Cardiomyopathy

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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

- Arrythmogenic – 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- coxackie, adeno -

erythro-viruses, and HIV -

BACTERIA- diphtheria, or fungal infection, -

PARASITES, -toxoplasma, trypanosome cruzi, -

chagas disease- sptirochaete- lyme disease

Toxines :- alcohol, metal (cobalt, lead, mercury - arsenic) and chemicals

- Drugs :- chemotherapy, cocaine –

Autoimmune :- SLE, dermatomyositis, systemic sclerosis

Metabolic- CKD - CARDIORENAL SYN.

- Endocrine :- DM, Thyroid heart disease hyperparathyroidism, acromegaly and phaochromocytoma

Neuromuscular :- Friedrich`s ataxia, -

muscular dystrophy , myotonic dystorphy , - neurofibromatosis

Causes cont

- Nutritional deficiency :- e.g. beriberi, pellagra, scurvy, kwashiorkor
- Inflamatory (granulomatous) e.g. sarcoidosos
- Infiltrative :- e.g. amyloidosis, Gausher disease, Hurler`s disease and Hunter disease

Storage :- haemachromatosis, - glycogen-storage disease type11 -

OTHER-

Tachycardia cardiomyopathy-

peripartum cardiomyopathy

Clinical features

congestive heart failure ,
in addition they may present with
syncope, arryhthmia, 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**

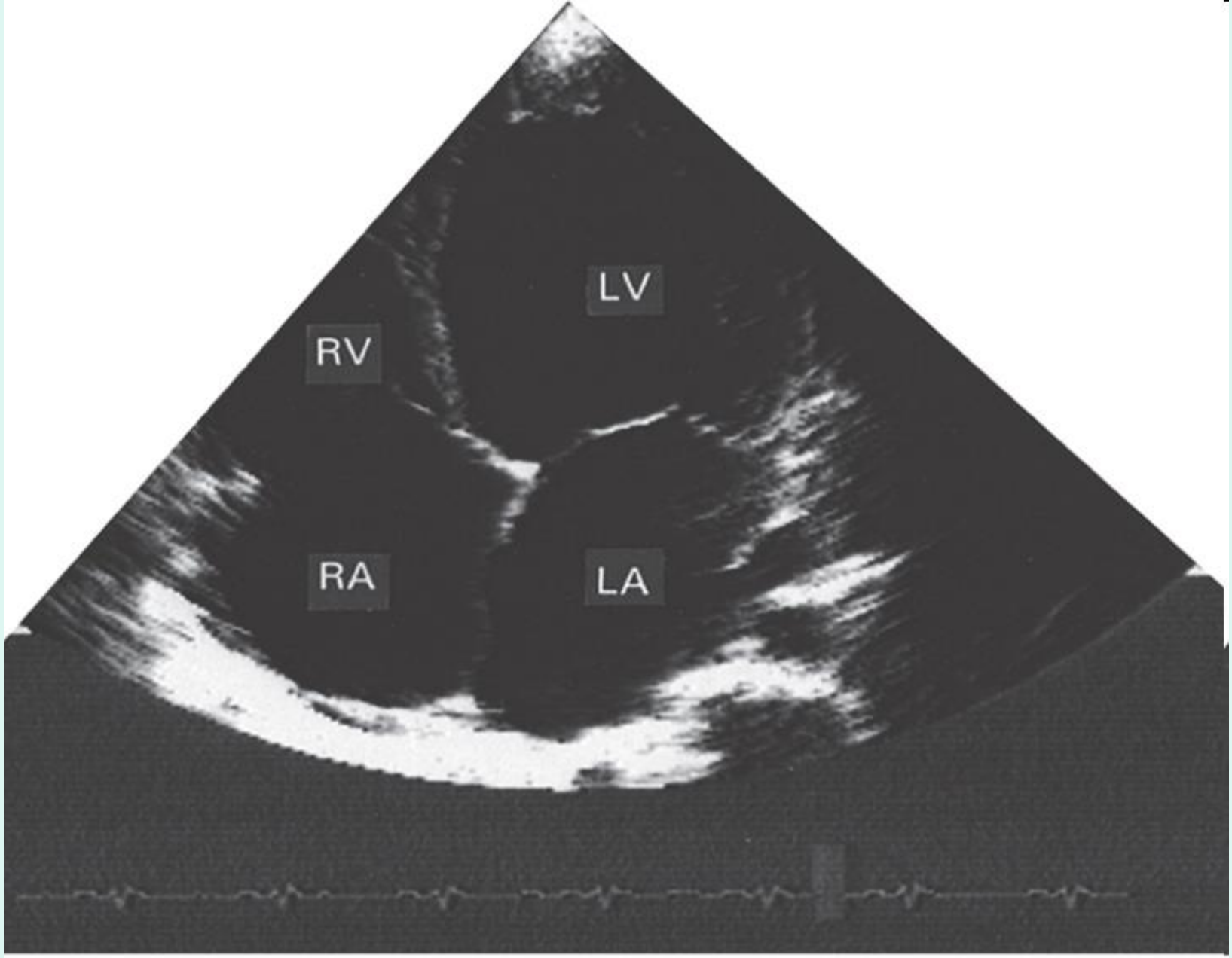
-Angiography –CATH- **should be done to exclude coronary artery disease**

