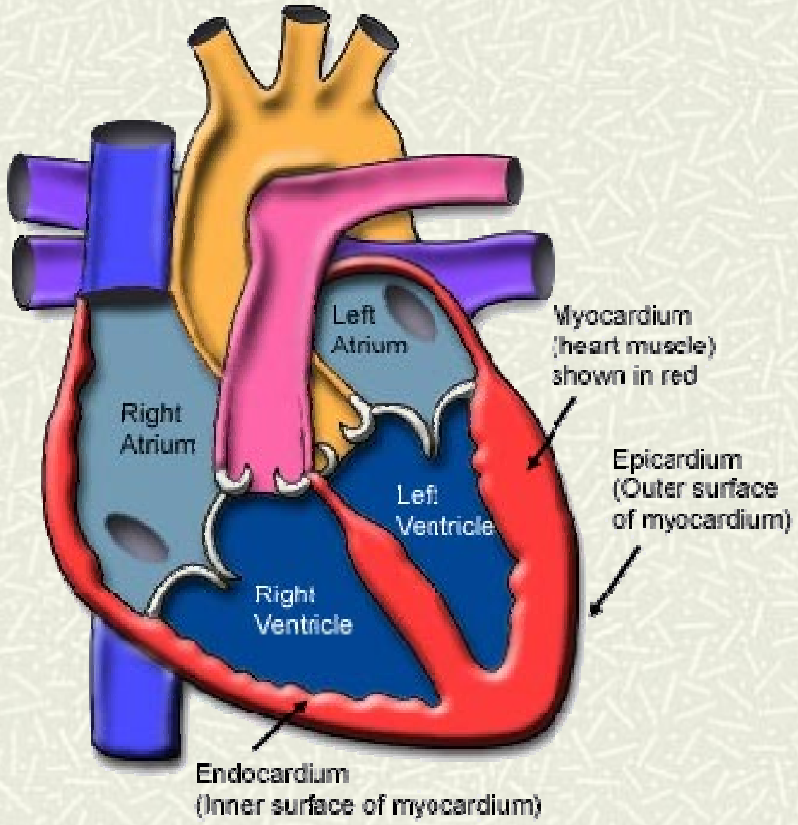
# Objectives

- Discuss the pathophysiology and etiologies of dilated, hypertrophic, and restrictive cardiomyopathy
- Review current medical and nursing management of patients with cardiomyopathy
- Discuss future trends in management

# Cardiomyopathy

- Irreversible primary, progressive disease of the heart muscle
- Damage to the myocardial cells
- Progressive deterioration




Predominant affect is on the myocardium

# Cardiomyopathy

"A diverse group of conditions whose final, common pathway is myocardial dysfunction"

Let's talk!

WHO/ISFC classification: Based on pathophysiological features

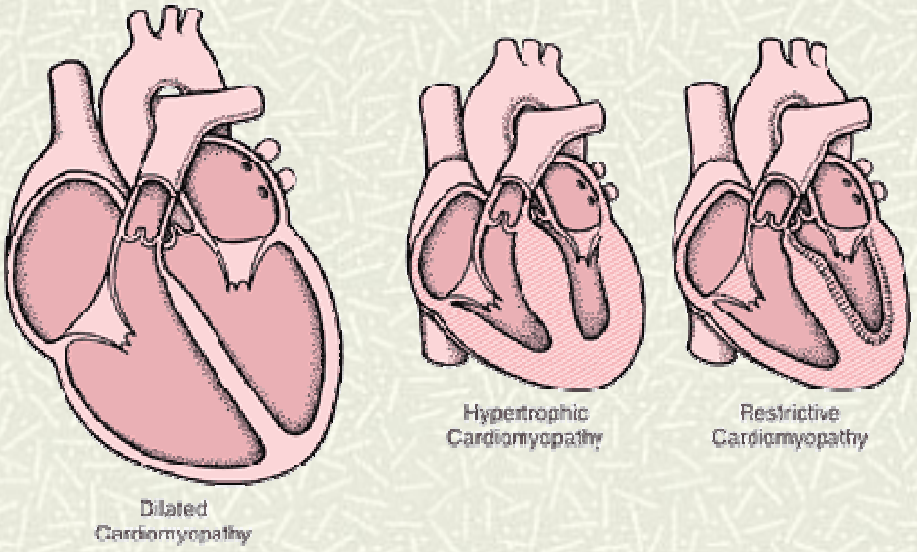
Primary & Secondary: CM & Specific CM

# Etiology

1. Cardiovascular disease (remodeling)
2. Infectious
3. Toxins
4. Systemic connective tissue diseases
5. Infiltrative and proliferative diseases
6. Nutritional deficiencies
7. Idiopathic

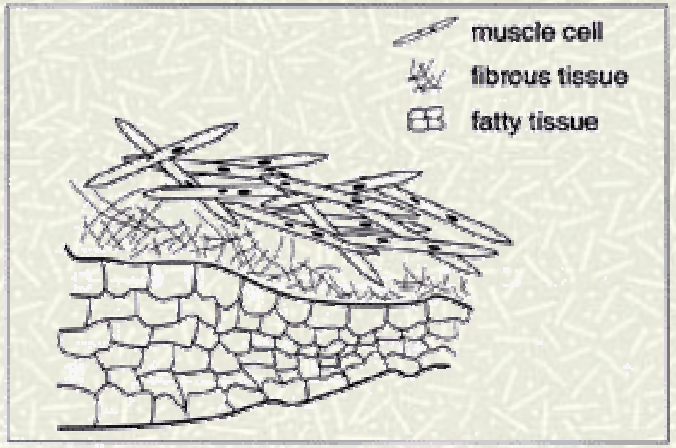
# Classification

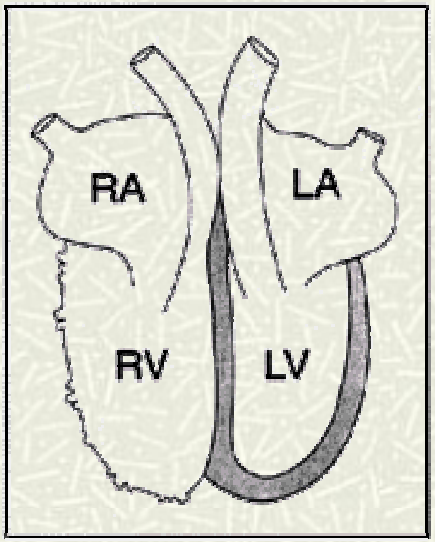
- Arrhythmogenic Right Ventricular CM
- Dilated
- Hypertrophic
- Restrictive
- Unclassified CM

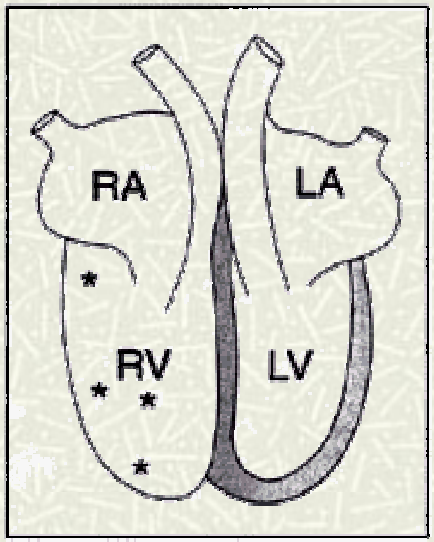


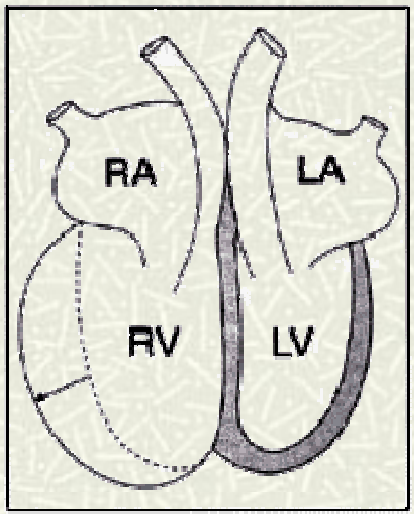
# Arrhythmogenic Right Ventricular Cardiomyopathy

Heart muscle disease characterized by replacement of the muscle by fibrous scar and fatty tissue. RV tends to be most affected.



