CARDIOMYOPTHY

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- A heterogeneous group of diseases of the myocardium associated with mechanical and/or electric dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilatation due to a variety of etiologies that are frequently genetic.
- Cardiomyopathies are either confined to the heart or part of generalised systemic disorders and often lead to cardiovascular death or progressive heart failure-related disability.

- Constitutes a group of diseases that directly affect the structural or functional ability of the myocardium.
- Cardiomyopathies are a mixed group of diseases of the myocardium (cardiac muscle) defined by structural or functional abnormalities that negatively affect the pump function of the heart. In some types, there is obstruction to the outflow of blood during the cardiac cycle.

CAUSES

- Causes of cardiomyopathy are classified as PRIMARY or SECONDARY

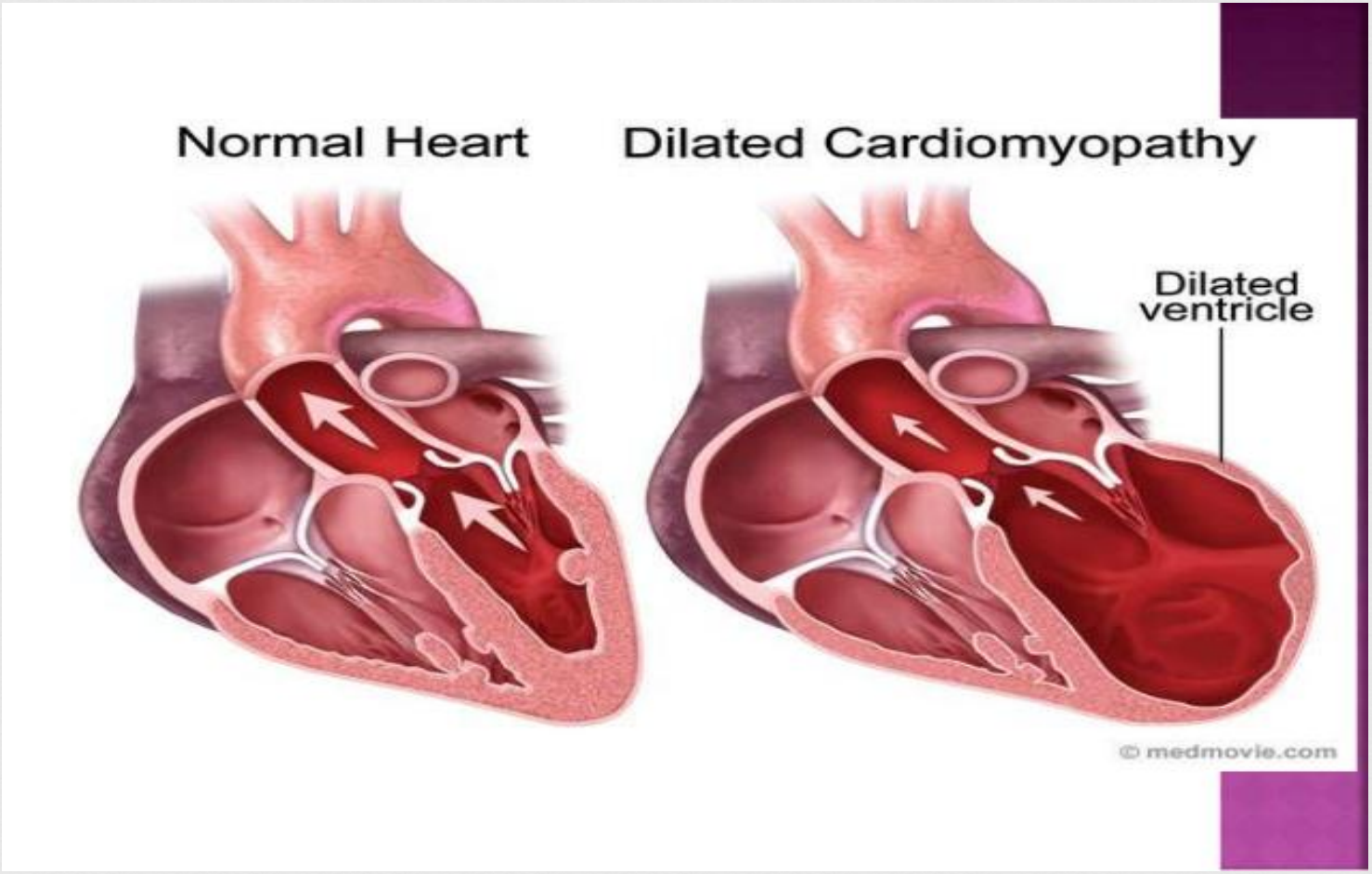
- 1. Primary cardiomyopathies have genetic, mixed, or acquired etiologies.
- 2. Secondary cardiomyopathies have infiltrative, toxic or inflammatory causes

