A study on Peripartum Cardiomyopathy

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ABSTRACT

Peripartum Cardiomyopathy (PPCM) is an idiopathic and reversible form of dilated cardiomyopathy which is of unknown aetiology with absence of determinable cause for cardiac failure. The disease is relatively uncommon with an estimated rate of 1 in 3,000-4,000 deliveries and its incidence is getting increased nowadays due to many reasons. The diagnosis of this entity is often difficult and it requires a high degree of clinical suspicion since majority of the cases in puerperium with cardiac failure (acute LVF or cardiac arrhythmias) are often the cause for maternal mortality. Early diagnosis facilitates immediate intervention, which will prevent maternal morbidity, mortality and also helps to team up with an internist or cardiologist to provide effective treatment to the patient. In this regard the present study was conducted at Hanamkonda, Warangal with the aim to review the maternal complications influencing mortality in peripartum cardiomyopathy. It is suggested that a protocol has to be evolved for early diagnosis, effective management and for better maternal and perinatal outcome.

Keywords: Peripartum Cardiomyopathy (PPCM), idiopathic, LVF, Cardiac arrhythmias, maternal mortality

I. INTRODUCTION

Peripartum cardiomyopathy is a life-threatening disorder of unknown aetiology that occurs in the last month of pregnancy or in first five months post-partum with absence of determinable cause for cardiac failure and marked by left ventricular dysfunction. Heart failure associated with pregnancy and the peripartum period was recognized in the literature as early as the 1800s by Virchow and others [1,2] but in 1970's it has been defined by Scientists Demakis et al., 1971, and Demakis and Rahimtoola [3,4]. The working group on PPCM of the European Society of Cardiology recently provided an updated operational definition of PPCM as cardiomyopathy with reduced ejection fraction (EF), usually <45%, presenting toward the end of pregnancy or in the months after delivery in a woman without previously known structural heart disease [5].

The timing of PPCM is also not certain. This timing differs strikingly from the onset of the major hemodynamic shifts of pregnancy, including reduced afterload, increased blood volume, and up to 40% increase in cardiac output, all of which occur early in the second trimester [6]. However, PPCM also can present well before and up to months after delivery. Because of these uncertainties, not all PPCM studies define the disease equivalently, raising caution in comparisons of results between studies.

Peripartum cardiomyopathy (PPCM) is a devastating form of cardiac failure affecting women mainly in their last months of pregnancy or early puerperium and often complicating their obstetrics as well as anesthetic

management. Setting aside several historical expressions, "peripartum cardiomyopathy" is now the term widely used to describe this clinical situation. In 1971, Demakis et al [3] first defined PPCM with three distinctive criteria (Table 1). The strict time limit used in their diagnostic criteria was intended to exclude congenital and acquired causes of heart failure that usually manifest by the second trimester. Specific echocardiographic diagnostic criteria have been proposed (Table 2) and their addition has resulted in easier differentiation between PPCM and other causes of cardiac failure.

Table 1. Diagnostic Criteria for PPCM

Diagnostic criteria for PPCM:

- Development of heart failure within last month of pregnancy or six month postpartum.
- Absence of any identifiable cause for heart failure.
- Absence of any recognizable heart disease before last month of pregnancy.

Table 2. Additional Echocardiographic Diagnostic Criteria

Demonstrable echocardiographic criteria of left ventricular dysfunction:

- Ejection fraction < 45%
- Left ventricular fractional shortening < 30%
- Left ventricular end-diastolic dimension > 2.7 cm/m2 body surface area.

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Epidemiology:

The incidence of PPCM varies worldwide due to many reasons [7]. The incidence of PPCM outside the United States is less well documented. Data in Africa and Asia suggest an incidence of ≈1 in 1000 live births [8-10]. There are, however, striking "hot spots" of PPCM, the cause of which remains unclear. In Haiti, the incidence of PPCM may be as much as 1 in 300 live births [11], possibly related to racial background, nutritional deficiencies, or a high prevalence of preeclampsia. In northern Nigeria, the incidence of PPCM has been reported as high as 1 in 100 live births [12], originally ascribed to indigenous customs of hot baths and high salt intake in the peripartum period, although a recent case-control study of 39 PPCM cases does not support this conclusion [13].

Overall, recent reports from various parts of the World show an incidence of 1 in 1,485 to 4,000 live births and the trend is increasing. Although it seems likely that women of reproductive age all over the world have some risk of developing PPCM, good data about incidence are unavailable because so few population-based registries exist. The reasons for this variation in incidence between countries remain unknown.