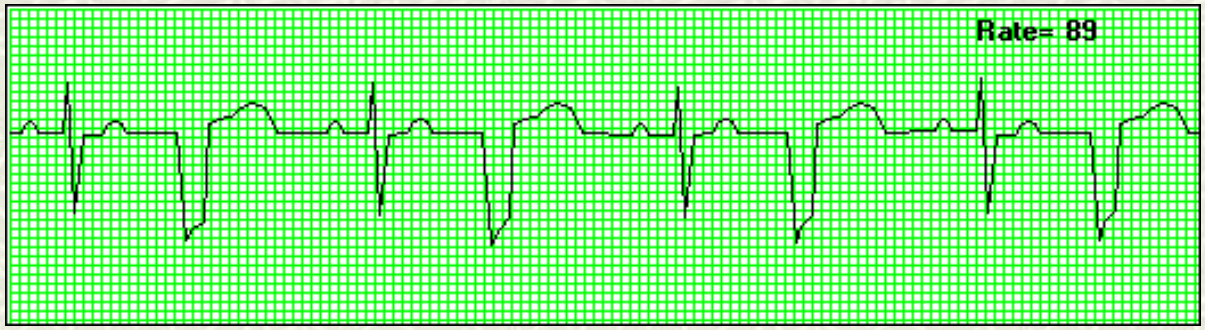



# Arrhythmogenic Right Ventricular Cardiomyopathy

- Symptoms early teens to second decade
- Etiology unknown (familial). Incidence: 1:3000-10,000
- Arrhythmia most prominent
- CHF, tricuspid regurgitation
- Symptoms
  - Palpitations
  - Syncope
  - Fatigue
  - Heart failure
  - Ventricular arrhythmias



# Arrhythmogenic Right Ventricular Cardiomyopathy

- Antiarrhythmics – Amiodarone/Sotolol
- ACE
- Anticoagulation
- Digoxin
- Diuretics

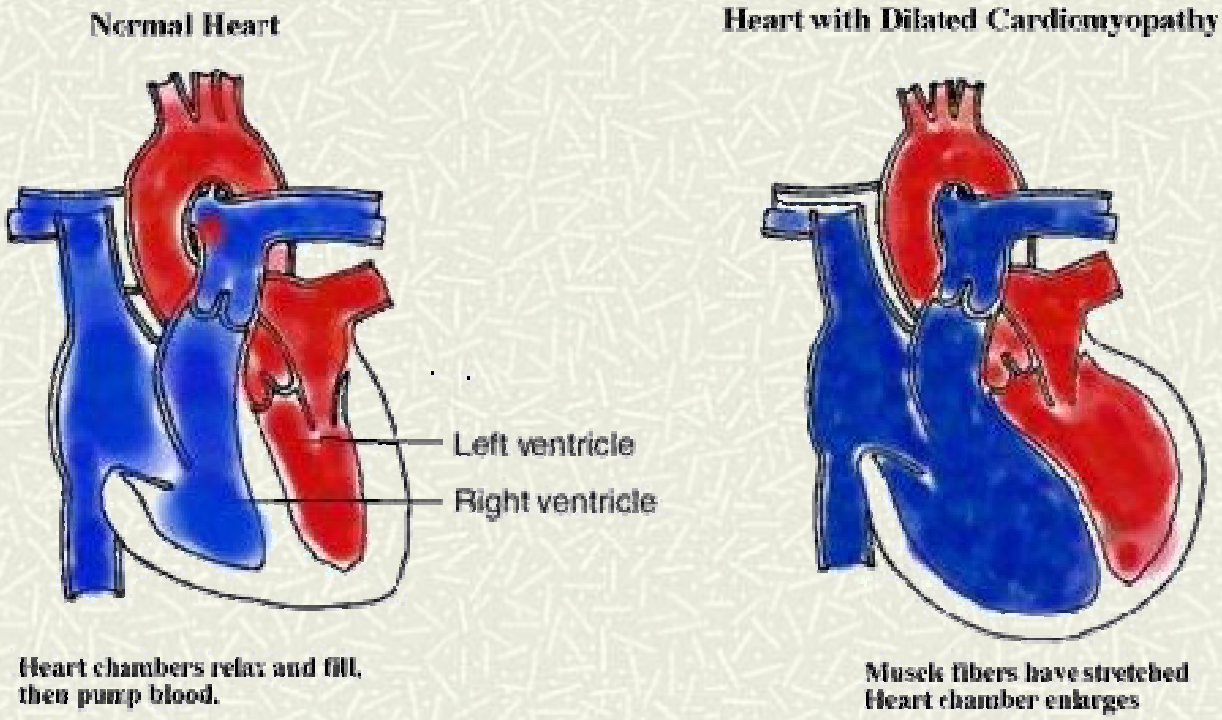
Arrhythmia most prominent
CHF, tricuspid regurgitation, embolus

# Arrhythmogenic Right Ventricular Cardiomyopathy

- Antiarrhythmics – Amiodarone/Sotolol
- ACE
- Anticoagulation
- Digoxin
- Diuretics
- Cardioversion
- Ablation
- Pacemaker
- ICD
- Surgery

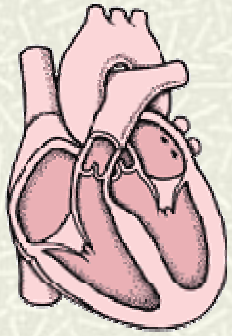
Arrhythmia most prominent
CHF, tricuspid regurgitation, embolus

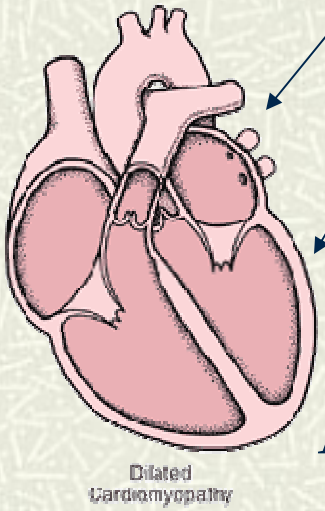
# Dilated Cardiomyopathy (DCM)




EF less than 40% in the presence of increased LV dimensions

# Dilated Cardiomyopathy (DCM)




Dilation and impaired contraction

- Cardiac enlargement
- Hypertrophy?
- Impaired **systolic** function of either or both ventricles

Apoptosis - necrosis - fibrosis

Laplace's law

# Dilated Cardiomyopathy (DCM)

- Most prominent CM
- Incidence – 36 cases/100,000 per year (Diagnostic criteria are lacking)
- Males and Africans
- Middle age
- IDCM - accounts for 25% of all heart failure cases

# DCM: Etiologies

- Primary - Idiopathic
- Secondary:
  - Electrolyte abnormalities
  - Endocrine abnormalities
  - Hypertension*
  - Infectious causes
  - Infiltrative diseases
  - Ischemia*
  - Neuromuscular diseases
  - Nutritional abnormalities
  - Rheumatologic diseases
  - Tachyarrhythmias
  - Toxins
  - Valvular heart disease*

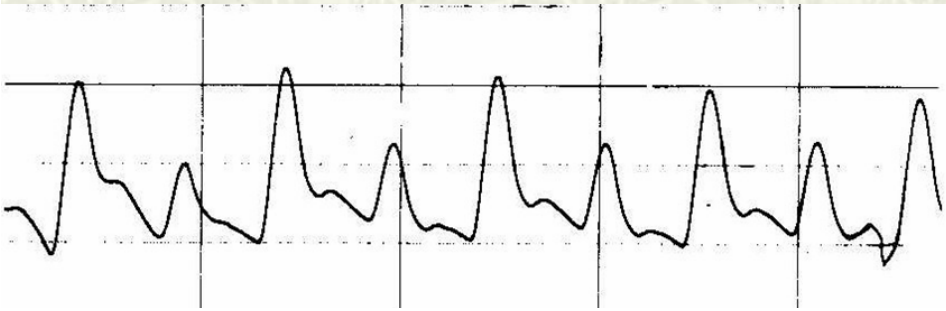
*WHO classified as specific cardiomyopathies

