MSD MANUAL
Professional Version

Overview of Cardiomyopathies

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A cardiomyopathy is a primary disorder of the heart muscle. It is distinct from structural cardiac disorders such as coronary artery disease, valvular disorders, and congenital heart disorders. Cardiomyopathies are divided into 3 main types based on the pathologic features (see figure Forms of Cardiomyopathy):

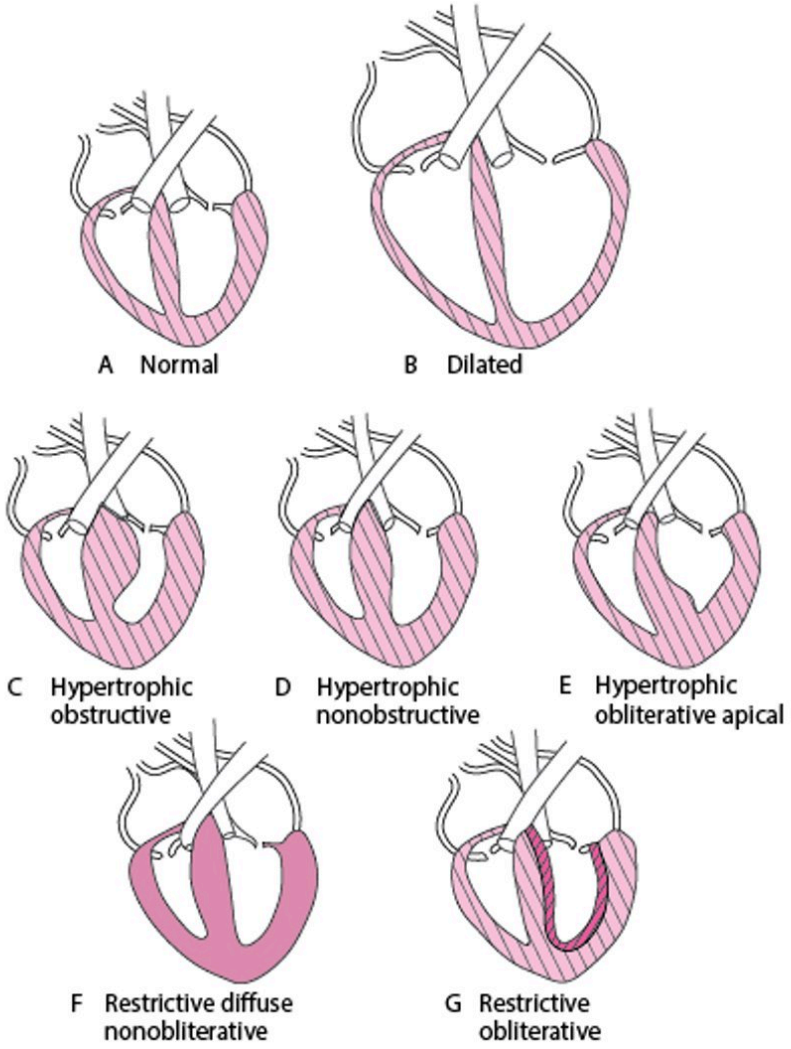
- Dilated
- Hypertrophic
- Restrictive

The term ischemic cardiomyopathy refers to the dilated, poorly contracting myocardium that can occur in patients with severe coronary artery disease (with or without areas of infarction). It is *not* classically considered to be in the above-listed categories because it does not describe a primary myocardial disorder.

Manifestations of cardiomyopathies are usually those of heart failure and vary depending on whether there is systolic dysfunction, diastolic dysfunction, or both. Some cardiomyopathies may also cause chest pain, syncope, arrhythmias, or sudden death.

Evaluation typically includes family history, blood tests, ECG, chest x-ray, echocardiography, and cardiac MRI. Some patients require endomyocardial biopsy. Other tests are done as needed to determine the cause. Treatment depends on the specific type and cause of cardiomyopathy (see table Diagnosis and Treatment of Cardiomyopathies).

Forms of Cardiomyopathy


A Normal
B Dilated
C Hypertrophic obstructive
D Hypertrophic nonobstructive
E Hypertrophic obliterative apical
F Restrictive diffuse nonobliterative
G Restrictive obliterative

TABLE

Diagnosis and Treatment of Cardiomyopathies

Feature or Method	Dilated Cardiomyopathy	Hypertrophic Cardiomyopathy	Restrictive Cardiomyopathy
Pathophysiology	Systolic dysfunction	Diastolic dysfunction ± outflow obstruction	Diastolic dysfunction
Clinical findings	LV and RV failure, sudden death Cardiomegaly Functional AV valve regurgitation S3 and/or S4	Exertional dyspnea, angina, syncope, sudden death Systolic murmur ± mitral regurgitation murmur, S4 Bifid carotid pulse with a brisk upstroke and rapid downstroke	Exertional dyspnea and fatigue LV ± RV failure Functional AV valve regurgitation
ECG	Nonspecific ST- and T-wave abnormalities Q waves ± BBB	LV hypertrophy and ischemia Deep septal Q waves	LV hypertrophy or low QRS voltage
Echocardiography	Dilated hypokinetic ventricles ± mural thrombus Low EF and, frequently, functional AV valve regurgitation	Hypertrophied ventricle, high, normal or low EF ± mitral systolic anterior motion ± asymmetric hypertrophy ± LV gradient	Increased wall thickness ± cavity obliteration LV diastolic dysfunction
X-ray	Cardiomegaly Pulmonary venous congestion	No or mild cardiomegaly	No or mild cardiomegaly
Hemodynamics	Normal or high EDP, low EF, diffusely dilated hypokinetic ventricles ± AV valve regurgitation	High EDP ± outflow subvalvular gradient ± mitral regurgitation Normal or low CO	High EDP, dip and plateau diastolic LV pressure curve Normal or low CO

	Low CO		
Prognosis	20% mortality in first year, and about 10%/year thereafter	About 1% annual risk of sudden death	70% 5-year mortality
Treatment	Diuretics, ACE inhibitors, angiotensin II receptor blockers, ARNI, SGLT2 inhibitor, beta-blockers, spironolactone or eplerenone, digoxin, ICD, cardiac resynchronization therapy, anticoagulants	Beta-blockers, ± verapamil, disopyramide, mavacamten, septal myotomy, catheter alcohol ablation or some combination	Phlebotomy for hemochromatosis Endocardial resection Hydroxyurea for hypereosinophilia

ACE = angiotensin-converting enzyme; ARNI = angiotensin II receptor blocker and neprilysin inhibitor; AV = atrioventricular; BBB = bundle branch block; CO = cardiac output; EDP = end-diastolic pressure; EF = ejection fraction; ICD = implantable cardioverter-defibrillator ; LV = left ventricular; RV = right ventricular; S3 = 3rd heart sound; S4 = 4th heart sound; SGLT2 = sodium glucose cotransporter 2 protein; ± = with or without.