conduction defects are seen in myocarditis associated with Lyme disease.

Echocardiography is recommended in the initial diagnostic evaluation to identify ventricular remodeling, including increasing chamber size and ventricular systolic dysfunction. Cardiac MRI is a promising technique to detect myocardial inflammation and injury based on small, observational clinical studies.

Transvenous endomyocardial biopsy should be performed only when there is rapid deterioration of the clinical condition (level of evidence B). Histopathologic abnormalities such as infiltrating white cells (i.e., macrophages, lymphocytes, and eosinophils), evidence of myocardial damage, and a finding of interstitial fibrosis are used to establish acute myocarditis, but the determination is subject to significant intraobserver and interobserver variability, and the biopsy often does not provide a conclusive diagnosis. The endomyocardial biopsy is helpful in diagnosing giant cell myocarditis (i.e., multinucleated giant cells are seen) or hypersensitivity myocarditis (i.e., eosinophilic infiltrate is seen). The polymerase chain reaction can detect specific viral genomes in the myocardium.

Treatment

Supportive care is the mainstay of treatment. A few patients with fulminant or acute myocarditis require an intensive level of hemodynamic support and aggressive pharmacologic intervention similar to that for patients with advanced heart failure.

After initial hemodynamic stabilization, treatment should follow current American College of Cardiology and American Heart Association (ACC/AHA) recommendations for the management of left ventricular systolic dysfunction. Treatment includes β-adrenergic blockers, angiotensin-converting enzyme inhibitors, aldosterone receptor blockers, and diuretics.

No evidence-based guided therapy for viral myocarditis has been established. Clinical trials of various forms of antiviral or immunosuppressive therapy (e.g., prednisone, cyclosporine, azathioprine, intravenous immunoglobulin, interferon immunoadsorption) have not resulted in conclusive evidence of benefit. Treatment of nonviral myocarditis is aimed at eradication of the specific infectious agent. For Chagas disease, treatment with antiprotozoal therapy, if initiated early in the course of infection, may be beneficial.

Hypersensitivity myocarditis and myocarditis associated with toxins respond to withdrawal of the offending agent. Immunosuppressive therapy has been effective in giant cell myocarditis.

Prognosis

Understanding the natural history of myocarditis has been limited by its diverse clinical presentations and causes. It is thought that one third of the patients fully recover, one third of the patients have some sequelae in the form of left ventricular systolic dysfunction but are stable on medical therapy, and one third of patients progress to advanced heart failure. Patients who progress to chronic DCM have 5-year survival rates of less than 50%.

Cardiomyopathies

Cardiomyopathies are a heterogeneous group of diseases in which the major structural abnormality is limited to the myocardium. The four main cardiomyopathic groups are dilated, hypertrophic, restrictive, and arrhythmogenic right ventricular cardiomyopathy. Familial (genetic) and nonfamilial (acquired) forms of the diseases have been described.

Dilated Cardiomyopathy

Definition and Epidemiology

Cardiac enlargement and systolic dysfunction in DCM result from a wide spectrum of genetic, inflammatory, toxic, and metabolic causes (Table 10-2), although most cases are idiopathic. Abnormal loading conditions such as hypertension, valvular disease, or coronary artery disease can lead to similar structural and functional changes; these conditions are not considered to be part of the DCM group and are discussed elsewhere.

Most cases are thought to result from acute viral myocarditis, a process described earlier. Exposures to cardiac toxins such as chemotherapeutic agents, alcohol, cocaine, and radiation, along with deficiency of nutrients such as thiamine (causes beriberi), vitamin C (causes scurvy), carnitine, selenium, phosphate, and calcium, can cause DCM. Peripartum cardiomyopathy is a rare cause of DCM that can develop during the last month of pregnancy and up to 6 months after delivery.

The pathogenesis of this often life-threatening disease is not completely understood, and it is a diagnosis of exclusion. Risk factors include older maternal age, being African American, and having multiple pregnancies. Prolonged periods of supraventricular or ventricular tachycardia can lead to idiopathic DCM (i.e., tachycardia-induced cardiomyopathy). The structural and functional changes usually reverse after the rapid heart rhythm is controlled.

Familial forms of DCM may be responsible for 20% to 30% of cases. Specific mutations described involve genes that encode proteins of the sarcomere, cytoskeleton, nuclear membrane, and mitochondria; many mutations remain unknown. The mode of inheritance is typically autosomal dominant, but it can be an X-linked or mitochondrial pattern.

Pathology

Marked enlargement of all four cardiac chambers is typical of DCM, although the disease sometimes is limited to the left or right chambers. The dilation is out of proportion to the ventricular thickness. Histology reveals evidence of myocyte degeneration with irregular hypertrophy and atrophy of myofibers with often extensive interstitial and perivascular fibrosis.

Clinical Presentation

DCM usually manifests with symptoms of heart failure, including fatigue, weakness, dyspnea, and edema. In some patients, the presenting episode is related to arrhythmia or an embolic event. On physical examination, signs of decreased cardiac output are often found, including cool extremities, narrow pulse pressure, and tachycardia. The cardiac examination reveals a laterally displaced apex. An S_3 gallop is common, along with murmurs of mitral and tricuspid regurgitation. Pulmonary edema manifests as auscultatory crackles over the lung fields, and breath sounds may be diminished if there are pleural effusions. In some patients, the clinical features of right ventricular heart failure may