Types of Cardiomyopathies

- Cardiomyopathies classified according to the structural abnormalities of the heart muscle.
- 1. Dilated cardiomyopathy (DCM)
- 2. Hypertrophic cardiomyopathy (HCM)
- 3. Restrictive or Constrictive cardiomyopathy (RCM)
- 4. Arrhythmogenic right ventricular cardiomyopathy (ARVC) and
- 5. Unclassified cardiomyopathy

Dilated Cardomyopathy

- It is a condition in which the hearts ability to pump blood is decreased because the hearts main pumping chamber, the left ventricle, is enlarged or weakened.
- Dilated cardiomyopathy is the most common form of cardiomyopathy.


Normal Heart
Dilated Cardiomyopathy
Dilated ventricle
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Pathophysiology

- It can be divided into two types;
- **1. Ischemic cardiomyopathy** :
- a) It is caused by inadequate oxygen supply due to obstruction in coronary artery.
- b) The lack of oxygen interrupts both mechanical & electrical function of the cells , decrease contractility , and causes dysrhythmias.

- **2) Non ischemic cardiomyopathy**:
- a) Cause is idiopathic (unknown)
- b) 10 to 50 % of cases are identified by genetic mutation
- c) Both the RV & LV enlarge significantly, causing a decrease in the ability of the heart to pump blood efficiently to the body.

Causes

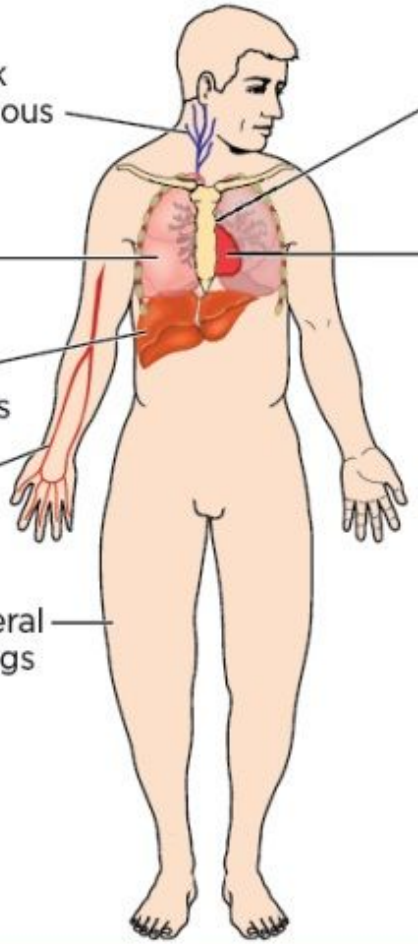
- Coronary Artery Disese
- Alcohol abuse
- Chemotherapy
- Chemical agents
- Myocarditis
- Pregnancy (third trimester & postpartum)
- Valve disease, endocrine disorders, and
- Infections such as HIV can cause dilated cardiomyopathy.

Clinical Features

- Decreased exercise capacity, Fatigue, Dyspnea, Paroxysmal nocturnal dyspnea ,Orthopnea
- As the disease progresses, Dry cough, palpitations , Abdominal bloating, Nausea, vomiting , anorexia
- Abnormal S3 and S4 sound ,Tachycardia or bradycardia, Edema , Pulmonary crackles, Weak peripheral pulses, Hepatomegaly, Jugular venous distention.

