

CARDIOMYOPATHY

What is cardiomyopathy?

Cardiomyopathy (*KAR-de-o-mi-OP-a-the*) refers to diseases of the heart muscle. These diseases have a variety of causes, symptoms, and treatments. In cardiomyopathy, the heart muscle becomes enlarged or abnormally thick or rigid. In rare cases, the muscle tissue in the heart is replaced with scar tissue.

As cardiomyopathy progresses, the heart becomes weaker and less able to pump blood through the body. This can lead to heart failure, arrhythmias (*ah-RITH-me-ahs; abnormal heart rhythms*), fluid buildup in the lungs or legs, and, more rarely, endocarditis (*a bacterial infection of the lining of the heart*). The weakening of the heart also can lead to other severe complications.

The four main types of cardiomyopathy are:

- Dilated cardiomyopathy
- Hypertrophic (hi-per-TROF-ik) cardiomyopathy
- Restrictive cardiomyopathy
- Arrhythmogenic (a-rith-mo-JEN-ik) right ventricular dysplasia (ARVD)

Cardiomyopathy can have a specific cause, such as damage to the heart from a heart attack, high blood pressure, or a viral infection. Some types of cardiomyopathy are caused by a gene mutation and run in families. In many cases, the cause is unknown.

Cardiomyopathy can affect people of all ages, from babies to older adults. However, certain age groups are more likely to have certain types of cardiomyopathy. Treatment may involve medicines, surgery, nonsurgical procedures, and lifestyle changes.

Outlook

Some people live long, healthy lives in spite of having cardiomyopathy. Some people don't even realize that they have the disease because they have no symptoms. In other people, the disease develops rapidly, symptoms are

severe, and serious complications develop. Current treatments can reduce symptoms and complications of cardiomyopathy.

Types of Cardiomyopathy

Dilated cardiomyopathy

Dilated cardiomyopathy is the most common form of cardiomyopathy. It generally occurs in adults aged 20 to 60 years. Men are more likely than women to develop dilated cardiomyopathy.

Dilated cardiomyopathy affects the heart's ventricles (*VEN-trih-kuls*) and atria. The ventricles are the two lower chambers of the heart, and the atria are the two upper chambers. Dilated cardiomyopathy usually starts in the left ventricle, where the heart muscle begins to dilate or stretch and become thinner. This leads to enlargement of the inside of the ventricle. The problem often spreads to the right ventricle and then to the atria as the disease gets worse.

When the chambers dilate, the heart can't pump blood very well. The heart tries to cope by dilating the chambers even more. Over time, the heart becomes weaker and heart failure can occur. Symptoms of heart failure include feeling tired, swelling of the legs and feet, and shortness of breath. Dilated cardiomyopathy also can lead to heart valve problems, arrhythmias, and blood clots in the heart. Having advanced dilated cardiomyopathy is a common reason for needing a heart transplant.

Up to one-half of all cases of dilated cardiomyopathy may be hereditary (*passed down in the genes from parent to child*). These cases are called familial dilated cardiomyopathy. Dilated cardiomyopathy also can be a complication of many conditions, including coronary artery disease and high blood pressure. It also can be caused by viral infections, excessive use of alcohol, and exposure to certain drugs (including cocaine, amphetamines, and some drugs used in cancer treatments). In some cases, no cause can be found.

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy occurs when the heart muscle thickens abnormally. The thickening generally happens in the left ventricle, the

heart's main pumping chamber. This type of cardiomyopathy can affect people of any age.

Hypertrophic cardiomyopathy can be obstructive or nonobstructive. In the obstructive type, the septum (the wall that divides the left and right sides of the heart) thickens and bulges into the left ventricle. This bulge blocks the flow of blood out of the ventricle. The ventricle must work much harder to pump blood past the blockage and out to the body. Symptoms can include chest pain, dizziness, shortness of breath, or fainting.

Obstructive hypertrophic cardiomyopathy also can affect the heart's mitral (*MI-trul*) valve, causing blood to leak backward through the valve.

In nonobstructive hypertrophic cardiomyopathy, the thickened heart muscle does not block the flow of blood out of the ventricle. The entire ventricle may become thicker (*symmetric ventricular hypertrophy*) or it may happen only at the bottom of the heart (*apical hypertrophy*). The right ventricle also may be affected.