# Alcoholic Cardiomyopathy (DCM)

- CHF, HTN, CVA, arrhythmia, sudden death
- Major cause of secondary, nonischemic CM
  - 1/3 of all cases
- Three mechanisms
  - direct toxic effect of alcohol/metabolites
  - nutritional (thiamine)
  - alcohol additives (cobalt)
- Men 30-55 years of age >10 year consumption

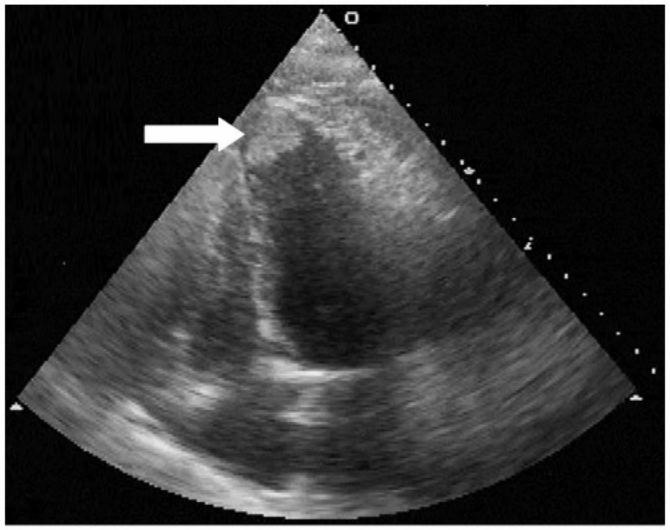
# DCM: Clinical Presentation

- Fatigue/weakness
- Weight loss
- Dyspnea on exertion
- Peripheral edema
- BP
- Pulsus alternans
- Pulsatile jugular veins
- Apical displacement
- S3 / S4
- Murmurs



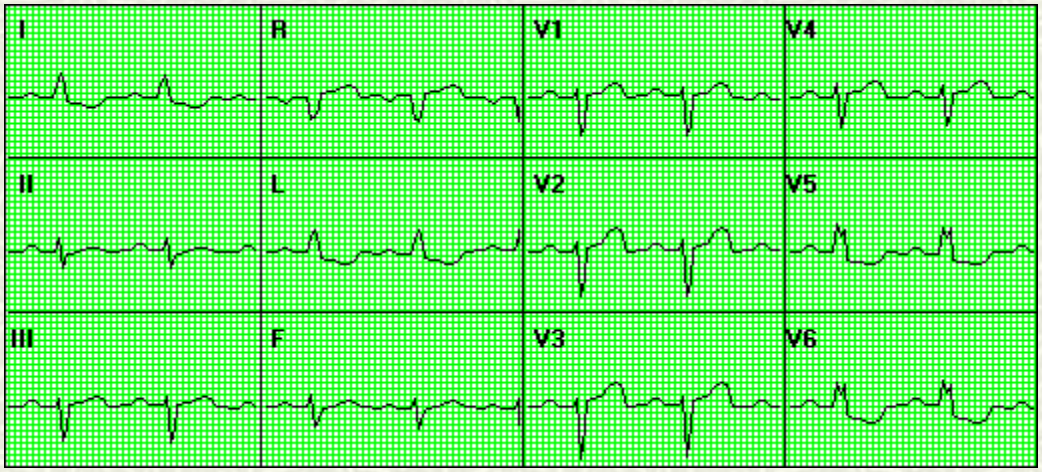
# DCM: Clinical Presentation

- Orthopnea
- PND
- Chest pain
- Abdominal pain
- Emboli
- Dysrhythmias
- Syncope
- Sudden death

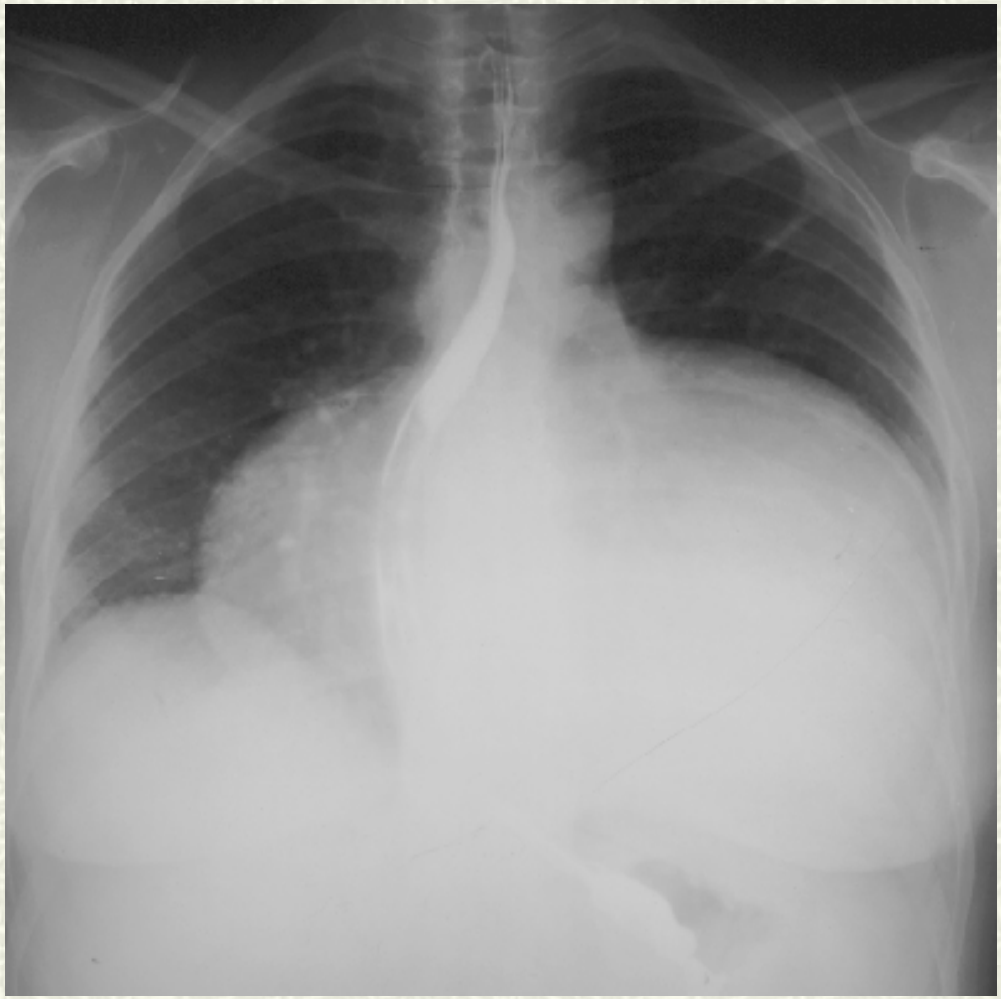


# DCM: Diagnostic Tests

- CXR - enlargement
- EKG - tachyarrhythmias, Q waves, R-wave
- Echocardiography - diffuse global dysfunction. (MV?)
- Catheterization



