CARDIOMYOPATHY

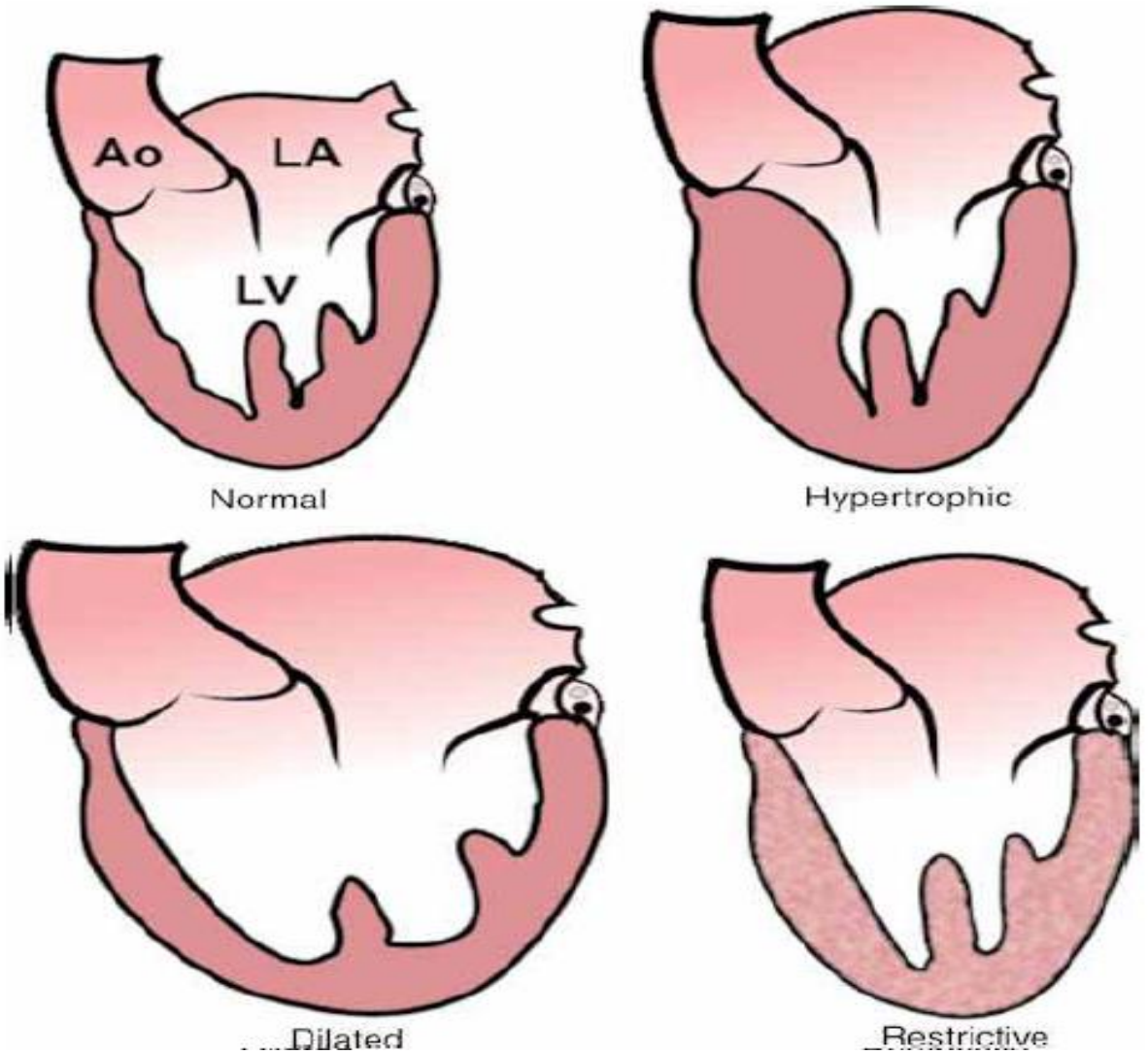
Etiology, Classification & Management

Sheba Medical Center
Cardiology Department
Matthew Wright
St. George's University of London

