THREE-YEAR PROSPECTIVE STUDY OF THE PROGNOSIS OF CARDIOMYOPATHY WITH PREGNANCY AND ITS OUTCOME AT A SINGLE INSTITUTION

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ABSTRACT

Objectives: To describe the characters of pregnancy with cardiomyopathy and its outcome.

Study Design: A case series study.

Place and Duration of Study: This study was done in the Armed Forces institute of Cardiology (AFIC) and Military Hospital Rawalpindi from October 2003 to October 2006.

Patients and methods: The study of case series consisted of thirteen consecutive women who were admitted with diagnoses of Cardiomyopathy with pregnancy. For the diagnosis of Peripartum Cardiomyopathy (PPCM), strict 4 point criteria were used.

Results: During the study period we managed a total of 13 pregnancies with Cardiomyopathy as outdoor cases, 08 with PPCM 4 with hypertrophic Cardiomyopathy and one due to thyroid disease. The number of patients were only 11 as 2 were back again within a year. We had one mortality with peripartum Cardiomyopathy who presented within 20 days of delivery with sudden cardiac failure. For peripartum Cardiomyopathy the average age at presentation was 28 years. Most patients presented with dyspnoea 4 cases, palpitations 7 and one patient with syncope. Left Ventricular Ejection Fraction (LVEF) was 0.35 on the average. The time of diagnosis was antepartum for three patients and postpartum for five. There were no cases of multiple pregnancies in this series. In hypertrophic cardiomyopathy majority of the patients had caesarean sections, less so in other categories .These were performed under general anaesthesia due to uncertainty about the safety of regional anaesthesia in these cases.

Conclusion: Cardiomyopathy is a poorly understood ominous complication of pregnancy. It accounts for a rising proportion of maternal mortality. Diagnosis should be considered whenever women present with heart failure in peripartum period. Improved case ascertainment requires heightened awareness among obstetricians and cardiologists.

Keywords: Peripartum Cardiomyopathy, Pregnancy associated Cardiomyopathy, Hypertrophic Cardiomyopathy.

INTRODUCTION

Cardiomyopathy in pregnancy and the post partum though rare accounts for a rising proportion of reported pregnancy related deaths. This has shown a rise since 1980's[1]. Cardiomyopathy can be divided into 2 groups. First group is peripartum, described in 1971. It is a distinct syndrome of unknown etiology with onset in last month of pregnancy or within 5 months of delivery[2]. The second group is of Cardiomyopathy other than peripartum Cardiomyopathy. Its onset is before pregnancy or before late pregnancy or those with specific underlying causes. These could have any of the three classically defined pathological presentations (dilated, restrictive or hypertrophic) or due to specific causes[3].

Peripartum Cardiomyopathy also called late pregnancy associated or early puerperium myopathy. It is a disorder of unknown etiology with potentially devastating consequences[4]. Following four criteria are needed to meet the definition of peripartum Cardiomyopathy [2].

1. Development of cardiac failure in the last month of pregnancy or within 5 months of delivery.
2. Absence of recognizable causes of cardiac failure.
3. Absence of recognizable disease prior to last month of pregnancy.
4. Left ventricular dysfunction demonstrated by classic criteria LVEF< 0.45, reduced shortening <25%.

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Its exact incidence is unknown. Much of the reported discrepancy is due to wide geographical variation. 1: 15000 in USA to 1: 1000 in S. Africa to 1: 100 in Nigeria. This is related to eating Kanwa dry lake salt for 40 consecutive days after delivery[5]. Etiology remains largely unknown. Inflammatory cytokines may play a role[6]. Myocarditis could be a possible cause based on biopsy. Familial clustering of Peripartum Cardiomyopathy (PPCM) has been observed. Hemodynamics of pregnancy could cause left ventricular hypertrophy and dysfunction[7].

PATIENTS AND METHODS

The study was conducted at Armed Forces Institute of Cardiology (AFIC) from October 2003 to October 2006. Obstetric patients are dealt with in a dedicated obstetric ward and labour ward based in AFIC. Obstetric cover is provided by Gynaecology and Obstetric department of MH Rawalpindi an allied hospital in the neighborhood.

Indoor patients diagnosed with Cardiomyopathy (PPCM or other) were included in the study. Patients who only reported in outdoor were not included. For PPCM four criteria were needed to meet the diagnosis as already mentioned in introduction. All patients with "other Cardiomyopathy" had echocardiographically proven disease.

All patients were evaluated by history, thorough physical examination. Relevant investigations like ECG & also chest X-ray with shielding in addition to echocardiography were performed where indicated. Patients with valvular and other heart disease in pregnancy were excluded from the study.

RESULTS

We managed a total of 13 pregnancies with Cardiomyopathy as indoor patients. The total number of patients was 11 as two were back again with in a year one with PPCM and 2nd with hypertrophic Cardiomyopathy.

In this series 8 patients met the 4 point criteria for PPCM. For PPCM, 3 patients presented ante partum after 36 weeks and five after delivery mostly within 6 weeks of delivery. The average age at presentation was 28 years. Most of them presented with dyspnoea (4 cases) and palpitations (7 cases). One patient (12% presented with syncope. LVEF on the average was 0.35 with a range of (0.22-0.45) (Table-1).

