MYOCARDIAL DISEASE

Myocardial disease, is a disaese of the myocardium that is not due to an ischaemic, valvular or hypertensive heart disease, It may be caused by:

- an acute or chronic inflammatory pathology (myocarditis)
- idiopathic myocardial disease (cardiomyopathy).

Myocarditis

- This is an acute inflammatory condition that can have an infectious, toxic or autoimmune aetiology.
- Myocarditis can complicate many infections in .\ which inflammation may be due directly to infection of the myocardium or the effects of circulating toxins.
- Viral infections are the most common causes, such as Coxsackie and influenza A and B viruses.
- Myocarditis may occur several weeks after the -
- initial viral symptoms and susceptibility is increased by corticosteroid treatment, immunosuppression, radiation, previous myocardial damage and exercise.

SOME BACTERIAL AND PROTOZOAL INFECTIONS
TOXIC AETIOLOGIES INCLUDE DRUGS
AUTOIMMUNE CONDITIONS SUCH AS SYSTEMIC
LUPUS ERYTHEMATOSUS AND RHEUMATOID
ARTHRITIS ARE ASSOCIATED WITH MYOCARDITIS

Pathology

- In the acute phase myocarditic hearts are flabby with focal haemorrhages;
- in chronic cases they are enlarged and hypertrophied.
- Histologically an inflammatory infiltrate is present
- lymphocytes predominating in viral causes;
- Polymorphonuclear cells in bacterial causes; —
- eosinophils in allergic and hypersensitivity causes

Clinical features

- Myocarditis may be an acute or chronic process;
 - its clinical presentations range from an asymptomatic state associated with limited and focal inflammation
 - to fatigue, palpitations, chest pain, dyspnoea and fulminant congestive cardiac failure due to diffuse myocardial involvement.
- Physical examination includes soft heart sounds, a prominent third sound and often a tachycardia.
 - A pericardial friction rub may be heard. —

Investigations

Chest X-ray may show some cardiac enlargement, depending on the stage and virulence of the disease.

ECG demonstrates ST- and T wave abnormalities and arrhythmias.

Heart block may be seen with diphtheritic myocarditis, Lyme disease — and Chagas' disease.

Cardiac enzymes are elevated TROPONIN . •

Viral antibody titres may be increased. •

However, since enteroviral infection is common in the general — population, the diagnosis depends on the demonstration of acutely rising titres.

Echocardiography may reveal left ventricular dysfunction that is sometimes regional (due to focal myocarditis)

Endomyocardial biopsy may show acute inflammation

Viral RNA can be measured from biopsy material using polymerase chain reaction (PCR).

BIOPSY

Treatment

- The underlying cause must be identified, treated, eliminated or avoided.
- Bed rest is recommended in the acute phase of the illness and athletic activities should be avoided for 6 months.
- Heart failure should be treated conventionally with the use of diuretics, ACE inhibitors/ARB, beta-blockers, spironolactone ± digoxin.
- Antibiotics should be administered immediately where appropriate.
- NSAIDs are contraindicated in the acute phase of the illness but may be used in the late phase.
- The use of corticosteroids is controversial and no studies have demonstrated an improvement in left ventricular ejection fraction or survival following their use.
- The administration of high-dose intravenous immunoglobulin on the other hand appears to be associated with a more rapid resolution of the left ventricular dysfunction and improved survival

CARDIOMYOPATHY

cardiomyopathies are defined as "a • heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilatation and are due to a variety of causes that frequently are genetic."

CARDIOMYOPATHY

1-DILATED CARDIOMYOPATHY

THIS CONDITION IS CHARACTERISED BY DILATATION AND IMPAIRED CONTRACTION OF THE LEFT (AND SOMETIMES THE RIGHT) VENTRICLE; LEFT VENTRICULAR MASS IS INCREASED BUT WALL THICKNESS IS NORMAL OR REDUCED

2-HYPERTROPHIC CARDIOMYOPATHY

THIS IS THE MOST COMMON FORM OF CARDIOMYOPATHY, WITH A PREVALENCE OF APPROXIMATELY 100 PER 100 000, AND IS CHARACTERISED BY INAPPROPRIATE AND ELABORATE LEFT VENTRICULAR HYPERTROPHY WITH MALALIGNMENT OF THE MYOCARDIAL FIBRES. THE HYPERTROPHY MAY BE GENERALISED OR CONFINED LARGELY TO THE INTERVENTRICULAR SEPTUM (ASYMMETRIC SEPTAL HYPERTROPHY

3-RESTRICTIVE CARDIOMYOPATHY IN THIS RARE CONDITION, VENTRICULAR FILLING IS IMPAIRED BECAUSE THE VENTRICLES ARE 'STIFF' (FIG. 18.105). THIS LEADS TO HIGH ATRIAL PRESSURES WITH ATRIAL HYPERTROPHY, DILATATION AND LATER ATRIAL FIBRILLATION. AMYLOIDOSIS IS THE MOST COMMON CAUSE OF RESTRICTIVE CARDIOMYOPATHY IN THE UK

DILATED CARDIOMYOPATHY (DCM)

DCM has a prevalence of 1 in 2500 and is • characterized by dilatation of the ventricular chambers and systolic dysfunction with preserved wall thickness.