Introduction

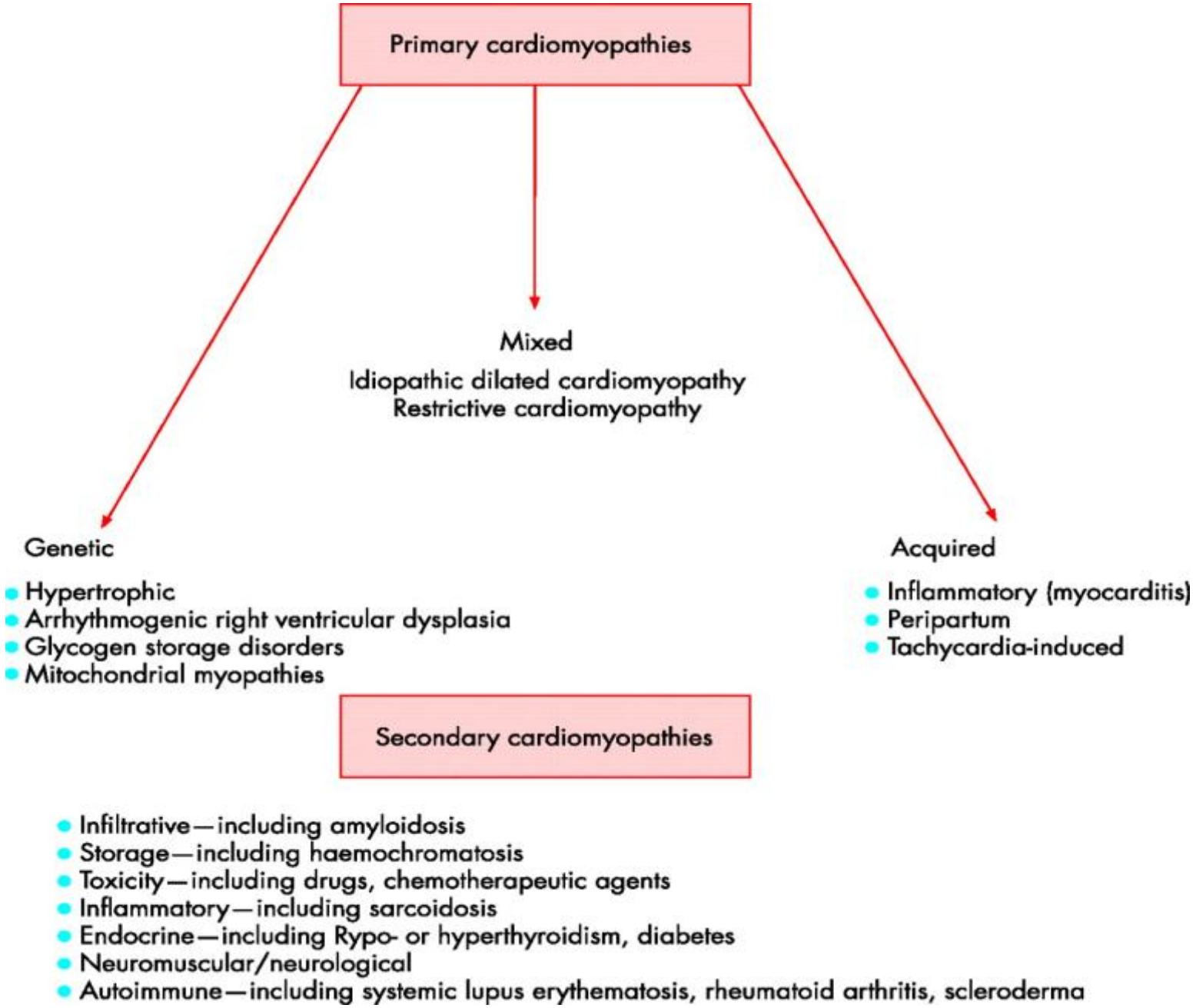
- World Health Organization (1995):
- *Diseases of myocardium (heart muscle) associated with cardiac dysfunction*
- Heart loses ability to pump blood
- Heart muscle becomes enlarged or abnormally thick or rigid
- Progression of cardiomyopathy leads to:
  - Heart failure
  - Arrythmias
  - Systemic/pulmonary oedema
- 3 Main Types:
  - Dilated Cardiomyopathy (DCM)
  - Hypertrophic Cardiomyopathy (HOCM)
  - Restrictive Cardiomyopathy (RCM)

Fig. 1. Cardiomyopathies: classification.



Classification

- Primary/Intrinsic Cardiomyopathy
  - Disease involving the myocardium and of unknown etiology
  - Genetic
    - HOCM
    - ARVC
  - Idiopathic
    - DCM
    - RCM
  - Acquired
    - Peripartum CM
    - Takotsubo CM
- Secondary/Extrinsic Cardiomyopathy
  - Systemic disease and/or known cause
    - Metabolic (Amyloid, HH)
    - Inflammatory (Chagas)
    - Endocrine (Hyperthyroidism)
    - Toxicity (Alcoholic CM)


Primary cardiomyopathies
Mixed
Idiopathic dilated cardiomyopathy
Restrictive cardiomyopathy
Genetic
Hypertrophic
Arrhythmogenic right ventricular dysplasia
Glycogen storage disorders
Mitochondrial myopathies
Acquired
Inflammatory (myocarditis)
Peripartum
Tachycardia-induced
Secondary cardiomyopathies
Infiltrative—including amyloidosis
Storage—including haemochromatosis
Toxicity—including drugs, chemotherapeutic agents
Inflammatory—including sarcoidosis
Endocrine—including Rypo- or hyperthyroidism, diabetes
Neuromuscular/neurological
Autoimmune—including systemic lupus erythematosis, rheumatoid arthritis, scleroderma

Etiology

- Ischemic cardiomyopathy
- Idiopathic cardiomyopathy
- Hypertensive cardiomyopathy
- Infectious cardiomyopathy
- Alcoholic cardiomyopathy
- Toxic cardiomyopathy
- Peripartum cardiomyopathy
- Radiotherapy (cobalt)
- Diabetes
- Thyroid Disease

