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Physiotherapy cardio-respiratory Clinic Course

L 19 : Cardiomyopathies

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Contents

- Definition
- Classification
- Types
- Clinical features
- Investigations
- Management

- cardiomyopathies are a group of diseases that affect the heart muscle itself and are not the result of hypertension or congenital or acquired valvular, coronary, or pericardial abnormalities.

Classification

1.Etiologic Classification

(i) a primary CARDIOMYOPATHY

(ii) a secondary CARDIOMYOPATHY

2.Anatomic and Physiologic Classification

1.Etiologic Classification

(i) a primary type:_

heart muscle disease of unknown cause

- Idiopathic (D,R,H)
- Familial (D,R,H)
- Eosinophilic endomyocardial disease (R)
- Endomyocardial fibrosis (R)

(ii) a secondary type:_

myocardial disease of known cause or associated with a disease involving other organ systems

2.Anatomic and Physiologic

1_ Dilated cardiomyopathy (DCM)

2_Hypertrophic cardiomyopathy (HCM)

3_ Restrictive cardiomyopathy (RCM)

4_ Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D)

5_ Unclassified cardiomyopathies

SECONDARY MYOCARDIAL INVOLVEMENT

Infective (D)
Viral myocarditis
Bacterial myocarditis
Fungal myocarditis
Protozoal myocarditis
Metazoal myocarditis
Spirochetal
Rickettsial

Metabolic (D)

Familial storage disease (D,R)
Glycogen storage disease
Mucopolysaccharidoses
Hemochromatosis
Fabry's disease

Deficiency (D)
Electrolytes
Nutritional

Peripartum heart disease (D)

Connective tissue disorders (D)
Systemic lupus erythematosus
Polyarteritis nodosa
Rheumatoid arthritis
Progressive systemic sclerosis
Dermatomyositis

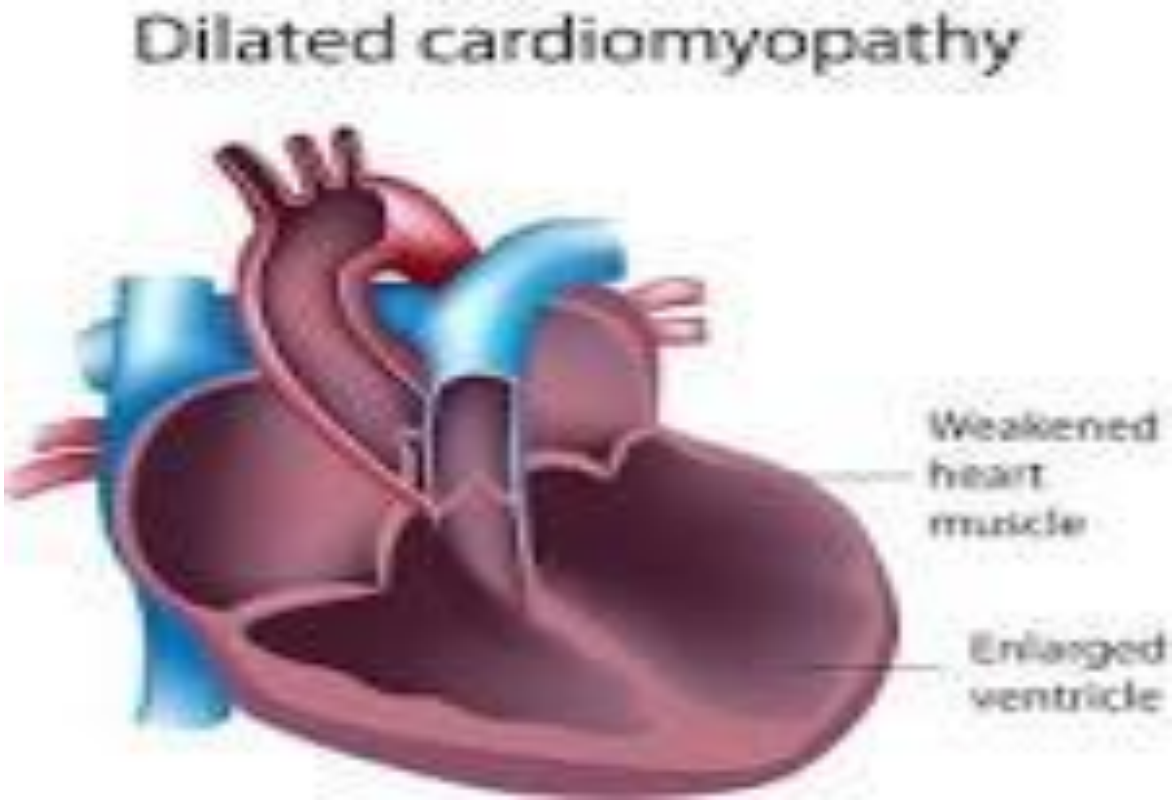
Infiltrations and granulomas (R,D)
Amyloidosis
Sarcoidosis
Malignancy

