ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 6 | June - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PERIPARTUM CARDIOMYOPATHY

Cardiology

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KEYWORDS

INTRODUCTION

Since the 18th century, peripartum cardiomyopathy (PPCM) has been recognized as a serious complication of pregnancy. History dates back to 1849, when Ritchie reported heart failure (HF) in late pregnancy and late puerperium1. Myocardial degeneration as a cause of death during puerperium was reported by Virchow and Porak in 1870.2 The disease was forgotten for almost 60 years. In 1930, PCM was described as a separate clinical entity. In 1971, Demakis et al. published a series of 27 cases of PPCM and defined the criteria for diagnosis of PPCM.3

DEFINITION

Peripartum cardiomyopathy was initially defined as the development of HF within the last month of absence of any identifiable cause for HF, and any recognizable heart disease before the last month o The National Heart , Lung and Blood Institue (NHLBI) and National Institute of Health (NIH) of United States, in April 19974 added echocardiographic criteria to the above definition as below: Depressed left ventricular shortening fraction (LVSF) less than 30% Or Depressed left ventricular ejection fraction (LVEF) less than 45 The most recent definition comes from Heart Failure Assosiation of European Society of Cardiology Working Group on PPCM in 2010 who felt that time frame of PPCM is arbitrary an d may lead to under diagnosis of PPCM and proposed a simplified definition - Peripartum cardiomyopathy is an idiopathic cardiomyopathy presenting with heart failure secondary to left ventricular systolic dysfunction towards the end of pregnancy or in the months following delivery, where no other cause of heart failure is found. It is a diagnosis of exclusion. The left ventricle may not be dilated but the LVEF is nearly always reduced below 45%." The definition has broadened the scope of disease and has taken into account similar presentation in early pregnancy.[4]

EPIDEMIOLOGY

The incidence of PPPCM higher in developing countries as compared to the developed nations. Incidence is lowest in Japan, being 1:6,000 live births and highest in Nigeria5 being 1%. In India6 the incidence is 1:1,374 live births.

NOMENCLATURE

Ever since the description of the disease since 1930s, various names have been given to this condition such as "toxic postpartum heart disease", "postpartum heart failure", "postpartum cardiomyopathy", "postpartum heart disease",

"postpartum myocardits", etc. However, PPCM is the most preferred term as it encompasses the whole spectrum of the disease.7

RISK FACTORS

Although the aetiology of peripartum cardiomyopathy remains unclear, the following are among the factors associated with increased risk of peripartum cardiomyopathy:

- Multiple pregnancy
- Multiparous women
- Women older than 30 years
- Black women
- Women with gestational hypertension or preeclampsia
- Women treated with tocolytic theray-Long term oral tocolytic tyerapy (more than 4 weeks therapy) with beta-adrenergic agonist such as terbutalinePrevious history of peripartum cardiomyopathy
- Maternal cocaine abuse
- Selenium deficiency: Recent case-control study of 39 patients in Nigeria revealed that levels of selenium in peripartum cardiomyopathy were half of that in control (P = .001)
- MHigher incidence in African countries has been attributed to the post-partum high-salt consumption of kanwa, though not validated in subsequent studies US Nationwide Inpatient Sample from 2004 to 2011, a sudy of total of 1337 peripartum cardiomyopathy cases suggested new associated causes8:

- Smoking (OR 33.6; 95% CI 9.3-157.4; P < .0001)
- Anaemia (OR 2.0; 95% CI 1.6-2.5; P < .0001)
- Asthama (OR 2.2; 95% CI 1.5-3.2; P = .0002)
- Thyroid disease (OR 5.9; 95% CI 1.5-2.13; P = .01)

ETIOPATHOGENESIS

Though the disease is present for more than one and a half century, still the etiology is obscure.9 It was thought to be idiopathic dilated cardiomyopathy (IIDM) unmasked by the hemodynamic stress of pregnancy. However, it has been refuted because of the following reasons:

- Maximum hemodynamic stress of pregnancy is in 2nd trimester and PPCM presents in the last trimester.
- Complete recovery occours in approximately 30% of the patients with partial recovery in many. Recovery in IDCM is rare.
- PPCM occurs in young women, while IDCM occurs late.

