

# HEART MUSCLE DISEASE

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Compared with other cardiovascular diseases, heart muscle disease (cardiomyopathy) is relatively rare. In its most common form, the disease accounts for only 50,000 new cases in the United States each year, while the annual number of stroke cases, for example, reaches 500,000. Unlike many other cardiovascular disorders that tend to affect the elderly, cardiomyopathy commonly occurs in the young and can have a tragically brief course.

Cardiomyopathy (*cardio* meaning heart, *myopathy* meaning muscle disease) refers to a group of disorders that directly damage the muscle of the heart walls. In these disorders, all chambers of the heart are affected. The heart's function as a pump is disrupted, leading to an inadequate blood flow to organs and tissues of the body. Depending on the nature of the injury or abnormality in the heart muscle and the resulting structural changes in the heart chambers, one of three types of nonischemic (not caused by heart attack) heart muscle disease may be present: dilated congestive, hypertrophic, or restrictive. (See Table 15.1.)

Massive or multiple heart attacks may also lead to severe heart damage as a result of a disruption of blood supply to heart muscle. The damage can result in functional impairment and structural abnormalities similar to those found in the other types of cardiomyopathy. This type of heart disease, resulting from coronary artery disease, is called ischemic car-

diomyopathy. When used alone, however, the term "cardiomyopathy" refers to heart muscle disease that is not caused by heart attacks.

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## DILATED CONGESTIVE CARDIOMYOPATHY

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This is the most common type of heart muscle disease. It is generally called either dilated or congestive cardiomyopathy. This type of disease damages the fibers of the heart muscle, weakening the walls of the heart's chambers. Usually, all chambers are affected, and depending on the severity of the injury, they lose some of their capacity to contract forcefully and pump blood through the circulatory system. To compensate for the muscle injury, the heart chambers enlarge or dilate. The dilation is often more pronounced in the left ventricle, the heart's main pumping chamber. (See Figure 15.1.)

Dilated cardiomyopathy causes heart failure—an inability of the heart to provide an adequate supply of blood to the body's organs and tissues—which, if left untreated, is always associated with excess fluid retention, congestion in the lungs and liver, and swelling of the legs. Fluid retention occurs during heart failure because many organs fail to receive suf-

**MAJOR CARDIOVASCULAR DISORDERS**

Table 15.1  
Types of Heart Muscle Disease (Cardiomyopathy)

Type	Therapy
Dilated congestive: Cavity of the heart is enlarged and stretched. Cause is usually unknown.	When underlying cause is unknown, treatment focuses on relieving symptoms and improving function. Drugs used include digitalis and digoxin (Lanoxin and others), diuretics such as furosemide (Lasix and others), steroids to relieve inflammation, and ACE inhibitors such as captopril (Capoten). When symptoms cannot be relieved, heart transplant may be considered.
Hypertrophic: Muscle mass increases, causing chest pain, palpitations, and possibly fainting during physical activity. May be genetically acquired.	Limit stressful physical activity, and use medication, including beta blockers or a calcium channel blocker such as verapamil (Calan, Isopton, Verelan). If medication does not relieve symptoms, undergo surgical removal of excess muscle tissue that obstructs blood flow in the heart chambers. If surgery does not help, heart transplant may be considered.
Restrictive: Abnormal cells, proteins, or scar tissue infiltrate the heart, causing the chambers to become thick and bulky. Most common cause in the United States is a disease (amyloidosis) that is associated with cancers of the blood.	Treated with medications that alleviate symptoms (see dilated congestive section above). No cure exists.

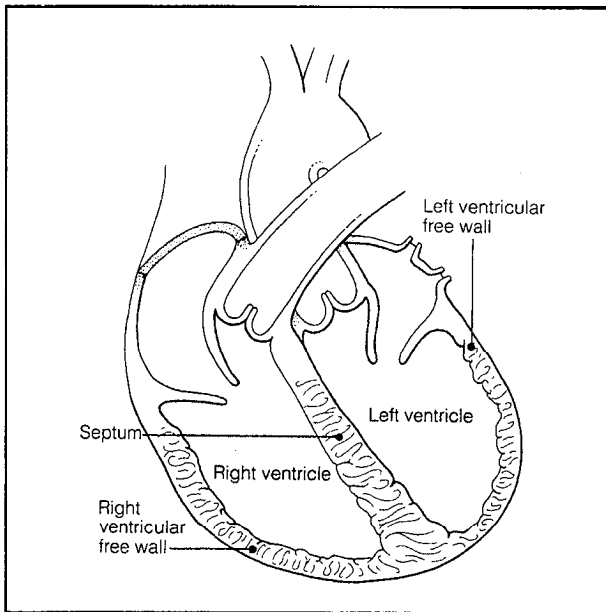
Type	Therapy
Ischemic (related to coronary artery disease):	Treated with medications that relieve symptoms of heart failure and coronary artery disease (see above). Angioplasty and coronary artery bypass grafting may help increase blood flow to the heart, enhancing heart muscle function. When neither drug therapy nor surgery helps, heart transplant may be considered.

icient blood flow. The kidneys respond to this lack of blood supply by retaining more than the usual amount of salt and water. With time, excess fluid retention leads to congestion in the lungs and other organs. At the end of the day, much of the retained fluid gravitates to the lower portions of the body and causes swelling in the legs. (See Chapter 14 for more information on heart failure and its symptoms.)