Neuromuscular (D)
Muscular dystrophy
Myotonic dystrophy
Friedreich's ataxia (H,D)

Sensitivity and toxic reactions (D)
Alcohol
Radiation
Drugs (anthracycline_cyclophosphamide)

1_Dilated cardiomyopathy (DCM

- This is characterised by dilatation and impaired of the Left and/or right ventricular systolic pump function(contraction)
- leading to progressive cardiac enlargement and congestive heart failure, arrhythmias and emboli


Dilated cardiomyopathy
Weakened heart muscle
Enlarged ventricle

- The common causes viruses and gene mutations.

-alcoholic cardiomyopathy

_peripartum cardiomyopathy

_neuromuscular disease(cardiac involvement)

_drugs cardiomyopathy.

- Coronary artery disease and valve disease are commonly called “ischemic cardiomyopathy” or “valvular cardiomyopathy

Clinical features

DCM can present with:_

_Symptomof left- and right-sided congestive heart failure develop gradually

Fatigue.dyspnoea.

_conduction defects or cardiac arrhythmias

_Syncope due to arrhythmias and systemic embolism

_sudden death.

_Some patients have left ventricular dilatation for months or even years before becoming symptomatic.

PHYSICAL EXAMINATION

_Variable degrees of cardiac enlargement

_ findings of CHF are noted

oedema.jaundice.hepatomegaly.ascites

_ the pulse pressure is narrow

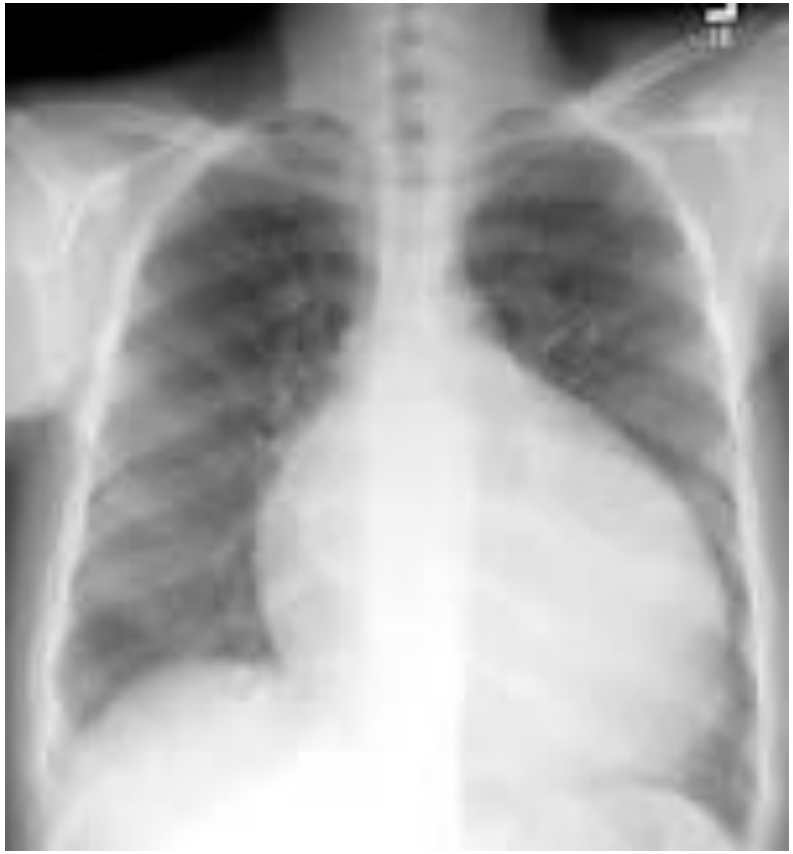
_ the jugular venous pressure is elevated.