Risk Factors:

Common reported risk factors for PPCM are advanced maternal age[15-18], multiparity, multiple gestations [19-21], black race [15&17], obesity, malnutrition, preeclampsia &gestational hypertension[22-24], poor antenatal care, breast feeding, caesarean section, alcohol, cocaine and tobacco abuse, low socioeconomic condition and family history. PPCM has been reported mostly in women older than 30 years, but it may occur in various age groups. Though PPCM has been reported in primigravida, it is found to occur more commonly with multiparity. Twin pregnancy appears to cause a higher risk of developing PPCM. Typical etiological nature points towards hypertensive heart failure caused by fluid overload rather than a true variety of PPCM. Preeclampsia and hypertension have been associated with a significant number of PPCM cases. Many authors

even report it as a variety of hypertensive heart failure. There are also reports of other rare risk factors such as maternal cocaine, alcohol and tobacco abuse. The question of the risks of carrying a second pregnancy often looms large in women who have had PPCM. A recent comprehensive review of the literature on this topic [25] showed that the risk of relapse in patients with persistent LV dysfunction before their recurrent pregnancy is much higher than in those who have normalized LV function. In fact, one idea was proposed in the past that PPCM is a vascular disease triggered by the hormonal changes of late pregnancy [26],

In this regard true association of these risk factors with PPCM needs serious re-evaluation in the modern context since the publications mentioning these risk factors are several years old and with inadequate data based on small numbers of patients with older diagnostic criteria.

Aetiology:

The actual aetiology of PPCM is unknown. Several hypotheses like myocarditis, viral infection, autoimmune factors, inflammatory cytokines, abnormal hemodynamic response to physiological changes in pregnancy, prolonged tocolysis and selenium deficiency have been postulated. Symptoms vary including Dyspnoea on exertion, cough, orthopnoea and paroxysmal nocturnal dyspnoea are commonly seen in patients with PPCM and often mimic left ventricular failure (LVF). Cardiac thrombus formations are not uncommon and they may present with embolic features like chest pain, haemoptysis and hemiplegia. Though extremely rare, single or multiple coronary embolisms (and myocardial infarctions) have taken place in patients with PPCM. Nonspecific symptoms like palpitations, fatigue, malaise and abdominal pain may be present in 50% of cases. Most PPCM patients present in NYHA class III or IV, but the use of NYHA classification may not accurately reflect severity because of the normal occurrence of these features in advanced pregnancy.

Blood pressure may be normal, elevated or low. Tachycardia, gallop rhythm, engorged neck veins and pedal edema are commonly found Clinically, the heart may be normal or there may be mitral and/or tricuspid regurgitation with pulmonary crepitations and hepatomegaly. Patients may even present with seizures associated with cerebral edema and cerebellar herniation.

Symptoms of peripartum cardiomyopathy are similar to symptoms of heart failure including:

- -rapid heartbeat or palpitations
- -chest pain
- -excessive fatigue
- -tiredness during physical activity
- -shortness of breath
- -swelling of feet and ankles
- -increased urination at night

Diagnosis:

PPCM has often been ascribed to a failed hemodynamic stress test during pregnancy. Indeed, pregnancy triggers large hemodynamic shifts that significantly increase cardiac workload[4]. Diagnosis of PPCM is based on excluding common causes of cardiac failure such as infection, toxins and metabolic, ischemic or valvular heart disease. Early diagnosis of PPCM may be difficult because many of the similarities of its presenting features

with that of advanced pregnancy. Complications of late pregnancy (like anaemia, toxaemia and amniotic fluid embolism) have similar manifestations that must be kept in mind. The commonest presentation of PPCM is in the postpartum period when most of these features are disappearing. Echocardiography and other laboratory evaluations strengthen the clinical diagnosis. Common differential diagnoses include accelerated hypertension, preeclampsia, IDCM, pulmonary embolism, anaemia and thyrotoxicosis, among others. Engorgement of the neck veins, pulmonary crepitations, hepatomegaly, and pedal edema may also be present. [27--36].

Additional Echocardiographic Diagnostic Criteria like Ejection fraction < 45%, Left ventricular fractional shortening < 30% and Left ventricular end-diastolic dimension > 2.7 cm/m2 body surface area etc. helps to diagnose the disease in addition to consider the other Diagnostic criteria i.e., Development of heart failure within last month of pregnancy or six month postpartum, Absence of any identifiable cause for heart failure and Absence of any recognizable heart disease before last month of pregnancy.

PPCM is associated with higher rates of thromboembolism than other forms of cardiomyopathy [37]. The peripartum period is a hypercoagulable state [38] likely an evolutionary adaptation to minimize postpartum haemorrhage (historically the most common cause of maternal death). Cardiac dilation, endothelial injury, and immobility additionally contribute to clotting propensity in PPCM and thromboembolic events can sometimes constitute the presenting symptoms of PPCM [39-40].

