MEDICINE

432 Team

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Extra: Myocardial and pericardial Diseases

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COLOR GUIDE: • Females' Notes • Males' Notes • Important • Additional

Objectives

Not Given

Myocardial disease:

Myocardial disease that is not due to ischaemic, valvular or hypertensive heart disease, or a known infiltrative, metabolic/ toxic or neuromuscular disorder may be caused by:

- An acute or chronic inflammatory pathology (myocarditis)
- Idiopathic myocardial disease (cardiomyopathy)

Myocarditis

Acute inflammation of the myocardium has many causes. Establishment of a definitive aetiology with isolation of viruses or bacteria is difficult in routine clinical practice.

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.

Investigations

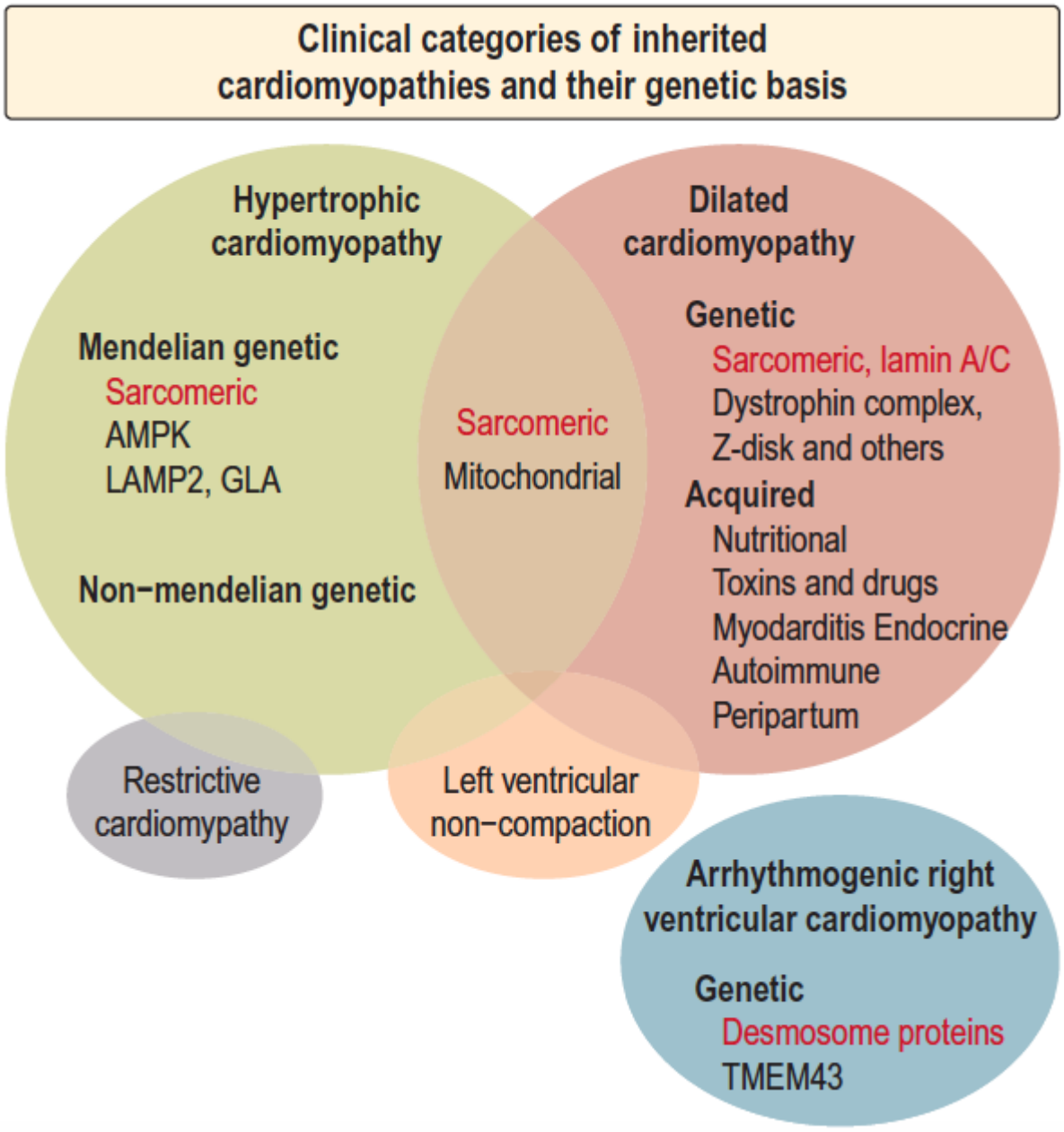
- Chest X-ray may show some cardiac enlargement, depending on the stage and virulence of the disease.
- ECG
- Cardiac enzymes are elevated.
- Viral antibody titres may be increased
- Endomyocardial biopsy may show acute inflammation but false negatives are common by conventional criteria. Biopsy is of limited value outside specialized units.
- Viral RNA.

