Cardiomyopathies
and
Myocarditis

Dr. Ali KHADDAM,MD,MRCP(UK)
Interventional Cardiology
Damascus University
Syrian Private University

Cardiomyopathies - Definition

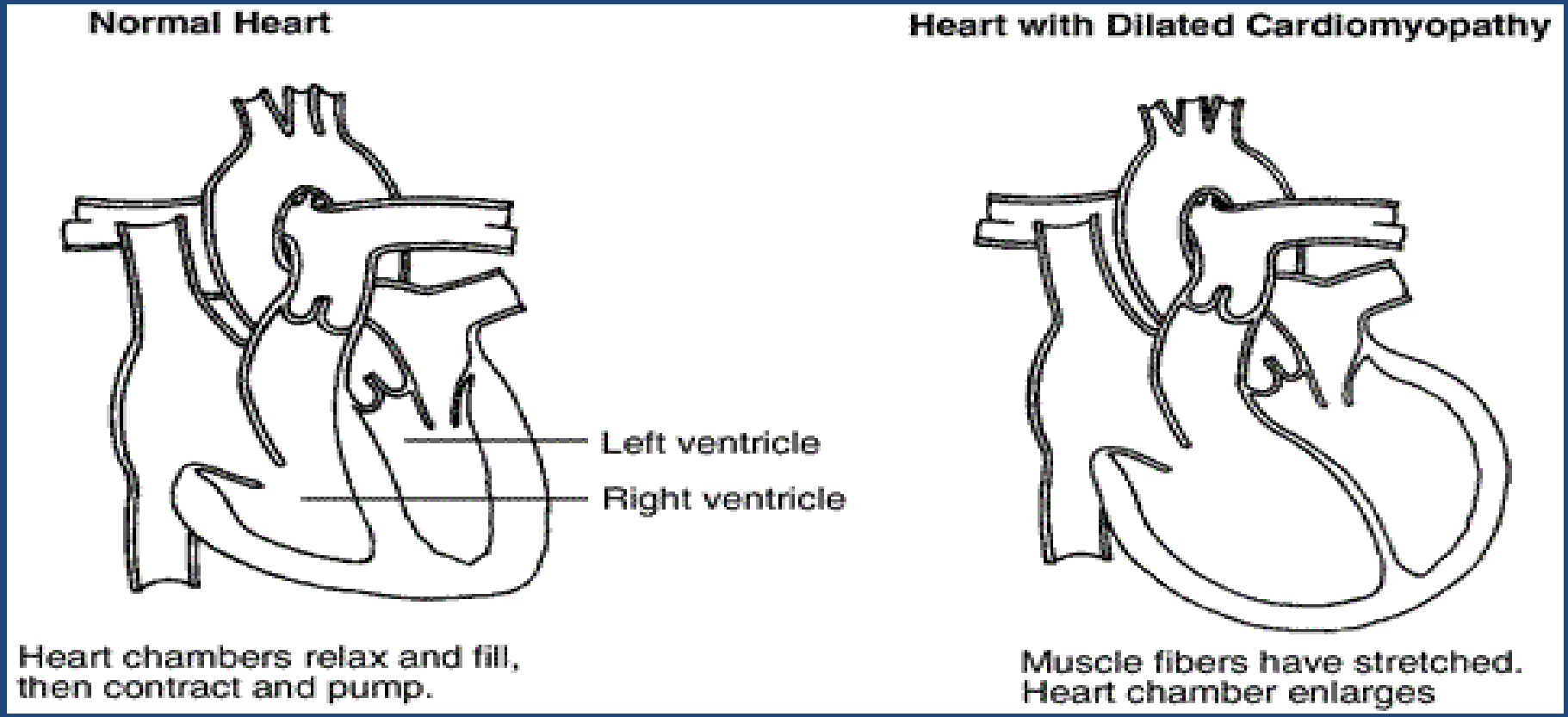
- Group of entities that affect the myocardium primarily and are **NOT** associated with another major cause of heart disease (like coronary artery disease, valvular heart disease)
- They are either confined to the heart or could be part of generalized systemic disorder, often leading to progressive heart failure- related disability or CV death .

Cardiomyopathy

- **Classification** (2 systems)
  - Based on Suspected Etiology
    - Primary
      - may be genetic, mixed (genetic or non genetic), or acquired
    - Secondary
      - which are accompanied by other organ system involvement
  - **WHO** classifies on the basis of their pathologic or pathophysiologic features (more accepted system)
    - All are based on echocardiography
      - Dilated – usually mostly Systolic dysfunction
      - Restrictive – usually mostly Diastolic dysfunction
      - Hypertrophic – usually mostly Diastolic dysfunction

Dilated Cardiomyopathy

Schematic of Dilated CM



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Dilated Cardiomyopathy

- Dilation of 4 chambers (V>A)
- Depressed systolic function and pump failure with low cardiac output
- Most of DCM cases are idiopathic
- African Americans and males have 2.5x increased risk
- Most common age of diagnosis 20-50yrs

Causes of **DCM**

- Idiopathic
- Toxin induced: EtOH, Anthracycline, Cobalt, Cocaine
- Infectious: Viral (coxsackievirus, adenovirus, parvovirus, human immunodeficiency virus [HIV]) , Parasites (Chagas' Disease)
- High Ouput States: Anemia, Peripartum, Thiamine deficiency, Thyrotoxicosis
- Radiation
- Sarcoidosis
- Amyloidosis
- Hemochromatosis
- Genetic

Primary dilated CM

- Incidence 5-8 cases per 100,000
- Prevalence 36 per 100,000
- mortality rate 25% at 1 year, 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

Idiopathic DCM-diagnosis of exclusion

- Responsible for half of DCM diagnoses
- Exclusion of > 50% obstruction of one or more coronary arteries
- Exclusion of active myocarditis
- Exclusion of a primary or secondary form of heart muscle disease

DCM - Genetic causes

- About 20-30% of dilated cardiomyopathy cases have been reported as familial
- Autosomal Dominant trait with variable penetrance

DCM - 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 echo .
- After initial screen, repeat every 3-5 years

DCM - Clinical Presentation

- Signs and symptoms of CHF-dyspnea on exertion, orthopnea and pnd.
- Chest pain can occur due to low coronary vascular reserve
- Mural thrombi formation can occur (Manifestations of embolizaiton: neurologic deficits, flank pain, hematuria, pulseless cyanotic extremity)
- Fatigue (decreased cardiac output)
- LL edema

DCM - Sequelae

- Ventricular and supraventricular arrhythmias
- Conduction system abnormalities
- Thromboembolism
- Sudden or heart failure–related death.

