

Figure 12-32 Dilated cardiomyopathy. **A**, Four-chamber dilatation and hypertrophy are evident. There is a mural thrombus (*arrow*) at the apex of the left ventricle (on the *right* in this apical four-chamber view). The coronary arteries were patent. **B**, Histologic section demonstrating variable myocyte hypertrophy and interstitial fibrosis (collagen is highlighted as blue in this Masson trichrome stain).

increasingly performed, and long-term ventricular assist can be beneficial. Interestingly, in some patients, relatively short-term mechanical cardiac support can induce durable improvement of cardiac function.

Arrhythmogenic Right Ventricular Cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited disease of myocardium causing right ventricular failure and rhythm disturbances (particularly ventricular tachycardia or fibrillation) with sudden death. Left-sided involvement with left-sided heart failure may also occur. Morphologically, the right ventricular wall is severely thinned due to loss of myocytes, accompanied by extensive

fatty infiltration and fibrosis (*Fig. 12-33*). Although myocardial inflammation may be present, ARVC is not considered an inflammatory cardiomyopathy. Classical ARVC has autosomal dominant inheritance with a variable penetrance. The disease has been attributed to defective cell adhesion proteins in the desmosomes that link adjacent cardiac myocytes. *Naxos syndrome* is a disorder characterized by arrhythmogenic right ventricular cardiomyopathy and hyperkeratosis of plantar palmar skin surfaces specifically associated with mutations in the gene encoding the desmosome-associated protein plakoglobin.

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is a common (incidence, 1 in 500), clinically heterogeneous, genetic

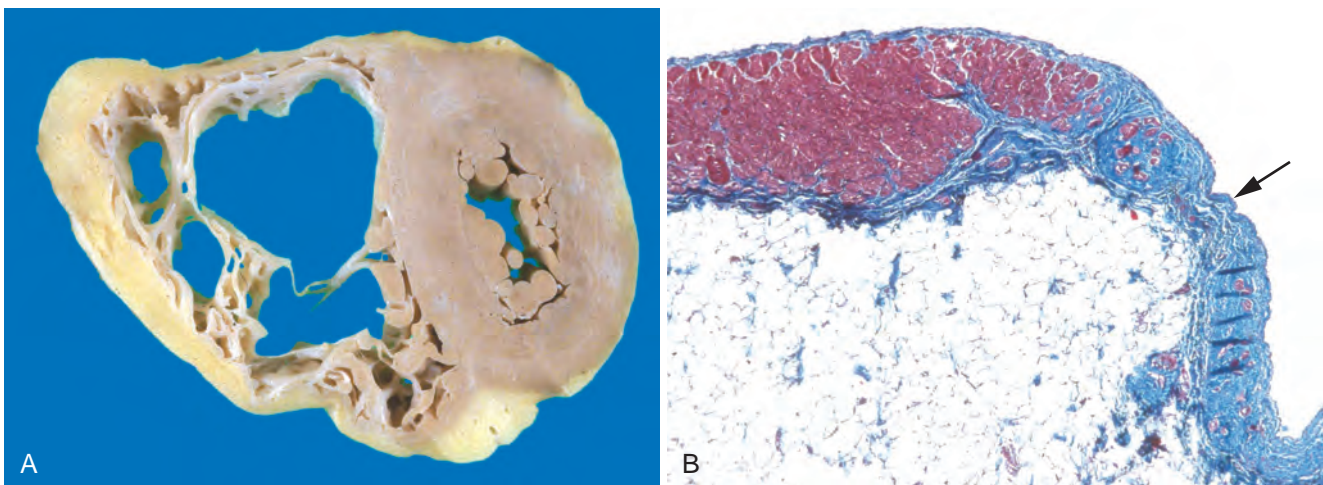


Figure 12-33 Arrhythmogenic right ventricular cardiomyopathy. **A**, Gross photograph, showing dilation of the right ventricle and near-transmural replacement of the right ventricular free-wall by fat and fibrosis. The left ventricle has a virtually normal configuration in this case, but can also be involved by the disease process. **B**, Histologic section of the right ventricular free wall, demonstrating replacement of myocardium (red) by fibrosis (blue, *arrow*) and fat (Masson trichrome stain).