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.

Cardiomyopathy

Cardiomyopathies are a group of diseases of the myocardium that affect the mechanical or electrical function of the heart. They are frequently genetic and may produce inappropriate ventricular hypertrophy or dilatation and can be primarily a cardiac disorder or part of a multisystem disease. Nevertheless, there is considerable heterogeneity and overlap between the conditions. Abnormal myocardial function produces systolic or diastolic heart failure; abnormal electrical conduction results in cardiac arrhythmias and sudden cardiac death.



I. Hypertrophic cardiomyopathy (HCM)

HCM includes a group of inherited conditions that produce hypertrophy of the myocardium in the absence of an alternate cause (e.g. aortic stenosis or hypertension). It is the most common cause of sudden cardiac death in young people and affects 1 in 500 of the population. The majority of cases are familial autosomal dominant, due to mutations in the genes encoding sarcomeric proteins. The most common causes of HCM are mutations of the β-myosin heavy chain MYH7 and myosin-binding protein C MYBPC3.

Symptoms:

- Many are asymptomatic and are detected through family screening of an affected individual or following a routineECG examination.
- Chest pain, dyspnoea, syncope or pre-syncope (typically with exertion), cardiac arrhythmias and sudden death are seen.
- Sudden death occurs at any age but the highest rates (up to 6% per annum) occur in adolescents or young adults. Risk factors for sudden death are discussed below.
- Dyspnoea occurs due to impaired relaxation of the heart muscle or the left ventricular outflow tract obstruction that occurs in some patients. The systolic cavity remains

Investigations:

- ECG
- Echocardiography
- Cardiac MR
- Genetic analysis

Treatment:

The management of HCM includes treatment of symptoms and the prevention of sudden cardiac death in the patient and relatives.

II. Arrhythmogenic (right) ventricular cardiomyopathy (AVC)

AVC is an uncommon (1 in 5000 population) inherited condition that predominantly affects the right ventricle with fatty or fibro-fatty replacement of myocytes, leading to segmental or global dilatation has been reported in up to 75% of cases. The fibro-fatty replacement leads to ventricular arrhythmia and risk of sudden death in its early stages, and right ventricular or biventricular failure in its later stages.

Clinical features:

Most patients are asymptomatic. Symptomatic ventricular arrhythmias, syncope or sudden death occur.

Investigations:

- ECG
- Echocardiography
- Cardiac MR
- Genetic analysis

Treatment:

Beta-blockers are first-line treatment for patients with nonlife- threatening arrhythmias. Amiodarone or sotalol are used for symptomatic arrhythmias but for refractory or lifethreatening arrhythmias an ICD is required.

III. 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. Familial DCM is predominantly autosomal dominant and can be associated with over 20 abnormal loci and genes. Many of these are genes encoding cytoskeletal or associated myocyte proteins (dystrophin in X-linked cardiomyopathy; actin, desmin, troponin T, beta myosin heavy chain, sarcoglycans, vinculin and lamin a/c in autosomal dominant DCM). Many of these have prominent associated features such as skeletal myopathy or conduction system disease and therefore differ from the majority of cases of DCM.

Clinical features:

DCM can present with heart failure, cardiac arrhythmias, conduction defects, thromboembolism or sudden death. Increasingly, evaluation of relatives of DCM patients is allowing identification of early asymptomatic disease, prior to the onset of these complications.

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.

PERICARDIAL DISEASE

The pericardium acts as a protective covering for the heart. It consists of an outer fibrous pericardial sac and an inner serous pericardium made up of the inner visceral epicardium that lines the heart and great vessels and its reflection the outer parietal pericardium that lines the fibrous sac. The normal amount of pericardial fluid is 20–49 mL that lubricates the surface of the heart. Presentations of pericardial disease include:

- Acute and relapsing pericarditis
- Pericardial effusion and cardiac tamponade
- Constrictive pericarditis.

