

Cardiomyopathy

Blackpool Victoria Hospital

Definition

- Disorders of the cardiac muscle of unknown aetiology

Specific types

- Infective/viral
- Metabolic/infiltrative (cardiac amyloidosis)
- Systemic/multistystem disease
- Familial neuro-muscular disorders eg muscular dystrophy
- Sensitivity or toxic reactions eg. Alcohol (dilated) and chemotherapy

Infective

- Viral eg. Coxsackie virus (dilated)
- Tuberculosis (restrictive)
- HIV/AIDS (dilated)
- Chagas disease (restrictive) - South America
- hypereosinophilic syndrome - could be parasitic, mostly unknown (restrictive - iron deposits)

Metabolic diseases

- Primary cardiac amyloidosis - restrictive (protein deposits)
- cardiac sarcoidosis - cardiac granulomas
- hemochromatosis

Classification

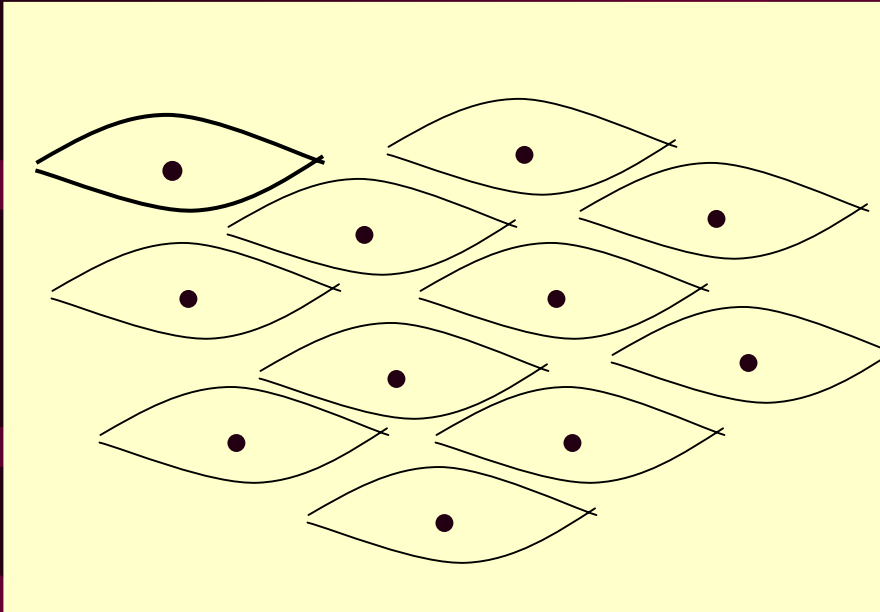
- Hypertrophic
 - HCM
 - HOCM
 - Obliterative
- Dilated (Congestive)
- Restrictive

