cardiomyopathies

BY

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Cardiomyopathies:

Cardiomyopathies are primary diseases of the myocardium, which are classified according to their effects on cardiac structure and function .They can be inherited or be caused by infections or exposure to toxins. In some cases no cause is identified.

Functional Classification

- Dilatated (congestive, DCM, IDC)
  - ventricular enlargement and syst dysfunction
- Hypertrophic (IHSS, HCM, HOCM)
  - inappropriate myocardial hypertrophy in the absence of HTN or aortic stenosis
- Restrictive (infiltrative)
  - abnormal filling and diastolic function

Dilated cardiomyopathy

- In North America and Europe, symptomatic dilated cardiomyopathy has an incidence of 20 per 100 000 and a prevalence of 38 per 100 000.
- Men are affected more than twice as often as women
- **pathology**

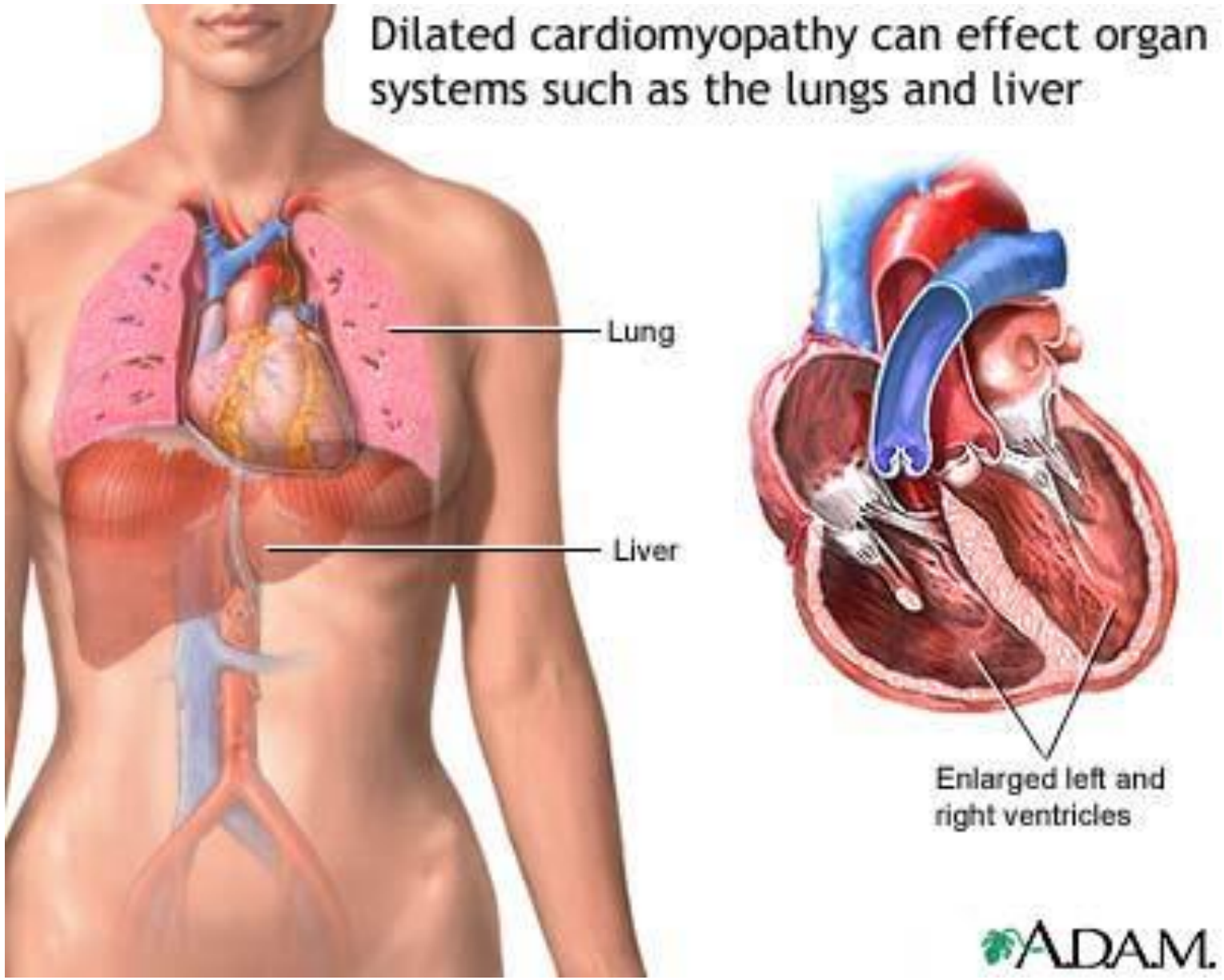
  increased heart size and weight

  ventricular dilatation, normal wall thickness

  heart dysfunction out of portion to fibrosis

Dilatation of the valve rings can lead to functional mitral and tricuspid incompetence.

