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# Cardiomyopathy

Cardiomyopathy refers to diseases of the heart muscle. These diseases have many causes, signs and symptoms as well as treatments. In most cases, cardiomyopathy causes the heart muscle to become enlarged, thick or rigid. In rare instances, diseased heart muscle tissue is replaced with scar tissue.

# What Is Cardiomyopathy in Adults?



Cardiomyopathy refers to diseases of the heart muscle. These diseases have many causes, <u>signs and symptoms</u> as well as <u>treatments</u>. In most cases, cardiomyopathy causes the heart muscle to become enlarged, thick or rigid. In rare instances, diseased heart muscle tissue is replaced with scar tissue.

As cardiomyopathy worsens, the heart becomes weaker. The heart becomes less able to pump blood throughout the body and incapable of maintaining a normal electrical rhythm. The result can be heart failure or irregular heartbeats called arrhythmias. A weakened heart also can cause other complications, such as heart valve problems.

### **Overview**

The main types of cardiomyopathy are:

- Dilated cardiomyopathy
- <u>Hypertrophic cardiomyopathy</u>
- <u>Restrictive cardiomyopathy</u>
- <u>Arrhythmogenic right ventricular dysplasia</u>
- Transthyretin amyloid cardiomyopathy (ATTR-CM)

Some other types of cardiomyopathy are called "unclassified cardiomyopathy." Yet another type is "stress-induced cardiomyopathy," also known as <u>broken heart</u> <u>syndrome</u>.

Cardiomyopathy can be "acquired," meaning it develops because of another disease, condition or factor. Or, cardiomyopathy can be "inherited," meaning the gene for the disease was passed on from a parent.

In many cases, the cause of cardiomyopathy isn't known. This is often the case <u>when the</u> <u>disease occurs in children</u>.

Cardiomyopathy can affect all ages, although certain age groups are more likely to have certain types of cardiomyopathy.

### Approaches to treatment

Some cases of cardiomyopathy have no signs or symptoms, and need no treatment. But in other cases, cardiomyopathy develops quickly with severe symptoms, and serious complications occur. Treatment is required in these instances.

Treatments include lifestyle changes, <u>medications</u>, <u>surgery</u>, <u>implanted devices to</u> <u>correct arrhythmias</u> and other nonsurgical procedures. These treatments can control symptoms, reduce complications and prevent the disease from worsening.

# **Dilated Cardiomyopathy (DCM)**

Dilated cardiomyopathy (DCM) is the most common type, occurring mostly in adults 20 to 60. It affects the heart's ventricles and atria, the lower and upper chambers of the heart, respectively.

Frequently the disease starts in the left ventricle, the heart's main pumping chamber. The heart muscle begins to dilate, meaning it stretches and becomes thinner. Consequently, the inside of the chamber enlarges. The problem often spreads to the right ventricle and then to the atria.

As the heart chambers dilate, the heart muscle doesn't contract normally and cannot pump blood very well. As the heart becomes weaker <u>heart failure</u> can occur. Common <u>symptoms of heart failure</u> include shortness of breath, fatigue and swelling of the ankles, feet, legs, abdomen and veins in the neck.

heartbeats) and blood clots in the heart.

## Other Names for Dilated Cardiomyopathy

- Alcoholic cardiomyopathy. (A term used when overuse of alcohol causes the disease)
- Congestive cardiomyopathy
- Diabetic cardiomyopathy
- Familial dilated cardiomyopathy
- Idiopathic cardiomyopathy
- Ischemic cardiomyopathy (A term used when <u>coronary heart disease</u>, also called coronary artery disease or <u>heart attack</u> cause the disease. Not all forms of DCM are ischemic in origin.)
- <u>Peripartum cardiomyopathy</u> (A term used when the disease develops in a woman shortly before or after she gives birth.)
- Primary cardiomyopathy

## What Causes Dilated Cardiomyopathy

Often, cause of dilated cardiomyopathy isn't known. Up to one-third of the people of those who have it inherit it from their parents.

Some diseases, conditions and substances also can cause the disease, such as:

- <u>Coronary heart disease</u>, <u>heart attack</u>, <u>high blood pressure</u>, <u>diabetes</u>, thyroid disease, viral hepatitis and HIV
- Infections, especially viral infections that inflame the heart muscle
- <u>Alcohol</u>, especially if you also have a poor diet
- Complications during the last month of pregnancy or within 5 months of birth
- Certain toxins such as cobalt
- Certain drugs (such as <u>cocaine</u> and amphetamines) and two medicines used to treat cancer (doxorubicin and daunorubicin)

## Other Types of Cardiomyopathy:

- <u>Hypertrophic Cardiomyopathy</u>
- <u>Restrictive Cardiomyopathy</u>
- <u>Arrhythmogenic Right Ventricular Dysplasia</u>

Learn more:

- Family History and Heart Disease
- Alcohol and Heart Disease
- <u>Cocaine, Marijuana and Other Drugs and Heart Disease</u>

# Hypertrophic Cardiomyopathy (HCM)



Hypertrophic cardiomyopathy is most often caused by abnormal genes in the heart muscle. These genes cause the walls of the heart chamber (left ventricle) to contract harder and become thicker than normal.