DCM - Physical Exam

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

DCM - Diagnostics

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

DCM - Medical Management

- Similar to congestive heart failure : **diuretics**, **inotrops .**
  - Clinical outcome has improved with **ACEI** and more recently **B-Blocker** therapy
  - **Anticoagulants**: all with mural thrombi; evidence of pulmonary or systemic emboli; AF
- **Supportive**: avoidance of exercise, salt and fluid restriction, ECG monitoring for arrhythmias.

DCM – Invasive Treatment

- **ICD**
- **Cardiac transplantation**
  - This disorder is the most common indication for cardiac transplantation
  - Survival after transplant is
    - 80% one year
    - 70% 5 years
- **Left Ventricular Reduction Procedures**
  - LV-reshaping

DCM - 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

Hypertrophic Cardiomyopathy

HCM - Historical Perspective

- HCM was initially described by Teare in 1958
  - Found massive hypertrophy of ventricular septum in small cohort of young patients who died suddenly
- Braunwald was the first to diagnose HCM clinically in the 1960s
- Most common form of cardiomyopathy
- Many names for the disease
  - Idiopathic hypertrophic subaortic stenosis (IHSS)
  - Muscle subaortic stenosis
  - Hypertrophic obstructive cardiomyopathy (HOCM)

HCM

- Hypertrophy of the LV, and occasionally of the RV
- Prevalence of HCM in the absence of aortic valve disease or systemic HTN is at least 1:500 of the adult
- Interventricular septum is typically more prominently involved than the LV free wall
- LV volume is normal or reduced in HCM, and diastolic dysfunction is usually present
- 50% of cases are inherited via an autosomal dominant transmission
- Most common acquired causes are HTN & Aortic Stenosis

HCM - Pathophysiology

- Dynamic LV outflow tract obstruction
  - Outflow tract gradient (≥30 mm Hg), considered severe if >50 mm Hg (occurs in 25-30% of cases leading to name hypertrophic obstructive cardiomyopathy)
- Diastolic dysfunction
  - Impaired diastolic filling, ↑ filling pressure
- Myocardial ischemia
- Mitral regurgitation
- Arrhythmias

HCM - Pathophysiology

- **Diastolic Dysfunction**
  - Contributing factor in 80% of patients
  - Impaired relaxation
    - High systolic contraction load
    - Ventricular contraction/relaxation not uniform
  - Accounts for symptoms of exertional dyspnea
    - Abnormal diastolic filling → increased pulmonary venous pressure

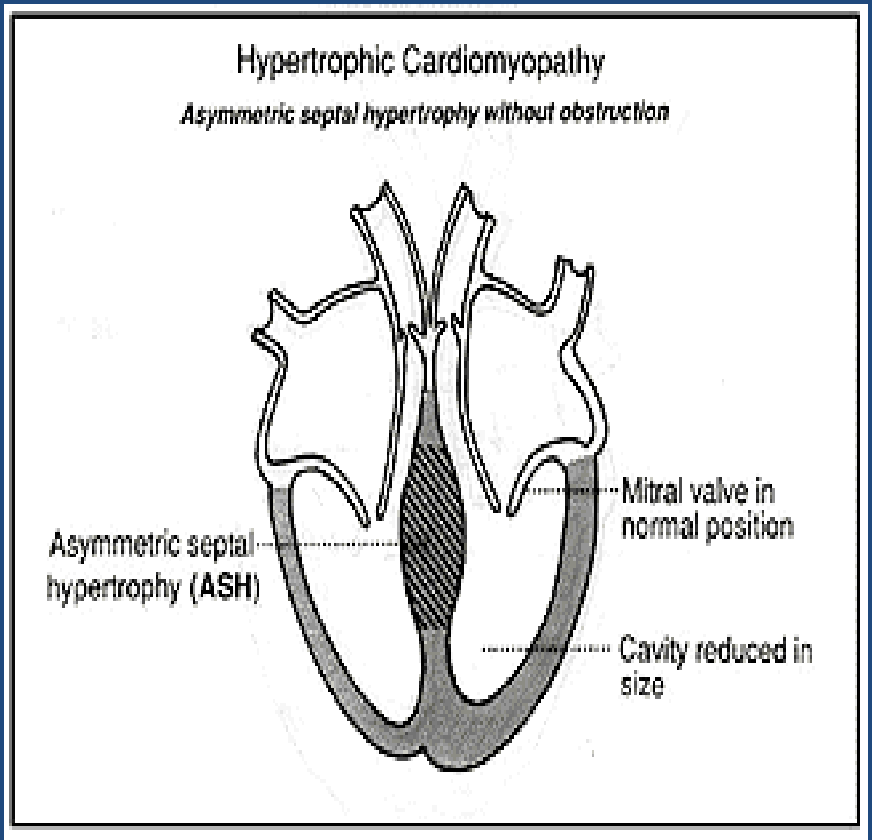
HCM - Pathophysiology

- **Myocardial Ischemia**
  - Often occurs without atherosclerotic coronary artery disease
  - Postulated mechanisms
    - Abnormally small and partially obliterated intramural coronary arteries as a result of hypertrophy
    - Inadequate number of capillaries for the degree of LV mass

