



Cardiomyopathy- the facts



What is it?

Cardiomyopathy means disease of the heart muscle.

If you have cardiomyopathy it may mean that your Heart muscle has become enlarged, thicker or stiff. If this happens your heart is less able to pump blood through your body and you may get abnormal heart rhythms. This can lead to heart failure or arrhythmia. In turn, heart failure can cause fluid to build up in the lungs, ankles, feet, legs, or abdomen. The weakening of the heart can also cause other severe complications, such as heart valve problems.

Different types of cardiomyopathy?

- Dilated cardiomyopathy
- Hypertrophic cardiomyopathy

You can get cardiomyopathy at any age, however if you have Barth Syndrome you are most likely to get this as a baby. Although you can get different types of cardiomyopathy if you have Barth Syndrome the most common type is dilated cardiomyopathy.



Tell me more about dilated cardiomyopathy?

Having dilated cardiomyopathy means that the left ventricle becomes dilated (stretched). When this happens, the heart muscle becomes weak and thin and is unable to pump blood efficiently around the body. This can lead to fluid building up in the lungs and a feeling of being breathless. This collection of symptoms is known as heart failure.

Dilated cardiomyopathy develops slowly, so most people have quite severe symptoms before they are diagnosed. There may also be 'mitral regurgitation'. This is when some of the blood flows in the wrong direction through the mitral valve, from the left ventricle to the left atrium.



How do you get it?

Cardiomyopathy can be acquired or inherited. "Acquired" means you aren't born with the disease but you develop it due to another disease, condition, or infection. "Inherited" means your parents passed the gene for the disease on to you. In Barth Syndrome it is inherited.

What about the future?

For most children that have cardiomyopathy the heart will start to work better over time but you may need to stay on medications to help your heart function well. You may find exercise becomes a bit more difficult but you can usually do normal day to day activities. A very small number of young people with dilated cardiomyopathy do get severe symptoms and this may mean they are more likely to get an abnormal heart rhythm. By regularly coming to clinics your medical team can check how well your heart is working and answer any questions you may have.

What about treatments?

At present there is no cure for dilated cardiomyopathy, but treatment with medicines helps to control symptoms and reduces the risk of the condition getting worse or of new symptoms. In some very rare cases, ventricular assist getting devices or a heart transplant may be considered. In cardiomyopathy associated with an abnormal heart rhythm some people may need to have a pacemaker or an ICD (implantable defibrillator) fitted.

Testing how well your heart is coping



At your local hospital or Barth Syndrome Clinic you may be asked to do a number of tests to look at how well your heart is coping.

What is an Echocardiogram (Echo)?

The echocardiogram or 'echo' looks at the structure of your heart and the heart valves, and also gives information on the function and pumping action of your heart.

You will have the tests in a private room as you will be asked to take your top off. You will be asked to lie on a bed and jelly will be rubbed onto your chest to help the probe make contact with your skin. The probe will be placed on different areas of your chest and it gives off pulses of high frequency sound waves which pass through your skin to your heart. The ultrasound waves either reflect or are absorbed against the side of your heart and the probe picks up the reflections and shows them in pictures on the machine. The tests can last from 15 minutes to an hour and is safe and easy to do.



What is an Electrocardiogram (ECG)?

An ECG can help detect problems with your heart rate or— called arrhythmias. Small sticky patches called electrodes are put on your arms, legs and chest. These are connected by wires to an ECG recording machine which picks up electrical signals that make your heart beat.

The electrical activity is recorded and printed out onto paper. The test is about 5 minutes long and is painless. Sometimes we will ask to do a signal averaged ECG, this works exactly the same way but takes a bit longer and we need to ask you to lie very still for around 20 minutes.



What is a 24hr tape?

You may be asked to wear a small ECG monitor machine that records your heart rhythm continuously for 24 hours. This can provide a lot of useful information on your heart rate, in particular any tendency to fast or slow heart rhythms or extra beats that may require treatment.

Three sticky pads (electrodes) are placed on your chest and connected by fine leads to a lightweight recorder. This is worn in a small pouch around the neck or on a belt for 24 hours. The physiologist will let you know when you need to return the recorder. You will also be given a diary card to record any symptoms such as dizziness or palpitations. These can be checked against your heart rhythm when the recorder is analysed.

It can take around 15 minutes to set up.

If you have any questions about tests or how your heart is working please contact your medical team

