CARDIOMIOPATHIAS

There are many causes of cardiomyopathy that can be categorized in several ways. One method of defining cardiomyopathy is based on the official definition by the American Heart Association, which are broken into two categories, primary and secondary. Another method of categorizing cardiomyopathy causes are extrinsic and intrinsic. The official definition of cardiomyopathy of the American Heart Association in 2006 is as follows:

"Cardiomyopathies are a heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilatation and are due to a variety of causes that frequently are genetic. Cardiomyopathies either are confined to the heart or are part of generalized systemic disorders, which may lead to cardiovascular death or progressive heart failure-related disability."

The definition divides heart disease into 1) primary cardiomyopathies, those that usually affect the heart alone (primary) and 2) secondary cardiomyopathies, those that are a result of an underlying condition affecting many areas of the body. The primary cardiomyopathies are further divided into inherited (genetic) diseases, those that are acquired, and those that are a combination of both.

Some causes are listed below of primary and secondary cardiomyopathies are listed below.

**Primary cardiomyopathy**

- **Genetic**: hypertrophic cardiomyopathy with conduction abnormalities
  - prolonged QT syndrome
  - Brugada syndrome
- **Mixed**: dilated cardiomyopathy
  - restrictive cardiomyopathy

**Secondary cardiomyopathies**

- **Infiltrative**: amyloidosis, Gaucher disease
- **Toxicity**: drugs/ Alcohol, heavy metals, chemicals
- **Inflammatory**: sarcoidosis
- **Endocrine**: diabetes mellitus, thyroid problems

The clinical diagnosis of hypertrophy cardiomyopathy (HCM) is established most easily and reliably with two-dimensional echocardiography by demonstrating left ventricular hypertrophy (LVH) (typically asymmetric in distribution, and showing virtually any diffuse or segmental pattern of left ventricular (LV) wall thickening).

Left ventricular wall thickening is associated with a nondilated and hyperdynamic chamber (often with systolic cavity obliteration) in the absence of another cardiac or systemic disease (e.g., hypertension or aortic stenosis) capable of producing the magnitude of hypertrophy evident, and independent of whether or not LV outflow obstruction is present.
Although the **usual clinical diagnostic criteria** for HCM is a maximal wall thickness greater than or equal to 15 mm, genotype-phenotype correlations have shown that virtually any wall thickness (including those within normal range) are compatible with the presence of a HCM mutant gene. Mildly increased LV wall thicknesses of 13 mm to 14 mm potentially due to HCM should be distinguished from certain extreme expressions of the physiologically-based athlete's heart.

The advent of contemporary **magnetic resonance** imaging that provides high-resolution tomographic images of the entire LV may represent an additional diagnostic modality particularly in the presence of technically suboptimal echocardiographic studies or when segmental hypertrophy is confined to unusual locations within the LV wall.

Since the modern description by Teare in 1958, HCM has been known by a confusing array of names that largely reflect its clinical heterogeneity, relatively uncommon occurrence in cardiologic practice, and the skewed experience of early investigators. This problem in nomenclature has been an obstacle to general recognition of the disease within the medical and non-medical community.

**Hypertrophic cardiomyopathy** (or HCM) is now widely accepted as the preferred term because it describes the overall disease spectrum without introducing misleading inferences that LV outflow tract obstruction is an invariable feature of the disease, such as is the case with hypertrophic obstructive cardiomyopathy, muscular subaortic stenosis, or idiopathic hypertrophic subaortic stenosis.

Indeed, most patients with HCM do not demonstrate outflow obstruction under resting (basal) conditions, although many may develop dynamic subaortic gradients of varying magnitude with provocative maneuvers or agents.

Of note, even though the absence of obstruction (at rest) is common, both in patients with and without symptoms, most treatment modalities have targeted those symptomatic HCM patients with outflow obstruction.

**Obstruction to Left Ventricle Outflow**

It is of clinical importance to distinguish between the obstructive or nonobstructive forms of HCM, based on the presence or absence of a LV outflow gradient under resting and/or provokable conditions. Indeed, in most patients, management strategies have traditionally been tailored to the hemodynamic state. Outflow gradients are responsible for a loud apical systolic ejection murmur associated with a constellation of unique clinical signs, hypertrophy of the basal portion of ventricular septum and small outflow tract, and an enlarged and elongated mitral valve in many patients.