- Biopsy

Treatment :- the aim is

to relieve symptoms,
retard disease progression
prevent complications of heart failure

- ACE- inhibitors, ARB- ARNI , SGTI2
-- Spironolactone- MRA- ivabradin- beta- blocker
- Anti-arrhythmic therapy or
- Pace maker- CRT- ICD - anticoagulant therapy
- LV-RV- mechanical assistant device
cardiac transplantation



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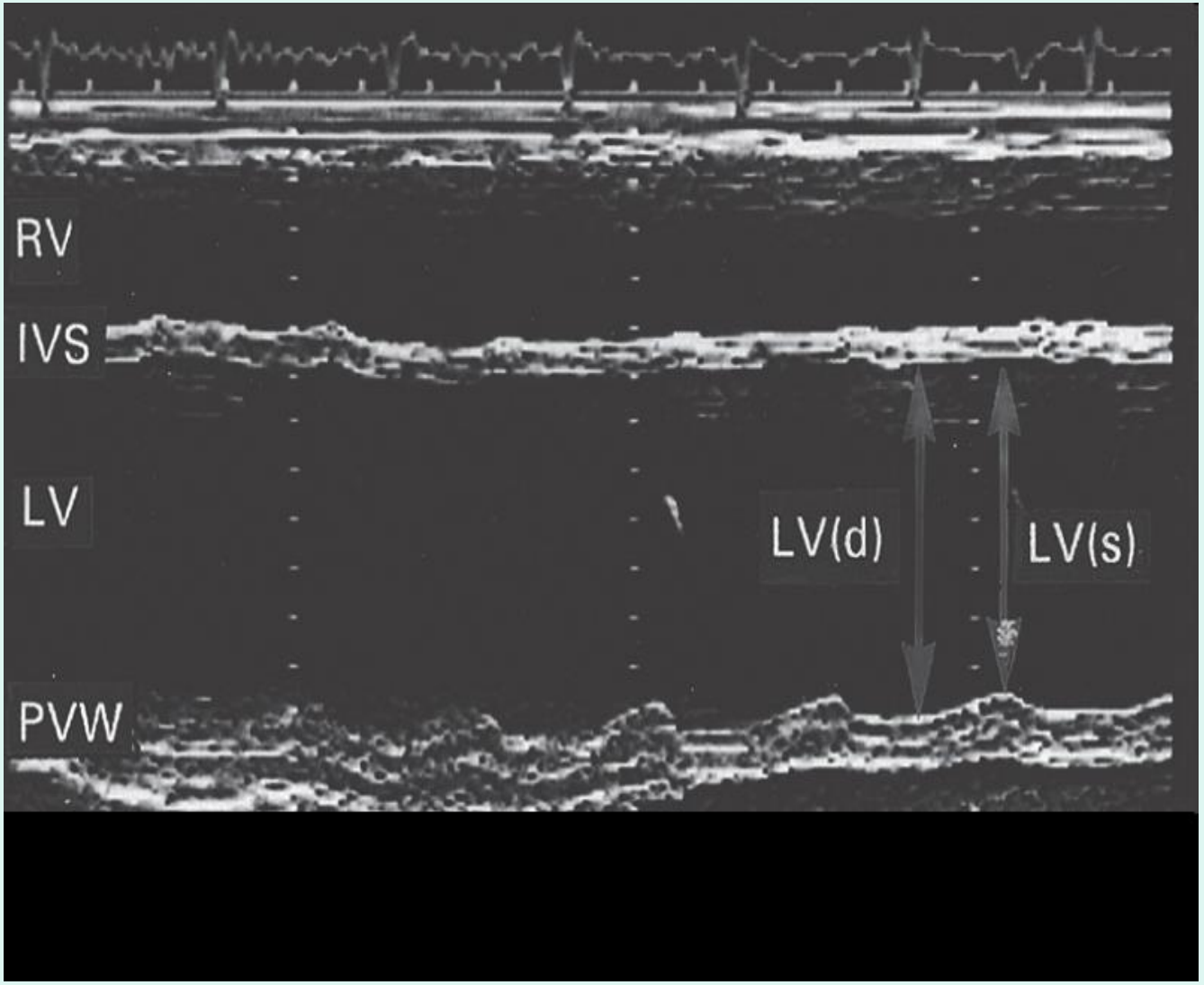

RV
IVS
LV
LV(d)
LV(s)
PVW

Hypertrophic cardimyopathy(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 protiens

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 <30YEAR

5-FAMILY HISTORY OF SUDDEN DEATH

The classic physical finding are

- double apical pulsation

jerky carotid pulse- -

rapid ejection and sudden LV out flow obstruction -

- fourth heart sound

- ejection systolic murmur

- pansystolic murmur of mitral regurgitation

Investigation

- Chest X- ray- **is usually unremarkable**

- ECG- **shows left ventricular hypertrophy, ST and T wave changes and Q wave in inferolateral leads**

