Cardiomyopathy

Dr. Jamal Dabbas
Interventional cardiologist & internist

Definition:

- Group of diseases that primarily affect the heart muscle and are not the result of congenital, acquired, valvular, hypertensive , coronary arterial or pericardial abnormalities

Classification:

1- Dilated

2- Restrictive

3- Hypertrophic CMP

4- Arrhythmogenic RV dysplasia

primary myocardial involvement

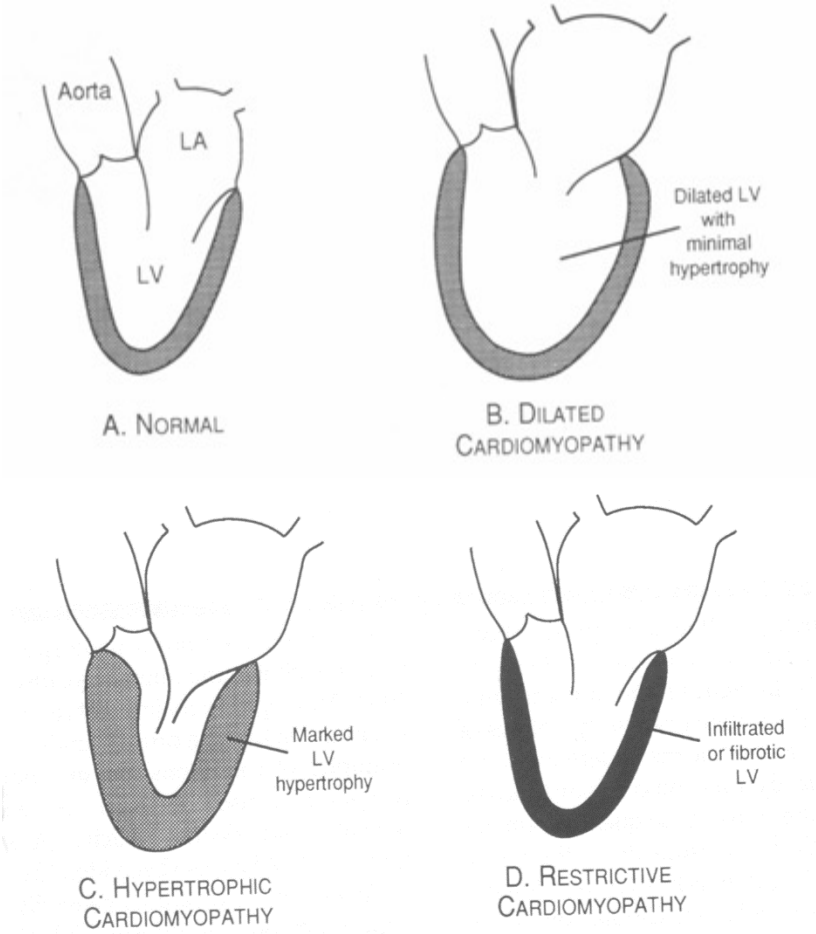
secondary myocardial involvement:
(infective, Metabolic, Infiltration, toxin, peripartum)

Cardiomyopathy

<u>WHO Classification</u>

anatomy & physiology of the LV

1. Dilated
   - Enlarged
   - Systolic dysfunction
2. Hypertrophic
   - Thickened
   - Diastolic dysfunction
3. Restrictive
   - Diastolic dysfunction
4. Arrhythmogenic RV dysplasia
   - Fibrofatty replacement



Cardiomyopathies are diseases of the **muscle tissue** of the **heart**. There are four major morphological types: dilated, **hypertrophic**, restrictive, and **arrhythmogenic right ventricular cardiomyopathy**.

In **dilated cardiomyopathy**, the most common type, ventricular output decreases eventually leading to failure of the left and right **heart**. The dilation can be seen on **echocardiography**, the most important diagnostic tool for all cardiomyopathies. Treatment is mostly similar to that of **heart failure** from other causes.

Hypertrophic cardiomyopathy can occur with or without left ventricular outflow obstruction. It is often asymptomatic, although **arrhythmias** and even sudden cardiac death can occur.

The obstructive type is also notable for signs of reduced blood flow (**dyspnea**, vertigo, **syncope**).
It is important that patients avoid strenuous exercise. Symptomatic patients should be treated with **beta blockers**.