Obstruction may either be subaortic or mid-cavity in location. Subaortic obstruction is caused by systolic anterior motion (SAM) of the mitral valve leaflets and mid-systolic contact with the ventricular septum. This mechanical impedance to outflow occurs in the presence of high velocity ejection in which a variable proportion of the forward blood flow may be ejected early in systole.

Systolic anterior motion is probably attributable to a drag effect or possibly a Venturi phenomenon and is responsible not only for subaortic obstruction, but also the concomitant mitral regurgitation (usually mild-to-moderate in degree) due to incomplete leaflet apposition, which is typically directed posteriorly into the left atrium.

When the mitral regurgitation jet is directed centrally or anteriorly into the left atrium, or if multiple jets are present, independent abnormalities intrinsic to the mitral valve should be suspected (e.g., myxomatous degeneration, mitral leaflet fibrosis, or anomalous papillary muscle insertion). Occasionally (perhaps in 5% of cases), gradients and impeded outflow are caused predominately by muscular apposition in the mid-cavity region—usually in the absence of mitral-septal contact—involving anomalous direct insertion of anterolateral papillary muscle into
the anterior mitral leaflet, or excessive mid-ventricular or papillary muscle hypertrophy and malalignment.

Although it has previously been subject to periodic controversy, there is now widespread recognition that the subaortic gradient (30 mm Hg or more) and associated elevations in intra-cavity LV pressure reflect true mechanical impedance to outflow and are of pathophysiologic and prognostic importance to patients with HCM.

Indeed, outflow obstruction is a strong, independent predictor of disease progression to HCM-related death (relative risk vs. nonobstructed patients, 2.0), to severe symptoms of New York Heart Association (NYHA) class III or IV, and to death due specifically to heart failure and stroke (relative risk vs. nonobstructed patients, 4.4).

However, the likelihood of severe symptoms and death from outflow tract obstruction was not greater when the gradient was increased in magnitude above the threshold of 30 mm Hg.

Disease consequences related to chronic outflow gradients are likely to be mediated by the resultant increase in LV wall stress, myocardial ischemia and eventually cell death and replacement fibrosis.

Therefore, the presence of LV outflow obstruction justifies intervention to reduce or abolish significant subaortic gradients in severely symptomatic patients who are refractory to maximum medical management.

Obstruction in HCM is characteristically dynamic (i.e., not fixed): the magnitude (or even presence) of an outflow gradient may be spontaneously labile and vary considerably with a number of physiologic alterations as diverse as a heavy meal or ingestion of a small amount of alcohol.

Different gradient cut-offs have been proposed for segregating individual patients into hemodynamic subgroups, but rigorous partitioning into such hemodynamic categories according to gradient can be difficult because the unpredictable dynamic changes that may occur in individual patients.

Nevertheless, it is reasonable to divide the overall HCM disease spectrum into hemodynamic subgroups, based on the representative peak instantaneous gradient as assessed with continuous wave Doppler:

1) obstructive gradient under basal (resting) conditions equal to or greater than 30 mm Hg (2.7 m/s by Doppler),
2) latent (provocable) obstructive—gradient less than 30 mm Hg under basal conditions and equal to or greater than 30 mm Hg with provocation
3) nonobstructive—less than 30 mm Hg under both basal and (provocable) conditions.

By current clinical convention, LV outflow gradients are routinely measured noninvasively with continuous wave Doppler echocardiography, generally obviating the need for serial cardiac catheterizations in this disease (except when atherosclerotic CAD or other associated anomalies such as intrinsic valvular disease are suspected).

**General considerations for natural history and clinical course**

Hypertrophic cardiomyopathy is a unique cardiovascular disease with the potential for clinical presentation during any phase of life from infancy to old age (day one to over 90 years). The clinical course is typically variable, and patients may remain stable over long periods of time with up to 25% of a HCM cohort achieving normal longevity (75 years of age or older). However, the course of many patients may be punctuated by adverse clinical events, largely related to sudden, unexpected death, embolic stroke, and the consequences of heart failure.

