

CARDIOMYOPATHY

DEFINITION

Cardiomyopathy is a term which indicates **disease of cardiac muscle** of unknown cause

It is **classified on clinical presentation** into :-

- **DCM** - Dilated cardiomyopathy
- **HCM**- Hypertrophic cardiomyopathy
- **Restrictive** cardiomyopathy
- **Arrhythmogenic** – RV- cardiomyopathy
- **OTHER** rare cardiomyopathy-conduction defect- mitochondrial- LQTS- BRUGADA

DILATED CARDIOMYOPATHY

- **DCM** is characterized by **dilatation** and **impaired systolic function** of the left and /or right ventricle, in the absence of underlying disease e.g., IHD- hypertension, valve disease
- The **cause** in the majority is **unknown (idiopathic)**
- At least 25% of the idiopathic are familial; **more than 20 abnormal chromosome loci and genes are involved**
- The **majority** are inherited as **autosomal dominant** , but X-linked or recessive cases can be found .

CAUSES OF SECONDARY CARDIOMYOPATHY

■ INFECTION:

- **VIRAL**- Influenza, Corona, Coxsackie, Adeno erythro-viruses, and HIV
- **BACTERIAL**- Diphtheria
- **FUNGAL INFECTION, PARASITES**- Toxoplasma, Trypanosome cruzi, Chagas disease- Spirochaete- Lyme disease

■ TOXINES: Alcohol, Metal (Cobalt, Lead, Mercury Arsenic) and Chemicals

■ DRUGS: Chemotherapy, Cocaine

■ AUTOIMMUNE: SLE, dermatomyositis, systemic sclerosis Metabolic- CKD - Cardiorenal SYN

CAUSES OF SECONDARY CARDIOMYOPATHY

- **ENDOCRINE** :- DM, Thyroid heart disease hyperparathyroidism, acromegaly and pheochromocytoma
- **NEUROMUSCULAR** :- Friedrich`s ataxia, muscular dystrophy , myotonic dystrophy , neurofibromatosis
- **NUTRITIONAL DEFICIENCY**: e.g., Beri-beri, Pellagra, Scurvy, Kwashiorkor
- **INFLAMMATORY** (granulomatous) e.g., Sarcoidosis
- **INFILTRATIVE**: e.g., Amyloidosis, Gaucher disease, Hurler`s disease and Hunter disease
- **STORAGE**: Hemochromatosis, Glycogen-storage disease type11
- **OTHER**: Tachycardia cardiomyopathy- Peripartum cardiomyopathy

CLINICAL FEATURES

- Congestive heart failure , **in addition** they may present with:
- Syncope, Arrhythmia, Conduction defect, Heart block
- Systemic embolism –CVA, Pulmonary embolism - Sudden Death