The thickened walls become stiff. This reduces the amount of blood taken in and pumped out to the body with each heartbeat.

## Obstructive and Nonobstructive HCM

In obstructive HCM, the wall (septum) between the two bottom chambers of the heart thickens. The walls of the pumping chamber can also become stiff. It may block or reduce the blood flow from the left ventricle to the aorta. Most people with HCM have this type.

In nonobstructive HCM, the heart's main pumping chamber still becomes stiff. This limits how much blood the ventricle can take in and pump out, but blood flow is not blocked.

# Signs, Symptoms and Risks

Some people with hypertrophic cardiomyopathy don't have symptoms. Others may not have signs or symptoms in the early stages of the disease but may develop them over time. Knowing the signs and symptoms of HCM is important. It can help with getting an early diagnosis, when treatment may be most effective. Signs and symptoms of HCM include:

- Chest pain, especially with physical exertion
- Shortness of breath, especially with physical exertion
- Fatigue
- <u>Arrhythmias</u> (abnormal heart rhythms)
- Dizziness
- Lightheadedness
- Fainting (syncope)
- Swelling in the ankles, feet, legs, abdomen and veins in the neck

HCM is a chronic disease that can get worse over time. This can lead to poorer function and quality of life, long-term complications and more financial and social burden.

People with HCM often need to make lifestyle changes, such as limiting their activity, to adjust for their disease.

As HCM progresses, it can cause other health problems. People with HCM are at higher risk for developing <u>atrial fibrillation</u>, which can lead to blood clots, <u>stroke</u> and other

heart-related complications. HCM may also lead to <u>heart failure</u>. It can also lead to <u>sudden cardiac arrest</u>, but this is rare. HCM has been cited as the most common reason for sudden cardiac death in young people and athletes under the age of 35.

## Diagnosis

Hypertrophic cardiomyopathy is most often inherited. HCM is the most common form of genetic heart disease. It can happen at any age, but most receive a diagnosis in middle age.

It's estimated that 1 in every 500 people have HCM, but a large percentage of patients are undiagnosed. Of those diagnosed, two-thirds have obstructive HCM and one-third have non-obstructive HCM.

A cardiologist or pediatric cardiologist often diagnoses and treats HCM. You may also be referred to a cardiomyopathy center where the health care team has specialized training.

HCM is diagnosed based on your medical history, family history, a physical exam and diagnostic test results.

### **Medical and Family Histories**

Knowing your medical history and any signs and symptoms you may have is an important first step. Your physician will also want to know if anyone in your family has been diagnosed with HCM, <u>heart failure</u> or <u>cardiac arrest</u>.

### Physical Exam

Your heart and lungs will be checked. Your physician will listen for certain sounds with a stethoscope. For example, the loudness, timing and location of a heart murmur may suggest obstructive HCM.

### **Diagnostic Tests**

Diagnosis is typically done by <u>echocardiogram</u>. It checks the thickness of the heart muscle and blood flow from the heart. In some cases, another type of echocardiogram, <u>transesophageal echo</u> (or TEE), may be performed. A TEE is done using a probe inserted in the throat while the patient is under sedation. Other diagnostic tests include:

- Electrocardiogram (ECG)
- <u>Cardiac MRI</u>
- <u>Stress tests</u>
- Holter and event monitors
- Genetic testing

#### **Diagnostic Procedures**

Confirming diagnosis or preparing for surgery may also involve one or more medical procedures including:

- <u>Cardiac catheterization</u>
- <u>Coronary angiography</u>

## Treatment and Management of HCM

There are currently no disease-specific medications for hypertrophic car diomyopathy.

For people with HCM who don't have symptoms, <u>lifestyle changes</u> and medications for conditions that may contribute to cardiovascular disease are recommended. For those with symptoms, the focus is on symptom management using medications and procedures.

#### Medications

Medications called beta-blockers, calcium channel blockers and diuretics offer limited and varying relief of symptoms. They may help with function but may also have adverse side effects.

