affecting either or both ventricles. This condition shares with the previous condition the partial obliteration of the ventricular apex with fibrosis extending into the valvular inflow tract and leaflets; however, it is not clear that the etiologies are the same for all cases. Pericardial effusions frequently accompany endomyocardial fibrosis but are not common in Löffler's endocarditis. For endomyocardial fibrosis, there is no gender difference, but a higher prevalence in African-American populations. While tropical endomyocardial fibrosis could represent the end-stage of previous hypereosinophilic disease triggered by endemic parasites, neither prior parasitic infection nor hypereosinophilia is usually documented. Geographic nutritional deficiencies have also been proposed as an etiology.

Medical treatment focuses on glucocorticoids and chemotherapy to suppress hypereosinophilia when present. Fluid retention may become increasingly resistant to diuretic therapy. Anticoagulation is recommended. Atrial fibrillation is associated with worse symptoms and prognosis, but may be difficult to suppress. Surgical resection of the apices and replacement of the fibrotic valves can improve symptoms, but surgical morbidity and mortality and later recurrence rates are high.

The serotonin secreted by *carcinoid* tumors can produce fibrous plaques in the endocardium and right-sided cardiac valves, occasionally affecting left-sided valves, as well. Valvular lesions may be stenotic or regurgitant. Systemic symptoms include flushing and diarrhea. Liver disease from hepatic metastases may play a role by limiting hepatic function and thereby allowing more serotonin to reach the venous circulation.

HYPERTROPHIC CARDIOMYOPATHY

Hypertrophic cardiomyopathy is defined as left ventricular hypertrophy that develops in the absence of causative hemodynamic factors, such as hypertension, aortic valve disease, or systemic infiltrative or storage diseases **(Figs. 287-15 and 287-16)**. It has previously been termed *hypertrophic obstructive cardiomyopathy* (HOCM), *asymmetric septal hypertrophy* (ASH), and *idiopathic hypertrophic subaortic stenosis* (IHSS). However, the accepted terminology is now hypertrophic cardiomyopathy with or without obstruction. Prevalence in North America, Japan, and China is about 1:500. It is the leading cause of sudden death in the young and is an important cause of heart failure. Although pediatric presentation is associated with increased early morbidity and mortality, the prognosis for patients diagnosed as adults is generally favorable.

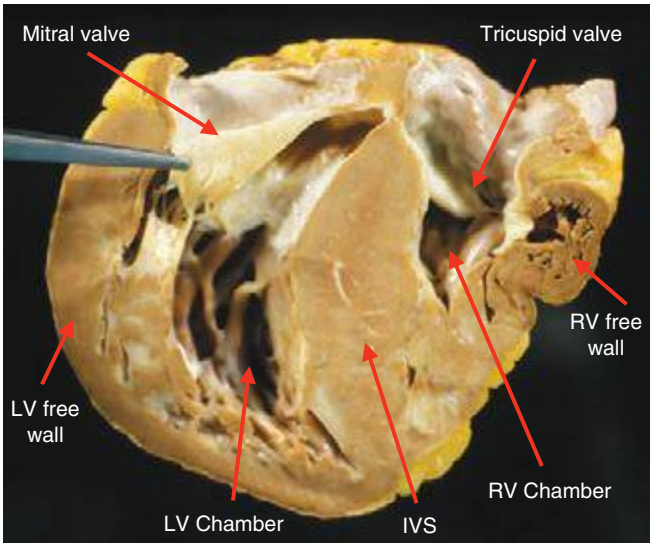

FIGURE 287-15 Hypertrophic cardiomyopathy. Gross specimen of a heart with hypertrophic cardiomyopathy removed at the time of transplantation, showing asymmetric septal hypertrophy (septum much thicker than left ventricular free wall) with the septum bulging into the left ventricular outflow tract causing obstruction. The forceps are retracting the anterior leaflet of the mitral valve, demonstrating the characteristic plaque of systolic anterior motion, manifest as endocardial fibrosis on the interventricular septum in a mirror-image pattern to the valve leaflet. There is patchy replacement fibrosis, and small thick-walled arterioles can be appreciated grossly, especially in the interventricular septum. IVS, interventricular septum; LV, left ventricle; RV, right ventricle. *(Image courtesy of Robert Padera, MD, PhD, Department of Pathology, Brigham and Women's Hospital, Boston.)*

