

MYOCARDIAL DISEASES

MYOCARDITIS

Disease of the myocardium - Inflammatory
Infiltrate

Damage to myocytes

focal/diffuse

Unknown, asymptomatic, resolves in time.

Causes of myocarditis

Idiopathic

- Giant cell Myocarditis

Infectious

- Viral
- Rickettsial
- Bacterial
- Fungal
- Parasitic

Non - infectious:

- Hypersensitivity/ Imm - med
- Drugs & Toxins
- Radiation
- Others

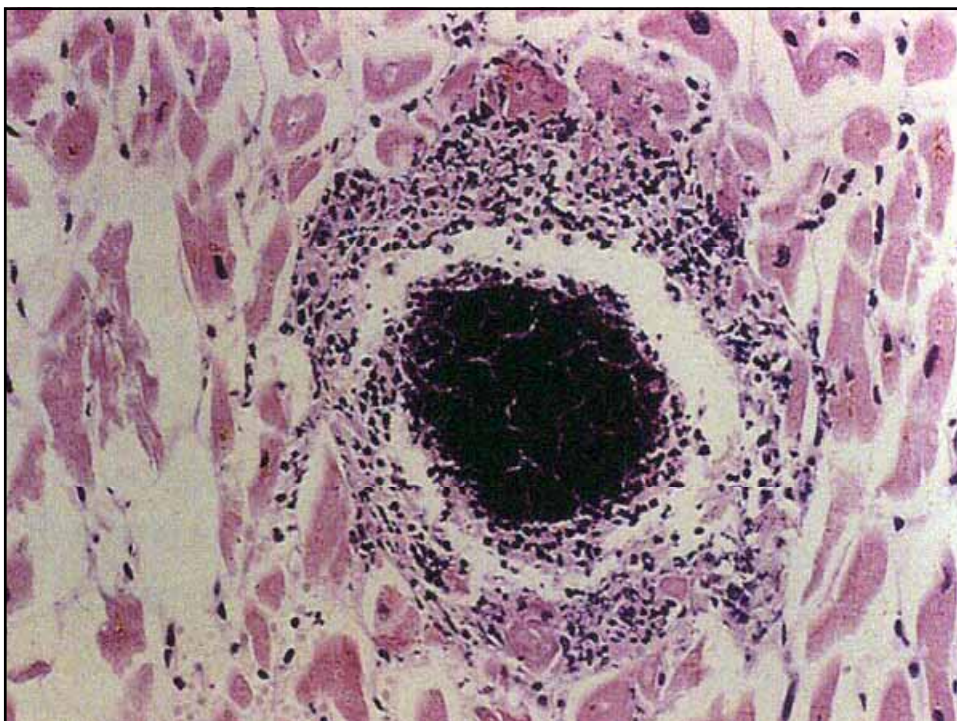
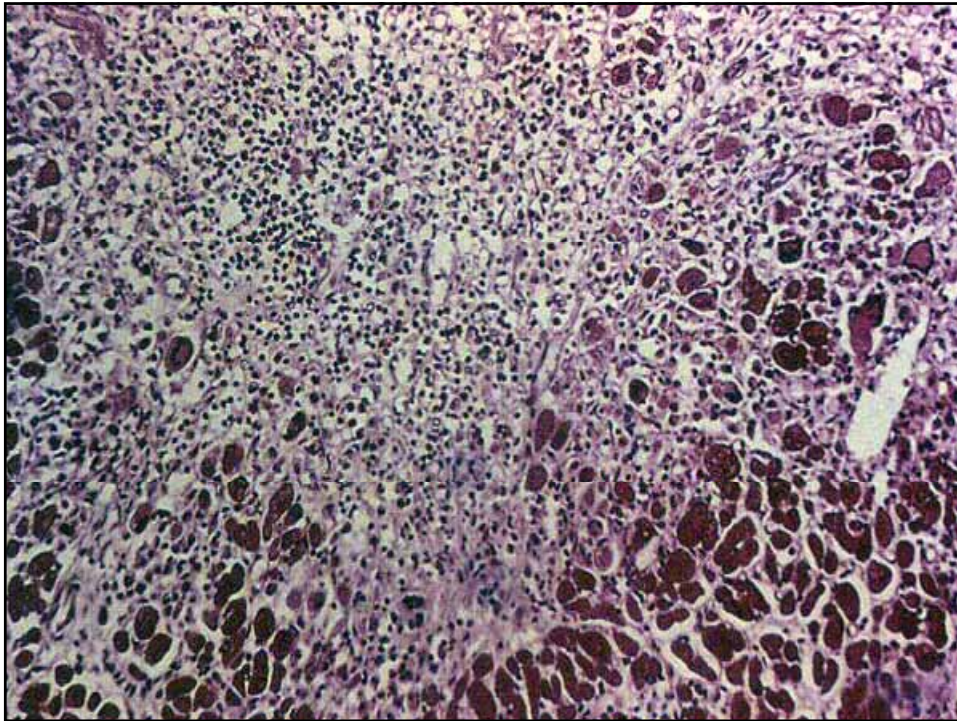
Clinical Features