#### Procedures

A range of surgical and nonsurgical procedures can be used to treat HCM:

- Septal myectomy Septal myectomy is open-heart surgery. It's considered for people with obstructive HCM and severe symptoms. This surgery is generally reserved for younger patients and for people whose medications aren't working well. A surgeon removes part of the thickened septum that's bulging into the left ventricle. This improves blood flow within the heart and out to the body.
- Alcohol septal ablation (nonsurgical procedure) In this procedure, ethanol (a type of alcohol) is injected through a tube into the small artery that supplies blood to the area of heart muscle thickened by HCM. The alcohol causes these cells to die. The thickened tissue shrinks to a more normal size. The risks and complications of heart surgery increase with age. For this reason, ablation may be preferred to myectomy in older patients with other medical conditions.
- **Surgically implanted devices** Surgeons can implant several types of devices to help the heart work better, including:
  - Implantable cardioverter defibrillator (ICD) An ICD helps maintain a normal heartbeat by sending an electric shock to the heart if an irregular heartbeat is detected. This reduces the risk of sudden cardiac death.
  - **Pacemaker** This small device uses electrical pulses to prompt the heart to beat at a normal rate.
  - **Cardiac resynchronization therapy (CRT) device** This device coordinates contractions between the heart's left and right ventricles.
- Heart transplant In HCM patients with advanced, end-stage disease, a heart transplant may be considered. In this procedure, a person's diseased heart is replaced with a healthy donor heart.

## Other types of cardiomyopathy:

- Dilated cardiomyopathy
- <u>Restrictive cardiomyopathy</u>
- <u>Arrhythmogenic right ventricular dysplasia</u>
- <u>Transthyretin amyloid cardiomyopathy (ATTR-CM)</u>

# Peripartum Cardiomyopathy (PPCM)



## What is peripartum cardiomyopathy?

Peripartum cardiomyopathy (PPCM), also known as postpartum cardiomyopathy, is an uncommon form of heart failure that happens during the last month of pregnancy or up to five months after giving birth. Cardiomyopathy literally means heart muscle disease.

PPCM is a <u>dilated</u> form of the condition, which means the heart chambers enlarge and the muscle weakens. This causes a decrease in the percentage of blood ejected from the left ventricle of the heart with each contraction. That leads to less blood flow and the heart is no longer able to meet the demands of the body's organs for oxygen, affecting the lungs, liver, and other body systems.

PPCM is rare in the United States, Canada, and Europe. About 1,000 to 1,300 women develop the condition in the U.S. each year. In some countries, PPCM is much more common and may be related to differences in diet, lifestyle, other medical conditions or genetics.

## How is it diagnosed?

PPCM may be difficult to detect because <u>symptoms of heart failure</u> can mimic those of third trimester pregnancy, such as swelling in the feet and legs, and some shortness of breath. More extreme cases feature severe shortness of breath and prolonged swelling after delivery.

During a physical exam, doctors will look for signs of fluid in the lungs. A stethoscope will be used to listen for lung crackles, a rapid heart rate, or abnormal heart sounds. An echocardiogram can detect the cardiomyopathy by showing the diminished functioning of the heart.

PPCM is diagnosed when the following three criteria are met:

- 1. <u>Heart failure</u> develops in the last month of pregnancy or within 5 months of delivery.
- 2. Heart pumping function is reduced, with an <u>ejection fraction</u> (EF) less than 45% (typically measured by an echocardiogram). EF is how much blood the left ventricle pumps out with each contraction. A normal EF can be between 55 and 70.
- 3. No other cause for heart failure with reduced EF can be found.

Laboratory blood tests are a standard part of the evaluation. This includes tests to assess kidney, liver and thyroid function; tests to assess electrolytes, including sodium and potassium; and a complete blood count to look for anemia or evidence of infection. In

addition, markers of cardiac injury and stress can be used to assess level of risk.

## Symptoms of the condition include:

- Fatigue
- Feeling of heart racing or skipping beats (palpitations)
- Increased nighttime urination (nocturia)
- Shortness of breath with activity and when lying flat
- Swelling of the ankles
- Swollen neck veins
- Low blood pressure, or it may drop when standing up.

The severity of symptoms in patients with PPCM can be classified by the New York Heart Association system:

- Class I Disease with no symptoms
- Class II Mild symptoms/effect on function or symptoms only with extreme exertion
- Class III Symptoms with minimal exertion
- Class IV Symptoms at rest

### What are the causes?

The underlying cause is unclear. Heart biopsies in some cases show women have inflammation in the heart muscle. This may be because of prior viral illness or abnormal immune response. Other potential causes include poor nutrition, coronary artery spasm, small-vessel disease, and defective antioxidant defenses. Genetics may also play a role.