**THE COURSE OF DILATED CARDIOMYOPATHY**

When the chambers dilate, the muscle fibers in the heart walls stretch, enabling them to contract more forcefully. (This is characteristic of all muscles.) Growth of muscle tissue, which can to some extent rebuild damaged areas of the heart wall, also helps to keep up normal function. If the injury to the heart muscle is relatively mild, new muscle growth and the process of fiber stretching, which occurs roughly in proportion to the muscle damage, can partially restore cardiac function. If, however, injury is severe, the heart's function deteriorates. When damage to the heart is chronic or recurrent, as may occur with a prolonged exposure to excessive amounts of alcohol or infection, chamber dilation maybe slow and progressive. Eventually, the enlarged, thin-walled ventricles become flabby and cannot generate sufficient pressure to pump blood effectively throughout the body.

Dilated cardiomyopathy typically leads to a steady deterioration in heart function, although the course



**Figure 15.1**  
*In dilated cardiomyopathy, the cavity of the heart is enlarged and stretched.*

of the decline varies greatly and is difficult to predict for any given patient. Most patients go through periods of relatively stable heart function that may last several months or even years. However, the majority eventually succumb to complications of the disease. Most commonly, they die of progressive heart failure that is not amenable to treatment, although some die suddenly and unexpectedly.

Most instances of sudden death are believed to result from ventricular fibrillation—an abnormally fast and irregular heart rhythm with ineffective contractions that causes death within minutes. Patients with dilated cardiomyopathy are at risk of sudden death because the underlying disease process disrupts the normal electrical pathways of the heart, possibly causing rhythm disturbances.

Less often, sudden death may result from an embolus—a blood clot that dislodges from one of the heart chambers, travels to another vital organ, such as the brain or lungs, and obstructs the blood supply. Poor circulation and stagnation of blood in the dilated heart chambers provide favorable conditions for blood clot formation.

### **SYMPTOMS AND CAUSES OF DILATED CARDIOMYOPATHY**

The main symptoms of dilated cardiomyopathy are those of congestive heart failure—breathlessness or fatigue during physical activity and swelling of the lower legs. Some patients, especially those who lead

a sedentary life, may experience few symptoms and be unaware that the heart is failing. With advanced disease, symptoms may occur with minimal activity or even in the absence of physical exertion.

In more than 80 percent of cases, the cause of dilated cardiomyopathy is unknown. Major causes of the disease are inflammation of the heart muscle (myocarditis), excessive alcohol use, poor nutrition, and, rarely, complications arising shortly before or after childbirth (peripartum) and genetic disorders. (See box, “Causes of Dilated Congestive Cardiomyopathy.”)

### **HEART MUSCLE INFLAMMATION (MYOCARDITIS)**

Most cases of dilated cardiomyopathy probably result from inflammation of the heart muscle (myocarditis), but not all cases of heart muscle inflammation lead to dilated cardiomyopathy. In fact, myocarditis is often categorized as heart muscle disease in its own right. In Western Europe and the United States, myocarditis occurs most often as a complication of a viral disease, but it is a rather rare complication. Viral infections are believed to cause indirect damage to the heart. The invading virus provokes proteins that normally are confined within heart muscle cells to become exposed to the bloodstream. This sets off an inflammatory process as the body mistakenly assumes these newly exposed proteins belong to foreign cells and attacks them in the same way it fights viruses and bacteria. The unfortunate result is inflammation and injury to the body’s own tissues—in the case of myocarditis, the tissues of the heart.

Many organisms can infect and injure the heart muscle. Coxsackie Type B, a virus among those that

### **Causes of Dilated Congestive Cardiomyopathy**

In many cases, the cause cannot be identified. When causes are known, they include:

- Inflammation of the heart muscle (myocarditis), either infectious or noninfectious
- Excessive alcohol consumption
- Nutritional deficiencies
- Complications arising shortly before or after childbirth (peripartum)
- Genetic disorders

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usually infect the gastrointestinal tract, is believed to be the most common offending agent. Many other viruses, such as those of polio, rubella, and influenza, have been associated with myocarditis. It is not clear why the same viruses cause myocarditis in some patients and different diseases—gastroenteritis, pneumonia, or hepatitis, for example—in others.