Hypertrophic cardiomyopathy is also a rare cause of severe heart failure in infants and very young children, and presentation in this age group itself constitutes an unfavorable prognostic sign.
In general, adverse clinical course proceeds along one or more of several of the following pathways, which ultimately dictate treatment strategies:

1) high risk for premature sudden and unexpected death;

2) progressive symptoms largely of exertional dyspnea, chest pain (either typical of angina or atypical in nature), and impaired consciousness, including syncope, near-syncope or presyncope (i.e., dizziness/light headedness), in the presence of preserved LV systolic function;

3) progression to advanced congestive heart failure (the “end-stage phase”) with LV remodeling and systolic dysfunction;

4) complications attributable to AF, including embolic stroke.

However, full appreciation of the clinical implications of HCM (and its treatment strategies) requires an awareness of the unique patterns of patient referral and selection biases that have had an important impact on our perceptions of this disease.

Perhaps to a far greater extent than other cardiovascular diseases, much of the published clinical data assembled over four decades have emanated largely from a few selected tertiary centers in North America and Europe, disproportionately comprised of patients referred because of their high-risk status or severe symptoms requiring highly specialized care (such as surgery).

On the other hand, clinically stable, asymptomatic, or elderly patients were often under-represented. Over-dependence on frequently cited, ominous mortality rates of 3% to 6% per year for HCM-related premature death from tertiary centers may have led to an exaggeration of the overall risk and impact of this disease on patients and, thereby, contributed to a misguided perception that HCM is invariably an unfavorable disorder with inevitable, adverse consequences frequently requiring major therapeutic intervention.

However, more recent reports from non-tertiary centers with fewer selected, regional, and community-based cohorts not subject to tertiary center referral bias are probably more representative of the overall disease state, citing annual mortality rates in a much lower range of about 1%, with the survival of patients not dissimilar to that of the general adult U.S. population.

Nevertheless, of note, there are subgroups of patients within the broad HCM spectrum with annual mortality rates far exceeding 1% and conform to the rates of up to 6% per year previously attributed to the overall disease.

Hypertrophic cardiomyopathy attributable to sarcomere protein mutations also occurs in the elderly and should be distinguished from non-genetic hypertensive heart disease or age-related changes in persons of advanced age. The determinants of extended survival in some patients with HCM are largely unresolved. It is possible that benign genetic substrates may convey favorable prognosis and normal life expectancy. However, at present, genotype data are available for only a limited number of elderly patients, with mutations in the cardiac myosin-binding protein C gene being most common.

Older patients with HCM characteristically show relatively mild degrees of LVH and may not experience severe symptoms. Some even have large resting subaortic gradients that are often caused by the SAMseptal contact associated with normal-sized mitral leaflets greatly displaced anteriorly, seemingly by calcium accumulation posteriorly in the mitral annulus, within a particularly small LV outflow tract.

Definitive clinical diagnosis of HCM in older patients with LVH and systemic hypertension is often difficult to resolve, particularly when LV wall thickness is less than 20 mm and SAM is absent. In the absence of genotyping, marked LVH disproportionate to the level of blood pressure elevation, unusual patterns of LVH unique to HCM, or an obstruction to LV outflow at rest represents presumptive evidence for HCM.

Not uncommonly, HCM coexists with other cardiac conditions such as systemic hypertension and/or CAD. In such patients, the management of HCM should be considered independent of any co-morbidity, and each of the disease entities should be treated on its own merit. For example, specific concerns that may arise include avoidance of angiotensin-converting
enzyme (ACE) inhibitors to control hypertension in the presence of HCM-related resting or provable LV outflow tract obstruction and failure to exclude the diagnosis of CAD in those HCM patients with angina pectoris.

In summary, it is probably most appropriate to regard HCM as a complex disease capable of producing important clinical consequences and premature death in some patients, while many other patients reach normal longevity and life expectancy with mild or no disability and without major therapeutic interventions. Many individuals affected by HCM may not require treatment for most or all of their natural lives, and they therefore deserve reassurance with regard to their prognosis.

Symptoms and pharmacological management strategies

A fundamental goal of treatment in HCM is the alleviation of symptoms related to heart failure. Pharmacological therapy has traditionally been the initial therapeutic approach for relieving disabling symptoms of exertional dyspnea (with or without associated chest pain) and improving exercise capacity for more than 35 years, since the introduction of beta-blockers in the mid-1960s. Also, drugs are often the sole therapeutic option available to the many patients without obstruction to LV outflow, under resting or provokable conditions, who constitute a substantial proportion of the HCM population. Indeed, it is the convention to empirically initiate pharmacologic therapy when symptoms of exercise intolerance intervene, although there have been few randomized trials to compare the effect of drugs in HCM.