Hypertrophic Cardiomyopathy

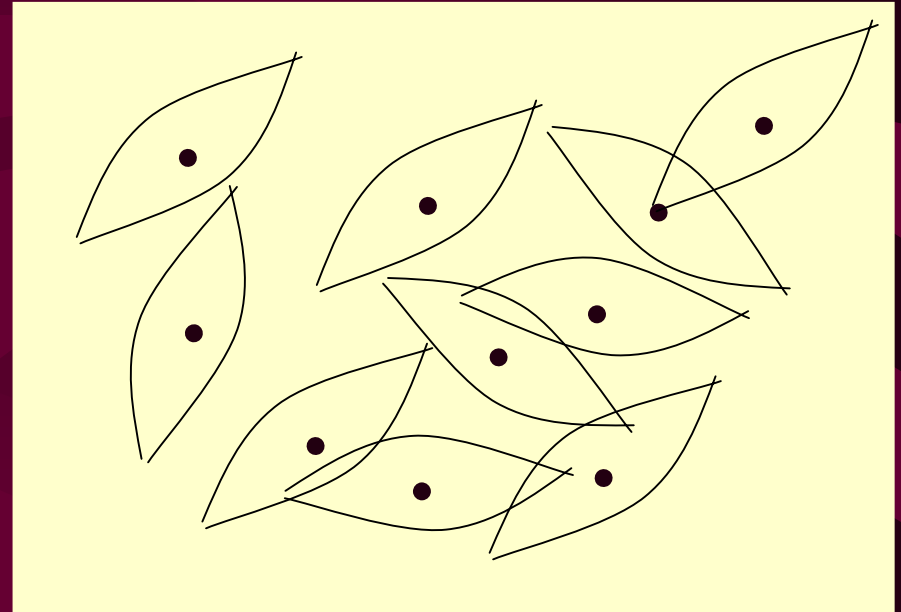
Hypertrophic Cardiomyopathy

- Pathological changes are of muscular hypertrophy associated with fibrosis
- Muscle fibres hypertrophy
- Myocardial disarray - some parts of the heart
- coronary arteries tendancy - large calibre

Myocardial Disarray



Normal Muscle Structure
(Parallel alignment)

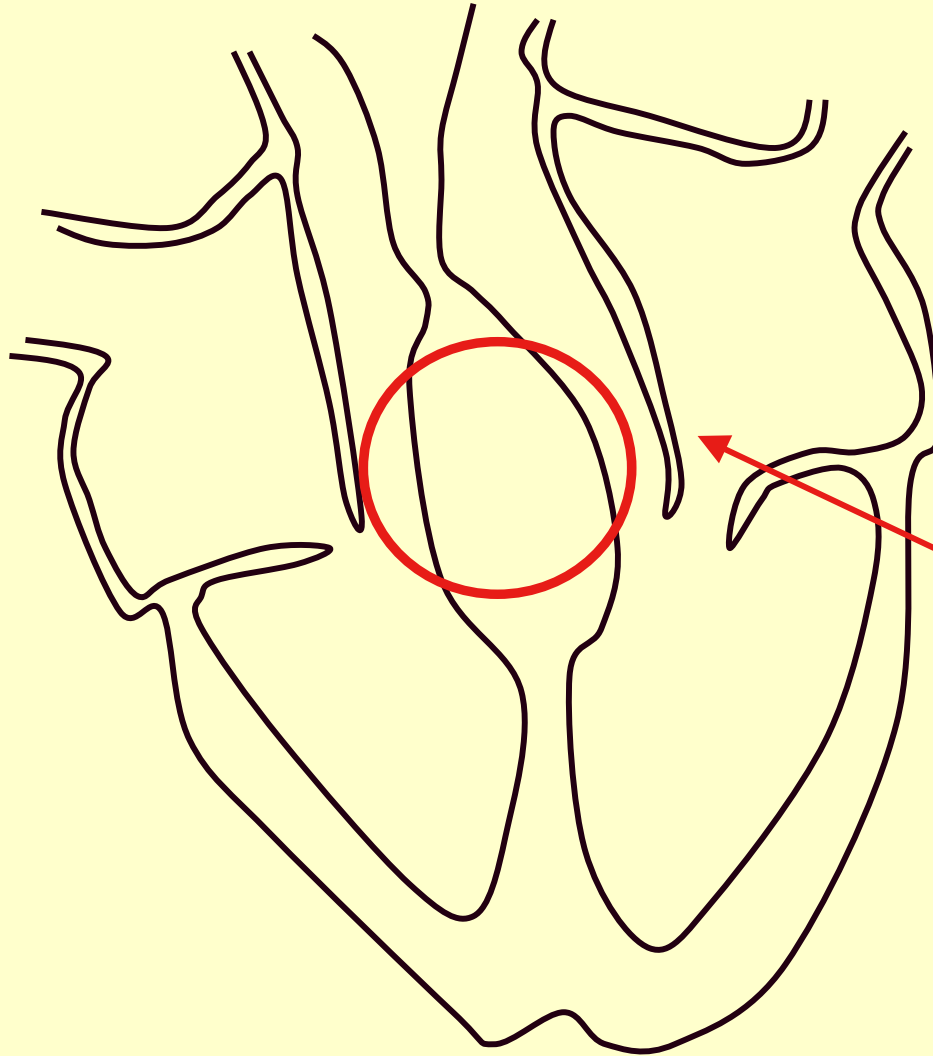


Myocardial Disarray
(Disorganised alignment)

