

cardiomyopathy had a 60% cumulative risk of sudden death within 5 years of the syncopal event ([Spirito, Autore, Rapezzi, et al, 2009](#)). Presentation in infancy includes signs of HF and has a poor prognosis. The ECG demonstrates left ventricular hypertrophy, often with ST-T changes. The echocardiogram is most helpful and demonstrates asymmetric septal hypertrophy and an increase in left ventricular wall thickness, with a small left ventricle cavity.

Restrictive cardiomyopathy, which is rare in children, describes a restriction to ventricular filling caused by endocardial or myocardial disease or both. It is characterized by diastolic dysfunction and absence of ventricular dilation or hypertrophy. Symptoms are similar to those of HF (see earlier in this chapter).

Therapeutic Management

Treatment is directed toward correcting the underlying cause whenever feasible. However, in most affected children, this is not possible, and treatment is aimed at managing HF (see earlier in this chapter) and dysrhythmias. Digoxin, diuretics, and aggressive use of afterload reduction agents have been found to be helpful in managing symptoms in those with dilated cardiomyopathy. Practice guidelines for the management of HF in children have been outlined and provide an in-depth review of available therapies ([Rosenthal, Chrisant, Edens, et al, 2004](#); [Rossano and Shaddy, 2014](#)). Digoxin and inotropic agents are usually not helpful in the other forms of cardiomyopathy because increasing the force of contraction may exacerbate the muscular obstruction and actually impair ventricular ejection. Beta-blockers (such as propranolol) and calcium channel blockers (such as verapamil) have been used to reduce left ventricular outflow obstruction and improve diastolic filling in those with hypertrophic cardiomyopathy.

Careful monitoring and treatment of dysrhythmias are essential. The placement of an automatic implantable cardioverter defibrillator (AICD) should be considered for patients at high risk of sudden death because of ventricular dysrhythmias. Anticoagulants may be given to reduce the risk of thromboemboli, a complication of the sluggish circulation through the heart. For worsening HF and signs of poor perfusion, IV inotropic or vasodilating drugs may be needed. Severely ill children may require mechanical ventilation, oxygen administration, and IV