Exertional dyspnea and disability (often associated chest pain), dizziness, presyncope and syncope usually occur in the presence of preserved systolic function and a nondilated LV.

Symptoms appear to be caused in large measure by diastolic dysfunction with impaired filling due to abnormal relaxation and increased chamber stiffness, leading in turn to elevated left atrial and LV end-diastolic pressures (with reduced stroke volume and cardiac output), pulmonary congestion, and impaired exercise performance with reduced oxygen consumption at peak exercise. The pathophysiology of such symptoms, due to this form of diastolic heart failure, may also be intertwined with other important pathophysiologic mechanisms such as myocardial ischemia, outflow obstruction associated with mitral regurgitation, and AF. Indeed, many patients may experience symptoms largely from diastolic dysfunction or myocardial ischemia in the absence of outflow obstruction (or severe hypertrophy). Other patients (i.e., those with LV outflow obstruction) are more disabled by elevated LV pressures and concomitant mitral regurgitation than by diastolic dysfunction, as is evidenced by the often dramatic
symptomatic benefit derived from major therapeutic interventions that reduce or obliterate outflow gradient (most frequently myectomy or alcohol ablation).

Chest pain in the absence of atherosclerotic CAD may be typical of angina pectoris or atypical in character. Most chest discomfort is probably due by bursts of myocardial ischemia, evidenced by the findings of scars at autopsy, fixed or reversible myocardial perfusion defects and the suggestion of scarring by magnetic resonance imaging, net lactate release during atrial pacing, and impaired coronary vasodilator capacity.

Myocardial ischemia is probably a consequence of abnormal microvasculature, consisting of intramural coronary arterioles with thickened walls (from medial hypertrophy) and narrowed lumen, and/or a mismatch between the greatly increased LV mass and coronary flow. Because typical anginal chest pain may be part of the HCM symptom complex, associated atherosclerotic CAD (which may complicate clinical course) is often overlooked in these patients. Therefore, coronary arteriography is indicated in patients with HCM and persistent angina who are over 40 years of age or who have risk factors for CAD, or when CAD is judged possible prior to any invasive treatment for HCM such as septal myectomy (or alcohol septal ablation).

**Treatment**

**Beta-adrenergic blocking agents**

Beta-blockers are negative inotropic drugs that have traditionally been administered to HCM patients with or without obstruction, usually relying on the patient's own subjective and historical perception of benefit.

However, judgments regarding treatment strategies in HCM with beta-blockers are often difficult, taking into account the frequent day-to-day variability in magnitude of symptoms. Treadmill or bicycle exercise—with or without measurement of peak oxygen consumption—have proved helpful in targeting patients for therapy or determining when changes in dosage or drugs are appropriate. If limiting symptoms progress, drug dosage may be increased within the accepted therapeutic range. Patient responses to drugs are highly variable in terms of magnitude and duration of benefit, and the selection of medications has not achieved widespread standardization and has been dependent, in part, on the experiences of individual
practitioners, investigators, and centers.

Verapamil
In 1979, the calcium antagonist verapamil was introduced as another negative inotropic agent for the treatment of HCM, and has been widely used empirically in both the nonobstructive and obstructive forms, with a reported benefit for many patients, including those with a component of chest pain.

Verapamil in doses up to 480 mg per day (usually in a sustained release preparation) has favorable effects on symptoms, probably by virtue of improving ventricular relaxation and filling as well as relieving myocardial ischemia and decreasing LV contractility.

However, aside from the mild side-effects of constipation and hair loss, verapamil may also occasionally harbor a potential for clinically important adverse consequences and has been reported to cause death in a few HCM patients with severe disabling symptoms (orthopnea and paroxysmal nocturnal dyspnea) and markedly-elevated pulmonary arterial pressure in combination with marked outflow obstruction.