Peripartum cardiomyopathy is a distinct clinical entity and various hypotheses have been proposed for its causation.

- Myocarditis hypothesis: Myocarditis as a cuase of PPCM in humans seems to be a most probable factor. Various authors have reported enlarged hearts with focal areas of necrosis and fibrosis consistent with myocarditis in women dying of heart failure in the puerperium.10 Pregnancy is immunocompromised state and there is likelihood of increased chances ofviral replication. Treatment with prednisone and azathioprine resulted in clinical improvement. However, prevelance of myocarditis has been reported from none to 100% and the variability in the prevelance may be because of so many reasons – geographical variability, various inclusions criteria, difficulty in performing endomyocardial biopsy and timing of endomyocardial biopsy.

- Hemodynamic stress of pregnancy hypothesis: There is considerable increase in cardiac output in first trimester (22%) and further increase in 2nd and 3rd trimester of pregnancy.There is drop in systemic vascular resistance, hence, pregnancy is high volume , low resistance state. However, there is no convincing data to support this hypothesis as a cause of PPCM.

- Abnormal immune response to pregnancy hypothesis: Fetal cells may escape in maternal circulation and may reside in cardiac tissue and initiate an autoimmune response at an appropriate time. Autoantibodies have been found in more than 50% of PPCM patients and this hypothesis may explain increased incidence of PPCM in subsequent pregnancies.

- Increased myocyte apoptosis hypothesis: Fas and Fas ligand are the cell surface proteins and play key role in apoptosis,

programmed cell death. They are significantly elevated in patients with PPCM as compared to normal volunteers.[1]

- Inflammation hypothesis: Inflammatory markers like hs-CRP nave been found to be elevated in patients of PPCM. Cytokines, the hormone-like molecules tumor necrosis factor-alpa (TNF-α), interleukin-1 (IL-1), interleukin-6 (IL-6), interferon-gamma (IFN-γ) play an important role in inflammation and have found to be more in sera of patients of PPCM. These proinflammatory cytokines at higher concentrations result in lef ventricular dysfunction, pulmonary edema, left ventricular remodelin and cardiomyopathy. Drugs like ACE inhibitors, pentoxifylline, statins and IV immunoglobulins result in decreasing the levels of circulating cytokines and have beneficial therapeutic effects in patients of PPCM. However, casual link between cytokines and PPCM needs to be established.12

- Genetic hypothesis: There are several reports of clustering of PPCM cases in families suggesting genetic link of disease.

- Malnutrition hypothesis: Malnutrition was thought to be one of the factors in the etiopathogenesis of PPCM as the disease is more prevelant in developing nations. Occurrence of PPCM in well nourished patients led this hypothesis in doubt. Selenium deficiency in PPCM patients has reported from Sahel region of Africa.13 Excess salt consumption is a known cause o P PCM. In Nigeria, pregnant women ingest kanwa; a dried lake salt while lying on heated mud beds and has found to have the highest incidence of PPCM worldwide.14

- Prolactin hypothesis: Association of prolactin and increased blood volume, reduced renal excretion of water, sodium and potassium and increased levels of erythropoetins leading to increased hematocrit exist.15 Serum prolactin levels are elevated in patients of PPCM. Patients treated with bromocriptine resulted decrease in prolactin levels and significant improvement in patients' status.16 Bromocriptine has serious adverse effects when used in pregnant women to suppress lactation and have been withdrawn. Use of bromocriptine in patients of PPCM has not been established.

- Myocardial ishchemia hypothesis: Though some authors have suggested myocardial ischemia as the possible cause for PPCM, however, coronary arteries have been found to be normal, refuting this hypothesis.

- Heightened adrenergic tone hypothesis: Heightened adrenergic tone has been found in pregnancy that can lead to fluid overload and transient LV dysfunction. Beta blockers (β-blockers) have been used in management of PPCM. Role of β-1 adrenergic receptor antibodies has also been considered in the pathogenesis of PPCM.17

- Hormonal hypothesis: Relaxin, an ovarian hormone, produced during pregnancy has been implicated in the pathogenesis of PPCM which can cause excessive relaxation of cardiac musculature. However, no convincing evidence has been found to prove the hypothesis.18

CLINICAL FEATURES

Patient present with dyspnea, orthopnea, paroxysmal nocturnal dyspnea, cough and hemoptysis. Other symptoms are faigue, paipitation, nonspecific chest pain abdominal discomfort and postural hypotension. Most of the patients are in New York Heart Association (NYHA) class III or IV. Signs include features of cardiomegaly and biventricular failure in form of displaced apical impulse, evidence of mitral and tricuspid regurgitation, presence of S3,basal crackles, raised jugular venous pressure (JVP), pedal edema and hepatomegaly.