# Dilated Cardiomyopathy (DCM)

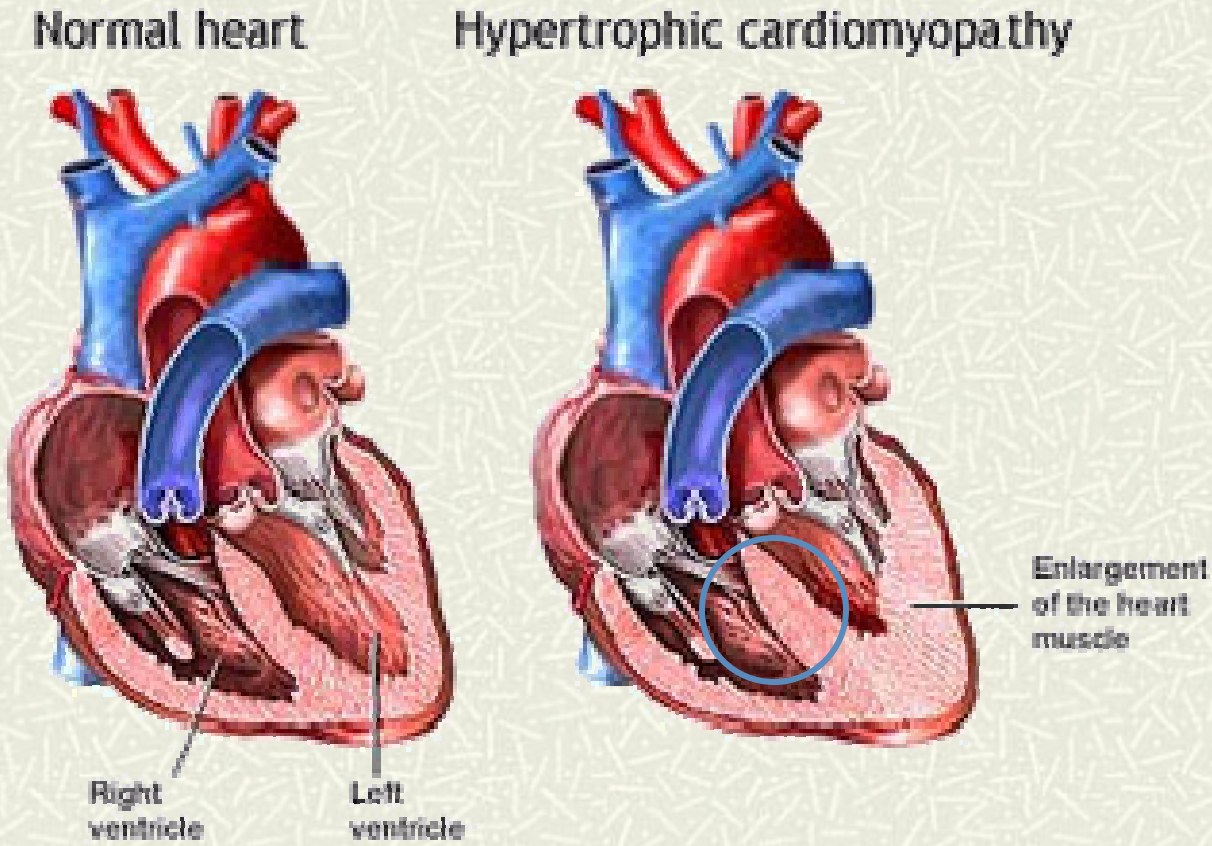


# DCM: Management

- Sodium restriction
- Vasodilators (arterial/venous)
- ACE, ARB
- Beta-Blockers
- Cardioversion
- Pacemakers
- Diuretics
- Anticoagulation
- Antiarrhythmics (amiodarone)
- Heart transplant

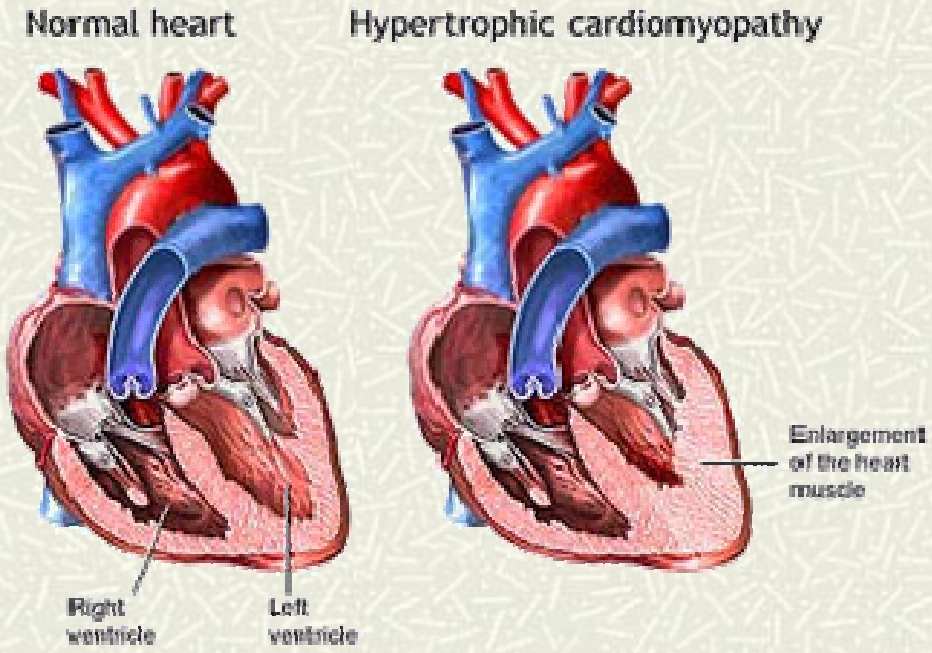
Adrenergic and renin-angiotensin systems

# Hypertrophic Cardiomyopathy




Stiffness of the LV with resultant impaired ventricular filling

# Hypertrophic Cardiomyopathy




Myocardial mass

Ventricular cavities

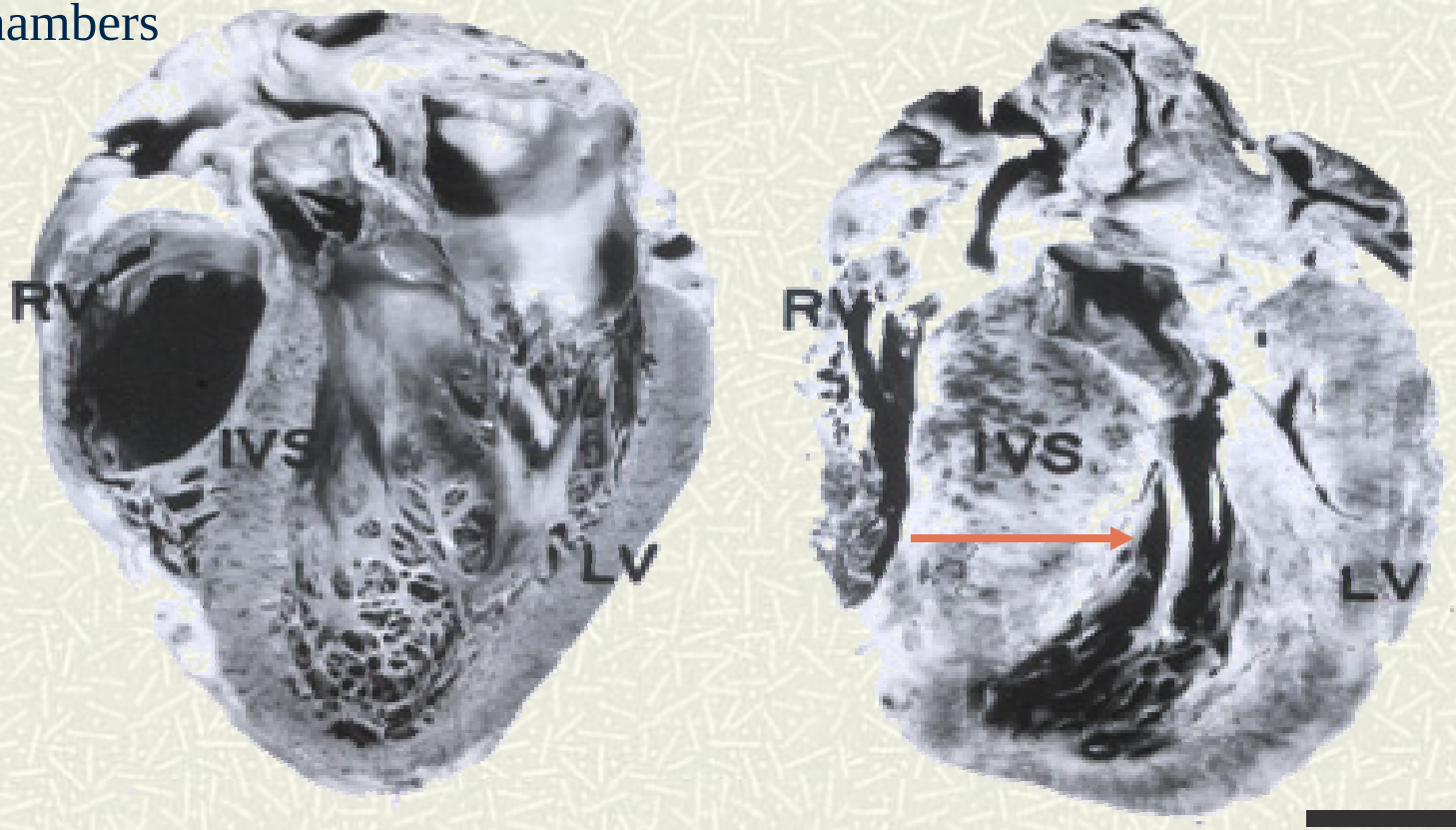
LV over RV

Atria

Heterogeneous

# Hypertrophic Cardiomyopathy

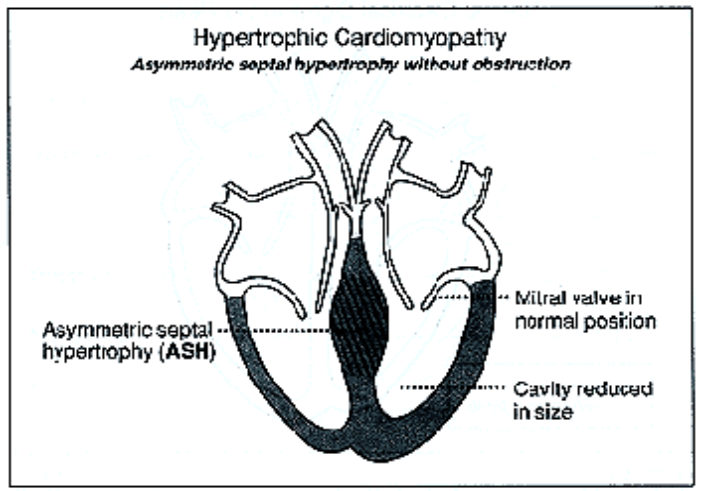
Disproportionate thickening of the of the intraventricular septum. Greater hypertrophy of the ventricular septum than of the ventricular chambers

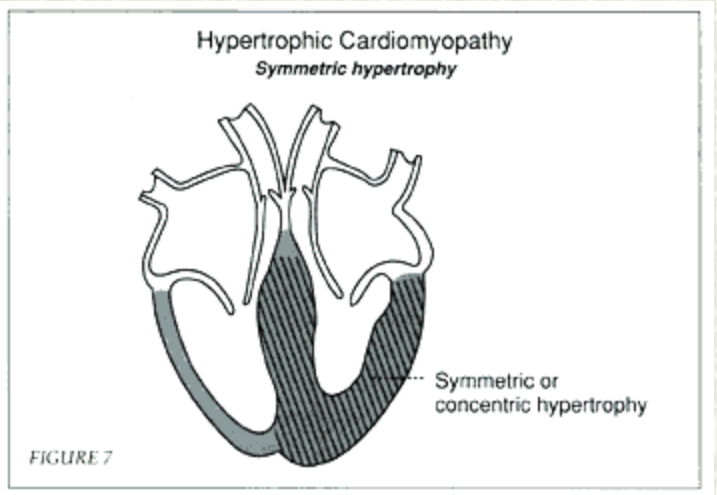


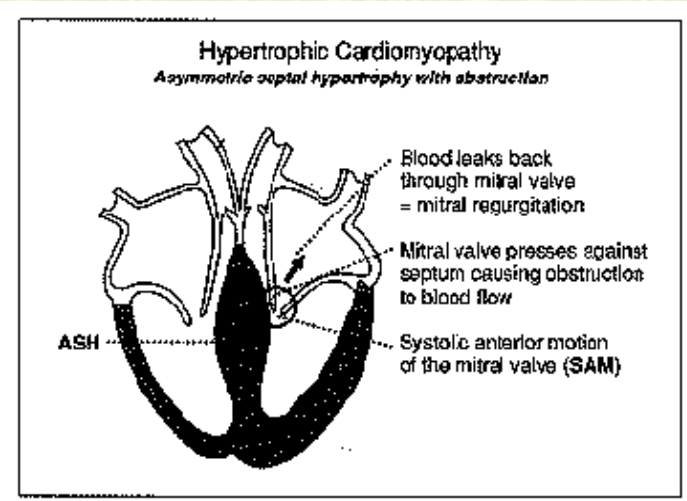
# Hypertrophic Cardiomyopathy

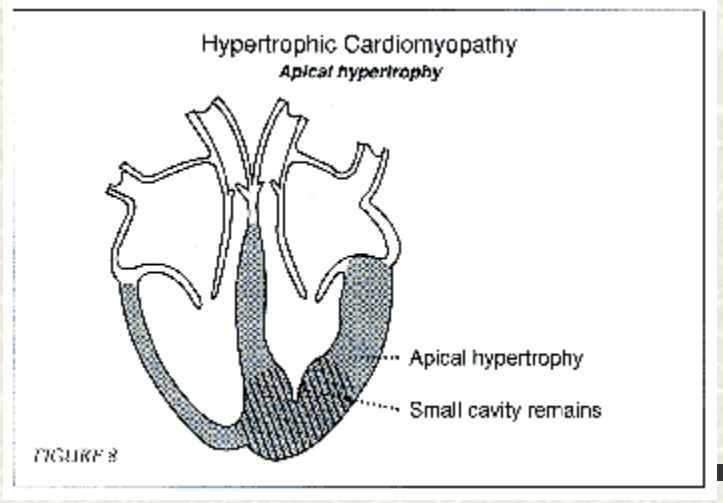
- Excessive thickening of the heart muscle.
- Myocardial disarray - normal alignment of muscle cells is absent
- Abnormalities of collagen deposition and altered contractile proteins in the myocytes (whole structure changes)
- Fibrosis – visible scar
- Myocardial ischemia - abnormal intramural coronary arteries

# Hypertrophic Cardiomyopathy



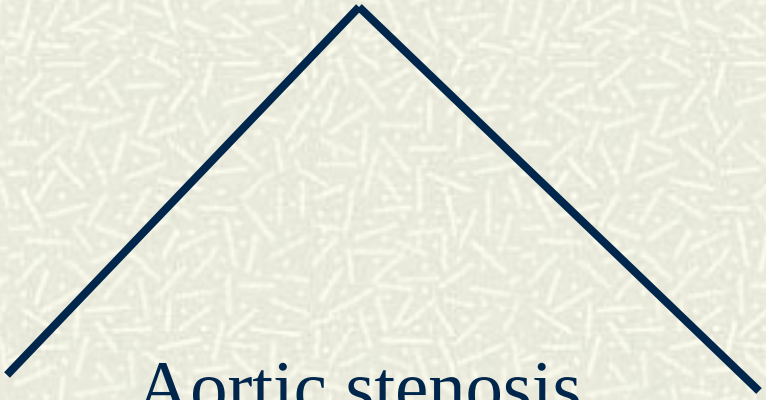
# Hypertrophic Cardiomyopathy

- ✓ **Rare genetic disease**

IHSS - Idiopathic hypertrophic subaortic stenosis

Asymmetric septal hypertrophy

Muscular subaortic stenosis



Aortic stenosis

Hypertension

HCM vs. physiological hypertrophy

**HCM in the elderly**

# Hypertrophic Cardiomyopathy

1. Hyperdynamic state – septal thickening
2. Diastolic dysfunction – thickened muscle usually contracts well but doesn't relax. Higher pressures result to allow expansion for the inflow of blood.
3. Possible outflow obstruction (~25%): MV involvement
4. Myocardial ischemia