Initially thought to be more common in women older than 30, PPCM has since been reported across a wide range of age groups. Risk factors include:

- Obesity
- History of cardiac disorders, such as myocarditis (inflammation of the heart muscle)
- Use of certain medications
- Smoking
- Alcoholism
- Multiple pregnancies
- African-American descent
- Poor nourishment

### How can PPCM be treated?

The objective of peripartum cardiomyopathy treatment is to keep extra fluid from collecting in the lungs and to help the heart recover as fully as possible. Many women recover normal heart function or stabilize on medicines. Some progress to severe heart failure requiring mechanical support or heart transplantation.

There are several classes of medications a physician can prescribe to treat symptoms, with variations that are safer for women who are breastfeeding.

- Angiotensin converting enzyme, or ACE, inhibitors Help the heart work more efficiently
- Beta blockers Cause the heart to beat more slowly so it has recovery time
- Diuretics Reduce fluid retention
- **Digitalis** Derived from the foxglove plant, it has been used for more than 200 years to treat heart failure. Digitalis strengthens the pumping ability of the heart
- Anticoagulants To help thin the blood. Patients with PPCM are at increased risk of

developing blood clots, especially if the EF is very low.

Doctors may recommend a low-salt diet, fluid restrictions, or daily weighing. A weight gain of 3 to 4 pounds or more over a day or two may signal a fluid buildup.

Women who smoke and drink alcohol will be advised to stop, since these habits may make the symptoms worse.

A heart biopsy may help determine if the underlying cause of cardiomyopathy is a heart muscle infection (myocarditis). However, this procedure is uncommon.

### How can women minimize their risk?

To develop and maintain a strong heart, women should avoid cigarettes and alcohol, eat a well-balanced diet and get regular exercise. Women who develop peripartum cardiomyopathy are at high risk of developing the same condition with future pregnancies.

### What's next?

Investigations are underway to understand the cause of PPCM and to develop new treatments. Treatments that alter the immune system such as intravenous  $\gamma$ -globulin and immunoabsorption have been tried but are not proven. Investigators also have focused on the role of prolactin in PPCM. Prolactin is a hormone released from the pituitary gland late in pregnancy and after delivery that stimulates breast milk production. Prolactin, however, may have adverse effects on the heart muscle by limiting its blood supply and causing cell death. Bromocryptine is a medication that inhibits the pituitary secretion of prolactin. Early studies suggest it helps treat PPCM, but more research is needed.

## **Restrictive Cardiomyopathy**

Restrictive cardiomyopathy tends to affect older adults. The heart's ventricles become rigid because abnormal tissue, such as scar tissue, replaces the normal heart muscle. Consequently, the ventricles can't relax normally and fill with blood, and the atria become enlarged. Blood flow in the heart is reduced over time. This can lead to problems such as <u>heart failure</u> or <u>arrhythmias</u>.

## Other Names for Restrictive Cardiomyopathy

- Idiopathic restrictive cardiomyopathy
- Infiltrative cardiomyopathy

## What Causes Restrictive Cardiomyopathy

Certain diseases, conditions and factors can cause restrictive cardiomyopathy, including:

- Hemochromatosis. (A disease in which too much iron builds up in your body. The extra iron is toxic to the body and can damage the organs, including the heart.)
- Sarcoidosis. (A disease that causes <u>inflammation</u> and can affect the body's organs. Researchers believe that an abnormal immune response may cause sarcoidosis. The abnormal response causes tiny lumps of cells to form in the body's organs, including the heart.)
- Amyloidosis. (A disease in which abnormal proteins build up in the body's organs, 163 | P a g e

including the heart.)

- Connective tissue disorders
- Some cancer treatments, such as radiation and chemotherapy

## Other Types of Cardiomyopathy:

- Dilated Cardiomyopathy
- Hypertrophic Cardiomyopathy
- <u>Arrhythmogenic Right Ventricular Dysplasia</u>

# Arrhythmogenic Right Ventricular Dysplasia

Arrhythmogenic right ventricular dysplasia (ARVD) is a rare type of cardiomyopathy. It occurs if the muscle tissue in the right ventricle dies and is replaced with scar tissue. This disrupts the heart's electrical signals and causes <u>arrhythmias</u>. Symptoms include palpitations and <u>fainting</u> after physical activity. Palpitations are feelings that your heart is skipping a beat, fluttering or beating too hard or too fast.

ARVD usually affects teens or young adults. It can cause sudden <u>cardiac arrest</u> (SCA) in young athletes.

## Other Names for Arrhythmogenic Right Ventricular Dysplasia

- Arrhythmogenic right ventricular cardiomyopathy
- Right ventricular cardiomyopathy
- Right ventricular dysplasia

### What Causes Arrhythmogenic Right Ventricular Dysplasia

• Researchers believe that arrhythmogenic right ventricular dysplasia is an inherited disease.