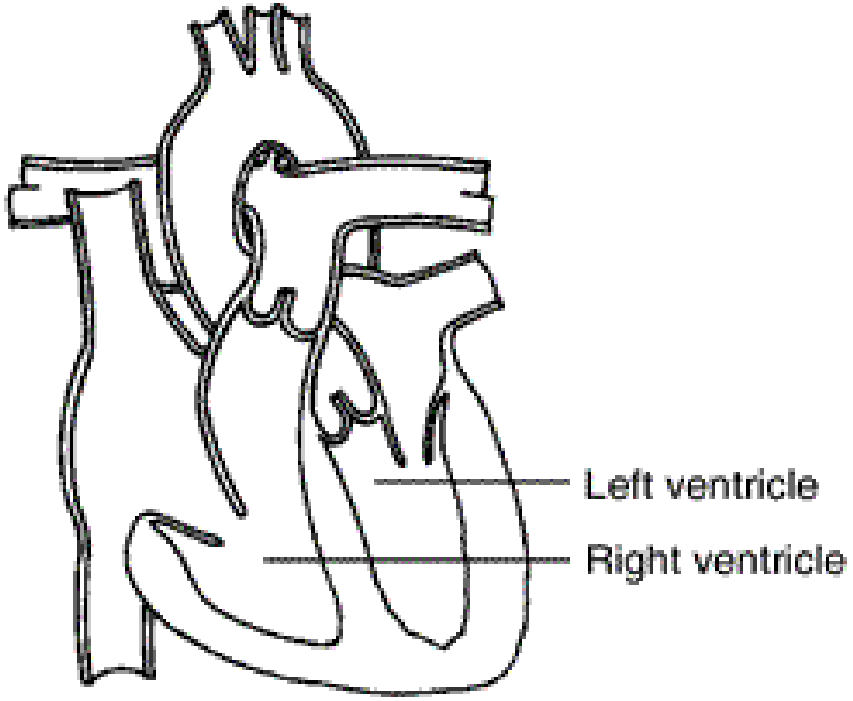
Management – General

- Treat underlying/contributing cause
- Symptom control
- Stop progression
- Reduce complications and chance of SCD
- Lifestyle Changes
- Pharmacological Therapy
- Surgical Therapy – Anaesthetic Considerations!!
  - Septal Myomectomy
  - LVAD
  - ICD
- Heart Transplant

Dilated Cardiomyopathy

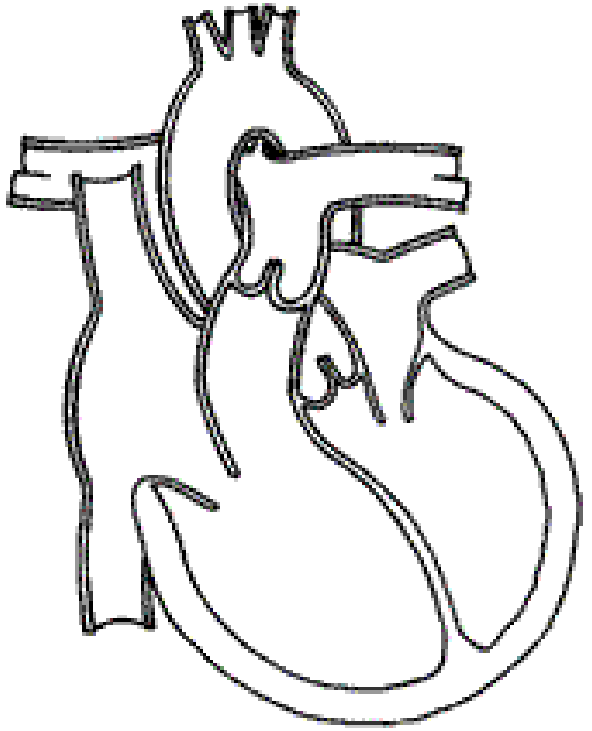
- Most common form of CM (60%)
- Age group: 20-60
- More common in men
- Ischaemic vs. Non-ischaemic
  - Ischaemic – CAD/IHD
  - Non-ischaemic – Myocarditis, Toxic, Peripartum
- Characterised by:
  - Dilation of the cardiac chambers
  - Reduction in ventricular contractile function

Normal Heart



Heart chambers relax and fill,
then contract and pump.