DIAGNOSIS

Diagnosis is often delayed as symptoms are not specific. Following laboratory tests are necessary, although they are not specific for peripartum cardiomyopathy:

- Complete blood count
- Blood urea creatinine electrolyte
- Liver function test
- Thyriod stimulating hormone leve
- Cardiac troponi
- Brain natriuretic peptide (BNP)
- Chest radiograph
- Electrocardiogram
- Transthoracic echocardiography
- Cardiac magnetic resonance imaging

Some biomarkers found to be specific for peripartum cardiomyopathy in recent experimental studies include19:

- The 16 kDa prolactin
- MicroRNA-146a
- Soluble Fms-like tyrosine kinase 1

Their diagnostic value in clinical practice, however, needs verification. In some cases, endomyocardial biopsy is warrented to exclude inflammatory aetiology of acute heart failure.

BNP or N-terminal pro-brain natriuretic peptide (NT-proBNP) plasma concentration has high sensitivity to include and high specificity to preclude heart failure. Despite significant hemodynamic stress, the concentration of BNP or NT-pro-BNP does not increase during normal pregnancy.

Normal BNP levels (<100 mcg/ml) show a very high negative predictive value for adverse maternal events in pregnant women with heart disease. NT-proBNP levels greater than 300 mcg/ml prior to pregnancy were associated with complications during peripartum period in women with dilated cardiomyopathy.20

INVESTIGATIONS

LABORATORY DIAGNOSIS

The laboratory tests are mostly normal, cardiac enzymes are usually not elevated, atrial and brain natriuretic peptides are elevated, while they not commonly rise in normal pregnancy. D-dimer will be elevated in patients with intracardiac thrombi.

No specific diagnostic biomarkers are there for diagnosing peripartum cardiomyopathy. MicroRNA, specifically miR-146a may serve as a noble biomarker, though more research is needed.17

ECG

The electrocardiogram usually shows sinus tachycardia often with multiple supraventricular or ventricular ectopics. While the ECG is not specific for PPCM, several findings including nonspecific ST-T wave abnormalities, QT interval prolongation, QRS widening, left ventricular hypertrophy and atrial fibrillation may be present. Arrhythmias are present in 18.7% of hospitalized PPCM patients. Ventricular tachycardia was most commonly seen in 4.2%, cardiac arrest was seen in 2.2%, electrocardioversion was done in 1.9%, permanent pacemaker implantation was done in 3.4% patients, ICD was implanted in 6.8% patients and the in-hospital mortality was three times more common in arrhythmia cohort (2.1% vs 0.7%).21

X-RAY

The chest radiograph shows enlarged heart and pulmonary congestion or frank pulmonary edema. The pleural effusions are frequent but small. It is important to remember fetal shielding and have informed consent prior to chest radiograph.

ECHOCARDIOGRAPHY

Ventricular dilation usually affects both the ventricles and marks generalized hypokinesia of myocardium; hypokinesia is found to be more local less often. Mitral regurgitation and tricuspid regurgitation are usually present. Apical thrombus is seen left ventricle and less often in both ventricles. Thrombi usually look shaggy, irregular and mobile in early cases.

Spontaneous echo contrast commonly shows up due to slow flow. All indices of contractility are reduced. A pericardial effusion is often present.

The original echocardiographic criteria for PPCM were strict left end-diastolic dimension more than 2.7 cm/m2 and impaired fractional shortening less than 30% or a left ventricular ejection fraction less than 45%. As per the latest guidelines for diagnonsis of PPCM, left ventricular dilatation is not a mandatory criterion.The common differential diagnosis like significant diastolic dysfunstion with

preserved systolic function as seen in preeclampsia can be excluded by echocardiography. Marked right ventricular dysfunction leads to pulmonary embolism and regional wall abnormality is seen in myocarditis or acute coronary syndrome.[22]

CARDIAC CATHETERIZATION

Raised pulmonary wedge pressure but normal or barely raised pulmonary artery pressure, raised left ventricular end- diastolic pressure and a normal coronary angiography are the findings on cardiac catheterization.