Dilated cardiomyopathy can effect organ systems such as the lungs and liver



Pathogenesis:

- **Idiopathic cardiomyopathy - the cause is unknown.**
- **At least 25% of cases are inherited as an autosomal dominant trait** and a variety of single-gene mutations have been identified. Most of these mutations affect proteins in the cytoskeleton of the myocytes, such as dystrophin, lamin A and C.
- Many are **also associated with abnormalities of skeletal muscle,** most of the X-linked inherited skeletal muscular dystrophies, such as Becker and Duchene ,are associated with cardiomyopathy.

Alcohol may be an important cause in some patients.

Infectious cardiomyopathy - HIV, viral myocarditis

Ischemic cardiomyopathy - caused by CAD & MI , w/c leave scars in the heart muscle

Toxic cardiomyopathy – due to cocaine, amphetamines, and some chemotherapy drugs (doxorubicin, daunorubicin)

Peripartum cardiomyopathy: This type appears in women during the last trimester of pregnancy or after childbirth.

Radiotherapy (cobalt)

diabetes and thyroid disease

Clinical features:

- Symptoms of heart failure
  - pulmonary congestion (left HF)
    dyspnea (rest, exertional, nocturnal), orthpnea
  - systemic congestion (right HF)
    edema, nausea, abdominal pain, nocturia
  - low cardiac output
    fatigue and weakness
  - lightheadedness, palpitation, syncope
- hypotension, tachycardia, tachypnea, JVD

Cardiac Imaging

- Chest radiogram (increased CTR,pulm.congestion)
- Electrocardiogram (LBBB,Sinus tachycardia,AF, etc).
- 24-hour ambulatory ECG (Holter).(Atrial and ventricular arrhythmia)
- Two-dimensional echocardiogram
- Cardiac catheterization
  - age >40, ischemic history, high risk profile, abnormal ECG

Management :

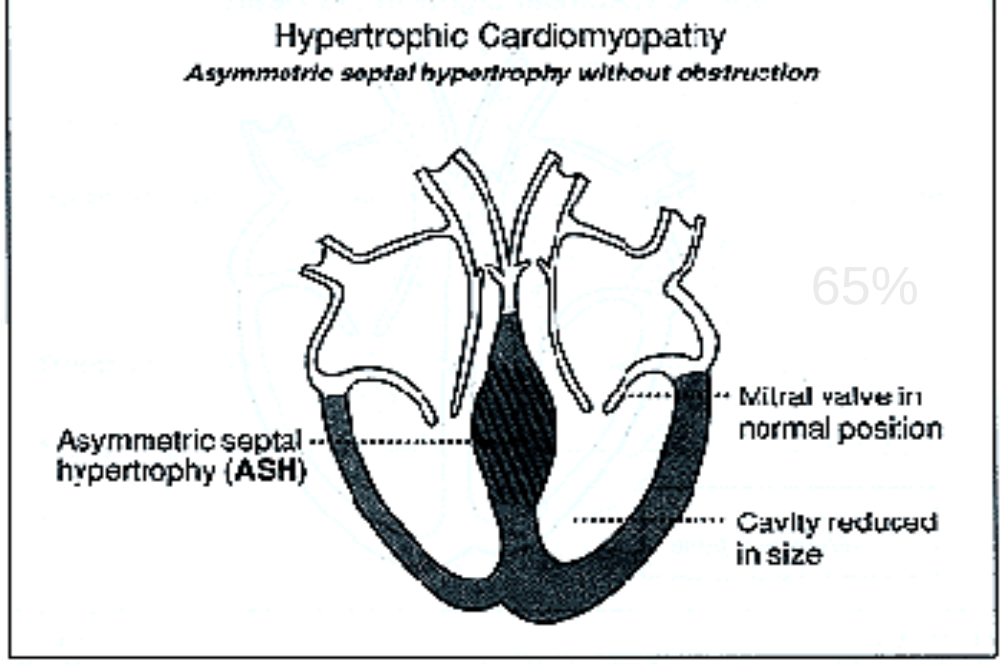
- salt restriction of a 2-g Na^+ (5g NaCl) diet
- Limit activity based on functional status
- fluid restriction for significant low Na+
- initiate medical therapy
  - ACE inhibitors, diuretics
  - hydralazine / nitrate combination

