Cardiomyopathies

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= **disease of heart muscle due to genetic defects, cardiac myocyte injury, or infiltration of myocardial tissues.**

Classification of cardiomyopathies

- ***Functional classification:***

1. Dilated cardiomyopathy
2. Hypertrophic cardiomyopathy
3. Restrictive cardiomyopathy
4. Arrhythmogenic right ventricular cardiomyopathy

- ***Etiologic classification:***

1. Primary cardiomyopathy, idiopathic (50%)
2. Secondary cardiomyopathy, due to:

   myocarditis, coronary artery disease, infiltrations, hypertension, connective tissue disease, toxins, drugs, valvular heart disease, endocrine disease etc.

- ***Specific classification:***

  ischemic cardiomyopathy
  valvular cardiomyopathy
  hypertensive cardiomyopathy
  inflammatory cardiomyopathy (eg myocarditis)
  metabolic cardiomyopathy
  peripartum cardiomyopathy

Dilated cardiomyopathy (DCM)

- Most common
- Characterized by heart dilatation with systolic dysfunction
- Causes = genetic, viral, immune, toxin etc.
- Symptoms = fatigue, weakness, thromboembolism, left heart failure
- Treatment = similar to heart failure

Hypertrophic cardiomyopathy (HCM)

- Characterized by inappropriate left ventricular hypertrophy (often asymmetrical interventricular septum) not due to another cardiac or systemic conditions such as aortic stenosis, hypertension etc.
- 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→ increased intraventricular pressures → heart failure.
- Diastolic dysfunction due to ventricular hypertrophy, scarring, fibrosis, disorganized architecture.
- Regional myocardial ischemia in absence of coronary artery disease, due to microvasculature abnormalities secondary to increased ventricular mass.
- 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, left heart failure.

Treatment =

- avoid strenuous exercise
- beta blocker = mainstay
- calcium antagonist – improve diastolic filling and regional myocardial blood flow
- antifailure drugs if heart failure
- amiodarone – for supraventricular and ventricular arrhythmias
- warfarin if atrial fibrillation
- pacemaker, ICD, alcohol infusion into selectively catheterized septal artery
- surgery : myectomy, myotomy-myectomy, mitral valve replacement (for severe drug refractory heart failure with outflow gradient $\geq$ 50 mmHg)

Restrictive cardiomyopathy

- least common
- abnormal diastolic function due to stiffness of ventricles
- ***etiology*** = amyloidosis (most common), myocardial fibrosis, infiltration, endomyocardial scarring, idiopathetic.
- ***symptoms*** = exercise intolerance, weakness, dyspnea, exertional chest pain, peripheral edema, ascite, anasarca
- ***treatment*** = symptomatic and poor prognosis

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.