Heart with Dilated Cardiomyopathy



Muscle fibers have stretched.
Heart chamber enlarges

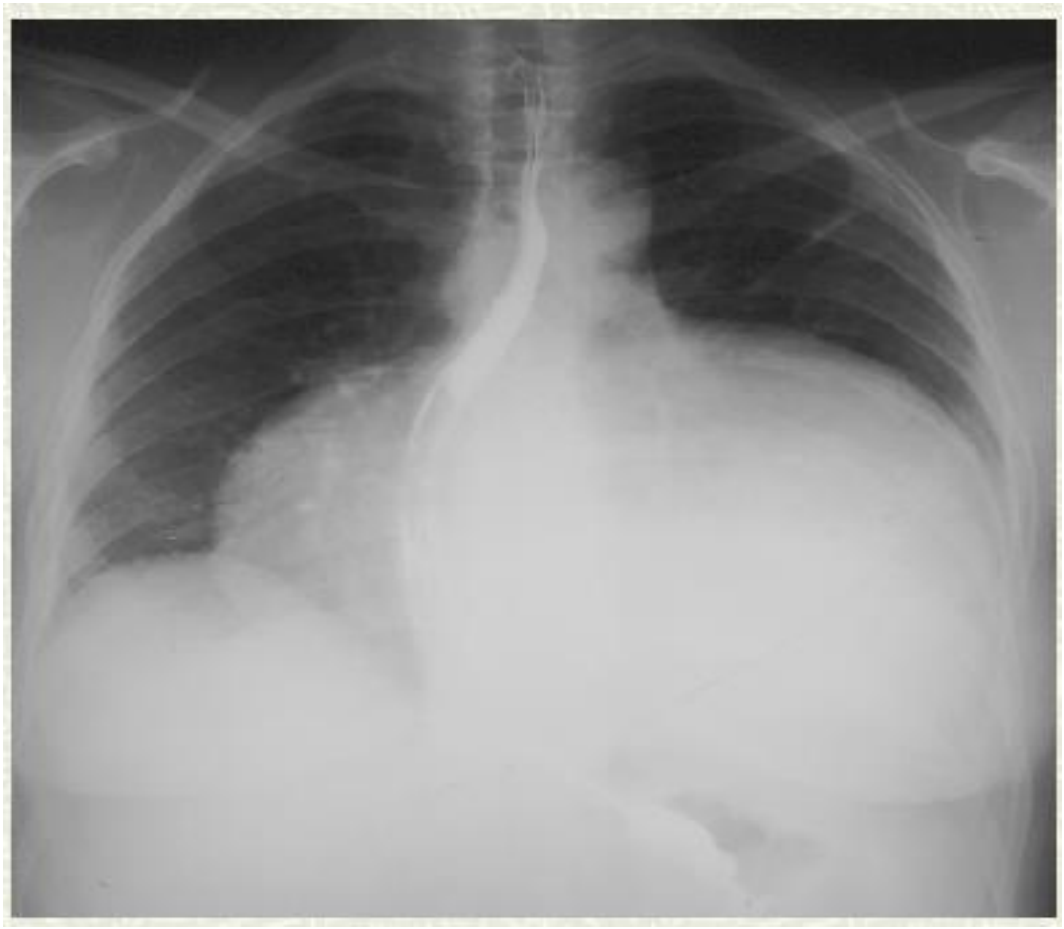
DCM - The Patient

- Clinical Presentation
  - Symptomatic HF (syncope, dyspnea, fluid overload)
  - Some patients are asymptomatic
  - ECG shows nonspecific changes
    - Sinus tachy/atrial fibrillation
    - Ventricular arrhythmias
    - Left atrial abnormality

- Physical Exam
  - Variable degrees of cardiac enlargement
  - Pulse pressure is narrow
  - JVP raised
  - 3^{rd} or 4^{th} heart sound are common
  - Mitral/ tricuspid regurgitation

DCM - Diagnosis

- CXR
  - Cardiomegaly, Pulmonary Congestion
- ECG
  - Low-voltage QRS, Abnormal Axis, ST Changes, LVH
- 2D Echo
- Coronary Angiography
  - Distinguish between ischaemic vs. non-ischaemic
- Endomyocardial Biopsy



Cardiac chamber dimensions	Dilated cardiac chambers, esp. the left ventricle; dilated atria; right sided chambers can be dilated as well
Wall dimensions	Normal, mildly increased, or decreased
Systolic indices and ejection fraction (normal >55%)	Reduced systolic function; decreased EF, <30%
Diastolic function	Impaired; may reflect volume overload
Valvular function: MR; TR	MR secondary to annular dilation and apical tethering; TR frequent

DCM - Management

- **Treatment goals**
  - Symptom management
  - Stop progression
  - Prevent complications
- **Pharmacological Therapy**
  - Mainstay of treatment
  - Symptomatic & reduce mortality
- **Surgical Therapy**
  - Reduction of afterload
  - Optimizing preload
  - Minimize myocardial depression

Pharmacological Therapy

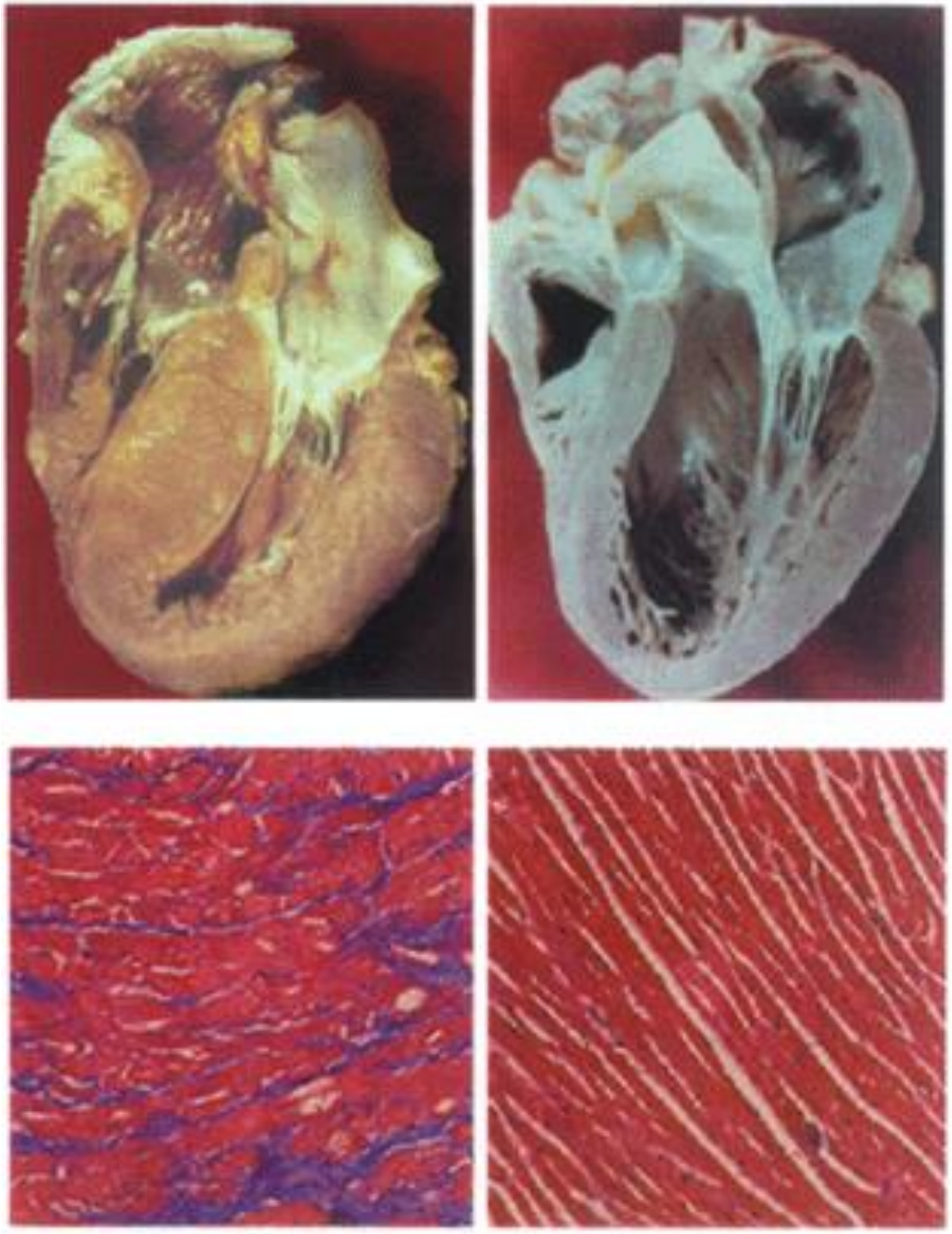
- Vasodilators
  - ACE-I
  - Milrinone
- Diuretics
  - Spironolactone
- Beta-Blockers
- Digoxin
- Amiodarone
- Anticoagulants

