

# MYOCARDIAL DISEASES

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# MYOCARDIAL DISEASE

## MYOCARDITIS

- Inflammatory process involving the myocardial wall, and may be caused virtually by any bacterial, viral, rickettsial, mycotic or parasitic organism.

### • INFECTIONS

#### (A). Viral

- Coxsackie-b, enterovirus, echovirus, adenovirus, cytomegalovirus, Herpes Simplex, H Zoster, Influenza, Mumps, Polio, Rubella, Viral Hepatitis

#### (B). Bacterial

- Diphtheria, Brucellosis, Actinomycosis, Scarlet fever, Rheumatic fever, Staphylococcus, Pneumococcal, Tuberculosis, Salmonella

# MYOCARDIAL DISEASE

## MYOCARDITIS

### (C.) SPIROCHAETAL

- Leptospirosis, Relapsing Fever, Syphilis, Q. Fever Typhus, Rocky mountain Spotted Fever.

### (D.) MYCOTIC (fungi)

- Aspergillosis, Blastomycosis, Candidiasis Histoplasmosis

### (E.) PROTOZOAL

- Amoebiasis, Chaga's Disease, Toxoplasmosis, Malaria, Leishmaniasis, Trypanosomiasis

### (F.) HELMINTHIC

- Ascariasis, Cysticercosis, Echinococcus, Filariasis, Trichinosis, schistosomiasis .

## NON INFECTIOUS

- Drugs
- Radiation



## CAUSES OF MYOCARDITIS

### Infectious

#### Viruses

Coxsackievirus, echovirus, HIV, Epstein-Barr virus, influenza, cytomegalovirus, adenovirus, hepatitis (A and B), mumps, poliovirus, rabies, respiratory syncytial virus, rubella, vaccinia, varicella zoster, arbovirus

#### Bacteria

*Corynebacterium diphtheriae*, *Streptococcus pyogenes*, *Staphylococcus aureus*, *Haemophilus pneumoniae*, *Salmonella* spp., *Neisseria gonorrhoeae*, *Leptospira*, *Borrelia burgdorferi*, *Treponema pallidum*, *Brucella*, *Mycobacterium tuberculosis*, *Actinomyces*, *Chlamydia* spp., *Coxiella burnetii*, *Mycoplasma pneumoniae*, *Rickettsia* spp.

#### Fungi

*Candida* spp., *Aspergillus* spp., *Histoplasma*, *Blastomyces*, *Cryptococcus*, *Coccidioidomyces*

#### Parasites

*Trypanosoma cruzii*, *Toxoplasma*, *Schistosoma*, *Trichina*

### Noninfectious

#### Drugs causing hypersensitivity reactions

Antibiotics: sulfonamides, penicillins, chloramphenicol, amphotericin B, tetracycline, streptomycin

Antituberculous: isoniazid, para-aminosalicylic acid

Anticonvulsants: phenindione, phenytoin, carbamazepine

Anti-inflammatories: indomethacin, phenylbutazone

Diuretics: acetazolamide, chlorthalidone, hydrochlorothiazide, spironolactone

Others: amitriptyline, methyldopa, sulfonylureas

#### Drugs not causing hypersensitivity reactions

Cocaine, cyclophosphamide, lithium, interferon alpha

#### Nondrug causes

Radiation, giant-cell myo

# MYOCARDIAL DISEASE

## MYOCARDITIS

### NATURAL HISTORY & PROGNOSIS

#### GENERAL CONSIDERATION

- True frequency of Myocarditis is uncertain as most patients with mild disease recover spontaneously
- Few patients blend into idiopathic dilated cardiomyopathy
- Patients with large heart & cardiac failure have poor prognosis
- Myocarditis during infancy, child-hood & pregnancy is often fulminant & fatal.

#### CHRONIC MYOCARDITIS

- Is difficult to diagnose clinically,
- Abnormalities usually nonspecific



## NATURAL HISTORY & PROGNOSIS

### GENERAL CONSIDERATION

- Prognosis generally favourable & most patients recover rapidly
- Significant number have recurrent chronic myocarditis
- Some patients succumb to a fulminant acute illness, is not unusual cause of sudden death in young adults
- If Heart failure, dysrhythmias and conduction disturbances prognosis is guarded.

# TREATMENT

- Admission to hospital if acute Myocarditis is suspected
- Patients with pericardial effusion, dysrhythmias, cardiac failure, evidence of myocardial ischemia, hypotension & shock need intensive care management
- Specific therapy for underlying infection
- General measures to decrease cardiac work.
- control of complications:
  - CARDIAC FAILURE
  - DYSRRHYTHMIAS
  - SHOCK
- > Admit in C.C.U.
- > Steroids beneficial in later stage
- > Bed rest
- > General analgesics & antipyretics
- > Prophylactic heparin to prevent thrombo-embolic complications



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In the Name of **Allah** the Most Beneficent and Merciful

# **Cardiomyopathies**

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# Cardiomyopathies

- **Primary**

“Heart muscle disease of unknown cause”

- **Secondary**

Myocardial involvement in systemic disease “ (rare specific heart muscle disease)”



# Primary Cardiomyopathies

1. Dilated (congestive)

2. Hypertrophic  
(I.H.S.S)

H.O.C.M.

H.N.O.C.M.

3. Restrictive / obliterative

SYSTOLE

NORMAL

CONGESTIVE

RESTRICTIVE

HYPERTROPHIC



DIASTOLE





## II. New WHO classification<sup>dis</sup> (1995)

### A. Functional classification of cardiomyopathy

1. Dilated cardiomyopathy
2. Hypertrophic cardiomyopathy
3. Restrictive cardiomyopathy
4. Arrhythmogenic right ventricular cardiomyopathy
5. Unclassified cardiomyopathies

### B. Specific cardiomyopathies

1. Ischemic cardiomyopathy
2. Valvular cardiomyopathy
3. Hypertensive cardiomyopathy
4. Inflammatory cardiomyopathy
  - a. Idiopathic
  - b. Autoimmune
  - c. Infectious

Cont.

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Cont.



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**5. Metabolic cardiomyopathy**

- a. Endocrine
- b. Familial storage diseases and infiltrations
- c. Deficiency
- d. Amyloid

**6. General system disease**

- a. Connective tissue disorders
- b. Infiltrations and granulomas

**7. Muscular dystrophies**

**8. Neuromuscular disorders**

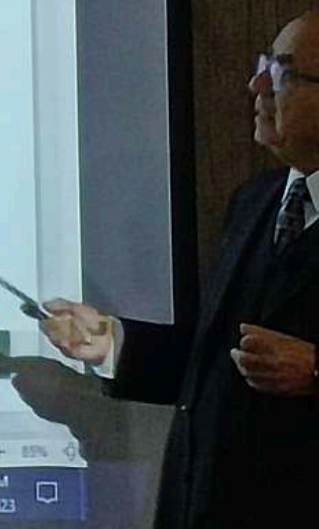
**9. Sensitivity and toxic reactions**

**10. Peripartal cardiomyopathy**

[Cont](#)

Notes 85%

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## Functional Classification of Cardiomyopathies