Disopyramide
The negative inotropic and Type I-A antiarrhythmic agent disopyramide was introduced into the treatment regimen for patients with obstructive HCM in 1982. There are reports of disopyramide producing symptomatic benefit (at 300 mg to 600 mg per day with a dose-response effect) in severely limited patients with resting obstruction, because of a decrease in SAM, outflow obstruction, and mitral regurgitant volume.

Drugs in end-stage phase
A small but important subgroup of patients with nonobstructive HCM develops systolic ventricular dysfunction and severe heart failure, usually associated with LV remodeling demonstrable as wall thinning and chamber enlargement. This particular evolution of HCM occurs in only about 5% of patients and has been variously known as the “end-stage”, “burnt-out”, or “dilated” phase. Drug treatment strategies in such patients with systolic failure differ substantially strategies for in HCM patients with typical LVH, nondilated chambers, and preserved systolic function (i.e., involving conversion to after loadreducing agents such as ACE inhibitors or angiotensin II-receptor blockers or diuretics, digitalis, beta-blockers or spironolactone). There is no evidence, however, that beta-blockers prevent or convey a benefit to congestive heart failure and ventricular systolic dysfunction of the “end-of-stage” (by contrast with the experience in dilated cardiomyopathy and CAD). Ultimately, patients with end-stage heart failure may become candidates for heart transplantation, and they represent the primary subgroup within the broad disease spectrum of HCM for when this treatment option is considered.

Asymptomatic patients
Data from largely unselected cohorts and genotyping studies in families suggest that most HCM patients, including many who are not even aware of their disease, probably have no symptoms or only mild symptoms. While most of the asymptomatic patients do not require treatment, some represent therapeutic dilemmas because of their youthful age and the consideration for prophylactic therapy to prevent SCD or disease progression. Prophylactic drug therapy in asymptomatic (or mildly symptomatic) patients to prevent or delay development of symptoms and improve prognosis has been the subject of debate for many years, but it remains on an entirely empiric basis without controlled data to either support or contradict its potential efficacy.
Conclusion

Hypertrophic cardiomyopathy is a genetic or familial disease where muscle in the left ventricle has a predisposition to thicken and prevent normal flow of blood out of the heart. Hypertrophic cardiomyopathy is the most common cause of sudden death in young people, such as exercising athletes.

Dilated cardiomyopathies occur when the heart muscle fibers are abnormally stretched when the heart chambers increase in size and volume. The stretched muscles lose their ability to contract strongly, similar to a slinky or an elastic band that has been overstretched and loses its shape and function. As the heart walls continue to stretch, they can also cause damage to the heart valves between the chambers of the heart causing blood to regurgitate or backwash, and as a result there is decreased cardiac output and heart failure. There are many causes of dilated cardiomyopathy including: infection, alcohol, cancer therapies, chemical poisonings (for example, lead and arsenic), neuromuscular disorders such as muscular dystrophy, and a variety of genetic diseases.

Dilated Cardiomyopathy

Dilated cardiomyopathy is a serious condition that weakens your heart muscle and causes it to stretch, or dilate. When your heart muscle is weak, it can't pump out blood as well as it should, so more blood stays in your heart after each heartbeat. As more blood fills and stays in the heart, the heart muscle stretches even more and gets even weaker.

Most of the time, this leads to heart failure. Heart failure does not mean that your heart stops pumping. It means that your heart can't pump enough blood to meet your body's needs.

The causes of dilated cardiomyopathy
The most common type of dilated cardiomyopathy develops after a heart attack has damaged the heart muscle. But it can also be caused by many diseases or problems that may or may not be related to your heart. Sometimes the cause is not known.

Some of the things that can lead to dilated cardiomyopathy include:

1. Coronary artery disease and heart attack.
2. High blood pressure, which can put stress on the heart walls.
3. Heart valve diseases, including aortic valve regurgitation and mitral valve regurgitation.
4. Myocarditis, which is inflammation of the heart muscle. It is caused by a virus or an immune system problem.
5. Drinking too much alcohol, using certain illegal drugs such as cocaine, or taking certain medicines such as chemotherapy.
6. Being exposed to toxic metals, such as lead or mercury.
7. Being pregnant. In rare cases, dilated cardiomyopathy develops toward the end of pregnancy or during the first 6 months after a woman gives birth. Experts don't know why this happens.

The symptoms

You may not have any symptoms at first. Or you may have mild symptoms, such as feeling very tired or weak.