Types Hypertrophic Cardiomyopathy

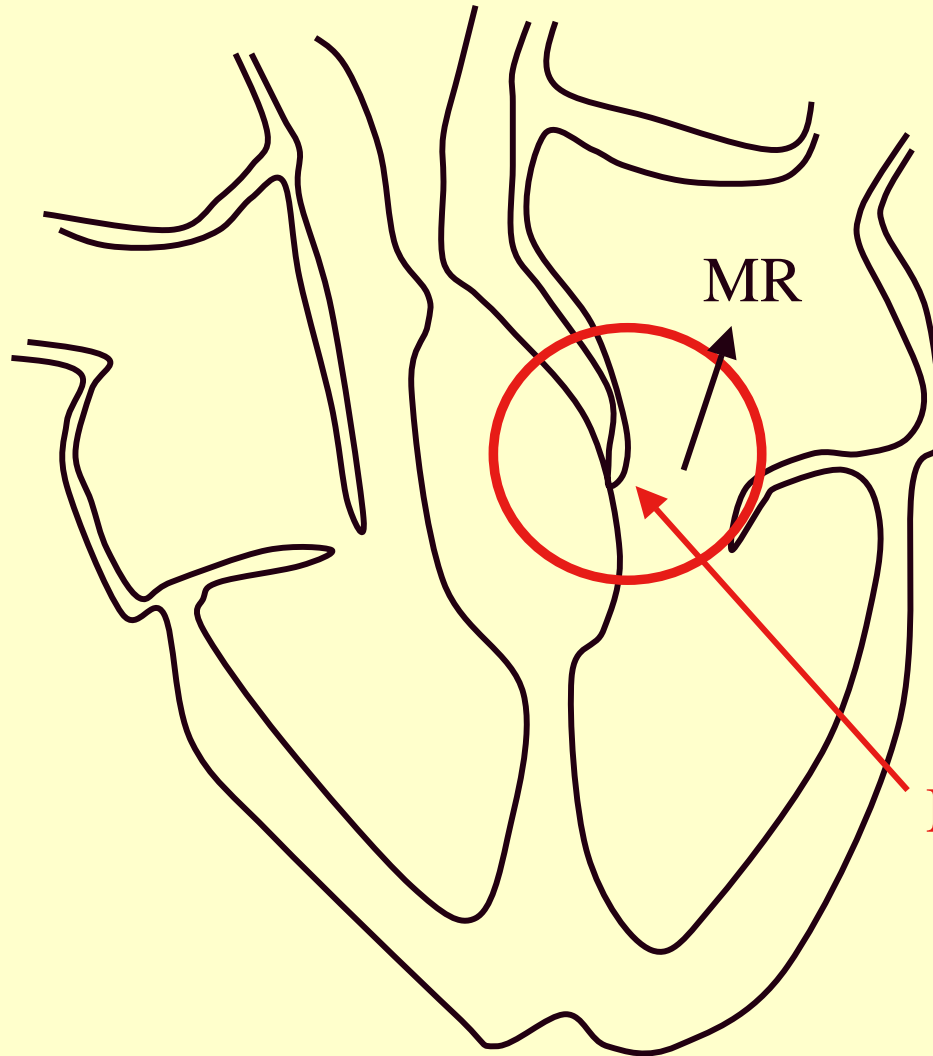
- HCM or HOCM
- Asymmetric septal (ASH) - without obstruction
- Asymmetric septal (ASH) - with obstruction
- Symmetric hypertrophy - concentric
- Apical hypertrophy