ENDOMYOCARDIAL BIOPSY

Endomyocardial biopsy should be taken from the right ventricle, only if it is free from thrombus. However, the biopsy results are uncertain and hence the current guidelines do not recommend them routinely. If the clinical course deteriorates with standard medical management, endomyocardial biopsy can be considerd to evaluate the presence of acute myocarditis or other forms of cardiomyopathy. Endomyocardial biopsy is recommended in the following cases:

1. Heart failure of less than 2 weeks duration with hemodynamic compromise
2. Heart failure of less than 3 months duration if associated with heart block, new ventricular arrhythmias
3. Refractory heart failure

CARDIAC MAGNETIC RESONANCE IMAGING (MRI)

The late gadolinium enhancement–cardiac MRI noted in 5% women at baseline; most women demonstrated significant improvement at 6 months consistent with low prevelance of late gadolinium enhancement. High prevelance of late gadolinium enhancement occoured in 71% of preipartum cardiomyopathy patient. The late gadolinium enhancement can occour even when ejction fraction is preserved. The late gadolinium enhancement is generally associated with persistent myocardial dysfunction despite medical therapy, arrhythmias and mortality. The late gadolinium detects foci irreversible myocardial damage manifested by enhancement regardless of mechanism or time of injury.[23]

The cardiac MRI complements echocardiography particularly in patients with suboptimal echocardiography. Global and segmental left ventricular contraction can be analysed to differentiate from acute myocarditis or ischemic heart disease.

As the gadolinium can cross the placenta, the benefit to risk ratio should be determined before subjecting to cardiac MRI with gadolinium enhancement.

DIFFERENTIAL DIAGNOSIS

Peripartum cardiomyopathy needs to be differentiated from certain pre-existing conditions such as:

- Dilated cardiomyopathy or other forms of cardiomyopathy
- Valvular diseases particularly valvular stenosisCongenital heart disease(preeclampsia and eclampsia)
- Acute myocarditis
- Acute pulmonary embolism
- Thyrotoxicosis
- Maternal sepsis
- Takotsubo syndrome
- Acute coronary spasm, dissection, thrombosis, myocardial infarction

DILATED CARDIOMYOPATHY

Previous familial and genetic studies have noted significant overlap in dilated cardiomyopathy and peripartum cardiomypathy. Peripartum cardiopathy resembles dilated cardiomyopathy in clinical presentation and cardiac imaging. Peripartum cardiomyopathy is a different disease and is not reversible form of dilated cardiomyopathy.Time frame is important as the dilated cardiomyopathy heart failure develops in the second trimester and the hemodynamic stress increases rapidly. Typical form peripartum cardiomyopathy develops early postpartum.

ACUTE MYOCARDITS

Viral and other types of myocarditis can occour during the peripartum period; they rapidly progress to acute heart failure mimicking peripartum cardiomyopathy. There is hesitation to endomyocardial biopsy because of low yield and the risks involved in pregnant women. Hence cardiac MRI is a good alternative method with high likelihood of diagnosis of acute myocarditis.

PREECLAMPSIA WITH HEART FAILURE

Severe hypertension in pregnancy including preeclampsia may cause a critical diastolic dysfunction and obvious heart failure. The pregnancy related volume overload precipitates cardiac decompensation hypertensive heart disease and preeclampsia. Echocardiography is used to differentiate the two diseases – left ventricular ejection fraction is lower in peripartum cardiomyopathy but is preserved in preeclampsia. Significant diastolic dysfunction and findings suggestive of elevated left atrial filling pressure indicate preeclampsia.

PULMONARY CUASES

Pneumonia may be facilitated by immune tolerance elicited during pregnancy and pulmonary embolism can result from the hypercoagulable peripartum period. Acute pulmonary edema can result from prolonged tocolysis or preeclampsia.

CARDIAC CAUSES

Myocardial infarction and Takotsubo cardiomyopathy are diagnosed by cardiac imaging modalities.