HCM - Pathophysiology

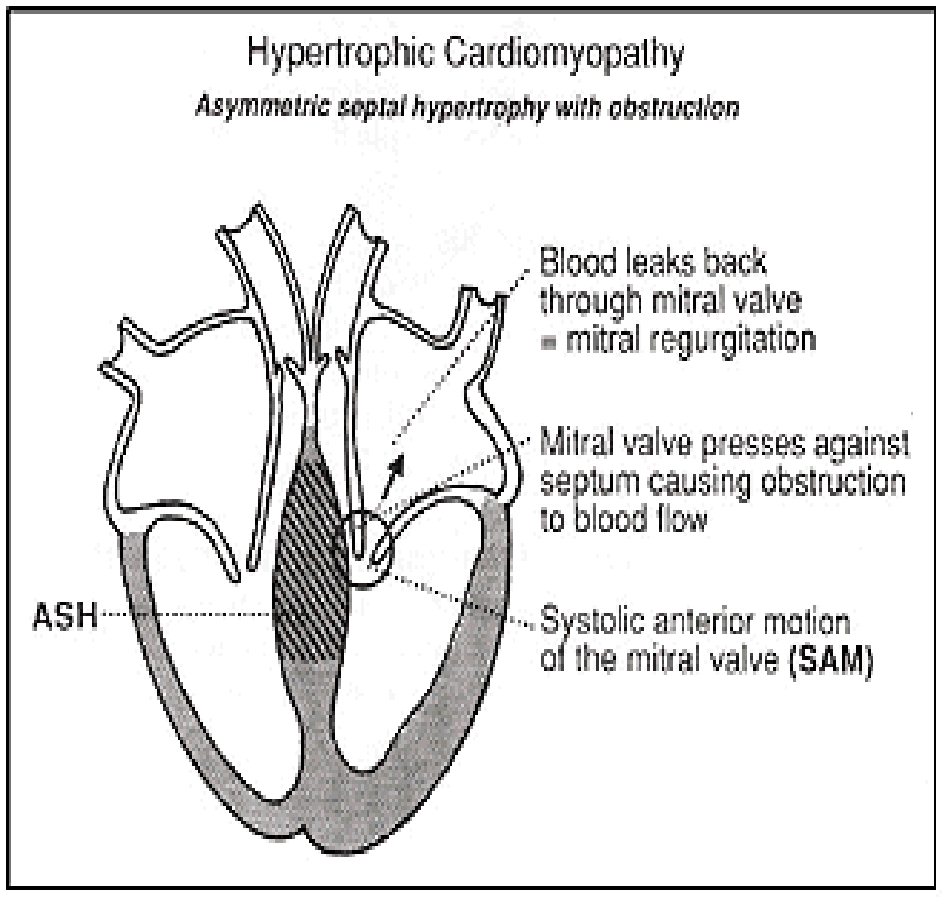
- **Mitral Regurgitation**
  - Results from the systolic anterior motion of the mitral valve
  - Severity of MR directly proportional to LV outflow obstruction
  - Results in symptoms of dyspnea, orthopnea in HCM patients

HCM – ASH Without Obstruction



- The major abnormality of the heart in HCM is an excessive thickening of the muscle. Thickening usually begins during early adolescence and stops when growth has finished.
- LV is almost always affected, and in some patients the muscle of RV also thickens
- Hypertrophy is usually greatest in the septum. The muscle thickening in this region may be sufficient to narrow the LVOT. This thickening is associated with obstruction to the flow of blood out of the heart into the aorta

HCM – ASH With Obstruction



- Asymmetric septal hypertrophy with obstruction to the outflow of blood from the heart may occur. The mitral valve touches the septum, blocking the outflow tract. Some blood is leaking back through the mitral valve causing mitral regurgitation

HCM - LVOT Gradient

- Approximately 25% of patients with HCM have a dynamic systolic pressure gradient in the left ventricular outflow tract caused by contact between the mitral valve leaflet(s) and the interventricular septum under resting conditions
- Outflow tract gradient in excess of **30 mmHg** is an important cause of symptoms

HCM - LVOT Gradient (Cont)

- Gradient greater than **50 mmHg**, the percentage of systolic volume ejected before the beginning of SAM is greatly reduced - responsible for patients' symptoms
- when severe, **outflow tract gradient** can cause dyspnea, chest pain, syncope, and predisposes to the development of atrial arrhythmias - independent predictor of disease progression and adverse outcome, including sudden death

HCM - LVOT Obstruction

- **Physiological Consequences of Obstruction**
  - Elevated intraventricular pressures
  - Prolongation of ventricular relaxation
  - Increased myocardial wall stress
  - Increased oxygen demand
  - Decrease in forward cardiac output

HCM - Clinical Presentation

- Asymptomatic (only discovered on echo)
- Dyspnea on exertion (90%), orthopnea, PND
- Angina (70-80%)
- Syncope (20%), Presyncope (50%)
  - outflow obstruction worsens with increased contractility during exertional activities
- Palpitations due to arrhythymias
- Symptoms severity increase with age
- Sudden cardiac death (due to v-tach/fib)
  - HCM is most common cause of SCD in young people, including athletes

