

Peripartum cardiomyopathy

Pregnancy-related cardiomyopathy

- Peripartum cardiomyopathy is a rare type of heart muscle disease which occurs during the last part of, or in the first few months after, pregnancy.
- The heart becomes enlarged and weakened, and less able to pump blood than normal.
- Peripartum cardiomyopathy is serious, but around 70% of women recover by 1 year following diagnosis (based on their ejection fraction, rather than their symptoms).

What is peripartum cardiomyopathy (PPCM)?

Peripartum cardiomyopathy (or PPCM) happens during pregnancy, usually starting between the final month of pregnancy and the first five months after giving birth. In PPCM the woman's heart becomes enlarged and its pumping action weakened.

PPCM is similar to dilated cardiomyopathy, in that it affects the left ventricle of the heart. This is the main pumping chamber of the heart: pumping blood out of the heart and to the rest of the body. Normally the left ventricle pumps blood out of the heart efficiently, with enough pressure to reach all areas of the body.

In PPCM, the left ventricle chamber becomes enlarged. As the ventricle becomes bigger than normal, the muscular wall becomes stretched and thinner. This makes the muscle weaker which, in turn, means that it works less well. This results in less blood being pumped out of the heart (see 'ejection fraction' below), and being pumped out at less force than normal. Due to blood not circulating effectively this can cause fluid to build up in the tissues, particularly the lungs, which means that less oxygen gets to the body's organs. When the impact of this causes symptoms such as breathlessness, coughing and extreme fatigue this is referred to as 'heart failure' as the heart is failing to meet the body's normal demands for oxygen.

♥ See our factsheets on how the heart works and DCM.

The ejection fraction is a measure of the amount of blood that is pumped out (ejected) from the heart each time the left ventricle contracts. It is usually measured as a percentage of the amount of blood within the ventricle. A normal ejection fraction is 55– 60%. Ejection fractions lower than this can mean that the heart is working less well.

What causes PPCM and how common is it?

PPCM is rare. Figures vary, but it is thought to affect around 1 in 5,000 to 1 in 10,000 women or 1 in every 2000 women who give birth. Although it can affect women at any age, it seems to be more common in those over 30.

It is not clear why PPCM develops, and it can develop in women with no history of heart conditions. It may be that the condition is caused by the increased pressures on the heart during pregnancy, or when someone has cardiomyopathy in the family (because cardiomyopathy can be genetic). However, the causes are currently not fully understood.

It is thought that women who are overweight, older, have had previous children, or a history of hypertension (high blood pressure) may be at increased risk.

What happens to the heart during pregnancy?

During pregnancy, the body goes through lots of changes to cope with the development of the baby and the demands this places on the mother's body. This includes changes that happen to the heart in response to the pregnancy. These changes are normal, and include:

- reduced blood pressure (during early stages of pregnancy, which increases slightly nearer the end);
- water retention;
- approximately 40% increase in total blood volume;
- increased heart rate (starting within the first few weeks); and
- increased cardiac output (a measure of the volume of blood that is pumped by the heart during one minute) which can increase by 40 – 50% by the first six weeks of pregnancy.

What are the symptoms of PPCM?

Although the symptoms of PPCM are clear, they are also similar to symptoms that can also happen in pregnancy. However, it is important that they are recognised as early on in pregnancy as possible so that they can be responded to with appropriate treatment.

It is important be aware of any symptoms that are worse than what would *normally* be expected during pregnancy, or if they get worse over time.

PPCM usually causes heart failure. This happens because the heart isn't working as well as it should, and fails to pump enough blood, at the right pressure, to meet the body's needs.



The symptoms of heart failure include:

- breathlessness, especially when resting or lying down;
- a cough;
- water retention (causing swelling in the ankles and tummy);
- palpitations (a change in the heart rate or rhythm that the person may become aware of, particularly tachycardia – an abnormally fast heart rate);
- extreme fatigue (as oxygen levels are low);
- finding it hard to exercise or be active; and
- some women may also have swollen veins in their neck due to blood not being pumped efficiently.

There is also a risk of blood clots forming in the lungs or heart. Although blood clots are a risk in all pregnancies, in PPCM it is an added risk due to the heart not pumping properly.

How is it diagnosed?

When the potential symptoms of PPCM are seen, the woman should be urgently referred to a cardiologist for investigation. There are several tests that might be done to diagnose PPCM. These tests are used to check how well the heart is working, and whether the symptoms experienced are due to the pregnancy, to other conditions, or due to cardiomyopathy.

- A **medical history** – to look at possible causes for any symptoms, such as an existing heart condition. This might include when the symptoms first started, and when they happen, such as feeling tired, breathless and having palpitations.
- A **physical exam** – to look for any possible causes for symptoms. This might include listening to the lungs, seeing whether breathing becomes more difficult when lying flat, whether there is discomfort in the chest and any coughing.
- **Blood tests** – to check how the kidney, liver and thyroid are working, to check for hormones which may indicate the heart is under stress, and to check for other causes of the symptoms such as anaemia or an infection.
- **Urine test** – to test for pre-eclampsia (a pregnancy-related high blood pressure) or urine infections.
- An **ECG** (electrocardiogram) – to look at how electrical impulses are conducted in the heart, and check for arrhythmias (abnormal heart rhythms).
- A **24-hour ECG (Holter)** - this checks for any episodes of heart rhythm disturbances.
- A **chest x-ray** – to look for fluid on the lungs.
- An **echocardiogram** – similar to an ultrasound scan to look at the structure and function of the heart muscle and valves, and check for any blood clots within the heart chambers. This uses a 'transducer' to create ultrasound waves, which form an image of the heart.