Myocarditis can occur as a rare complication of bacterial infections, including diphtheria, tuberculosis, typhoid fever, and tetanus. Other infectious organisms, such as rickettsiae and parasites, may also cause inflammation in the heart muscle. In Central and South America, myocarditis is often due to Chagas disease, an infectious illness that is transmitted by insects.

Noninfectious causes of myocarditis are numerous, but all of them are rare. They include systemic lupus erythematosus, a disease in which the body attacks its own organs and tissues; adverse or toxic drug reactions; and radiation-induced heart injury as a complication of cancer radiotherapy.

Often myocarditis, particularly in its mild form, produces no symptoms at all. However, it is frequently accompanied by an inflammation of the heart's outer membrane—the pericardium. Inflammation of the heart lining is called pericarditis and, unlike myocarditis, can cause severe pain that typically gets worse when the person takes a deep breath or changes position.

Myocarditis may start as a flulike illness that lingers longer than the usual several days. If significant muscle damage and weakening of the heart's chambers occur, symptoms of heart failure may develop. A month or two later, the symptoms of flu-weakness and malaise—merge with symptoms of heart failure—fatigue during physical activity and shortness of breath. If the illness is persistent and progressive, symptoms eventually become disabling enough for the person to consult a physician. By this time, however, the infecting organisms usually cannot be detected or cultured from the heart or other places in the body. By the time the patient seeks medical help, all traces of the infecting organism or disease process that may have triggered the condition may be undetectable.

In some cases, the injury to the heart muscle is mild but persists or recurs intermittently over many years. Symptoms of heart failure sometimes appear 20 to 30 years after the initial viral illness. Patients usually do not recall having had a viral infection and often mistakenly interpret the symptoms as a sign of age until progressive heart disease produces more obvious signs of congestion and heart failure.

Myocarditis is usually diagnosed after it has reached an advanced stage and produces heart failure. Physical examination and a chest X-ray usually reveal signs of lung congestion and heart enlargement. An electrocardiogram may show changes of heart damage, and an echocardiogram demonstrates the characteristic abnormalities of severe myocarditis—enlargement of all heart chambers and poor contraction of the heart muscle. In acute myocarditis, a heart biopsy, in which a small sample of muscle tissue is removed from the heart chamber for laboratory examination, may be performed to document the presence of an ongoing inflammatory process. In cases of infectious myocarditis, however, it is usually impossible to grow the infecting organism from samples of the heart tissue.

Mild cases of myocarditis with no signs of heart failure are usually not diagnosed and consequently remain untreated. When treatment is given, it is aimed at eliminating the underlying cause. When the cause is unknown, steroids (cortisone) are sometimes prescribed to reduce inflammation. (This approach to therapy has not yet been shown to be beneficial but is currently under study.) Medications are also prescribed to relieve the symptoms of heart failure. (See Chapters 14 and 23.) During the acute phase of myocarditis, patients are advised to rest and gradually return to a more active life-style once evidence disappears of ongoing inflammation and heart injury. (See box, “Guidelines for Cardiomyopathy Patients.”)

Many cases of myocarditis cause minimal heart damage. Heart function fully recovers in these mild cases. Occasionally, severe cases of myocarditis also clear up spontaneously and leave little permanent damage. More typically, however, severe inflammation produces chronic, progressive, and irreversible heart damage.

Left untreated, myocarditis may lead to a severe form of pulmonary edema, or lung congestion, in which fluid leaks from the blood into the tissues and air spaces of the lung. The onset of this can be quite rapid, often waking the patient from sleep. Such patients are severely disabled and require emergency treatment. It must be emphasized, however, that myocarditis is rare and that viral infections rarely result in heart muscle damage.

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## ALCOHOL

In Western countries, excessive alcohol consumption is a major cause of cardiomyopathy. Alcohol can damage the heart directly by exerting a toxic effect on heart muscle cells. Severe heart damage may also