Echocardiography **- diagnostic it shows –** -

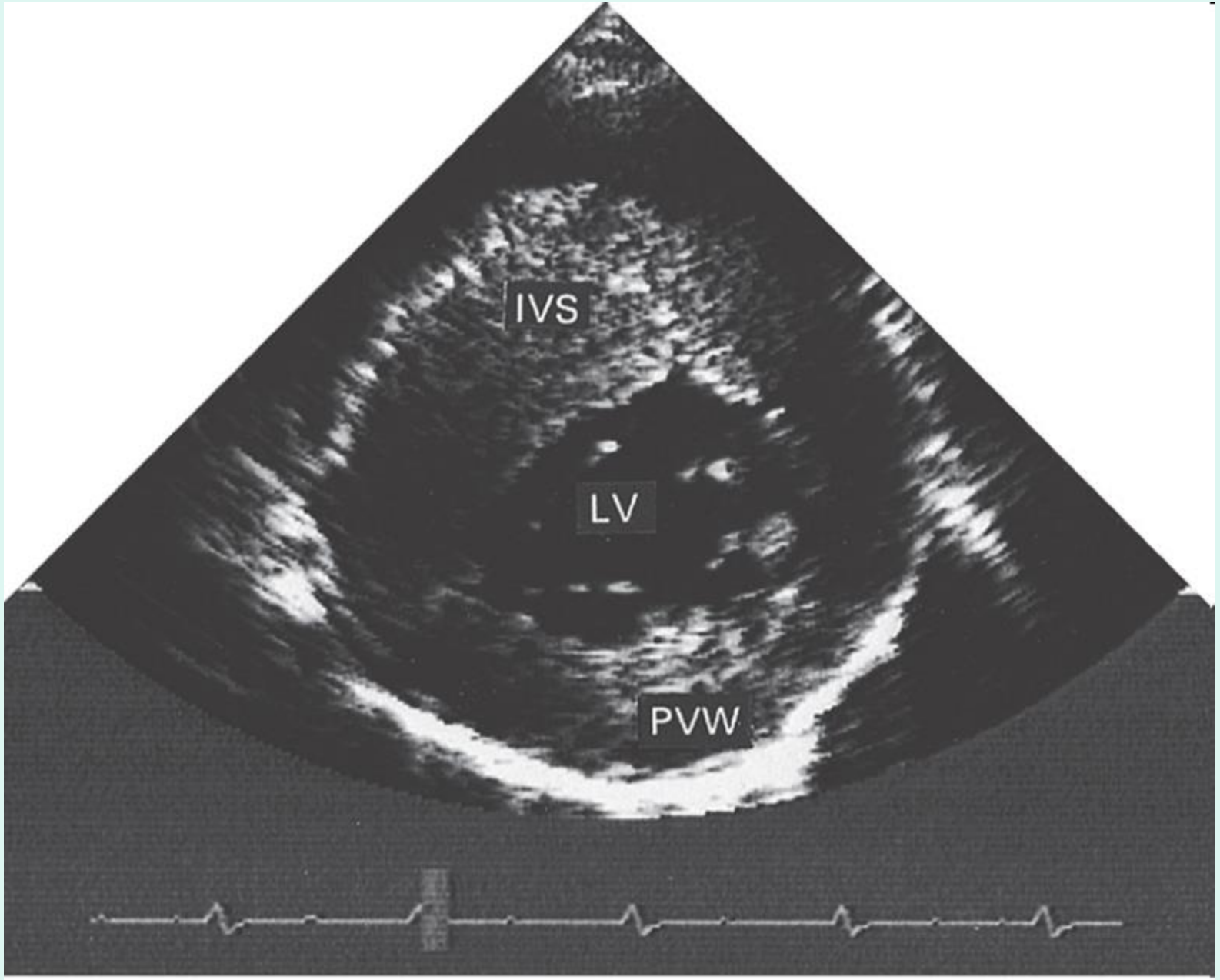
THICK –IVS-

SAM-systolic anterior motion of anterior mitral valve leaflet

- asymmetric left ventricular hypertrophy-

HIGH PRESSURE GRADIENT ACROSS LV-out flow tract

-- Pedigree analysis - Exercise testing



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Treatment

Chest pain and dyspnea -

treated beta-blockers and verapamil , -
pre-syncopy- and syncopy- arryhythmia-

anti-arryhthmic – -

AMOIDERON- FLECANIDE-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 cardimyopathy

There is no ventricular dilatation or muscular hypertrophy but there is resistance to DIASTOIC-ventricular filling resulting - HF.

SECONDARY Causes :-

amyloidosis , sarcoidsis, Loeffler`s endocarditis , endo myocardial fibrosis .

Clinical PRESENTATION-

Dyspnea, fatigue, and THROMBO- embolic

ANASRCA- elevated venous pressure, hepatomegaly, ascitis and general edema , the signs are similar to CHRONIC CONSTRICTIVE PERICARDITIS

Investigation :-

Chest X-ray, ECG, Echocardiogam

Cardiac catheterization

is 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