Surgical Therapy

- Dual Chamber Pacing
- Cardiomyoplasty
- LV Assist Devices
- Cardiac Transplantation
- Cardiac procedures
  - Correction of AV valve insufficiency
  - Placement of ICD device
  - LV Assist device placement
  - Allograft Transplantation

Hypertrophic Cardiomyopathy

- Most common cause of sudden death in young, healthy individuals (VT/VF)
- Prevalence- 0.2% population
- Causes:
  - Genetic – Autosomal Dominant
  - Acquired
  - Idiopathic
- Obstructive vs. Non-obstructive
- Characterized by:
  - Inppropriate LV hypertrophy
  - Myocardial disarray
- Hypertrophy may be generalized or confined largely to interventricular septum.


HCM
Normal

HCM - Pathophysiology

- Subaortic Obstruction
- Diastolic Dysfunction
- Myocardial Ischemia
- Mitral Regurgitation
- Arrythmias

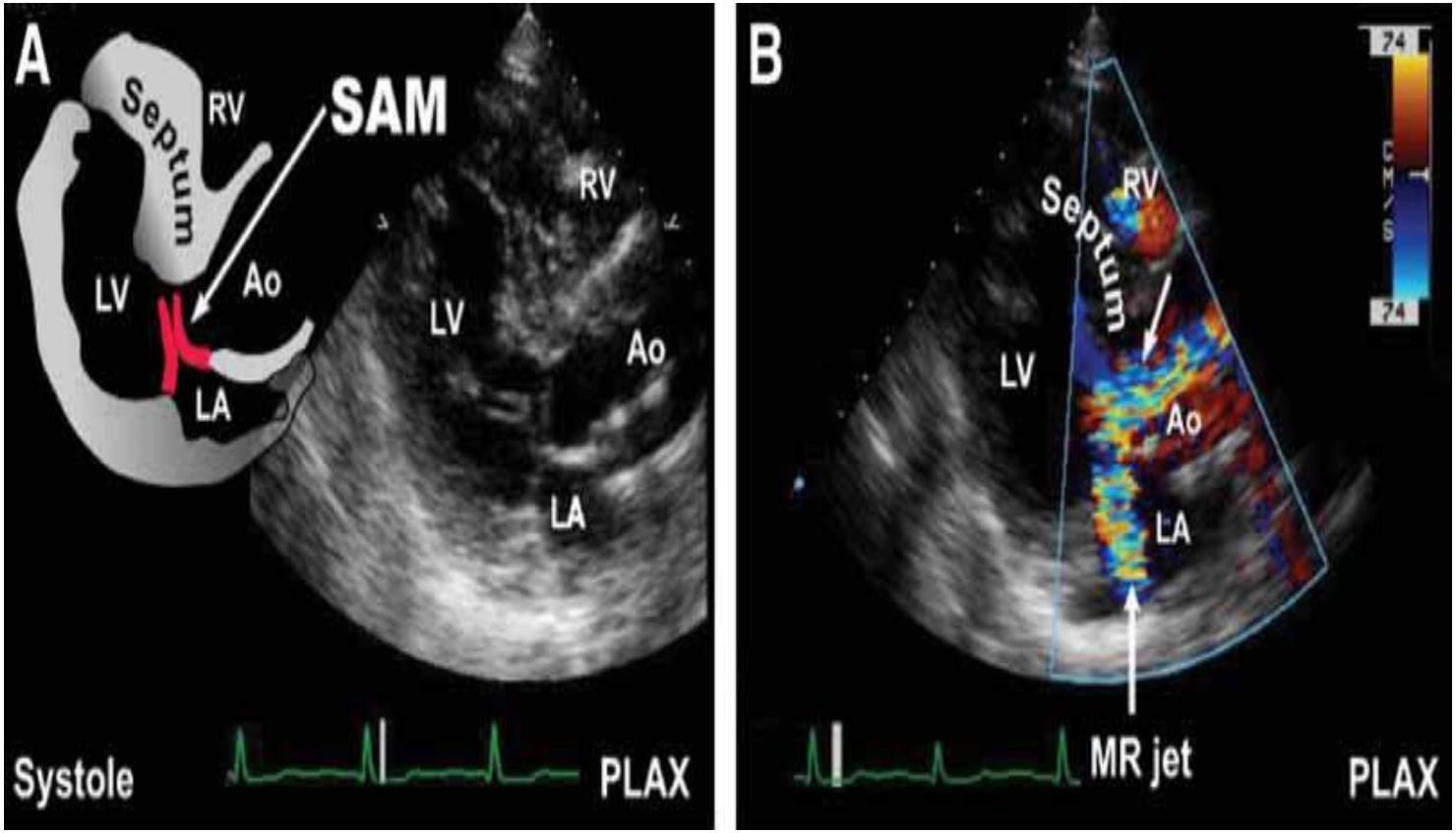
HCM – The Patient

- Symptoms
  - Angina on effort
  - Dyspnea on effort
  - Syncope on effort
  - Sudden death

