

Outcomes

Prognosis is poor in patients with RCM with a 90% mortality rate at 10 years (Kavinsky & Parrillo, 2000). Amyloidosis carries the highest mortality, with a 2-year mortality rate being greater than 80% (Darovic, 2002).

Linking Knowledge to Practice

- ✓ *Anything that would normally cause the heart rate to increase, including activity, decreased blood pressure, fever, shivering, and low blood volume, results in a further decrease in stroke volume in patients with RCM.*
- ✓ *Patients with RCM should be closely monitored for signs of decreased CO that may result from overdiuresis. Signs include hypotension, especially orthostatic hypotension, lethargy, increased heart rate, and increased blood urea nitrogen levels.*

HYPERTROPHIC CARDIOMYOPATHY

HCM is characterized by hypertrophy of the myocardium (Figure 10.1). Associated with the increase in muscle mass is a decrease in ventricular filling (diastolic dysfunction) and a decrease in CO. Other causes of ventricular hypertrophy, including long-standing hypertension and aortic stenosis, must be ruled out before a diagnosis of HCM can be made. This disease process has had many names in the past, including idiopathic hypertrophic subaortic stenosis (IHSS). The World Health Organization's recommendation of "hypertrophic cardiomyopathy" as the correct terminology for this disease process has been widely accepted. HCM is a general term that covers all cases; however, a subgroup of patients with HCM develop HCM with obstruction. Once obstruction develops, the process is referred to as hypertrophic obstructive cardiomyopathy or HOCM.

Prevalence

HCM is found in one in every 500 people and affects women and men equally (Maron et al., 2003). It is the most common reason for sudden cardiac death in young adults.

Causes

The cause of HCM is unknown; however, over 50% of HCM cases are transmitted genetically (Kavinsky & Parrillo, 2000). An abnormal sympathetic nervous system response or abnormal catecholamine levels may cause the other half of the cases (Shah, 2003).

Pathophysiology

In HCM, a generalized disarray of the cardiac myofibrils occurs, along with hypertrophy of the myocytes (Figure 10.7). Cardiac cells take on a variety of shapes, and myocardial scarring and fibrosis occur. These changes result in myocardial walls that become very thick and stiff. The left ventricle is usually affected, with little effect on the right ventricle. The changes may be symmetrical; however, in many cases, asymmetrical septal hypertrophy is the most common finding (Figure 10.8). In asymmetrical hypertrophy, the ventricular septum experiences the greatest increase in wall thickness, often up to twice its normal size.

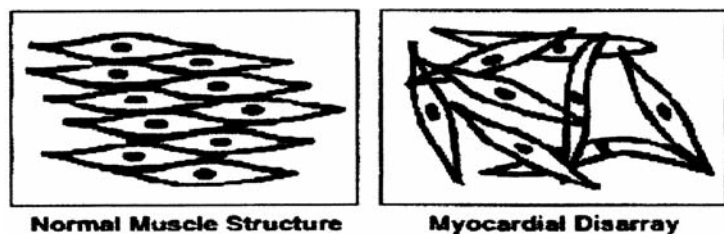


Figure 10.7: The difference between normal myocardial cells and the myocardial disarray with HCM.
Courtesy of the Hypertrophic Cardiomyopathy Association www.4hcm.org