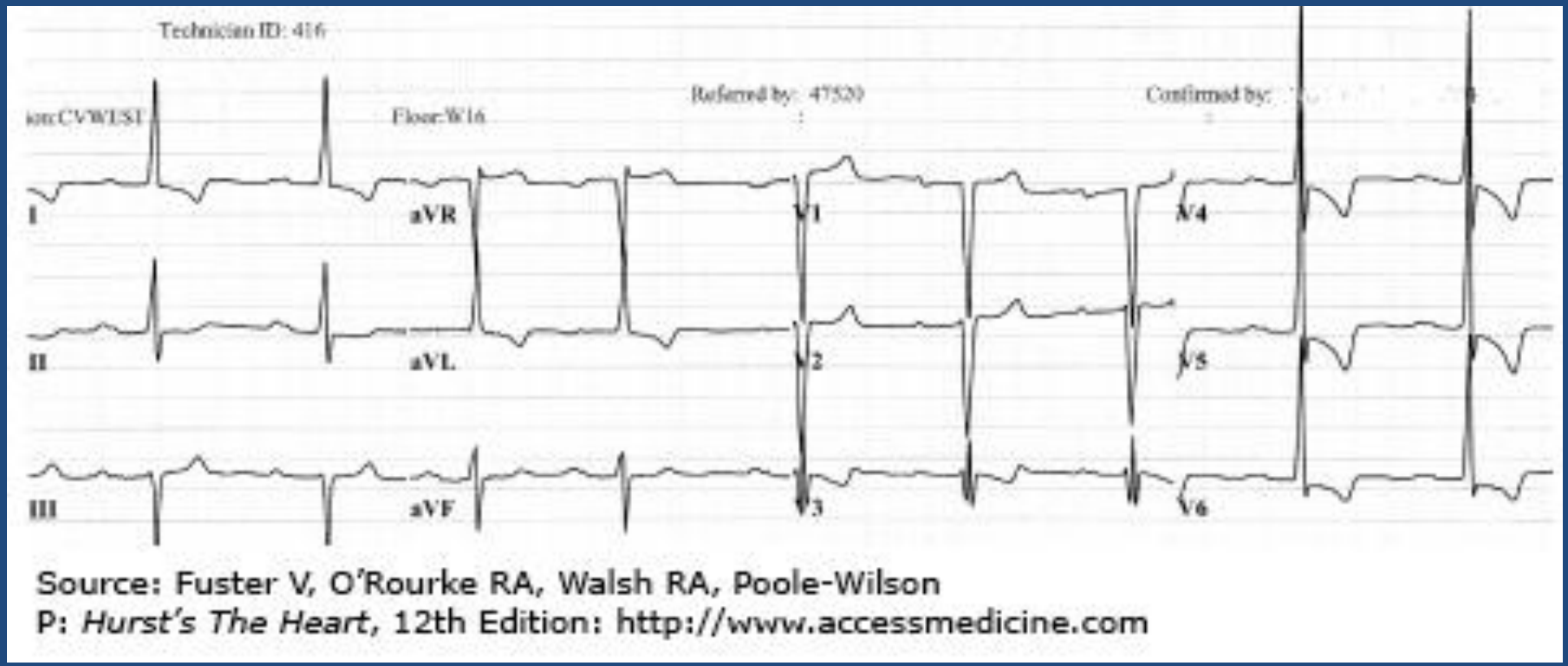
HCM - Physical Examination

- Pulse : jerky, bifid (short upstroke & prolonged systolic ejection)
- JVP : Prominent *a wave* - decreased ventricular compliance
- Double Apical Impulse
- Heart Sounds : S4 usually present due to hypertrophy
- Systolic Murmur : medium-pitch along LLSB, intensity increases with decreased preload (i.e. valsalva) and decreases with increased preload (i.e. hand grip, squatting)

HCM - ECG Findings

- Abnormal in 85-90% of cases
- LVH, Strain pattern
- Abnormal ST-T's, giant T wave inversions
- Abnormal Q's,
- Bundle Branch Block
- Left atrial enlargment
- Ventricular arrhthymias
- * think of HCM in any young person whose EKG shows large septal Q waves*

Electrocardiogram in HCM

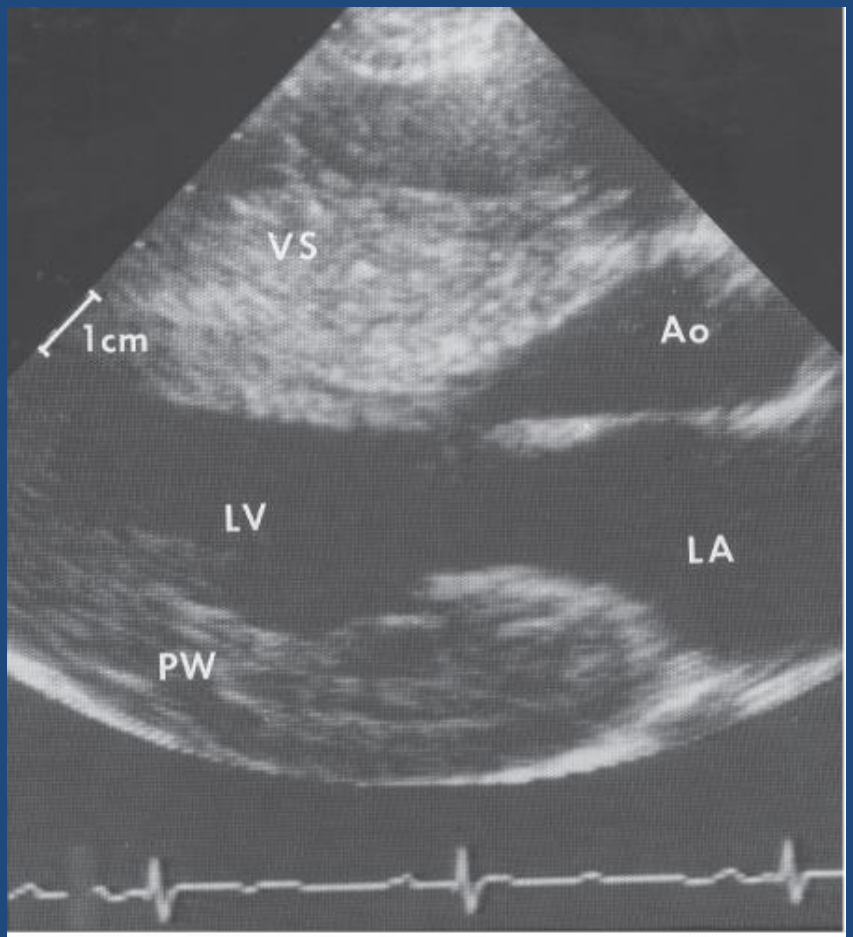


Source: Fuster V, O'Rourke RA, Walsh RA, Poole-Wilson P: *Hurst's The Heart*, 12th Edition: http://www.accessmedicine.com