- Famelial whichis autosomal domenant
- Sporadic DCM can be caused by multiple conditions:
 - myocarditis Coxsackie, adenoviruses, erythroviruses, HIV, bacteria, fungae, mycobacteria, parasitic (Chagas' disease)
- toxins alcohol, chemotherapy, metals (cobalt, lead, mercury, arsenic)
 - autoimmune
 - endocrine -
 - neuromuscular. —

Clinical features

DCM can present with heart failure, cardiac • arrhythmias, conduction defects, thromboembolism or sudden death

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Investigations

- **Chest X-ray** demonstrates generalized cardiac enlargement.
- **ECG** may demonstrate diffuse non-specific ST segment and T wave changes.
 - Sinus tachycardia, conduction abnormalities and arrhythmias (i.e. atrial fibrillation, ventricular premature contractions or ventricular tachycardia) are also seen.
 - **Echocardiogram** reveals dilatation of the left and/or right ventricle with poor global contraction function .
 - **Cardiac MR** may demonstrate other aetiologies of left ventricular dysfunction (e.g. previous myocardial infarction) or demonstrate abnormal myocardial fibrosis .
 - **Coronary angiography** whenever there is evidence of ischemia
 - **Biopsy** is generally not indicated outside specialist care. •

Treatment

- Treatment consists of the conventional management of heart failure with the option of cardiac resynchronization therapy and ICDs in patients with NYHA III/IV grading.
 - Cardiac transplantation is appropriate for certain patients.

CLINICAL FEATURES OF HYPERTROPHIC CARDIOMYOPATHY SYMPTOMS

ANGINA ON EFFORT DYSPNOEA ON EFFORT SYNCOPE ON EFFORT SUDDEN DEATH

SIGNS

JERKY PULSE*

PALPABLE LEFT VENTRICULAR HYPERTROPHY

DOUBLE IMPULSE AT THE APEX (PALPABLE FOURTH HEART SOUND

DUE TO LEFT ATRIAL HYPERTROPHY)

MID-SYSTOLIC MURMUR AT THE BASE*

PANSYSTOLIC MURMUR (DUE TO MITRAL REGURGITATION) AT THE

APEX

RISK FACTORS FOR SUDDEN DEATH IN HYPERTROPHIC CARDIOMYOPATHY•

A history of previous cardiac arrest or sustained ventricular tachycardia •

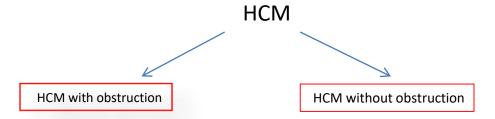
Recurrent syncope •

An adverse genotype and/or family history •

Exercise-induced hypotension •

Multiple episodes of non-sustained ventricular tachycardia on ambulatory ECG • monitoring

Marked increase in left ventricular wall thickness.



BB .\
BB +/- disopyramide if obstruction persist .\(\text{`Y} \)
Diuretics for congestive symptom .\(\text{`Y} \)
Verapamide in stead of disopyramide .\(\text{`Surgical myectomy or alcohol septal ablation } \text{.0} \)
Dual chamber cardiac pacing .\(\text{`1} \)

BB &/or verapamil .\
Diuretics for congestion .\
Transplantation .\(^{\mathcal{T}}

SPECIFIC DISEASES OF HEART MUSCLE

- 1-INFECTIONS VIRAL, E.G. COXSACKIE A AND B, INFLUENZA, HIV BACTERIAL, E.G. DIPHTHERIA, *BORRELIA BURGDORFERI* PROTOZOAL, E.G. TRYPANOSOMIASIS
- 2-ENDOCRINE AND METABOLIC DISORDERS E.G. DIABETES, HYPO- AND HYPERTHYROIDISM, ACROMEGALY, CARCINOID SYNDROME, PHAEOCHROMOCYTOMA, INHERITED STORAGE DISEASES
- 3-CONNECTIVE TISSUE DISEASES E.G. SYSTEMIC SCLEROSIS, SYSTEMIC LUPUS ERYTHEMATOSUS (SLE), POLYARTERITIS NODOSA
- 4-INFILTRATIVE DISORDERS E.G. HAEMOCHROMATOSIS, HAEMOSIDEROSIS, SARCOIDOSIS, AMYLOIDOSIS
- 5-TOXIN E.G. DOXORUBICIN, ALCOHOL, COCAINE, IRRADIATION
- 5-NEUROMUSCULAR DISORDERS E.G. DYSTROPHIA MYOTONICA, FRIEDREICH'S
- **ATAXIA**
- 7-IDIOPATHIC
- 8-FAMELIAL

Peri-partum cardiomyopathy

- This rare condition affects women in the last trimester of pregnancy or within 5 months of delivery.
- It presents as a dilated cardiomyopathy, is more common in obese, multiparous women over 30 years old and is associated with preeclampsia.
- Nearly half of patients will recover to normal function within 6 months but in some patients it can causes progressive heart failure and sudden death.

Alcohol and tachycardia C M P