Restrictive cardiomyopathy is caused by the **proliferation** of **connective tissue** with subsequent atrial enlargement (but normal ventricles). Like dilated myopathy, **restrictive cardiomyopathy** produces left and **right heart failure**. **Ejection fraction** is usually normal, but diastolic filling is reduced on **echocardiography**.

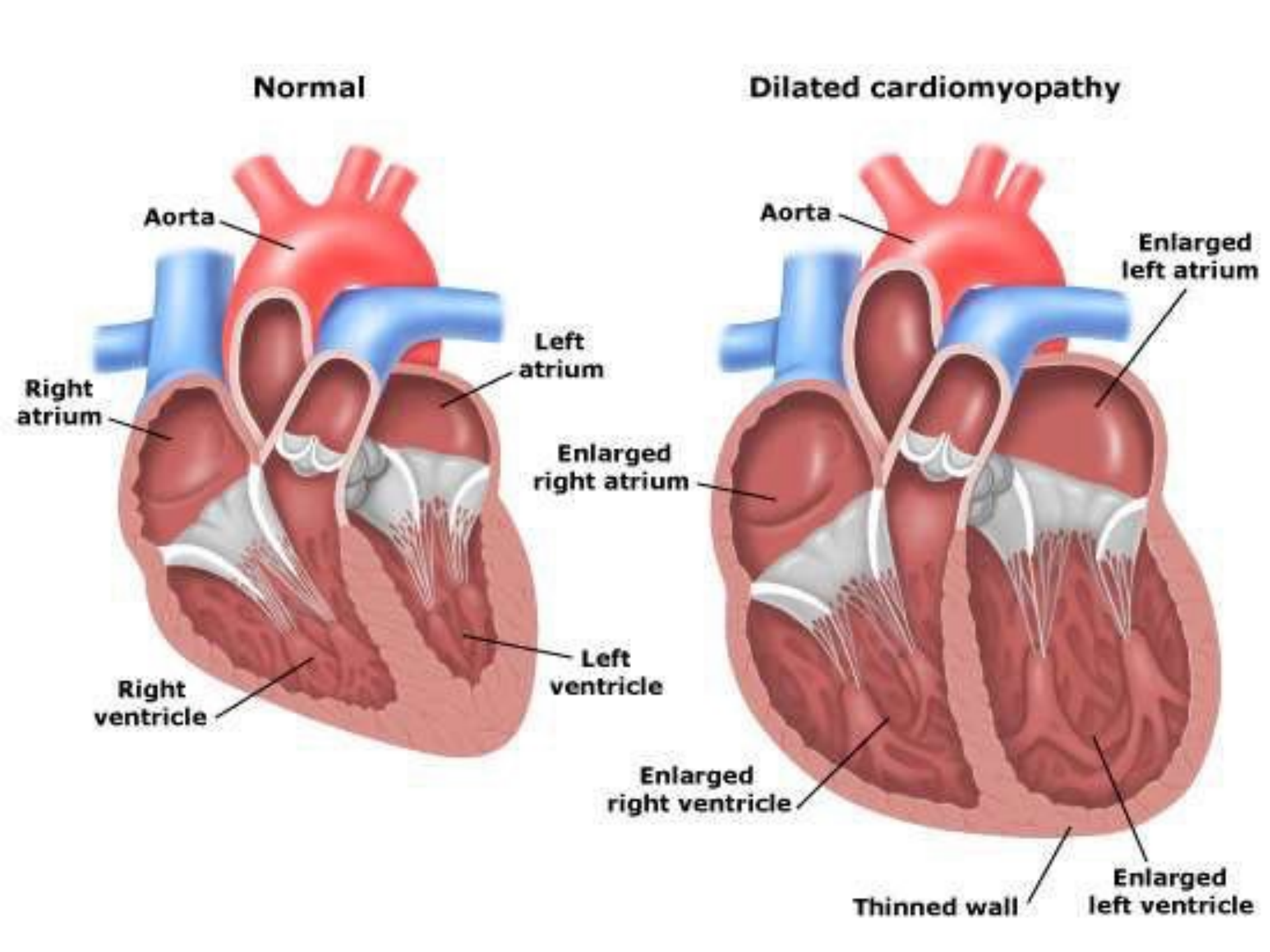
Arrhythmogenic right ventricular cardiomyopathy affects primarily the **right ventricle** and is characterized by cellular breakdown and subsequent ventricular dilation. Symptoms are very variable although the hallmark finding is **arrhythmia**. There is no general best treatment as it depends entirely on individual factors such as the extent of the disease. All patients should avoid strenuous exercise.