HCM - Echocardiogram

- **LVH** especially septal >1.3 cm (usually >1.5 cm)
- Mitral regurgitation (**MR**)
- Systolic anterior motion of the mitral valve (**SAM**)
- Premature midsystolic closure of the aortic valve
- Asymmetric septal hypertrophy (**ASH**)
- **Diastolic dysfunction**
- LVOT obstruction (**LVOT gradient**)
- A normal ECHO in a young child does not R/O the diagnosis
- Serial ECHOs are recommended up to the age of 20 yr where there is a family history of HCM

Echocardiography in HCM



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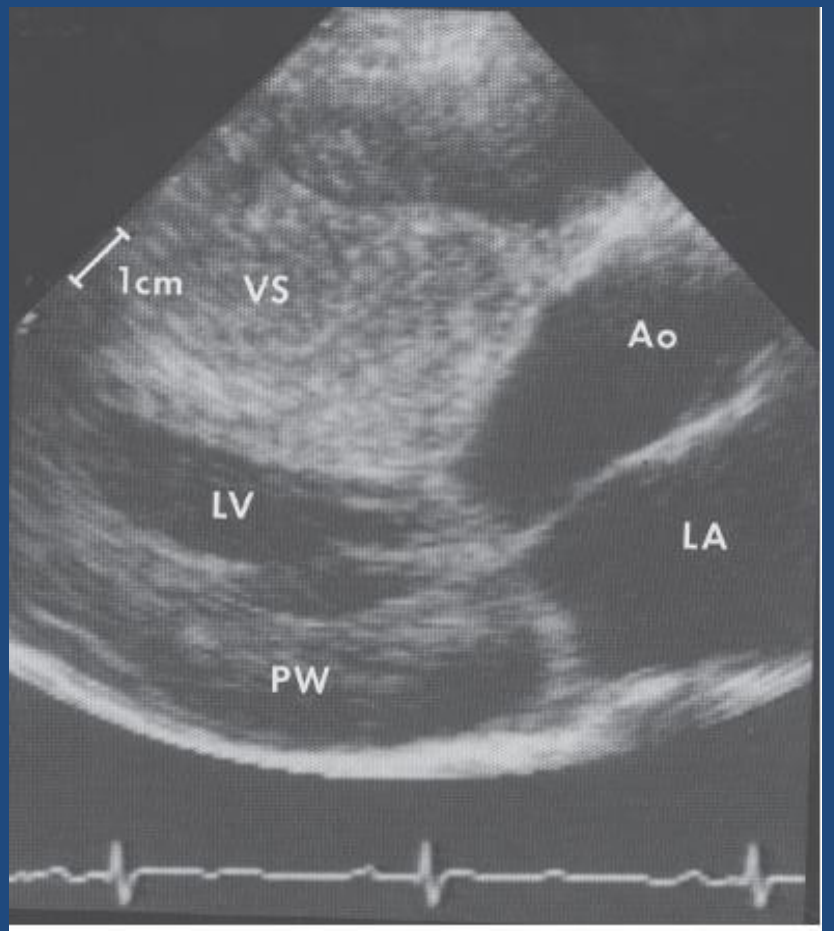

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HCM - Risk Factors For SCD

- Young age (<35 years)
- “Malignant” family history of sudden death
- Aborted sudden cardiac death
- Sustained VT or SVT
- Non-sustained VT on holter monitoring
- Atrial fibrillation
- Dilated left ventricle
- NYHA classes III or IV
- Syncope
- Severe hypertrophy (≥3.0 cm)
- Abnormal BP response to exercise
- Coronary artery disease
- Strenuous exercise or work

HCM - Treatment

- Medical therapy
- Device therapy
- Surgical septal myectomy
- Alcohol septal ablation

HCM - Treatment

- Avoid agents that reduce ventricular volume (nitrates) or increase myocardial contractility (digoxin)
- Antibiotic prophylaxis for dental procedures
- Avoidance of competitive athletics
- Anticoagulation if in afib

HCM - Medical Therapy

- **Beta-blockers**
  - Increase ventricular diastolic filling/relaxation
  - Decrease myocardial oxygen consumption
  - Have not been shown to reduce the incidence of SCD
- **Verapamil**
  - Augments ventricular diastolic filling/relaxation
  - Negative inotrope
- **Diuretics** (pulmonary congestion)
- **Amiodarone**

