ROLE OF ROUTINE 2D ECHO IN HIGH RISK CASES OF PERIPARTUM CARDIOMYOPATHY: A CASE STUDY

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Abstract: 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 absence of determinable heart disease. The disease is relatively uncommon with an estimated rate of 1 in 3,000-4,000 deliveries and its incidence is getting increased nowadays. 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 arrhythymias) and are often the cause for maternal mortality. Therefore it is essential that when aetiological risk factor is present in a case or whenever clinical suspicion arises, 2D echocardiogram be suggested to diagnose peripartum cardiomyopathy. Early diagnosis facilitates immediate intervention, which will prevent maternal morbidity, mortality and it 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 to review the role of routine 2D echo in high risk antenatal women and it also throws light on the role of this noninvasive modality to diagnose cardiomyopathy of pregnancy and puerperium. The aims and objectives of the study are to evaluate the role of 2D ECHO in diagnosing peripartum cardiomyopathy and to assess its prognostic value for a better maternal and fetal outcome.

Key words: Peripartum cardiomyopathy, puerperium, mortality, 2D echo.

INTRODUCTION:

Peripartum cardiomyopathy is a potentially life-threatening pregnancy-associated disease that typically arises in the peripartum period and is marked by left ventricular dysfunction and heart failure. 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.
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, cesarean 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 reevaluation 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 Dyspnea on exertion, cough, orthopnea and paroxysmal nocturnal dyspnea 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, hemoptysis and hemiplegia. Though extremely rare, single or multiple coronary embolisms (and myocardial infarctions) have taken place in patients with PPCM. Non-specific 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.

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 of many of the similarities of its presenting features with that of advanced pregnancy. Complications of late...
pregnancy (like anemia, toxemia 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, anemia 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%
- Left ventricular end-diastolic dimension > 2.7 cm/m² body surface area etc.

It 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 hemorrhaging (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].

**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 [41-42]

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[43-47].

**Material and Methods:**

The present study was done 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
- Parturition register.

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

**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 cardiographic abnormality suggestive of peripartum cardiomyopathy. The total number of deliveries conducted in the hospital in 2 years of study were 12,102, out of which 18 cases of peripartum cardiomyopathy were noted i.e., an incidence of 1 in 672 cases.

The present study revealed that increasing maternal age has got a positive influence, as borne out by 25% of patients in the age group of 40 and above. Majority of the study done in literature testify that increase in maternal age is a risk factor in peripartum cardiomyopathy. In our study regarding the etiological/associated risk factors for peripartum cardiomyopathy, it was noted that 25% of the patients had multiple gestation, 23% had advanced maternal age, 19% had severe preeclampsia, 15% had obesity and gestational diabetes is seen in 14% of cases. This is in conformity with the study of Silva et al.,2005 and Witlin et al., 1997 who postulated that increased maternal age, preeclampsia and multiple gestation are associated with increased incidence of peripartum cardiomyopathy. All the conditions related above are high risk pregnancies and can be diagnosed early and investigated by 2D ECHO.

Our study also shows positive correlation between increasing parity and increased risk of peripartum cardiomyopathy as evidenced by 25% of peripartum cardiomyopathy being gravida 3 or above which is in conformity with studies in literature especially Silva et al. in 2005 says that the most common risk factors are poor socioeconomic status, increasing maternal age and multiparity.

Regarding the clinical presentation in this study, 27% of patients presented with dyspnoea, 27% were asymptomatic, 17% presented with fatigue and edema and 6% presented with chest pain and palpitations. This analysis shows that we have to investigate high risk patients with 2D ECHO because 27% of them are asymptomatic. Various studies have shown that 25% of patients presented with dyspnoea. In some studies all of the patients showed sinus tachycardia, raised JVP and pedal edema.

In our study 72% of women presented in gestational age between 28 weeks to term and 22% of cases presented in postpartum period. The cardiovascular changes occurring in pregnancy take place between 28 to 36 weeks with increased cardiac output, increased stroke volume and decreased peripheral vascular resistance. Therefore the symptoms of peripartum
cardiomyopathy manifest during this period. It is also very significant that in 22% of cases peripartum cardiomyopathy presents in postpartum period. In a study done by Lori and Karen in 2011[48], it was reported that most of the patients in their series developed symptoms during 1st month after delivery. Therefore we have to screen the patients of high risk by 2D-ECHO between 28 weeks to postpartum period.