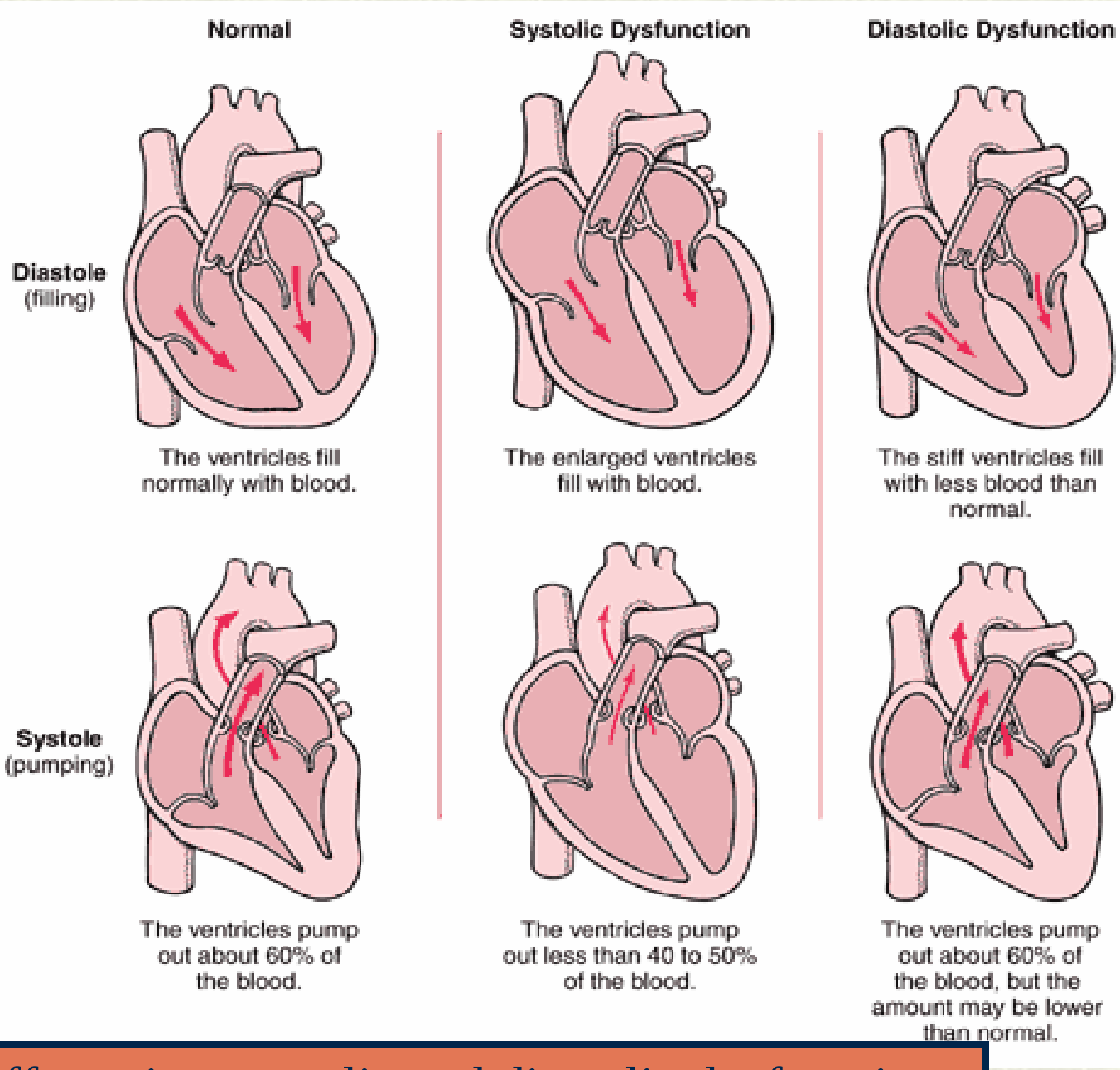
Systolic dysfunction

(pump)

vs.

Diastolic dysfunction

(fill)




Need to differentiate systolic and diastolic dysfunction

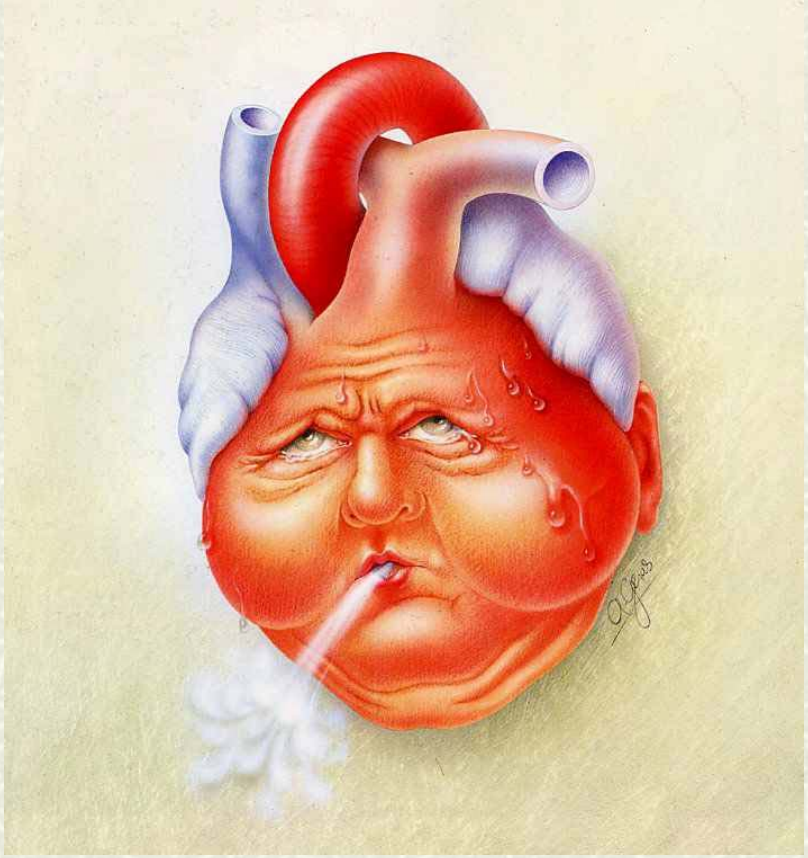
# Hypertrophic Cardiomyopathy

## Clinical Manifestations

- Mild to asymptomatic – screenings
  - Sudden death

- Dyspnea – most common from diastolic dysfunction

# Hypertrophic Cardiomyopathy



Clinical Manifestations:

- Angina, fatigue, syncope, dysrhythmias (more common)
- Palpitations, PND, CHF, dizziness (less common)

