

## CARDIOMYOPATHY

### **What is cardiomyopathy?**

Cardiomyopathy (kar"de-o-mi-OP'ah-the) is a serious disease in which the heart muscle becomes inflamed and doesn't work as well as it should. There may be multiple causes including viral infections.

Cardiomyopathy can be classified as primary or secondary. Primary cardiomyopathy can't be attributed to a specific cause, such as hypertension, heart valve disease, artery diseases or congenital heart defects. Secondary cardiomyopathy is due to specific causes and is often associated with diseases involving other organs as well as the heart. There are three principal types of cardiomyopathy -- dilated, hypertrophic and restrictive.

### **What is dilated (congestive) cardiomyopathy?**

This is the most common form. The heart cavity is enlarged and stretched (dilated) in contrast to the thickening of the walls in the hypertrophic form. The heart is weak and doesn't pump normally, and most patients develop congestive heart failure. Arrhythmias and disturbances in the heart's electrical conduction also may occur.

Since blood flows more slowly through an enlarged heart, blood clots easily form. A blood clot is also called a thrombus. A clot that breaks free, circulates in the bloodstream and blocks a small blood vessel is called an embolus.

- If the clot breaks off in the right ventricle, it can be carried into the pulmonary circulation in the lung, forming a pulmonary emboli.
- Blood clots formed in the left side of the heart may be dislodged and carried into the body's circulation to form cerebral emboli in the brain, renal emboli in the kidney, peripheral emboli or even coronary artery emboli.

### **How is dilated or congestive cardiomyopathy treated?**

A person with cardiomyopathy may suffer an embolus before any other symptom of cardiomyopathy appears, and anti-clotting (anticoagulant) drug therapy may be needed. Arrhythmias may require antiarrhythmic drugs. More rarely, "heart block" may develop, requiring an artificial pacemaker. Therapy for dilated cardiomyopathy is sometimes disappointing, however. If the person is young and otherwise healthy, and if the disease gets worse and worse, a heart transplant may be considered.

When cardiomyopathy results in marked cardiac dilation, the leaflets of the mitral and tricuspid valves may not be able to close properly, resulting in murmurs. Blood pressure may increase because of increased sympathetic nerve activity. These nerves can also cause arteries to constrict (narrow). This mimics hypertensive heart disease (high blood pressure). That's why some people have high blood pressure readings. Because the level of blood pressure determines the workload and oxygen needs of the heart, one approach in treatment is to use

vasodilators (drugs that "relax" the arteries). They lower blood pressure and thus the left ventricle's workload.

### **What is hypertrophic cardiomyopathy?**

In this condition, the muscle mass of the left ventricle enlarges or "hypertrophies." In one form of the disease, the wall between the two ventricles (septum) becomes enlarged and obstructs the blood flow from the left ventricle. The syndrome is known as hypertrophic obstructive cardiomyopathy (H.O.C.M.) or asymmetric septal hypertrophy (A.S.H.). It's also called idiopathic hypertrophic subaortic stenosis (I.H.S.S).

Besides obstructing blood flow, the thickened wall sometimes distorts one leaflet of the mitral valve, causing it to leak. In over half the cases, the disease is hereditary. Close blood relatives (parents, children or siblings) of such persons often have enlarged septums, although they may have no symptoms. This disease is most common in young adults. In the other form of the disease, non-obstructive hypertrophic cardiomyopathy, the enlarged muscle doesn't obstruct blood flow.

The symptoms of hypertrophic cardiomyopathy include shortness of breath on exertion, dizziness, fainting and angina pectoris (chest pain or discomfort caused by reduced blood supply to the heart muscle). Some people have cardiac arrhythmias, abnormal heart rhythms that in some cases can lead to sudden death. The obstruction to blood flow from the left ventricle increases the work the ventricle must do, and a heart murmur may be heard.

### **How is hypertrophic cardiomyopathy treated?**

A drug known as a beta-blocker (such as propranolol) or a calcium channel blocker is the usual treatment. If a person has an arrhythmia, an antiarrhythmic drug may also be used. Surgical treatment of the obstructive form is possible in some cases if the drug treatment fails.

Alcohol ablation is another nonsurgical treatment being developed for hypertrophic obstructive cardiomyopathy. It involves injecting alcohol down a small branch of one of the heart arteries to the extra heart muscle. The procedure results in the extra heart muscle being destroyed without having to cut it out surgically.

People undergoing this procedure usually suffer chest pain during the alcohol injection. The alcohol can also disrupt normal heart rhythms and require the insertion of a pacemaker. Alcohol ablation is a relatively new procedure being performed at only a few specialized centers in the United States. However, it's too soon to know whether this treatment will result in long-term benefit. **It's still considered experimental.**

### **What is restrictive cardiomyopathy?**

This is the least common type in the United States. The myocardium of the ventricles becomes excessively "rigid," so it's harder for the ventricles to fill with blood between heartbeats. A person with restrictive cardiomyopathy often complains of being tired, may have swollen hands and feet, and may have difficulty breathing on exertion. This type of cardiomyopathy is usually due to another disease process.