- Signs
  - Jerky pulse
  - Palpable LV heave
  - Double impulse at apex
  - mid-systolic murmur –sign of LV outflow obstruction
  - Pansystolic murmur

DCM – Diagnosis

- ECG
  - ↑ QRS voltage, ST-T changes, Axis deviation, LVH
- CXR
  - Left atrial enlargement or normal
- Echo
  - Aymmetric hypertrophy
  - SAM of mitral valve
  - LVOT obstruction
- Invasive Cardiac Catheterisation
  - Indications: Suspected CAD or Severe mitral valve disease
  - LV pressure gradient, ↓ ventricular volume, ↑ LVEDP


A
Septum
RV
SAM
LV
Ao
LA
RV
LV
Ao
LA
Systole
PLAX
B
74
CM/S
-74
RV
Septum
LV
Ao
LA
MR jet
PLAX

HCM – Management

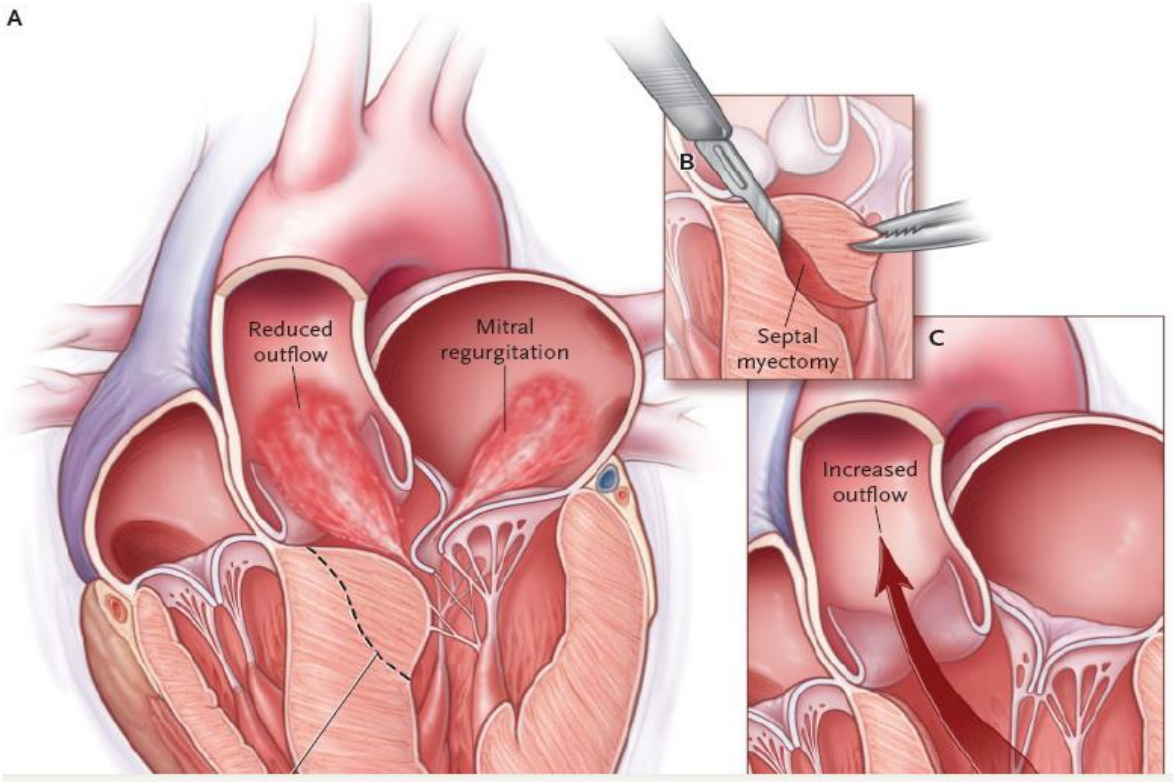
- Early identification for High Risk Patients
- Pharmacological
- Surgical
- Non Surgical Alternatives
- Implantable Cardioverter Defibrillator(ICD)
- Cardiac Transplantation

Pharmacological Therapy

- Beta-Blockers
  - Mainstay of therapy
  - Symptomatic relief
- Calcium Channel Blockers
  - Improves diastolic function
- Disopyramide
  - Negative inotropic & Vasoconstrictive effect
  - Reduce LVOTO
- Arrhythmias respond well to amiodarone