# Hypertrophic Cardiomyopathy

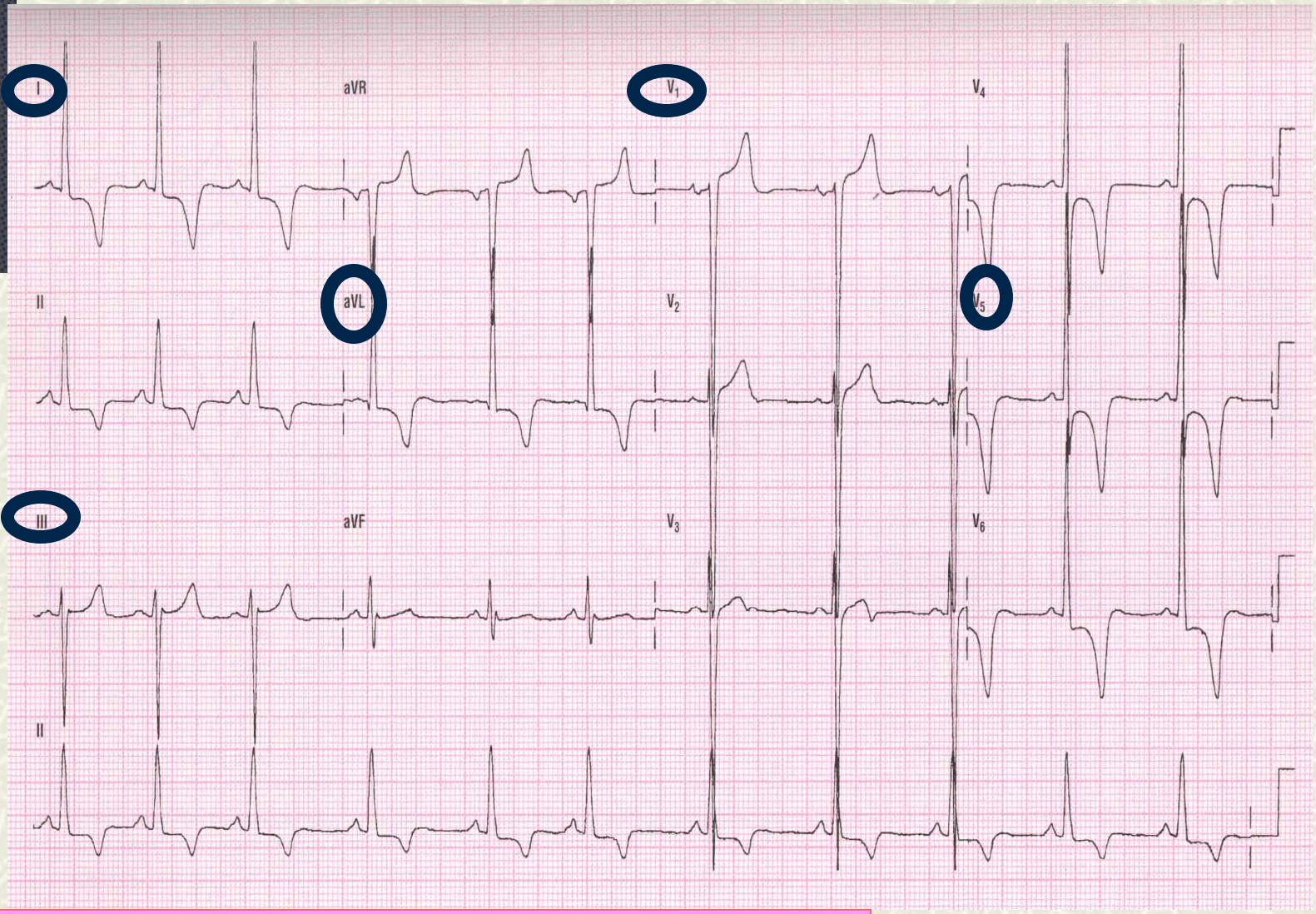
Clinical Manifestations

- S4 / S3 may be heard with outflow obstructions as well as a systolic murmur
- Ventricular arrhythmias – ¾ of patients
- SVT – ¼ to ½ of patients. Less tolerated
- EP testing – limited predictive value
- CXR – normal to cardiomegaly
- Squat position

# Hypertrophic Cardiomyopathy

## Echocardiogram – screening and diagnosis

- Cardinal sign is LV hypertrophy of septum and anterolateral free wall
- Variability in hypertrophy
- Dilated left atrium
- Normal to near-normal EF
- Septum at least 1.3 to 1.5 times the thickness of the posterior wall (15 mm). Average finding is 20 mm
- Outflow tract obstructions; MV / pressure gradient changes
- Diastolic dysfunction




R-wave in AVL >11mm;

R wave height in Lead I plus the S wave depth in Lead III > 25 mm

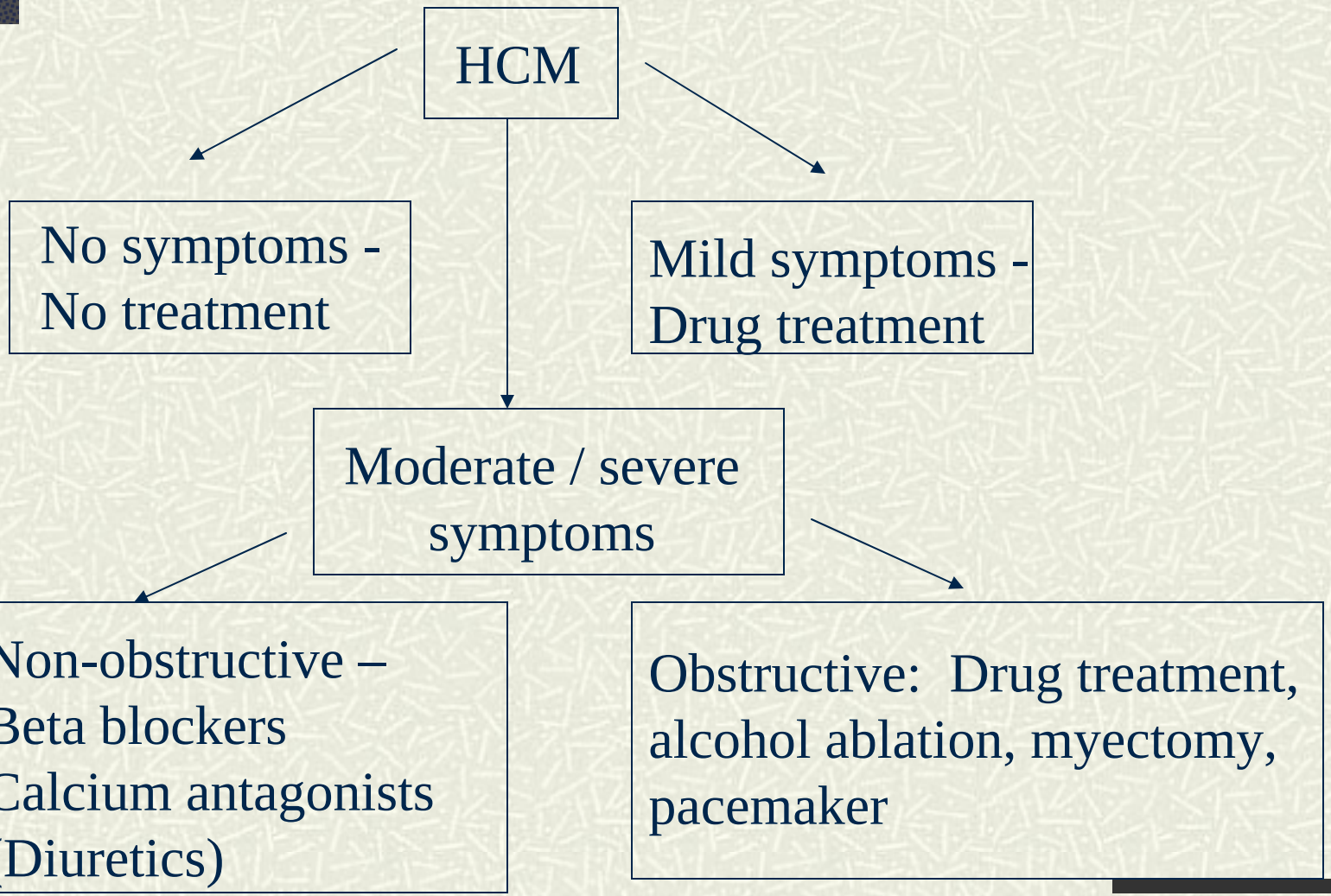
*S wave depth in V1 plus the height in V5 that exceeds 35 mm

# Hypertrophic Cardiomyopathy

## Morbidity / Mortality

- Mortality – 1%-3% per year
- Some remain stable or improve. Clinical deterioration is slow
- Sudden death – higher in children/ adolescents
- Patients with gradients are more likely to deteriorate
- Atrial fibrillation – may lead to increase symptoms (LA dilation)
- LV dilation and dysfunction (DCM) occurs in 5-10%. Wall thinning and scar formation

# Hypertrophic Cardiomyopathy



# Restrictive Cardiomyopathy

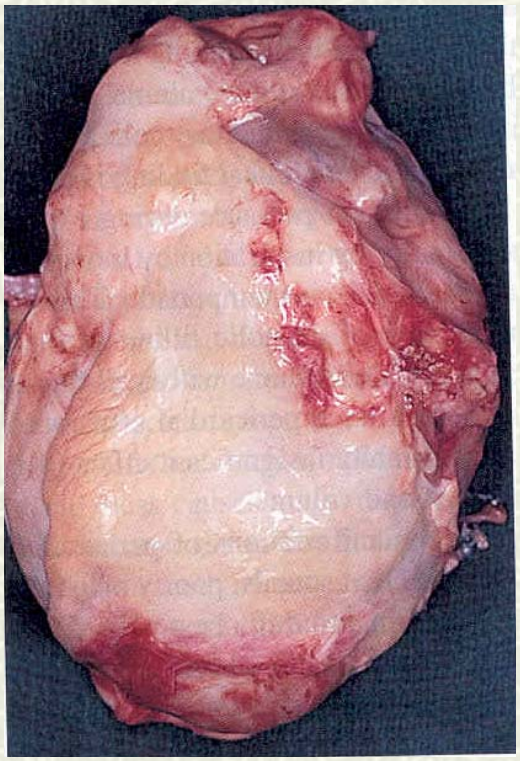
**Heart with Hypertropic Cardiomyopathy**

Growth and arrangement of muscle fibers are abnormal.
Heart walls thicken, especially in the left ventricle

**Heart with Restrictive Cardiomyopathy**

Ventricle walls stiffen and lose flexibility.

