Cardiomyopathies

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= disease of heart muscle (diagnosed by endomyocardial biopsy using bioptome), but not the result of pericardial, hypertensive, congenital, valvular or ischemic diseases.

- ***Classification of cardiomyopathies:-***

1. ***Dilated cardiomyopathy (DCM)*** = most common, characterized by ventricular dilatation, contractile dysfunction, heart failure symptoms.
2. ***Hypertrophic cardiomyopathy (HCM)*** = characterized by inappropriate left ventricular hypertrophy (often asymmetrical interventricular septum), preserved or enhanced contraction (until late in the course)

- ***Restrictive cardiomyopathy =*** least common. Characterized by impaired diastolic filling.

- ***Specific cardiomyopathies:*** usually DCM pattern. eg. ischemic,valvular,hypertensive, inflammatory cardiomyopathies

- ***Arrhythmogenic right ventricular cardiomyopathy =*** progressive fibrofatty replacement of right ventricular myocardium associated with reentrant ventricular tachyarrhythmias of right ventricular origin (producing electrocardiographic left bundle branch block configuration of QRS complex) and sudden death. Familial common.

Dilated cardiomyopathy (DCM)

= final common pathway of myocardial damage due to cytotoxic, metabolic, immunological, familial, infectious mechanisms.

- Characterized by: cardiac enlargement, impaired systolic function, heart failure symptoms
- Treatment similar to heart failure

Hypertrophic cardiomyopathy (HCM)

= most common genetically transmitted cardiac disorder. Mutation of cardiac myosin-binding protein C gene.

- Usually diastolic dysfunction characterized by abnormal stiffness of left ventricle with resultant impaired ventricular filling → increase left ventricular end-diastolic pressure → pulmonary congestion and dyspnea
- Histology = myocardial hypertrophy and gross disorganization of muscle bundles and disarray of cells resulting in characteristic whorled pattern. Prominent fibrosis. Abnormal intramural coronary arteries with stenosis and thickening.

- Pressure gradient across left ventricular outflow tract (due to septal hypertrophy and abnormal location of mitral valve) by systolic anterior motion (SAM) of often elongated mitral valve leaflet against the hypertrophied septum.
- Impaired diastolic filling, due to fibrosis and cellular disorganization.
- Apical HCM = predominant involvement of apex, characterized by spade-like configuration of left ventricle, electrocardiographic giant T wave, no intraventricular pressure gradient, mild symptoms and usually benign course.

- ***Etiology =*** autosomal dominant mendelian-inherited disease or mutations of cardiac myosin heavy chain gene.
- ***Symptoms =*** majority asymptomatic. However, first manifestation may be sudden death (risk factors = young age, family history, abnormal blood pressure response to exercise, presence of severe symptoms, presence of nonsubstained ventricular tachycardia or conduction system disorders, marked hypertrophy and left atrial dilatation).
- Dyspnea (most common), angina, fatigue, dizziness, syncope, palpitation, paroxysmal nocturnal dyspnea, heart failure.

Treatment =

- avoid strenuous exercise
- beta blocker = mainstay
- calcium antagonist – improve diastolic filling and regional myocardial blood flow
- amiodarone – for supraventricular and ventricular arrhythmias
- pacemaker, ICD, alcohol infusion into selectively catheterized septal artery
- surgery : myectomy, myotomy-myectomy, mitral valve replacement

Restrictive cardiomyopathy

- abnormal diastolic function
- ***etiology*** = myocardial fibrosis, infiltration, endomyocardial scarring, idiopathetic.
- ***symptoms*** = exercise intolerance, weakness, dyspnea, exertional chest pain, peripheral edema, ascite, anasarca
- ***treatment*** = symptomatic and poor prognosis