

PERIPARTUM CARDIOMYOPATHY **(A CASE REPORT)**

Dr Shiranthi Fernando M.D.
Dr Athma Prasanna M.D. M.N.A.M.S.
ROYAL HOSPITAL
Dept of Anesthesia
P O Box 1131, SEEB PC 111
Muscat, Sultanate of Oman.

Correspondence:
Dr Athma Prasanna

INTRODUCTION

Peripartum cardiomyopathy is a rare form of heart failure of unknown etiology occurring during pregnancy or within five months of postpartum period¹⁻³, with a potential for fatal outcome in about 20 percent. It has an incidence of 1 in 3000^{4,5} patients and 1 in 30,000 deliveries⁶. The majority recover partially or completely, with some, requiring heart transplant. Although the advocacy for these patients is not to conceive, because of an increased risk of recurrence of cardiomyopathy with worsening signs and symptoms, it is not uncommon for these women to have subsequent pregnancies in societies that do not encourage family planning measures. These women with subsequent pregnancies do pose a challenge to their anesthetic management.

We report the anaesthetic management of a case with post partum cardiomyopathy, for caesarean section with successful outcome both for the baby and the mother.

CASE REPORT

Thirty three years old multigravida (gravida 8, para 5) of African lineage was scheduled for an elective Lower segment caesarian section with a principal diagnosis of RH isoimmunization, dilated cardiomyopathy and history of two previous Lower segment caesarian sections.

She presented to the hospital with history of decreased fetal movements for two days and palpitation on severe exertion. There was no chest pain or dyspnoea. She had been diagnosed to have post partum dilated cardiomyopathy during the second month of the postpartum period of the previous pregnancy, four years earlier.

The echocardiographic findings at the time of diagnosis was, a non dilated but globally hypokinetic left ventricle with moderately impaired overall left ventricular systolic function, with an ejection fraction of 40 percent. There was trivial mitral regurgitation, but no atrial stenosis or regurgitation. The right atrium and right ventricle were mildly dilated.

The repeat echocardiography two months after the initial diagnosis, showed the findings of non dilated, mildly hypo-contractile left ventricle. There was overall global, moderately impaired left ventricular systolic function with trivial mitral regurgitation and an ejection fraction of 35 percent. The right atrium and right ventricle were not dilated. The impression was post partum cardiomyopathy.

She did not visit the hospital for her cardiac complaint at any time after that, until the present pregnancy. Antenatal history of the present pregnancy followed in Primary Health Center at 15 and 25 weeks of gestation showed that the foetal ultrasound scans were normal, with middle cerebral artery Doppler around 95 centile. The repeat ultrasound scan at admission at 28 weeks revealed high velocity middle cerebral artery Doppler, suggestive of moderate to severe anaemia in the foetus.

The past obstetric history had no eventful episodes regarding cardiac status. However all babies had developed jaundice post delivery and had been treated with phototherapy. The previous caesarian sections were done for obstetric causes under general anaesthesia. The pelvimetry done in 1995 showed a pelvic inlet of 12.5 cms and an outlet of 15cms. The detailed obstetric history is as below

- (1) abortion at 1990
- (2) Spontaneous vaginal delivery- 1991
- (3) Spontaneous vaginal delivery – 1992
- (4) Spontaneous vaginal delivery – 1995
- (5) Lower segment caesarian section for transverse lie in 1998.
- (6) Spontaneous vaginal delivery–. H/o Antenatal haemorrhage and assisted breech delivery with fresh still birth in 1999. Had received one unit of blood transfusion after delivery.
- (7) Lower segment caesarian section in 2000 for breech, with abnormal CTG, had anti D titre of 1.256 and anti C of 1-32, amniocentesis done.

The Primary indication for Lower segment caesarian section in this admission was, previous two Lower segment caesarian sections and the secondary indication was breech presentation with RH isoimmunization and Cardiomyopathy. There was hypertrophied abdominal scar from the previous surgery.

In the preoperative phase, she needed a 15degree head up position while supine, even at rest. She was on frusemide, dexamethasone, and heparin, in addition to salt and fluid restriction. The echocardiography at admission with gestation age of 28 weeks, revealed post partum dilated cardiomyopathy with moderate Left ventricular dysfunction with compensated heart. She was not in Left ventricular failure and had an ejection fraction of 25 percent. The comparison of echocardiographic findings at diagnosis and at recent admission revealed that the post partum cardiomyopathy had worsened over the four year period. The other investigation parameters are shown in the Table:

	27/12 (preop)	29/12 (day of op)	30/12 (post op)
Hb	12.1	11.5	11.4
platelets	242	235	224
WBC/DC	6.9	6	7.5
PT coagulation	-	13.2	-
APTT	-	27.4	-
Fibrinogen	-	3.4	-
TT	-	15.0	-
Urea	-	2.4	1.5
Creatinine	-	49	46
NA	-	137	138
K	-	3.9	3.9
Glucose	-	4.5	-
Protein	-	54	-
Alb/Glob	-	29/25	26
bilirubin	-	16	-
ALT	-	13	-
AP	-	84	77
Calcium	-	2.44	2.44
Megnesium			0.68 (0.65-1.25)
Tropanin	-	0.011	
Myoglobin		69.33nanno (N=25- 58ng/ml)	

ANAESTHETIC MANAGEMENT

The anaesthetic management for lower segment caesarian section was planned under epidural anaesthesia after a detailed explanation to the patient in the preoperative period. The epidural catheter was inserted at L2-3 level in the left lateral decubitus position through an 18 gauge Tuohy needle and fixed at 11 centimeters at skin. The test dose of 3 millilitres of 2% xylocaine with adrenaline was administered, followed by 50 micrograms of fentanyl. The total dose of 12 millilitres of 2% xylocaine with adrenaline was administered in small increments of 4 millilitres to prevent any sudden haemodynamic instability. After ensuring the adequacy of the block, up to T8 dermatomal level, the surgery was performed. A top up dose of 4millilitre of 0.5% bupivacaine was administered after the delivery of the baby.

Intraoperative monitoring, with Agilant HP monitor, included percutaneous arterial saturation (SaO₂), continuous electrocardiogram (ECG), fractional inspired Oxygen concentration (FIO₂), direct arterial blood pressure (ABP), by cannulation of right radial artery, and central venous pressure (CVP) through cannulation of right internal jugular. The procedures for invasive monitoring were done under awake condition after local infiltration with 1% plain Xylocaine.

There was no intra operative haemodynamic instability through out the surgical procedure, either with the rate or rhythm. The heart rate was maintained between 74-78 beats per minute, and the blood pressure around 130/70 mm Hg.

Twenty international units of drug syntocinon to facilitate uterine contraction was infused slowly after the delivery of a premature female baby, to prevent sudden drop in systemic vascular resistance, and an increase in heart rate. The diastolic dysfunction aggravated transiently during the extraction of the baby and while obtaining uterine haemostasis. Minimal pressure on the uterus would elevate the CVP to 18-22 cms of water from a base level of 12 cms with a 10-15% drop in the blood pressure. However this did not require any intervention.

Post operatively she was monitored in Intensive care unit, without any inotropic support at any time. Post operative analgesia was through continuous epidural infusion of 0.125% bupivacaine with 2 micrograms fentanyl at a rate of 5ml per hour. She was discharged a week later on iron capsules, vitamins and paracetamol.

DISCUSSION

Peripartum Cardiomyopathy is defined clinically as the onset of cardiac failure with no identifiable cause in the last month of pregnancy or within five months after delivery, in the absence of heart disease. The finding of left ventricular systolic dysfunction by echocardiography is an important criterion for making the diagnosis. Stricter echocardiography criteria have been recommended such as a left ventricular ejection fraction of less than 45 percent, fractional shortening of less than 30 percent of an M-mode echocardiographic scan or both and a left ventricular end diastolic dimension of more than 2.7 cm per square meter of body surface area⁷.

Some of the risk factors for development of post partum dilated cardiomyopathy include multigravida, multiple gestation, pregnancy induced hypertension or preeclampsia, older women with a history of several previous pregnancies, and of African descent.

The diagnosis is made by exclusion criteria as the disease has no pathognomic features⁸. The aetiology and pathogenesis, such as viral infection associated with altered immune status during pregnancy triggering an autoimmune response in susceptible individuals^{5,9,10} is mostly theoretical. The increased blood selenium levels and increased salt intake, as other causes have been hypothesized. Long standing post partum cardiomyopathy may damage other systems such as liver and the kidney.

Our case fulfilled all the criteria for diagnosis of post partum dilated cardiomyopathy done at two months into the post partum period without any evidence of heart disease during the previous pregnancy. The echocardiography had revealed left ventricular systolic dysfunction and global moderate hypokinesia with an ejection fraction of 40 percent. She had associated risk factors of being multigravida, and of African descent.

In the absence of any cardiac symptoms, one of the early indicators about this condition, is revealed during evaluation of the foetus with a foetal monitor and ultrasound. Since the foetal growth is dependent on good blood flow to the uterus and placenta, an insufficient blood flow, show signs of inadequate oxygenation by slowed growth. This might prompt further investigation to discover heart disease. In our case the diagnosis of peripartum cardiomyopathy

had already been established and the fetal monitor with Doppler revealed RH isoimmunization leading to anaemia.