In both kinds of hypertrophic cardiomyopathy, the thickened muscle makes the inside of the left ventricle smaller so that it holds less blood. The walls of the ventricles also may become stiff. As a result, they are less able to relax and fill with blood. This causes increased pressure in the ventricles and the blood vessels of the lungs. Changes also occur to the cells in the damaged heart muscle. This may interfere with the heart's electrical signals, leading to arrhythmias.

Some people with hypertrophic cardiomyopathy have no symptoms, and the condition does not affect their lives. Others have severe symptoms or develop complications such as serious arrhythmias. A few people with the condition have sudden cardiac arrest because of dangerous arrhythmias.

Hypertrophic cardiomyopathy can be inherited because of a gene mutation or develop over time because of high blood pressure or aging. Often, the cause is unknown.

Restrictive Cardiomyopathy

Restrictive cardiomyopathy tends to mostly affect older adults. In this cardiomyopathy, the ventricles become stiff and rigid due to replacement of the normal heart muscle with abnormal tissue, such as scar tissue. As a result, the ventricles cannot relax normally and expand to fill with blood, which causes the atria to become enlarged. Eventually, blood flow in the heart is reduced, and complications such as heart failure or arrhythmias occur.

Restrictive cardiomyopathy can occur for no known reason, or it can develop because the person has another disease. Some of the diseases that can cause restrictive cardiomyopathy include hemochromatosis, sarcoidosis, amyloidosis, and connective tissue disorders. Restrictive cardiomyopathy also can occur as a result of radiation treatments, infections, or scarring after surgery.

Arrhythmogenic Right Ventricular Dysplasia

Arrhythmogenic right ventricular dysplasia (ARVD) is a rare type of cardiomyopathy. ARVD develops when the muscle tissue in the right ventricle dies and is replaced with scar tissue. This process causes problems in the heart's electrical signaling, resulting in arrhythmias. Symptoms include a feeling of strong or irregular heartbeats (palpitations) and fainting after exercise.

ARVD usually develops in teens or young adults and is often the cause of sudden cardiac death in young athletes. ARVD is thought to be an inherited disease.

Other Names for Cardiomyopathy

- Dilated cardiomyopathy
- Familial dilated cardiomyopathy
- Congestive cardiomyopathy
- Idiopathic dilated cardiomyopathy
- Hypertrophic cardiomyopathy
- Hypertrophic obstructive cardiomyopathy
- Asymmetric septal hypertrophy

- Idiopathic hypertrophic subaortic stenosis
- Familial hypertrophic cardiomyopathy
- Arrhythmogenic right ventricular dysplasia
- Right ventricular dysplasia
- Right ventricular cardiomyopathy
- Restrictive cardiomyopathy
- Arrhythmogenic ventricular cardiomyopathy

What causes cardiomyopathy?

Many times, the cause of cardiomyopathy is unknown. When this happens, the disease is called idiopathic (*or primary*) cardiomyopathy. The majority of cardiomyopathies in children are idiopathic.

Sometimes, cardiomyopathy is inherited (passed down in the genes from parent to child) or caused by another disease or condition.

Dilated Cardiomyopathy

Dilated cardiomyopathy can be inherited. It also can be caused by certain diseases, conditions, and substances, including:

- Coronary artery disease and heart attacks (ischemic cardiomyopathy)
- Infections, especially viral infections that cause the heart muscle to become inflamed (myocarditis)
- Alcohol, especially when a person has a poor diet (alcoholic cardiomyopathy)
- Complications during the last month of pregnancy or within 5 months of birth (peripartum cardiomyopathy)
- Certain toxins, such as cobalt
- Certain drugs, such as cocaine, amphetamines, and two medicines used to treat cancer (doxorubicin and daunorubicin)
- Diseases such as diabetes and thyroid disease

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy can be inherited. It also can develop over time because of high blood pressure or aging. Often, the cause of hypertrophic cardiomyopathy is unknown.

Restrictive Cardiomyopathy

Certain diseases and conditions can cause restrictive cardiomyopathy, including:

- Hemochromatosis, a condition in which too much iron is deposited into tissues, including heart tissue
- Amyloidosis, a disease in which abnormal proteins are deposited into heart tissue
- Sarcoidosis, a disease in which inflammation produces tiny lumps of cells in various organs in the body, including the heart
- Connective tissue disorders

Arrhythmogenic Right Ventricular Dysplasia

Arrhythmogenic right ventricular dysplasia is thought to be an inherited disease.