# Transthyretin Amyloid Cardiomyopathy (ATTR-CM) For Professionals

## Eliminate the What-If. Learn When to Rule Out ATTR-CM.

When patients present with preserved ejection fraction and no clear cause, it's time to consider Transthyretin Amyloid Cardiomyopathy (ATTR-CM). The symptoms can be similar, but ATTR-CM has a different prognosis and requires unique management strategies.

Learn when to test and more about ATTR-CM on our podcast series.

# Is Broken Heart Syndrome Real?



# When Your Heart Breaks ... (Literally).

"You can die of a broken heart — it's scientific fact — and my heart has been breaking since that very first day we met. I can feel it now, aching deep behind my rib cage the way it does every time we're together, beating a desperate rhythm: Love me. Love me. Love me." —Abby McDonald, Getting Over Garrett Delaney

When you think of a broken heart, you may picture a cartoon drawing with a jagged line through it. But a real-life broken heart can actually lead to cardiac consequences. There are established ties between depression, mental health and heart disease. Read on for more information about how an extremely stressful event can have an impact on your heart.

### Breakdown of a Broken Heart

Broken heart syndrome, also called stress-induced cardiomyopathy or takotsubo cardiomyopathy, can strike even if you're healthy. (Tako tsubo, by the way, are octopus traps that resemble the pot-like shape of the stricken heart.)

Women are more likely than men to experience the sudden, intense chest pain— the reaction to a surge of stress hormones — that can be caused by an emotionally stressful event. It could be the death of a loved one or even a divorce, breakup or physical separation, betrayal or romantic rejection. It could even happen after a good shock (like winning the lottery.)

Broken heart syndrome may be misdiagnosed as a heart attack because the symptoms and test results are similar. In fact, tests show dramatic changes in rhythm and blood substances that are typical of a heart attack. But unlike a heart attack, there's no evidence of blocked heart arteries in broken heart syndrome.

In broken heart syndrome, a part of your heart temporarily enlarges and doesn't pump well, while the rest of your heart functions normally or with even more forceful contractions. Researchers are just starting to learn the causes, and how to diagnose and treat it.

failure.

**The good news:** Broken heart syndrome is usually treatable. Most people who experience it make a full recovery within weeks, and they're at low risk for it happening again (although in rare cases in can be fatal).

### What To Look For: Signs and Symptoms

The most common signs and symptoms of broken heart syndrome are angina (chest pain) and shortness of breath. You can experience these things even if you have no history of heart disease.

Arrhythmias (irregular heartbeats) or cardiogenic shock also may occur with broken heart syndrome. Cardiogenic shock is a condition in which a suddenly weakened heart can't pump enough blood to meet the body's needs, and it can be fatal if it isn't treated right away. (When people die from heart attacks, cardiogenic shock is the most common cause of death.)

### Heart attack and broken heart syndrome: What's the difference?

Some signs and symptoms of broken heart syndrome differ from those of heart attack. In broken heart syndrome, symptoms occur suddenly after extreme emotional or physical stress. Here are some other differences:

- EKG (a test that records the heart's electric activity) results don't look the same as the EKG results for a person having a heart attack.
- Blood tests show no signs of heart damage.
- Tests show no signs of blockages in the coronary arteries.
- Tests show ballooning and unusual movement of the lower left heart chamber (left ventricle).
- Recovery time is quick, usually within days or weeks (compared with the recovery time of a month or more for a heart attack).

### Learn More About Broken Heart Syndrome

If your doctor thinks you have broken heart syndrome, you may need coronary angiography, a test that uses dye and special X-rays to show the insides of your coronary arteries. Other diagnostic tests are blood tests, EKG, echocardiography (a painless test that uses sound waves to create moving pictures of your heart) and cardiac MRI.

To keep tabs on your heart health, your doctor may recommend an **echo** about a month after you're diagnosed with the syndrome. Talk with your doctor about how often you should schedule follow-up visits.

### Learn more:

- Taking Care of Yourself After a Cardiac Event
- Three Tips to Manage Stress
- Heart Attack Symptoms in Women

# **Pediatric Cardiomyopathies**



Patient education material brought to you by the American Heart Association and the <u>Children's C ardi omyop at hy Found at ion (link opens in new</u> window).

Cardiomyopathy is rare in children. For that reason alone, a diagnosis of cardiomyopathy can rattle parents, and possibly the child too. Fortunately, our understanding of how the heart works under normal and abnormal conditions is increasing each year.

As you familiarize yourself with pediatric cardiomyopathy, you'll find yourself in a better position to evaluate your child's treatment options. Knowledge is power. Learn all that you can as you work with your child's doctor to identify the best course of action.