Dilated cardiomyopathy (DCM)

Dilated cardiomyopathy

Definition: Dilated cardiomyopathy (DCM) is characterized by left ventricular dilatation and systolic dysfunction in the absence of hypertension, coronary artery disease, valve disease, congenital heart disease.

Atrial dilation as well as right ventricular dilation and dysfunction can also develop.


Normal
Aorta
Left atrium
Right atrium
Left ventricle
Right ventricle
Dilated cardiomyopathy
Aorta
Enlarged left atrium
Enlarged right atrium
Enlarged right ventricle
Thinned wall
Enlarged left ventricle

DCM - Incidence and Prognosis

- Most common cardiomyopathy
- Complete recovery is rare
- 50% die within 2yrs and 25% survive longer than 5yrs

- Reversible form may be found with alcohol abuse, pregnancy, thyroid disease, cocaine use and chronic uncontrolled tachycardia.

Clinical features

- symptoms of left and right sided CHF.
- some patients have LV dilation for months or even years before becoming symptomatic
- vague chest pain (typical angina is unusual and suggestive of IHD)
- syncope due to arrhythmias
- systemic embolism

Physical examination

- Narrow pulse pressure and elevated JVP in advanced disease
- S3
- S4
- MR
- TR

Laboratory examination

- CXR : cardiomegaly , pulmonary congestion
- ECG : Sinus tachycardia or AF , Ventricular arrhythmias, Non specific changes
- Echocardiography : LV dilation , systolic dysfunction
- Angiography : to exclude IHD

Diagnostics

Patient history (especially family history)

ECG: **Left bundle branch block** (unfavorable prognosis)

Laboratory

B-type natriuretic peptide (**BNP**)

Troponin, **creatine kinase** (CK)-MB to rule out **myocardial infarction**

Chest x-ray

Cardiomegaly → left-sided **hypertrophy** with a balloon appearance.

Signs of **heart failure** decompensation: pulmonary edema

Pathology

Eccentric hypertrophy: dilated **left ventricle** with normal or decreased wall thickness.

Treatment

See treatment of heart failure

Avoiding cardiotoxic agents

Abstain from alcohol

Anticoagulation

Treat the underlying disease

Interventional/surgical

If LVEF < 35%: an ICD might prevent sudden cardiac death caused by ventricular fibrillation

Main indication for heart transplantation

Complications

Progressive left ventricular failure → global heart failure

Systemic thromboembolism → stroke, pulmonary embolism, mesenteric infarct

Ventricular tachycardia → ventricular fibrillation

Sudden cardiac death

DILATED CARDIOMYOPATHY
PROVEN THERAPEUTIC OPTIONS

TREATMENT	INDICATIONS
ACE Inhibitors	Symptomatic heart failure and asymptomatic LV dysfunction
ARBs	ACE intolerance ACE intolerance
Hydralazine - nitrates	Add on treatment for resistant CHF
Diuretics	Volume overload
Beta-blockers	Symptomatic heart failure in addition to ACE inhibitor
Digoxin	Persistent heart failure despite diuretics, ACE inhibitor
Warfarin	Chronic or paroxysmal atrial fibrillation LV thrombus or prior embolic
ICD	event Cardiac arrest; uncontrolled VT

Myocarditis

- myocarditis is most commonly the result of infectious process

- Most common cause is viruses especially coxsakievirus B

  adenovirus

  hepatitis C

  and HIV

- Patients with viral myocarditis may give a history of upper respiratory febrile illness or a flu like syndrome ,

  and viral nasopharyngitis or tonsillitis may be evident.

Clinical manifestation

- clinical spectrum ranges from an asymptomatic state to fulminant condition with arrhythmias, acute CHF and death.

- sometime myocarditis simulates an acute coronary syndrome (chest pain, ECG change and elevated troponin level) but typically in patients younger than those with coronary atherosclerosis.

- Viral myocarditis is most often self-limited and without sequelae but sometime may progress to a chronic from and to DCM.

Alcoholic cardiomyopathy

- large quantities (>90g/d) of alcohol over many years cause DCM
- Risk of developing CMP is partially genetically
- patient with severe CHF have a poor prognosis (< 1/4 of such patients survive 3 years)

- Second presentation of alcoholic cardiotoxicity is recurrent supraventricular or ventricular tachyarrhythmia’s
- Holiday heart syndrome: AF, Atrial flutter or frequeut PVC after a drinking binge.