Although opinions differ as to the optimal method of anaesthesia for caesarean section, be it elective or emergency, any technique employed should avoid increases in afterload and use of negative inotropic agents. The use of vasodilator infusion and availability of inotropic support with invasive monitoring would be helpful.

General anaesthetic techniques, involve the use of either intravenous cardiodepressant drugs such as thiopentone and /or the inhalational anaesthetic agents such as Isoflurane, sevoflurane or desflurane or high dose narcotics, for maintaining haemodynamic stability. The latter technique may necessitate post-operative ventilation for both mother and infant. Since the endpoint at induction of anaesthesia with narcotics is not well defined, there is an increased risk of gastric aspiration. The management of a failed intubation may become difficult by the longer acting nature of these drugs with mask ventilation. This may be compounded further if associated with obesity.

The considerations for central neuraxial anaesthesia in these patients are similar to those with other causes of heart failure. Subarchnoid block may better be avoided in these patients because of sudden onset of haemodynamic instability. Epidural anaesthesia may be a better choice particularly when incremental doses of local anaesthetic are administered along with opioids¹¹.

The gradual and controlled induction of anaesthesia, may improve myocardial performance and the cardiac output by decreasing the systemic vascular resistance, thus reducing the afterload on the left ventricle without impairing contractility, although not all authors agree¹². The presence of a pulmonary artery catheter¹³ can guide fluid and inotrope requirements, with minimal change in haemodynamic parameters. These women do not need additional volume before induction of central neuraxial block. These considerations made us to prefer epidural anesthesia in this case.

Small bolus doses or an incremental infusion of bupivacaine 0.5% with fentanyl 4-5 mcg/ml have been advocated as suitable for these purposes. In our case we preferred to use 12 millilitres of 2% Xylocaine with adrenaline in incremental doses in addition to 50mcg of fentanyl as bolus dose in epidural space. The choice for the 2% xylocaine instead of 0.5% bupivacaine is its rapid onset and intense motor block. The cause of transient residual neuropathy is not yet proven conclusively. The anaesthesia was prolonged by administering 0.5% bupivacaine as the top up dose. We did not administer additional intravenous fluid volume before induction of neuraxial block.

Intra operative monitoring depends on the preoperative signs and symptoms. If the cardiomyopathy is asymptomatic, a central venous catheter is adequate with non invasive blood pressure monitoring, with a provision for using a per cutaneous catheter introducer to allow rapid insertion of a pulmonary artery catheter if needed¹³. In symptomatic cardiomyopathy or with echocardiographic findings of left ventricular dysfunction, a pulmonary artery catheter and an arterial line need to be inserted. We preferred to monitor central venous pressure with triple lumen catheter in the right internal jugular vein and direct arterial pressure

through right radial artery cannulation, inserted under local anaesthesia since the patient was mildly symptomatic preoperatively. We did not use any inotropic support as there was haemodynamic stability and no tachycardia.

The syntocinon (pitocin) after delivery was used as an infusion, to prevent sudden vasodilatation, resulting in hypotension and tachycardia requiring rapid fluid infusion. This also helped in reducing the after load maintaining the haemodynamic stability.

We monitored this patient in the intensive care unit, because, the post operative management also requires intensive monitoring measures similar to the intraoperative period until the patient is stabilized. The retention of water due to the antidiuretic effect of syntocinon (pitocin), and the reabsorption of the third space fluid after 48 hrs of the caesarian section, may increase the preload, worsening the patient's condition.

These women may develop a reduction in left ventricular systolic function during subsequent pregnancies. This reduction would be greater in those with persistent left ventricular dysfunction at the start of the pregnancies as was in this case. Symptoms of heart failure develop in about 20 percent of women whose systolic function is normal at the start of the subsequent pregnancy and in almost half of the women who have persistent left ventricular dysfunction¹⁴.

Our patient had dyspnoea on exertion and was comfortable at rest with a 15 degree head up position. This is probably due to the impaired contractile reserve which is not revealed during rest. The pregnancy induced haemodynamic stress may unmask impaired contractile reserve that is not apparent at rest. This deterioration in cardiac function probably could be determined with help of dobutamine echocardiography¹⁵ which we did not perform. The requirement of a head up tilt at rest in this case may have been due to the combination of heart problem and the abdominal distension pushing the diaphragm due to gravid uterus, than due to orthopnea. These women with persistent cardiac dysfunction, also have increased frequency of premature birth¹⁴ as was in this case.

The medical management of these patients is similar to patients with heart failure. Salt restriction, diuretics are advocated to decrease pulmonary congestion, and volume overload. Our patient was on oral frusemide, and salt restriction.

These patients are at increased risk for thromboembolic events since there is association between impaired cardiac function, pregnancy and prothrombotic state. Patients on oral anticoagulants require change to parenteral anticoagulants with short half life and the dose adjusted according to the Partial-thromboplastin time which may be discontinued before delivery. After delivery warfarin may be used. Our patient was on prophylactic heparin from the day of hospitalization. We discontinued the heparin twelve hours prior to the surgery. The coagulation profile particularly the activated Partial-thromboplastin time was within the normal range.

