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Peripartum Cardiomyopathy: An Uncommon Enigmatic Disease Causing Significant Maternal Mortality and Morbidity

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ABSTRACT:
Peripartum cardiomyopathy (PPCM) is a rare,potentially life threatening condition, which is idiopathic, systolic dysfunction that presents in late pregnancy or, more commonly, the early postpartum period(1)(2).It is an important and steadily increasing cause of pregnancy-associated morbidity and mortality.
The incidence of peripartum cardiomyopathy in the United States has been estimated at between 1 in 900 and 1 in 4000 live births(3,4,5,6)and approximately 1 in 1,000 births worldwide. The aetiology of peripartum cardiomyopathy remains elusive. Although the condition is prevalent worldwide, women with black origin seem to be at a greater risk, and the condition is particularly prevalent in Nigeria and Haiti, 1 in 299 for Haiti(12).
Major risk factors include pre-eclampsia, advanced maternal age, and multiple gestation pregnancy(1,2,7-9). Although the complete pathophysiology of peripartum cardiomyopathy is unclear, research over the past decade suggests the importance of familial cardiomyopathies.
More than half of affected women recover systolic function, but some may become sufferers of chronic cardiomyopathy. There are no disease-specific therapies, management of peripartum cardiomyopathy is based on treatment of heart failure and its symptoms, and preventing long-term morbidity. Ventricular function recovery and rates of recurrence of peripartum cardiomyopathy vary by ethnicity and geography. Mortality rates associated with peripartum cardiomyopathy range from 3% to 40%, depending on geographic location.
The echocardiographic findings suggestive of peripartum cardiomyopathy are:

- Global or generalised reduced left ventricular wall motion(decreased contractility), rather than regional
- Dilatation of all four chambers of the heart (particularly left ventricle)
- Left ventricle end diastolic dimension exceeds 52mm (normal 36 to 52mm)
- Valvular compromise including moderate to severe MR or TR
- Reduced ejection fraction <45%
- LV thrombus

OBJECTIVE:
To study the incidence, diagnostic method, pathophysiology ,feto-maternal outcome, associated morbidity and the management of peripartum cardiomyopathy.
METHOD AND MATERIAL:
STUDY SETTING:
This isaretrospective study that was carried out in the Department of Obstetrics and Gynecology of Vadilal Sarabhai General Hospital, Ahmedabad, Gujarat from April 2018 to June 2019

SOURCE OF DATA:
Analysis of 9 cases of peripartum cardiomyopathy, between April 2018 to June 2019 was done in this tertiary care hospital.

INCLUSION CRITERIA:
All patients >18 years of age who were diagnosed with postpartum/peripartum cardiomyopathy at this centre were studied. The medical records of these patients were identified.
All the cases having peripartum cardiomyopathy were taken irrespective of presence or absence of any associated heart lesion or any previous history of cardiac lesion.
All delivery records and follow-up encounters were reviewed for clinical and demographic information. Patients with a history of prior cardiomyopathy attributable to other causes or structural heart disease were excluded.
Each patient was followed up for any improvement in EF at 6 weeks. Time to recovery was noted for patients who had improvement in LV function.

Assessment of EF: EFs at the time of diagnosis of PPCM were recorded and considered baseline EFs. Each patient was followed up over time to assess EF, and the EF from the last echocardiogram report was noted for those without LV improvement. For patients who had an improvement in EF, the EF and the time to improvement in EF were noted.
Definition of improvement: An EF >50% at follow-up visit was considered complete recovery. If the EF remained <35%, it was considered as no improvement. If the follow-up EF was between 35% and 50%, the improvement was considered to be partial provided that there was a >10% absolute increase from baseline EF value.

I. INTRODUCTION:

Peripartum cardiomyopathy (PPCM) is a potentially life-threatening condition commonly presenting as heart failure with reduced ejection fraction in the last month of pregnancy or in the months following delivery in women without other known cause of heart failure.

As shortness of breath, fatigue and pedal oedema are common feature in the peripartum period, a high index of suspicion is required to not miss the diagnosis of peripartum cardiomyopathy. Measurement of natriuretic peptides, electrocardiography and echocardiography are advised to promptly diagnose or exclude heart failure/PPCM.

Important differential diagnoses include pulmonary embolism, myocardial infarction, hypertensive heart disease during pregnancy, and pre-existing heart disease. A genetic factor is present in up to 20% of PPCM. PPCM is associated with high morbidity and mortality if spontaneous recovery does not occur, but also with a high chance of partial and often full recovery if diagnosed and treated promptly.