## Cardiomyopathy and its incidence among children

Cardiomyopathy refers to a diseased state of the heart involving abnormalities of the muscle fibers, which contract with each heartbeat. It can be considered "primary" or "secondary":

- In primary cases, cardiomyopathy occurs because the muscle cells themselves are abnormal (usually due to a gene mutation).
- Secondary cases of cardiomyopathy involve healthy heart muscle cells that are adversely affected by other conditions. Precipitating conditions include low blood flow to the heart, low blood oxygen, high blood pressure and certain infections.

According to the Pediatric Cardiomyopathy Registry, one in every 100,000 children in the U.S. under the age of 18 is diagnosed with cardiomyopathy. The majority of diagnosed children are under 12 months, followed by children 12 to 18 years old.

# Types of cardiomyopathy

Cardiomyopathies can be grouped into four broad categories. The clinical features and treatment options differ for each.

- Dilated cardiomyopathy (PDF)
- Hypertrophic cardiomyopathy (PDF)
- <u>Restrictive cardiomyopathy (PDF)</u>
- Miscellaneous (rare) cardiomyopathies (PDF)

### Learn more:

• <u>Overview of inheritance for cardiomyopathies (PDF)</u>

# **Understand Your Risk for Cardiomyopathy**

Cardiomyopathy can affect all ages and races, but certain types of the disease are more common in certain groups.

<u>Dilated cardiomyopathy</u> is more common in African-Americans than in Caucasians. It is also more prevalent in men than in women. Teens and young adults are more likely than older people to have <u>arrhythmogenic right ventricular dysplasia</u>, although it's rare in both groups.

## Major risk factors

Certain diseases, conditions or factors can raise your risk for cardiomyopathy. Among the major risk factors are:

- <u>Family history</u> of cardiomyopathy, <u>heart failure</u> or <u>sudden cardiac arrest</u> (SCA)
- A disease or condition that can lead to cardiomyopathy, such as <u>coronary heart disease</u>, <u>heart attack</u> or a viral infection that inflames the heart muscle
- <u>Diabetes</u> or other <u>metabolic diseases</u>, or severe obesity
- Diseases that can damage the heart, such as hemochromatosis, sarcoidosis or amyloidosis
- Long-term <u>alcoholism</u>
- Long-term high blood pressure

# **Symptoms and Diagnosis of Cardiomyopathy**



It's important to identify those who may be at high risk for cardiomyopathy. After all, some people with cardiomyopathy never have signs or symptoms. Others don't have signs or symptoms in the early stages of the disease.

If people without symptoms recognize their heightened risk for cardiomyopathy, there's a better chance of diagnosing it early, when treatment may be most effective.

Signs and symptoms of cardiomyopathy include:

- Shortness of breath or trouble breathing, especially with physical exertion
- Fatigue
- Swelling in the ankles, feet, legs, abdomen and veins in the neck
- Dizziness
- Lightheadedness
- Fainting during physical activity
- <u>Arrhythmias</u> (irregular heartbeats)

- Chest pain, especially after physical exertion or heavy meals
- <u>Heart murmurs</u> (unusual sounds associated with heartbeats)

Signs and symptoms of heart failure usually occur in the later stages of cardiomyopathy, as the heart weakens.

## How is cardiomyopathy diagnosed?

Your doctor will diagnose cardiomyopathy based on your medical history, family history, a physical exam and diagnostic test results.

Often, a cardiologist or pediatric cardiologist diagnoses and treats cardiomyopathy. These doctors specialize in heart diseases.

## Medical and family histories

Your doctor will want to learn about your medical history as well as any signs and symptoms you may have. Your physician will also want to know whether anyone in your family has been diagnosed with cardiomyopathy, <u>heart failure</u> o <u>cardiac arrest</u>.

## Physical exam

Using a stethoscope, your doctor will listen to your heart and lungs for sounds that may suggest cardiomyopathy. Particular sounds may even suggest a certain type of the disease.

For example, the loudness, timing and location of a heart murmur may suggest <u>obstructive hypertrophic cardiomyopathy</u>. A "crackling" sound in the lungs may be a <u>sign</u> <u>of heart failure</u>.

Certain physical signs also help your doctor diagnose cardiomyopathy. Swelling of the ankles, feet, legs, abdomen or veins in your neck suggests fluid buildup, a sign of heart failure.

