Dilated cardiomyopathy

Cardiomyopathy UK
the heart muscle charity

An introduction to dilated cardiomyopathy or 'DCM'

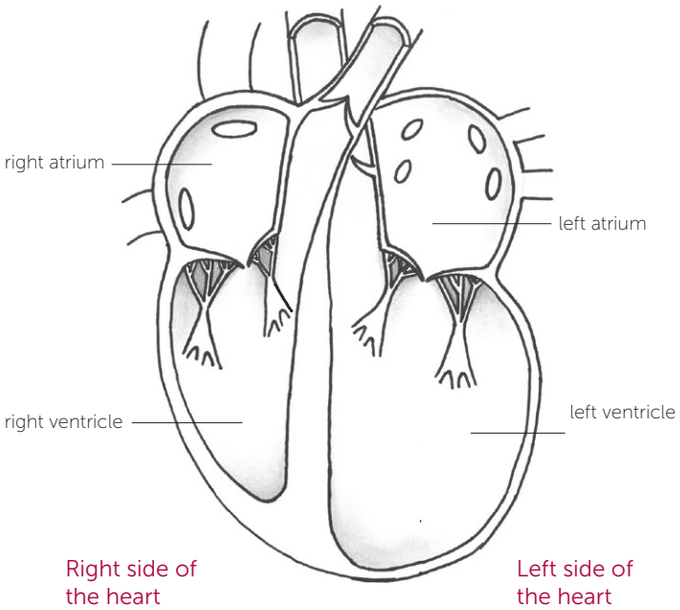
- DCM is a condition where the heart chambers become enlarged, which affects its ability to pump.
- The left ventricle of the heart becomes enlarged (dilated) and the muscle wall becomes thinner. This gives the heart a more rounded (rather than the normal cone) shape.
- The left ventricle becomes weaker and not able to pump as effectively as normal.

What is DCM?

The ventricles are the bottom chambers of the heart, which receive blood from top chambers and pump it to the lungs and to the rest of the body. The walls of both ventricles are thick and muscular. However, the wall of the left ventricle is thicker than the right as it is the main pumping chamber: pumping blood at high pressure to all areas of the body.

In DCM, the left ventricle becomes stretched and enlarged. The walls of the ventricle become thinner and can appear 'baggy'. This gives the heart a rounded or spherical shape, rather than its usual 'cone' shape. The thinner walls are weakened, and so are not able to contract or 'squeeze' as normal. This makes the heart less effective at pumping blood out.

The structure of the heart



How many people have DCM?

There is no definite figure for the number of people with DCM. However, recent studies suggest that as many as 1 in 250 people have a cardiomyopathy.

What causes DCM?

There are many different causes of DCM.

The genetics of DCM

It is thought that around 30% of DCM has a genetic cause. This might be a direct cause that means DCM develops, or a genetic *predisposition* to DCM (an underlying tendency to developing it), which is then triggered by an event such as a viral infection or pregnancy. However, there are up to 100 genes that, either alone or in combination, can cause DCM. In addition, there are some genetic conditions, such as Duchenne muscular dystrophy, that can cause DCM.

Where DCM is genetic, it is usually inherited as an 'autosomal dominant trait'. This means that the mutated gene is on one of the non-sex chromosomes (called the autosomes), and a child of an affected parent will have a 50% chance of inheriting the gene mutation. More rarely, it can be a mutation on the X (female sex) chromosome. Where DCM has a genetic cause, it is recommended that first-degree relatives (children, siblings and parents) of the person with DCM are also screened for DCM. This usually involves having an echocardiogram and an Electrocardiogram (see page 3), under the care of a cardiologist. Genetic testing of relatives may also be considered if there is a known gene mutation. This involves looking for the same genetic mutation responsible for DCM in a first degree relative.

For many individuals with DCM genetic testing does not have a direct impact on their condition or how it is managed. However, an exception to this is a mutation in a gene called 'lamin A/C'. It is important to identify individuals with this mutation as it increases the risk of sudden cardiac death (SCD). In individuals with this specific mutation an implantable defibrillator is usually implanted as a preventative measure, which prevents SCD (see page 3).

Other causes of DCM

- Auto-immune disease – a condition of the immune system, where the normal immune response doesn't work properly, is over-active, and attacks the body's healthy cells and organs. Antibodies in the blood can be used to help diagnose auto-immune conditions.
- Viral infections that affect the heart – such as viral myocarditis, which can cause DCM during infection or because the virus does not clear from the heart.