### I. Cardiac dilatation

#### A. With systolic failure

1. Idiopathic dilated cardiomyopathy
2. Late cardiac amyloidosis
3. Tachycardia-induced congestive failure

#### B. Without systolic failure

1. High cardiac output state
2. Bradycardia-induced congestive failure

### II. Cardiac hypertrophy

#### A. With obstruction

1. Hypertrophic obstructive cardiomyopathy

#### B. Without obstruction

1. Hypertrophic cardiomyopathy
2. Left ventricular hypertrophy due to systemic hypertension

### III. Cardiac restriction

- #### A. Early cardiac amyloidosis
- #### B. Myocardial fibrosis



## Etiological Classification of Cardiomyopathies

### I. Infective/inflammatory

- Idiopathic lymphocytic myocarditis
- Peripartum myocarditis
- Eosinophilic myocarditis
- Giant-cell myocarditis
- Viral myocarditis
- Rickettsial myocarditis
- Bacterial myocarditis
- Mycobacterial heart disease
- Spirochetal heart disease
- Fungal myocarditis
- Protozoal myocarditis
- Metazoal myocarditis
- Helminthic myocarditis
- Chemical or drug hypersensitivity
- Autoimmune myocarditis

### II. Metabolic

#### A. Endocrine

1. Thyroid disease
  - Thyrotoxicosis
  - Hypothyroidism
2. Pheochromocytoma
3. Acromegaly
4. Diabetes mellitus
5. Carcinoid heart disease

#### B. Uremia

#### C. Hyperoxaluria

#### D. Gout

#### E. Storage diseases and infiltrative processes

## Etiological Classification of Cardiomyopathies

1. Lysosomal storage diseases
  - GM1 gangliosidosis
  - Tay-Sachs disease and variants
  - Sandhoff's disease
  - Niemann-Pick disease
  - Gaucher's disease
  - Fabry's disease
  - Farber's disease
  - Fucosidosis
  - Hurler's syndrome
  - Scheie's syndrome
  - Hunter's syndrome
  - Sanfilippo
  - Morquio
  - Moroteaux-Lamy
2. Glycogen storage diseases
  - Pompe's disease
  - Cori's disease
  - Andersen's disease
  - Dominantly inherited cardioskeletal myopathy with lysosomal glycogen storage and normal acid maltase levels
3. Refsum's syndrome
4. Hand-Schüller-Christian
5. Adipositas cordis
6. Hemochromatosis
7. Deficiency of carnitine
  - 1. E...



## Etiological Classification of Cardiomyopathies

### 2. Nutritional

- Kwashiorkor
- Beriberi
- Pellagra
- Scurvy
- Selenium
- Carnitine

### III. Amyloid

- AL (primary amyloid, myeloma-associated amyloid)
- AA (secondary amyloid, familial Mediterranean fever-associated amyloid)
- AF (familial amyloid)
- SSA (senile cardiac amyloid, senile systemic amyloid)
- IAA (atrial amyloid)

### IV. General system disorders

- A. Collagen vascular (connective tissue)
  - Systemic lupus erythematosus
  - Polyarteritis nodosa
  - Rheumatoid arthritis
  - Scleroderma
  - Dermatomyositis
  - Whipple's disease
  - Kawasaki's disease

- B. Sarcoid

- C. Neoplasms

### V. Muscular dystrophies, and neuromuscular disorders

Cont.

## Etiological Classification of Cardiomyopathies

- A. Muscular dystrophies
  - Duchenne's muscular dystrophy
  - Becker's muscular dystrophy
  - Myotonic dystrophy
  - Facioscapulohumeral muscular dystrophy
  - Limb girdle dystrophy
  - Scapulohumeral dystrophy, including Emery-Dreifuss
  - Congenital muscular dystrophy
  - Distal muscular dystrophy
- B. Congenital myopathies
  - Central-core disease
  - Nemaline myopathy
  - Myotubular myopathy (centronuclear)
  - Congenital fiber-type disproportion
- C. Mitochondrial myopathies, including Kearns-Sayre syndrome
- D. Neuromuscular disorders, Friedrich's ataxia
- VI. Toxicity, hypersensitivity, and physical agent effects
  - A. Toxic effects
    - 1. Caused by drugs, heavy metals, and chemical agents
      - Alcohol (ethyl)
      - Amphetamine/methamphetamine
      - Anthracyclines
      - Anticancer agents

Cont.



## Etiological Classification of Cardiomyopathies

- Carbon monoxide
- Catecholamines
- Chloroquine
- Cobalt
- Cocaine
- Cyclophosphamide
- Emetine
- 5-Fluorouracil
- Hydrocarbons
- Interferon
- Lead
- Lithium
- Mercury
- Methysergide
- Paracetamol
- Phenothiazines
- Phosphorus
- Reserpine

2. Caused by scorpions, spiders, arthropods, and snakes

- Scorpions
- Arthropods
- Black widow spider
- Snakes

B. Hypersensitivity reactions

- Acetaminophen

- Amphotericin B

- Amphotericin B

- Amphotericin B

[Cont.](#)

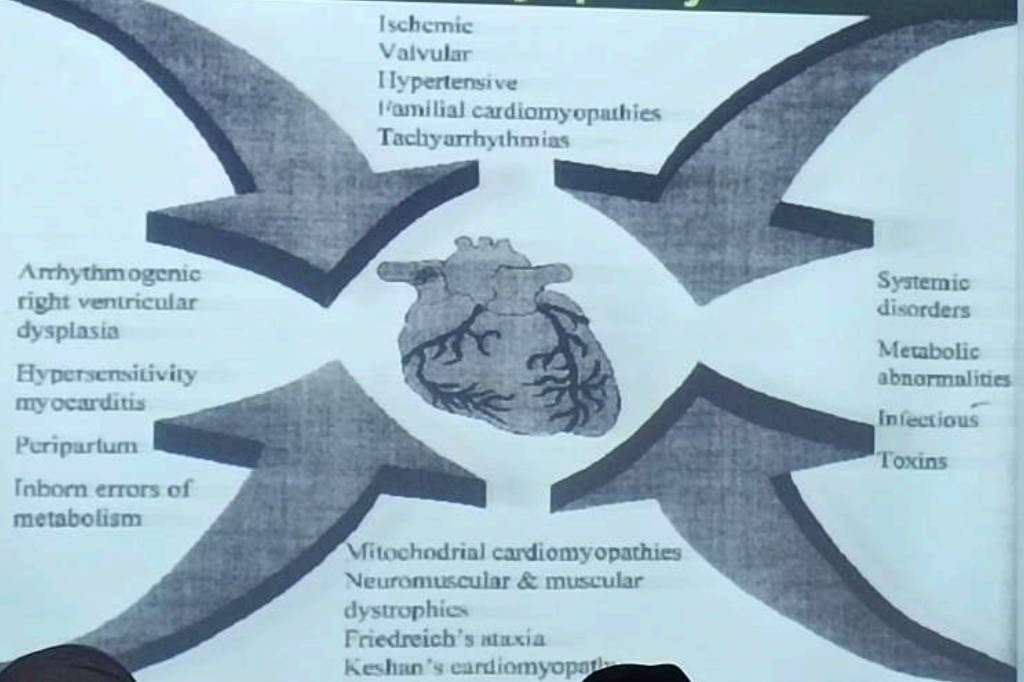
## Etiological Classification of Cardiomyopathies

Chlorthalidone  
Hydrochlorothiazide  
Indomethacin  
Isoniazid  
Methyldopa  
Oxyphenbutazone  
Para-aminosalicylic acid  
Penicillin  
Phenindione  
Phenylbutazone  
Phenytoin  
Streptomycin  
Sulfadiazine  
Sulfisoxazole  
Sulfonylureas  
Tetracycline  
C. Physical agents  
Heat  
Hypothermia  
Radiation  
VII. Miscellaneous  
Peripartum heart disease  
Tachycardia-induced cardiomyopathy  
Ectodermal dysplasia-associated cardio-  
myopathy  
Idiopathic endocardial fibrosis  
Endocardial fibroelastosis  
Infantile cardiomyopathy  
Arrhythmogenic right ventricular  
dysplasia

Cont.