HCM



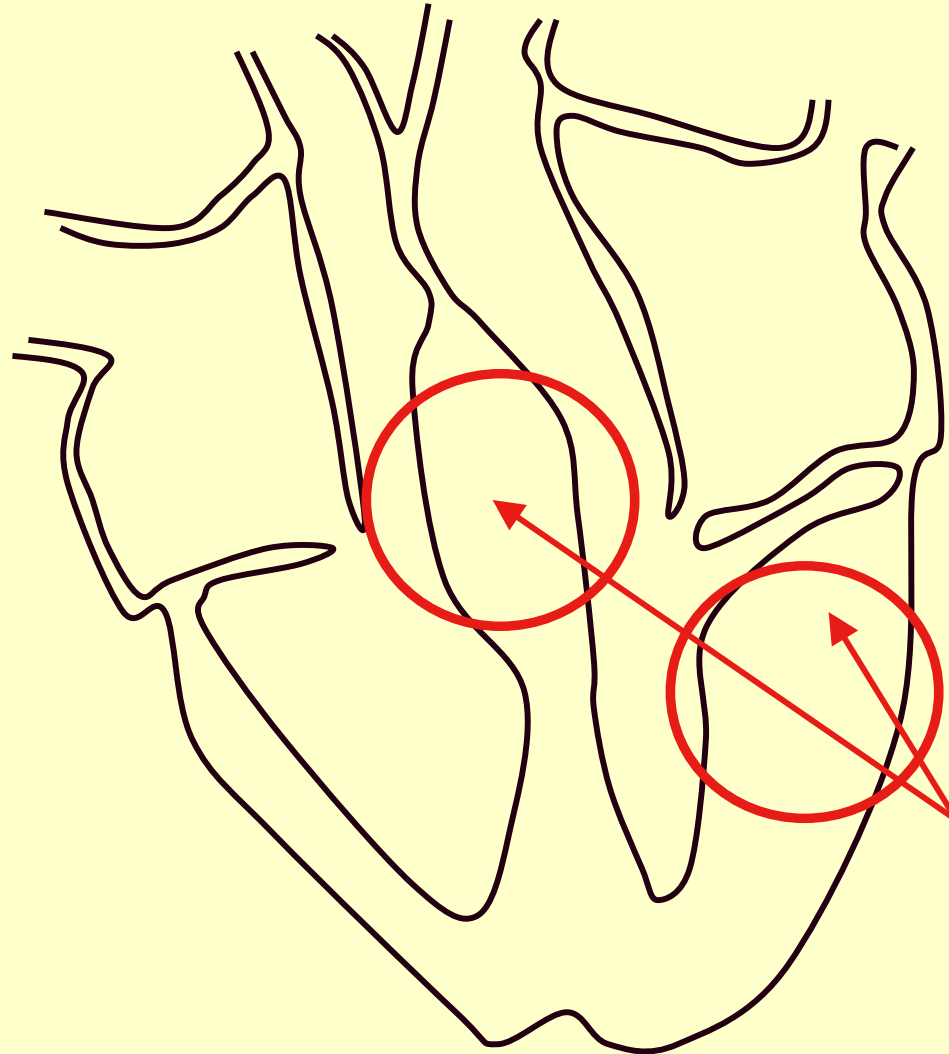
Mitral
Valve in
normal
position

HOCM



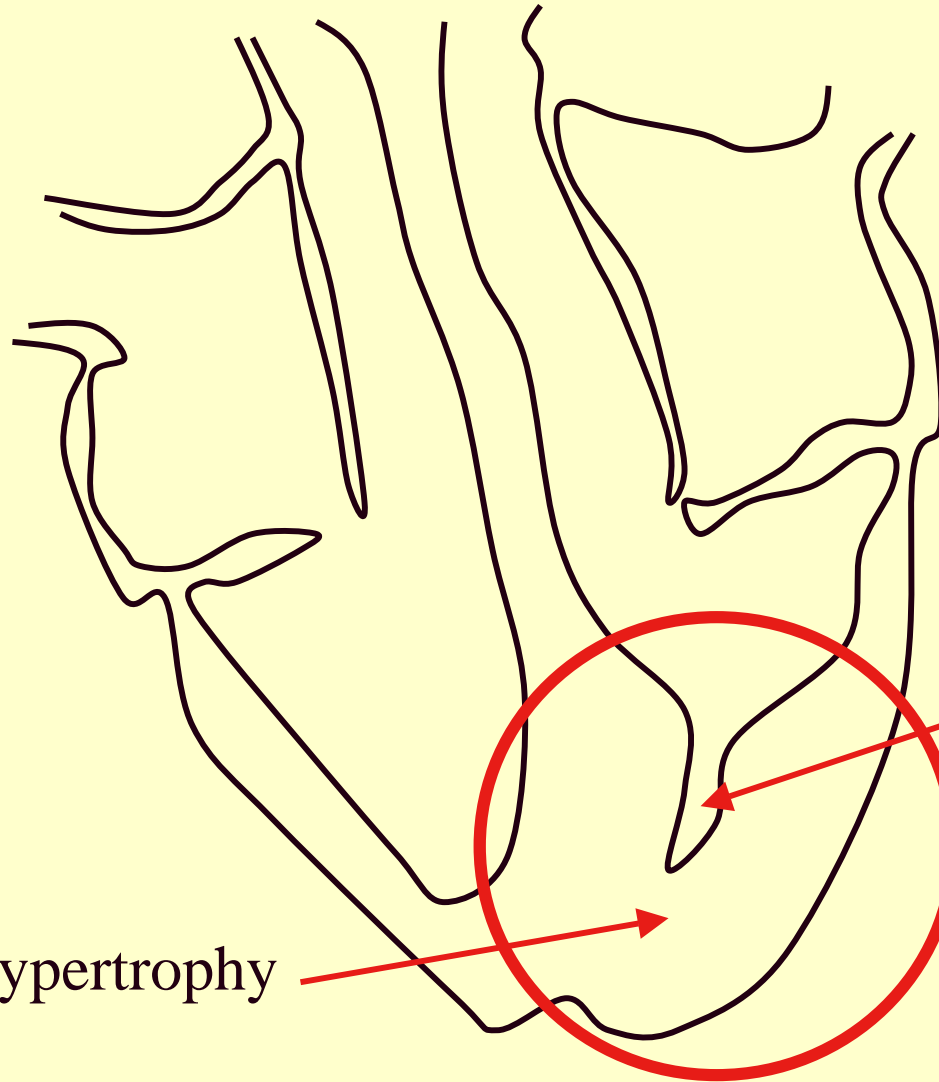
Mitral valve
presses
against
septum

SYMMETRIC



symmetric
or
concentric

APICAL



Apical Hypertrophy

Small
cavity
remains

Symptoms

- Dyspnoea
- angina
- palpitation
- dizzy spells
- syncope

- Gene is present from birth
- linked to growth hormone
- Myopathy can develop at any age - mostly under age 25, but not always
- diagnosis at early age increases risk of poor prognosis

Prognosis

- Adults - 2-3% SCD per year
- Adolescents - 4-6% SCD per year
- Infants (less than 1 yr old), mortality = 50%

Identification of patients at risk

- Diagnosis in childhood/adolescence
- Family history of HCM or SCD
- Syncope

Mechanism of death

- Mostly unknown
- ?VT or VF
- AF with fast ventricular response
- Any arrhythmia or increased obstruction causing Haemodynamic compromise

Dilated Cardiomyopathy

Dilated Cardiomyopathy

- Dilated left ventricle
- poorly contracting
- little difference between systole and diastole
- low cardiac output

- Difficult to distinguish between dilated cardiomyopathy & dilated left ventricle due to severe CAD
- Usually post MI/CAD - local dysfunction
- Dilated myopathy - global dysfunction

Right ventricular dilatation

- Uhl's disease
 - RV dysplasia or myopathy
 - congenital
 - associated ventricular arrhythmias

Restrictive Cardiomyopathy

Restrictive Cardiomyopathy

- Idiopathic
- Infiltrative eg - amyloidosis
- Cardiac transplant rejection
- Endomyocardial fibrosis

Characteristics

- Abnormal compliance of the left ventricle
- short relaxation time

Summary

- Muscle disease
- various types
- diagnosis
 - ECHO
 - BLOOD TESTS
 - LINK TO MEDICAL CONDITIONS
- Classification difficult
 - discrepancies between countries (terminology & nomenclature)