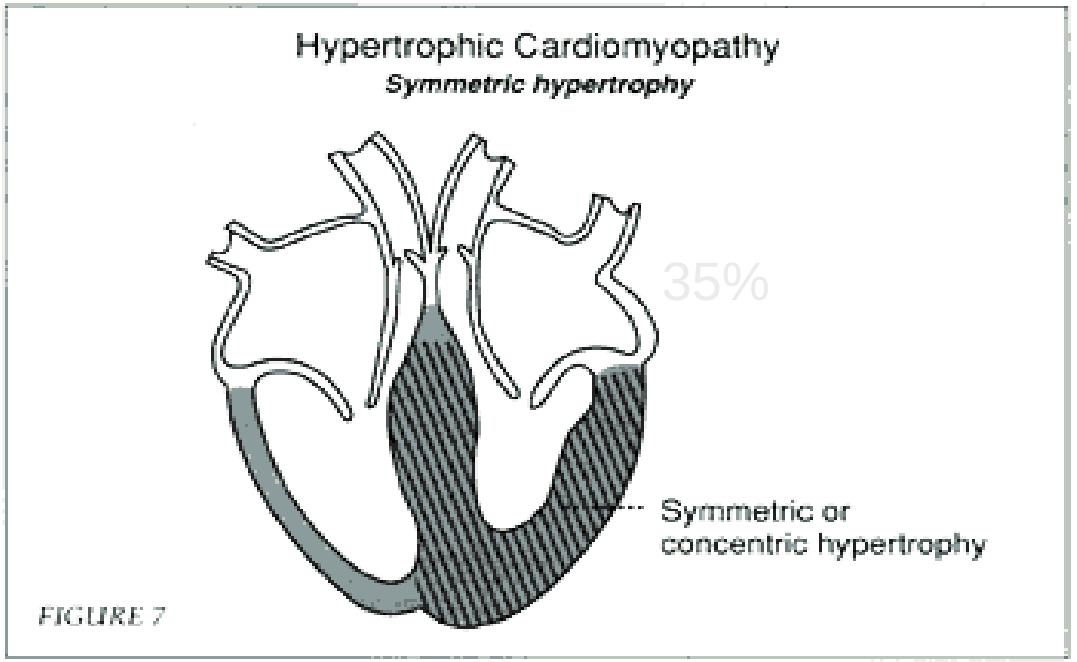
Management :

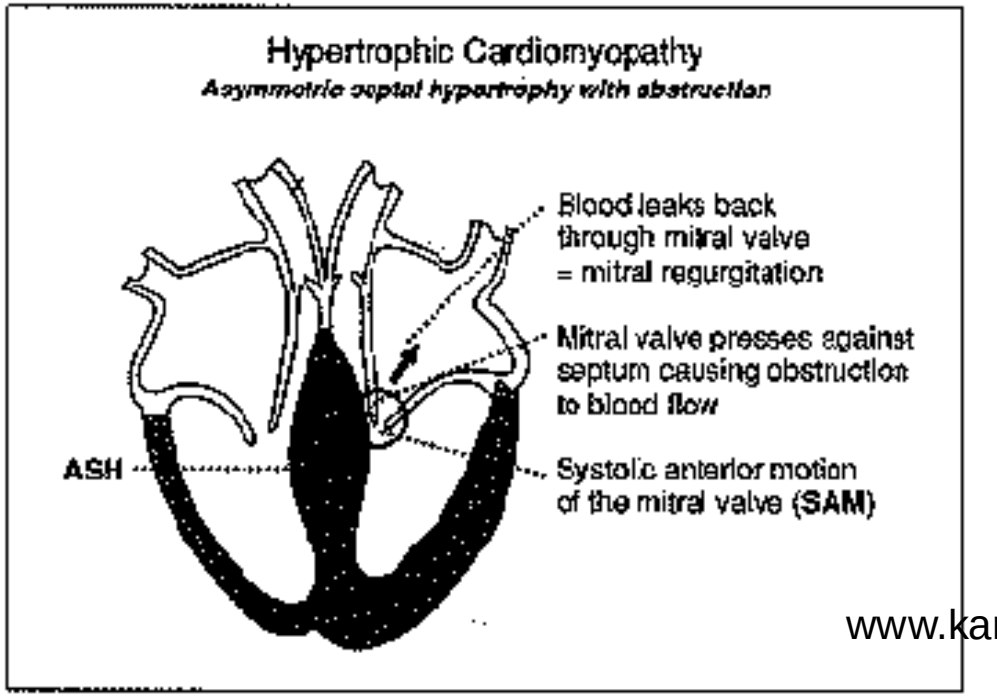
- consider adding ß-blocking agents if symptoms persists
- anticoagulation for EF <30%, history of thromboemoli, presence of mural thrombi
- cardiac transplantation