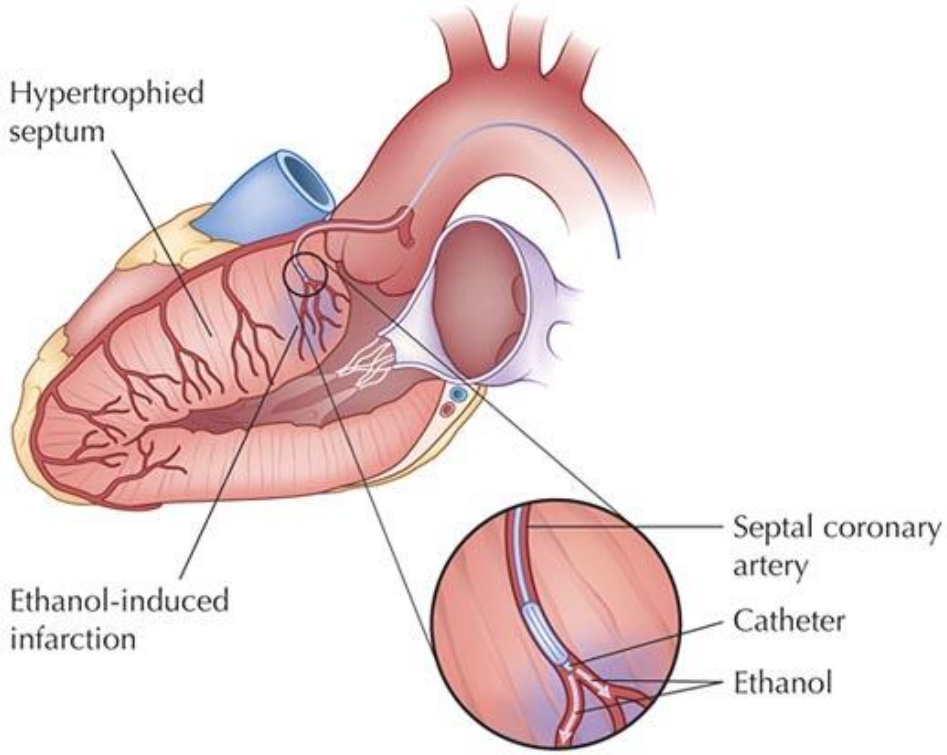
Surgical Therapy

- Surgical Septal Myectomy
  - Gold standard
  - Operative mortality: <1%
  - Complications rare (heart block, VSD, aortic regurg)



Non-Surgical Alternatives

- Alcohol Septal Ablation
  - Non surgical septal reduction therapy
  - Reduce LVOT grad in 85-90% immediately
  - Permanent heart blocks (5-10%)
- Dual Chamber or AV Sequential Pacing
  - Exact mechanism unknown
  - Rarely recommended

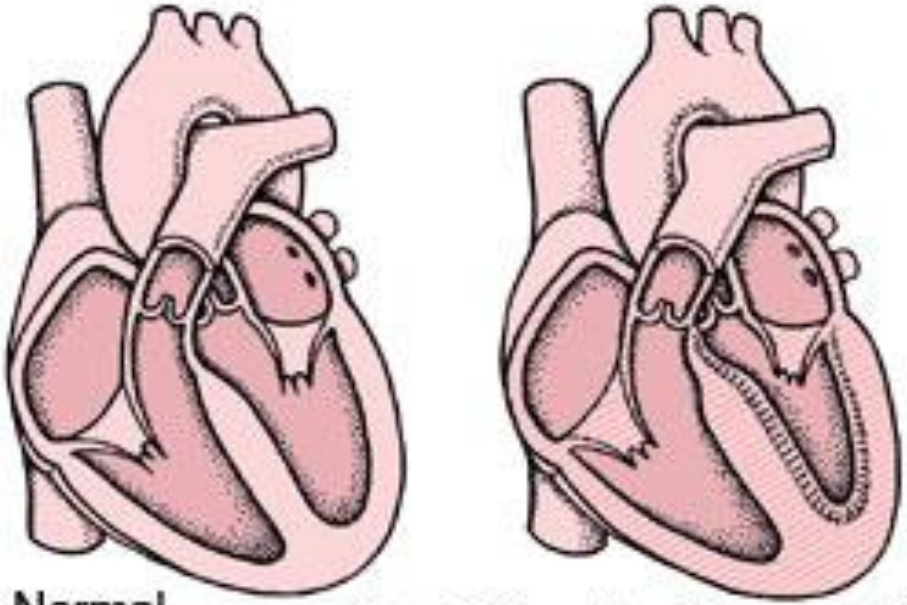


Restrictive Cardiomyopathy

- WHO 1995 Defintion:
  - *Restrictive filling & reduced diastolic volume of either or both ventricles with normal or near normal systolic function & wall thickness*
- Rare condition
- Primary Causes:
  - Loeffler's Endocarditis
  - Endocardial Fibroelastosis
- Myocardial
  - Non-infiltrative – Idiopathic, Familial, HCM, DM
  - Infiltrative – Amyloidosis, Sarcoidosis
  - Storage Disease- HH, Glycogen
- Endomyocardial
  - Fibrosis
  - Carcinoid
  - Radiation
  - Drug-Induced – Serotonin, Busulfan, Ergotamine

RCM - Pathophysiology

- Ventricles become stiff and rigid
  - Replacement of the normal heart muscle with abnormal tissue, such as scar tissue.
- Ventricles cannot relax – Diastolic Dysfunction
- Increased atrial pressure, atrial hypertrophy/dilatation
- Complications
  - Heart Failure
  - Arrhythmias



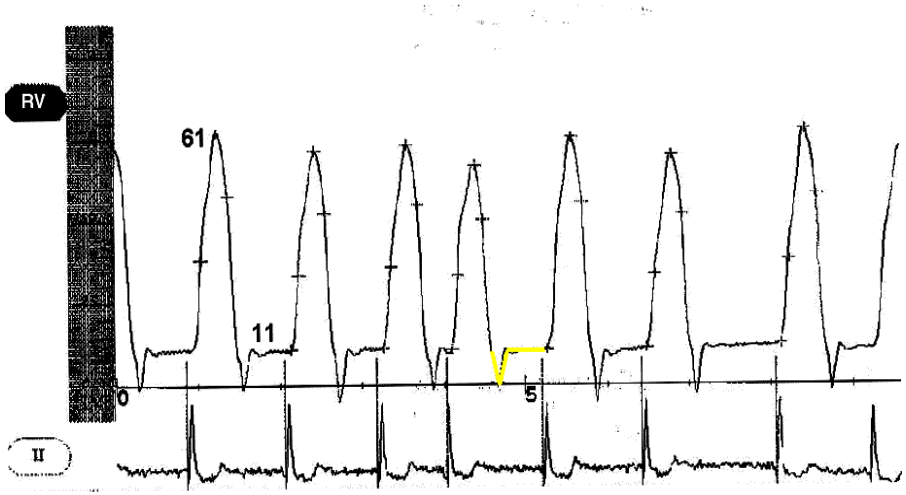
Normal

Restrictive Cardiomyopathy

In restrictive cardiomyopathy, the walls of the ventricles become stiff, but not necessarily thickened.