One pregnant patient had Cardiomyopathy due to thyroid disease. Is she pregnant was a 40 year old lady who presented with orthopnoea and paroxysmal nocturnal dyspnoea. Four patients were admitted with hypertrophic Cardiomyopathy. Two were diagnosed before pregnancy, one during pregnancy and the last incidentally in the post partum period. All cases were diagnosed due to symptomatology and in one patient it was incidental finding (Table-1). Out of the 13 pregnancies 7 were diagnosed ante partum, whereas in cases of PPCM 3 were ante-partum and 5 postpartum. Out of these 50% were delivered by caesarean section. Medications given to patients during pregnancy included beta blockers, digoxin and in one patient anticoagulants. Caesarean section rate was highest for patients with Hypertrophic Cardiomyopathy (3 out of 4 cases). These were performed under general anaesthesia. For the remaining, the caesarian section was done in only 2 cases and rest had vaginal delivery (Table-2). Pain relief during labour was in the form of Opiates. Postpartum decompensation was prevented by diuretics. Luckily there were no cases of atrial fibrillation in this series. There was one mortality in this series. One patient with PPCM after delivery presented with cardiac failure had LVEF of 0.26 and died within 20 hours of admission despite intensive care.

Regarding neonatal outcome we lost one baby to abortion in a case of Hypertrophic cardiomyopathy (HCM). One of the ladies with postpartum PPCM had delivered a dead baby at home. Lastly one pregnancy had to be terminated due to previous history of PPCM and recurrent cardiac dysfunction (Table-3).

Table-1: Clinical Features of PPCM, n=8

Features	Number
Age	28 yrs(22-38)
Dyspnoea NHYA >II	4
Syncope	1
Palpitations	7
LVEF	0.35% (0.2%-045%)

Table-2: Mode of Delivery n=13

Type of Cardiomyopathy	No. of pregnancies	LSCS	SVD
HCM n=4	4(30%)	3(75%)	1(25%)
PPCM n=8	8(61.5%)	2 (25%)	6* (75%)
Thyroid Cardiomyopathy n=1	1(8.5%)	Nil	1

*One termination of pregnancy at 16 weeks.

Table-3: Neonatal outcome

Type of Cardiomyopathy	Alive babies	Peri-natal death
HCM	3	1(Abruptio placentae)
Ante-partum PPCM	2	1*
Postpartum PPCM	4	1**
Thyroid	1	Nil

Note:
* Pregnancy terminated due to cardiac compromise.
** Dead baby delivered at home.

DISCUSSION

Cardiomyopathy with pregnancy though rare accounts for an increasing proportion of pregnancy related mortality. Incidence seems to be rising due to better diagnosis[8]. The increased reporting of death may be due to better case ascertainment.

In our series age was 28 years, other larger series of the world by El-Kayam[9] report as 30 years. In this case series there were no cases of multiple pregnancy or hypertension.

The cases in this series presenting with palpitations, dyspnoea & syncopal attacks, and same is observed in most of the other series[9]. The average LVEF was higher in our patients 0.35 versus 0.32.

All patients in this study with PPCM met the 4 point criteria described by National Heart Lung and Blood institute (NHLBI) 1992[10]. With this threshold for diagnosis the echo findings also included pericardial effusion and mitral regurgitation in some of the cases. Endomyocardial biopsy was not done in this case series as it does not help in management.

Most patients were managed by combination of Digoxin for symptom control, diuretics for removing excess fluid and Beta-blockers for after load reduction.

In this series, in cases of hypertrophic Cardiomyopathy, majority of the patients had operative delivery. Regional anaesthesia was avoided in these cases due to the gradient that exists across left heart. In PPCM antepartum vaginal delivery was preferred unless the mother decompensated. Vaginal delivery is associated with lower rates of complications as there is no 3rd spacing of fluids as occurs after caesarean section.

Prognosis of Cardiomyopathy must take into account maternal, obstetric and neonatal outcomes. Largest series of 153 cases describes a mortality of 10% with a cardiac transplant rate of 4%[9]. We had one mortality in this series (1/13) in a case of PPCM post partum. Adverse prognostic factors are black women and multiparity. According to the biggest study on pregnancy related mortality in Cardiomyopathy in USA, mortality was found to be 6 times higher in black women, 1.8 times higher with age from 20-35 years, 3 times more common with parity more than 3 and twice as common in multiparity[11-13].

Pregnancy after cardiomyopathy–to be or not to be? Well the answer is that pregnancy is not a reasonable option in 50 % of the patients who do not recover from ventricular dysfunction. The mortality here is high. In our series one patient after recovering from post partum PPCM was pregnant again within a year. She had to be offered a termination of pregnancy due to recurrent cardiac dysfunction. The second lady had hypertrophic Cardiomyopathy and did well. Such patients should be monitored for signs of ventricular dysfunction if they choose to become pregnant.

CONCLUSION

To sum up Cardiomyopathy is an ominous complication of pregnancy about which little is known. The diagnosis of peripartum Cardiomyopathy should be considered whenever women present with heart failure during the peripartum period. It accounts for an increasing proportion of maternal mortality. Though our study suggests that patients with

Cardiomyopathy, also hypertrophic Cardiomyopathy generally tolerate pregnancy well. No particular feature was associated with poor outcome. A limitation of the study is the small case number which prevented an assessment of confounding variables. Hence bigger multicentric studies need to be conducted to improve our assessment of this poorly understood condition.

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