_Third and fourth heart sounds are common

_mitral or tricuspid regurgitation may occur.

Investigations

- **Chest X-ray**
- generalized cardiomegaly
- pulmonary venous hypertension
- interstitial or alveolar edema.



■ **ECG may**

- diffuse non-specific ST segment and T wave changes
- Sinus tachycardia
- arrhythmias
- atrial fibrillation,
- ventricular premature contractions

- ■ **Echo**

- dilatation
- low ejection fraction
- sinus tachycardia or atrial fibrillation
- ventricular arrhythmias
- left atrial abnormality

■ **Cardiac MR**

demonstrate other aetiologies of left ventricular dysfunction (e.g):_

_previous myocardial infarction or myocardial fibrosis

BNP

sensitive and specific in diagnosis of HF

■ Cardiac catheterization and coronary angiography

are often performed to exclude ischemic heart disease, (generally patients > 40 years or younger if symptoms or risk factors are present).

■ Biopsy

not indicated outside specialist care

treatment

- Bed rest
- Standard therapy of heart failure with salt restriction
- diuretics
- digitalis
- anticoagulation.
- (ACE) inhibitors
- An angiotensin II receptor blocker in ACE-intolerant pt.
- Most patients should be treated with adrenergic blocker.
- Spironolactone should be added for most patients with recent or current advanced heart failure.

- biventricular pacing

- Insertion of an implantable cardioverter-defibrillator (ICDs)in patients with NYHA III/IV