PRACTICAL MANAGEMENT OF PERIPARTUM CARDIOMYOPATHY

Only two RCTs consisting of total 44 patients have been reported; therefore, treatment is largely based on clinical experience and extrapolation fromdata with other forms of systolic heart failures for the drugs that can be safely used during pregnancy.24,25

- Intravenous immunoglobulin – Given intravenous globally shows important improvement compared with 11 historical control subjects; no further studies were conducted since 1999.
- Pentoxifylline – A 2002 South African study reported improved outcomes in 30 patients receiving pentoxifylline compared to 29 historical control subjects; no further studies conducted.
- A 2011 randomized control trial of 24 women with peripartum cardiomyopathy; use of calcium channel sensitzer showed no difference.
- Bromocriptine – The prolactin inhibitor is given 2.5 mg BID for 2 weeks, then 2.5 mg daily for 2 weeks. Bromocriptine still remains an experimental drug being tried in a pilot study and an on going trail. Anticoagulation is mandatory during Bromocriptine treatment.
- Anticoagulation is necessitated by the high risk of thrombo-embolism in patients with left ventricular ejection fraction below 35%.
- Referral for a ventricular assist device or cardiac transplant should be considered for the selected patients.
- Atrial fibrillation, though rare, is best treated by cardioversion under full heparinization and after exclusion of thrombus.
- For cardiac transplant, the peripartum cardiomyopathy patient should be top priority because of the following:
- Young age
- Needs of the newborn
- Possibly other young children at home

The contraindication for cardiac transplant is pulmonary infarction due to embolism; reinforcing full anticoagulation is must in every mother.

MANAGEMENT DURING PREGNANCY

- ACE inhibitors are contraindicated
- Loop diuretics
- Digoxin in small doses
- Beta blockers are the drug of choice and recommended for at least 6 months after full recovery; due to concerns regarding their influence on uterine tone, β-1 selective blockers are preferred
- A combination of hydralazine and nitrate is a safe alternative during pregnancy
- Anticoagulation with heparin

MANAGEMENT POSTPARTUM

- Loop diuretics
- Digoxin
- ACE inhibitor benazepril, captropril and enalapril are safe in nursing mothers. Angiotensin receptor blockers (ARB) are not to be used in nursing mothers.

- Beta blockers: For nursing mothers, metoprolol is the best suited agent.

- Amlodipine has been shown to improve survival in nonischemic cardiomyopathy, as plasma levels of IL-6 were reduced in amlodipine recipients in PRAISE trail.26

- Immunomodulation therapy with higher doses of immunoglobulins (2g/kg) in six women series was associated withb marked mprovement of left ventricular function. However, a randomized trial with acute cardiomyopathy did not prove efficacious. Currently there is no trail in peripartum cardiiomyopathy; therefore, status is unknown.27

- Mechanical circulatory support – In 10 patients between 2010 and 2015, with median age of 29 years and left ventricular ejection fraction of 10% at femoral extracorporeal membrane oxygenator (EMCO), outcomes suggest that an early stage peripheral mechanical circulatory offers safe and effective therapy for critical patients in the setting of cardiogenic shock to deteriorate despite maximal treatment. Central rescue cannulation has to be considered in patient unresponsive to peripheral support. Left ventricular assist devices have been used for 6 months with successful recovery in postpartum cardiomyopathy patients.28

MANAGEMENT OF DECOMPENSATED HEART FAILURE

- Optimal status of ABC (airway, breathing and circulation).

- Noninvasive ventilation with positive end-expiratory pressure of 5-7.5 cm H2O.

- Endotracheal intubation when needed.

- Supplementary oxygen to maintain arterial saturation greater than 95%.

- Monitoring with pulse oximetry is mandatory and frequent blood gas analysis is indicated until breathing is stabilized.

- Cardiac monitoring inclusive of heart rate and rhythm and blood pressure monitoring.

- Invasive hemodynamic monitoring in hemodynamically unstable peripartum cardiomyopathy patient is indicated which is central line for blood pressure monitoring to maintain a blood pressure more than 90-100 mmHg and central venous pressure line for pulmonary wedge pressure for refractory heart failure or with uncertain left ventricular filling pressures.