A variety of imaging tests can measure the heart and the rate of blood flow. Some of these imaging tests can also view potential lung damage. Tests may include:

- -X-ray of the entire chest
- -CT scan for detailed pictures of the heart
- -nuclear heart scan to show heart chambers
- -sound waves to create moving pictures of the heart (echocardiogram)

Echocardiography: Echocardiography is generally sufficient to differentiate from these causes and usually shows LV dilatation of variable degrees, LV systolic dysfunction, right ventricular and biatrial enlargement, mitral and tricuspid regurgitation, and pulmonary hypertension [41-43]. Patients with PPCM can present with severe depression of LV function and demonstrate a rapid deterioration. Inotropes, intra-aortic balloon pumps, LV and biventricular assist devices, and extracorporeal membrane oxygenation should be considered in these cases and have been used successfully [44-48].

Material and Methods:

The present study was carried out at The Govt. Maternity Hospital, Hanamkonda in the department of Obstetrics and Gynaecology attached to Kakatiya Medical College, Warangal between Jun 2007 and Nov 2009 which comprises of 100 suspected cases of Peripartum cardiomyopathy.

This was a prospective and observational study done with the following tools.

- Case records of antenatal and labour wards.
- 2D Echo given by the cardiologist
- Partuirition register.

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In the present study, 17% of the women were in the age group of 15 – 19 Yrs, 24% between 20 – 24 Yrs, 17% between 25 – 29 Yrs, 13% between 30 – 34 Yrs, 17% between 35 – 39 Yrs, 25% between 40 – 44 Yrs.

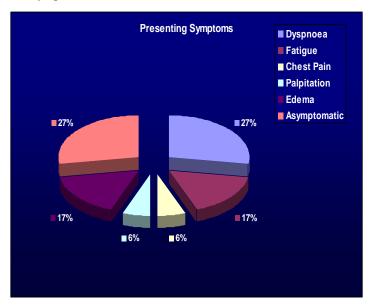
Table 3. Number of cases observed for the study				
Category	No of cases	Percentage		
Normal	74	74		
PPCM cases	18	18		
Other Heart diseases	8	8		
Total	100	100		

The data collected from the study were recorded and the standard statistical methods were followed by using the protocols established.

II. RESULTS AND DISCUSSION

In this study of 100 high risk cases of suspected peripartum cardiomyopathy, 18 cases were diagnosed as peripartum cardiomyopathy by 2D ECHO, 8 cases were confirmed cases of rheumatic heart disease with valvular disease and 74 cases did not show any ECHO cardio graphic abnormality suggestive of peripartum cardiomyopathy. The total number of deliveries conducted in the hospital in 2 years of study period were 12,102, out of which 18 cases of peripartum cardiomyopathy were noted i.e., an incidence of 1 in 672 cases [49]. In the present study major complications observed were dyspnea, fatigue, chest pain, palpitation, edema and asymptomatic.

Among all dyspnoea was the presenting symptom in 27% of cases, fatigue in 17%, chest pain in 6%, palpitation in 6%, edema in 17% and asymptomatic in 27% of cases.



In the present study, 22% of cases presented with LVF in antenatal women, 33% presented with LVF during intrapartum period and 44% cases presented with LVF in postpartum period.

Table: 4 Cases with LVF				
Presenting With LVF	No of Cases	No of Deaths	% of Mortality	
Antenatal	2	1	22	
Intrapartum	3	1	33	
Postpartum	4	2	44	
Total	9	4		

Table: 5 Maternal Complications Vs Mortality				
Complication	No of Cases	No. of Deaths	% of Mortality	
Thromboembolism	1	1	6	
Arrhythmias	3	0	0	
Congestive Heart Failure	7	2	11	
Pulmonary Edema	1	1	6	
Organ Dysfunction	0	0	0	
No complication	6	0	0	
Total	18	4		

In the present study it was observed that 11% of deaths were due to congestive heart failure followed by thromboembolism and pulmonary edema which caused 6% mortality. It was also observed that 75% of maternal mortality was seen in patients with Ejection Fraction, EF<50% and 25% was seen in patients with EF>50%. Hence showing the importance of EF in prognosis.

The present results are in support with the study of [50-51] which reported that PPCM is associated with higher rates of thromboembolism than other forms of cardiomyopathy. Another study [52] reported LV thrombus in 30% of the patients. Hence, it was suggested that high risk of thromboembolism, anticoagulation is advisable in PPCM at least during pregnancy and the first 2 months postpartum. Heparin and unfractionated heparin are safe during pregnancy, and the former is preferred near term because of its shorter half-life [53].

III.CONCLUSION

The increasing incidence of peripartum cardiomyopathy has to be realised and hence a protocol has to be evolved for early diagnosis, effective management and for better maternal and perinatal outcome. Early diagnosis of peripartum cardiomyopathy, adequate labour analgesia, early management of complications and institution of thromboprophylaxis in postpartum period reduces maternal mortality significantly and improves pregnancy outcome. Women who receive a diagnosis of peripartum cardiomyopathy are at risk for developing

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the condition with future pregnancies. In these cases, it is suggested that women to consider taking birth control to prevent pregnancies.

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