## Guidelines for Cardiomyopathy Patients

- To detect cardiomyopathy in its early stages, watch out for its early symptoms. Shortness of breath may constitute the first warning and therefore should never be disregarded as simply a sign of aging.
- Because hypertrophic cardiomyopathy can be passed on as a genetic trait, children of people with this condition should be evaluated by a cardiologist.
- When possible, eliminate all factors causing or contributing to heart muscle disease, such as excessive alcohol consumption.
- Keep salt intake to a minimum to decrease the tendency toward lung congestion and possibly reduce the need for diuretics. “
- Losing weight is an effective way to decrease the workload of an impaired heart, but it may not eliminate all symptoms.
- Quitting tobacco use is essential, since smoking constricts blood vessels and increases the work of the heart.
- Any disorder that can impair heart function, such as high blood pressure, should be treated and controlled.
- While rest is recommended during the acute inflammatory stage of myocarditis, it is essential that patients lead as normal a life as possible after that. Adopts life-style as vigorous as possible within the capacity of the heart muscle—that is, its ability to provide inadequate blood supply to meet the demands of the body. In other words, do not go beyond the body's limits. Avoid sudden stresses, like lifting heavy objects or exercising with free weights. Instead, opt for types of exercise—such as walking, bicycling, swimming—that are ideal for patients with cardiovascular disease.  
*Caution:* People with hypertrophic cardiomyopathy should consult physician before choosing an exercise regimen.
- Inquire about heart transplant if symptoms are not well control led with medications. Modern scientific advances have made heart transplantation a feasible alternative for some patients with severe cardiomyopathy.

result from nutritional deficiencies that occur when alcohol is the person's main source of caloric intake. In some drinkers, alcohol primarily attacks the liver, causing cirrhosis, and in others, mainly the heart, but severe damage usually does not occur in both organs at the same time.

Alcoholic cardiomyopathy can develop after five to ten years of excessive alcohol use. Examples of excessive amounts are two-thirds of a pint of whiskey or gin, one quart of wine, or two quarts of beer daily, although individual susceptibility to and tolerance of alcohol varies greatly. Most patients with this type of cardiomyopathy are males, possibly because there are more heavy drinkers among men than women.

### NUTRITIONAL ABNORMALITIES

Cardiomyopathy can also be caused by severe nutritional deficiency (which can also be related to alcohol use). The heart muscle, like any other muscle, can be damaged by chronic deficiency in certain vitamins, particularly vitamin B-1, or in minerals. In some developing countries, nutritional-deficiency-related cardiomyopathy is more common than coronary artery disease, the predominant form of heart disease in the United States.

### PERIPARTUM CARDIOMYOPATHY

For an unknown reason, cardiomyopathy sometimes develops in connection with pregnancy and is referred to as peripartum (*peri* meaning around or at the time of, *partum* meaning labor or childbirth) cardiomyopathy. During the last month of pregnancy or within several months following delivery, the woman develops heart muscle inflammation that appears to be unrelated to any infection or other known causes. The condition may result in severe and irreversible heart failure, although many patients recover completely. Women who survive the illness are at a high risk of developing this type of heart muscle disease in subsequent pregnancies. Peripartum cardiomyopathy occurs with particularly high frequency in African-American women in the United States and is a common cause of heart failure among women of childbearing age in some African countries.

### GENETIC DISORDERS

Dilated cardiomyopathy is known to develop in patients with some genetic disorders that affect the muscles or nerves of the back, arms, and legs. (Such diseases include progressive muscular dystrophy, myo-

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tonic muscular dystrophy, and Friedreich's ataxia.) There are also cases when dilated cardiomyopathy is not associated with muscle disorders but appears to be genetic in origin because several members of the same family are affected. However, because no abnormal gene has been identified, it is uncertain whether clustering of this condition within some families results from genetic or environmental factors.

### DIAGNOSIS OF DILATED CARDIOMYOPATHY

In most cases, dilated cardiomyopathy is preceded by heart muscle inflammation that produces flu-like symptoms such as fever, chills, and muscle aches. These symptoms are so common and vague that the cardiomyopathy is usually not diagnosed until heart muscle injury has caused impaired heart function and produced symptoms of heart failure.

Diagnosis is based on assessing the size and function of the heart chambers. A chest X-ray typically reveals the main features of dilated cardiomyopathy: an enlarged heart and fluid congestion in the lungs. An electrocardiogram may show evidence of heart damage. Characteristic abnormalities can also be detected using echocardiography or radionuclide angiography, often called the MUGA scan (equilibrium radionuclide angiogram).

If diagnosis remains in doubt, heart catheterization, sometimes accompanied by a heart biopsy, may be performed. Catheterization allows a physician to measure pressures in the heart chambers and to see the heart's structures when a contrast dye is injected into its chambers and vessels through the catheter—a thin plastic tube that is inserted in an artery or vein and threaded through it to the heart. During the procedure, X-ray images of the heart are recorded on film or videotape. In biopsy, a small sample of tissue is removed from the heart wall and examined under a light or electron microscope. The combined findings of catheterization and heart biopsy usually make it possible to distinguish dilated cardiomyopathy from other forms of heart disease.

Tests may also be used to rule out recognized causes of dilated cardiomyopathy. In most cases no cause can be established. However, blood tests, for example, may sometimes show that the patient has had a recent viral infection known to be associated with cardiomyopathy.