- Antepartum fetal heart rate monitoring to detect early stages of fetal distress.

- Intravenous administration of loop diuretics is often required to reduce cardiac preload and pulmonary edema while avoiding of utero-placental hypoperfusion.

- Intravenous vasodilation with use of nitroglycerine and hydralazine to reduce preload and afterload while watching for hypertension and utero-placental hypoperfusion.

- Intravenous nitroprusside is not recommended during pregnancy, as it causes fetal thiocyanate toxicity.

- Intravenous dobutamine is preferred in low systolic blood pressure; it can cause sinus tachycardia and myocardial ischemia.

- Milrinone can increase inotropy in cardiomyocytes and induces vasorelaxation in vascular smooth muscles. It is appropriate where systolic blood pressure is more than 90 mm Hg and with concurrent beta blocker treatment. Its recommendation is unclear because of higher risk of mortality.

- Severe cases of refractory heart failure may need mechanical cardiovascular support in form of intra-aortic balloon pump, extracorporeal oxygenation, cardiac transplant and left ventricular assist devices.

- Delivery should be managed in high-risk care setting by a multidisciplinary team; timing and mode of delivery is chosen based maternal hemodynamic stability, obstetric indications including fetal condition and the women or couples' wishes. Stillbirth was commom (odds ratio, 3.8 with $p < .001$) in a population study of 535 patients. Spontaneous vaginal delivery is preferred in hemodynamically stable patient the advantages being avoiding abdominal surgery, less blood loss, early recovery; regional anesthesia does not cause left ventricular depression.

- Heparin should be stopped when uterine contractions start or 24 hours before a planned caesarean section.

- Pain management is critical and therefore epidural anaesthesia is preferred for vaginal delivery.

- It is recommended to shorten the second stage of labour by assisting delivery by vacuum or by application of forceps.
- A single centre study in Germany of 7 women with a mean follow-up of 81 days and severely reduced ejection fraction (mean 18%), who received wearable cardioverter-defibrillator Life Vest, documented four events of ventricular fibrillation with successful shock therapy in three of these women.29 A larger retrospective registry study of wearable cardioverter-defibrillator post market release in 107 women with peripartum cardiomyopathy with average ejection fraction of 22% revealed no shocks for ventricular tachycardia after an average of 4 months follow up.

- In the third stage of labour, auto transfusion of blood products from the contarcted uterus increases cardiac preload and therefore IV furosemide may be needed for decongestion.

- Planned caesarean section is preferable in hemodynamically unstable or critically ill women, in which a combination of spinal and epidural anaesthesia is preferred.

- In general, termination of pregnancy is not indicated in peripartum cardiomyopathy.

RECURRENCE

Recurrence with subsequent pregnancy is common and therefore it is advised to avoid pregnancy in postpartum cardiomyopathy patients. The commonly seen complications in subsequent pregnancy in these patients include:

- A decrease in ventricular function
- Clinical heart failure
- Death

The risk is higher in women with persistent left ventricular dysfunction. Current consensus advises strongly against subsequent pregnancy for all women experiencing peripartum cardiomyopathy, particularly those who have not fully recovered left ventricular systolic or diastolic functions. If already pregnant, the decision of woman and then of her family should be a priority. A comprehensive recent review covering 191 recurrent pregnancies show the risk of relapse in persistent LV dysfunction (48% vs 27%) and death (16% vs nil).30

Raised concentration of TNFα could be a marker of deteriorating left ventricular function in subsequent pregnancies.

PROGNOSIS

The US National Inpatient Database revealed overall mortality of 1.3%, with long-term follow-up studies showing 11% mortality over 8 years. In the USA, out of total maternal mortality, 10%-15% are attributable to peripartum cardiomyopathy.31 Mortality is increased in patients with persistent left ventricular dysfunction for more than 6 months after delivery, mainly precipitated by three causes – left ventricular dysfunction, thromboembolic events and arrhythmias.

Improvement of left ventricular function within 6 months of delivery is expected in 50% of women. An early echocardiographic study of women with recovered left ventricular function revealed decreased contractile reserve in response to dobutamine indicating presence of persistent subclinical myocardial dysfunction. Therefore, women with recovered left ventricular ejection fraction remain at high risk of recurrence in subsequent pregnancies. The cellular and molecular recovery may significantly lag behind the echocardiographic recovery. An approach with caution is advocated even in patients who have made

full recovery. Therefore, patient should continue her heart failure therapy for a year after recovery to allow time for optimal remodeling. They should be closely monitored for 6 months postpartum and during the next pregnancy.

