CARDIOMYOPATHY:

→ They are heterogeneous group of diseases of Myocardium Associated with Mechanical or electrical dysfunction that usually exhibit inappropriate ventricular hypertrophy.

CLASSIFICATION:

Based on etiology: 1° / 2nd

⇒ Primary cardiomyopathy: confined to myocardium genetic or acquired
⇒ 2ndary cardiomyopathy: multiorgan disorders → Inflammatory disorder, Immunological disorder, Muscular dystrophies.

Based on clinical, Functional & Pathological Patterns:

1) Dilated cardiomyopathy: ~90% (Most common)
2) Hypertrophic cardiomyopathy (HCM)
3) Restrictive cardiomyopathy (least common)

DILATED CARDIOMYOPATHY:

DCM is characterized morphologically by progressive cardiac dilation & functionally by contractile dysfunction (systolic)

GENETIC CAUSES →
→ DCM is hereditary in About 20-50% cases
→ Autosomal dominant pattern.
↓
caused by loss of function mutation of cytoskeletal protein or protein that links the sarcomere to the cytoskeleton.

X-linked DCM is most frequently associated with mutation in dystrophin.

NON-GENTIC CAUSES:

→ Infections: Adenovirus, parvovirus B-19, human herpesvirus 6.
→ Alcohol or exposure to other toxins → Alchol Abuse, DCM
→ Cheonic Alcohol use can produce thiamine deficiency of can Result in beri-beri heart-disease.
→ Pregnancy
→ Iron-overload:

MORHOLOGY:

→ Heart: enlarged, heavy (2-3 times the n.w), flabby with dilation of all chambers
→ Thickness of dilated ventricles: (less then / equal to or great than normal)
→ Mural theombi ⇒ Apex of heart.
→ Absence of secondary cause of cardiac dilation
→ Microscopy:
Hypertrophy of myocytes → hypertrophy with enlarged nuclei.
→ Interstitial & endocardial fibrosis → Scattered Area of Replacement fibrosis; myocytic ischemia nenosis due to hypoperfusion.
→ DCM secondary to iron overload:
Accumulation of intramyocardial hemosiderin
This pigment can be demonstrated by staining with Prussian blue

At end stage → defect in DCM is ineffective contraction.
cardiac ejection is less than 25%
muffled heart sound
↳ Beck's Triad

DIAGNOSIS
→ ECHO
→ chest X Ray
→ t/t cardiac transplantation.

NON - HODGKIN'S LYMPHOMA

HYPERTROPHIC CARDIOMYOPATHY:

genetic disorder characterized by inappropriate myocardial hypertrophy & defective diastolic filling.

→ obstructive ventricular outflow obstruction.

→ the heart in HCM is thick-walled, heavy & hypercontractile

→ myocardium donot Relax & show primary diastolic dysfunction

CAUSES

GENETIC: mutated sarcomere protein include
- β myosin heavy chain
- myosin binding protein C
- Troponin T.

MORPHOLOGY:

Heart is Always enlarged & show massive myocardial hypertrophy without dialated of ventricle dilation.

→ Asymmetric septal hypertrophy: classical features is disproportion thickening of the ventricular septum.
(Ratio of septum to free wall greater than 3:1)
most marked in the subaortic Region.

→ left ventricular wall is thick, cavity is small
→ banana-like (sometime only slit configuration)

→ endocardial mural plaque → Thickned hypertrophied subaortic septum bulges

MICROSCOPY:

- Marked hypertrophy of myocytes.

Myofibres disarray →

Haphazard myocyte & myofibres disarray.

Normally the myocytes are arranged in to muscle bundles.
In disarray orientation of adjacent hypertrophic myocytes.