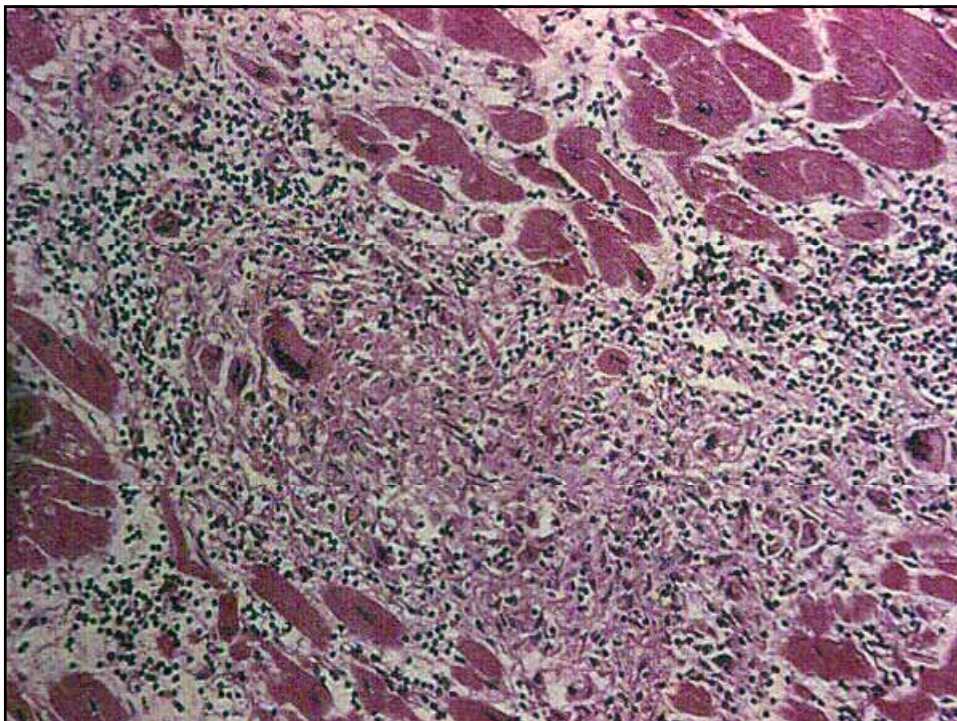
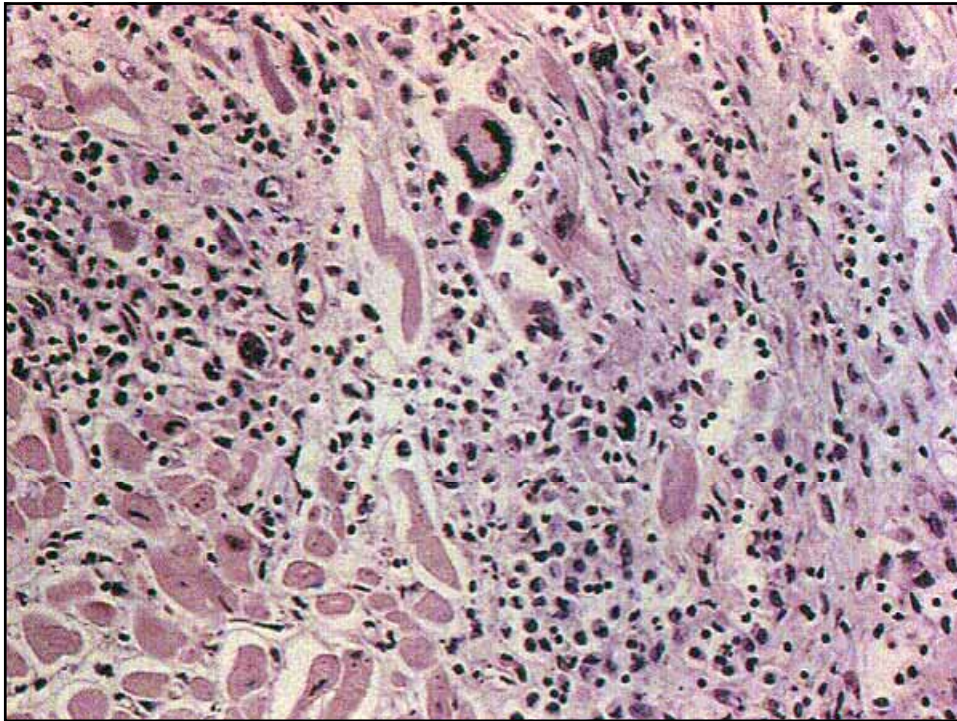
- Asymptomatic - fulminant fatal course
fever, dyspnea, fatigue, palpitation, CHF
- Enzyme & EKG changes are seen

