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Cardiomyopathy

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Cardiomyopathy

- a condition *primarily* nvolving the myocardium
- *not* the result of congenital, acquired valvular, hypertensive, coronary arterial, or pericardial abnormalities
- 2 forms:
  - Primary type
  - Secondary type

Primary cardiomyopathy

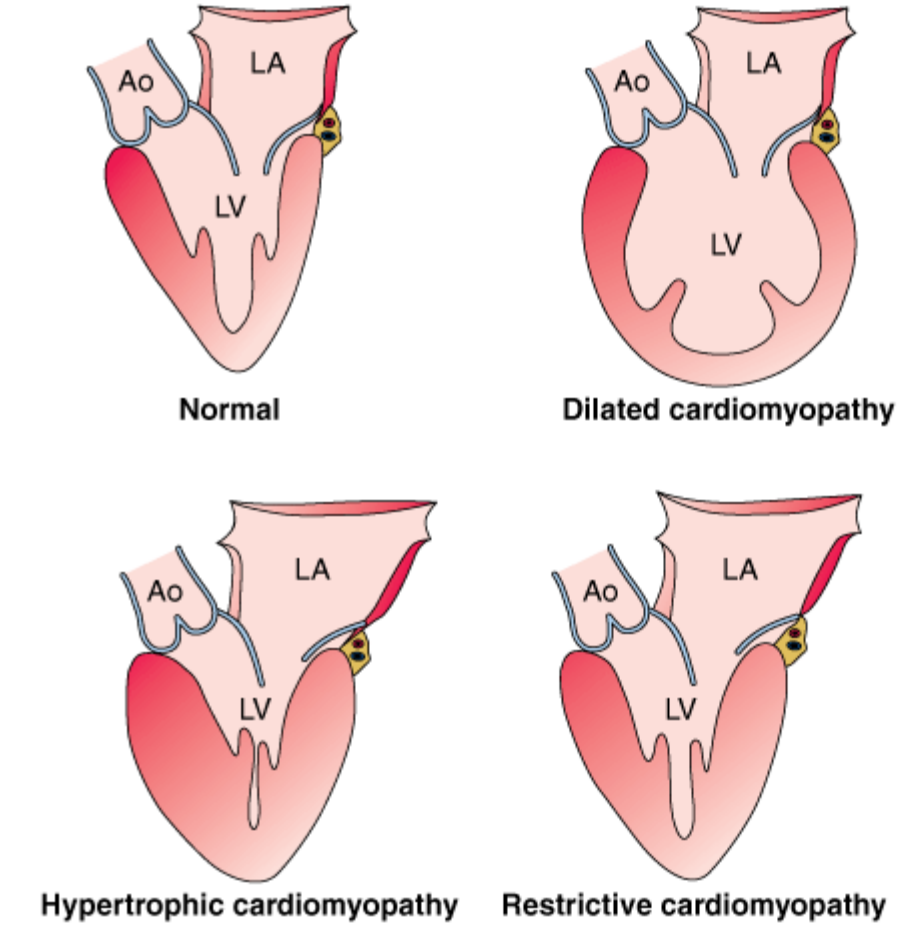
- consisting of heart muscle disease predominantly involving the myocardium and/or of unknown cause

Secondary cardiomyopathy

- consisting of myocardial disease of known cause or associated with a systemic disease such as amyloidosis or chronic alcohol use (

three morphologic types

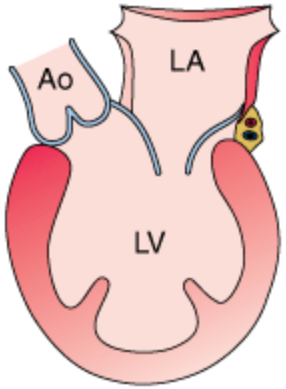
- dilated, restrictive, and hypertrophic



three morphologic types

- 1. **Dilated**: Left and/or right ventricular enlargement, impaired systolic function, congestive heart failure, arrhythmias, emboli
- 2. **Restrictive**: Endomyocardial scarring or myocardial infiltration resulting in restriction to left and/or right ventricular filling
- 3**. Hypertrophic**: Disproportionate left ventricular hypertrophy, typically involving septum more than free wall, with or without an intraventricular systolic pressure gradient; usually of a nondilated left ventricular cavity

Dilated cardiomyopathy



Dilated cardiomyopathy

- 1/3 of all cases
- LV and/or right ventricular (RV) systolic pump function is impaired, leading to progressive cardiac dilatation (remodeling)
- Symptoms of heart failure (HF) typically appear only after remodeling has been ongoing for some time (months or even years)