### TREATMENT

When the cause of dilated cardiomyopathy is known, therapy is aimed at treating the underlying disorder,

such as a curable infection or nutritional deficiency. For example, in the case of heart muscle disease caused by alcohol consumption, treatment entails total abstinence. Alcoholic cardiomyopathy is one of the few for which there exists a specific treatment, and patients with this type of heart muscle disease have a good prognosis if they follow the prescribed treatment.

Because in most cases the cause of dilated cardiomyopathy is unknown, treatment focuses on relieving the symptoms and improving the function of the injured heart chambers. Patients receive medications that enhance the contraction capacity of the heart muscle. The few drugs that produce this effect work indirectly, by increasing the level of calcium inside the heart cells. (Calcium initiates heart muscle contractions.) Digitalis and its derivatives such as digoxin (Lanoxin and others), the oldest and best-known of such drugs, are usually administered orally but may, in some circumstances, be given by an intravenous injection. More potent cardiac stimulants such as dobutamine (Dobutrex), dopamine (Intropin), and amrinone (Inocor), can be given only intravenously and are therefore primarily reserved for use in the hospital in more serious situations. Oral forms of such medications are currently being developed.

Diuretics are prescribed to relieve lung congestion and remove excess body fluid. Commonly referred to as "water pills," they facilitate the kidney's excretion of excess salt and fluid into the urine. While these drugs, which reduce congestion and swelling, are an essential part of heart failure therapy, patients can help the physician decrease the dosages of diuretics by limiting the amount of salt in their diet.

Function of the impaired heart can be significantly improved by altering the conditions under which it must work. Because the body responds to heart failure by constricting blood flow to all but the most vital organs, drugs that dilate blood vessels (vasodilators) reduce the work of the heart by decreasing resistance to bloodflow. Angiotensin-converting enzyme (ACE) inhibitors, a class of vasodilators, are particularly effective in heart failure treatment.

In the presence of an active inflammation in the heart (usually confirmed by heart biopsy), anti-inflammatory drugs such as steroids (cortisone) may be prescribed. Although these medications have been extensively studied, it has not been proved whether they benefit patients with newly diagnosed dilated cardiomyopathy.

In the early, acute stages of cardiomyopathy, when signs of heart inflammation and ongoing muscle injury are present, patients are told to rest. With re-

covery from acute illness, they are advised to engage in regular physical activity to the extent that their heart function permits.

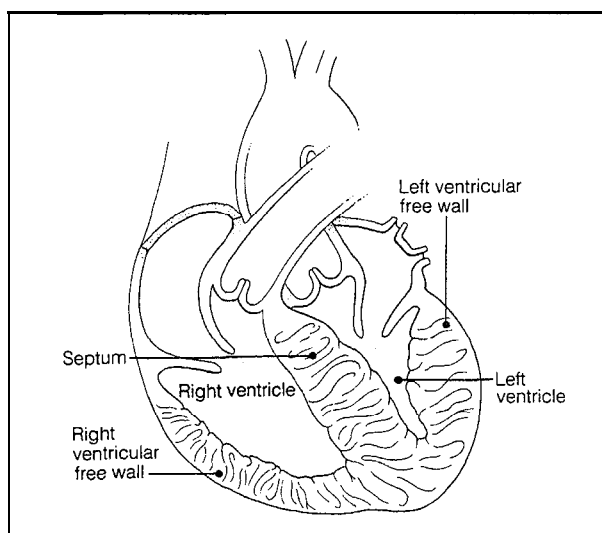
In extreme cases, when cardiomyopathy has progressed to the point when medical treatment can no longer relieve the symptoms, a heart transplant may be considered. Transplants increase life expectancy in persons with advanced heart failure who might otherwise be expected to live less than six months. Currently, 80 to 90 percent of heart transplant recipients survive at least one year, and more than 75 percent survive five years. The scarcity of donor organs and the high level of sophisticated care required for a successful transplant make this option available only to a small number of patients—approximately 2,000 per year in the United States.

## PROGNOSIS

By the time patients with dilated cardiomyopathy develop heart failure symptoms, the disease has usually reached an advanced stage and prognosis is poor. About 50 percent of patients achieve the average survival rate five years after initial diagnosis, and 25 percent survive ten or more years after diagnosis. These statistics have not changed significantly in several decades, although current forms of therapy for heart failure offer promise that this bleak prognosis may improve.