Morphology

Gross: normal /Dilatation of ventricles
- May mimic MI

Micro: -Infiltrates → Ly, Mo, Neu & Eos
Granuloma & giant cells
Interstitial Edema
Myofiber Necrosis .





DEFINITIVE DIAGNOSIS

Endomyocardial biopsy & Histology

Recombinant DNA Tech. -Viral genome

CARDIOMYOPATHY (C.M.P.)

Disease of the Heart Muscle (WHO/IFOC)

**Heart Muscle Diseases of unknown cause
excluding those due to Cardiac Valves,
Coronary Artery Disease, Cong. H. D.
Pericardial Disease, etc.**

- **Non inflammatory, non ischemic, non
hypertensive cardiac muscle disease**

Primary/Idiopathic Cardiomyopathy

Profound and Fatal C. H. F

Three categories of CMP: based on
Structure & Patho-physiology

Dilated - most common

Hypertrophic

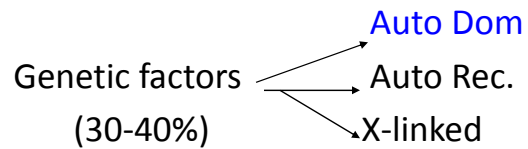
Restrictive - least common

New entity: Arrhythmogenic RV CMP/dysplasia

DILATED CARDIOMYOPATHY (DCM)

- **Gradual development of cardiac failure associated with four-chamber hypertrophy and dilatation of heart of unknown cause.**

Aetio pathogenesis



Suspected underlying mechanisms

Alcohol/toxic agent (Adriamycin)