Incidence in India is not known. The incidence of PPCM is not known because population based estimates are not available, and the diagnosis of this rare disease is not always straight forward. The reported incidence rates range from 1 per 1485 to 1 per 15,000 .The currently accepted estimate of incidence is approximately 1 per 3000 to 1 per 4000 live births and tends to be higher in developing countries(10,11).
RiskfactorsforPPCMincludemultiparty,advancedmaternalage, multifetal pregnancy, preeclampsiaandgestational hypertension,andAfricanAmericanrace,butanycausalassociationhas notbeenshown[1,2,3,4].
The role of selenium and zinc deficiency in PPCM is controversial.

Hypertensive disorders coexist with peripartum cardiomyopathy. Antiangiogenic factors which are already associated with pathogenesis of preeclampsia can induce changes to cause peripartum cardiomyopathy.

II. ETIOLOGY:

A number of possible causes have been proposed for PPCM,(none of them are exclusively associated with PPCM) including myocarditis, maladaptive response to the hemodynamic stresses of pregnancy, abnormal immune response to pregnancy, stress-activated cytokines, and prolonged tocolysis(>4weeks) in pregnancy.
There have been reports of familial PPCM, raising the possibility that some cases of PPCM are actually familial dilated cardiomyopathies that get unmasked by pregnancy.

III. DIAGNOSIS:

The diagnosis of PPCM is based on the echocardiographic findings of new left ventricular systolic dysfunction during the peripartum period and absence of any positive history of cardiac disorder earlier during the antenatal period.

Many women in the last month of a normal pregnancy experience difficulty in breathing, fatigue, weakness and pedal oedema, which are symptoms identical to early congestive heart failure. Peripartum cardiomyopathy may, therefore, go unrecognized, leading to underestimation of incidence.

The symptoms and signs that raise the suspicion of heart failure include paroxysmal nocturnal dyspnoea, changes in the pulse rate and rhythm, sudden onset of chest pain, cough, neck vein distention, new murmurs consistent with atrioventricular valve regurgitation, and pulmonary crackles[9–11].

The diagnosis of PPCM requires excluding other causes of cardiomyopathy and is confirmed by standard echocardiographic assessment of left ventricular systolic dysfunction, including depressed fractional shortening and ejection fraction.

PROGNOSIS:
The prognosis for women with PPCM depends on the normalization of left ventricular size and function within 6 months after delivery.
Currently,thereisno available large populationstudyregardingrecommendationsforfuturepregnancyafterPPCM. Patientswhoseleftventricularsizeorfunctiondoesnotreturntonormalshouldbe counselled stronglytoavoidsubsequentpregnancyandtreatedaccordingly, including adoptingaheart-healthydietandlifestyle. Patientswhosecardiomyopathyapparentlyresolvescompletelyareamoredifficultgrouptocounsel.BecausePPCMhas beenassociatedwithmultipartyinsomestudies,theriskofirreversiblecardiac damage mayincreasewitheachsubsequentpregnancy.

Eventhoughtheleftventricularsizeandfunctionreturntonormal,thereisevidencethatcontractilereserve of the heartisimpaired,andrecurrenceofPPCMdespiterapidreturnofheartsizeandfunctiontonormalintheprioraffectedpregnancyhasbeenreported.

IV. RESULTS:

In the present study, there were 9 cases of peripartum cardiomyopathy out of total 10,802 deliveries in the given timeline giving an incidence of 0.83 cases per 1000 deliveries

Table 1: Incidence Of Peripartum Cardiomyopathy With Regard To Age

AGE (IN YEARS)	NUMBER OF CASES	PERCENTAGE
<20	1	11%
20-29	6	66%
> 30	3	33%

Table 1 shows increased incidence of PPCM with increasing age, our study corresponds with the study of Pillarisetti et al which shows PPCM to be common in the age group 30±7

Table 2: Table Showing The Increased Incidence Of Peripartum Cardiomyopathy In Case Of Multiparous Females

PARITY	**NUMBER OF CASES**	**PERCENTAGE**
PRIMIPARA	**2**	**22%**
1-3	**5**	**44%**
>3	**2**	**22%**

Table 2 shows increased risk of PPCM with multiparous state, which corresponds with the study of Pillarisetti et al which also showed the increased incidence of PPCM in multiparous patients.