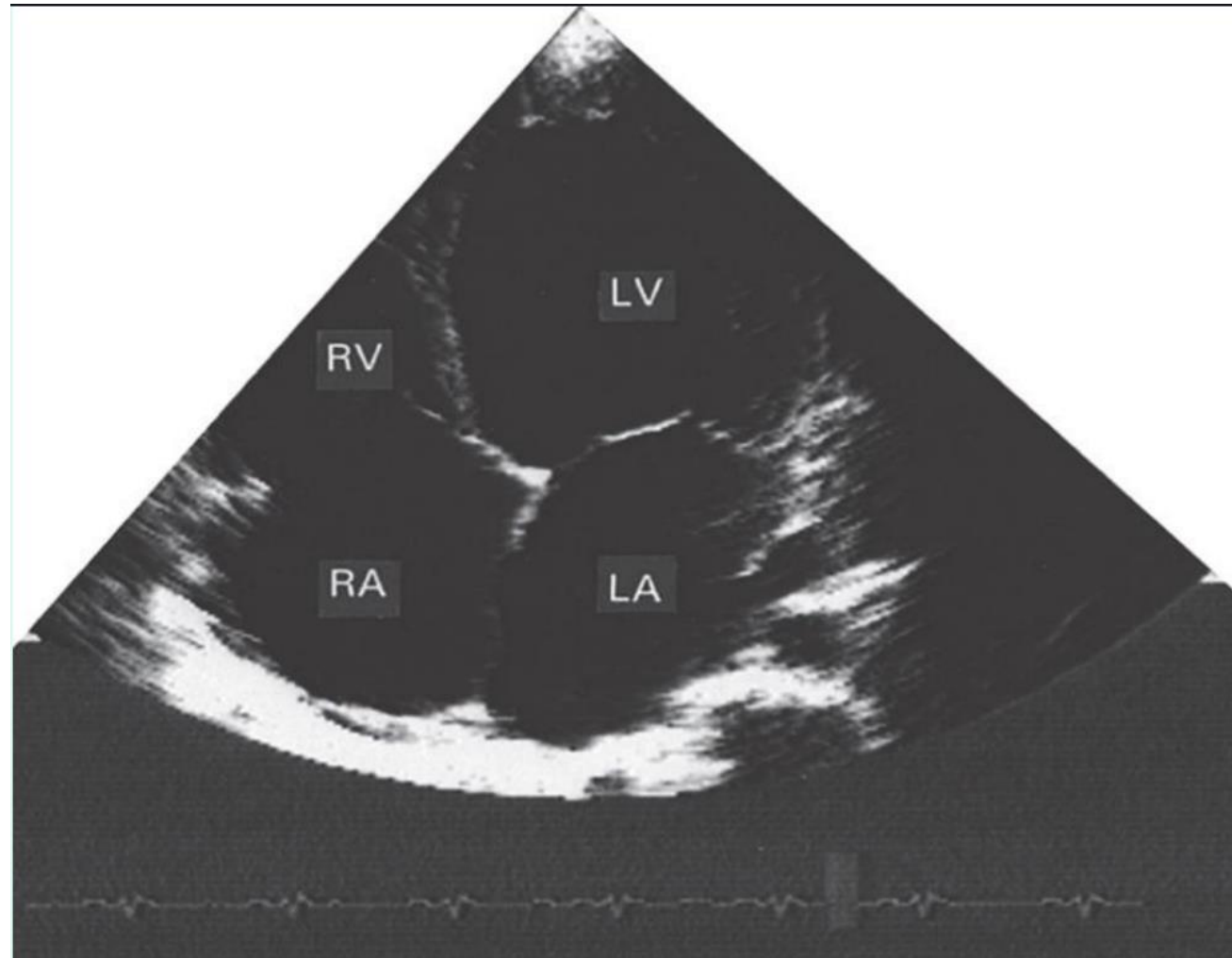
INVESTIGATION

- Chest X-ray shows **enlarged cardiac shadow**
- ECG shows **diffuse non-specific ST segment and T wave changes**
- Also, may show **sinus tachycardia, conduction defects, and arrhythmia**
(i.e., **atrial fibrillation, ventricular premature beats or ventricular tachycardia**)

- Echocardiography reveals **dilatation** of the left and /or right ventricle
- Angiography –CATH- should be done to **exclude coronary artery disease**
- Biopsy