RCM – The Patient

- Symptoms
  - Symptoms of R/L Heart Failure
  - Kussmaul's Sign – Increased JVP during inspiration
  - Pulsus paradoxus
- Signs
  - CXR- pulmonary congestion, small heart size
  - ECG- BBB, low voltage QRS, Square Root Sign
  - 2D Echo – Dilated atria, reduced ventricles, diastolic dysfunction



RCM – Diagnosis

- Diagnosis is difficult
- Requires complex Doppler echocardiography
- CT and MRI
- Endomyocardial biopsy

- Differential Diagnosis – Constrictive Pericarditis
  - Paradoxical Pulse
  - MR/TR usually absent
  - CXR – Pericardial calcification
  - ECG – usually normal
  - Echo - Minor atrial enlargement
  - MRI – Thickened pericardium

RCM - Management

- Depends on aetiology
- Poor prognosis
- Transplantation is the best treatment
- Idiopathic
  - Diuretics
  - B-blockers, Amiodarone, CCB
  - Anticoagulation
  - CCBs, ACEi
  - Dual Chamber Pacing
- Amyloidosis
  - Melphelan, prednisone, H+L transplant
- Haemochromatosis
  - Phlebotomy, Desferrioxamine
- Carcinoid
  - Somatostatin analogs, Valvuloplasty/Valve replacement
- Sarcoidosis
  - Steroids , Pacing, ICD, Transplantation

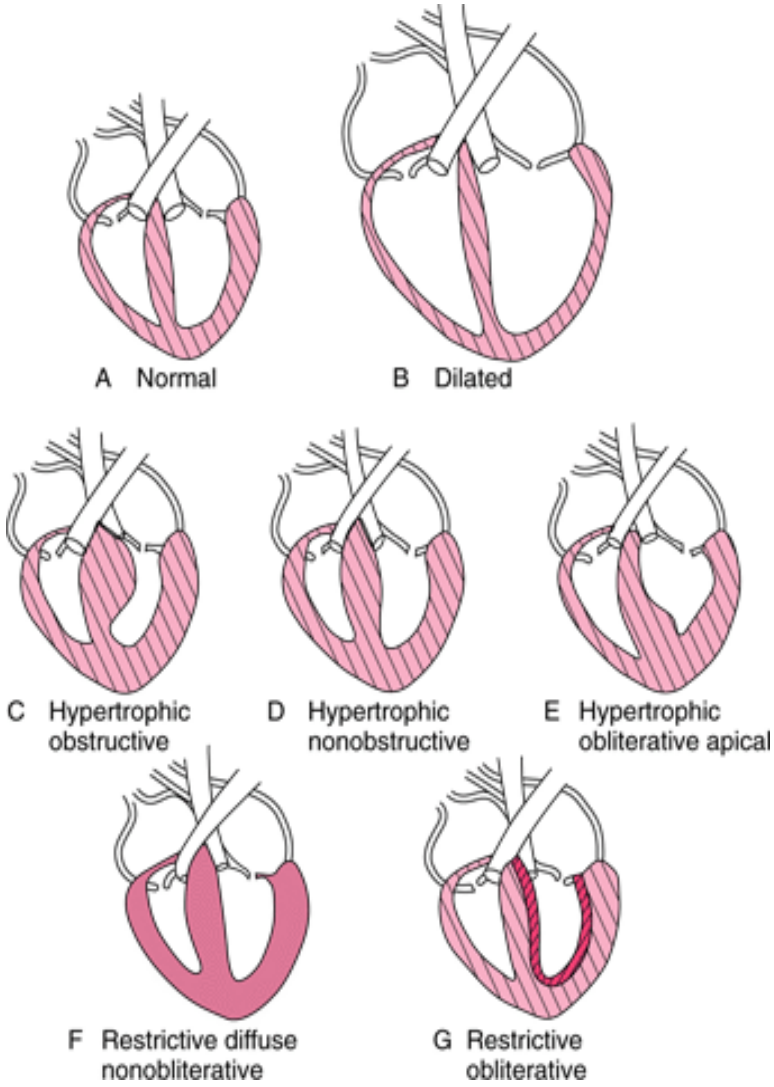
Arrhythmogenic Right Ventricular Dysplasia

- In this condition, patches of the right ventricular myocardium are replaced with fibrous and fatty tissue.
- Inherited as autosomal dominant trait
- Dominant clinical problems are:
  - Ventricular arrhythmia
  - Sudden death
  - Right sided cardiac failure
- ECG shows inverted T waves in the right precordial leads.
- MRI useful diagnostic tool and used to screen 1st degree relatives from having the same pathology

Takotsubo Cardiomyopathy

- Transient, reversible, left ventricular dysfunction
- Temporary weakening of myocardium
- Causing severe hypotension – can mimic an acute coronary event
- Rare condition
- Associated with emotional stress – ‘broken heart syndrome’
- Cardiac catheterization often reveals normal coronary arteries
- Treatment supportive

SUMMARY


A Normal
B Dilated
C Hypertrophic obstructive
D Hypertrophic nonobstructive
E Hypertrophic obliterative apical
F Restrictive diffuse nonobliterative
G Restrictive obliterative

THANK YOU