Peripartum cardiomyopathy



Pregnancy-related cardiomyopathy

- Peripartum cardiomyopathy is a rare type of heart muscle disease which occurs during the last part of pregnancy, or in the first few months after pregnancy.
- Peripartum cardiomyopathy affects the function of the left ventricle of the heart.
- Peripartum cardiomyopathy is a serious and life-threatening condition, but around 70% of women recover by 1 year following diagnosis.

What is peripartum cardiomyopathy (PPCM)?

Peripartum cardiomyopathy (or PPCM) is a type of cardiomyopathy that only occurs during or following pregnancy. It is usually diagnosed either during the last trimester of pregnancy or the first five months after giving birth.

PPCM affects the left ventricle of the heart - it's main pumping chamber, which pumps blood out of the heart and to the rest of the body. In the normal heart the left ventricle pumps blood out of the heart efficiently, with enough pressure to reach all areas of the body. However, in a heart affected by PPCM the muscle walls of the left ventricle can become stretched and weakened and so it becomes less efficient at pumping blood out of the heart (as in dilated cardiomyopathy). The heart is then pumping less blood at a lower force around the body. This is referred to as a reduced 'ejection fraction' and causes symptoms of heart failure, such as breathlessness, coughing and extreme fatigue (because the heart is failing to meet the body's normal oxygen requirements).

♥ 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. In PPCM the ejection fraction is generally less than 45%.

What causes PPCM and how common is it?

PPCM is rare and can only happen during or following pregnancy. The incidence of PPCM varies globally from around 1 in 100 pregnancies in African countries to 1 in 10,000 pregnancies in Denmark. It is not currently known what the incidence of PPCM is in the UK. The reasons why PPCM develops are still not fully understood and more research is needed. However, PPCM is more common in women who have a history of high blood pressure or who develop preeclampsia during pregnancy. PPCM can develop in women with no previous history of heart conditions. Other risk factors include ethnicity, family history, being pregnant at an older age (over 30-years), obesity (having a raised BMI), having multiple pregnancy (such as carrying twins) as well as lifestyle factors (such as smoking and malnutrition). Pregnancy is a natural stress test for the body and this may trigger PPCM, particularly where other factors also exist.

What happens to the heart during pregnancy?

During pregnancy, the body goes through lots of changes. Changes to the cardiovascular system occur from the early weeks of pregnancy and continue as the pregnancy progresses, including further changes to cope with labour, and again as the body returns to normal after the baby is born. Changes during pregnancy include:

- 40% increase in total blood volume.
- increased heart rate.
- 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.
- blood also become more susceptible to clotting.

What are the symptoms of PPCM?

Symptoms of PPCM include:

- feeling breathless
- having a fast heart rate and/or palpitations
- a persistent cough, mainly at night, which feels like "gasping for air"
- being unable to lie flat in bed (needing to be propped up with pillows or to sit up)
- noticing sudden and severe swelling of feet, ankles and hands (due to fluid retention)
- sudden and significant weight gain (due to fluid retention)
- excess fatigue
- feeling or looking 'not quite right'

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PPCM usually causes heart failure as 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 PPCM are symptoms of heart failure.

However, in pregnancy these symptoms can often be ignored or considered to be a normal part of pregnancy as they overlap with common symptoms of pregnancy.

It is really important to be aware if these symptoms occur, particularly if they are getting worse over time. If you have symptoms, please talk to your midwife, GP or obstetrician immediately. Describe your symptoms and let them know that you are worried. As PPCM is relatively rare, you may want to ask them directly to consider whether your symptoms could be caused by PPCM.

How is it diagnosed?

If any symptoms of heart failure develop, several investigations will be undertaken to check how well the heart is working and whether the symptoms experienced are due to the pregnancy, other conditions, or if they may be due to cardiomyopathy. Once other causes for these symptoms are ruled out, the woman should be referred to a cardiologist for specialist review. There is no specific test to confirm PPCM, so it can only be diagnosed when all other causes for the symptoms of heart failure are ruled out. Examinations include:

- A medical history the clinician will ask questions about your past as well as present health and enquire whether anyone in your family has (or had) heart disease, or if there have been any unexplained deaths in your family. They will also ask you to describe your symptoms in detail, including when they first started, when and how often you notice them and how much they affect your everyday life.
- 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.
- A physical exam this might include listening to your heart and lungs, checking your blood pressure and pulse, measuring your oxygen saturations (with a probe on your finger) and looking for any noticeable swelling.
- 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.
- 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 at the shape of the heart.