Acute pericarditis

This refers to inflammation of the pericardium. Classically, fibrinous material is deposited into the pericardial space and pericardial effusion often occurs. Acute pericarditis has numerous aetiologies (Table 14.49) although in most cases a cause is not identified (idiopathic).

Table 14.49 Aetiology of pericarditis

I.	Infectious pericarditis
	Viral (Coxsackievirus, echovirus, mumps, herpes, HIV)
	Bacterial (*Staphylococcus, Streptococcus, Pneumococcus, Meningococcus, Haemophilus influenzae, Mycoplasmosis, Borreliosis, Chlamydia*)
	Tuberculous
	Fungal (*Histoplasmosis, Coccidioidomycosis, Candida*)
II.	Post-myocardial infarction pericarditis
	Acute myocardial infarction (early)
	Dressler's syndrome (late)
III.	Malignant pericarditis
	Primary tumours of the heart (mesothelioma)
	Metastatic pericarditis (breast and lung carcinoma, lymphoma, leukaemia, melanoma)
IV.	Uraemic pericarditis
V.	Myxoedematous pericarditis
VI.	Chylopericardium
VII.	Autoimmune pericarditis
	Collagen-vascular (rheumatoid arthritis, rheumatic fever, systemic lupus erythematosus, scleroderma)
	Drug-induced (procainamide, hydralazine, isoniazid, doxorubicin, cyclophosphamides)
VIII.	Post-radiation pericarditis
IX.	Post-surgical pericarditis
	Post-pericardiotomy syndrome
X.	Post-traumatic pericarditis
XI.	Familial and idiopathic pericarditis

Clinical features:

Pericardial inflammation produces sharp central chest pain exacerbated by movement, respiration and lying down. It is typically relieved by sitting forward. It may be referred to the neck or shoulders. The main differential diagnoses are angina and pleurisy. The classical clinical sign is a pericardial friction rub occurring in three phases corresponding to atrial systole, ventricular systole and ventricular diastole. It may also be heard as a biphasic 'to and fro' rub. The rub is heard best with the diaphragm of the stethoscope at the lower left sternal edge at the end of expiration with the patient leaning forward. There is usually a fever, leucocytosis or lymphocytosis when pericarditis is due to viral or bacterial infection, rheumatic fever or myocardial infarction. Features of a pericardial effusion may also be present. Large pericardial effusion can compress adjacent bronchi and lung tissue and may cause dyspnoea.

Investigations:

ECG is diagnostic. Chest X-ray may demonstrate cardiomegaly (in cases with an effusion) which should be confirmed with echocardiography. CT and cardiac MR may be helpful for in cases with thickened (>4 mm) or inflamed (abnormal delayed enhancement) pericardium.

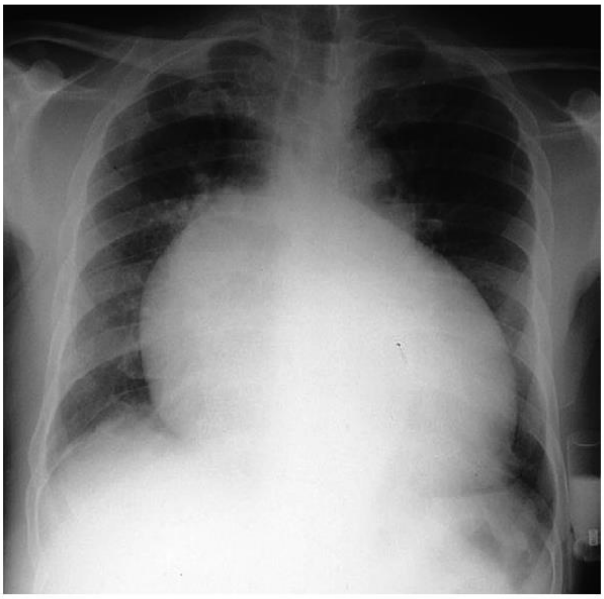


Figure 14.114 Chest X-ray showing a pericardial effusion; the heart appears globular.

Treatment

If a cause is found, this should be treated.
Bed rest and oral NSAIDs (high-dose aspirin indometacin or ibuprofen) are effective in most patients. Aspirin is the drug of choice for patients with a recent myocardial infarction.