Peripartum cardiomyopathy

- Cardiac dilation and CHF may develop during the last trimester of pregnancy or within 6 months of delivery.
- Mortality rate is 10%.
- Patient who recover from peripartum CMP should be encouraged to avoid further pregnancy .

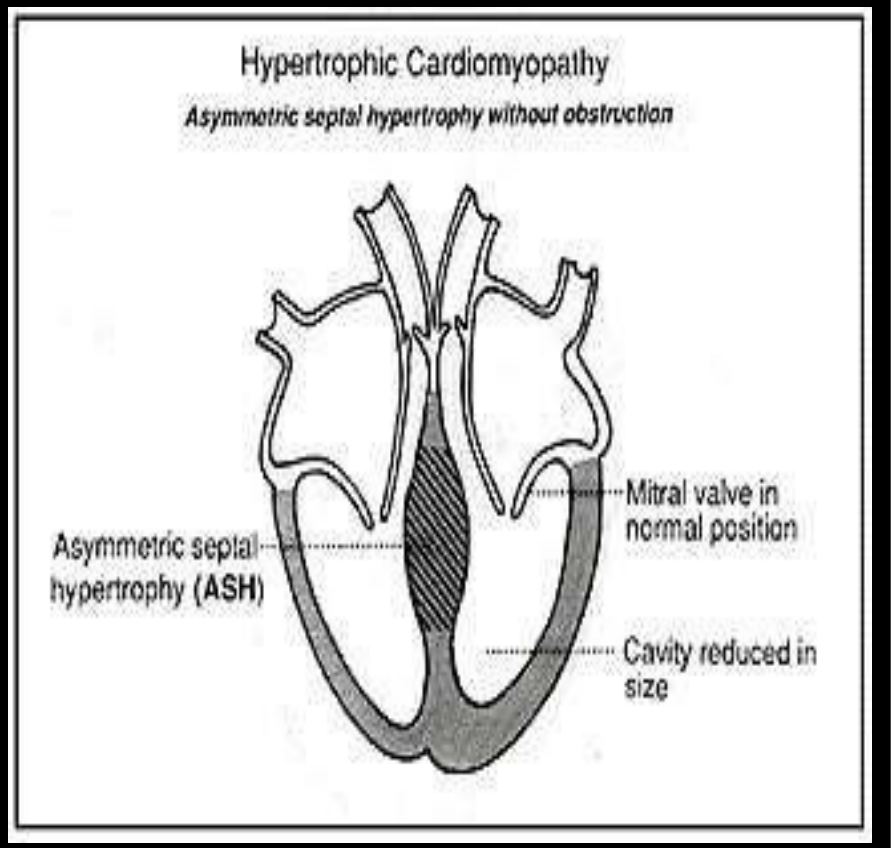
Drugs

- A variety of drugs may damage the myocardium acutely (myocarditis) or they may lead to chronic damage (like DCM)
- Anthracycline derivatives :
- Doxorubicin cardiotoxicity may occur acutely but more commonly develops 3 months after the last dose.
- TCA – antidepressants, phenothiazines , lithium
- Cocaine abuse (SCD , myocarditis , DCM, acute MI)