Fig 2. **Cardiomyopathies: signs and symptoms**

General: cachexia, fatigue, dyspnoea, patient unable to tolerate lying flat (needs to be propped up by pillows), plus any other signs and symptoms of underlying systemic disease



Neck: distended neck veins from raised venous jugular pressure

Lungs: pulmonary crackles

Gastrointestinal: enlarged liver, ascites

Pulse: irregular

Extremities: peripheral oedema involving legs (up to sacrum)

On palpation: parasternal lift or systolic thrills; apex beat may be displaced

On auscultation: valvular murmurs (for example, pan-systolic murmur from mitral regurgitation); gallop rhythm caused by extra heart sound

Diagnostic Measures

- History,
- Echocardiography,
- Chest x-ray: shows the signs of cardiomegaly,
- ECG: reveals tachycardia, bradycardia and dysarrythmias.
- Cardiac catheterization: it is performed to confirm Coronary Artery Disease.

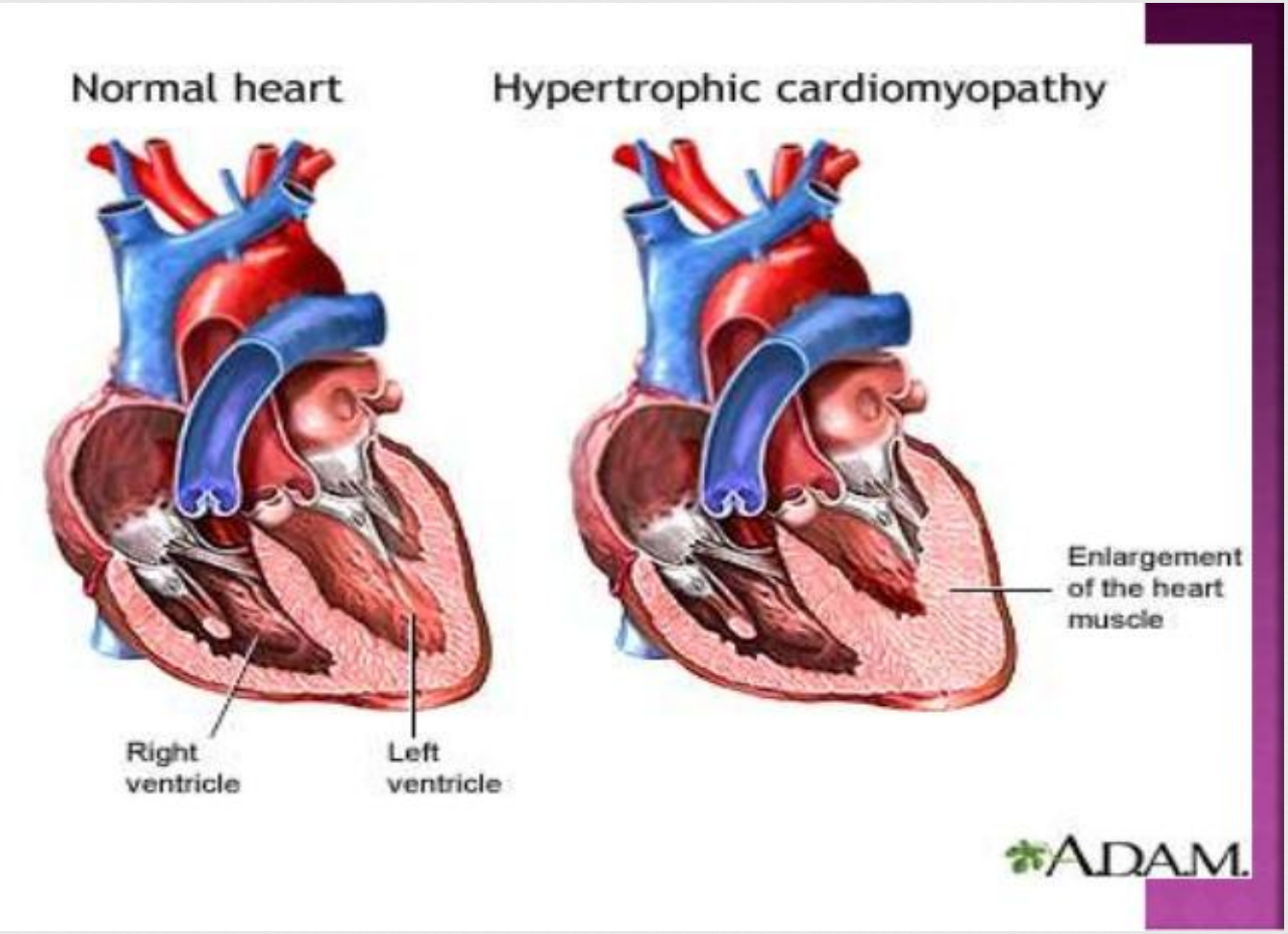
Management

- Nitrates: isosorbitrate,
- Loop diuretics: furosemide,
- ACE inhibitors: captopril,
- Beta adrenergic blockers: atenolol,
- Aldosterone agonists: spironolactone,
- Cardiac glycoside : digoxin,
- Anticoagulation therapy
- Cardiac Transplant

Hypertrophic Cardiomyopathy

