

Restrictive Cardiomyopathy

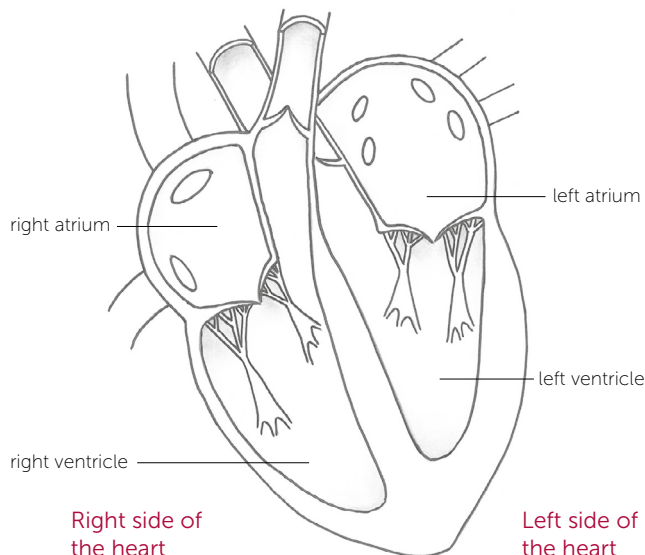
An introduction to restrictive cardiomyopathy

- In restrictive cardiomyopathy the heart's ventricles become stiff, which restricts their contraction.
- It affects how the ventricles fill with blood, and reduces the blood circulation through the heart and to the body.
- Treatment aims to support the heart and reduce symptoms.

What is restrictive cardiomyopathy?

In restrictive cardiomyopathy (RCM), the muscle cells in the heart become replaced with abnormal tissue (such as scar tissue). This causes the muscle walls of the ventricles (the lower chambers of the heart) to become stiff, which restricts the movement of the heart. The pumping action of the heart is not usually affected, and the heart can contract to pump blood around the body. However, it is the filling function of the heart, where the heart muscle relaxes and the ventricles fill with blood, that is affected.

The stiffened muscle of the ventricles does not relax properly which means that the ventricles can't fill with blood. This poor filling function means that the ventricles receive less blood than normal, and so blood flow around the body and the heart is reduced. Also, blood gets 'backed up' as it cannot enter the heart as normal, which causes a build-up of pressure. This can cause the atria (top chambers of the heart) to enlarge.



Who gets RCM?

RCM is relatively rare compared to other types of cardiomyopathy. It is most often diagnosed in children (rather than adults) at 5 – 6 years of age, although it can develop at any age.

What causes RCM?

Although the cause of RCM is often unknown, it can be genetic and run in families. It can also develop due to other conditions such as:

- scar tissue in the heart;
- amyloidosis – a condition where there is too much of an abnormal protein (called amyloid) in the body, and when it builds up in the heart muscle it can cause RCM;
- haemochromatosis – a condition where too much iron from the diet is absorbed and it builds up in the organs, including the heart; and
- following some treatments for cancer, such as radiation therapy.

What are the symptoms of RCM?

Some people have no symptoms of RCM, or very mild symptoms. But over time, the restricted function of the heart can lead to heart failure (where the heart fails to pump enough blood, at the right pressure, to meet the body's needs). This can cause symptoms, which develop as the heart's function reduces.

Symptoms can include the following:

- tiredness due to reduced blood flow and oxygen levels;
- shortness of breath due to fluid around the lungs (which might be when exercising, but can develop and happen even when resting);



- swelling in the abdomen and ankles due to oedema (a build up of fluid caused by reduced blood flow through the heart);
- arrhythmias (abnormal heart rhythms, where the heart beats too fast, too slow or erratically) caused by disrupted electrical signals in the heart;
- palpitations (feeling your heart beating or like it is 'fluttering'), caused by arrhythmias;
- chest pain (which may be caused by reduced blood flow from the heart);
- a cough;
- loss of appetite;
- weight gain; and
- fainting (due to reduced oxygen and blood flow to the brain).

In some people, how the electrical messages are transmitted through the heart can become slowed or blocked. This is called 'heart block', and can cause symptoms such as breathlessness, dizziness and palpitations.

How is RCM diagnosed?

As symptoms can be subtle to start with, diagnosis may be difficult. A diagnosis might involve some or all of the following tests.

- **Medical history** – to look at symptoms and whether other family members have this condition (as it can be genetic).
- **Physical exam** – to see what physical symptoms, if any, are happening.
- **ECG (electrocardiogram)** – to look at the conduction of electrical impulses through the heart and see if arrhythmias are happening.
- **Echo (echocardiogram) and MRI (magnetic resonance imaging) scan** – types of scan that show the structure of the heart. They can also show how the heart is working and the flow of blood through the heart.

Additional tests are not always needed, but might include the following.

- **Cardiac catheterisation** – this involves a thin plastic tube (a catheter) being inserted through the blood vessels into the heart. This allows tests to be done such as measuring the pressure within the heart's chambers, and using ultrasound or x-rays to show the structure and function of the heart.
- **Myocardial biopsy** – this is rarely done, but might help to identify the cause of the someone's RCM. It involves surgically removing a small piece of heart muscle, which is then examined. This is more likely to be done in adults than in children.

- A **bone scan** might be suggested if the RCM could be caused by amyloidosis. (See 'What causes RCM?')

There may be other tests suggested, depending on what the doctors think could be the cause of the RCM. If this is the case, they will talk to you about what the other tests are, and what they are used for.

How is RCM treated and managed?

Although RCM cannot be cured, treatment aims to reduce and control symptoms, particularly those of heart failure. Some treatments may not be suitable, depending on the underlying cause of the RCM. Treatment may involve any of the following.

Medication

- **ACE inhibitors (angiotensin-converting enzyme inhibitors)** – relax the smooth muscle around the blood vessels to reduce the workload on the heart, and reduce the volume of the blood, making it easier for the heart to work.
- **Anti-arrhythmic medication** – reduces abnormal heart rhythms.
- **Anticoagulants (blood thinners)** – may be used in people with arrhythmias to reduce the risk of blood clots forming, which could lead to a stroke.
- **Beta-blockers** – reduce the rate and force of the heart's contraction, by reducing stimulation of adrenalin (which would normally make the heart beat faster).
- **Calcium channel blockers** – reduce the force at which the heart contracts. This can help to reduce blood pressure, chest pain and arrhythmias.
- **Diuretics (water tablets)** – reduce the build-up of fluid on the lungs or the ankles by encouraging the kidneys to get rid of water as urine.
- **Vasodilators** – relax and widen the blood vessels, helping blood to flow more easily.

Other treatments

- **Life-style changes** – such as a healthy diet, low salt intake, exercise, and avoiding alcohol, caffeine and smoking, may all help symptoms of heart failure.
- **Diet** – some children with RCM have difficulties gaining weight, which can affect their growth. They might need a special high-calorie diet to help them to gain weight.
- **Pacemaker** – may be recommended for people who have heart block due to RCM (which makes the heart rate slow down). Pacemakers take over the electrical signalling of the heart to keep a normal heart rhythm.



- **LVADs** – these artificial pumps are used to help the heart to pump blood around the body. They are used for severe heart failure, and often for people while they are waiting for a transplant.
- **Heart transplant** – in some cases, people with RCM require a heart transplant, if their heart function is very poor.
- **Treatment for other conditions** – if the RCM is caused by another underlying condition, this might be treated separately.

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