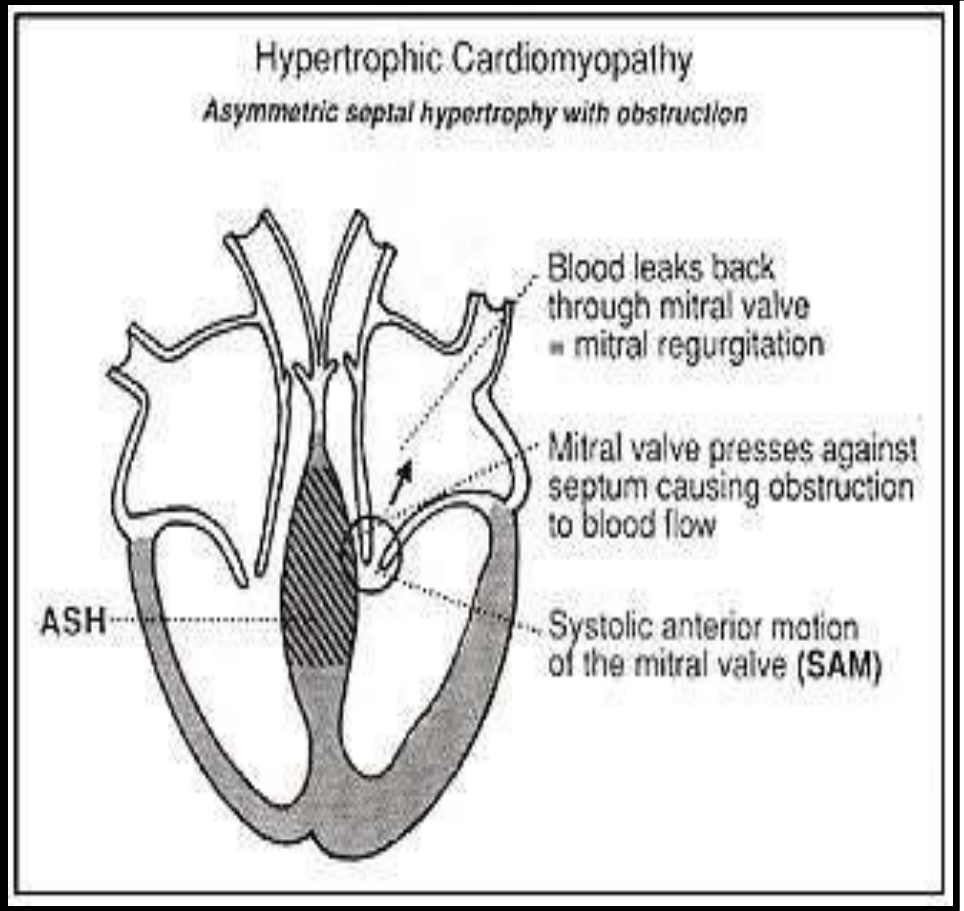
Arrhythmogenic right ventricular cardiomyopathy / Dysphasia (ARVC/D)

- ARVD is a familial CMP with progressive fibrofatty replacement of the RV and to a much lesser degree LV.
- Patients may present by RV failure or ventricular tachycardia

