Cardiomyopathy: An Overview

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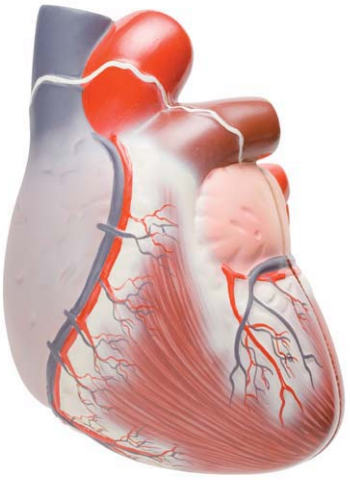

ABSTRACT

Cardiomyopathies and their resultant systolic and diastolic heart failure remain the main cause of cardiovascular morbidity and mortality in both children and adults and are a frequent indication for cardiac transplantation. According to the American Heart Association 2005 Heart and Stroke Update, more than 26,000 deaths each year in the United States are caused by cardiomyopathy. Cardiomyopathy is second to coronary artery disease for the most common direct cause of sudden death in the United States and is a leading cause of heart failure. This article provides an overview of the pathophysiology, causes, signs and symptoms, diagnosis, and treatment of the different types of cardiomyopathies. Newer therapeutic modalities and pharmacologic interventions are discussed, with an emphasis on improving symptoms and long-term survival.

Keywords: cardiomyopathy, diastolic dysfunction, dilated cardiomyopathy, hypertrophic cardiomyopathy, restrictive cardiomyopathy, systolic, systolic dysfunction

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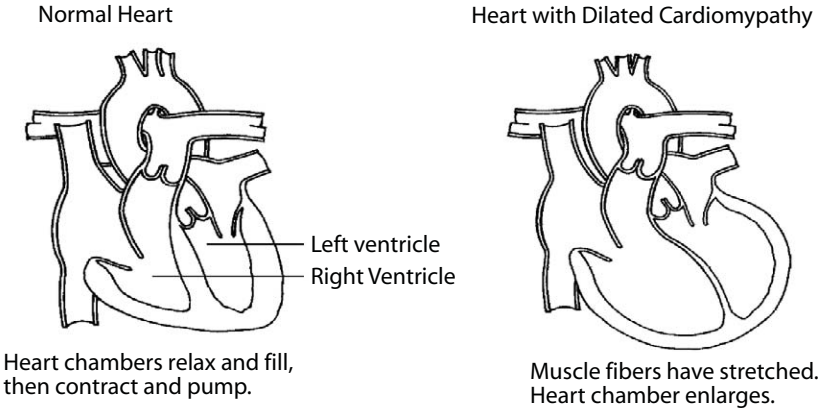
In 1995, the World Health Organization/International Society and Federation of Cardiology Task Force on the Definition and Classification of the Cardiomyopathies defined cardiomyopathies as "diseases of the myocardium associated with cardiac dysfunction."[2]

The purpose of this article is to provide the advanced practice nurse with an overview of the pathophysiology, causes, signs and symptoms, diagnosis, and management of the different types of cardiomyopathies. Newer therapeutic modalities and pharmacologic interventions are discussed, with an emphasis on improving symptoms and long-term survival.

SYSTOLIC VERSUS DIASTOLIC DYSFUNCTION

Systolic dysfunction is characterized by a decrease in myocardial contractility. A reduction in the left ventricular ejection fraction (LVEF) results when myocardial contractility is decreased throughout the left ventricle. Cardiac output is maintained in two ways: left ventricular enlargement results in a higher stroke volume, and the Frank-Starling relationship (an increase in contractility in response to an increase in stretch). However, these compensatory mechanisms are eventually exceeded and cardiac output decreases, resulting in the physiologic manifestations of heart failure.[3] The left heart cannot pump with enough force to push a

Figure 1. Diagrams of a normal heart and a heart with dilated cardiomyopathy.
Adapted from: *www.nscardiology.com/factscardiomyopathy.htm*.



sufficient amount of blood into the systemic circulation. This leads to fluid backing up into the lungs and pulmonary congestion. Systolic dysfunction is a characteristic of dilated cardiomyopathy (DCM). It is also seen in some patients with hypertrophic cardiomyopathy (HCM) who develop progressive left ventricular dilatation and a decrease in LVEF. In general terms, systolic dysfunction is defined as an LVEF less than 40%.[4]

Diastolic dysfunction refers to cardiac dysfunction in which left ventricular filling is abnormal and is accompanied by elevated filling pressures. The diastolic phase of cardiac function includes two components. Left ventricular relaxation is a process that takes place during isovolemic relaxation (the period between aortic valve closure and the mitral valve opening) and then during early rapid filling of the ventricle. Later in diastole, after relaxation is complete, further left ventricular filling is a passive process that depends on the compliance, or distensibility, of the myocardium. The ventricles are unable to relax, and subsequent muscle hypertrophy occurs which then leads to inadequate filling.[3] Diastolic dysfunction may lead to fluid accumulation, especially in the feet, ankles, and legs, and some patients may also have pulmonary congestion. For patients with heart failure but without systolic dysfunction, diastolic dysfunction is the presumed cause. Diastolic dysfunction is characteristic of both HCM and restrictive cardiomyopathy (RCM).[3] However, some component of diastolic dysfunction is also common in patients with DCM. In general terms, diastolic dysfunction is defined as an LVEF of greater than 40%.

Diastolic dysfunction is more difficult to identify with echocardiograph scanning than systolic dysfunction, and it may be missed or underestimated in many cases. Doppler scan assessment of transmitral flow is the standard approach to detect diastolic dysfunction, although a variety of other measurements can be used.[3] It is important to understand that some of the symptoms of systolic and diastolic heart failure are similar.

DILATED CARDIOMYOPATHY

Anatomic and Physiologic Classification

DCM, the most common form of cardiomyopathy, is characterized by enlargement of one or both ventricles accompanied by systolic and diastolic contractile dysfunction and symptoms of heart failure.[5]

In DCM, myocardial muscle mass is increased and ventricular wall thickness is reduced. The heart assumes a globular shape, and there is pronounced ventricular chamber dilatation, diffuse endocardial thickening, and atrial enlargement often with thrombi in the appendages.[6] The heart muscle becomes thin and weakened and is unable to pump the blood efficiently. The heart muscle stretches and dilates so that it can hold more but in time becomes even weaker, leading to symptoms of heart failure. These structural changes decrease the amount of blood ejected from the ventricle with systole and allow more blood in the ventricle after contraction. A smaller volume of blood enters the ventricle during diastole and increases end-diastolic pressure and pulmonary pressures. The enlarged stretched ventricle alters valvular function, usually resulting in regurgitation. Left ventricular dilatation occurs as venous return and systemic vascular resistance rise. Eventually, the atria also dilate as more work is required to pump blood into the full ventricles. Cardiomegaly occurs as a consequence of dilatation of the atria and ventricles. Blood pooling in the ventricles increases the risk of emboli (Figure 1).