

Peripartum Cardiomyopathy: A five year hospital-based analytical study

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Objective: To review the incidence, epidemiological characteristics, diagnosis and management of peripartum cardiomyopathy.

Design: Retrospective analytical study of all cases, which were diagnosed as peripartum cardiomyopathy (PPCM) among the maternity admissions between January 1st 1999 and December 31st 2003.

Setting: Salmaniya Medical Complex (SMC), which is the main referral hospital in Bahrain; where approximately 80% of all deliveries in the Kingdom are performed.

Methods: We reviewed all maternity admissions who developed cardiac or respiratory complications in the five year period, including those happened in the last four weeks of antepartum, intrapartum, or the first week of the postpartum periods. These have been drawn by diagnosis from the records departments, the labor room records and the ICU register and the information's then transferred to special forms.

Result: The incidence is comparatively low at 1/17,500. The age ranged between 23 to 45 years and the onset was between 38 weeks of pregnancy and early post partum period. No evidence of underlying cardiac disease has been found in any of the patients reviewed.

Conclusion: Among 52,806 deliveries conducted in the SMC and associated hospitals during the five years of this study there were three confirmed cases of PPCM and two doubtful cases. There has been no mortality cases, but the condition is life threatening and costly to treat. Research for the prediction of PPCM among high risk groups like sicklers are being investigated to develop better diagnosis and prevention.

Bahrain Med Bull 2004;26(3):

Peripartum cardiomyopathy (PPCM) is a rare disorder of unknown etiology. It was first described by Demakis and Rahimatolla in 1971¹. The incidence varies from 1/100 deliveries in Nigeria to 1/1300 in Europe, to 1/15000 in the United States (USA)².

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It is defined as a heart failure occurring during the last month of pregnancy, or during the first five postpartum months. A requisite for this definition is the absence of any identifiable cause for the cardiac failure and the absence of any recognizable heart disease prior to the last month of pregnancy. An additional prerequisite is the presence of left ventricular dysfunction demonstrated by echocardiography showing depressed shortening fraction, or reduced ejection fraction, abnormal cardiac wall motion and severe congestive cardiac failure³.

The pathologic findings are cardiac hypertrophy and focal myocardial fibrosis. Mural thrombi in either or both ventricles, endocardial fibrosis, degenerative, necrotic and fibrotic myocardial changes may be associated findings. Although myocardial fibrosis may be diffuse, it is only focal and microscopic and cannot provide sufficient explanation for either the hypertrophy or heart failure and other manifestations. It was suggested that the above lesions in and around the small ventricular arteries may lead to repeated episodes of closure with consequent focal ischemic necrosis, degeneration and fibrosis, with ultimate cardiac dilatation and failure. However in most cases, the intra myocardial vessels are normal⁴.

The etiology remains unknown but several hypotheses have been postulated. An autoimmune theory based on immune reaction to the fetal tissue was reported. Other explanations include underlying occult heart disease such as myocarditis, or maladaptive response to the hemodynamic stresses of pregnancy, stress activated cytokines, tocolytic drugs, cocaine abuse, selenium deficiency, chlamydia infection and enterovirus infections⁵.

The risk factors include: age above thirty-five year and multiparity, although the condition can be found in primiparas. Peripartum cardiomyopathy occurs in all races but is more common among black in USA. It seems also to be common among those who are pregnant with twins or triplets due to overloaded circulation⁶. Many associated conditions are reported: in Nigeria where the consumption of large quantities of salt in the peripartum period, may lead to overloaded circulatory disturbances⁶. Type 1 diabetes, anemia, pregnancy induced hypertension, hemoglobinopathies, and uses of tocolytic drugs in treatment of pre-term labor are other risk factors.

There is no “cure” for peripartum cardiomyopathy. However, variety of drugs were used to ease its symptoms. This included: digoxin, heparin and ACE inhibitors such as hydralazine. A low salt diet and an exercise program can be helpful; bed rest is not usually advised. In deteriorating severe peripartum cardiomyopathy, a heart transplant is an option⁷.

The mortality rate of this condition is very high, but it varies geographically. In the USA the reported mortality rate varies widely between 6 to 50 percent, with nearly half of these deaths occurring in the first three months after delivery of the baby or babies.

