

# PERIPARTUM CARDIOMYOPATHY

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Peripartum cardiomyopathy is defined as the onset of acute heart failure without demonstrable cause in the last trimester of pregnancy or within the first 5 months after delivery. It is a form of Dilated Cardiomyopathy with left ventricular systolic dysfunction that results in signs and symptoms of heart failure. It is often unrecognized, as symptoms of normal pregnancy commonly mimic those of mild heart failure.

The National Heart, Lung and Blood Institute and the Office of rare diseases (1997) have established the following diagnostic criteria for Peripartum cardiomyopathy:

- 1 Development of Cardiac failure in the last month of pregnancy or within 5 month after delivery.
- 2 Absence of an identifiable cause for the cardiac failure.
- 3 Absence of recognizable heart disease prior to the last month of pregnancy.
- 4 Left ventricular systolic dysfunction demonstrated by classic echo cardio Graphic criteria such as depressed shortening fraction or ejection fraction.

The reported incidence in the west ranges from 1 in 4000 deliveries, representing less than 1% of cardiovascular problems associated with pregnancy. Sixty percent present within the first 2 months postpartum but up to 7% may present in the last trimester of pregnancy. Geographic variations exist with a higher incidence reported in areas of Africa because of malnutrition and local customs in the puerperium.

Etiology is still unknown. It may be postulated that it may be due to nutritional deficiencies, small vessel coronary artery abnormality, hormonal effects, toxemia, maternal immunologic response to fetal antigen or myocarditis.

Predisposing factors include maternal age greater than 30 yr, multiparous or eclamptic patients, twinning, racial origin (black), hypertension and nutritional deficiencies. In the majority of cases there is no family history.

Peripartum cardiomyopathy usually presents with symptoms of worsening cardiac failure. These include dyspnoea on exertion, fatigue, ankle oedema, embolic phenomena, atypical chest pains and haemoptysis.

Examination may reveal evidence of a raised CVP, tachycardia, cardiomegaly with a gallop rhythm (S3), mitral regurgitation, pulmonary crackles and peripheral oedema.

Chest radiographs may show cardiomegaly with pulmonary oedema and pulmonary venous congestion.

The electrocardiogram may show nonspecific ST and T wave changes, atrial or ventricular arrhythmias and conduction defects.

Echocardiography / Doppler examination may reveal enlargement of all four chambers with marked reduction in left ventricular systolic function, small to moderate pericardial effusion and mitral, tricuspid and pulmonary regurgitation may be evident. Ventricular wall motion, ejection fraction and cardiac output are decreased and pulmonary wedge pressure is increased.

Symptoms like decreased exercise capacity, tiredness, dyspnoea, orthopnoea and palpitations may occur even in normal pregnancy and can be mistaken for a diseased state.

Physical signs like hyperventilation, edema, and distended neck veins can be seen in normal pregnancy.

On auscultation of the heart, one can hear loud first heart sound, exaggerated splitting, mid systolic murmur and continuous venous hum. These physical signs may confuse the anesthesiologist and there could be mistakes in the form of over diagnosis or disregarding of heart disease.

The clinical presentation and hemodynamic features in PPCM are indistinguishable from those of other forms of dilated cardiomyopathy. In the absence of any cardiac symptoms, one of the early indications about this condition is revealed during evaluation of the fetus with a fetal monitor and ultrasound. Since fetal growth is dependent on good blood flow to the uterus and placenta, an insufficient blood flow, show signs of decreased oxygenation by slowed growth. This might prompt further investigation to discover heart disease.

The clinical course of PPCM varies with approximately 50-60% patients showing complete or near complete recovery of clinical status and cardiac function, usually within the first 6 months postpartum. The remaining patients demonstrate either continued clinical deterioration leading to early death or persistent left ventricular dysfunction and chronic heart failure. There appears to be an initial high risk period with mortality of 25-50% in the first 3 months postpartum. Patients with persistent cardiomegaly at 6 months have a reported mortality of 85% at 5 years.

Subsequent pregnancies in women with PPCM are often associated with relapses and high risk for maternal morbidity and mortality. For this reason subsequent pregnancy should be discouraged in women with PPCM who have persistent cardiac dysfunction. Management of PPCM should include vigorous treatment of acute heart failure.