## HYPERTROPHIC CARDIOMYOPATHY

This rare disease is the second most common type of cardiomyopathy. Hypertrophic cardiomyopathy is also known as idiopathic hypertrophic subaortic stenosis (IHSS) or asymmetric septal hypertrophy (ASH). The disease is characterized by a disorderly growth of heart muscle fibers causing the heart chambers to become thick-walled and bulky. All the chambers are affected, but the thickening is generally most striking in the walls of the left ventricle. Most commonly, one of the walls, the septum, which separates the right and left ventricles, is asymmetrically enlarged. The distorted left ventricle contracts, but the supply of blood to the brain and other vital organs may be inadequate because blood is trapped within the heart during contractions. Mitral valve function is often disrupted by the structural abnormalities in the left ventricle with backward leakage of blood. (See Figure 15.2.)



*Figure 15.2*  
in hypertrophic cardiomyopathy, the muscle mass of the left and occasionally the right ventricle is enlarged, causing the heart chambers to stiffen. The septum between the ventricles often becomes disproportionately thickened; this is noticeable when it is compared to the freestanding portion of the left ventricular wall.

Fainting during physical exertion is often the first and most dramatic symptom of hypertrophic cardiomyopathy. During periods of exertion, as the body stimulates the heart to beat more forcefully, blood has a greater tendency to become trapped within the vigorously contracting chambers. As a result, the person may faint or, in extreme cases, die. Unexplained death during athletic activity always leads physicians to suspect undiagnosed hypertrophic cardiomyopathy. Several world-class athletes have suffered this type of sudden death in the past few years.

## COMPLICATIONS OF HYPERTROPHIC CARDIOMYOPATHY

The walls of the hypertrophied heart are not always adequately nourished by blood vessels. This leads to muscle injury and scarring—a common complication of hypertrophic cardiomyopathy. In advanced stages of the disease, the thickened, deformed, and scarred walls of the heart prevent the chambers from contracting effectively and filling up completely, leading to severe loss of heart function.

Sudden death—the most unpredictable and devastating complication of this type of cardiomyopathy—is most commonly caused by ventricular fibrillation, abnormally fast and disorganized contractions of the left ventricle that interfere with effective pumping. The hypertrophied heart is prone to fibrillation because the disorderly overgrowth of muscle creates abnormal sites and pathways for the heart's electrical activity.

### **CAUSES OF HYPERTROPHIC CARDIOMYOPATHY**

Hypertrophic cardiomyopathy is usually congenital. In half of the cases, the patient has inherited an abnormal gene from one parent. The pattern of genetic transmission is termed "autosomal dominant." This means that a copy of the gene from only one parent is needed for the disease to develop in the child. However, some siblings in the family may carry the gene but have hardly any trace of hypertrophic cardiomyopathy while others may die at a young age.

In the rest of the cases, neither parent carries a gene for hypertrophic cardiomyopathy, and the child is believed to develop the disease because of a spontaneous gene mutation.

### **SYMPTOMS OF HYPERTROPHIC CARDIOMYOPATHY**

People with hypertrophic cardiomyopathy may experience a variety of symptoms during the course of the disease. Some have absolutely no symptoms for years and are only diagnosed after having an electrocardiogram for another reason, or when they come for an examination because a family member is known to carry the disease. Sometimes the disorder is detected for the first time at autopsy after the patient's death.

Although predisposition for developing hypertrophic cardiomyopathy is present from birth, the abnormal heart muscle proliferation may actually begin during the adolescent growth spurt. In such cases, symptoms may develop rather abruptly during teenage years.

Fainting upon exertion and chest pain similar to the angina of coronary artery disease may be early symptoms of hypertrophic cardiomyopathy. Some patients experience palpitations because of abnormal heart rhythms. Eventually, symptoms of congestive heart failure may become prominent.

### **DIAGNOSIS OF HYPERTROPHIC CARDIOMYOPATHY**

The most important diagnostic tool in assessing hypertrophic cardiomyopathy is echocardiography. It provides images and reveals blood flow patterns that allow physicians to identify the distinctive abnormalities in the heart walls and valves. Other characteristic features of the disease are often recorded on chest X-rays, on electrocardiograms, and during cardiac catheterization.

### **TREATMENT**

When hypertrophic cardiomyopathy is severe, patients are advised to limit stressful physical activity, particularly strenuous competitive sports. They may also be given drugs to relieve symptoms. Traditionally, drugs called beta blockers have been used to prevent a rapid heartbeat and decrease the excessive force of contractions. Antiarrhythmic drugs are often prescribed to treat abnormal heart rhythms. In the past decade, calcium channel blockers, particularly verapamil (Calan), have been shown to be especially effective for relief of symptoms. Like beta blockers, calcium antagonists reduce the force of the heart's contractions, but they also increase the flexibility of the bulky heart chambers. These combined effects increase the efficiency of pumping and reduce congestion.

Surgery may be performed in people with hypertrophic cardiomyopathy whose symptoms are not relieved by medications. The surgeon may remove the excess muscle tissue that obstructs the blood flow in the heart chamber.