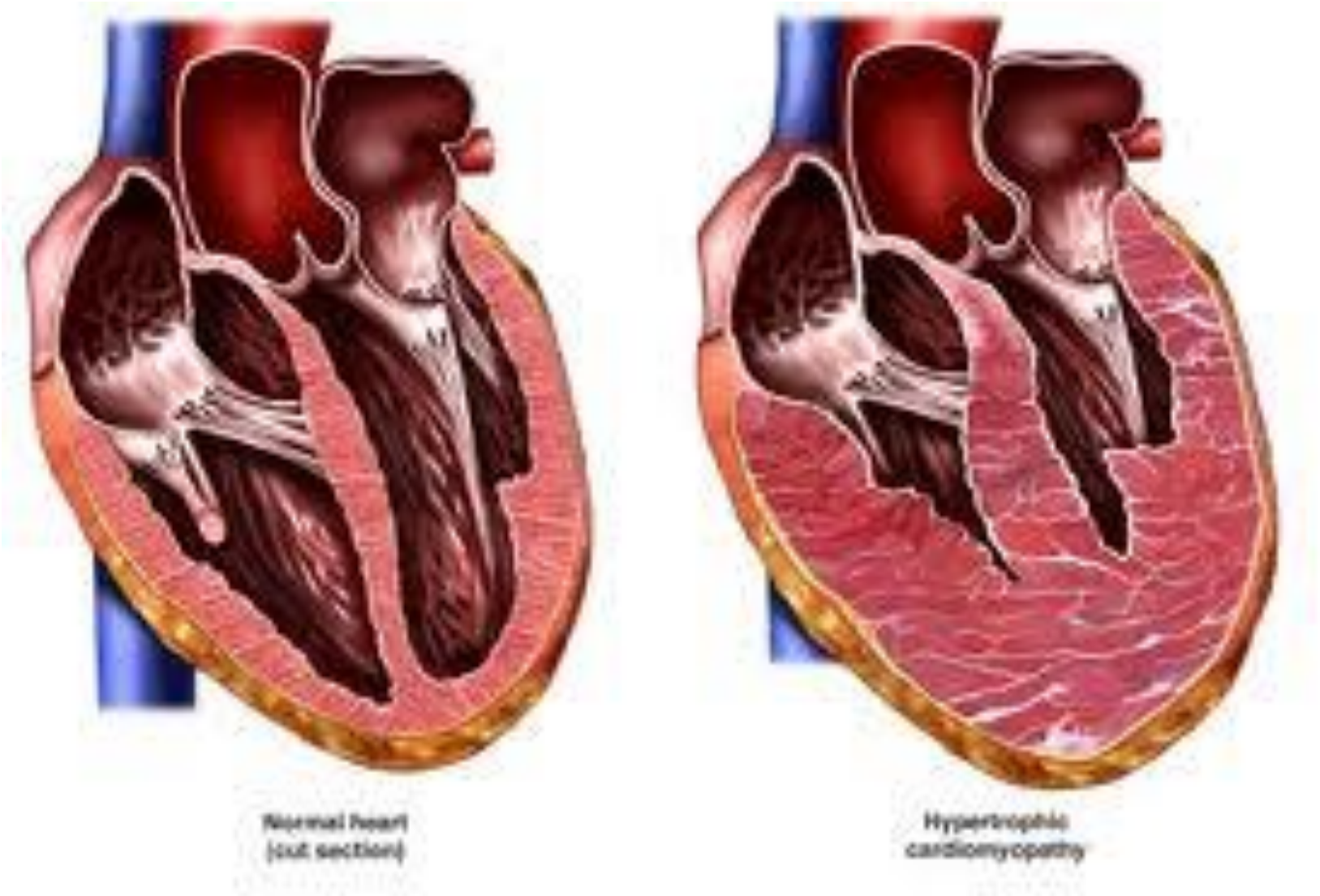
cardiac transplantation

- if refractory to medical therapys

should be avoided

- Alcohol (its cardiac toxic effects)
- calcium channel blockers
- nonsteroidal anti-inflammatory drugs
- Antiarrhythmic agents unless they are needed to treat symptomatic or serious arrhythmias).

2_Hypertrophic cardiomyopathy (HCM)



- HCM is hypertrophy of the myocardium and diastolic dysfunction which reduced ventricular volume, with Characteristic histologic changes include myocyte hypertrophy and disarray
- hypertrophy of the myocardium occur in the absence of an alternate cause (e.g. aortic stenosis or hypertension)
- The interventricular septum is typically more prominently involved than the left ventricular free wall, but concentric and apical hypertrophy can occur
- It is the most common cause of sudden cardiac death in young
- . The majority of cases are familial autosomal dominant, due to mutations of the β-myosin heavy chain and myosin-binding protein C. and Other mutations
- First-degree relatives of patients with HCMshould bescreened by echocardiograph

HEMODYNAMICS

(1) increased left ventricular contractility >>(*elevated left ventricular* diastolic pressure)

(2) decreased ventricular volume (preload),

(3) decreased aortic impedance and pressure (afterload).

CLINICAL FEATURES

- **The clinical course of HCMis highly variable.**
- . Unfortunately, the first clinical manifestation of the disease may be sudden death, frequently occurring in children and young adults, often during or after physical exertion.
- many are asymptomatic and are detected through family screening or ECG examination
- In symptomatic patients:_

1_ dyspnea

. the most common complaint

. due to increased stiffness of the left ventricular walls

. impairs ventricular filling and left ventricular outflow tract obstruction

- 2_ Other symptoms include:_
- (chest pain,palpitation, syncope or pre-syncope (typically with exertion)
- ***Signs***
- a double or triple apex beat
- fourth heart sound)
- a rapidly rising carotid arterial pulse
- Systolic thrillat lower left sternal edge
- Harsh ejection systolic murmur

  * Is The hallmark of obstructive HCMis, which is typically harsh, diamond-shaped, and usually begins well after the first heart Sound

Investigations

- ■ ECG
- left ventricular hypertrophy
- T wave changes invertion
- deep, broad Q waves especially in the infero-lateral leads. suggest an old myocardial infarction
- arrhythmias
- both atrial (supraventricular tachycardia or atrial fibrillation) and ventricular (ventricular tachycardia), during ambulatory (Holter) monitoring

- **■ Echocardiography**

- The mainstay of the diagnosis of HCMis the *echocardiogram, which demonstrates:_*
- . left ventricular hypertrophy
- apical hypertrophy
- The septum may demonstrate an unusual “ground-glass” appearance,
- The left ventricular cavity typically is small in HCM,