Who is at risk for cardiomyopathy?

Populations Affected

People of all ages can develop cardiomyopathy, but certain cardiomyopathies are more common in certain groups:

- African Americans are more likely to have dilated cardiomyopathy compared to Caucasians.
- Men are more likely to have dilated cardiomyopathy compared to women.
- Teens and young adults are more likely to have arrhythmogenic right ventricular dysplasia compared to older people.

Major Risk Factors

Major risk factors for developing cardiomyopathy include:

- Having a family history of cardiomyopathy, heart failure, or sudden cardiac death
- Having a disease or condition that can lead to cardiomyopathy, such as:
 - Coronary artery disease
 - A previous heart attack
 - Myocarditis
- Diseases that can damage the heart (for example, hemochromatosis, sarcoidosis, or amyloidosis)

- Long-term alcoholism
- Long-term high blood pressure
- Diabetes and other metabolic diseases

Some people with cardiomyopathy never have symptoms. That is why it's important to identify people who may be at high risk for this disease so that potential problems (such as serious arrhythmias or sudden cardiac death) can be prevented.

What are the signs of symptoms of cardiomyopathy?

Some people with cardiomyopathy never have symptoms, and others have no symptoms in the early stages of the disease. As cardiomyopathy progresses and the heart weakens, signs and symptoms of heart failure usually appear. These signs and symptoms include:

- Tiredness
- Weakness
- Shortness of breath after exercise or even at rest
- Swelling of the abdomen, legs, ankles, and feet

Other signs and symptoms can include dizziness, lightheadedness, fainting during exercise, abnormal heart rhythms (arrhythmias), and an extra or unusual sound heard during the heartbeat (heart murmur).

How is cardiomyopathy diagnosed?

If cardiomyopathy is suspected, the diagnosis is based on a person's:

- Symptoms and medical history
- Family history of cardiomyopathy, heart failure, or sudden cardiac arrest
- Physical exam
- Results on diagnostic tests and procedures

The physical exam will identify a number of findings in patients with cardiomyopathy. The doctor will use a stethoscope to listen to the person's heart and lungs for sounds that may suggest the presence of cardiomyopathy. These sounds may even indicate a certain type of cardiomyopathy. For example, the loudness, timing, and location of a heart murmur may suggest that a person has hypertrophic obstructive cardiomyopathy. A "crackling"

sound in the lungs may be a sign of heart failure, which often develops in the later stages of cardiomyopathy.

Physical signs also help the doctor diagnose cardiomyopathy. Swelling of the abdomen, legs, or feet may indicate excess fluid, which is a sign of heart failure.

Sometimes, doctors discover cardiomyopathy during a routine exam if they hear a heart murmur or if the patient has an abnormal electrocardiogram.

Specialists Involved

Often a cardiologist or pediatric cardiologist is involved in the care of a patient with cardiomyopathy. A cardiologist is a doctor who specializes in heart diseases. A pediatric cardiologist specializes in children's heart diseases.

Diagnostic Tests and Procedures

Doctors may order one or more tests to diagnose cardiomyopathy, including:

- EKG (electrocardiogram). This test measures the rate and regularity of the heartbeat and can detect arrhythmias.
- Holter monitor (ambulatory EKG/ECG). This test records the EKG readings for a continuous 24-hour period. It can detect arrhythmias that occur only rarely throughout the day. A small monitoring device is attached to patches (electrodes) that are placed on the patient's chest. The device is carried in a pouch around the neck or attached to a belt.
- Echocardiogram. This test uses sound waves to create a moving picture of your heart. Echocardiogram provides information about the size and shape of your heart and how well your heart chambers and valves are functioning. The test also can identify areas of poor blood flow to the heart, areas of heart muscle that are not contracting normally, and previous injury to the heart muscle caused by poor blood flow.
- There are several different types of echocardiograms, including a stress echocardiogram. During this test, an echocardiogram is done both before and after your heart is stressed either by having you exercise or by injecting a medicine into your bloodstream that

makes your heart beat faster and work harder. A stress echocardiogram is usually done to find out if you have decreased blood flow to your heart (coronary artery disease).

Echocardiogram is a good way to diagnose hypertrophic cardiomyopathy because it shows the thickened walls of the heart.