Hypertrophic cardiomyopathy

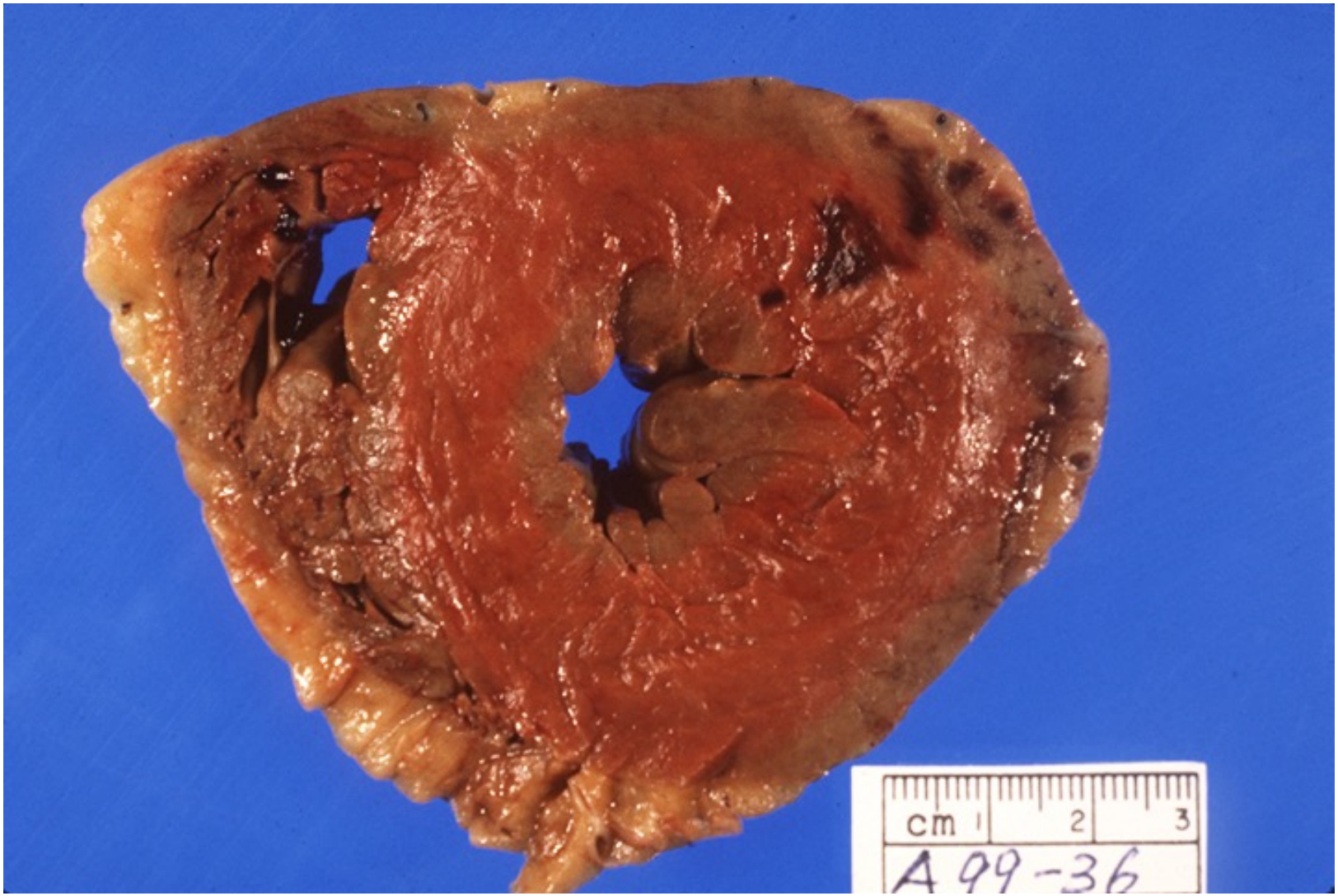


- The major abnormality of the heart in HCM -- excessive thickening of the muscle. Thickening usually begins during early adolescence and stops when growth has finished. uncommon for thickening to progress after this age
- left ventricle almost always affected
- Hypertrophy is usually greatest in the septum, associated with obstruction to the flow of blood into the aorta



- 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

Hypertrophic Cardiomyopathy



Epidemiology

Prevalence: 200/100,000

Obstructive type/**hypertrophic obstructive** cardiom yopathy **(HOCM)**: ~ 70% of cases

Nonobstructive type: ~ 30% of cases

Alongside myocarditis, hypertrophic cardiomyopathy is one of the most frequent causes of **sudden cardiac death in young patients**, especially young athletes.

Etiology

Primary hypertrophic cardiomyopathy (60–90% of cases)

Most common hereditary heart disease

Autosomal dominant inheritance with varying penetrance (familial occurrence in > 50% of cases)

Secondary hypertrophic cardiomyopathy (e.g., in Friedreich ataxia, or amyloidosis)
Pressure related hypertrophy (some medical schools consider HTN one type of HCM though this is not agreede upon yet)
Chronic hypertension → increased afterload → increased myo cardial wall tension → sarcomeres laid down in parallel
→ increased left ventricular thickness → decreased left ventricular size → diastolic dysfunction.

Pathophysiology

Obstructive and nonobstructive types: hypertrophy of the left (possibly also the right) ventricle resulting in:

Reduced diastolic compliance and filling of the ventricle → reduced diastolic filling volume → reduced systolic output volume

Obstructive type (HOCM) also involvesLeft ventricular outflow tract obstruction (LVOT) due to interventricular septum hypertrophy and systolic anterior motion (SAM) of the mitral valve
Increasing mitral regurgitation

Obstruction exacerbated by
Positive inotropic drugs (e.g., digoxin)
Physical exercise/stress
Reduction of preload or afterload

Clinical features

Frequently asymptomatic (especially the nonobstructive type)

Obstructive type

(Exertional) dyspnea

Angina pectoris

Vertigo, **syncope**

Obstructive and nonobstructive type

Cardiac arrhythmias and palpitations

Sudden cardiac death (particularly during or after **intense physical activity**)

HOCM is an important cause of sudden cardiac death in young patients!

Auscultation findings

Systolic ejection murmur (crescendo-decrescendo)

Increases with valsalva maneuver and standing

Decreases with hand grip, squatting, or passive leg elevation

Apex (laterally displaced due to myocardial hypertrophy)

S4 gallop due to thickened, non-compliant ventricle

Possible murmur from mitral regurgitation

ECG

Signs of **left ventricular hypertrophy**

Left bundle branch block

Ventricular tachycardia or atrial fibrillation

Treatment

All patients should avoid strenuous exercise. Asymptomatic patients usually do not require further treatment.

