Cardiomyopathy

Definition

- Disorders of the cardiac muscle of unknown aetiology

Specific types

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

Infective

- Viral eg. Coxackie virus (dilated)
- Tuberculosis (restrictive)
- HIV/AIDS (dilated)
- Chagas disease (restrictive) - South America
- Hypersensitivity 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 tendency - large calibre

Types Hypertrophic Cardiomyopathy

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

Myocardial Disarray

Normal Muscle Structure (Parallel alignment) Myocardial Disarray (Disorganised alignment)

Symptoms

- Dyspnoea
- Angina
- Palpitation
- Dizzy spells
- Syncope
CARDIOMYOPATHY

Cardiomyopathies are diseases whose dominant feature is direct involvement of the heart muscle itself.

In this classification, the cardiomyopathies were defined as "diseases of the myocardium associated with cardiac dysfunction." They were classified according to anatomy and physiology into the following types, each of which has multiple different causes:

- Dilation and impaired contraction of one or both ventricles
- Impaired systolic function
- Dilation cardiomyopathy
- Hypertrophic cardiomyopathy
- Restrictive cardiomyopathy
- Arrhythmogenic right ventricular cardiomyopathy
- Unclassified cardiomyopathies (this includes endomyocardial fibroelastosis and ventricular non-compaction)

PATHOPHYSIOLOGY

Depletion of myocardial energy stores: reduced myocardial levels of creatine, phosphocreatine, and adenosine triphosphate (ATP), and diminished activity of the Na-K-ATPase pump

Myocardial ischemia: Abnormalities in subendocardial to subepicardial flow ratios and impaired coronary flow have been found in tachycardia mediated cardiomyopathy. The impaired coronary blood flow occurs in association with elevation in cardiac metabolic demands, possibly related to coronary vasoconstriction.

Abnormal calcium handling and beta adrenergic responsiveness

Oxidative stress and injury

Is myocarditis the cause of Idiopathic DCM?

- Around 15 percent of patients with myocarditis progress to DCM
- Only around 10 percent (or less) of patients with DCM have biopsy evidence of myocarditis

Although there is the presence of high antibody viral titers, viral-specific RNA sequences, and apparent viral particles in patients with “idiopathic” DCM, the more rigorous technique of polymerase chain reaction generally has not confirmed the presence of viral remnants in the myocardium of most cardiomyopathy patients.
ALCOHOLIC CARDIOMYOPATHY

The pathophysiology of ACM is complex and may involve cell death (possibly due to apoptosis) and changes in many aspects of myocyte function. Changes in mitochondrial, sarcoplasmic, and extracellular matrix components may contribute to ACM.

LABS

The consumption of alcohol may result in myocardial damage by three basic mechanisms:

1. A presumed direct toxic effect of alcohol or its metabolites;
2. Nutritional effects, most commonly in association with thiamine deficiency that leads to beriberi;
3. A generalized cardiomegaly.

Failure supervenes in these patients, usually in the absence of nutritional deficiencies.

Echocardiography

Echocardiography reveals increased end-diastolic and end-systolic left ventricular volumes, reduced cardiac function in early left ventricular inflow, and reduced diastolic function. It can also reveal the presence of valvular abnormalities, pericardial effusion, and areas of abnormal wall motion.

Histological specimens obtained by right ventricular endomyocardial biopsy usually reveal normal vessels, although coronary vasodilatory capacity may be impaired. In patients with idiopathic DCM, the coronary blood flow may be normal or increased, and the arteriolar resistances may be normal or increased.

Endomyocardial Biopsy

The examination may be of particular value in excluding the presence of myocarditis on biopsy. Advanced age, specific endomyocardial biopsy features (such as loss of intracellular myofilaments), poor R wave progression, and intraventricular conduction abnormalities, especially left bundle branch block, may help to identify patients more likely to have myocarditis on biopsy.

There are also disease-independent predictors of survival.

Cardiac catheterization and angiography

Only certain patients with DCM require cardiac catheterization for therapeutic purposes. Some patients with known or suspected coronary artery disease may benefit from arteriography, especially those with chest pain and a suspicion of ischemic disease or patients thought to have a treatable systemic disease such as sarcoidosis or hemochromatosis, where myocardial biopsy is an important part of the catheterization procedure.

The sensitivity of endomyocardial biopsy is at best around 35%.

These prognostic determinants assume that the cause of the myocardial dysfunction cannot be treated.

There are also disease-independent predictors of survival.

A variety of clinical phenotypes and patterns of presentation of ACM have been identified.
Management

Because the cause of idiopathic DCM, by definition, is unknown, specific therapy is not possible. Treatment, therefore, is for heart failure.

- Regular physical exercise (as tolerated) increases exercise capacity by improving endothelial dysfunction and augmenting blood flow in skeletal muscles. Exercise training improves exercise tolerance and LV function. This is accompanied by a decrease in biventricular oxidative metabolism and enhanced forward work efficiency. Therefore, exercise training elicits an energetically favorable improvement in myocardial function and exercise tolerance in patients with DCM.

- Diet- low salt, low fat, heart healthy

- Only cardiac transplantation and specific pharmacological therapy (the vasodilators enalapril or hydralazine plus nitrates, the beta-adrenoceptor blocker carvedilol, and the aldosterone receptor blocker spironolactone) have been shown to prolong life.

- DUAL CHAMBER PACING.

  - This has been used in some patients with DCM and intact atrioventricular conduction. Changes the sequence of ventricular depolarization, reduces functional mitral regurgitation, improves clinical status, and some symptomatic and hemodynamic improvement has been reported. It is more effective in patients with atrial fibrillation. Atrial pacing using two pacemaker leads, one in the right atrium and the other in the coronary sinus, may elicit atrial fibrillation in the absence of atrial flutter.

- SURGICAL TREATMENT.

  - Mitral annuloplasty or replacement of regurgitant valves has been attempted in some patients with DCM and prominent atrioventricular valvular regurgitation.

  - The results of surgical repair are usually not satisfactory because of the degree of remodeling, valve distortion, and the often pre-existing heart failure. However, if the mitral valve is moderately regurgitant and is not associated with mitral stenosis, a low threshold should be entertained for valve repair (mitral valve annuloplasty).

  - In appropriately selected patients, cardiac transplantation may be an effective alternative to medical therapy, with a 1-year survival rate of about 70%.

  - Surgical interventions of the infrarenal aorta to revascularize the heart and augment cardiac output by cardiac transplantation have not been performed in the United States or Europe. However, cases of partial or total atrioventricular block (AVB) have been reported as complications of heart transplantation. Associated conduction disturbances and heart failure may limit the technical adequacy of the procedure.

- 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)

Restrictive Cardiomyopathy

- ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

  - This unique cardiomyopathy, which is also called arrhythmogenic right ventricular dysplasia (ARVD) is marked by scar-related left heart and right-sided ventricular dysfunction.

  - ARVD is associated with severe ventricular arrhythmias and the risk of sudden death.

  - In about one third of the cases there is abnormal dominant inheritance of the disease, and several distinct genetic mutations have been reported. One variant, found on the Greek island of Naxos, is inherited as a recessive trait but with a high degree of penetrance.

  - Nonsustained and sustained ventricular tachycardia are common. Patients with ARVD have right ventricular outflow tract hypertrophy, although the left ventricle is morphologically normal.

  - Magnetic resonance imaging (MRI) shows promise for identifying patients with ARVD. It may also help in identifying arrhythmias that are not visible on electrocardiogram.

  - Cardiac arrhythmia is the result of abnormalities of ventricular myocardium, sympathetic innervation, or impaired pigmentation.