### **PROGNOSIS**

The course of hypertrophic cardiomyopathy varies. The condition may remain stable over decades or may progress slowly. Approximately 4 percent of people with the disease die annually, most of them from sudden death, which may occur at any stage of the disease. The younger the age at which the disease appears, the higher the risk of sudden death.

### **SPECIAL RISKS OF HYPERTROPHIC CARDIOMYOPATHY FOR YOUNG ATHLETES**

People who regularly participate in strenuous physical exercise, such as professional athletes or those who train for marathons, may experience changes in the function and structure of their hearts. These adaptations, which include a slower than normal heartbeat and an overall enlarged heart, enable the heart to deliver more oxygen to the tissues in the limbs in order to sustain and enhance athletic performance.

Heart muscle thickness may increase in an athlete's heart. Generally, this is nothing to worry about, but athletes—especially teenagers—should be monitored to ensure that hypertrophic cardiomyopathy is not an underlying cause of heart enlargement.

A good deal of publicity was given to this disease when Hank Gathers, the college basketball star, died

suddenly in 1990 while playing in a conference championship game for Loyola Marymount in Los Angeles. Gathers was known to have an enlarged heart and was taking medication at the time of his death. Most often, though, a young athlete with this condition is not aware of it.

Although these incidents seem to make headlines frequently, they are relatively rare. According to the *American Journal of Diseases of Children*, each year there are 1 or 2 cases per 200,000 athletes 30 years old and younger. Of those few cases, hypertrophic cardiomyopathy is involved about 60 percent of the time. Even so, anyone who begins participating in a new sport should undergo a complete medical history and physical examination—including orthopedic, neurologic, and cardiovascular assessment. If hypertrophic cardiomyopathy is diagnosed, the athlete should avoid strenuous sports.

### **HYPERTROPHIC CARDIOMYOPATHY CAUSED BY DRUG THERAPY**

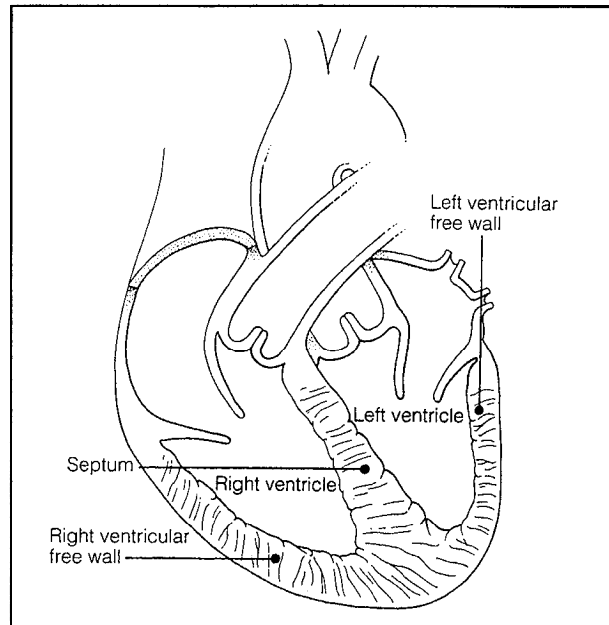
A number of drugs can have toxic effects on the heart. Perhaps most damaging are the chemotherapy drugs doxorubicin (Adriamycin and Rubex) and daunorubicin (Cerubidine). Although they are effective in the treatment of leukemia and other cancers, large doses can be toxic to heart muscle. Changes in the radio-nuclide angiogram (See Chapter 10) may help detect this type of reaction.

Drugs used to treat emotional and psychiatric problems can also alter heart function. Phenothiazine drugs such as chlorpromazine (Thorazine) and thioridazine (Mellaril) and tricyclic antidepressant drugs such as imipramine (Tofranil) and amitriptyline (Endep and Elavil) may cause electrocardiographic abnormalities and heart rhythm disorders.

If possible, these drugs should be avoided by anyone with a history of heart disease. In any case, people taking these drugs, especially large or continuous doses of them, should undergo heart examinations, including electrocardiograms, to detect toxic effects. If these are detected, the drugs will most often be discontinued, and alternate treatment will be instituted.

## **RESTRICTIVE CARDIOMYOPATHY**

Restrictive cardiomyopathy is extremely rare. In this type of heart muscle disease, abnormal cells, proteins,



*Figure 15.3*

*In restrictive cardiomyopathy, abnormal cells, proteins, or scar tissue infiltrate the heart, causing the chambers to become thick and bulky.*

or scar tissue infiltrate the muscle and structures of the heart, causing the chambers to become stiff and bulky. The heart may initially contract normally, but the rigid chambers restrict the return of blood to the heart. As a consequence, high pressures are needed to fill the heart chambers, forcing the blood back into various tissues and organs—the lungs, abdomen, arms, and legs. Eventually, heart muscle is damaged and contractions impaired. (See Figure 15.3.)