- Transesophageal (tranz-ih-sof-uh-JEE-ul) echocardiography (TEE). In this test, the doctor inserts an ultrasound probe into the throat after the patient is sedated. TEE provides a view of the back of the heart.
- Stress Test. Some heart problems are easier to diagnose when your heart is working harder and beating faster than when it's at rest. During stress testing, you exercise (or are given medicine if you are unable to exercise) to make your heart work harder and beat faster while heart tests are performed.
- During exercise stress testing, your blood pressure and EKG readings are monitored while you walk or run on a treadmill or pedal a bicycle. Other heart tests, such as nuclear heart scanning or echocardiography, also can be done at the same time. These would be ordered if your doctor needs more information than the exercise stress test can provide about how well your heart is working.
- If you are unable to exercise, a medicine can be injected through an intravenous line (IV) into your bloodstream to make your heart work harder and beat faster, as if you are exercising on a treadmill or bicycle. Nuclear heart scanning or echocardiography is then usually done.
- During nuclear heart scanning, radioactive tracer is injected into your bloodstream, and a special camera shows the flow of blood through your heart and arteries. Echocardiography uses sound waves to show blood flow through the chambers and valves of your heart and to show the strength of your heart muscle.
- Your doctor also may order two newer tests along with stress testing if more information is needed about how well your heart works. These new tests are magnetic resonance imaging (MRI) and positron emission tomography (PET) scanning of the heart. MRI shows detailed images of the structures and beating of your heart, which may help your doctor better assess if parts of your heart are weak or damaged. PET scanning shows the level of chemical activity in different areas of your heart. This can help your doctor

determine if enough blood is flowing to the areas of your heart. A PET scan can show decreased blood flow caused by disease or damaged muscles that may not be detected by other scanning methods.

- Chest x ray. A chest x ray takes a picture of the organs and structures inside the chest, including the heart, lungs, and blood vessels. This test can show whether the heart is enlarged or whether fluid is building up in the lungs.
- Blood tests, such as complete blood count, blood chemistries, and cardiac enzymes. These tests are done to provide information on the condition of the heart and to rule out other conditions.

The doctor may order additional tests to confirm the diagnosis or if surgery is planned. These tests may include:

- Cardiac catheterization. With this test doctors can check the pressure and blood flow in the heart's chambers, collect blood samples from the heart, and examine the arteries of the heart using x ray. A thin, flexible tube (catheter) is passed through an artery in the upper thigh (groin) or in the arm to reach the coronary arteries. This allows the doctor to study the inside of the arteries to look for blockages.
- Coronary angiography. This test is usually performed along with cardiac catheterization. Angiography enables the doctor to see the flow of blood to the heart muscle. A dye that can be seen on an x ray image is injected into the coronary arteries. Dye also can be injected into the chambers to evaluate the pumping function of the heart.
- Myocardial biopsy. In this test, the doctor removes a piece of heart muscle to look at under a microscope. The biopsy can be done during a cardiac catheterization and is useful in diagnosing some types of cardiomyopathy.

Because some types of cardiomyopathy run in families, the doctor may recommend looking for the disease in the parents, brothers and sisters, and children of people with cardiomyopathy.

Genetic counseling may be recommended. Genetic counseling is useful to help define and explain how the disease runs in families and to determine the chances of parents passing it on to their children.

How is cardiomyopathy treated?

Not everyone with cardiomyopathy needs treatment. People who have no symptoms may not need treatment. In some cases, dilated cardiomyopathy that comes on suddenly may even go away on its own. For other people with cardiomyopathy, treatment is necessary.

Specific treatments depend on the type of cardiomyopathy, how severe the symptoms and complications are, and the age and overall health of the person.

Goals of Treatment

The main goals of treating cardiomyopathy are to:

- Manage any conditions that cause or contribute to the cardiomyopathy
- Control symptoms so that the person can live as normally as possible
- Stop the disease from getting worse
- Reduce complications and the chance of sudden cardiac death

Specific Types of Treatment

Treatments for cardiomyopathy may include medicines, surgery, nonsurgical procedures, and lifestyle changes.

Medicines

A number of medicines may be used to treat cardiomyopathy, including:

- Diuretics, which remove excess fluid and sodium from the body.
- Angiotensin-converting enzyme (ACE) inhibitors, which lower blood pressure and reduce stress on the heart.
- Beta-blockers, which slow the heart rate by reducing the speed of the heart's contractions. These medicines also lower blood pressure.