- Assymetric left ventricular hypertrophy without ventricular dilation. Hypertrophic cardiomyopathy is a genetically transmitted disorders. Hypertrophic cardiomyopathy is primarily due to the abnormal thickening of the ventricular septum of the heart without ventricular dilation.
- When the septum between two ventricles become enlarged and obstructs the blood flow from left ventricle, it is known as hypertrophic obstructive cardiomyopathy.

- **CAUSES :**
- Aortic stenosis, Genetic, Hypertension, More common in men between ages 30 to 40
- **CLINICAL FEATURES :**
- Exertional dyspnea (Shortness of breath during exercise), Decreased cardiac output, Fatigue, Angina, Syncope, Hypertension


Normal heart
Hypertrophic cardiomyopathy
Enlargement of the heart muscle
Right ventricle
Left ventricle
A.D.A.M.

PATHOPHYSIOLOGY

Thickened intra-ventricular septum and ventricular wall

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ventricular hypertrophy

↓

diastolic dysfunction

↓

impaired ventricular filling and obstruction to decreased outflow

↓

decreased cardiac output

Diagnostic Studies

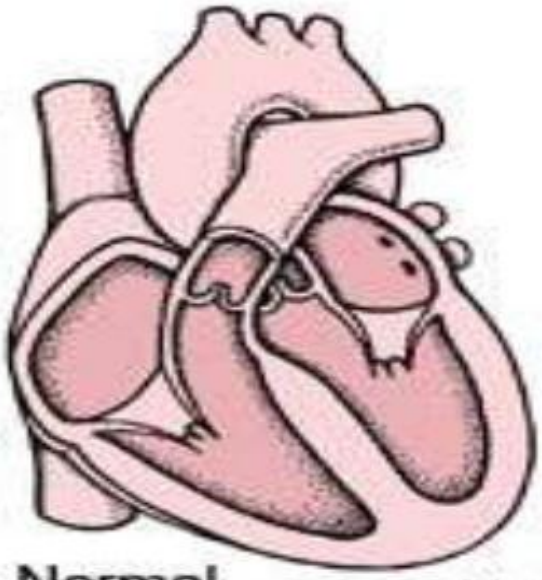
- History and physical examination
- Electrocardiogram (ECG)
- Cardiac MRI.
- Cardiac catheterization