Prognosis also appears to be related to the severity of the left ventricular dysfunction. If a woman’s heart recovers normal beat function during the six months after the diagnosis, her chances of survival improve significantly⁸.

The objective of this study is to obtain the incidence of PPCM in our practice, review the methods of diagnosis and management of this condition.

METHODS

We have reviewed a number of resources: hospital ICU registers, the labor suite log books, the Health Information's Directorate (HID) data bank, and the hospital Records Department for the period starting 1st January 1999 to 31st December 2003.

During the study out of 52806 deliveries, three patients were diagnosed with PPCM and two were strongly suspected, but not confirmed with echocardiography. The case histories of the three confirmed cases will be reported.

THE CASES

CASE 1

Forty-five year old (G6, P4, Ab2) Bahraini woman was admitted through accident and emergency at 38 weeks of gestation. Her main complaint was shortness of breath, cough with frothy sputum, and lower chest discomfort. She was obese with dark complexions and cyanotic. She was restless and sweating profusely. Vital signs revealed a pulse rate to be 118 per minute, blood pressure 170/120 mmHg, temperature normal and respirations 28 breaths per minute.

Previous pregnancies had been uneventful and she gave no history of heart disease. The last pregnancy ended in caesarean section, the indication of which was unknown. She has also had an ectopic gestation treated by partial salpingectomy. The current pregnancy was uneventful apart from mild pregnancy induced hypertension (PIH), which required no treatment. Examination of the chest revealed bilateral crepitations. The heart was enlarged and had a gallop rhythm at the apex. The uterus was relaxed and showed no contractions.

She was treated with nasal oxygen, Lasix (frusemide) 40 mg, given rapidly by IV and a nitroglycerin drip 0.25 mg/Kg/mL. A blood sample was sent for arterial blood gases (ABG) and thyroid function tests, and echocardiography was requested. A review of her case record indicated that she was a known case of PPCM diagnosed by echocardiography during a previous pregnancy, five year earlier. This time, she was transferred to the ICU where she was put on a morphine drip, antibiotic cover Rocephine (ceftriaxone), digoxin 0.25 mg daily, heparin and Zantac (cimetidine).

Her condition improved and two days later she underwent caesarean section and sterilization with spinal anesthesia because of development of PIH and a non-reactive stress test on fetal monitoring. Her postoperative progress was uneventful and she was discharged home on the sixth postoperative day. Currently, her condition is stable and being followed up in the Cardiac Clinic.

CASE 2

Twenty-six year old Bahraini primigravida, was admitted at 38 weeks of gestation for elective caesarean section because of pregnancy induced hypertension (PIH) and triplets. She was treated for primary infertility in Jordan. The pregnancy was achieved with assisted reproductive technique (IVF) and resulted in triplet. Her admission at 13 weeks gestation showed hyperemesis and slight vaginal bleeding.

Cervical cerclage was performed under anesthesia. The post-operative period was smooth. At the 24th week she was referred to SMC for further care because of threatened pre-term labor and high risk of severe prematurity. She was also found an impaired glucose tolerance test illnesses or major surgery. Uterine contractions resolved with rest and hydration. At 33 weeks of gestation she was re-admitted to hospital with painful uterine contractions and was given an injection of Decadron (dexamethasone) 8 mg to stimulate production of fetal pulmonary surfactant and the cerclage was removed. Ultrasound showed the first fetus presenting as a breech. There was no evidence of any congenital abnormality and their birth weights were normal. The contractions ceased spontaneously, but the blood pressure was around 140/90 mmHg. She was discharged home with an appointment to be readmitted at the 38th week for an elective caesarean section.

At the 39th week she had a lower uterine segment caesarean section under general anesthesia. Live, mature and healthy triplets (two female and one male) were born. The early post-operative period was complicated by low grade fever and a rise in the blood pressure of 160/100mmHg. She was started on Keflex (cephalexin) 500 mg Q6h, and put on apresoline drip. Chest X ray revealed bilateral infiltration of both lungs, minimal pleural effusion and cardiomegaly. Blood gases showed lower oxygenation. Despite treatment with antibiotics there was minimum response. Echocardiograph showed a dilated heart with left ventricular strain and a reduced ejection component, confirming the diagnosis of PPCM.