- Calcium channel blockers, which slow a rapid heartbeat by reducing the force and rate of heart contractions. These medicines also lower blood pressure.
- Digoxin, which increases the force of heart contractions and slows the heartbeat.
- Anticoagulants, which prevent blood clots from forming. Anticoagulants are often used in the treatment of dilated cardiomyopathy.
- Antiarrhythmia medicines, which keep the heart beating in a normal rhythm.
- Antibiotics, which are used before dental or surgical procedures. Antibiotics help to prevent endocarditis, an infection of the heart walls, valves, and vessels.
- Corticosteroids, which reduce inflammation.

Surgery

Doctors can use several different types of surgery to treat cardiomyopathy, including removing part of the enlarged heart muscle (septal myectomy) and implanting devices that help the heart beat more effectively. Heart transplant is sometimes used in cases of severe heart failure.

Septal myectomy

Septal myectomy (also called septal myomectomy) is open-heart surgery for people with hypertrophic obstructive cardiomyopathy and severe symptoms. It is generally used in younger patients and when medicines aren't working well.

In septal myectomy, a surgeon removes part of the thickened septum that is bulging into the left ventricle. This widens the pathway in the ventricle that leads to the aortic valve and improves blood flow through the heart and out to the body. The tissue that is removed does not grow back. If necessary, the mitral valve can be repaired or replaced at the same time. This surgery is often successful, and the person can return to a normal life with no symptoms.

Surgically implanted devices

Surgeons can place several different types of devices in the heart to help it beat more effectively. One device is a pacemaker, which electronically helps

maintain normal heart rhythm. Sometimes, doctors choose to use a biventricular pacemaker, which coordinates contractions between the heart's left and right ventricles.

A left ventricular assist device (LVAD) helps the heart pump blood to the body. LVAD can be used as a long-term therapy or as a short-term treatment for people who are waiting for a heart transplant.

An implantable cardioverter defibrillator (ICD) is used in people who are at risk of life-threatening arrhythmia or sudden cardiac death. This small device is implanted in the chest and connected to the heart with wires. If the ICD senses a dangerous change in heart rhythm, it will send an electric shock to the heart to restore a normal heartbeat.

Heart transplant

In this surgery, a doctor replaces a person's diseased heart with a healthy heart from a person who has recently died. It is a last resort for people with heart failure when all other treatments have failed.

Nonsurgical Procedure

Alcohol septal ablation

In this procedure, a doctor injects ethanol (a type of alcohol) through a catheter into the small artery that supplies blood to the thickened area of heart muscle. The alcohol kills the cells and the thickened tissue shrinks to a more normal size. Blood can flow freely through the pathway in the ventricle that leads to the aortic valve, and symptoms improve.

Lifestyle Changes

The doctor may recommend lifestyle changes to manage a condition that is causing the cardiomyopathy. These changes may help reduce symptoms.

Lifestyle changes may include:

- Quitting smoking
- Losing excess weight
- Eating a low-salt diet
- Getting moderate exercise, such as walking, and avoiding strenuous exercise
- Avoiding the use of alcohol and illegal drugs
- Getting enough sleep and rest

- Reducing stress
- Treating underlying conditions, such as diabetes and high blood pressure

How can cardiomyopathy be prevented?

People can make lifestyle choices to reduce the risk of conditions that may lead to cardiomyopathy, such as coronary artery disease, high blood pressure, and heart attack. Examples of lifestyle choices include:

- Quitting smoking
- Eating a healthy diet and maintaining a healthy weight
- Getting regular physical exercise
- Avoiding the use of alcohol and illegal drugs

People also can control high blood pressure, high blood cholesterol, and diabetes by:

- Getting regular checkups with their doctors
- Following their doctors' advice about lifestyle changes
- Taking medicines as directed

Some types of cardiomyopathy, such as inherited forms, can't be prevented. Restrictive cardiomyopathy can't always be prevented because it occurs as the result of another disease. Sometimes, underlying diseases can be prevented or treated early enough to stop restrictive cardiomyopathy from developing.

It may be possible to prevent sudden cardiac death if doctors can identify a person at high risk of this event and treat him or her with an implantable cardioverter defibrillator.

*Source: National Heart Lung and Blood Institute, 8/2006
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