Inflammatory - Viral/Non-viral myocarditis

Pregnancy associated

Nutritional deficiency - Beri Beri

Immunologic

Mutations: sarcomere, Cytoskeleton, Nuclear env, Mito

MORPHOLOGY

Gross: Heart - Heavy - 1 1/2 - 2 N Weight

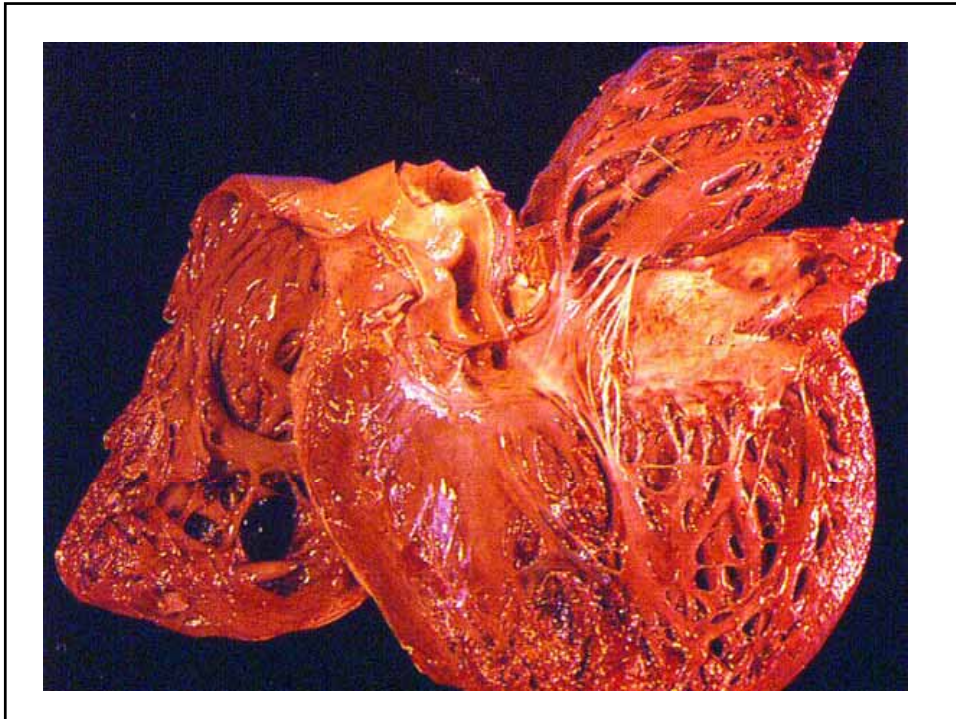
Large and flabby. All chambers are dilated