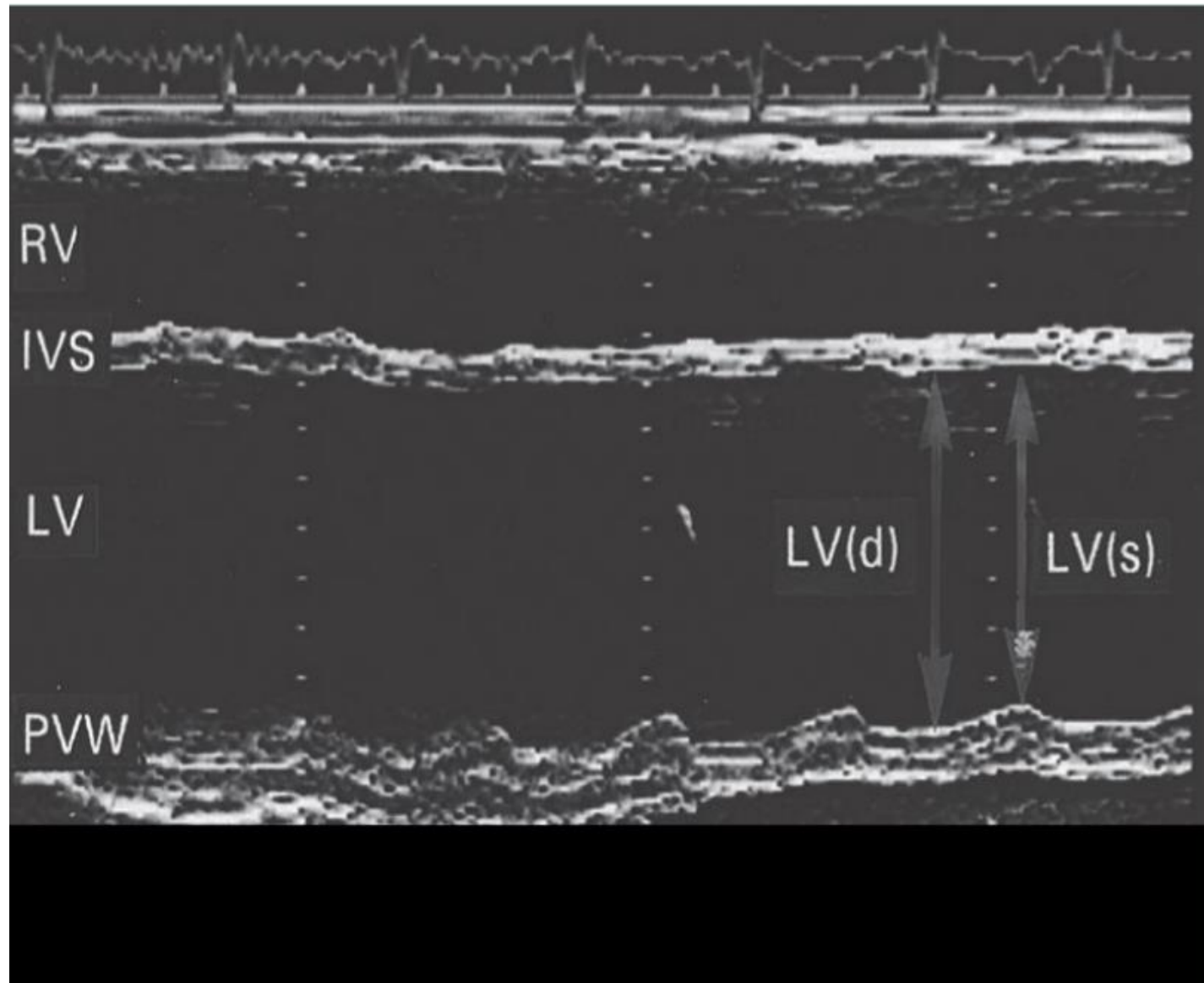
TREATMENT

- **The aim** is to relieve symptoms, retard disease progression prevent complications of heart failure
- ACE- inhibitors, **ARB, ARNI , SGT12**
- Spironolactone- MRA- **Ivabradine- BB**
- **Antiarrhythmic** therapy
- **Pace maker**- CRT- ICD - **Anticoagulant** therapy
- LV-RV- mechanical assistant device
- Cardiac transplantation



(a)

Kumar & Clark: Kumar and Clark's Clinical Medicine, 7th Edition.
Copyright © 2009 by Saunders, an imprint of Elsevier, Ltd. All rights reserved.



HYPERTROPHIC CARDIOMYOPATHY (HOCM) (HCM)

- This is **characterized by** variable myocardial hypertrophy **mostly involving inter-ventricular septum** with disorganization (disarray) of cardiac myocytes and myofibrils
- Absence of clear causes like - AS- HTN
- The **majority of cases** are **familial/autosomal dominant** mutation in genes encoding sarcomeric proteins

CLINICAL FEATURES

■ They usually presents with:

- Asymptomatic
- Chest pain
- Dyspnea
- Syncope
- Cardiac arrhythmia
- SUDDEN DEATH

■ More common in:

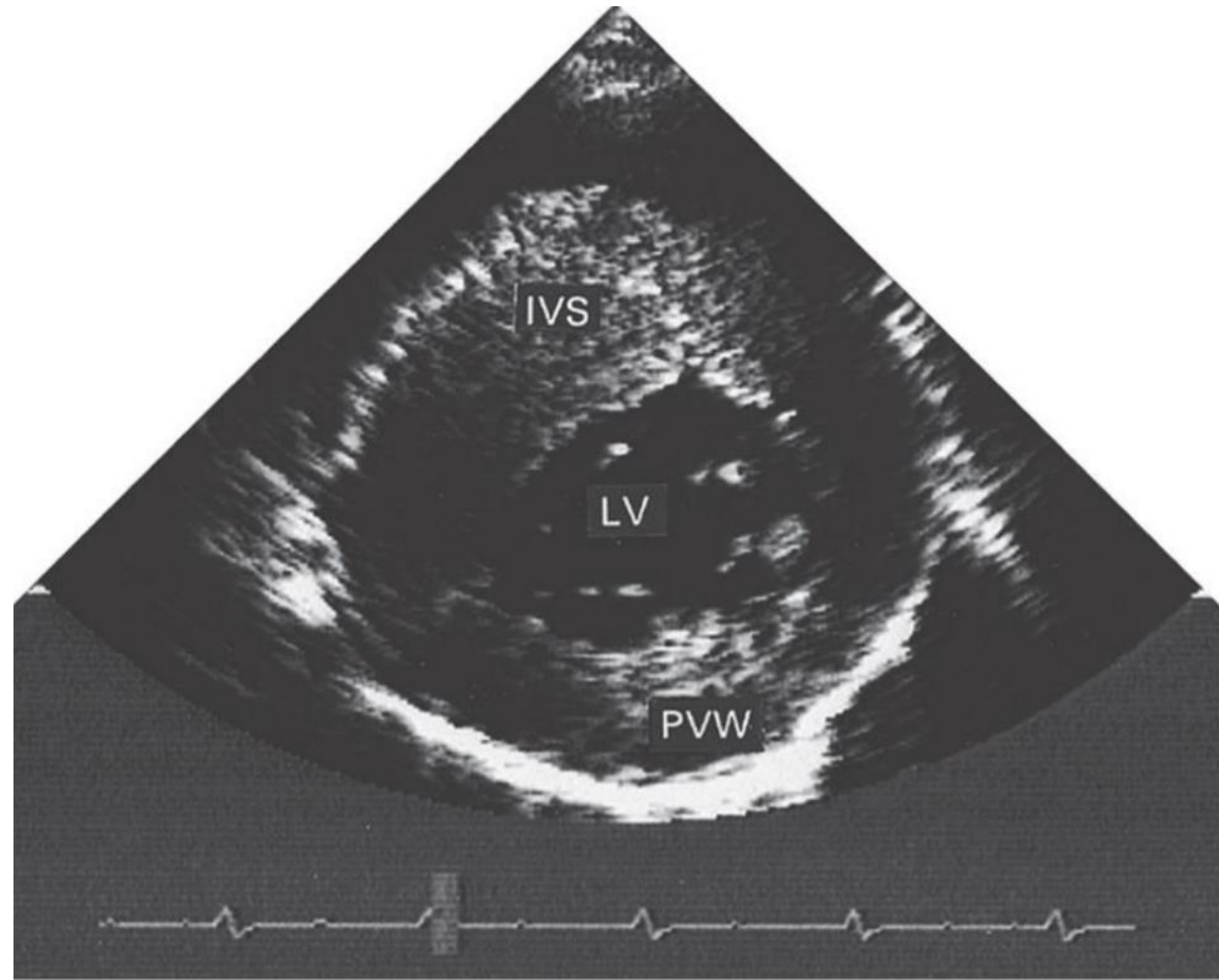
- 1- >3 CM thickness of IVS
- 2- Syncope
- 3- Arrhythmia
- 4- Young people < 30 years
- 5- Family history of sudden death

■ The classic physical finding are:

- Double apical pulsation
- Jerky carotid pulse
- Fourth heart sound
- Ejection systolic murmur
- Rapid ejection and sudden LV out flow obstruction
- Pansystolic murmur of mitral regurgitation

INVESTIGATION

- CXR- is usually **unremarkable**
 - ECG- shows **left ventricular hypertrophy**, ST and T wave changes and Q wave in inferolateral leads
 - Echocardiography - **diagnostic** it shows – THICK –IVS
 - SAM-systolic anterior motion of **anterior mitral valve leaflet**
- asymmetric left ventricular hypertrophy
- HIGH PRESSURE GRADIENT ACROSS LV- outflow tract**
- Pedigree analysis - Exercise testing



(a)

TREATMENT

- Chest pain and dyspnea by
 - Beta-blockers and Verapamil
- Pre-syncope & syncope- Arrhythmia: Antiarrhythmic – Amiodaron- Flecainide- Propafenon

INTERVENTION

- ICD- prevent sudden death - Dual- chamber pacing
- ALCOHOL ABLATION injection IN COR. Induce MI- to reduce IVS
- Surgical procedure - relieve obstruction
- Vasodilators should be avoided

RESTRICTIVE CARDIOMYOPATHY

- There is **no ventricular dilatation or muscular hypertrophy** but there is **resistance to DIASTOLIC-ventricular filling** resulting HF
- **SECONDARY CAUSES:** Amyloidosis, Sarcoidosis, Loeffler`s endocarditis ,
Endomyocardial fibrosis

CLINICAL PRESENTATION:

Dyspnea, fatigue, and Thrombo-embolic, Anasarca, Elevated venous pressure, hepatomegaly, ascites and general edema

THE SIGNS ARE SIMILAR TO CHRONIC CONSTRICTIVE PERICARDITIS

INVESTIGATION

- Chest X-ray, ECG, Echocardiogram
- Cardiac catheterizations used for distinction from Ch. constrictive pericarditis

TREATMENT

- No specific treatment
- It is only symptomatic
- Cardiac transplantation - in some cases
- In amyloidosis melphalan plus prednisolone with or without colchicine may be used