- ■ Cardiac MR can detect both the
- hypertrophy
- abnormal myocardial fibrosis
- Exercise test with or without Holter monitor to risk stratify
- Genetic analysis, where available, may confirm the

. *Radionuclide scintigraphy with thallium 201 frequently*

(myocardial perfusion defects even in asymptomatic patients)

- *Chest roentgenography may be normal, although* a mild to moderate increase in the cardiac silhouette is common
- cardiac catheterization

Interventions that increase ventricular volume and ameliorate the gradient and murmur (+ve)

1_ elevation of arterial pressure by:_

- Phenylephrine
- Squatting
- sustained handgrip

2_ augmentation of venous return by :_

- passive leg raising

(-ve) Interventions that increase in the gradient and the murmur

- 1_increase myocardial contractility:_
- exercise
- sympathomimetic amines
- digitalis glycosides

2_ that reduce ventricular volume

Valsalva maneuver

sudden standing

nitroglycerin

amyl nitrite

Risk factors for sudden death

- ■ massive left ventricular hypertrophy (> 30 mm on echocardiography)
- ■ family history of sudden cardiac death (< 50 years old)
- ■ non-sustained ventricular tachycardia on 24-hour Holter monitoring
- ■ prior unexplained syncope
- ■ abnormal blood pressure response on exercise (flat or hypotensive response).

*and patients with two or more should be assessed for implantable cardioverter–defibrillator (ICD).

*In patients in whom the risk is less, amiodarone is an appropriate alternative.

treatment

- The management of HCM includes treatment of symptoms and the prevention of sudden cardiac death in the patient and relatives.

- beta-blockers.(for angina pectoris and syncope)
- beta-blockers and verapamil either alone or in combination.
- Verapamil and diltiazem increase exercise tolerance and may reduce stiffness of the ventricle and diastolic pressures

_severity of outflow tract pressure gradient

- Amiodarone (arrhythmias)
- anticoagulate

.dual-chamber pacing In some pt with significant left outflow obstruction

.Alcohol (non-surgical) ablation of the septum

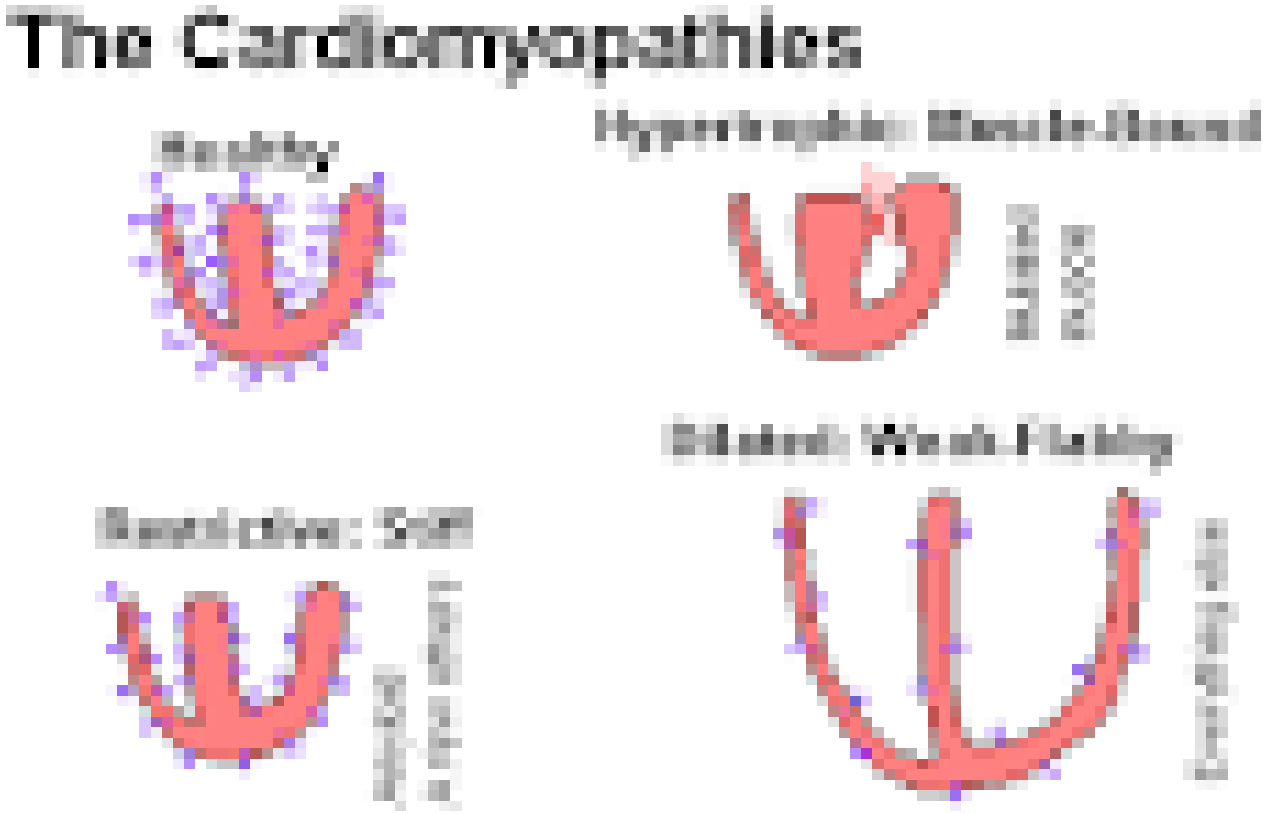
carries risks of heart block and myocardial infarction.