In some cases the following tests might also be done to help with reaching a 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 ruleout 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.

In conclusion, PPCM may be diagnosed when:

- symptoms of heart failure are experienced
- the heart's ejection fraction is 45% or lower
- the heart may be enlarged (although not always)
- 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 personalised to the individual, taking into account whether or not they are still pregnant. There are specific guidelines for the treatment of heart failure and these include medications such as:

- diuretics (water tablets) to reduce the build-up of any fluid on the lungs or the ankles by encouraging the kidneys to 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) 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.
- Anticoagulants (blood thinners) to prevent dangerous blood clots from forming.

Finding the right medical therapy for PPCM is a shared decision, taking into account individual circumstances - including whether the mother is still pregnant and whether she wishes to breastfeed her baby.

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 type of arrhythmia they experience.

♥ See our factsheets on arrhythmias.



www.cardiomyopathy.org Helpline 0800 018 1024 Mon-Fri 8.30am-4.30pm (Free from a landline, mobile costs vary) Although it is very rare, PPCM can be lifethreatening and needs to be treated promptly. The earlier PPCM is diagnosed and treatment is started, the better the woman's chances of making a good recovery. If the mother does not recover well, she may need ongoing treatment. The drug therapies for PPCM can be very effective, but other treatment options may be considered according to individual needs. For example, a small number of women affected may require a mechanical support device such as LVAD (Left Ventricular Assist Device) and possibly eventually be considered for a heart transplant.

Because pregnancy is a time of changes within the body, a variety of specialist healthcare professionals will work together to ensure the woman receives the best care. This is known as a multi-disciplinary team (MDT) approach. The MDT may include: cardiologists, obstetricians, obstetric physicians, midwives and specialist nurses (amongst others). Each member of the team will discuss the options available and work collaboratively to help the individual to be involved in making choices about their care.

Does PPCM affect pregnancy and delivery?

PPCM is often not diagnosed until after delivery, however signs and symptoms may have been present during the pregnancy. It is important that symptoms are recognised as early as possible and a diagnosis made, so that appropriate treatment can start at an early stage to prevent the condition from worsening. If PPCM is diagnosed during pregnancy, the MDT will decide on the most appropriate plan for delivery according to individual circumstances. The mother's wishes are a key consideration and will always be respected. It is usually possible to plan for a vaginal delivery with PPCM, however there may be instances in which a Caesarean birth is recommended. If the woman's condition deteriorates during pregnancy, it may be necessary to deliver the baby early, before the expected date of delivery. If the baby needs to be delivered preterm, a paediatrician (baby doctor) will explain how the baby needs to be looked after once it's born. This may involve the baby being admitted to the neonatal intensive care unit for a while.

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 a small number of women, recovery can take much longer due to the severity of their symptoms and the ability of their heart to recover.

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

If a woman has had PPCM during pregnancy, research suggests that she has an increased chance of getting it again in future pregnancies, even if she recovers completely. Pregnancy followed by PPCM therefore needs to be carefully planned and prevented until the heart has sufficiently recovered to be able to tolerate another pregnancy. A discussion about appropriate contraception will be part of the postnatal care, so that women with a history of PPCM are protected from the potential risks of an unplanned pregnancy. Becoming pregnant again before the heart has properly recovered can worsen the mother's health and can be potentially life-threatening. Very occasionally, in severe cases, where an unplanned pregnancy threatens the health of the mother early in pregnancy, the medical team may advise a termination of pregnancy in order to protect the mother. For this reason it is strongly advised to use reliable contraception to prevent pregnancy occurring before the heart is fully recovered.

Women who are considering another pregnancy should be referred for pre-pregnancy counselling with a specialist cardiologist and/or obstetrician/ obstetric physician with a background in cardiac disease in pregnancy. This will help to ensure that all aspects of care can be considered in advance and future pregnancies are carefully planned and managed to keep the mother and baby safe.

This information sheet 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 on our website.

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