The patient was transferred to the CCU for management of her heart failure. Her condition improved progressively and she was discharged home on the 14th post-operative day with follow up appointment to the Cardiology Clinic. Three years later she is still alive and well.

CASE 3

Twenty-three year old (G3, P2, Ab1) Bahraini woman at the 34th week of gestation with pre diagnosis of gestational diabetes was admitted through accident and emergency department with a history of severe pain in both knee joints, shortness of breath, cough and marked pallor. She was a known case of sickle cell disease, and PPCM had been diagnosed during her last pregnancy, six years earlier. On questioning, she said that she had no follow-ups in the interim period because she was free of any cardiac symptoms. Her vital signs were as follows 37.6⁰C, pulse 110 beats/minute, and blood pressure 150/90 mmHg. On examination, she was dyspneic, tachypnic and complaining of chest pain. The lungs had bilateral crackles. The fundal height of the uterus corresponded to dates. The fetus was in the longitudinal position, with cephalic presentation. The fetal heart was regular with a rate of 150 beats per minute. Arterial blood gases were reported as follows: pH 7.38, PaCO₂ 31 mmHg, PaO₂ 88.2 mmHg. Chest X ray was reported as showing cardiomegaly and bilateral infiltration of both lungs. She was put on Rocephine (ceftriaxone) 250 mg twice daily; as well as an injection of Decadron (dexamethasone phosphate) 8 mg and two units of packed red blood cells. Erythromycin 500 mg tablets 6 hourly and Aldomet (methyldopa) 250 mg twice daily were given. Her condition remained stable until the 11th day of admission when she showed signs of severe fetal distress on the non stress test (NST). An elective lower segment caesarean section was performed under spinal anesthesia and she was kept in the ICU during the early post-operative period.

The operation was uncomplicated and she gave birth to a live and healthy female baby. On the 2nd post-operative day in the ICU, her condition deteriorated following an exchange transfusion and she became cyanosed, dyspneic and tachypneic. She was given Lasix

(frusemide) 40mg by IV. The arterial blood gases showed signs of hypoxia; ECG showed left ventricular strain and echocardiogram confirmed the diagnosis of PPCM. The treatment consisted of oxygen, digoxin, frusemide and ACE inhibitors.

In the following days she progressively improved and was discharged home to be followed up in the Cardiac Clinic. Four years after her discharge she is still alive and well.

These two cases, of non-confirmed PPCM which have not been included in this series were as follows: a twenty-four year old primigravida with sickle cell disease (SCD) who was admitted at 32 weeks of gestation with shortness of breath, tachypnea and fever. Following chest radiographs, ABGs and ECG, she was diagnosed and treated as a case of pneumonia. Despite therapy her condition deteriorated and she was transferred to the ICU, where the diagnosis was adjusted to Acute Chest Syndrome and she was treated with antibiotics, oxygen, exchange blood transfusions, vasopressors and other relevant drugs.

She had a live pre-term labor and delivered a growth retarded pre-term baby in the ICU. Following delivery she had another episode of dyspnea and cyanosis. A pulmonary V/Q scan and ECG were non-specific, but echocardiograph revealed left ventricular strain and a reduced ejection component. Despite the high suspicion of PPCM, her discharge diagnosis was recorded as Acute Chest Syndrome secondary to SCD.

The second patient was a thirty two year old, grossly obese pregnant patient who came into the ANC at 32 weeks of gestation. She was a known case of gestational diabetes, who had not been compliant with her insulin therapy. She complained of breathlessness and chest discomfort. Her BP was 150/90 mmHg but other vital signs were normal. She was admitted to the labor suite, but her condition deteriorated rapidly. On auscultation the lungs were full of crepitations. Despite intense resuscitation she expired before a diagnosis was made. Therefore, they did not meet the diagnostic criteria.