Table 3 : Table Showing Increased Incidence Of PPCM With Following Associated Conditions:

ASSOCIATED CONDITIONS	NUMBER OF CASES	PERCENTAGE
1. PREECLAMPSIA	4	44%
2. GESTATIONAL DIABETES MELLITUS	1	11%
3. TOBACCO/ALCOHOL ABUSE	2	22%
4. ESSENTIAL HYPERTENSION	1	11%
5. HYPERLIPIDEMIA	3	33%
6. FAMILY HISTORY	0	0

Though peripartum cardiomyopathy can affect women in all age groups, more than half of the cases are in women older than 30 years of age.Infact,the incidence is 10 times higher in women older than 40 years old compared with those younger than 20 years old(15)

This table shows association of other obstetric and medical conditions with peripartum cardiomyopathy, particularly pregnancy associated hypertension-preeclampsia has a very high degree of association with peripartum cardiomyopathy(16)

Table 4 : Table Showing The Improvement In Ejection Fraction At 6 Week Follow Up

FOLLOW UP EF					
BASELINE EF	**<20%**	**20-30%**	**30-50%**	**>50%**	**IMPROVEMENT**
<20%	**2**	**0**	**1**	**1**	**2/4**
20- 30%	**0**	**0**	**1**	**2**	**3/3**
30-50%	**0**	**0**	**1**	**1**	**1/2**

Table 4 shows that majority of the patients had improvement in left ventricular function at follow up, which correspond with the study of Cunningham et al which showed similar result

A follow up of 6 week was carried out in the present study due to drop out of follow up patients beyond this period.

Table 5 : Table Showing Time At Which PPCM Was Diagnosed

TIME OF DIAGNOSIS	**NUMBER OF PATIENTS**	**PERCENTAGE**
ANTENATAL IN THIRD TRIMESTER	**2**	**22%**
POSTNATAL WITHIN 24HRS	**3**	**33%**
POSTNATAL WITHIN 7 DAYS	**4**	**44%**

Table 5 shows that 77% of the patients in this study were diagnosed after delivery which corresponds with the study of pillarisetti et al which showed similar results where 71% patients were diagnosed with PPCM post delivery

Table 6: Table Showing The Mode Of Delivery In Patients With PPCM

MODE OF DELIVERY	NUMBER OF PATIENTS	PERCENTAGE
VAGINAL DELIVERY	4	44%
CASEAREAN (LSCS)	5	55%

Table 6 shows that 55% of the patients in our study delivered by caesarean section, which corresponds with the study of Pillarisetti et al where caesarean section rate of 56% was noted.

A caesarean section rate of 32% was reported by Elkayam et al (13), for which obstetric pathogenesis was stated to be a reason in 70%, there was a cardiac cause in 10%, and the cause was unknown in 20%.

V. DISCUSSION:

The major findings of our study are as follows:
1) A large proportion of patients with PPCM recover LV function (55%), complete recovery occurred in 44% of patients in the present study.
2) The follow-up duration in this study was not long enough to note delayed complete recovery of EF.
3) The study had a mortality rate of 22%, which suggests high mortality associated with peripartum cardiomyopathy.
4) The study showed that majority of the cases were diagnosed postnatally 77%.
5)Maternal age of 30 years or more is a well described independent risk factor for PPCM, with an adjusted odds ratio of 1.7-1.8 compared with women less than 30 years.30 In a recent US analysis, the incidence of PPCM was one in 1200 live births among women aged 20-29 years, one in 790 live births among those aged 30-39 years, and one in 270 live births among those aged 40-54 years.
6) Pre-eclampsia and eclampsia are closely associated with PPCM

PPCM is a distinct clinical entity from other types of cardiomyopathy, although the symptoms resemble those of dilated cardiomyopathies.
Oxidative stress, genetic susceptibility, autoimmunity, and myocarditis have all been implicated in the pathogenesis (1,2).
Complete recovery of ventricular function usually occurs in a substantial proportion of women, which is unusual in other forms of cardiomyopathy (6).
Complete recovery was seen in 44% of patients in the present study, with partial recovery in 33%.
Factors predictive of recovery suggested in prior studies included baseline ejection fraction 30%.
A recent prospective study by McNamara et al. (14) that followed 100 patients in the United States showed complete recovery in 65% of women at 6 months and showed that baseline EF and race were predictors of recovery and outcomes.

VI. CONCLUSION:

Peripartum cardiomyopathy is an uncommon but serious medical condition that affects women throughout the world.

While the underlying pathophysiology remains unclear, vasculo-hormonal influences and genetic susceptibility probably play a role.

Although there have been significant advancements in understanding peripartum cardiomyopathy as well as development of improved management strategies, this uncommon enigmatic disease continues to be a cause of significant pregnancy-associated morbidity and mortality.

Careful diagnosis of peripartum cardiomyopathy is paramount in understanding the epidemiologic parameters of the disease and to improve appropriate treatment.

Currently, its management is limited to nonspecific heart failure treatment, given there are no proven disease-focused therapies. In current era of increasing incidence of peripartum cardiomyopathy, further research is needed to develop targeted therapies for better patient management and to reduce patient morbidity.

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