Symptomatic patients

Beta blockers (1st line) or verapamil (2nd line).

Treat possible ventricular tachycardia or atrial fibrillation

If there is high risk of sudden cardiac death consider an **implantable cardioverter defibrillator (ICD)**

If symptoms persist after medical therapy:

Myectomy

Transcoronary ablation of septal hypertrophy (TASH)

Contraindicated drugs :
Positive inotropic and afterload-reducing or preload-reducing drugs (e.g., digitalis, glyceryl trinitrate, calcium channel blockers of the dihydropyridine class, ACE inhibitors) are contraindicated in cases of obstructive hypertrophic cardiomyopathy!

- Competitive sports and very strenuous activities should be proscribed
- dehydration should be avoided.

- B- blockers ameliorate angina
- Varapamil and diltiazem may reduce the stiffness of LV
- Amiodaron reduced risk of SCD and arrhythmia
- Digitalis,diuretics,nitrates, dihydropyridine calcium blockers, vasodilators and B- agonists are best avoided.

Again :

- Surgical myotomy / myectomy of septum and ethanol injections into the septal artey are the invasive treatment.
- ICD should be considered in high- risk patients

- Screening by echocardiography of first- degree relatives between the age 12 and 20 should be done every 12-24 months unless diagnosis evaluated by genetic testing

Restrictive cardiomyopthy

Epidemiology

Very rare

Pathophysiology

Proliferation of connective tissue → ↓ elasticity of myocardium → ↓ ventricular compliance → ↓ diastolic filling → atrial congestion → atrial enlargement and severe diastolic dysfunction

Etiology

Idiopathic

Secondary to

endomyocardial fibrosis (e.g., eosinophilic endocarditis, postradiation fibrosis, endocardial fibroelastosis) or systemic disease

(e.g., scleroderma, **amyloidosis**, sarcoidosis, hemoc hromatosis) :

- Etiology

- Amyloidosis,
  hemochromatosis,
  glycogen storage disease,
  endomyocardial fibrosis,
  sarcoidosis,
  hypereosinophilic disease,
  scleroderma,
  following mediastinal irradiation

Mnemonic for etiology of restrictive cardiomyopathy: **P**uppy **LEASH:** P = **P**ostradiation fibrosis, L = **L**öffler endocarditis, E = **E**ndocardial fibroelastosis, A = **A**myloidosis, S = **S**arcoidosis, H = **H**emochromatosis

Clinical findings

Symptoms of left-sided heart failure

Symptoms of right-sided heart failure

- Inability of the ventricles to fill, caused decrease cardiac out put
- exercise intolerance and dyspnea
- elevated systemic venous pressure cause edema

  ascitis and elevated JVP

- Hallmark of the RCM is abnormal diastole function.
- Myocardial fibrosis,hypertrophy or infiltration caused rigid LV walls

Treatment

Treatment of underlying condition (to avoid progression)

Symptomatic: beta blockers, cardioselective calcium channel blockers, diuretics

Anticoagulation and maintenance of a sinus rhythm in patients with a history of AF

	Dilated cardiomyopathy	Hypertrophic cardiomyopathy	Restrictive cardiomyopathy	Arrhythmogenic right ventricular cardiomyopathy
Pathophysiology	•↓Ventricular contractility d ue to dilation → •↓left ventricular ejection fraction (LVEF)	•Hypertrophy of the left ventricle → ↓ diastolic fillin gand systolic o utput • Left ventricular outflow obstruction in obstructive type	•Proliferation of connective tissue → atrial enlargement and severe diastolic dysfunction	•Right ventricular myocardial cell death → ventricular dilation, arrhy thmia and dysfunction

	Dilated cardiomyopathy	Hypertrophic cardiomyopathy	Restrictive cardiomyopathy	Arrhythmogenic right ventricular cardiomyopathy
Distinctive clinical features	•Signs of left heart failure and right heart failure	•Frequently asymptomatic •Dyspnea, syncope, vertigo •Arrhythmias •Sudden death	•Signs of left heart failure and right heart failure	•Very variable •Exercise-induced ventricular tachycardia

	Dilated cardiomyopathy	Hypertrophic cardiomyopathy	Restrictive cardiomyopathy	Arrhythmogenic right ventricular cardiomyopathy
Other characteristics	•Most common cardiomyopathy	•Most common cause of sudden heart failure in athletes and teenagers	•Poor prognosis without heart transplant	•Most commonly in young adults