DISCUSSION

Between 1st January 1999 and 31st December 2003, the number of deliveries in SMC and its peripheral maternity units was 52,806. Among those we have traced three patients who were diagnosed to have PPCM, giving an incidence of 1/17,500. Even if we include the other two unconfirmed cases the incidence would still be 1/12,000. Considering the international incidence which varies between one percent in parts of Nigeria to 1/ 1,300 in Europe to 1/15,000 in the USA, the incidence in Bahrain is relatively low. The possibility of under diagnosis cannot be excluded because approximately 20% of deliveries are conducted outside this institution, however, at SMC an average of 20 cases are treated annually for non-pregnancy cardiomyopathies. Echocardiographic diagnosis was available in the hospital since the early nineteen eighties. We feel that further research of the cardiac function by Doppler studies, echocardiography, and measurement of tissue perfusion in high risk pregnant women for PPCM, may help to facilitate the diagnosis and to undertake preventive measures.

PPCM can succeed normal deliveries as well as Cesarean sections. The risk factors of cases of PPCM in this series were similar with those reported from elsewhere⁹. The combination of anemia and hyper-dynamic circulation are important contributory factors. Associated diabetes and PIH were also seen in these cases. Circulatory overload as in cases of twins or triplets are other risk factors. None of the patients had any drug therapy that could account for this condition. PPCM in this series is not exclusive to elderly grand multiparas in their

mid thirties, but can happen even in young women in their first pregnancy as has been seen above. The symptoms particularly in patients with SCD can often be misdiagnosed as pulmonary embolism or acute chest syndrome as it happened in one of the cases.

Although there can be some variations in the clinical presentations of PPCM, particularly in patients with pregnancy induced hypertension or SCD, other severe anemia, or pneumonia, the specific tool for diagnosing PPCM is early echocardiography. Myocardial needle biopsy was recommended in the past, but nowadays it is hardly practiced. Admission to the ICU or the CCU as soon as the diagnosis is suspected is imperative. Care should be taken not to overload these patients with fluid by monitoring the central venous pressure. Ventilatory support and intubation may be necessary in some cases with acute respiratory distress. Treatment of the heart failure is the same as in other cases and suitable antihypertension drugs can be added when required.

CONCLUSION

Peripartum cardiomyopathy is a rare condition in Bahrain with an incidence of 1/17,200 deliveries. Although, there was no mortality in this series the morbidity of PPCM is extensive and the treatment is costly. In our practice we have also a complicating factors such as SCD, which can mask the diagnosis of PPCM. The authors, are currently participating in research for studying the role of Doppler flowmetry, early echocardiography and tissue perfusion studies among pregnancies complicated by SCD who are at risk of PPCM, in an attempt, to find if these tests will help in detecting patients who are likely to develop this condition, thus facilitating prevention and /or diagnosis.

REFERENCES

1. Demakis JG, Rahimatoola S. Peripartum cardiomyopathy. *Circulation* 1971;44:964-8.
2. Berg CJ, Chang J, Callaghan WM, et al. Pregnancy-related mortality in the United States, 1991-1997. *Obstet Gynecol* 2003;101:289-96.
3. Mehta NJ, Mehta RN, Khan IA. Peripartum cardiomyopathy: clinical and therapeutic aspects. Review article. *Angiology* 2001; 52:759-62.
4. Pearson GD, Veille JC, Rahimatoola S, et al. Peripartum cardiomyopathy: National Heart, Lung and Blood Institute and Office of Rare Diseases (National Institute of Health) Workshop Recommendations and Review. *JAMA* 2000; 283:1183-8.
5. Hibbard JU, Lindheimer M, Lang RM. A modified definition for peripartum cardiomyopathy and prognosis based on echocardiography. *Obstet Gynecol* 1999;94:311-6.
6. Carson M, Jacob DE, Sander GE, et al. Peripartum cardiomyopathy: A review. Last up dated, November 10, 2003. <http://www.emedicine.com/med/topic292.htm>. Date of access: March 20, 2004.
7. Ford RF, Barton JR, O'Brian JM, et al. Demographics, management, and outcomes of peripartum cardiomyopathy in a community hospital. *Am J Obstet Gynecol* 2000;182:1036-8.
8. Futterman LG, Lemberg L. Peripartum cardiomyopathy: an ominous complication of pregnancy: A review. *Am J Critic Care* 2000;9:352-6.
9. Danel I, Berg C, Johnson CH, et al. Magnitude of maternal morbidity during labor and delivery: United States, 1993-1997. *Am J Public Health* 2003,93:631-4.