The outcome of these patients with peripartum cardiomyopathy is highly variable. In some the clinical and echocardiographic status improves rapidly and returns to normal. The initial severity of the left ventricular systolic dysfunction or

dilatation is not necessarily predictive of the long term functional outcome¹⁶. Few deteriorate rapidly requiring heart transplant¹⁷. Some will have persistent cardiac dysfunction as was in our case, while few have a slow return to normal cardiac function over several years.

Women with peripartum cardiomyopathy appear to have a better survival rate (94percent at 5 years) than patients with cardiomyopathy due to other causes¹⁸.

Although women with post partum dilated cardiomyopathy should ideally not have subsequent pregnancies, to prevent the worsening of the condition with a threat to their own survival, it is not always possible especially in societies which do not advocate family planning measures. Hence these women continue to pose a challenge to the medical community's (anaesthetist) skill and knowledge to have a successful outcome.

REFERENCES

- (1) Lang RM, Lampert MB, Poppas A, Hameed A, Elkayum U. Peripartum cardiomyopathy. In: Elkayum U, Gleicher N, eds. Cardiac problems in pregnancy. 3rd ed. New York:Wiley-Lisa, 1998:87-100.
- (2) Heider AL, Kuller JA, Strauss RA, Wells SR. Peripartum Cardiomyopathy: a review of the literature. *Obstet Gynecol Surv* 1999;54:526-531.
- (3) Pearson GD, Veille J-C, Rahimtoola S et al. Peripartum cardiomyopathy: National Heart, Lung and Blood Institute and office of Rare diseases (National Institutes of Health) Workshop recommendations and review. *JAMA* 2000;283:1183-88
- (4) Ventura SJ, Peters KD, Martin JA, Maurer JD. Births and Deaths: United States, 1996. *Mon. Vital Stat Rep* 1997;46(1): Suppl 2.
- (5) Veille JC. Peripartum Cardiomyopathy - A Review. *Am J Obstet Gynecol* 148; 805-818, 1984.
- (6) Humans DC: Peripartum Cardiomyopathy *N. Engl J. Med* 1985; 312:1432
- (7) Hibbard JU, Lindheimer M, Lang RM. A modified definition for peripartum cardiomyopathy and prognosis based on echocardiography. *Obstet Gynecol* 1999;94:311-316.
- (8) Julian DG, Szekely P. Peripartum cardiomyopathy. *Progress in Cardiovasc Dis* 1985;27:223-240.
- (9) Demakis JG, Rahimtoola SH, Sutton GS et al. Natural Course of Peripartum Cardiomyopathy. *Circulation* 44; 1053-1061, 1971.
- (10) Demakis JG, Rahimtoola SH. Peripartum Cardiomyopathy. *Circulation* 44; 964- 968, 1971.
- (11) Hawthorne L, Lyons G. Cardiac arrest complicating spinal anesthesia for caesarian section. *Int J Obstet Anesth* 1997;6:126.
- (12) Brown G, O'Leary M, Douglas I, Herkes R. Perioperative Management of a Case of Severe Peripartum Cardiomyopathy. *Anaesth. Intens. Care* 20; 80- 83, 1992.

- (13) Breen TW, Janzen JA. Pulmonary hypertension and cardiomyopathy: anesthetic management for caesarean section. *Can J Anaesth* 1991;38:895.
- (14) Elkayam U, Tummala PP, Rao K, Akhter MW, Karaalp I S, Wani OR, Hameed A, GviabdaBS, Shotan A . Maternal and foetal outcomes of subsequent pregnancies in women with peripartum cardiomyopathy. *N Engl J Med* 2001;344;21:1567-1571
- (15) Lampert MB, Weinert L, Hibbard J, Korkorz C, Lindheimer M, Lang RM. Contractile reserve in patients with peripartum cardiomyopathy and recovered left ventricular function. *Am J Obstet Gynecol* 1997;176:189-195.
- (16) Cole P, Cook E, Plappert T, Saltzman D, St John Sutton M,. Longitudinal changes in left ventricular architecture and function in peripartum cardiomyopathy. *Am J Cardiol* 1987;60:871-876.
- (17) Aziz TM, Burgess MI, Acladious NN et al. Heart transplantation for peripartum cardiomyopathy: a report of three cases and a literature review. *Cardiovascular Surg* 1999;7:565-567.
- (18) Felkar GM, Thompson RE, Hare JM et al. Underlying causes and long term survival in patients with initially unexplained cardiomyopathy. *N Engl J Med* 2000,342:1077-84.