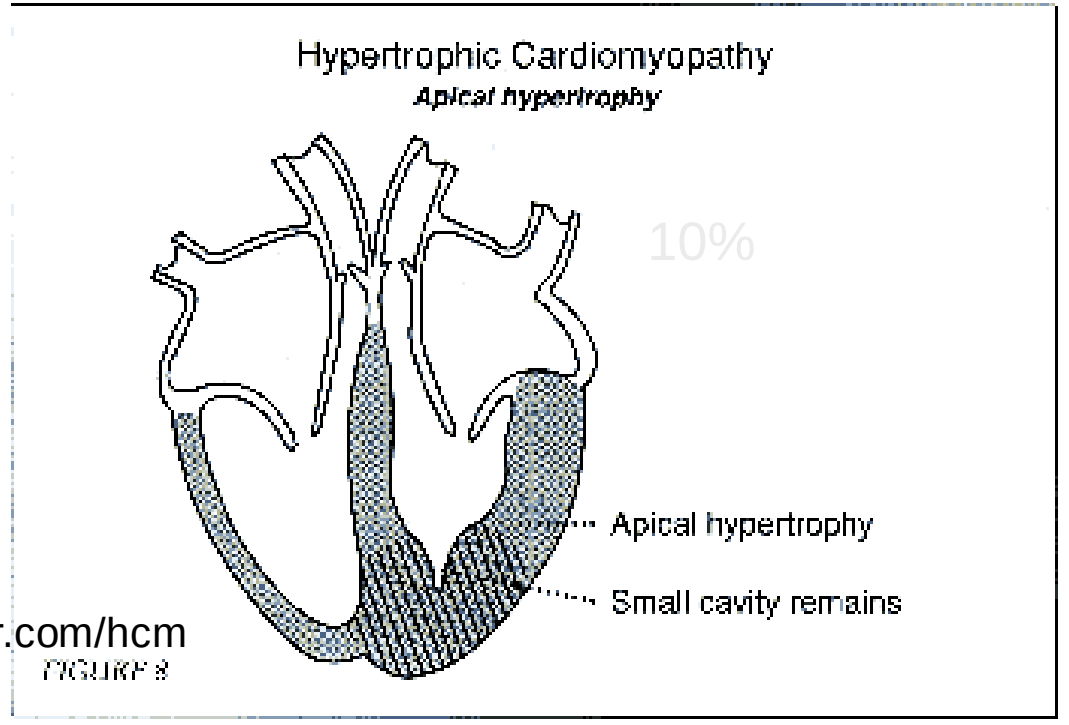
Hypertrophic cardiomyopathy

- This is the most common form of cardiomyopathy, with a prevalence of approximately 100 per 100 000.
- It is characterized by inappropriate left ventricular hypertrophy with malalignment of the myocardial fibres and myocardial fibrosis.
- The hypertrophy may be generalised or confined largely to the interventricular septum (asymmetric septal hypertrophy) or other regions of the heart.
- A specific variant termed apical hypertrophic cardiomyopathy is common in the Far East.