### Diagnostic tests

Your doctor may recommend one or more of the following tests to diagnose cardiomyopathy:

- **Blood tests**: A small amount of blood is taken usually drawn from a vein in your arm using a needle.
- <u>Chest X-ray</u>: A chest X-ray takes pictures of the organs and structures inside your chest, and can show whether your heart is enlarged. It can also reveal whether fluid is building up in your lungs.
- Electrocardiogram (EKG or ECG): An EKG records the heart's electrical activity, showing how fast the heart is beating and whether its rhythm is steady or irregular. An EKG can be used to detect cardiomyopathy as well as other problems, including <u>heart attacks</u>, <u>arrhythmias</u> (irregular heartbeats) and <u>heart failure</u>. To diagnose heart problems that come and go, your doctor may have you wear a portable EKG monitor.
- Holter and event monitors: Both of these are portable devices that record your heart's electrical activity during your normal daily activities. A <u>Holter monitor</u> records the heart's electrical activity for a full 24- or 48-hour period. An event monitor records your heart's electrical activity only at certain times.
- <u>Echocardiogram</u> (Echo): An echocardiogram (echo) is a test that uses sound waves to

create a moving picture of your heart. It shows how well your heart is working as well as its size and shape. There are several types of echocardiography, including "stress echo," which is administered as part of a stress test. Another type, <u>transesophageal echo</u> (or TEE), provides a view of the back of the heart.

 <u>Stress test</u>: In a stress test, the aim is to make your heart work hard (and beat fast) while tests are performed. These tests may include nuclear heart scanning, <u>echo</u>, and <u>positron</u> <u>emission tomography</u> (PET) scanning. You'll be asked to walk in place on an inclined treadmill. If you are unable to exercise, you may be given medicine to simulate the effects of exertion.

### Diagnostic procedures

Confirming a diagnosis may involve one or more medical procedures. Or, if surgery is planned, a diagnostic procedure may be performed in preparation for surgery. Such diagnostic procedures may include:

- <u>Cardiac catheterization</u>: Cardiac catheterization checks the pressure and blood flow in your heart's chambers. In this procedure, a long, thin, flexible tube called a catheter is inserted through a blood vessel and threaded to the heart. This allows the doctor to collect blood samples and check your heart's arteries for blockages using X-ray imaging.
- <u>Coronary angiography (PDF)</u>: This procedure often is done during cardiac catheterization. Dye that can be seen on an X-ray is injected into your coronary arteries. The dye allows your doctor to study blood flow within your heart and blood vessels.
- **Myocardial biopsy**: In this procedure, your doctor removes a piece of your heart muscle, which can be done during cardiac catheterization. The muscle is studied under a microscope to see whether changes in cells have occurred, which may suggest cardiomyopathy.
- **Genetic testing**: Your doctor may suggest genetic testing to look for signs of cardiomyopathy in your parents, brothers and sisters or other family members. Genetic testing can show how the disease runs in families and can reveal the chances of parents passing the genes for the disease on to their children. Genetic testing may also be useful if your doctor suspects that you may have cardiomyopathy, but you don't yet exhibit signs or symptoms.

#### Learn more:

- Family history and heart disease, stroke
- <u>Understand your risk for heart failure</u>

# **Prevention and Treatment of Cardiomyopathy**

People who have cardiomyopathy but no signs or symptoms may not need treatment. Sometimes, <u>dilated cardiomyopathy</u> that comes on suddenly may even go away on its own.

In other instances, treatment is needed. Treatment hinges on a few factors: the type of cardiomyopathy, the severity of your symptoms and complications as well as your age and overall health.

## Treatment goals

When treating cardiomyopathy, objectives include:

- Stopping the disease from getting worse
- Managing any conditions that cause or contribute to the disease
- Reducing complications and the risk of sudden cardiac arrest (SCA)

• Controlling symptoms so that you can live as normally as possible

## Treatments for cardiomyopathies

Treatment for cardiomyopathy may include one or more of the following:

#### Lifestyle changes

Lifestyle changes may help to manage a condition that's causing your cardiomyopathy.

#### Healthy diet and physical activity

- A <u>healthy diet</u> and <u>physical activity</u> are part of a healthy lifestyle. A healthy diet includes a variety of <u>fruits</u>, <u>vegetables</u> and grains. Half of your grains should come from whole-grain products.
- Choose foods that are low in <u>saturated and trans fats</u>. Healthy choices include <u>lean</u> <u>meats</u>, poultry without skin, <u>non-fried fish</u>, beans as well as fat-free or low-fat milk and milk products.
- Choose and <u>prepare foods with little salt</u> (sodium). Too much salt can raise your <u>risk of high blood pressure</u>. Studies show that following a <u>Dietary Approaches to Stop Hypertension</u> (DASH) eating plan can lower blood pressure.
- Choose foods and beverages that are low in <u>added sugar</u>. Avoid drinking alcohol. <u>If you</u> <u>drink alcoholic beverages</u>, do so in moderation.
- Aim for a <u>healthy weight</u> by staying within your daily calorie needs. Balance the calories you take in with the calories you use during physical activity.
- Be as physically active as you can. But talk to your doctor before increasing your physical activity if:
  - you are taking medications
  - you have an ongoing health problem
  - $\circ$   $\;$  you have symptoms such as chest pain, shortness of breath or dizziness