Dilated cardiomyopathy

- In most cases, no cause is apparent
- Familial
- end result of myocardial damage
- may be the late consequence of acute viral myocarditis
- most commonly becomes apparent clinically in the third or fourth decades
- reversible form of DCM may be found with alcohol abuse, thyroid disease, cocaine use and chronic uncontrolled tachycardia

Dilated cardiomyopathy

CLINICAL FEATURES

- Symptoms of left- and right-sided CHF usually develop gradually
- vague chest pain may be present
- Syncope due to arrhythmias and systemic embolism (often emanating from a ventricular thrombus) may occur

Dilated cardiomyopathy

PHYSICAL EXAMINATION

- Variable degrees of cardiac enlargement
- findings of Congestive heart failure
    - rales, edema
- pulse pressure is narrow
- jugular venous pressure is elevated

Dilated cardiomyopathy

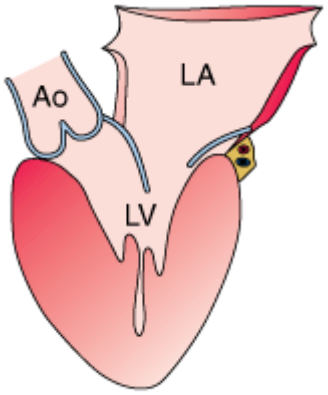
LABORATORY

- Chest Xray
  - enlargement of the cardiac silhouette due to LV dilatation
  - pulmonary vascular redistribution
  - alveolar edema
- ECG
  - Sinus tachycardia,atrial fibrillation, ventricular arrhythmias, left atrial abnormality, low voltage

Treatment

- Most patients die within 4 years
- ¼ have spontaneous improvement
- patients should be considered for chronic anticoagulation
- Standard therapy for heart failure
- Alcohol should be avoided - cardiotoxic effects

Hypertrophic cardiomyopathy



Hypertrophic cardiomyopathy

- characterized by LV hypertrophy, typically of a nondilated chamber, without obvious cause
- 1 in 500 of the general population
- pathophysiologic abnormality is *diastolic* dysfunction

Hypertrophic cardiomyopathy

- majority demonstrate a ventricular septum whose thickness is disproportionately increased when compared with the free wall
- half of all patients with HCM have a positive family history
- genetic testing may allow a definitive diagnosis of HCM

Hypertrophic cardiomyopathy

CLINICAL FEATURES

- Variable
- asymptomatic or mildly symptomatic
- first clinical manifestation may be SCD (sudden cardiac death)
    - occurring in children and young adults during or after physical exertion
    - most common cause of SCD in young competitive athletes

Hypertrophic cardiomyopathy

PHYSICAL EXAMINATION

- hallmark of obstructive HCM is a systolic murmur, which is typically harsh, diamond-shaped, and usually begins well after the first heart sound

Hypertrophic cardiomyopathy

LABORATORY

- ECG:
  - LV hypertrophy and widespread deep, broad Q waves
- Chest Xray
  - May be normal or mild increase in silhouette
- Echocardiogram
  - Mainstay in diagnosis
  - LV hypertrophy, often with the septum 1.3 times the thickness of the posterior LV free wall

Hypertrophic Cardiomyopathy

TREATMENT

- Dehydration avoided
- Diuretics used in caution
- Amiodarone appears to be effective in reducing the frequency of supraventricular as well as of life-threatening ventricular arrhythmias

Restrictive Cardiomyopathy

- abnormal diastolic function
- walls are excessively rigid and impede ventricular filling
- Myocardial involvement with *amyloid* is a common cause of secondary restrictive cardiomyopathy
- transplanted heart, in hemochromatosis, glycogen deposition, endomyocardial fibrosis, sarcoidosis, hypereosinophilic disease, and scleroderma

Restrictive Cardiomyopathy

CLINICAL FEATURES

- inability of the ventricles to fill limits cardiac output and raises filling pressures
- exercise intolerance and dyspnea
- dependent edema, ascites, and an enlarged, tender, and often pulsatile liver
- heart sounds may be distant, and third and fourth heart sounds are common

Restrictive Cardiomyopathy

LABORATORY

- ECG
  - low-voltage, nonspecific ST-T-wave abnormalities and various arrhythmias
- Echocardiography, CTI, and CMRI
  - symmetrically thickened LV walls and normal or slightly reduced ventricular volumes and systolic function