- surgical resection of septal .
- implantable cardioverter defibrillator

pt must avoid

- sports
- Dehydration
- diuretics (used with caution)
- Nifedipine
- . Digitalis,nitrates_Vasodilators
  exacerbate an outflow pressure gradient)
- adrenergic agonists
- social alcohol (produce sufficient vasodilatation)

3_ Restrictive cardiomyopathy (RCM)



Restrictive cardiomyopathies is abnormal diastolic function the ventricular walls are excessively rigid and impede ventricular filling. Myocardial fibrosis, hypertrophy, or infiltration

must be distinguished from constrictive pericarditis, which causes a similar impairment in ventricular fillingPrimary

Causes of restrictive cardiomyopathy

- although restriction is also seen in
- amyloidosis
- hemochromatosis,
- glycogen deposition
- endomyocardial fibrosis
- Loeffler's endocarditis
- sarcoidosis
- Fabry's disease
- the eosinophilias,
- scleroderma in the transplanted heart
- following mediastinal irradiation

(eg, idiopathic or familial
The idiopathic form of restrictive cardiomyopathy may be familial (**Noninfiltrative**)

Clinical features

- symptom and signs of heart failure(dyspnoea, fatigue) and embolic symptoms.
- On clinical examination
- there will be elevated jugular venous pressure with diastolic collapse (Friedreich's sign)
- and elevation of venous pressure with inspiration (Kussmaul's sign),
- an enlarged, tender, and often pulsatile liver.
- ascites and dependent oedema.
- Third and fourth heart sounds may be present

Investigations

- ■ Chest X-ray

_ pulmonary venous congestion.

- The cardiac silhouette can be normal or show cardiomegaly and/or atrial enlargement.
- ■ ECG
- _low-voltage QRS and ST segment and T wave abnormalities.
- ■ Echocardiography

_ symmetrical myocardialthickening

_normal systolic ejection fraction

_but impaired ventricular filling.

- ■ Cardiac MR

abnormal myocardial fibrosis in amyloidosis or sarcoidosis

- ■ Cardiac catheterization and haemodynamic studies
- may help distinguish between restrictive cardiomyopathy and constrictive pericarditis

- ■ Endomyocardial biopsy

- . The electrocardiogram often shows
- low-voltage, nonspecific ST-T-wave changes and various arrhythmias.

- x-ray
- Pericardial calcification which occurs in constrictive pericarditis, is absent

- . Echocardiography
- typically reveals symmetrically thickened left ventricular walls and normal or slightly reduced ventricular volumes and systolic function
- . Cardiac catheterization
- a decreased cardiac output
- , elevation of the right and left ventricular end-diastolic pressures

constrictive pericarditis

- may be operation. and right ventricular transvenous endomyocardial biopsy
- computed tomography or magnetic resonance
- imaging (by demonstrating a thickened pericardium in constrictive
- pericarditis).