If your heart gets weaker, you will develop heart failure. When this happens, you will feel other symptoms, including:

- Shortness of breath, especially with activity.
- Tiredness.
- Trouble breathing when you lie down.
- Swelling in your legs.
- Chest pain.

You may get these symptoms slowly, over months or years. Or you may get them suddenly, such as after pregnancy or an illness caused by a virus.

Heart failure that suddenly gets worse is an emergency. Get medical help right away if:

- You have severe trouble breathing.
- You cough up pink, foamy mucus.
- You have a new irregular or rapid heartbeat.

When you have heart failure, keeping track of your symptoms every day is important. Call your doctor if:

- You have a sudden weight gain such as 3 lb (1.4 kg) or more in 2 to 3 days.
- Your ability to exercise changes.
- You have any change in your symptoms.

The diagnose of dilated cardiomyopathy

Your doctor will ask questions about your symptoms and past health. He or she will want to know about recent illnesses and about heart disease in your family. Your doctor will listen to your heart and lungs and check your legs for fluid buildup.
You may also have other tests, including:

- An electrocardiogram, also known as an ECG or EKG.
- A chest X-ray.
- An echocardiogram.
- Coronary catheterization (angiogram).
- Routine blood tests.

In some cases, a doctor may want to look at a small sample of heart tissue, called a biopsy, to make a definite diagnosis.

**Treatment**

Treatment for dilated cardiomyopathy focuses on relieving your symptoms, improving heart function, and helping you live longer.

You will probably need to take several medicines to treat heart failure caused by dilated cardiomyopathy. It is very important to take your medicines exactly as your doctor tells you to and to keep taking them. If you don't, your heart failure could get worse.

Your doctor may suggest a mechanical device to help your heart pump blood or to prevent life-threatening irregular heart rhythms. Such devices include a pacemaker for heart failure (also called cardiac resynchronization therapy or CRT), an implantable cardioverter-defibrillator (ICD), or a combination pacemaker and ICD. If your condition is very bad, a heart transplant may be an option.

Self-care is an important part of your treatment. Self-care includes the things you can do every day to feel better, stay healthy, and avoid the hospital.

**The medicines as prescribed.** This gives them the best chance of helping you. Medicines for heart failure include:

- Angiotensin-converting enzyme (ACE) inhibitors or angiotensin II receptor blockers (ARBs). These make it easier for blood to flow.
- Diuretics. These help remove excess fluid from the body.
- Beta-blockers. These slow the heart rate and can help the heart fill with blood more completely.

**Live a healthy lifestyle.** It can help slow down heart failure. Limit salt, try to get regular exercise, and don't smoke.

**Prognosis**

Most of the time, dilated cardiomyopathy leads to heart failure. Heart failure usually gets worse over time, but treatment can slow the disease and help you feel better and live longer. In more and more cases, the problem is being found earlier, when it can be better managed.

Some people develop other problems, including:

- Stroke.
- Heart attack.
- Sudden cardiac death, which means the heart suddenly stops working. This may be more likely to happen to people who have serious rhythm problems (arrhythmias) in one of the lower heart chambers (ventricles).
If a woman gets dilated cardiomyopathy from pregnancy, she should not get pregnant again. This is true even if her heart problem gets better.

If your disease is getting worse, you may want to think about making end-of-life decisions. It can be comforting to know that you will get the type of care you want.

**RESTRICTIVE CARDIOMIOPAHY**

**Chambers of the Heart**

The heart has four chambers: two atria and two ventricles.

- The right atrium receives oxygen-poor blood from the body and pumps it to the right ventricle.
- The right ventricle pumps the oxygen-poor blood to the lungs.
- The left atrium receives oxygen-rich blood from the lungs and pumps it to the left ventricle.
- The left ventricle pumps the oxygen-rich blood to the body.

**Restrictive cardiomyopathy** is a serious problem that makes your heart muscle stiff. When your heart muscle is stiff, it can't stretch to allow enough blood to enter its lower chambers, the ventricles. So blood that would normally enter the heart backs up in your circulatory system.

Most of the time, this leads to heart failure. Heart failure doesn't mean that your heart stops pumping. It means that your heart can't pump enough blood to meet your body's needs.