- Toxins - exposure to toxins such as excessive alcohol and some drugs used in chemotherapy treatment. However, this cause is rare, and it may be that the person has a genetic predisposition towards DCM.
- Pregnancy – called peripartum cardiomyopathy, DCM develops during, or within three months from birth.

DCM and ejection fractions

The heart's ejection fraction – or EF – is often used in DCM as one measure of how effectively the heart is pumping. The EF is a measure of the percentage of blood that is pumped out (or 'ejected') from the heart when the left ventricle contracts, compared to the amount of blood within the ventricle. The EF of a healthy heart is usually around 50-60%. In someone with DCM, their EF is usually under 45% (considered 'abnormal'). However, it is normal to have variations in EF and it can vary a little from one test to another.

Although EF is commonly used to monitor how the heart is working over a time, it is considered alongside the symptoms someone experiences and the results of other tests. As the EF doesn't always match the severity of someone's symptoms, it is usually not considered as an accurate measure of heart function on its own.

What are the symptoms of DCM?

DCM can be hard to recognise. The typical symptoms, such as breathlessness, can build up slowly, and can be caused by other conditions. For example, asthma also causes breathlessness. This can delay diagnosis of the condition, which means that the heart may be severely affected by the time DCM is diagnosed.

Typical symptoms include the following.

- **Breathlessness** (or dyspnoea) – fluid builds-up in the lungs, making it harder to breathe.
- **Swollen legs, ankles and tummy** – build-up of fluid in the tissues, because the heart isn't pumping effectively, can cause swelling (called 'oedema').
- **Fatigue (tiredness)** – as the heart's function is reduced, less energy is delivered to the tissues, causes excessive tiredness.
- **Palpitations** (feeling your heart beating too fast, too hard or like it is 'fluttering') – this is caused by arrhythmias (when the electrical messages which control the heart's rhythm are disrupted).
- **Chest pain** – caused by reduced oxygen levels getting to the heart.
- **Mitral regurgitation** – a small amount of blood leaks back through the mitral valve when the left ventricle contracts (instead of flowing through the aorta out of the heart). This is caused by the mitral valve not closing tightly shut when the ventricle contracts. When the blood flows in the wrong direction it can be heard through a stethoscope, and is referred to as a heart murmur.

Arrhythmias

DCM can cause arrhythmias (abnormal heart rhythms) as the stretched heart muscle can develop scars. Scars can disrupt the normal electrical signalling that control the heart beat. This disruption causes a change in the heart's rhythm, making it beat too fast, too slow or erratically. Arrhythmias include the following.

- **Atrial fibrillation** (AF) – the atria beat very quickly and erratically, and are uncoordinated. This can make the blood flow around the atrium 'turbulent', and the heart less efficient at pumping out blood. AF can cause palpitations and increase the risk of blood clots forming, which can increase the risk of a stroke. Most people with AF will take blood thinning drugs (anticoagulants) to prevent clots.
- **Left bundle branch block** (LBBB) – the electrical impulses that cause the heart to beat, and the ventricles to contract, normally travel through the ventricles via a pathway called the bundle of His. The bundle has branches in the left and right ventricle walls. In LBBB the electrical messages are blocked, or slowed down, through the left side of the bundle. This affects the transmission of electrical messages and reduces how efficiently the heart works. This reduces the amount of blood that leaves the heart and circulates around the body.
- **Ventricular ectopic beats** (VEB) – VEB are when a premature or extra beat starts in the ventricles (rather than the normal contraction which starts in the atria) and causes the heart to beat too early. This causes an irregular heart rhythm.
- **Ventricular tachycardia** (VT) – VT is caused by abnormal electrical activity in the ventricles, which makes the heart contract abnormally fast (over 100 beats per minute). This can lead to loss of consciousness as the heart's output (amount of blood leaving the heart) is reduced.
- **Ventricular fibrillation** (VF) – the heart's system for regulating the heart beat is interrupted by multiple abnormal impulses coming from the ventricle walls. This causes the ventricles to contract in an uncoordinated way, and they 'quiver' rather than contract normally, so blood is not pumped out of the heart effectively. This condition is life-threatening as it can lead to a cardiac arrest and requires urgent treatment with a defibrillator.