ENDOMYOCARDIAL FIBROSIS(EMF)

- **is a progressive disease of unknown** cause
- most commonly in tropical and subtropical Africa, particularly Uganda and Nigeria.
- In some ways this disease resembles eosinophilic endomyocardial
- The clinical picture depends on which ventricle and atrioventricular valve show predominant involvement;
- left-sided involvement results in symptoms of pulmonary congestion, while predominant right-sided disease presents features of a restrictive cardiomyopathy.
- Medical treatment is often disappointing, and surgical .
- Surgical treatment with endomyocardial resection and valve replacement may be beneficial

EOSINOPHILIC ENDOMYOCARDIAL DISEASE
(Loeffler's endocarditis)

_hypereosinophilic syndrome in which the heart is predominantly involved

_ Large mural thrombi may develop in e ventricl serving as a source of pulmonary and systemic emboli.

_ Hepatosplenomegaly and localized eosinophilic infiltration of other organs are usually present.

Management

diuretics, afterload-reducing agents

anticoagulation.

glucocorticoids

Cytotoxic drug

. Surgical treatment

amyloidosis

- **Involvement of the heart** is **the most frequent** cause of death in *primary amyloidosis .,while clinically* significant cardiac involvement is uncommon in the secondary form
- The heart is firm, rubbery, and noncompliant
- four clinical presentations (alone or in combination) are seen:_

(1) diastolic dysfunction (restrictive cardiomyopathy)
(2) systolic dysfunction
(3) arrhythmias and conduction disturbances
(4) orthostatic hypotension.

Diagnosis:_ two dimensional echocardiogram

treatment.

_ Chemotherapy with alkylating agents

_heart transplantion (often combined with bone marrow transplantation, or liver or kidney transplantation for hereditary amyloidosis) may help

_selected patients

, prognosis is poor, especially in the primary form with advanced cardiac involvement.

Hemochromatosis

_ *result of multiple transfusions* or ahemoglobinopathy

_the familial (autosomal recessive) form should be suspected if cardiomyopathy occurs in the setting of diabetes mellitus, hepatic cirrhosis, and increased skin pigmentation.

diagnosis endomyocardial biopsy.

Treatment :_ deferoxamine

Myocardial *sarcoidosis*

generally associated with other manifestations of systemic disease and may cause restrictive as well as congestive features

_ infiltration by sarcoid granulomas
_ increased stiffness of the myocardium
_ diminished systolic contractile function
_ . A variety of arrhythmias,

- common cardiac manifestation of systemic sarcoidosis is right heart overload due to pulmonary artery hypertension

Treated:_ glucocorticoids

4_Arrhythmogenic right ventricular cardiomyopathy

- heart muscle disease characterized by ventricular arrhythmias and a specific myocardial pathology, The myocardium of the right ventricular free wall (and frequently the left ventricle as well) is replaced by fibrous and/or fibro-fatty tissue, with scattered residual myocardial cells.

Laboratory Evaluation of the Cardiomyopathies

	Dilated	***Restrictive***	***Hypertrophic***
Chest roentgenogram	Moderate to marked cardiac silhouette enlargement Pulmonary venous hypertension	Mild cardiac silhouette enlargement	Mild to moderate cardiac silhouette enlargement
Electrocardiogram	ST-segment and T-wave abnormalities	Low voltage, conduction defects	ST-segment and T-wave abnormalities Left ventricular hypertrophy Abnormal Q waves
Echocardiogram	Left ventricular dilatation and dysfunction	Increased left ventricular wall thickness Normal or mildly reduced systolic function	Asymmetric septal hypertrophy (ASH) Systolic anterior motion (SAM) of the mitral valve
Radionuclide studies	Left ventricular dilatation and dysfunction (RVG)	Normal or mildly reduced systolic function (RVG)	Vigorous systolic function (RVG) Perfusion defect (201Tl)
Cardiac catheterization	Left ventricular dilatation and dysfunction Elevated left- and often right-sided filling	Normal or mildly reduced systolic function Elevated left- and rightsided	Vigorous systolic function Dynamic left ventricular outflow obstruction Elevated left- and

- Thank you