# Restrictive Cardiomyopathy

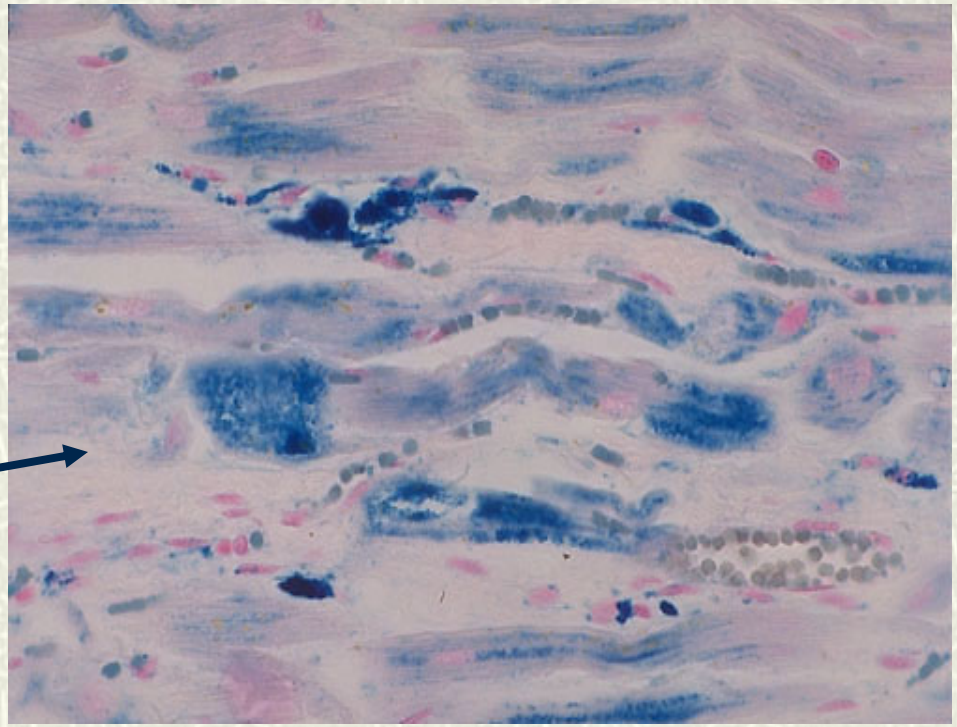


Restrictive pericarditis

- Myocardium becomes rigid, noncompliant
- Diastolic dysfunction
- Ventricular filling
- Systolic function preserved
- Resembles constrictive pericarditis
- Prevalence: <5% of CM in western world

# Restrictive Cardiomyopathy

- Idiopathic
- Noninfiltrative: Scleroderma
- Infiltrative: Amyloidosis, Sarcoidosis
- Storage Disease: Hemochromatosis
- Endomyocardial: Metastatic cancers Radiation



# Restrictive Cardiomyopathy

Clinical heart failure – right failure prominent

- JVD
- S3, S4, or both
- Elevation in CVP
- Peripheral edema, liver enlargement, ascites
- Exercise intolerance
- Weakness
- Dyspnea
- AV block
- Symptomatic bradycardia
- Atrial fibrillation

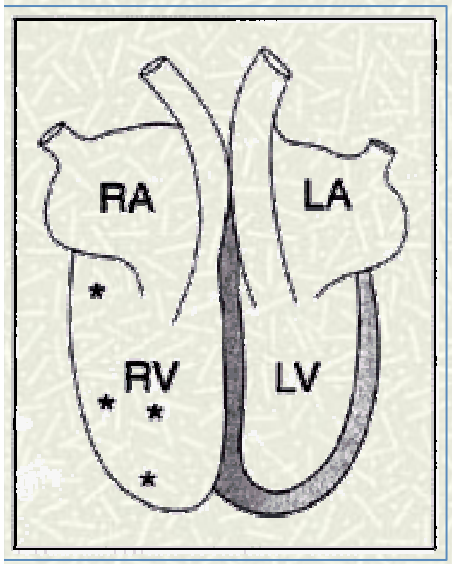
# Restrictive Cardiomyopathy

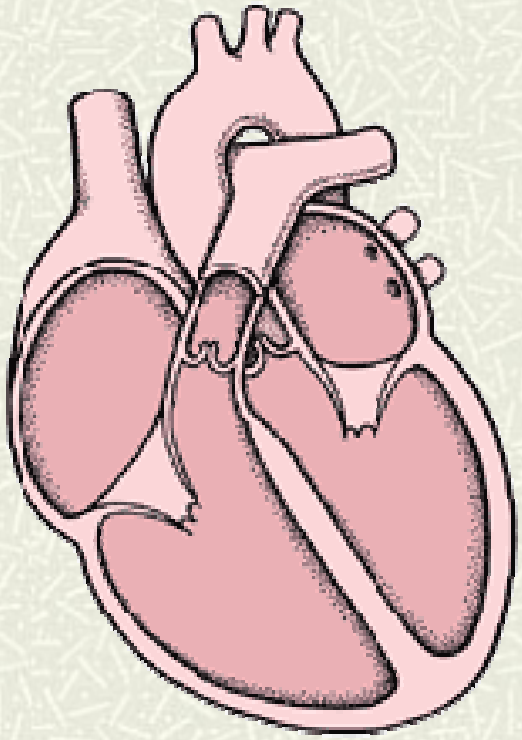
- CXR
- CT / MRI
- Echocardiogram – dilated atria, increased early LV filling velocity, decreased atrial filling velocity, and decreased isovolumetric relaxation time
- Endomyocardial biopsy

# Restrictive Cardiomyopathy

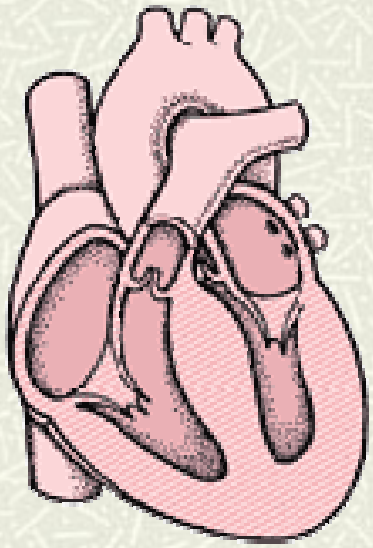
- No satisfactory medical therapy (treat secondary causes)
- Drug therapy must be used with caution:
  - Diuretics for extremely high filling pressures
  - Vasodilators may decrease filling pressure
  - ?Calcium channel blockers to improve diastolic compliance
  - Digitalis and other inotropic agents are not indicated

# Summary

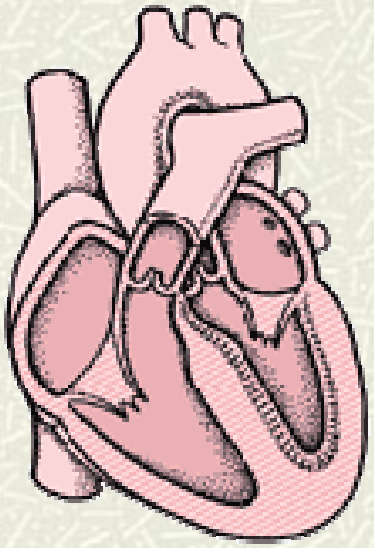


Dilated
Cardiomyopathy



Hypertrophic
Cardiomyopathy



Restrictive
Cardiomyopathy