# Various Etiologies That Can Lead to Cardiomyopathy



## Characteristics Of The Three Main Types Of Cardiomyopathy

Charateristics	Dilated	Hypertrophic	Restrictive Obliterative
Myocardial Mass	■■■■■	■■■■	nl ■■■
Ventricular Cavity Size	■■■■■■■	■■■■■ nl	■
Dilated Atrial Cavity	+	+	+
Endocardial Thickening	0 ■■ +	◆+	++++
Myocardial Infiltration	0	0	0 ■■ + ■
Asymmetric Septal Hypertrophy	0	+	0
Myocardial Fiber Disorientation	0	+	0
Abnormal Intramural Coronary Arteries	0	+	0
Endocardial Plaque, LV Outflow Tract	0	+	0
Contractile Function	↓↓↓	■■■■ ↓	nl ■■ ↓
Ventricular Inflow Resistance	0	++	+
Ventricular Outflow "Obstruction"	0	0 → +	0
LV Filling Pressure	■■■	nl	■
Mitral Regurgitation	+	+	+
Thickened Mitral Valve	0	+	±
Interventricular thrombi	+	0	1



## Dilated Cardiomyopathy (D.C)

- Cardiomegaly with dilatation of both ventricles.
- Impairment of systolic function.
- Increased myocardial mass.(dilatation more impressive than the hypertrophy)

### Factors

- Alcohol, hypertension, pregnancy, immunological disorders, viral infections, chemical agents.

## Three General Mechanism by Which Alterations in Gene Expression Can Influence the Development or Progression of a Dilated Cardiomyopathy

### Types of Process

### Examples

Gene mutation

Cardiac  $\alpha$ -actin,<sup>34</sup> desmin,<sup>35</sup> dystrophin,<sup>36,37</sup> lamin<sup>38,39</sup>

Polymorphic variation in modifier genes

Angiotensin converting enzyme (ACE),<sup>16,41,44</sup>  $\beta_2$ -adrenergic receptor<sup>46</sup>

Altered expression of a completely normal, wild type gene

Decreased expression:  $\beta_1$ -adrenergic receptors,<sup>8</sup>  $\alpha$ -MHC,<sup>47,48</sup> SERCA-2<sup>49</sup>

Increased expression: ANP,<sup>50</sup>  $\beta$ -MHC,<sup>47</sup> ACE,<sup>51,52</sup> TNF- $\alpha$ ,<sup>53</sup> endothelin,<sup>54</sup>  $\beta$ ARK<sup>55</sup>

ABBREVIATIONS: MHC = myosin heavy chain; TNF = tumor necrosis factor;  $\beta$ ARK =  $\beta$ -adrenergic receptor kinase; SERCA = sarcoplasmic reticulum calcium ATPase; ANP = atrial natriuretic



## Types of Dilated Cardiomyopathies

Ischemic insult (ischemic cardiomyopathy)

Valvular disease (mitral regurgitation, aortic regurgitation, aortic stenosis) (valvular cardiomyopathy)

Chronic hypertension (hypertensive cardiomyopathy)

Tachyarrhythmias (supraventricular, ventricular, atrial flutter)

Familial (autosomal dominant, X-linked)

Idiopathic

Toxins

Ethanol

Chemotherapeutic agents (anthracyclines such as doxorubicin and daunorubicin)

Cobalt

Antiretroviral agents (zidovudine, didanosine, zalcitabine)

Phenothiazines

Carbon monoxide

Lithium

Lead

Cocaine

Mercury

Metabolic abnormalities

Nutritional deficiencies (thiamine, selenium, carnitine, protein)

Endocrinologic disorders (hypothyroidism, acromegaly, thyrotoxicosis, Cushing's disease, pheochromocytoma, catecholamines, diabetes mellitus)

Electrolyte disturbances (hypocalcemia, hypophosphatemia)

Exsackie virus, cytomegalovirus, HIV)

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# Dilated Cardiomyopathy

## Epidemiology:

- World wide
- All ages and races
- More common in men than in women.
- ?genetic predisposition.

## Pathology

- Dilatation of both ventricles (Lt . more than Rt.)
- Poor vent .contraction and reduced ejection fraction.
- Relative stasis of blood and clot formation.
- Focal endocardial thickening
- Leaflets of cardiac valves usually normal (occasionally margins show focal thickening) .
- Valve ring dilation.

# Dilated Cardiomyopathy

## Clinical Manifestations

### History

- Insidious onset of left vent. Failure
- Followed by symptoms of Rt. Sided congestive failure
- Chest pain may occur specially with exertion



# Presentation

- Progressive congestive cardiac failure.
- Arrhythmias
- Atypical chest pain

## Physical examination

- Cardiomegaly
- Gallop rhythm
- C.C.F.
- Varying degree of mitral regurgitation.
- Atrial fibrillation 10-20%.

# Physical Examination

- Signs of C.C.F.
- Skin cold, pale, cyanosed
- Peripheral veins constricted
- Arterial pulse of small volume
- Pulsus alternans
- Tachycardia at rest
- J.V.P. is raised
- Tricuspid regurgitation
- Apex beat displaced out-side mid.C.line
- Rt.V & L.V. pulsation due to dilatations, systolic murmurs of mitral & tricuspid regurgitations
- Gallop rhythm –III H.S.& 4<sup>th</sup> H.S. P<sub>2</sub> loud.
- Reduced pulse pressure
- Pericardial effusion may be present
- Basal crepts in lungs with pleural effusion
- Hepatomegoly & liver may be pulsatile
- Periphral oedema & ascites



