

Peripartum Cardiomyopathy: Rare Albeit Significant

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Rec date: June 27, 2016; Acc date: June 29, 2016; Pub date: June 30, 2016

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Peripartum Cardiomyopathy

Peripartum cardiomyopathy is not a common disorder with an approximate incidence of 1 per 2500 to 4000 live births in western countries, more common in West Africa and Haiti [1-4]. It presents in the later part of pregnancy or up to first few months of the postpartum period. It is the cardiomyopathy of dilated type in which the heart musculature weakens and the amount of blood ejected from the left ventricle with each contraction decreases. Hence, it is difficult for the heart to cope up with the body's demand for oxygen, thereby affecting various organs.

Normal Physiological Changes during Pregnancy

Understanding physiological changes of pregnancy is important for the timely recognition of cardiovascular pathologies. Haemodynamic changes begin right in the initial months of pregnancy. Total blood volume increases by more than 40%, heart beat increase from baseline of 75 bpm to nearly 90 in the third trimester. There is significant decrease in the systemic vascular resistance and a reverse proportionate increase in cardiac output [5]. Stress of labour aggravates these changes to maximum and then returns to normal by about 42 days after delivery. The risk of thrombosis is 4-10 folds higher around pregnancy [6].

Diagnosing Peripartum Cardiomyopathy

More or less the symptoms replicate those of last trimester of pregnancy; however an ECG can detect diminished cardiac functioning. It is suspected when the following three conditions are met:

1. Cardiac failure develops in the last month of pregnancy or within first few months of delivery.
2. Lower than 45% of Ejection fraction
3. No other cause with reduced ejection fraction can be found [7].

Causes

Still unclear, heart biopsies have shown inflammation in 10-75% of cases. Others include poor diet, vasospasm of coronary artery, disease involving small-vessel, and defective antioxidant defense [1]. Genetics may have a role.

Risk Factors

- Elderly gravida (more than 30)
- Multiparty
- Multiple pregnancies

- African origin
- Hypertension
- Earlier exposure to toxin (cocaine), smoking
- Drugs to prevent preterm labour

Symptoms

Major symptoms are those of cardiac failure including fatigue, feeling of heart racing (palpitations), skipped beats, light headedness, or syncope, breathlessness, cough, chest pain, nocturia, abdominal pain and diminished appetite. Fluid retention may manifest itself in form of swelling in legs, abdominal bloating, chest congestion and weight gain. Rarely presentation may be of stroke or heart attack due to the blood clot clogging the vital organs.

Evaluation

Starts from seeking history and general examination, to uncover any underlying heart disease, assessing evidence of diminishing heart function and detecting complications. Investigations to assess hepatic, renal and thyroid functions, electrolytes, complete blood count, troponin and inflammatory markers [8]. Tests specific for PPCM include:

X-ray chest

To look for any increase in the size of heart and pulmonary oedema.

CT scan of chest

For evidence of any blood clots in the lungs.

Electrocardiogram

For heart beat count and regularity, electric conduction and to negate any possibility of heart attack.

Echocardiogram

For knowing the heart size and function.

Coronary angiography along with Cardiac catheterization

For any possibility of blockages or dissection of the coronary arteries

Other imaging studies for inflammation or scarring

MRI and nuclear scans.

Management

Provides a challenging situation for the treating physician. Medications used are to properly sustain heart function, improvement in blood circulation, managing fluid excess, to prevent and treat

complications. Presently, Peripartum Cardiomyopathy is managed concurrent with the guidelines issued by the ESC for cardiac failure in pregnancy [8]. Choice of medication is based upon whether PPCM is ante- or post- partum. Beta-blockers, thiazide diuretics, or furosemide may be a requirement in certain cases of pregnancy that may impair placental perfusion; however, lowest possible dosage should be used. Post- partum, recommendation is of standard therapy for cardiac failure that includes- beta-blockers, ACE-inhibitors/AT1-blockers, mineralocorticoid receptor antagonists and diuretics [9]. Infusion of an inotrope is a recommendation exclusively when there is significant low blood pressure.

Medications to treat PPCM

Vasodilators: For relaxation of blood vessels, so that it becomes easier for heart to pump blood.

Hydralazine is the drug of choice during pregnancy. After delivery ACE-inhibitors/AT1-blockers can be used safely.

Diuretics: For renal excretion of excess salt and water so as to relieve symptoms related to fluid overload.

Beta-blockers: To counter the role of catecholamines and thus reduce heart rate [10] and hypertension. Over time they help to heal and recover normal ejection fraction.

Digitalis: Strengthens pumping ability of heart and is safe during and after pregnancy. Spironolactone: mild diuretic that retains potassium and helps the heart to heal.

Antiarrhythmics: To maintain near normal heart rate and rhythm. During pregnancy beta blockers, sotalol and procainamide by intravenous route may be helpful. Amiodarone can be given during gestation or post-delivery, but it may be harmful to fetus. Severe symptoms may necessitate enthusiastic therapy in a background of intensive care facilities. For patients non-responsive to such management, support with pump of balloon or temporary cardiac pump is required.

Delivery

Mode of delivery for patients with PPCM is generally based on obstetric indications. The advantages for vaginal delivery are minimal blood loss, greater haemodynamic stability, avoidance of surgical stress, and less chance of post-operative infection and pulmonary complications. Effective pain management is a necessity to avoid further increase in cardiac output from pain and anxiety. Caesarean section is reserved for the indications such as fetal distress, failure to progress and other absolute indications. Delivery may not be hastened if both the mother and fetus are uncompromised. Complications may include preterm, intrauterine growth restriction and malformations. Patient may choose to breast-feed after delivery as most drugs are safe in this period.

Anesthesia

Regional anesthesia with continuous epidural is preferred. It prevents the complications of general anesthesia. Decreases after load, thus reducing the amount of work heart has to do. Whereas with combined spinal-epidural there's a risk of fall in blood pressure. During general anesthesia, if at all required, agents that preserve left ventricular contractility without significant vasodilatation are helpful and dreadful changes in pre- and after- load should be avoided.

Secondly, agents that cause tachycardia or increase contractility of the heart have to be shunned or used with caution.

Recurrence

These women are at increased probability of having the same condition in future conceptions. If the ejection fraction is stable at rest and changes with stress, the possibility of recurrence appears to be lower. 50% regain stable cardiac function, 25% have persistently diminished function but remain stable on treatment and rest 25% deteriorate to severe cardiac failure. If complete cardiac function is not regained another conception is generally not recommended. This can significantly damage the heart and may prove to be detrimental for the mother as also for the developing fetus.

And, if there is complete recovery of the cardiac function, another conception may be allowed but under careful supervision [11]. If women choose to move ahead, drugs that are potentially harmful or dangerous to the developing fetus should be discontinued and serial and meticulous monitoring becomes critical. When to discontinue treatment remains controversial but at least not before 1 year of stabilization of ejection fraction.

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