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Familial HCM

- First reported by Seidman et al in 1989
- occurs as autosomal dominant in 50%
- different genes mutation on at least 4 chromosome.
  - chromosome 14 (myosin)
  - chromosome 1 (troponin T)
  - chromosome 15 (tropomyosin)
  - chromosome 11

Clinical features of hypertrophic cardiomyopathy

- Symptoms

I. Angina on effort
II. Dyspnoea on effort
III. Syncope on effort
IV. Sudden death

- Signs

I. Jerky pulse*

II. Double impulse at the apex (palpable fourth heart sound due to left

III. atrial hypertrophy)

IV. Mid-systolic murmur at the base*

V. Pansystolic murmur (due to mitral regurgitation) at the apex

VI. *Signs of left ventricular outflow tract obstruction may be augmented by standing up (reduced venous return), inotropes and vasodilators (e.g. sublingual nitrate).

Risk factors for sudden death in hypertrophic cardiomyopathy

- A history of previous cardiac arrest or sustained ventricular tachycardia
- Recurrent syncope
- An adverse genotype and/or family history
- Exercise-induced hypotension
- Non-sustained ventricular tachycardia on ambulatory ECG monitoring
- Marked increase in left ventricular wall thickness

Investigations

- Echocardiography is the investigation of choice and is usually diagnostic.
- Sometimes the diagnosis is more difficult when another cause of left ventricular hypertrophy is present but the degree of hypertrophy in hypertrophic cardiomyopathy is usually greater than in physiological hypertrophy and the pattern is asymmetrical.
- The ECG is abnormal and shows features of left ventricular hypertrophy ,deep T-wave inversion
- Genetic testing can be performed and is helpful in screening relatives of affected individuals.

Management

- Beta-blockers, rate-limiting calcium antagonists can help to relieve symptoms and prevent syncopal attacks.
- Arrhythmias often respond to treatment with amiodarone.
- No pharmacological treatment has been identified that can improve prognosis, however. Outflow tract obstruction can be improved by partial surgical resection (myectomy) or by iatrogenic infarction of the basal septum (septal ablation) using a catheter-delivered alcohol solution.
- Digoxin and vasodilators may increase outflow tract obstruction and should be avoided.
- An ICD should be considered in patients with clinical risk factors for sudden death.

Restrictive cardiomyopathy

- In this rare condition, ventricular filling is impaired because the ventricles are ‘stiff’ .This leads to high atrial pressures with atrial hypertrophy, dilatation and, later, AF.
- Amyloidosis is the most common cause in the UK, although other forms of infiltration due to glycogen storage diseases, idiopathic peri-myocyte fibrosis and a familial form of restrictive cardiomyopathy can also occur.
- The diagnosis can be difficult and requires assessment with Doppler echocardiography, CT or MRI, and endomyocardial biopsy.
- Treatment is symptomatic but the prognosis is usually poor and transplantation may be indicated.

Exclusion "Guidelines"

- LV end-diastolic dimensions ≥ 7 cm
- Myocardial wall thickness ≥ 1.7 cm
- LV end-diastolic volume ≥ 150 mL/m2
- LV ejection fraction < 20%

Obliterative cardiomyopathy

- This is a rare form of restrictive cardiomyopathy, involving the endocardium of one or both ventricles; it is characterized by thrombosis and fibrosis, with gradual obliteration of the ventricular cavities by fibrous tissue .
- The mitral and tricuspid valves become regurgitant.
- Heart failure and pulmonary and systemic embolism are prominent features.
- can occur in eosinophilic leukemia and eosinophilic granulomatosis with polyangiitis (formerly known as Churg–Strauss syndrome).

- Mortality is high: 50% at 2 years. Anticoagulation and antiplatelet therapy are used, and diuretics may help symptoms of heart failure.
- Surgery (tricuspid and/or mitral valve replacement with decortication of the endocardium) may be helpful in selected cases.

Arrhythmogenic ventricular cardiomyopathy

- Arrhythmogenic ventricular cardiomyopathy (AVC) predominantly

affects the myocardium of the right ventricle.

- It is inherited in an autosomal dominant manner and has a prevalence of approximately 10 per 100 000.
- In some cases, the LV is also involved and this is associated with a poorer prognosis.
- The dominant clinical problems are ventricular arrhythmias, sudden

death and right-sided cardiac failure.

- The ECG typically shows a slightly broadened QRS complex and inverted T waves.
- Management is based on treating right-sided cardiac failure with diuretics and cardiac arrhythmias with β-blockers or, in patients at high risk of sudden death, an implantable defibrillator can be offered.

Takotsubo cardiomyopathy

Takotsubo cardiomyopathy (Takotsubo syndrome) is a form of acute left ventricular dysfunction characterized by dilatation of the left ventricular apex and adjacent myocardium, with associated left ventricular impairment.