What are the complications of DCM?

DCM can lead to additional complications.

- **Heart failure** – when the heart is not working effectively and it 'fails' to pump enough blood, at the right pressure, to meet the body's needs. It describes a collection of symptoms caused by a heart that is struggling to work effectively, such as weight gain (due to fluid retention), shortness of breath, a cough, oedema, palpitations, dizziness and tiredness.

See our factsheet 'About heart failure'.

- **Heart block** – the electrical activity that controls the heart beat is slowed or stopped, and stops the heart contracting normally. A pacemaker might be needed if the heart is unable to keep a normal rhythm.
- **Sudden cardiac death** (SCD) – this can happen due to dangerous arrhythmias, such as ventricular fibrillation (VF), where the contraction of the ventricles is uncoordinated, and they 'quiver' rather than contracting normally, so blood is not pumped out of the heart effectively. Although it is relatively rare, if VF is not controlled (using a defibrillator to shock the heart back into normal rhythm), it can cause the heart to stop beating (a cardiac arrest).

How is DCM diagnosed?

There are several tests that might be used to diagnose DCM, including the following.

- **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, the person may be experiencing.
- **Electrocardiogram** – this looks at the electrical activity of the heart and can identify arrhythmias (abnormal heart rhythms) or ectopics (extra beats) An ECG might also be done during exercise (called an exercise test).
- **Exercise tests** – these are tests done during exercise, such as on an exercise bike or a treadmill, to look at how the heart works during exertion. Sometimes oxygen consumption is measured at the same time (this is called Cardiopulmonary Exercise Test-ing - CPET).
- **Holter monitoring** – this is when an ECG is recorded over a period of time, such as a few hours or days, while carrying on with normal activities. The 'holter' is a ECG device which is worn around the waist or in a pocket which makes it possible to monitor whilst normal activities are undertaken.
- **Echocardiogram** (Echo) – this is a type of ultrasound scan, which uses sound waves to create echoes when they hit different parts of the body. This looks at the structure of the heart, including the walls and valves of the heart and how it is working.
- **Cardiac Magnetic Resonance Imaging** (Cardiac MRI) scan - this scan produces high quality images and is used to look at the structure of the heart and can identify the presence of fibrous tissue within the heart muscle walls.
- **Electrophysiology study** (EPS) – this test involves having a long tube called a catheter inserted into a blood vessel and fed up to the heart. Electrical signals are sent through the catheter to the heart which makes it beat at different rates, which is recorded. This can be used to find where in the heart arrhythmias are starting (and can be used to identify treatment options).

www.cardiomyopathy.org
Helpline 0800 018 1024 Mon-Fri 8.30am-4.30pm
(Free from a landline, mobile costs vary)

- **Coronary angiography/ cardiac catheter –** this is used to see the structure of blood vessels. It involves a catheter being inserted into a blood vessel in the groin and dye being injected into the vessel. An x-ray is then used to look for blockages.
- **Myocardial perfusion scan** (or **radionuclide test**) – this involves having a radioactive substance (called a tracer) injected into a blood vessel. A special type of scanner is used, called a CT scanner. This uses x-rays to produce images of the heart. The tracer assesses the flow of the blood to the heart muscle during exercise and rest.
- **Trans-oesophageal echocardiogram** (TOE)– this test is special kind of echo that involves swallowing a small probe (thin tube) which takes pictures of the heart and blood vessels. This procedure provides a more detailed picture of the inside of the heart than a transthoracic eachocardiogram.

How is DCM treated and managed?

Treatment for DCM aims to control symptoms and reduce complications, and the condition itself may improve with treatment. However, ongoing monitoring of treatment and symptoms is important. Treatment is individualised to the symptoms someone experiences. It may include any of the following.