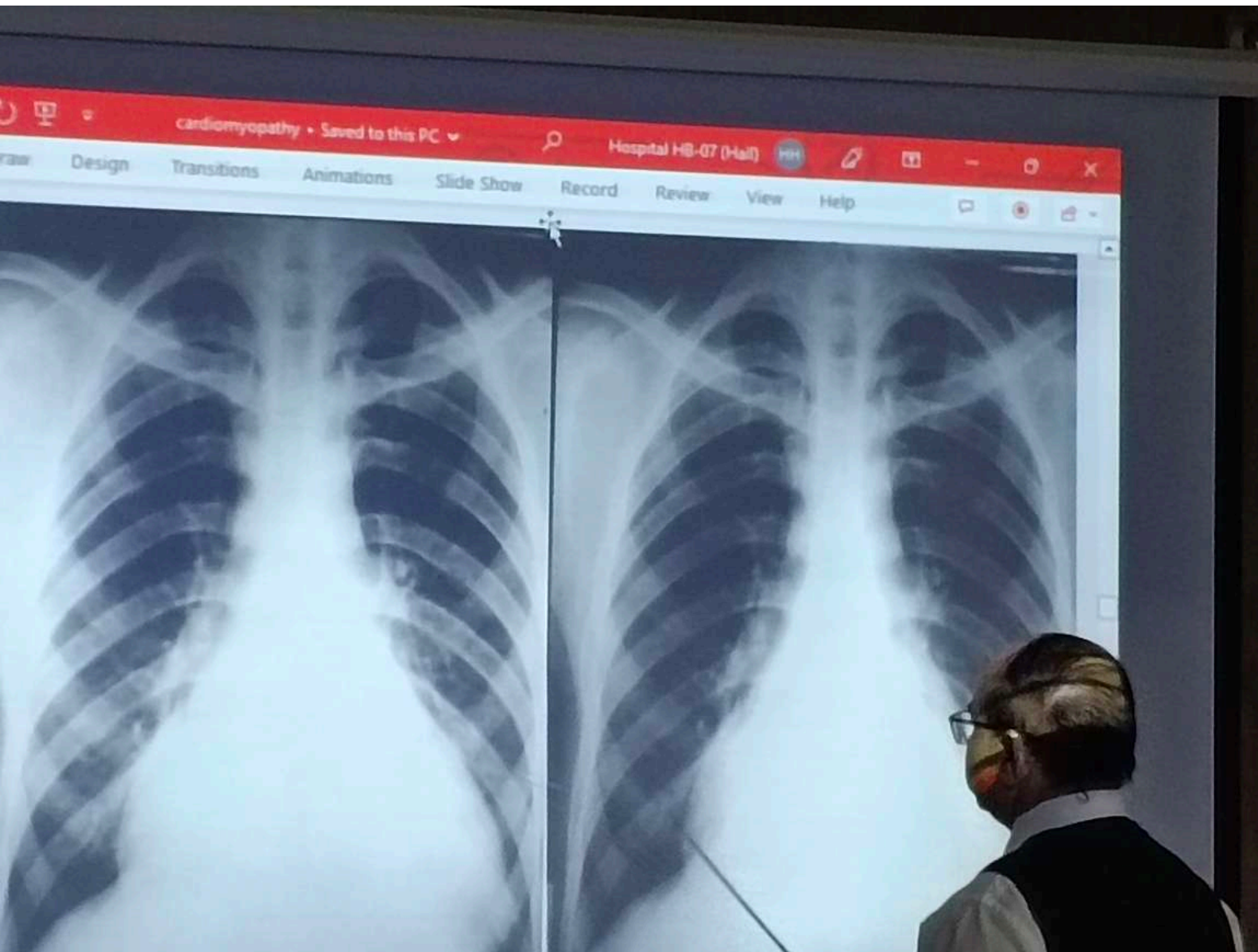
# Investigations

## 1. X-ray chest

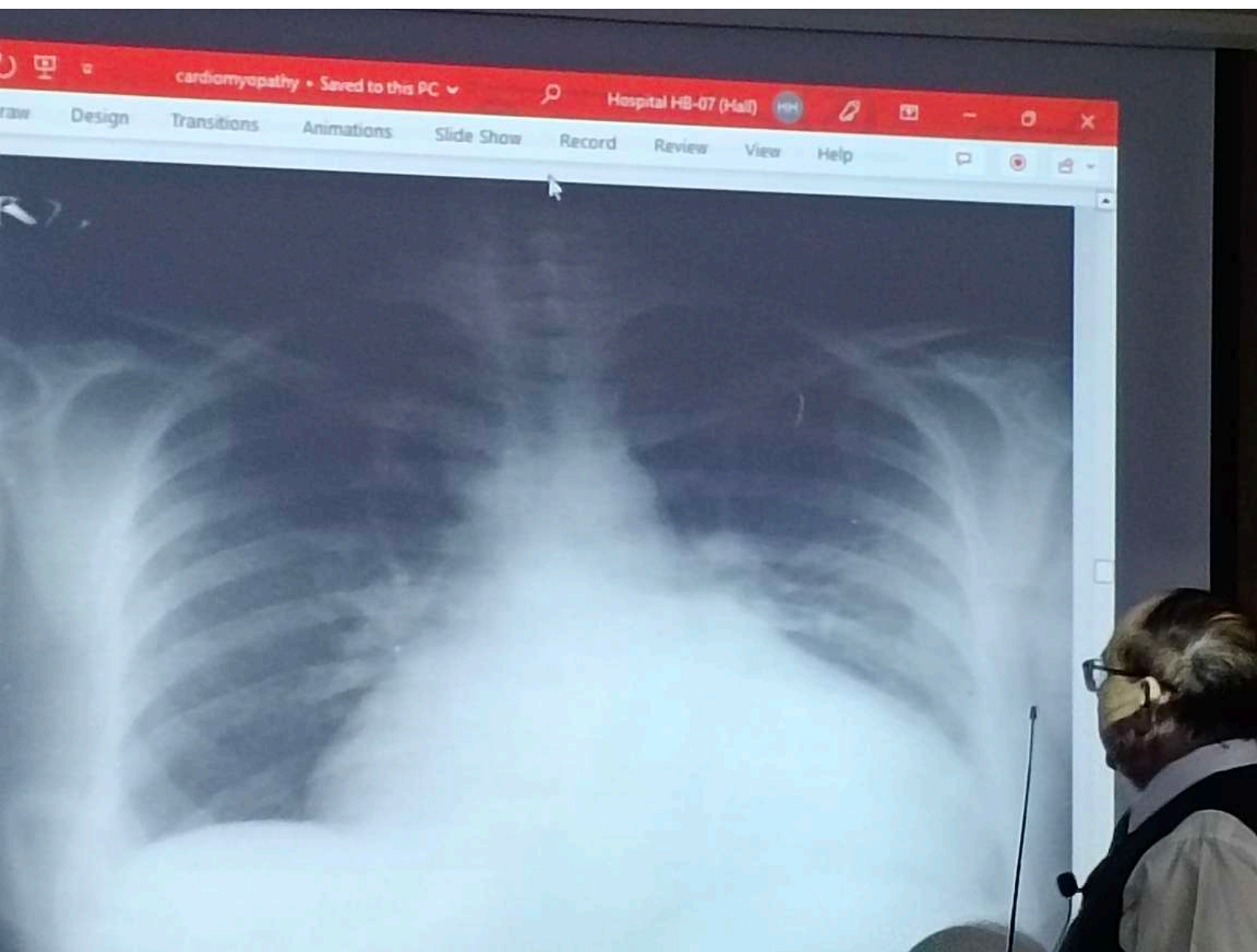
- Dilated and enlarged heart
- Pul. Venous hypertension
- Enlarged main Pul. Arteries
- Pleural effusion
- When pericardial effusion → water bottle shape heart