Dilatation - Ventricular wall - Thin of Thick

Mural Thrombi → Thromboembolism

M. V. R. - Valve ring dilatation

Coronaries - N or obstruction → healed ischaemic lesions



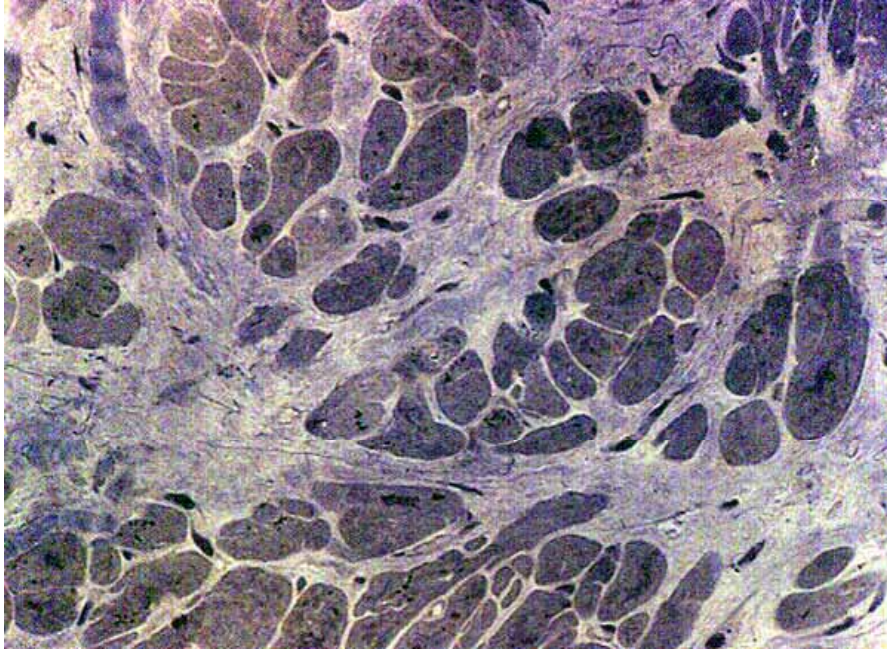
MORPHOLOGY

Micro:- Variation in myocyte size -
Hypertrophic, or attenuated

Nuclei - large, Interstitial & Endocardial
Fibrosis

Final Abnormality:

Flabby hypocontracting heart with →systolic
failure



Clinical Features & Treatment

- Any age (20 - 60 yr.), Dyspnoea, chest pain, fatigue, palpitation, features of CHF - progressive and unremitting
Rx: Cardiac transplantation

HYPERTROPHIC CARDIOMYOPATHY

- Idiopathic Hypertrophic Subaortic Stenosis/Hypertrophic obstructive CMP
- Ventricular muscle thickening
- Irregular hypertrophy
- Myocyte disarray