The mechanism is poorly understood but may involve noradrenergic coronary vasoconstriction and acute left ventricular outflow obstruction.

It is often associated with acute environmental or emotional stress and presents with chest pain, breathlessness and sometimes cardiac failure.

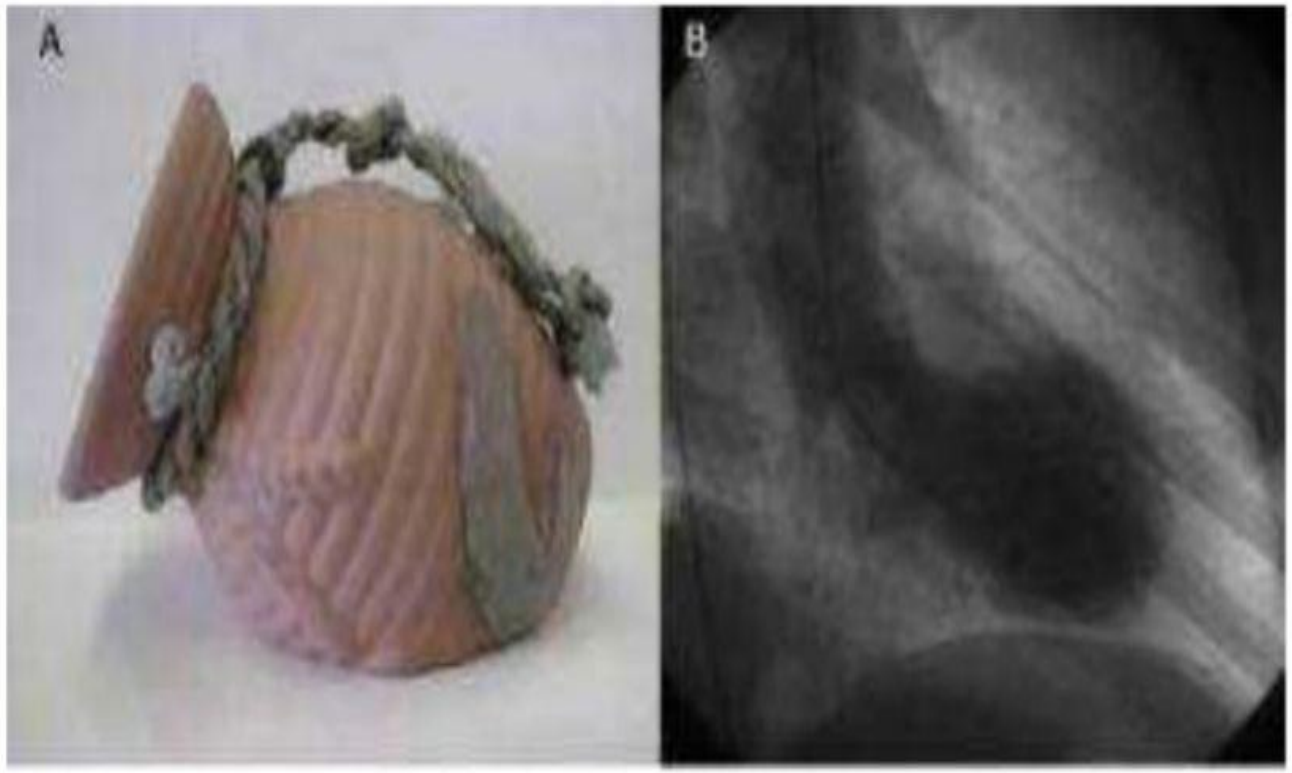
It occurs more frequently in women than in men.

- In terms of both symptoms and the ECG, the condition mimics acute

ST elevation acute coronary syndrome.

- The diagnosis is usually made at coronary angiography, when CAD is found to be absent or minimal.
- Echocardiography then shows characteristic ‘apical ballooning’ of the LV. The dilated apex and narrow outflow of the LV resemble a Japanese octopus trap, or takotsubo .

Comparison



Takatsubo = Octopus trap

- Left ventricular dysfunction usually recovers within 4–5 days, although this can take weeks in some cases.
- Treatment is with a β-blocker, to prevent arrhythmia, and an ACE inhibitor, to treat left ventricular dysfunction. These drugs are continued only until cardiac function has recovered.


Thank
you