## 2. ECG

- Sinus tachycardia
- Non specific changes flat & inverted T waves
- Atrial fibrillation common
- QRS low voltage & wide
- L.V.H. +
- Conductive disturbances







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I

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# Investigations

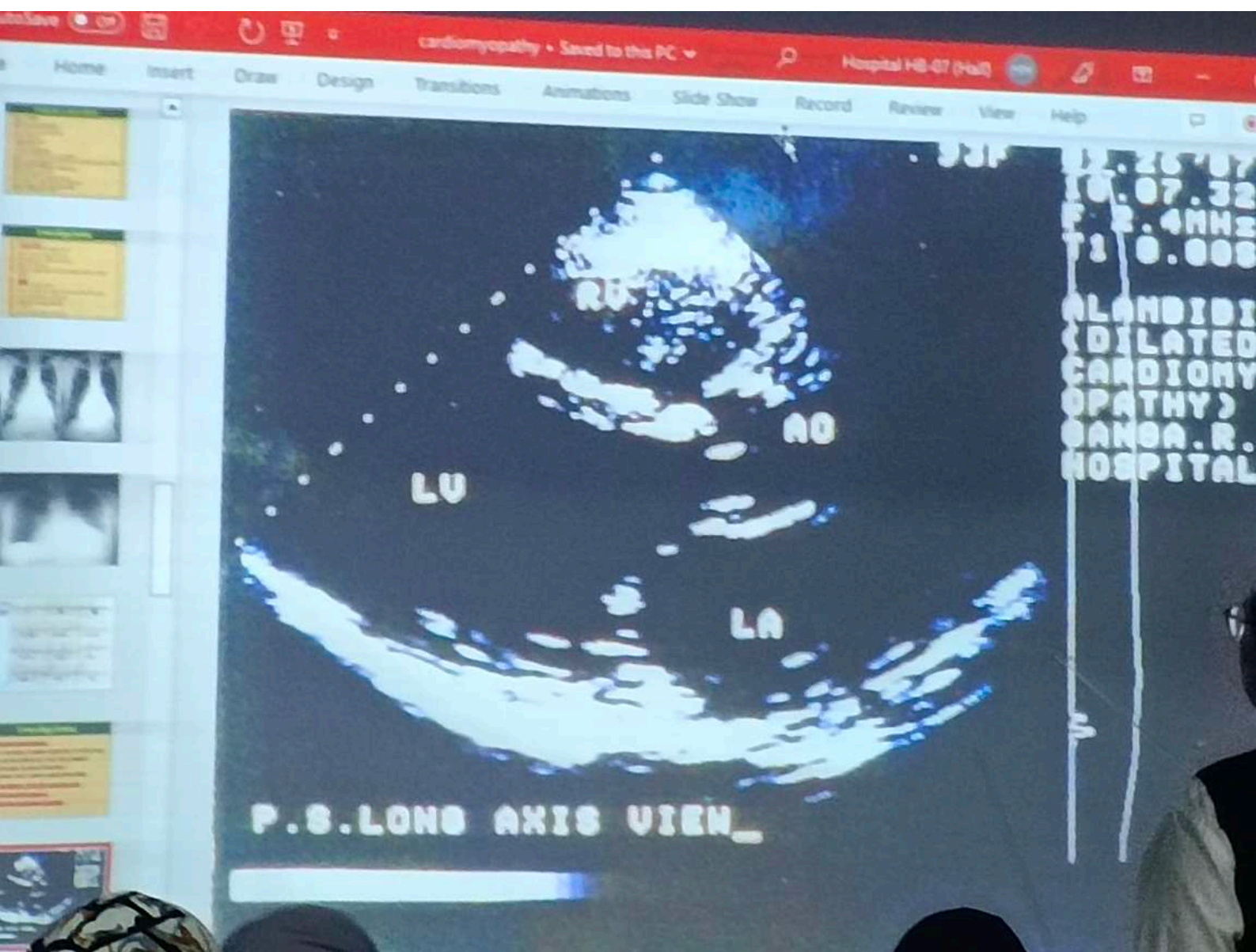
## 3. Echocardiography

- All chambers dilated & poorly contracting
- Pericardial effusion may be present
- Ventricular & atrial thrombi
- Regional wall motion abnormalities