#### Other lifestyle changes

Your doctor also may recommend other lifestyle changes, such as:

- Quitting smoking
- Losing excess weight
- Avoiding alcohol and illegal drugs
- Getting enough sleep and rest
- <u>Reducing stress</u>
- Treating underlying conditions, such as <u>diabetes</u> and <u>high blood pressure</u>

#### Medications

Many medications are used to treat cardiomyopathy. Your doctor may prescribe medicines to:

- Lower your blood pressure. ACE inhibitors, angiotensin II receptor blockers, beta blockers and calcium channel blockers are examples of <u>medicines that lower blood</u> <u>pressure</u>.
- Slow your <u>heart rate</u>. Beta blockers, calcium channel blockers, and digoxin are examples
  of medicines that slow the heart rate. Beta blockers and calcium channel blockers also are
  used to lower blood pressure.
- Keep your heart beating with a normal rhythm. These medicines, called <u>antiarrhythmics</u>, help prevent <u>arrhythmias</u> (irregular heartbeats).
- Balance electrolytes in your body. Electrolytes are minerals that help maintain fluid levels and the acid-base balance in your body. Electrolytes also help muscle and nerve tissues work properly. Medicines used to balance electrolytes include aldosterone blockers.

- **Remove excess fluid and sodium from your body.** Diuretics, or "water pills," are an example of a medicine that helps remove excess fluid and sodium from the body.
- **Prevent blood clots from forming.** <u>Anticoagulants (PDF)</u>, or \*blood thinners, help to prevents blood clots. Blood thinners often are used to prevent blood clots from forming in people who have <u>dilated cardiomyopathy</u>.
- **Reduce inflammation.** Medications used to reduce inflammation include corticosteroids.

#### Procedures for cardiomyopathy

A range of surgical and nonsurgical procedures can be used to treat cardiomyopathy:

- Septal myectomy Septal myectomy is open-heart surgery. It's considered for people who have <u>obstructive hypertrophic cardiomyopathy</u> and severe symptoms. This surgery generally is reserved for younger patients and for people whose medications aren't working well. A surgeon removes part of the thickened septum that's bulging into the left ventricle. This improves blood flow within the heart and out to the body.
- **Surgically implanted devices** Surgeons can implant several types of devices in the body to help the heart work better, including:
  - **Pacemaker** This small device uses electrical pulses to prompt the heart to beat at a normal rate.
  - <u>Cardiac resynchronization therapy (CRT) device</u> This device coordinates contractions between the heart's left and right ventricles.
  - <u>Left ventricular assist device</u> (LVAD) This implantable device helps the heart pump blood to the body. An LVAD can be used for long-term therapy or as an interim treatment for those awaiting a heart transplant.
  - **Implantable cardioverter defibrillator (ICD)** An ICD helps to maintain a normal heartbeat by sending an electric shock to the heart if an <u>arrhythmia</u>, or irregular heartbeat, is detected.
- **Heart Transplant** In a heart transplant surgery, a person's diseased heart is replaced with a healthy donor heart. A heart transplant is a last resort for people who have end-stage heart failure. ("End-stage" means that all other treatment options have been explored, without success.)
- Alcohol septal ablation (nonsurgical procedure) In this procedure, ethanol (a type of alcohol) is injected through a tube into the small artery that supplies blood to the area of heart muscle thickened by HCM. The alcohol causes these cells to die. The thickened tissue shrinks to a more normal size. The risks and complications of heart surgery increase with age. For this reason, ablation may be preferred to myectomy in older patients with other medical conditions.

### How can cardiomyopathy be prevented?

You cannot prevent inherited types of cardiomyopathy. But you can take steps to lower your risk for conditions that may lead to (or complicate) cardiomyopathy, such <u>coronary</u> <u>heart disease, high blood pressure</u> and <u>heart attack</u>.

Cardiomyopathy can be precipitated by an underlying disease or condition. Treating that initial problem early enough may help prevent the complications presented by cardiomyopathy. For example, to control the underlying conditions of high blood pressure, high blood cholesterol and diabetes:

- Get regular checkups with your doctor.
- Follow your doctor's advice about lifestyle changes.
- Take all of your medications exactly as prescribed by your doctor.

Just as some underlying conditions can bring about cardiomyopathy, cardiomyopathy in turn can cause other complications.

For instance, cardiomyopathy can increase the risk for <u>sudden cardiac arrest</u> (SCA). An implantable cardioverter defibrillator (ICD) can be used to mitigate this risk.