**The causes of restrictive cardiomyopathy?**

Often the cause is never found. But we do know that there are a number of diseases or problems that can lead to restrictive cardiomyopathy. These include:

1. Cardiac amyloidosis, a buildup of an abnormal protein in the heart muscle.
2. Hemochromatosis, a buildup of iron in the heart muscle.
3. Sarcoidosis, a rare type of heart inflammation.
4. Radiation therapy and chemotherapy, used to treat cancer.
5. Carcinoid syndrome, a rare disease that causes certain chemicals to be released into the blood stream. These chemicals can stiffen heart muscle.
6. Löeffler's syndrome and endomyocardial fibrosis, conditions that can cause scar tissue in the heart.
7. Genetic factors. You can inherit diseases, including Gaucher disease and Fabry's disease, that can lead to restrictive cardiomyopathy. But these diseases can be treated to prevent restrictive cardiomyopathy.

**The symptoms**

You may not have any symptoms at first. Or you may have mild symptoms, such as feeling very tired or weak.

If your heart gets weaker, you will develop heart failure. When this happens, you will feel other symptoms, including:

- Shortness of breath, especially with activity.
- Tiredness.
- Trouble breathing when you lie down.
- Swelling in your legs.
- Chest pain.

Heart failure that suddenly gets worse is an emergency. Get medical help right away if:

- You have severe trouble breathing.
- You cough up pink, foamy mucus.
- You have a new irregular or rapid heartbeat.

When you have heart failure, keeping track of your symptoms every day is important. Call your doctor if:

- You have a sudden weight gain such as 3 lb (1.4 kg) or more in 2 to 3 days.
- Your ability to exercise changes.
- You have any change in your symptoms.

**The diagnose of restrictive cardiopathy**

Your doctor will ask questions about your symptoms and past health. He or she will want to know about recent illnesses and about heart disease in your family. Your doctor will listen to your heart and lungs and check your legs for fluid buildup.

You may also have other tests, including:

- Electrocardiogram, also known as an ECG or EKG.
- Chest X-ray.
- Echocardiogram.
- Coronary catheterization (angiogram).
- Routine blood tests.

In some cases, a doctor may want to look at a small sample of heart tissue, called a biopsy, to make a definite diagnosis.
The treatment

Most of the time, treatment focuses on relieving symptoms, improving heart function, and helping you live longer. You may also have other treatment for the problem that is causing restrictive cardiomyopathy, such as medicines to get rid of too much iron in the heart muscle (hemochromatosis).

You will probably need to take several medicines to treat heart failure caused by restrictive cardiomyopathy. It's important to take your medicines exactly as your doctor tells you to and to keep taking them. If you don't, your heart failure could get worse.

Your doctor may suggest a mechanical device to help your heart pump blood or prevent life-threatening irregular heart rhythms. Such devices include a pacemaker, an implantable cardioverter-defibrillator (ICD), or a combination pacemaker and ICD. If your condition is very bad, a heart transplant may be an option.

Self-care is an important part of your treatment. Self-care includes the things you can do every day to feel better, stay healthy, and avoid the hospital.

**The medicines as prescribed.** This gives them the best chance of helping you.
Medicines for heart failure include:

- Angiotensin-converting enzyme (ACE) inhibitors or angiotensin II receptor blockers (ARBs). These make it easier for blood to flow.
- Diuretics. These help remove excess fluid from the body.
- Beta-blockers. These slow the heart rate and can help the heart fill with blood more completely.

**Live a healthy lifestyle.** It can help slow down heart failure. Limit salt, and don't smoke. Ask your doctor about how you can exercise safely. People with heart failure due to restrictive cardiomyopathy need to avoid doing too much, since their hearts can't increase blood flow during exercise.

The prognosis

Most of the time, restrictive cardiomyopathy leads to heart failure. Heart failure usually gets worse over time. But treatment can slow the disease and help you feel better and live longer. If your doctor finds the cause of your restrictive cardiomyopathy, then the cause will also be treated, if possible.

Some people develop other problems, including:

- Stroke.
- Heart attack.
- A blood clot in the lung, called a pulmonary embolism.
- Sudden cardiac death, which means the heart suddenly stops working. This may be more likely to happen to people who have serious rhythm problems (arrhythmias) in one of the lower heart chambers (ventricles).