Peripartum cardiomyopathy

Peripartum cardiomyopathy (or postpartum cardiomyopathy) is defined as the development of heart failure in the last month of pregnancy or first five months after delivery in the absence of any other cause. The heart’s ejection fraction (the volume of blood pumped out with each beat) is less than 40 per cent. (In a normal heart the volume would be 55 to 60 per cent).

PPCM is often difficult to diagnose because the symptoms, including breathlessness, exercise intolerance, cough and shortness of breath when lying flat, may mimic common symptoms in normal pregnancy. Diagnosis relies on the medical teams looking after pregnant women and new mothers being aware of the condition and that worsening breathlessness and excessive fluid retention are not normal in pregnancy. If PPCM is suspected, the mother should be immediately referred to a cardiologist.

There is evidence that PPCM is becoming more common, perhaps due to more awareness of the disease, the increasing age of mothers, and more multiple pregnancies.

Causes

The exact cause for PPCM is unknown but various factors have been implicated. These include the body’s immune responses being directed against its own tissues, inflammation in the heart muscle, genetic abnormalities in the baby, an increased rate of heart cell death, and having dilated cardiomyopathy in the family.

However, evidence for and against heart muscle inflammation being a cause remains uncertain. Dietary deficiencies in selenium and other micronutrients might also play a role. Reduced selenium increases susceptibility to viral infections in the heart, high blood pressure and an abnormally low level of calcium in the body.

Women with PPCM also have gene mutations associated with inherited dilated cardiomyopathy, suggesting there may be an overlap in the two diseases.

High levels of prolactin, a hormone that stimulates milk production in women, might also trigger PPCM. Using drugs to suppress prolactin production has prevented the disease developing in animals.

Symptoms

Symptoms of PPCM are breathlessness, palpitations, fluid retention, stomach pain, cough, a fall in exercise tolerance, and lethargy. They all require further investigation, particularly if symptoms are severe or getting worse.

Diagnosis

Women with PPCM are likely to have signs of heart failure. They may be breathing rapidly and have a fast heart rhythm. They may have low levels of oxygen in their blood and raised central venous pressure (the amount of blood returning to the heart and the ability of the heart to pump the blood into the arteries). Extra fluid may or may not be in the lungs. Normal pregnancy increases the risk of...
blood clots, but up to 30 per cent of women with PPCM may experience them. With their hearts already not pumping properly, they are vulnerable to blood clots in the lungs and left ventricle.

Tests for peripartum cardiomyopathy

**Lab tests**

Blood tests should be done to check potassium levels and liver and thyroid function. Other blood tests (troponin and B-type natriuretic peptide) should be done to detect heart problems but these may be normal. Urine should be checked to exclude pre-eclampsia (another pregnancy related condition) and a urine infection. Other causes of cardiomyopathy should also be ruled out.

**ECG**

An ECG will commonly show abnormalities, including a fast heartbeat, and there may be an underlying irregular rhythm.

**X-rays**

A chest x-ray will be performed to look for fluid in the lungs. With the use of an abdominal shield, x-rays are safe as the radiation dose to the baby is negligible.

**Echocardiogram**

An echo takes images of the heart through the chest wall. This provides a non-invasive, accurate and quick assessment of the overall health of the heart, including how it is pumping, the health of the valves and whether there are any blood clots.

**Magnetic resonance imaging (MRI)**

An MRI provides more information about how the heart is working and can look for heart inflammation, heart cell death and blood clots in the left ventricle. But this may not be feasible, particularly for women in advanced pregnancy.

**Angiogram**

A coronary angiogram may be done to rule out coronary artery disease or coronary artery dissection, but may be judged not to be suitable during pregnancy.

**Acute treatment**

In PPCM, care must focus on the mother, not the baby. Treatment should not be restricted because of potential harm to the baby. Care should be organised by a multidisciplinary team, including a cardiologist, obstetrician, midwife and a paediatrician with specialist knowledge looking after critically ill premature or full term babies.

Care is similar to that used in conventional heart failure. Excess fluid must be reduced using drugs such as diuretics which increase urine production. Oxygen levels in the blood should be kept above 95 per cent.
Drugs may be needed to maintain blood pressure in critically ill mothers. They should be on anti-clotting drugs to prevent blood clot complications. Low molecular weight heparin is used for pregnant women and after the birth warfarin for at least up to six weeks. Warfarin is continued long-term if the mother’s ejection fraction remains below 30 per cent. Beta-blockers, such as metoprolol or a similar type, should be started.

When PPCM is first diagnosed, decisions on continuing the pregnancy are based on the mother’s state of health, the degree of heart failure and how far the pregnancy has progressed. Where possible, pregnancy should continue until the outlook for the baby is good. But this cannot be at the expense of the mother’s life. The decision to end a pregnancy before the baby is viable, or to deliver prematurely is a decision for the mother and the multidisciplinary team.

How the baby is delivered is affected by the health of the mother and how near to full term the baby is. Vaginal delivery may be possible but an emergency c-section may be needed if the mother is critically ill.

Treatment after delivery

Drugs not suitable to be taken during pregnancy, such as ACE inhibitors, can now be safely given, including to breast feeding mothers. Diuretics, warfarin and beta-blockers are also safe. The preferred beta-blockers are propranolol and metoprolol which are excreted in small quantities. Others such as nadolol and sotalol are excreted in higher amounts which can lead to low blood pressure, slow heart rate and rapid breathing in the baby.

Prognosis

Prognosis depends on the mother’s recovery from heart failure, which is defined as having an ejection fraction of 50 per cent or more or the figure improving by 20 per cent or more. Recovery is usually between three and six months after the birth, but might be as long as four years afterwards.

Most women make a good recovery, and some recover completely.

Delayed diagnosis, poor heart function, black ethnicity, a blood clot in the left ventricle, multiple births and having other illnesses may make recovery slower. Mothers who show little sign of recovery often need a heart transplant.

Follow up echos should be performed, as well as pre-discharge scans. Six weeks and six monthly echos are advised afterwards.

Subsequent pregnancies

Even after a complete recovery from PPCM the risk of it recurring in future pregnancies remains high and heart failure can worsen again.

But having an ejection fraction of 55 per cent or more seemed to be the most important factor for avoiding a relapse in a post-PPCM pregnancy, one recent study showed.

It is difficult to predict how mothers with PPCM will do in a subsequent pregnancy. Much of the medical literature advises against future pregnancies. However, recurrence is unpredictable.
If a woman accepts a high risk and has another pregnancy, close monitoring of heart function (every four to six weeks) is crucial to detect problems early on. There is a risk of relapse which is higher in women who have had persistent heart failure problems before their SSP. There is also an increased risk of premature birth and terminations. Full recovery of heart function before the SSP is associated with better prognosis. However, an uneventful pregnancy is not guaranteed, and approximately 20 per cent of mothers will have a relapse of PPCM and a substantial decrease in heart function.

Relapse of PPCM may be associated with heart function deterioration, heart failure and irregular heart rhythms. It needs aggressive treatment, including the use of temporary and permanent heart devices. It may also have detrimental long-term consequences.

Conclusion

PPCM results in heart function problems that can sometimes continue to affect women’s lives. It remains a poorly understood, rare disease with symptoms that can easily be attributable to normal pregnancy. More awareness is vital for early diagnosis and prompt treatment by a specialist multidisciplinary team. Careful assessment of symptoms, early investigation and rapid treatment remain vital.