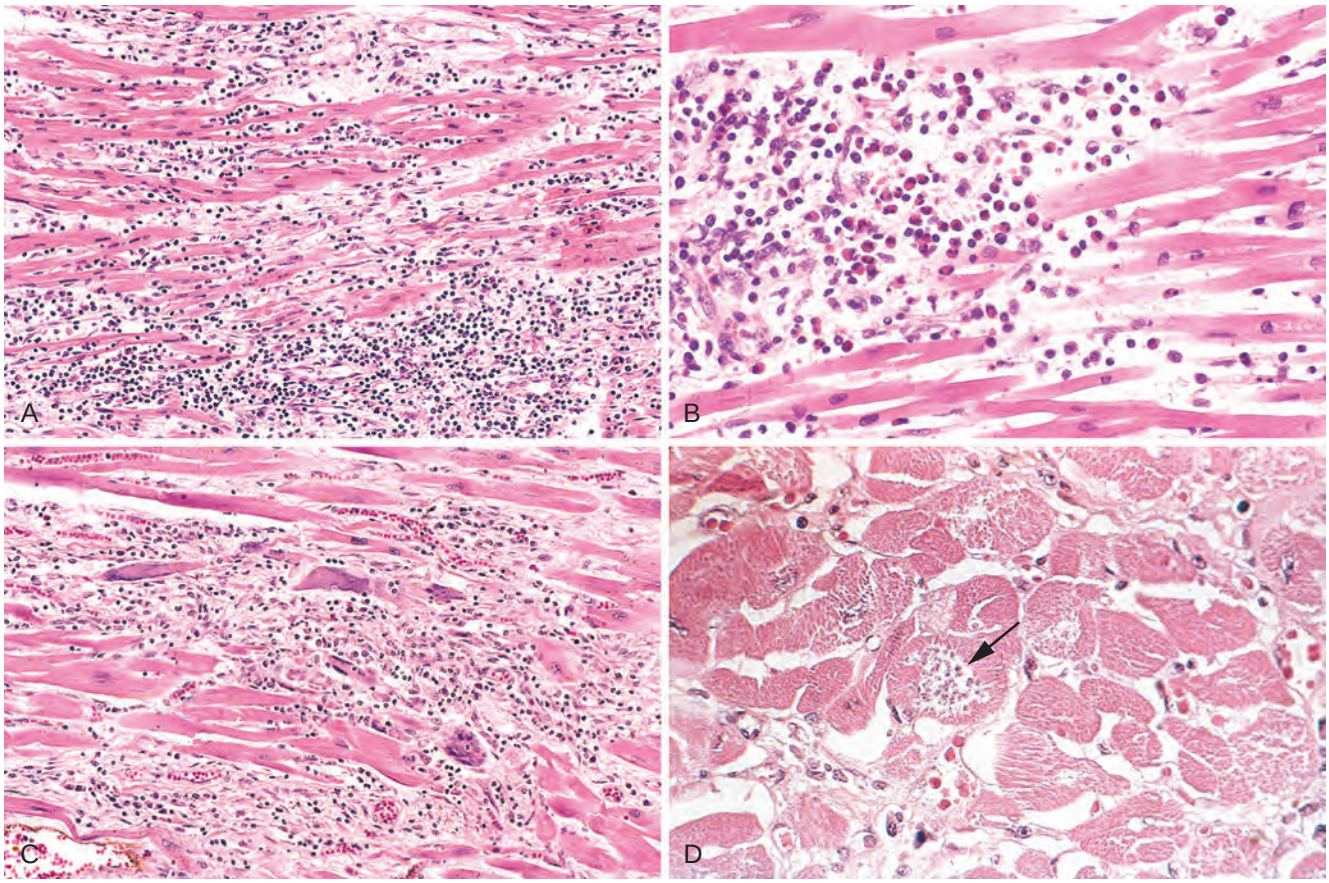

Figure 12-35 Myocarditis. **A,** Lymphocytic myocarditis, associated with myocyte injury. **B,** Hypersensitivity myocarditis, characterized by interstitial inflammatory infiltrate composed largely of eosinophils and mononuclear inflammatory cells, predominantly localized to perivascular and expanded interstitial spaces. **C,** Giant-cell myocarditis, with mononuclear inflammatory infiltrate containing lymphocytes and macrophages, extensive loss of muscle, and multinucleated giant cells (fused macrophages). **D,** The myocarditis of Chagas disease. A myofiber distended with trypanosomes *(arrow)* is present along with individual myofiber necrosis, and modest amounts of inflammation.

amyloidosis can appear as a consequence of systemic amyloidosis (e.g., due to myeloma or inflammation-associated amyloid) or can be restricted to the heart, particularly in the aged (*senile cardiac amyloidosis*). Senile cardiac amyloidosis characteristically occurs in individuals 70 years and older, and has a far better prognosis than systemic amyloidosis. Senile cardiac amyloid deposits are largely composed of *transthyretin*, a normal serum protein synthesized in the liver that *trans*ports *thy*roxine and *retin*ol-binding protein. Mutant forms of transthyretin can accelerate the cardiac (and associated systemic) amyloid deposition; 4% of African-Americans have a transthyretin mutation substituting isoleucine for valine at position 122 that produces a particularly amyloidogenic protein, responsible for autosomal dominant familial transthyretin amyloidosis. Isolated atrial amyloidosis can also occur secondary to deposition of atrial natriuretic peptide, but its clinical significance is uncertain.

Cardiac amyloidosis most frequently produces a restrictive cardiomyopathy, but it can also be asymptomatic, manifest as dilation or arrhythmias, or mimic ischemic or valvular disease. The varied presentations depend on the predominant location of the deposits, for example, interstitium, conduction system, vasculature, or valves.

MORPHOLOGY

In cardiac amyloidosis the heart varies in consistency from normal to firm and rubbery. The chambers are usually of normal size, but can be dilated and have thickened walls. Small, semitranslucent nodules resembling drips of wax may be seen on the atrial endocardial surface, particularly on the left. Histologically, hyaline eosinophilic deposits of amyloid may be found in the interstitium, conduction tissue, valves, endocardium, pericardium, and small intramural coronary arteries (Fig. 12-36); they can be distinguished from other deposits by special stains such as Congo red, which produces classic apple-green birefringence when viewed under polarized light (Fig. 12-36*B*). Intramural arteries and arterioles may have sufficient amyloid in their walls to compress and occlude their lumens, inducing myocardial ischemia ("small-vessel disease").

KEY CONCEPTS

Cardiomyopathy

- Cardiomyopathy is intrinsic cardiac muscle disease; there may be specific causes, or it can be idiopathic.