Medication

- **Diuretics** (water tablets) – reduce the build-up of fluid on the lungs or the ankles by encouraging the kidneys to get rid of excess fluid as urine.
- **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.
- **Beta-blockers** – reduce the rate and force of the heart's contraction, by reducing stimulation of adrenaline (which would normally make the heart beat faster). Ivabradine may be used for people unable to take beta blockers.
- **Anti-arrhythmic medication** – reduces abnormal heart rhythms and helps to control the normal rhythm.
- **Anticoagulants** (blood thinners) – may be used in people with arrhythmias to reduce the risk of blood clots forming, which could lead to a stroke.
- **Angiotensin II Receptor Blockers** (ARBs) – dilate (enlarge) the blood vessels which helps to reduce blood pressure and may be used if the person is not able to tolerate ACE inhibitors.
- **Dapagliflozin** - an SGLT2 inhibitor prescribed in addition to standard medication, used to treat heart failure. Like Entresto, prescription follows NICE Guidelines (see page 4).
- **Sacubitril** and **valsartan** (brand name Entresto) – contains the ARB valsartan and an anti-hypertensive drug called sacubitril. It is a relatively new medication and may be used in severe heart failure, where a combination of ACE inhibitors and beta blockers are not effective.

Devices

- **Pacemaker** – pacemakers either send electrical impulses on demand, or continuously, to take over the electrical signalling of the heart and keep a normal rhythm. This may be recommended for people with some types of arrhythmias.
- **Biventricular pacemaker** – this is a specific type of pacemaker used in cardiac resynchronisation therapy (CRT). It usually has three leads, connected to the right atrium, right ventricle and left ventricle. It takes over the electrical stimulation of the heart, sending electrical signals to both sides of the heart to make both ventricles beat in synchrony. It may be used for people with LBBB (see arrhythmias).
- **ICDs** (implantable cardioverter defibrillator) – these detect and correct dangerous arrhythmias which could otherwise lead to a cardiac arrest. An ICD combined with a biventricular pacemaker is used if both functions are required.
  Note: anyone with DCM and an ejection fraction of 35% or less will usually be considered for an ICD. If someone's DCM is caused by a lamin A/C gene mutation (see causes) an ICD is recommended.

Surgery

When medication and devices don't fully control symptoms, surgery might be considered.

- **LVAD** (Left ventricular assist device) – this mechanical device helps to pump blood out of the left ventricle (main pumping chamber of the heart) when it is not working properly. This is currently only used to support the heart while waiting for a transplant.
- **Heart transplant** – a small number of people may have a heart transplant if their heart doesn't respond to other treatment. This involves removing the failing heart and replacing it with a donor heart.

Lifestyle management

In addition to medication and devices, there may be ways to reduce the effect of DCM through lifestyle. The following are examples of what might help.

- **Minimise alcohol** – alcohol can raise your heart rate and increase blood pressure. You may not need to completely avoid it, but staying within recommended guidelines can reduce any potential affects.

The Chief Medical Officer reviewed these guidelines in 2016. You can read them at www.gov.uk and search 'alcohol guidelines'.

- **Healthy eating** – a balanced diet can help to keep a healthy weight, which will reduce the impact on the heart as well as helping with general health.
- **Minimising salt** – reducing salt intake can help to reduce water retention (which can cause swelling in the ankles and tummy) and blood pressure, which helps with heart function. Your specialist can give you guidance on your salt intake.
- **If you smoke** – stopping smoking is important to help your overall health as well as your heart and lung function (as it can reduce oxygen levels in the blood as well as narrowing blood vessels). Your GP or an NHS stop smoking service may be able to help.
- **Minimise caffeine** – some people are more sensitive to the effects of caffeine than others, and it can cause palpitations in some people. For anyone who *is* sensitive to caffeine, limiting tea and coffee with caffeine, and avoiding items high in caffeine (such as energy drinks, strong coffee and high cocoa content chocolate), may be helpful.
  If you are not sure whether caffeine might affect you, you might like to talk to your specialists about how to manage this.

A note on exercise

Exercise is often recommended for people with a heart condition. Moderate exercise can be important for people with DCM, depending on what symptoms they have. If you have DCM you might like to talk to your doctors about what exercise is suitable for you. You might also like to ask whether you can have cardiac rehabilitation, which offers practical advice about exercise.

We're here for you

At Cardiomyopathy UK we offer help and support for you and your family. We have information about each type of cardiomyopathy as well as diagnosis, treatment and lifestyle issues. Look on our website or call us for more information. Call our helpline to talk to our cardiomyopathy support nurses. We can put you in contact with other people affected by cardiomyopathy through our support groups, peer support volunteers and social media. Contact us for more about our services, or look online.

NICE guidelines

NICE (the National Institute for Health and Care Excellence) produce guidance on treatment, including the use of medication.

Visit www.nice.org.uk and search 'heart failure'.

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