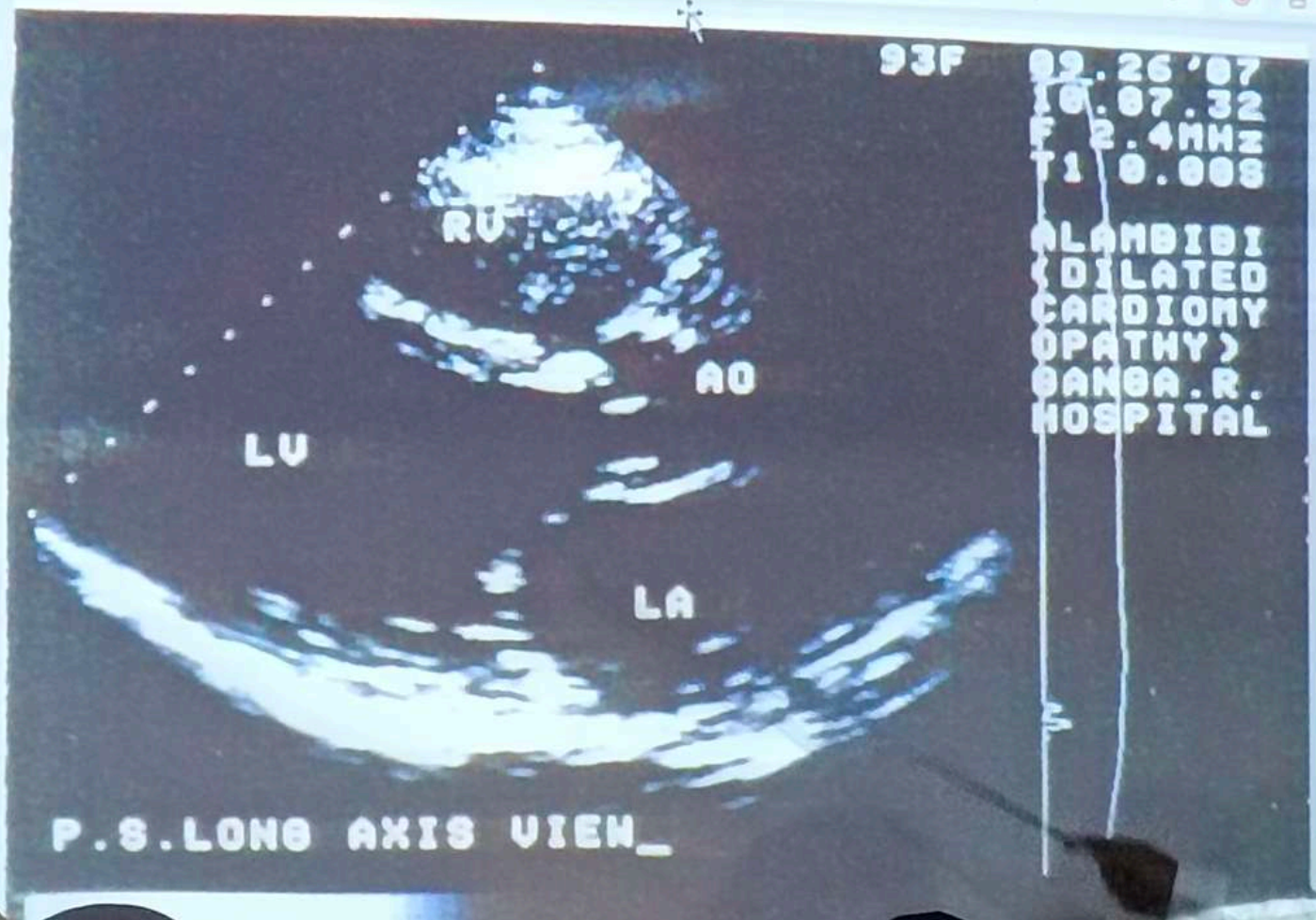
## 4. Ambulatory E.C.G monitoring

## 5. Angiocardiology

## 6. Endo-myocardial biopsy







## Natural History & Prognosis

- Course is usually steadily downhill
- Death-commonly within 6 months to several years
- Persistent cardiomegaly, L.B.B.B.and dysrrhythmia have poor prognosis
- Mean survival time 3 years from onset of symptoms.



## D.Diagnosis

1. I.H.D.
2. Hypertensive heart disease
3. Specific heart muscle disease
4. Hypertrophic cardiomyopathy

## Treatment

- Treatment of heart failure
- Vasodilators
- Salbutamol
- Positive inotropic agents
- Selective B blockers small dose
- Anticoagulants

# Hypertrophic Cardiomyopathy

- H.O.C.M.
- H.N.O.C.M., A.S.H.
- I.H.S.S.

**Etiology**

**Inheritance H.L.A. typing**

**Inherited abnormality of handling catecholamines by myocardium**



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## The Multitude of Terms Used to Describe HCM

Asymmetrical hypertrophic cardiomyopathy  
 Asymmetrical hypertrophy of the heart  
 Asymmetrical septal hypertrophy  
 Brock's disease  
 Diffuse muscular subaortic stenosis  
 Diffuse subvalvular aortic stenosis  
 Dynamic hypertrophic subaortic stenosis  
 Dynamic muscular subaortic stenosis  
 Familial hypertrophic cardiomyopathy  
 Familial hypertrophic subaortic stenosis  
 Familial muscular subaortic stenosis  
 Familial myocardial disease  
 Functional aortic stenosis  
 Functional hypertrophic subaortic stenosis  
 Functional obstructive cardiomyopathy  
 Functional obstruction of the left ventricle  
 Functional obstructive subvalvular aortic stenosis  
 Functional subaortic stenosis  
 Hereditary cardiovascular dysplasia  
**HYPER TROPHIC CARDIOMYOPATHY**  
 Hypertrophic constrictive cardiomyopathy  
 Hypertrophic hypertensive cardiomyopathy  
 Hypertrophic infundibular aortic stenosis  
 Hypertrophic nonobstructive cardiomyopathy  
 Hypertrophic obstructive cardiomyopathy  
 Hypertrophic stenosing cardiomyopathy  
 Hypertrophic subaortic stenosis  
 Idiopathic hypertrophic cardiomyopathy  
 Idiopathic hypertrophic obstructive cardiomyopathy  
 Idiopathic hypertrophic subaortic stenosis

Idiopathic hypertrophic subvalvular stenosis  
 Idiopathic muscular hypertrophic subaortic stenosis  
 Idiopathic muscular stenosis of the left ventricle  
 Idiopathic myocardial hypertrophy  
 Idiopathic stenosis of the flushing chamber of the left ventricle  
 Idiopathic ventricular septal hypertrophy  
 Irregular hypertrophic cardiomyopathy  
 Left ventricular muscular stenosis  
 Low subvalvular aortic stenosis  
 Muscular aortic stenosis  
 Muscular hypertrophic stenosis of the left ventricle  
 Muscular stenosis of the left ventricle  
 Muscular subaortic stenosis  
 Muscular subvalvular aortic stenosis  
 Non-dilated cardiomyopathy  
 Nonobstructive hypertrophic cardiomyopathy  
 Obstructive cardiomyopathy  
 Obstructive hypertrophic aortic stenosis  
 Obstructive hypertrophic cardiomyopathy  
 Obstructive hypertrophic myocardiopathy  
 Obstructive myocardiopathy  
 Pseudoaortic stenosis  
 Stenosing hypertrophy of the left ventricle  
 Stenosis of the ejection chamber of the left ventricle  
 Subaortic hypertrophic stenosis  
 Subaortic idiopathic stenosis  
 Subaortic muscular stenosis  
 Subvalvular aortic stenosis of the muscular type  
 Teare's disease

# Clinical Manifestations

## History

1. Dyspnoea
2. Chest pain
3. Palpitations
4. Dizziness
5. Syncope

## Physical signs

- A late onset systolic murmur
- Arterial pulse is abrupt, ill-sustained & jerky
- Cardiac apical impulse, powerful but ill-sustained



## Possible Mechanism for Ischemia in Hypertrophic Cardiomyopathy

### Increased myocardial oxygen demand

Myocardial hypertrophy  
Diastolic dysfunction

Myocyte disarray

Left ventricular outflow obstruction

arrhythmias

### Reduced myocardial perfusion

Small vessel disease

Abnormal vascular responses

Myocardial bridges

Increased coronary vascular resistance

# Investigations

## A. X-ray chest

### A. E.C.G

- L.V.H.
- Lt. Ant. Hemiblock
- Q waves in inferior and Lt. Precordial leads
- R.V.H. may occur
- Giant inverted T waves and high precordial QRS