Cardiac transplant graft survival is inferior, and age-adjusted survival in cardiac transplant patient is lower reflecting younger age and allosensitization. Cardiac transplant or left ventricular assist device treatment is performed in 4% - 11% in peripartum cardiomyopathy patients.32

CONTRACEPTION

Contraception such as using intrauterine device is preferred, as it is safe and effective; oestrogen based contraceptives raise the risk of thromboembolism and sterilization should be strongly considered.

PERIPARTUM CARDIOMYOPATHY IN INDIA

The studies of peripartum cardiomyopathy patients are growing in India.

- A prospective study of 16 cases of peripartum cardiomyopathy was conducted at Apple Saraswati Multispeciality Hospital and Dr D.Y.Patil Medical College and Hospital, Kolhapur, Maharashtra, India, from January 2006 to December 2012. The study included 9/16 (56%) primigravidae, 4/16 (25%) with preeclampsia and 6/16 (35%) had co-existing hypertension. There was a significant difference in the echocardiography parameters as were seen in recovered and nonrecovered patients. The left ventricular rend-diastolic dimension was higher in nonrecovered patients (5.6 vs 6.06 cm), while left ventricular ejection fraction (28.7% vs 22.4%) and left ventricular fractional shortening (17.5% vs 13.4%) were significantly reduced in this subgroup. 13 out of 16 patients were followed up for a period of 1 year, out of which 61% (8/13) patients recovered completely. Maternal mortality was 6.25% (1/16) in the study.33

- In another study with 14/36 (39%) primiparae, 26/36 (72%) improved clinically and in 7/36 (48%), the left ventricular function status returned to normal. Persistent cardiomyopathy developed in five cases (14%) while five women (14%) presented with thromboembolic events, and anticouagulation was used as a secondary prophylaxis. Maternal mortality was 14% (5/36). Six of these women underwent subsequent pregnancies, one of these patients who had persistent cardiomyopathy died after delivering a stillborn baby. The remaining five cases who had a normal left ventricular functional status had favourable fetal outcomes. However, morbidities such as symptoms of heart failure (2 cases) and progression to persistent cardiomyopathy (1 case) were seen in these women.[34]

- The clinical, echocardiographic profiles and outcomes were studied in 56 women, with a mean age of 31±5 years at presentation and a mean parity of 2.6 ± 1, presenting with peripartum cardiomyopathy. They were followed up to a mean period of 6.1 years. Of the 56 patients, 18 (32.1%) had NYHA class II, 24 (42.9%) had NYHA class III and 14 (25%) had NYHA class IV symptoms, and 21 (37.5%) and 35 (62.5%), respectively, presented with features of heart failure during pregnancy and postpartum. On follow-up, the left ventricular ejection fraction improved from 31% ± 7.2% to 43% ± 8% (p value 0.05). Nine patients (16.1%) became pregnant, with a mortality of 55.5% during pregnancy and 23.2% during follow-up.35

- In a study conducted at Kasturba Medical College Hospital, Manipal, the incidence of peripartum cardiopathy reported was 1 case per 1374 live births. Major risk factors for development of peripartum cardiomyopathy were multiparity and advanced maternal age. Standard therapy was instituted and most of patients showed good symptomatic improvement, both at the time of discharge and during follow-up. Clinical improvement correlated with echocardiographically assessed improvement of LV function. Although significant thrombotic complications were noticed, there were no mortality during and in the follow-up period of the study.6

CONCLUSION

Peripartum cardiomyopathy is rare, but serious disorder of pregnancy. Though having significant morbidity and mortality, a good number of patients recover completely. There is a significant risk of developing PPCM in subsequent pregnancies. There shall be very high index of suspicion for PPCM as it mimics so many conditions. Exact etiology is not known is not known and various causative factors have been considered. Management of PPCM revolves around managing congestive heart failure (CHF). However, certain specific targeted therapies have also been considered and are promising; however, none of them have been adopted as standard care of management.

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