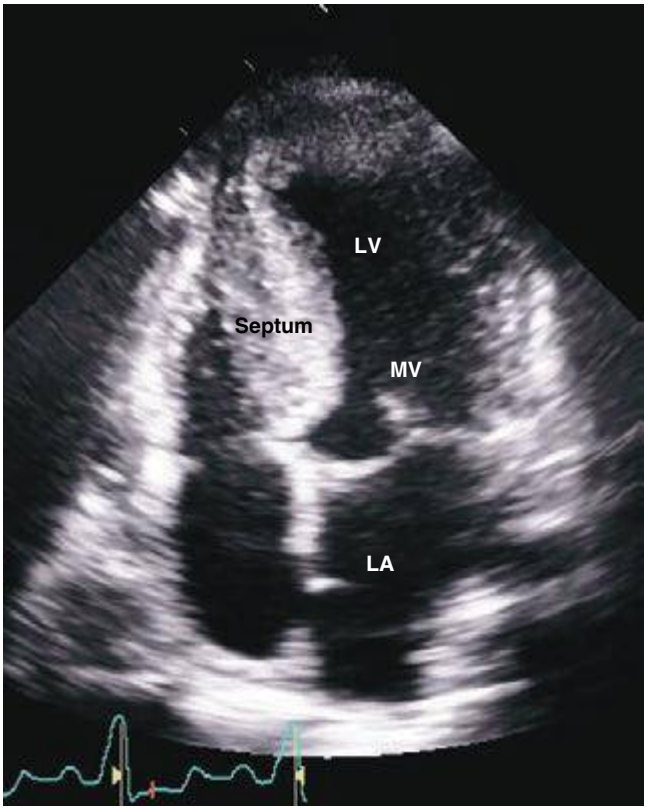

FIGURE 287-16 Hypertrophic cardiomyopathy. This echocardiogram of hypertrophic cardiomyopathy shows asymmetric hypertrophy of the septum compared to the lateral wall of the left ventricle (LV). The mitral valve (MV) is moving anteriorly toward the hypertrophied septum in systole. The left atrium (LA) is enlarged. Note that the echocardiographic and pathologic images are vertically opposite, such that the LV is by convention on the top right in the echocardiographic image and bottom right in the pathologic images. *(Image courtesy of Justina Wu, MD, Brigham and Women's Hospital, Boston.)*

The clustering of hypertrophic cardiomyopathy within families has been appreciated since recognition of the disease approximately 55 years ago. Echocardiographic screening of families revealed an autosomal dominant pattern of inheritance. Initial genetic studies using linkage analysis in large families identified disease-causing mutations in sarcomeric genes. A sarcomere mutation is present in ~60% of patients with hypertrophic cardiomyopathy and is more common in those with familial disease and characteristic asymmetric septal hypertrophy. More than nine different sarcomere genes with over 1400 mutations have been implicated, although ~80% of patients have a mutation in either *MYH7* or *MYBPC3* (Table 287-3), most of which are unique to individual families ("private" mutations).

Hypertrophic cardiomyopathy is characterized by age-dependent and incomplete penetrance. The defining phenotype of left ventricular hypertrophy is rarely present at birth and usually develops later in life. Accordingly, screening of family members should begin in adolescence and extend through adulthood. In *MYBPC3* mutation carriers, the average age of disease development is 40 years, while 30% remain free from hypertrophy after 70 years. Related individuals who carry the *same* mutation may have a different extent and pattern of hypertrophy (e.g., asymmetric versus concentric), occurrence of outflow tract obstruction, and associated clinical outcomes (e.g., sudden death, atrial fibrillation).