### C. ECHOCARDIOGRAPHY

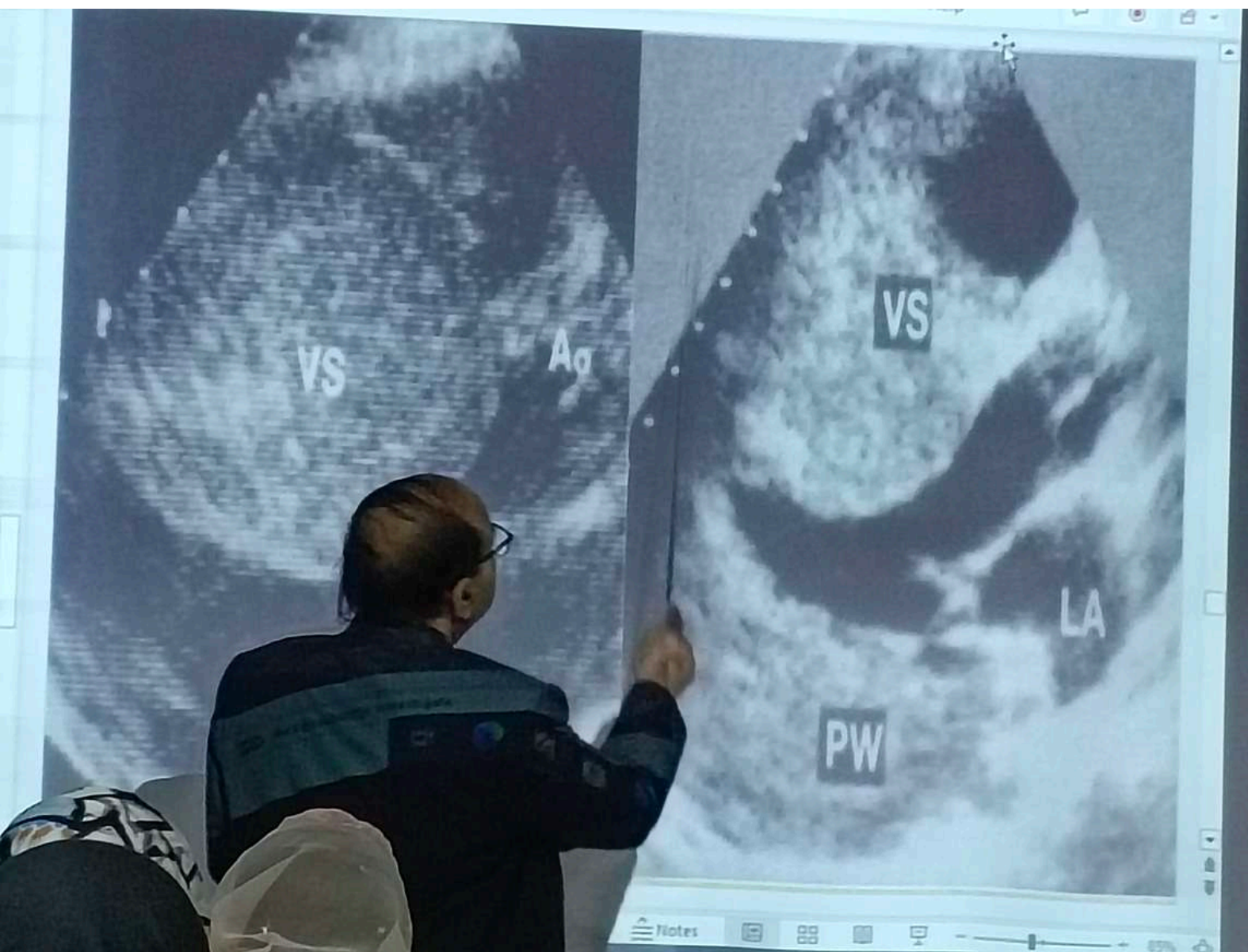
- A.S.H.
- Poor septal contraction
- Reduced systolic dimension of L.V. cavity
- S.A.M.

### D. Radionuclide imaging

### E. Cardiac catheterization & angiography

### F. Endomyocardial biopsy





# Natural History

- Extremely variable
- Life span-few years to many years
- Most patients-stable course about 10 years
- Half of patients die suddenly due to haemodynamic (mechanical) and arrhythmias
- C.C.F.- related to atrial fibrillation and extensive myocardial necrosis
- Systemic embolism
- Mitral regurgitation
- Infective endocarditis



# Treatment

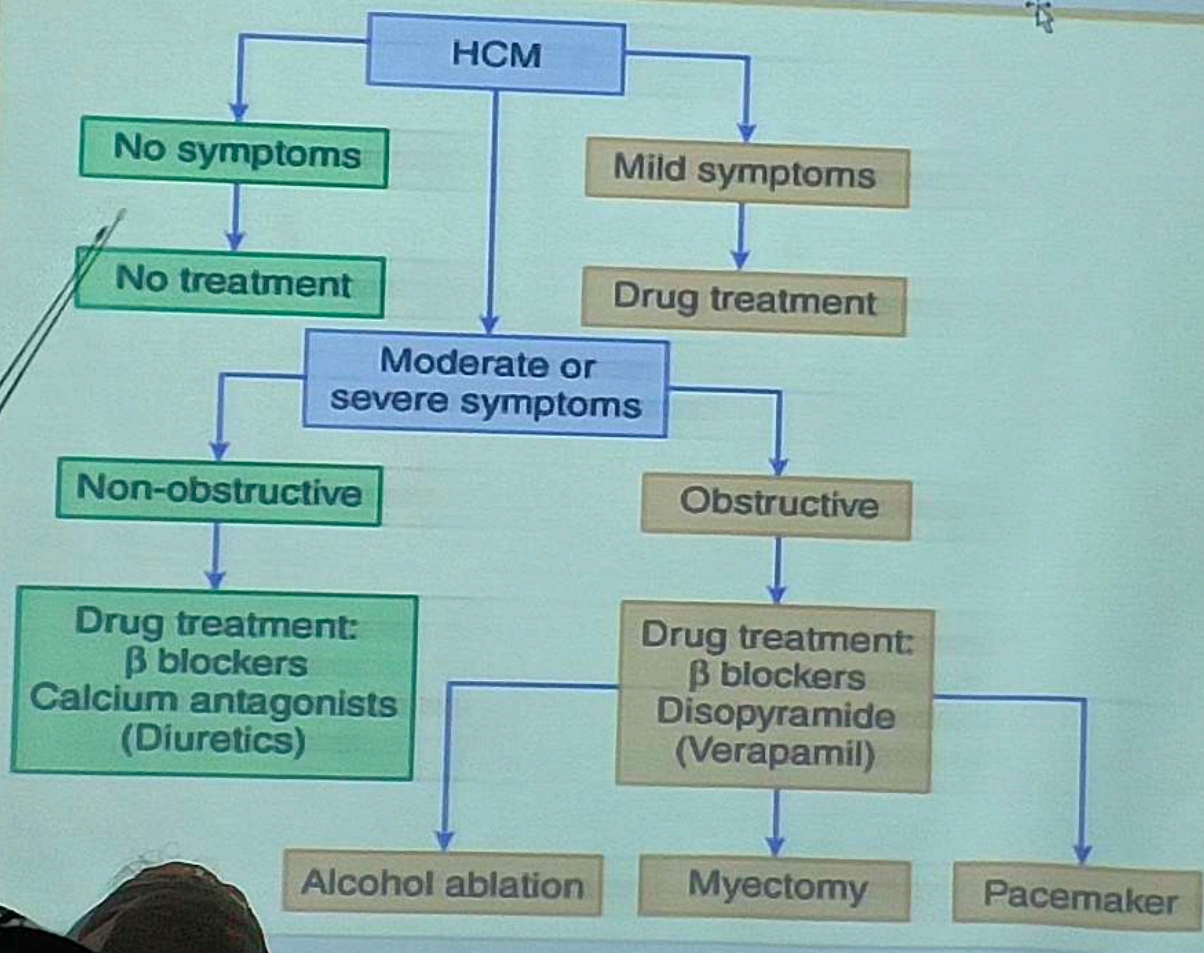
## **A. Medical treatment**

- B blockers
- Calcium blockers
- Amiodarone
- Treatment of C.C.F.