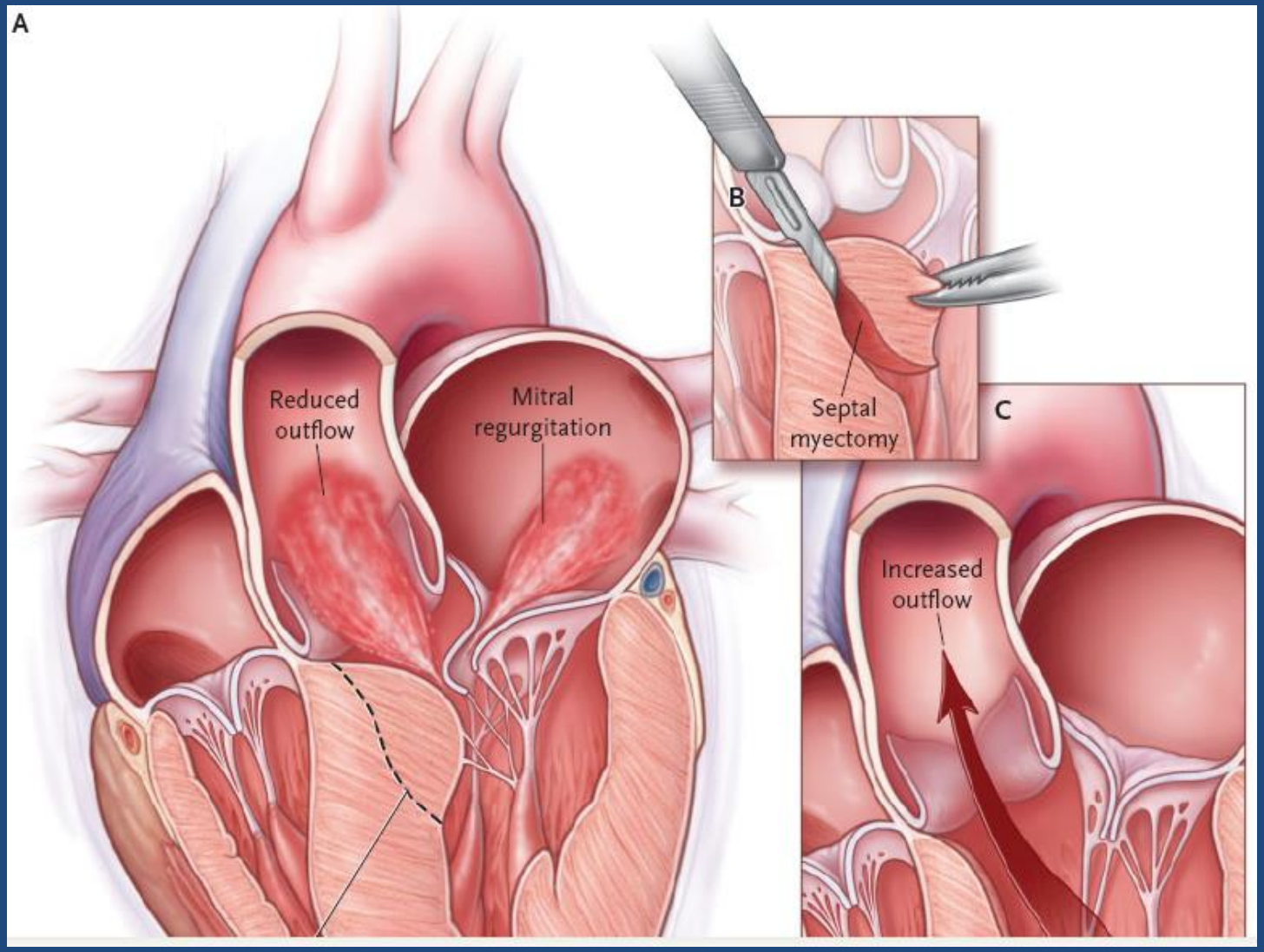
  treatment of choice for ventricular dysrhythmias

HCM – Device Therapy

- Dual-Chamber Pacing **:**
- Proposed benefit : pacing the RV apex will decrease the outflow tract gradient