Important Genes responsible for this familial disease

Genes are: Chr. 14 - β -heavy chain myosin

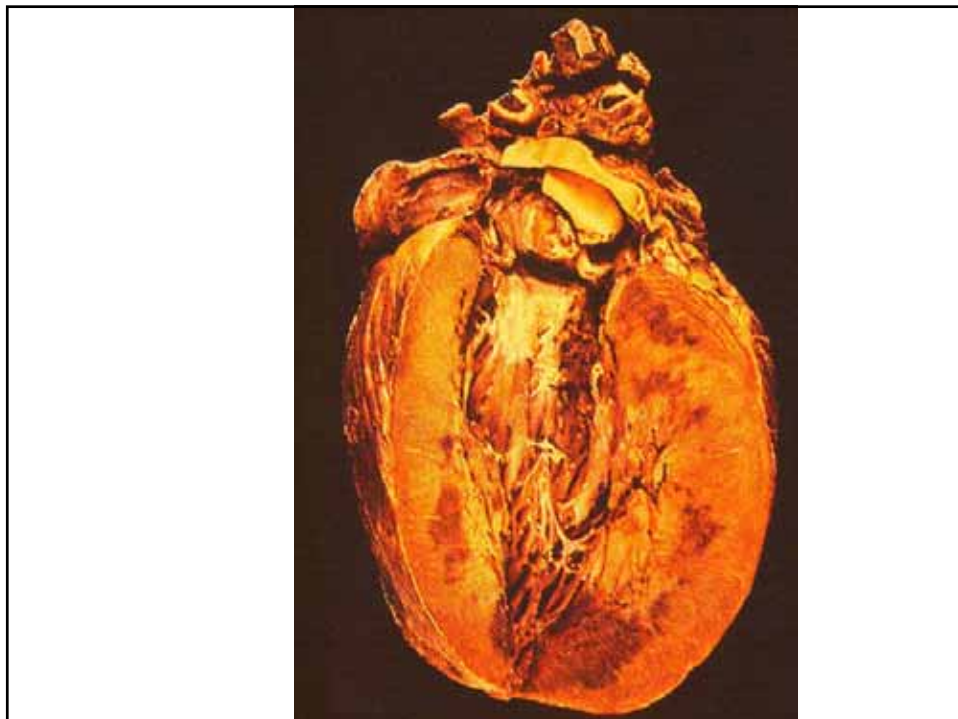
Chr. 1 - Troponin - T

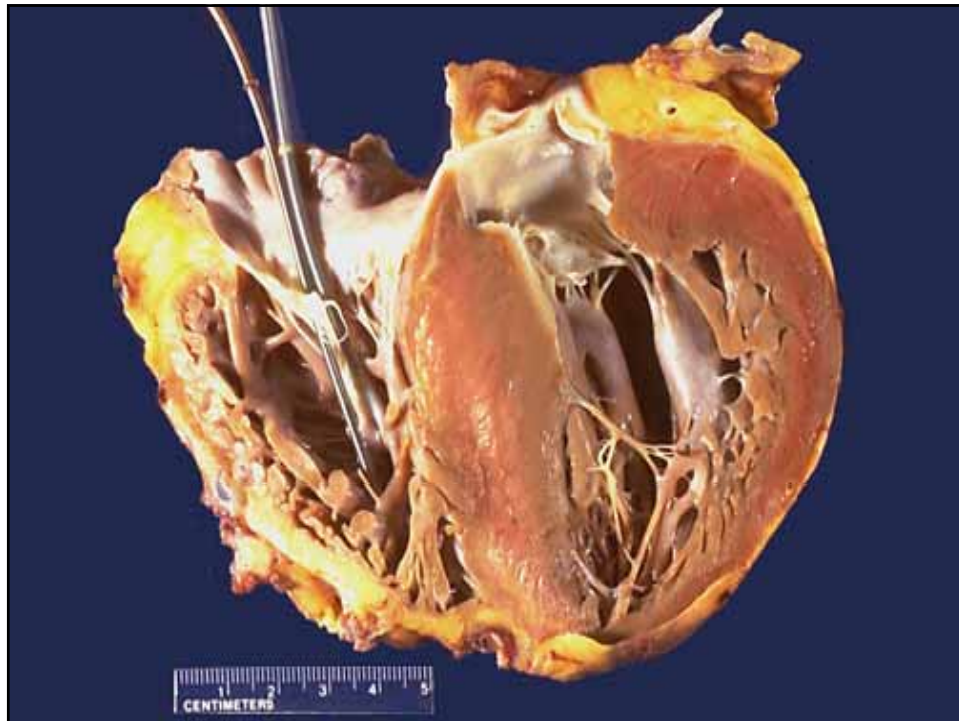
Chr. 15 - α - Tropomyosin

Responsible for abnormal myofibril formation and alignment

MORPHOLOGY

- Massive myocardial hypertrophy - ↑wt.
- Asymmetric septal hypertrophy - classic
- Ventricular cavity - ' Banana Shaped '
- Anterior mitral valve leaflet thickened
- L. V. outflow narrowed, endocardial thickening



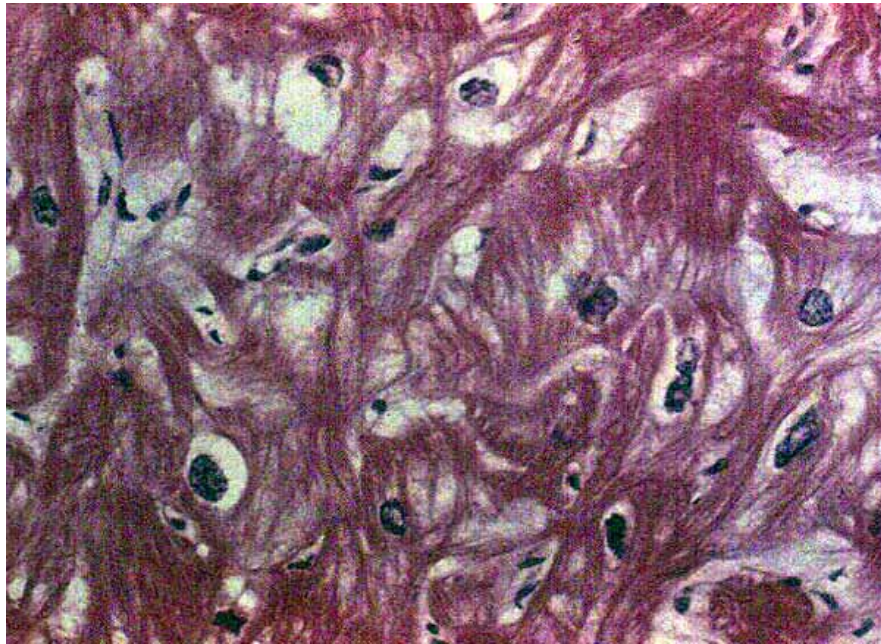


MORPHOLOGY

Micro:

Extensive myocyte hypertrophy
Disarray of bundles, myocytes and myofibres
Interstitial fibrosis
Small vessel disease

Functional Abnormality: - Heavy muscular
hypercontracting heart with diastolic
dysfunction



CLINICAL FEATURES

- Palpitation, dyspnoea, cardiac murmur, and sudden death in young males

TREATMENT : Septal surgery

RESTRICTIVE CARDIOMYOPATHY

Diastolic relaxation and L. V. chamber filling is impeded

Morphology: Gross:

Ventricles - N or slightly enlarged, cavity N

Myocardium – Firm, Biatrial dilatation

Micro: Patchy/diffuse Interstitial fibrosis

Functional Abnormality:

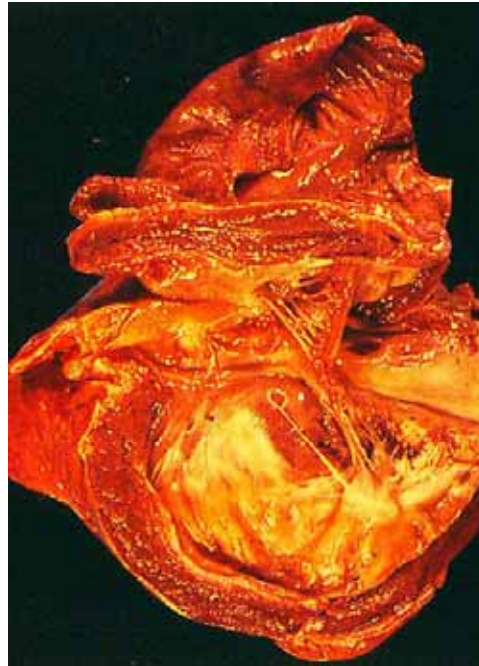
Diastolic function impaired, systolic - N or impaired. Variants/subtypes:-

ENDOMYOCARDIAL FIBROSIS

Unknown cause

Children, young adults - Africa and Tropics

- Fibrosis of ventricular endo/ Subendo, Apex & Inflow tract
 - Reduced compliance of chamber
 - Mural thrombi



Leoffler's Endomyocarditis

- Endomyocardial fibrosis & mural thrombi
- Eosinophilic infiltration - release toxic products - Major Basic Protein
- Myocardial damage
- Rx Endomyocardial stripping

Endocardial fibroelastosis

uncommon, unknown cause

Age: 1-2years

Focal/diffuse cartilage like fibroelastic thickening

Assoc. with Cong. Cardiac anomalies

- CCF

LAB DIAGNOSIS

X-ray, ECG, ECHO, Cardiac Cath & Endomyocardial Biopsy

Endomyocardial Biopsy:

Biopsy → **(R) Internal Jugular Vein (R)** →
Heart → **R. V. Septum - Snip (1-3mm)**

Study → **Virology**

E.M.

Imm. Histo.

Biochemical

Molecular

COMPLICATIONS OF CMP

Heart failure

Cardiac arrhythmia's

Atrial

Ventricular

Mural thrombi and embolism

Endocarditis and sudden death - HCM

SUMMARY

	<u>DCM</u>	<u>HCM</u>	<u>RCM</u>
L.V. Cavity	Dilated	Small	Normal
L.V. Wall Thickness	N, ↑ ↓	↑	N or ↑
Systolic Function	↓	↑ or N	N or ↓
Diastolic Function	N	↓	↓
Septal Asymmetry	-	+	-