In some cases, the following test might also be done to help with the diagnosis. This will depend on the results of the tests above, and whether they are felt to be suitable at the time of diagnosis.

- **Cardiac MRI** (type of scan) – to look at the structure and function of the heart. This might be used if the results from the echo are not clear, and to look for blood clots. It can also be used to rule-out other conditions that can affect the heart (such as infections or inflammation).
- **Coronary angiogram** – this looks at the blood supply to the heart (through the coronary arteries which supply the heart muscle with blood). It is used to check that the arteries are not narrowed or blocked due to coronary artery disease.

PPCM may be diagnosed when:

- the heart is found to be enlarged;
- symptoms of heart failure are seen;
- the ejection fraction is 40% or lower; and
- no other cause of the symptoms can be found.

How is it treated?

Treatment focuses on managing the symptoms of heart failure, and will be individualised to the symptoms the woman is experiencing, and the stage of pregnancy. There are specific guidelines for the treatment of heart failure, and these include:

- **diuretics** (water tablets) - to reduce the build-up of any fluid on the lungs or the ankles by encouraging the kidneys get rid of water as urine;
- **beta-blockers** – to reduce the rate and force of the heart's contraction, by reducing stimulation of adrenalin (which would normally make the heart beat faster); and
- **ACE inhibitors** (angiotensin-converting enzyme inhibitors) may be given after delivery – to 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. Unfortunately, treatment with ACE inhibitors means the mother is no longer able to breast feed.

Women who have arrhythmias during pregnancy (abnormal heart rhythms, which can affect how well the heart is pumping) may have specific treatment for this. This depends on the arrhythmia they experience.

♥ *See our factsheets on arrhythmias.*

Although it is very rare, PPCM can be life-threatening and needs to be treated promptly. If the mother does not recover well she may need ongoing treatment. The drug therapies for the condition can be very effective but other treatment options are considered according to the individual needs, for example, a small number of women affected may eventually need a heart transplant.



Treatment may involve many different healthcare professionals, including a cardiologist, obstetrician and midwife, all of whom will look after the mother and baby and will help the mother to make decisions about her treatment and care throughout the pregnancy and afterwards.

Does PPCM affect pregnancy and delivery?

Whether and how much PPCM affects a pregnancy and developing baby will depend on when the PPCM starts, and how the symptoms affect the mother. For this reason, it is important that symptoms are recognised as early as possible, and a diagnosis made, so that appropriate treatment can start.

How the baby is delivered will depend on the mother's symptoms. While a vaginal birth is often possible, a caesarean section might be considered if the mother is very unwell.

The possibility of a termination of the pregnancy may need to be considered. This is a difficult decision but may need to be discussed if there is an issue of substantial risk to the mother.

If someone has PPCM during pregnancy, will they always have it?

Although PPCM is a serious condition, it is worth remembering that many women will make a good recovery. Over half of women with PPCM will have normal heart function again by six months after giving birth.

For some women recovery can take several years due to the severity of their symptoms, and how well their heart can recover from heart failure. If they have any other conditions this can also delay their recovery. However, some women may continue to have symptoms that need ongoing treatment, and it may cause further complications.

It is recommended that women should have echo scans six weeks after giving birth, and every six months, to check how their heart is recovering.

If someone develops PPCM, will it happen with future pregnancies?

If a woman has had PPCM during a pregnancy, research suggests she has an increased chance of getting it again in future pregnancies, even if she recovers completely.

In future pregnancies, the symptoms of heart failure can be worse than before. This risk can be difficult to predict, and the causes are not clearly known. However, this may be important to know if considering becoming pregnant again.

In planning a future pregnancy it is advisable to discuss this with the cardiologist to clarify what potential risks this may entail, and what monitoring would be required during the pregnancy.

*This factsheet is specifically about peripartum cardiomyopathy – a condition that develops during, and because of pregnancy. Some women have cardiomyopathy that is not **due** to pregnancy, but may need particular treatment and management of their condition **during** pregnancy. We have information on other types of cardiomyopathy available on our website or by calling us.*

We are here for you

We understand that this factsheet contains some potentially worrying or frightening information. If you have, or think you have, peripartum cardiomyopathy, it is important to talk to the healthcare professionals involved in your care. If you would like to talk in confidence to someone about what is happening and how you are feeling, you may like to talk to our helpline. Or helpline is staffed by specialist nurses.

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. We can put you in contact with other people affected by cardiomyopathy through our support groups, support volunteers, social media and our online forum. Contact us for more about our services, or look online.

Cardiomyopathy^{UK}
the heart muscle charity

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