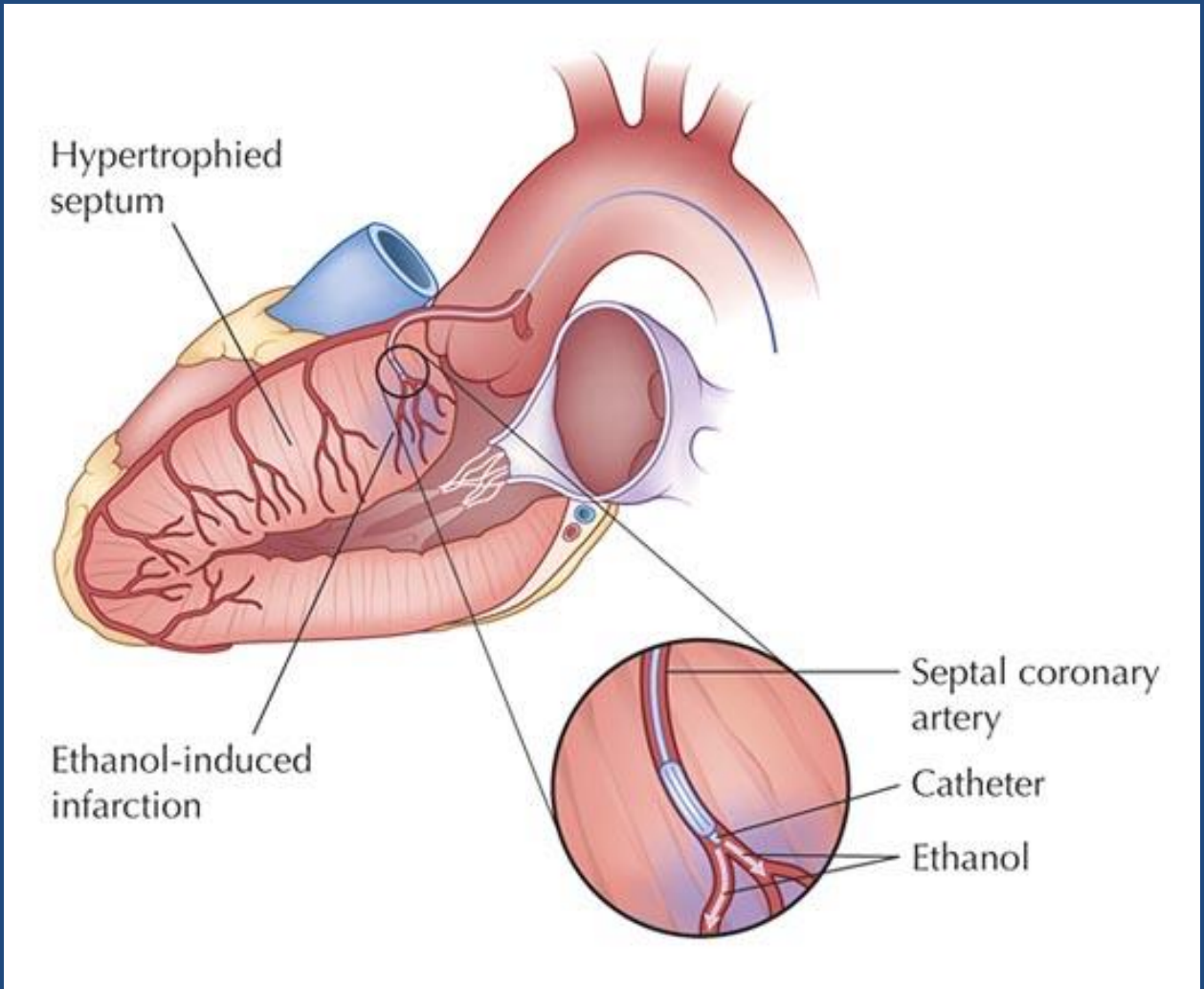
- **ICD** in cases of VT/VF

HCM - Surgical Septal Myectomy



Nishimura RA et al. NEJM. 2004. 350(13):1320.

HCM - Alcohol Septal Ablation



Braunwald. Atlas of Heart Diseases: Cardiomyopathies, Myocarditis, and Pericardial Disease. 1998.

HCM - Alcohol Septal Ablation

- Successful short-term outcomes
  - LVOT gradient reduced from a mean of 60-70 mmHg to <20 mmHg
  - Symptomatic improvements, increased exercise tolerance
- **Complications**
  - Complete heart block
  - Large myocardial infarctions

Restrictive Cardiomyopathy

RCM - Definition

- WHO defines RCM as a myocardial disease characterized by restrictive filling and reduced diastolic volume of either or both ventricles with normal or near-normal systolic function and wall thickness.
- The hallmark of the RCM is abnormal diastolic function; the ventricular walls are excessively rigid and impede ventricular filling.

RCM - Classification

- **Myocardial**
  - **Noninfiltrative**
    - Idiopathic
    - Scleroderma
  - **Infiltrative**
    - Amyloid
    - Sarcoid
    - Gaucher disease
    - Hurler disease
  - **Storage Disease**
    - Hemochromatosis
    - Fabry disease
    - Glycogen storage
- **Endomyocardial**
  - Endomyocardial fibrosis
  - Hyperesinophilic synd
  - Carcinoid
  - Metastatic malignancies
  - Radiation, anthracycline
- **Idiopathic**

RCM - Symptoms

- Progressive exertional dyspnea reflects the left heart failure.
- Fatigue and weakness are results of decreased stroke volume.
- distention of the abdomen & swollen feet (right heart failure).
- Angina like chest pain (observed only in patients with amyloidosis)
- Palpitations (atrial fibrillation)
- Thromboembolic complications.
- Syncopal attacks.
- Conduction disturbances particularly common in infiltrative RCM.
- Depending on the etiology, patients may have a prior history of radiation therapy, heart transplantation, chemotherapy, or a systemic disease.

RCM - Signs

- Low volume Pulse due to decreased stroke volume
- High JVP , but the most prominent wave is the Y descent (atrium emptying into the “stiff” ventricle)
- Elevation of JVP with inspiration (Kussmaul's sign).
- S4 in early disease (forceful atrial contraction against a stiff ventricle).
- S3 in advanced disease.
- Murmurs due to mitral and tricuspid valve regurgitation
- Pitting pedal Edema , palpable liver & ascites (in advanced cases)
- Reduced breath sounds, rales (LV failure)

RCM - Investigations

- **CXR**
  - Pulmonary venous congestion .The cardiac silhouette can be normal (familial) or show cardiomegaly and/or atrial enlargement.
- **ECG**
  - usually has low-voltage and ST segment and T wave abnormalities.
- **Echocardiogram**
  - symmetrical myocardial thickening and often a normal systolic ejection fraction, but impaired ventricular filling.

RCM - Investigations

- **Cardiac catheterization** **and** haemodynamic studies help distinction from constrictive pericarditis.
- **Endomyocardial biopsy** in contrast with other cardiomyopathies is often useful in this condition and may permit a specific diagnosis such as amyloidosis to be made.

RCM - Differential Diagnosis

Constrictive Pericarditis

- Pericardial calcification on x-ray, which occurs in constrictive pericarditis, is absent in RCM.
- Right ventricular transvenous endomyocardial biopsy (by revealing myocardial infiltration or fibrosis in RCM)
- CT scan or MRI (by demonstrating a thickened pericardium in constrictive pericarditis).

Clinical features of constrictive pericarditis and restrictive cardiomyopathy

Clinical Features	Constrictive Pericarditis	Restrictive Cardiomyopathy
History	Prior history of pericarditis or condition that causes pericardial disease	History of systemic disease (eg, amyloidosis, hemochromatosis)
General examination		Peripheral stigmata of systemic disease
Systemic examination - Heart sounds	Pericardial knock, high-frequency sound	Presence of loud diastolic filling sound S_4,S_3, Low-frequency sounds
Murmurs	No murmurs	Murmurs of mitral and tricuspid insufficiency
Prior chest x-ray	Pericardial calcification	Normal results of prior chest x-ray

RCM - Treatment & Prognosis

- The goal of treatment in RCM is to reduce symptoms by lowering elevated filling pressures without significantly reducing the cardiac output.
- No satisfactory specific medical therapy
- 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

RCM - Treatment & Prognosis

- Cardiac failure and embolic manifestations should be treated.
- Cardiac transplantation should be considered in some severe cases (especially the idiopathic variety).
- However, patients with cardiac amyloidosis have a worse prognosis than other forms, and the disease often recurs after transplantation
- Liver transplantation may be effective in familial amyloidosis and may lead to reversal of the cardiac abnormalities

