TABLE 10-2 CARDIOMYOPATHIES

DISORDER	DESCRIPTION AND CAUSE
Dilated cardiomyopathy	Dilation and impaired systolic function of the left or both ventricles
Familial (genetic)	Known or unknown genetic mutations
Nonfamilial	Viral myocarditis, nonviral infective myocarditis, idiopathic (immune) myocarditis
	Toxins (drugs, alcohol)
	Pregnancy (peripartum cardiomyopathy)
	Nutritional (thiamine deficiency [beriberi], vitamin C deficiency [scurvy], selenium deficiency)
	Endocrine (diabetes mellitus, hyperthyroidism, hypothyroidism, hyperparathyroidism, pheochromocytoma, acromegaly)
	Autoimmune (rheumatoid arthritis, systemic lupus erythematosus, dermatomyositis)
	Tachycardia induced
Hypertrophic cardiomyopathy	Left and/or right ventricular hypertrophy, often asymmetrical (usually more prominent hypertrophy of the interventricular septum)
Familial (genetic)	Mutations of sarcoplasmic proteins (several hundred described)
	Metabolic storage diseases of the myocyte
Restrictive cardiomyopathy	Restrictive filling of the ventricles; ventricles are usually small, atria are markedly enlarged
Familial (genetic)	Mutations of sarcomeric proteins
	Familial amyloidosis (transthyretin, apolipoprotein)
	Hemochromatosis
	Desminopathy, pseudoxanthoma elasticum, glycogen storage diseases
	Unknown genetic mutations
Nonfamilial	Amyloidosis, sarcoidosis, carcinoid, scleroderma
	Endomyocardial fibrosis (hypereosinophilic syndrome, idiopathic, chromosomal defect, drugs)
	Radiation, metastatic cancer, anthracycline toxicity
Arrhythmogenic right ventricular	Progressive fibrofatty replacement of the right and, to a lesser degree, left ventricular cardiomyopathy
Familial	Unknown gene mutation
	Mutations of intercalated disk protein, cardiac ryanodine receptor, transforming growth factor-β3
UNCLASSIFIED CARDIOMYOPATHIES	
Takotsubo (stress-induced) cardiomyopathy	Transient dilation and dysfunction of the distal parts of the left ventricle (apical ballooning) in the setting of a stressful situation; usually resolves within weeks
Left ventricular noncompaction	Characterized by prominent left ventricular trabeculae and deep intertrabecular recesses; familial in most cases, caused by arrest in the normal embryogenesis of the heart; apex and periapical regions of the left ventricle most affected; some patients remain asymptomatic, but others develop left ventricular dilation and systolic dysfunction
Cardiomyopathies associated with muscular dystrophies and neuromuscular disorders	Duchenne-Becker muscular dystrophy, Emery-Dreifuss muscular dystrophy, myotonic dystrophy, Friedreich's ataxia, neurofibromatosis, tuberous sclerosis
Ion channelopathies	Disorders caused by mutations in genes encoding ionic channel proteins; not considered cardiomyopathies because they are not associated with typical structural changes of the heart but rather manifest with electrical dysfunction; some classifications include these disorders as cardiomyopathies: long QT syndrome, short QT syndrome, Brugada syndrome, catecholaminergic polymorphic ventricular tachycardia

predominate, with jugular venous distention hepatomegaly, ascites, and peripheral edema.

Diagnosis

Standard diagnostic procedures include a chest radiograph, an electrocardiogram, serum markers, and echocardiography. The radiograph shows cardiomegaly, pulmonary venous congestion, and pleural effusions. The electrocardiogram may reveal enlargement of the heart chambers along with other nonspecific ST- and T-wave abnormalities. Serum B-type natriuretic peptide (BNP) levels are elevated.

Echocardiography provides a comprehensive evaluation of ventricular size and function and valvular function, and it can show a ventricular thrombus. Similar information can be obtained with MRI.

A complete work-up should rule out ischemic, valvular, and hypertensive heart disease as the cause of myocardial dysfunction, and it should include evaluation for potentially reversible causes of DCM (e.g., alcohol, nutritional deficiencies). Myocardial biopsy may be considered if the cause of DCM is in question. In patients with a strong family history, a referral for genetic testing should be considered.

Treatment

Potential reversible causes of DCM should be addressed (e.g., alcohol cessation, correction of nutritional deficiencies, removal of cardiotoxic agents). Treatment should follow current ACC/AHA recommendations for the management of left ventricular systolic dysfunction and include β-adrenergic blockers, angiotensin-converting enzyme inhibitors, aldosterone receptor blockers, and diuretics.

Patients with idiopathic DCM who have persistent, moderate to severe symptoms of heart failure and a QRS duration longer than 120 milliseconds may benefit from cardiac resynchronization therapy with a biventricular pacemaker. Survival of patients with a left ventricular ejection fraction less than 35% despite maximal medical management is improved with the use of implantable cardioverter-defibrillators (ICDs). Patients with limiting heart failure symptoms despite use of the previously described therapies may be considered for heart transplantation or support with a left ventricular assist device.

Prognosis

The prognosis of patients with DCM depends on the response to medical therapy. Some patients have a significant improvement