### **CAUSES OF RESTRICTIVE CARDIOMYOPATHY**

The most common cause of restrictive cardiomyopathy in the United States is amyloidosis, a disease that is sometimes associated with cancers of the blood. In amyloid heart disease, abnormal proteins are deposited around the heart cells, making the chambers thick, inflexible, and waxy in appearance. Other rare diseases can also fill the heart's walls with abnormal cells or excessive starlike tissue. For example, sarcoidosis (a disease characterized by the growth of numerous tumors called granulomas throughout the body), hemochromatosis (a metabolic disorder characterized by a buildup of iron in the body), endomyocardial fibrosis (an abnormality of heart muscle tissue), and some cancers that metastasize into the heart walls are all possible causes. (See box, "Possible Causes of Restrictive Cardiomyopathy.")

## Possible Causes of Restrictive Cardiomyopathy

- *Amyloidosis*: The heart muscle is infiltrated by amyloid, a fibrous protein, causing the heart chambers to stiffen.
- *Sarcoidosis*: An inflammatory disease that affects many tissues, especially the lungs.
- *Hemochromatosis*: Iron deposits form in the tissues, impairing heart function (and also resulting in liver disease and diabetes).
- *Endomyocardial fibrosis*: A progressive disease characterized by fibrous lesions on the inner walls of one or both ventricles. A frequent cause of heart failure in Africa.

### SYMPTOMS OF RESTRICTIVE CARDIOMYOPATHY

The major symptoms of restrictive cardiomyopathy stem from the stiffening of the chambers, which impedes blood return to the heart. Congestion occurs in the lungs but is typically most severe in the organs of the abdomen (the liver, stomach, and intestines) as well as the legs. Patients tend to tire easily and complain of swelling, nausea, bloating, and poor appetite. Symptoms of advanced restrictive cardiomyopathy typically include significant weight loss, muscle wasting, and abdominal swelling. Patients with this condition are commonly misdiagnosed initially as having cirrhosis or cancer.

### DIAGNOSIS AND TREATMENT OF RESTRICTIVE CARDIOMYOPATHY

It is occasionally possible for a physician to suspect the diagnosis based on a patient's symptoms of heart disease and the presence of an underlying disease. Imaging techniques that show details of the heart walls and function help a physician detect the restrictive movements of the heart chambers. Such techniques include echocardiography, computerized tomography (CT) scanning, and magnetic resonance imaging (MRI). Definitive diagnosis can be made by biopsy of the heart muscle.

Accurate diagnosis is important, since restrictive cardiomyopathy shares many clinical features and symptoms with a more treatable form of heart disease, constrictive pericarditis. Pericarditis is an inflammation and thickening of the membrane

surrounding the heart—the pericardium. Severe or chronic pericarditis may lead to pericardial constriction, which disrupts the filling of the heart in the same manner as do the stiff chambers of restrictive cardiomyopathy. Constrictive pericarditis can often be treated effectively with surgery. In contrast, most forms of restrictive cardiomyopathy cannot be cured, and treatment is focused on alleviating symptoms.

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## CARDIOMYOPATHY FROM THE LACK OF OXYGEN (ISCHEMIA)

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Severe heart injury caused by a major heart attack or multiple smaller heart attacks may result in heart enlargement and thinning of the chamber walls—abnormalities resembling those observed in dilated cardiomyopathy. This type of heart disease, called ischemic cardiomyopathy, typically develops in patients with severe coronary artery disease, often complicated by other conditions such as diabetes and hypertension.

Heart failure symptoms in ischemic cardiomyopathy are similar to those found in dilated cardiomyopathy. However, ischemic disease is more likely to be accompanied by symptoms of coronary artery disease, such as angina (chest pain). Diagnosis is typically based on a history of heart attacks and studies that demonstrate poor function in major portions of the left ventricle. The diagnosis can be confirmed by coronary angiography, which reveals areas of narrowing and blockage in the coronary blood vessels.

Patients with ischemic cardiomyopathy are treated with medications that relieve heart failure symptoms and improve blood flow through the diseased coronary arteries (nitroglycerine, some types of calcium channel blockers, and ACE inhibitors). When symptoms of heart failure and coronary artery disease cannot be controlled with medications, coronary angioplasty or surgery may be considered. Angioplasty and coronary artery bypass grafting may help increase blood flow to the heart, which in turn enhances heart muscle function.

When heart failure symptoms are advanced and cannot be improved by drug therapy or surgery, patients may be referred for a heart transplant. Patients with ischemic cardiomyopathy now account for approximately half of all heart transplant recipients.