- 1 Oxygen, diuretics, digoxin and vasodilators.
- 2 Use of ACE inhibitors in early pregnancy should be avoided as it has teratogenic effects on fetus
- 3 Anticoagulant therapy is recommended because of high incidence of thrombo embolic events in PPCM. Patient on oral anticoagulants require change to parenteral anticoagulants with short half life and the dose adjusted according to the PTT which may be discontinued before delivery. After delivery Warfarin may be used.
- 4 Since the disease may be reversible, the temporary use of intra aortic balloon pump or LV assist device may help to stabilize the patient's condition pending improvement.

Dobutamine stress echocardiography may have a role in evaluating contractile reserve in women with recovered systolic function who are contemplating further pregnancies.

A high number of patients with PPCM show evidence of myocarditis in biopsy specimens. Autopsy shows cardiac enlargement, often with mural thrombi along with histological evidence of myocardial degeneration and fibrosis.

**The anaesthetic considerations** for a patient with heart failure presenting for caesarian section are similar regardless of etiology. Hemodynamic goals include maintenance of normal to low heart rate to decrease oxygen demand, and prevention of large swings in blood pressure. Achievement of these goals have been undertaken by giving general and regional anesthesia. During GA important factors to keep in mind are:

1. Volatile agents that decrease LV contractility without dramatic vasodilatation is desirable.
2. Avoid agents that decrease preload and after load, eg. hypovolemia, nitroglycerine, nitroprusside.
3. Avoid agents that directly or indirectly increase heart rate and contractility eg. Pancuronium, atropine, epinephrine, ephedrine.
4. Blood loss to be replaced promptly.
5. Hypotension better treated with volume expansion and pure alpha adrenergic

agonist.

6. Remember that insertion of CVP / PAC may induce atrial or ventricular dysarrhythmias.

General anaesthetic techniques, involve the use of either intravenous cardiodepressant drugs such as thiopentone and /or the inhalational anaesthetic agents such as Isoflurane, Sevoflurane or Desflurane or high dose narcotics, for maintaining haemodynamic stability. The latter technique may necessitate post operative ventilation for both mother and infant. Since the end point at induction of anaesthesia with narcotics is not well defined, there is an increased risk of gastric aspiration. The management of a failed intubation may become difficult by the longer acting nature of these drugs with mask ventilation. This may be compounded further if associated with obesity.

The consideration for central neuraxial anaesthesia in these patients are similar to those with other causes of heart failure. Subarachnoid block may better be avoided in these patients because of sudden onset of haemodynamic instability. Epidural anaesthesia may be a better choice particularly when incremental doses of local anaesthetic are administered along with opioids. The gradual and controlled induction of anaesthesia, may improve myocardial performance and the cardiac output by decreasing the systemic vascular resistance, thus reducing the afterload on the left ventricle without impairing contractility. The presence of a pulmonary artery catheter can guide fluid and inotrope requirements, with minimal change in haemodynamic parameters. These women do not need additional volume before induction of central neuraxial block and preloading to be avoided..

Small bolus doses of 0.5% bupivacaine or 2.0% xylocaine ( 10 to 12 ml in L2 to L4) along with fentanyl upto 40 mics may be preferred.

Intra operative monitoring depends on the preoperative signs and symptoms. If cardiomyopathy is asymptomatic, a central venous catheter is adequate with non invasive BP monitoring. In symptomatic patients or with echo findings of left ventricular dysfunction, a PA catheter and an arterial line if available will be useful.

Oxytocin infusion is preferable since as an infusion it will not produce sudden vasodilatation and hypotension. It also helps to decrease the after load maintaining the hemodynamic stability.

It is better to monitor these patients in an ICU for post operative hemodynamic stabilization. Patient's hemodynamic stability may worsen due to retention of water due to antidiuretic effect of Oxytocin and re absorption of third space fluid after 48 hrs of the caesarian section, may increase the preload, worsening the patient's condition.

These women may develop a reduction in the left ventricular systolic function during subsequent pregnancies. This reduction would be greater in those with persistent left ventricular dysfunction at the start of the pregnancies. Symptoms of heart disease develop in about 20% of women whose systolic function is normal at the start of the subsequent pregnancy and in almost half of the women who have persistent left ventricular dysfunction.

The outcome of these patients with peripartum cardiomyopathy is highly variable. In some clinical and echocardiographic status improves rapidly and returns to normal. The initial severity of the left ventricular systolic dysfunction or dilatation is not necessarily predictive of the long term functional outcome.

Some will have persistent cardiac dysfunction while few have a slow return to normal cardiac function over several years. These patients appear to have a better survival rate (94% at 5 years) than patients with cardiomyopathy due to other causes

It is advisable to avoid subsequent pregnancies in these patients if possible to avoid possible threat to life for them if it occurs.