Management

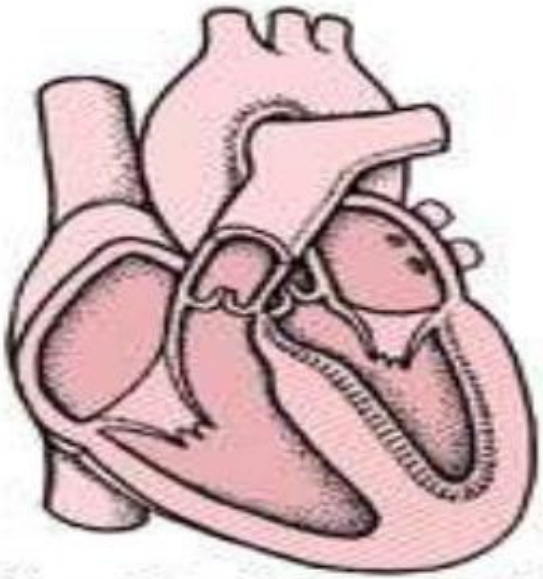
- Beta adrenergic blockers: atenolol,
- Calcium channel blocker: verapamil,
- Antidysrhythmic drugs : amiodarone
- **Septal myectomy:** It is an open heart surgical procedure in which the surgeon removes the part of thickened , over grown septum between the ventricles.
- Septal ablation
- Implantable cardioverter-defibrillator(ICD)

Restrictive Cardiomyopathy

- Disease of the heart muscle that impairs diastolic filling and stretch and the systolic function remains unaffected.
- The heart muscle becomes infiltrated by various substances , resulting in severe fibrosis.
- The heart muscle becomes stiff and non distensible, impairing the ability of the ventricle to fill with blood adequately.
- **ETIOLOGY :**
- Unknown etiology, Myocardial fibrosis, endocardial fibrosis, sarcoidosis and radiation to the thorax.



Normal



Restrictive Cardiomyopathy

In restrictive cardiomyopathy, the walls of the ventricles become stiff, but not necessarily thickened.

PATHOPHYSIOLOGY

etiologic factors

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Stiffness of the ventricular wall with loss of ventricular compliance

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Ventricles become resistant to filling

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decrease cardiac output

Clinical Manifestations

- Fatigue, Exercise intolerance, Dyspnea,
- Orthopnea(shortness of breath (dyspnea) which occurs when lying flat),
- Syncope, Palpitations,
- Peripheral edema, Jugular venous distention.

Diagnostic Studies

- Chest x-ray
- ECG: shows tachycardia
- Echocardiography : for the visualization of left ventricle
- CT-Scan and MRI Scan

Management

- Beta adrenergic blockers: atenolol
- Calcium channel blocker: verapamil
- Steriods: hydrocortisone
- Antidysrhythmic drugs : amiodarone
- **Heart transplantation** may be considered if the heart function is very poor and the symptoms are severe.

Arrhythmogenic Right Ventricular Cardiomyopathy

- Arrhythmogenic right ventricular cardiomyopathy (ARVC), also known as arrhythmogenic right ventricular dysplasia, is a heritable heart muscle disorder that predominantly affects the right ventricle.
- Progressive loss of right ventricular myocardium and its replacement by fibrofatty tissue is the pathological hallmark of the disease.

- The disease is caused by a genetic defect in the cardiac desmosomes reslting in mutations in the genes encoding desmosomal proteins, which are important in cell-to-cell adhesion, play a key role in the pathogenesis of ARVC.
- These proteins include Desmoplakin (DSP), Plakophilin 2 (PKP2), Desmoglein 2 (DSG2), and Desmocollin 2 (DSC2).

Clinical Presentation

- Autosomal dominant
- Prevalence 1 in 2000 to 1 in 5000
- Men to Women ratio 3:1,
- Symptoms appearing between 20-40 years old.
- 20% of sudden death – higher prevalence in young athletes
- Palpitations, syncope and sudden death
- HF in minority of patients

TREATMENT

- **Implantable Cardioverter Defibrillator :**
- For Sustained VT/VF on medical therapy
- As a preventPrimary prevention, patients at high-risk (syncope, severe RV dysfunction, LVdysfunction)
- **Exercise restriction,**
- **Beta Blockers,**
- **ACE Inhibitors.**

Other cardiomyopathies

- Peripartum cardiomyopathy
- Stress-induced cardiomyopathy
- Left ventricular non-compaction