Study went into the details of investigations and it showed that 100% of cases showed cardiomegaly, 89% showed ECG alterations, 44% of women had an EF<50% and 56% had EF>50%. A simple chest X-ray can be taken in second trimester of pregnancy which shows cardiomegaly and the ECG which is a non-invasive test is also useful. 2D-ECHO is mandatory for confirmation of peripartum cardiomyopathy.

Regarding 2D-ECHO findings, the review of literature says that M-Mode 2D doppler ECHO with EF<45% and LVEDD>57mm are the diagnostic criteria. In our study we noted that 39% of women had EF between 35-40%, 22% had EF between 45-55% and 39% had >55%. 70% of the patients had dilated cardiomyopathy with LVEDD>57mm which correlates well with all the studies.

From the present study one more important finding was that 50% of patients presented with LVF during different periods of gestation i.e., 44% in postpartum period, 33% in intrapartum period and 22% in antepartum period. The diagnosis is clinical and is also based on unexplained and sudden decrease in oxygen saturation for which the treatment modalities are IV nitroglycerine, diuretics and oxygen. Sometimes digoxin as well as nitrates and ACE inhibitors are used. Ventilatory support with Controlled Mode Ventilation and PEEP is also used. Also prophylactic anticoagulation with heparin is also used to prevent thrombosis and embolisation.

During intrapartum period emergency delivery with epidural analgesia/general anaesthesia if and when necessary is mandatory because delivery of fetus reduces haemodynamic stress on heart. The mode of delivery for patients with peripartum cardiomyopathy is generally based on obstetric indication and advantage of vaginal delivery is minimal blood loss, greater haemodynamic stability, avoidance of surgical stress and post operative complications. Regional anaesthesia has added advantage of decreasing preload and afterload and minimises fluctuation of cardiac output associated with labour. Regional anaesthesia is contraindicated in anticoagulated patients. In our study 84% of women delivered vaginally out of whom 13% (2 patients) were given epidural analgesia. 16% were delivered by caesarean for obstetric indications.

In our series, 22% of women i.e. 4 out of 18 had maternal mortality due to acute pulmonary edema in 17% of cases, thromboembolism complicating 6% of cases. The pulmonary edema of peripartum cardiomyopathy is sometimes intractable to treatment. Thromboembolism can be prevented by early institute of thromboprophylaxis.

In our series the maternal mortality in relation to EF was studied and it was found that in 75% of women EF was <40% and in 25% of women it was between 45-50% which was borne out by the study of Naseer et al in 2005, which have mentioned a mortality rate ranging between 18-66%. Study by Jeff et al (ACOG 2005) says a mortality rate between 25-50% with one half of deaths occurring within first 3 months of delivery.

Regarding the relation of NYHA class to our study, it was proved that 6% of women were in class III and IV as oppose to 39% in class I and II which correlates well with the study by Walkiria of Brasil in 2002. NYHA classification helps us to institute proper pharmacological management by B blockers, diuretics, ACE inhibitors and digoxin in class II and class III.

In our study the fetal outcome was good i.e., 50% of the neonates had apgar of 8-10, 25% had apgar of 6-8. In 19% of neonates, the fetal outcome was poor with apgar of 4-6, 6% were stillborn. This proves that early diagnosis and immediate intervention ensures a good fetal outcome and haemodynamic changes taking place in cardiovascular system do not effect the fetus adversely.

**Conclusion:**

Peripartum cardiomyopathy is a rare but serious condition of unknown cause that affects childbearing women. Diagnosis of peripartum cardiomyopathy requires heightened awareness among multidisciplinary patient care teams and a high degree of suspicion. Despite the frequent good outcomes in patients with PPCM, a significant fraction of patients eventually require transplantation, often with bridging via mechanically assisted circulatory support.

This study proves the role of routine 2D-ECHO in early diagnosis of high risk cases of peripartum cardiomyopathy, in relation to EF taken as a vital parameter and these patients can be monitored in a high risk pregnancy unit by senior obstetrician/internist/cardiologist to decrease the maternal mortality rate and improve pregnancy outcome. ECHO cardiography is an extremely valuable tool which is non invasive and useful for diagnosis and serial evaluation of peripartum cardiomyopathy in high risk women. Specifically, M-Mode 2D doppler ECHO cardiography is the instrument of choice. However, in depth research is required in this area as per the modern needs of the society.
References:


