



Figure 12-36 Cardiac amyloidosis. **A**, Hematoxylin and eosin stain, showing amyloid appearing as amorphous pink material around myocytes. **B**, Congo red stain viewed under polarized light, in which amyloid shows characteristic apple-green birefringence (compared with collagen, which appears white).

- There are three general pathophysiologic categories of cardiomyopathy: dilated (90%), hypertrophic, and restrictive (least common).
- Dilated cardiomyopathy results in systolic (contractile) dysfunction. Causes include myocarditis, toxic exposures (e.g., alcohol), and pregnancy. In 20% to 50% of cases, genetic cytoskeletal protein defects are causal, with titin mutations representing up to 20% of cases of dilated cardiomyopathy.
- Hypertrophic cardiomyopathy results in diastolic (relaxation) dysfunction. Virtually all cases are due to autosomal dominant mutations in the proteins comprising the contractile apparatus, in particular β -myosin heavy chain.
- Restrictive cardiomyopathy results in a stiff, noncompliant myocardium and can be due to deposition (e.g., amyloid), increased interstitial fibrosis (e.g., due to radiation), or endomyocardial scarring.
- Myocarditis is myocardial damage caused by inflammatory infiltrates secondary to infections or immune reactions. Coxsackie A and B viruses are the most common causes in the United States. Clinically, myocarditis can be asymptomatic, give rise to acute heart failure, or evolve into dilated cardiomyopathy.

Pericardial Disease

The most important pericardial disorders cause fluid accumulation, inflammation, fibrous constriction, or some combination of these processes, usually in association with other cardiac pathology or a systemic disease; isolated pericardial disease is unusual.

Pericardial Effusion and Hemopericardium

Normally, the pericardial sac contains less than 50 mL of thin, clear, straw-colored fluid. Under various circumstances the parietal pericardium may be distended by serous fluid (*pericardial effusion*), blood (*hemopericardium*),

or pus (*purulent pericarditis*). With long-standing cardiac enlargement or with slowly accumulating fluid, the pericardium has time to dilate. This permits a slowly accumulating pericardial effusion to become quite large without interfering with cardiac function. Thus, with chronic effusions of less than 500 mL in volume, the only clinical significance is a characteristic globular enlargement of the heart shadow on chest radiographs. In contrast, rapidly developing fluid collections of as little as 200 to 300 mL—e.g., due to hemopericardium caused by a ruptured MI or aortic dissection—can produce clinically devastating compression of the thin-walled atria and venae cavae, or the ventricles themselves; cardiac filling is thereby restricted, producing potentially fatal *cardiac tamponade*.

Pericarditis

Pericardial inflammation can occur secondary to a variety of cardiac, thoracic, or systemic disorders, metastases from remote neoplasms, or cardiac surgical procedures. Primary pericarditis is unusual and almost always of viral origin. The major causes of pericarditis are listed in [Table 12-14](#). Most evoke an acute pericarditis, but a few, such as tuberculosis and fungi, produce chronic reactions.

Acute Pericarditis

Serous pericarditis is characteristically produced by non-infectious inflammatory diseases, including rheumatic fever, SLE, and scleroderma, as well as tumors and uremia. An infection in the tissues contiguous to the pericardium—for example, a bacterial pleuritis—may incite sufficient irritation of the parietal pericardial serosa to cause a sterile serous effusion that can progress to serofibrinous pericarditis and ultimately to a frank suppurative reaction. In some instances a well-defined viral infection elsewhere—upper respiratory tract infection, pneumonia, parotitis—antedates the pericarditis and serves as the primary focus of infection. Infrequently, usually in young adults, a viral pericarditis occurs as an apparent primary infection that may be accompanied by myocarditis (*myopericarditis*). Tumors can cause a serous pericarditis by lymphatic