Myocarditis

Myocarditis - Definition

- It is an inflammation of the myocardium, sometimes involves the pericardium.
- Often follows URI
- **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

Myocarditis - 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
- In some patients depressed ventricular function may develop in the absence of symptoms
- Manifests months to years later as DCM

Infectious	Noninfectious
Viruses 1. Coxsackie B 2. HIV 3. Influenza A&B	**Systemic Diseases:** 1. SLE & RA 2. Sarcoidosis 3. Vasculitides(Wegener's) 4. Celiac disease
Bacteria 1. Corynebacterium diphtheriae	**Neoplastic infiltration**
Protozoa 1. Trypanosoma cruzi (Chagas disease)	**Drugs & toxins:** 1. Ethanol 2. Cocaine 3. Radiation 4. Chemotherapeutic agents - Doxorubicin
Spirochete 1. Borrelia burgdorferi (Lyme disease)	

Natural History of Acute (Viral) Myocarditis

- **Subclinical**, no sequelae
- **Fulminant**; cardiac dilation, heart failure, arrhythmias, death
- Self limited cardiac dysfunction with resolution in weeks/months
- **Chronic**, cardiomyopathy

Myocarditis - Signs & Symptoms

- Days to weeks after onset of acute febrile illness present with heart failure without any known antecedent symptoms; highly variable
- Fever, tachycardia, myalgia, headache, rigors
- Chest pain due to coexisting pericarditis
- Onset of heart failure may be abrupt and fulminant or gradual.
- May mimic acute MI with ST elevation, positive cardiac markers, on echo regional wall motion abnormalities

Myocarditis - ECG & CXR

- **ECG** - nonspecific ST-T changes or elevation and conduction delays are common, Sinus Tach, low voltage , AV block .
  - Ventricular ectopy may be only clinical finding
- **CXR** – could be normal, cardiomegaly is frequent, may have evidence for pulmonary venous hypertension & pulmonary edema

Myocarditis - Diagnostics

- Wbc's often elevated
- ESR increased
- Troponins elevated in 1/3
- Blood culture, viral serology
- Echocardiogram helps evaluate cardiac function & exclude other causes
- Endomyocardial biopsy of the right ventricle remains the gold standard for diagnosis

Myocarditis

- Think of myocarditis in a young male with recent viral prodrome & now presents with unexplained CHF, MI with normal angiogram, new dysrhythmia/conduction defect, or symptoms of pericarditis with heart failure
- Spectrum of presentation of myocarditis
  - Excessive fatigue
  - Chest pain
  - Unexplained sinus tachycardia
  - Acute pericarditis
  - S3, S4, or summation gallop
  - Abnormal electrocardiogram
  - Abnormal echocardiogram
  - New cardiomegaly on chest x-ray
  - Atrial or ventricular arrhythmia
  - Partial or complete heart block
  - New onset congestive heart failure
  - Atypical myocardial infarction
  - Cardiogenic shock

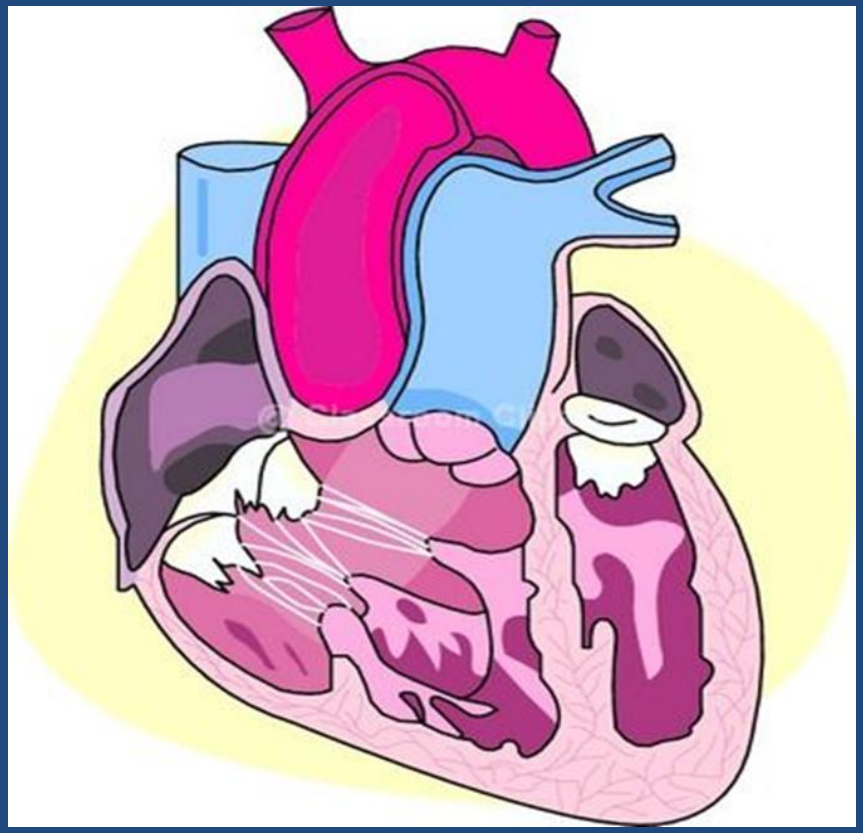
Myocarditis - Prognosis

- Fulminant myocarditis patients may present with cardiogenic shock (death or recovery)
- Subacute cases have dilated cardiomyopathy
- Chronic myocarditis patients may have mildly dilated LV and some of them have restrictive cardiomyopathy

Myocarditis - Treatment

- Antibiotics if specific agent is identified
- Standard HF therapy
- Arrhythmia suppression
- Limited exercise
- IVIG and steroids are controversial
- Fulminant myocarditis need aggressive short term support from IABP (intra-aortic balloon pumps) &/or LVAD (LV assistant device)

Questions?



“ Living isn’t just about doing for yourself, but what you do for others as well “

Nelson Mandela