## **B. Alternative to surgery**

- Dual chamber pacing
- Alcohol septal ablation

## **C. Surgical treatment**



Notes

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## **Classification Of The Restrictive Cardiomyopathies**

### **Myocardial**

#### **1. Noninfiltrative cardiomyopathies**

**Idiopathic**

**Familial**

**Pseudoxanthoma elasticum**

**Scleroderma**

#### **2. Infiltrative cardiomyopathies**

**Amyloidosis**

**Sarcoidosis**

**Gaucher's disease**

#### **3. Storage disease**

**Hemochromatosis**

**Fabry's disease**

**Glycogen storage diseases**

### **Endomyocardial**

#### **1. Obliterative**

**Endomyocardial fibrosis**

**Hypercosinophilic syndrome**

#### **2. Nonobliterative**

**Carcinoid**

**Malignant infiltration**

**Iatrogenic (radiation, drugs)**

## Classification of Types of Restrictive Cardiomyopathy According to Cause

### Myocardial

#### *Noninfiltrative*

- Idiopathic cardiomyopathy\*
- Familial cardiomyopathy
- Hypertrophic cardiomyopathy
- Scleroderma
- Pseudoxanthoma elasticum
- Diabetic cardiomyopathy

#### *Infiltrative*

- Amyloidosis\*
- Sarcoidosis\*
- Gaucher disease
- Hurler disease
- Fatty infiltration

#### *Storage Disease*

- Hemochromatosis
- Fabry disease
- Glycogen storage disease

#### *Endomyocardial*

- Endomyocardial fibrosis\*
- Hypereosinophilic syndrome
- Carcinoid heart disease
- Metastatic cancers
- Radiation\*
- Toxic effects of anthracycline\*
- Drugs causing fibrous endocarditis (serotonin, methysergide, ergotamine, mercurial agents, busulfan)



## Restrictive / Obliterative Cardiopathy

- Diastolic vent. Volume and stretch are impaired by morphologic endocardial sub-endocardial or myocardial lesions
- There is limitation of ventricular filling leading to increased filling pressure
- Systolic function in early stage is good
- The most common cause world wide is probably endomyocardial fibrosis with / without eosinophilia
- When process is extensive, ventricular cavity is reduced in size i.e., Obliterative cardiomyopathy

# Clinical Manifestations

## Acute E.M.F.

- Oedema
- Breathlessness
- Fever
- These symptoms commonly remit & patient is well for a period
- It is followed by progressive tiredness, general ill-health with prolonged fever
- Signs of Lt. And Rt. Heart failure dyspnea, cough, hepatomegaly
- Sinus tachycardia and atrial fibrillation common
- massive pericardial effusion may cause tamponade
- Clinical picture may mimic constrictive pericarditis
- Mitral regurgitation may occur



# Investigations

## A. X-rays chest

- Moderate enlargement of heart
- Enlarged L.A. Pulmonary Venous congestion
- Small pleural effusion
- Calcific linear deposit in the region of L.V. apex

## B. E.C.G.

- L.V.H.
- L.A.H.
- Occasional R.V.H.
- QRS voltage diminished

# Investigations

## **C. Echocardiography**

- Increase bright echoes in the endocardium
- May show pericardial effusion
- Defines functional consequence of disease
- Obliteration of apices of ventricles, presence of thrombus and calcification
- Preserved vent. contractility and atrial dilatation

## **D. Angiography**

## **E. Endomyocardial biopsy**



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# Treatment

## Medical Treatment

- Treatment of heart failure
- Anticoagulants
- Digitalis of little value
- Restrict fluids

## Surgical treatment

English (United Kingdom)

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Notes

# Classification of Cardiomyopathies

## Classification of the Cardiomyopathies

Disorder	Description
Dilated cardiomyopathy	Dilation and impaired contraction of the left or both ventricles. Caused by familial-genetic, viral, and/or immune, alcoholic-toxic, or unknown factors or is associated with recognized cardiovascular disease.
Hypertrophic cardiomyopathy	Left and/or right ventricular hypertrophy, often asymmetrical, which usually involves the interventricular septum. Mutations in sarcomeric proteins cause the disease in many patients.
Restrictive cardiomyopathy	Restricted filling and reduced diastolic size of either or both ventricles with normal or near-normal systolic function. Is idiopathic or associated with other disease (e.g., amyloidosis, endomyocardial disease).
Arrhythmogenic right ventricular cardiomyopathy	Progressive fibrofatty replacement of the right, and to some degree left, ventricular myocardium. Familial disease is common.
Unclassified cardiomyopathy	Diseases that do not fit readily into any category. Examples include systolic dysfunction with minimal dilation, mitochondrial disease, and fibroelastosis.

Notes



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# Functional Classification of Cardiomyopathies

## Functional Classification of the Cardiomyopathies

Dilated	Restrictive	Hypertrophic
<b>Symptoms</b> Congestive heart failure, particularly left-sided Fatigue and weakness Systemic or pulmonary emboli	Dyspnea, fatigue Right-sided congestive heart failure Signs and symptoms of systemic disease e.g., amyloidosis, iron storage disease	Dyspnea, angina pectoris Fatigue, syncope, palpitations
<b>Physical Examination</b> Moderate to severe cardiomegaly; S <sub>3</sub> , S <sub>4</sub> Atrioventricular valve regurgitation, especially mitral	Mild to moderate cardiomegaly; S <sub>3</sub> or S <sub>4</sub> Atrioventricular valve regurgitation; inspiratory increase in venous pressure (Kussmaul sign)	Mild cardiomegaly Apical systolic thrill and heave; brisk carotid upstroke S <sub>4</sub> common Systolic murmur that increases with Valsalva maneuver
<b>Chest Roentgenogram</b> Moderate to marked cardiac enlargement, especially left ventricular	Mild cardiac enlargement Pulmonary venous hypertension	Mild to moderate cardiac enlargement Left atrial enlargement



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51	<b>Electrocardiogram</b> Sinus tachycardia Atrial and ventricular arrhythmias ST segment and T wave abnormalities Intraventricular conduction defects	Low voltage Intraventricular conduction defects Atrioventricular conduction defects	Left ventricular hypertrophy ST segment and T wave abnormalities Abnormal Q waves Atrial and ventricular arrhythmias
52			
53	<b>Echocardiogram</b> Left ventricular dilation and dysfunction Abnormal diastolic mitral valve motion secondary to abnormal compliance and filling pressures	Increased left ventricular wall thickness and mass Small or normal-sized left ventricular cavity Normal systolic function Pericardial effusion	Asymmetrical septal hypertrophy (ASH) Narrow left ventricular outflow tract Systolic anterior motion (SAM) of the mitral valve Small or normal-sized left ventricle
54			
55	<b>Radiotracer Studies</b> Left ventricular dilation and dysfunction (RVG)	Infiltration of myocardium ( <sup>201</sup> Tl) Small or normal-sized left ventricle (RVG) Normal systolic function (RVG)	Small or normal-sized left ventricle (RVG) Vigorous systolic function (RVG) Asymmetrical septal hypertrophy (RVG or <sup>201</sup> Tl)
56	<b>Cardiac Catheterization</b> Left ventricular enlargement and dysfunction Mitral and/or tricuspid regurgitation Elevated left- and often right-sided filling pressures	Diminished left ventricular compliance "Square root sign" in ventricular pressure recordings Preserved systolic function Elevated left- and right-sided filling pressures	Diminished left ventricular compliance Mitral regurgitation Vigorous systolic function Dynamic left ventricular outflow gradient
57			