Idiopathic dilated cardiomyopathy presenting in pregnancy

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Purpose: To describe the clinical course and management of a patient who presented with idiopathic dilated cardiomyopathy in early pregnancy.

Clinical Features: A 27 yr old, previously well, Chinese primigravida presented at 18 wk gestation with a history of irregular heart beat and decreased exercise tolerance. Echocardiography showed moderate left ventricular dysfunction with left ventricular ejection fraction of 35-40%. Idiopathic dilated cardiomyopathy was diagnosed. She declined termination of pregnancy and was managed medically with furosemide, digoxin and potassium supplements. Low molecular weight heparin was prescribed. Emergency Cesarean delivery was performed at 31 wk gestation because of deteriorating liver function and a non-reassuring fetal heart rate pattern. General anesthesia was given because of relative urgency, the patient's wish, and concerns about potential risk of spinal hematoma. Invasive monitoring with pulmonary and radial artery catheters was used and low dose inotropic support was given. Postoperatively, she was managed in the intensive care and coronary care units where she was treated with dobutamine, furosemide, digoxin, potassium, captopril, losartan and warfarin. Her postoperative course was complicated by a severe embolic stroke five weeks after delivery and she died five months later.

Conclusion: Idiopathic dilated cardiomyopathy may rarely present in pregnancy. A multidisciplinary approach and close peripartum monitoring are important in management and termination of pregnancy should be considered. Thromboembolic complications are a major risk.

Objectif : Décrire l'évolution et le traitement cliniques d'une patiente qui présentait une hypertrophie cardiaque idiopathique au début de sa grossesse.

Éléments cliniques : Une Chinoise primigeste de 27 ans, auparavant en bonne santé, présentait à 18 sem de grossesse un pouls irrégulier et une diminution de tolérance à l'effort. L'échocardiographie a montré une dysfonction ventriculaire gauche modérée et une fraction d'éjection ventriculaire gauche de 35-40 %. Le diagnostic d'hypertrophie cardiaque idiopathique a été posé. La patiente a refusé de mettre fin à sa grossesse. Le traitement médical administré comprenait donc du furosémide, de la digoxine et des suppléments de potassium. De l'héparine de faible poids moléculaire a été prescrite. Une césarienne d'urgence a été réalisée à 31 sem de grossesse à cause d'une fonction hépatique altérée et d'un rythme cardiaque foetal inquiétant. On a utilisé une anesthésie générale en tenant compte de l'urgence relative, du désir de la patiente et du risque potentiel d'hématome rachidien. Un monitoring effractif, constitué de cathéters des artères pulmonaire et radiale, a été utilisé et une faible dose de soutien inotrope a été administrée. Après la césarienne, la patiente a été traitée aux unités de soins intensifs et de soins coronariens où elle a reçu : dobutamine, furosémide, digoxine, potassium, captopril, losartine et warfarine. L'évolution postopératoire s'est compliquée par une sévère embolie cinq semaines après l'accouchement et la patiente est décédée cinq mois plus tard.

Conclusion : L'hypertrophie cardiaque idiopathique se présente rarement pendant la grossesse. Une démarche multidisciplinaire et une surveillance étroite de la grossesse sont des composantes importantes du traitement et même l'interruption de la grossesse est à considérer. Les complications thromboemboliques sont un risque majeur.

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IDIOPATHIC dilated cardiomyopathy occurring during pregnancy has been rarely described.¹ Although it clinically resembles peripartum cardiomyopathy, the latter occurs in late pregnancy or in the puerperium. We describe the anesthetic management and course of a 27 yr old primigravida who first presented with signs of idiopathic dilated cardiomyopathy at 18 wk gestation and proceeded to Cesarean section at 31 wk gestation.

Case Report

A previously well 27 yr old primigravida presented to her family practitioner at 18 wk gestation with irregular heart beat and decreased exercise tolerance. After referral to a cardiologist, she was admitted to hospital for management. Echocardiography showed moderate left ventricular dysfunction with left ventricular ejection fraction of 35-40%, and mitral regurgitation. A 12 lead electrocardiogram (ECG) showed left bundle branch block and first degree atrioventricular block. Serum glutamic-oxaloacetic transaminase and erythrocyte sedimentation rate were mildly increased, but other investigations, including thyroid function tests and screening for viral infection were normal. As no underlying or etiological cause for cardiomyopathy could be identified, a diagnosis of idiopathic dilated cardiomyopathy was made. Termination of pregnancy was discussed with the patient and her husband but was declined. She was discharged home with arrangements for regular medical and obstetric follow-up as an outpatient.

The patient's condition was stable until 24 wk gestation when she was readmitted complaining of dyspnea on mild exertion and orthopnea. Examination revealed a third heart sound but no lung crepitations. Echocardiogram showed left ventricular ejection fraction was 31%. Medical treatment included 0.125 mg digoxin daily, 80 mg furosemide daily, potassium supplements and low molecular weight heparin (0.3 ml fraxiparine *sc*) daily. The patient remained in hospital with a plan for elective Cesarean delivery nearer to term. Anesthetic consultation was requested. At 31 wk gestation, because

of signs of deteriorating liver function (alkaline phosphatase $114 \text{ IU}\cdot\text{l}^{-1}$ and total bilirubin $20 \text{ }\mu\text{mol}\cdot\text{l}^{-1}$) and fetal heart rate decelerations, Cesarean section was decided upon as an emergency. Anesthetic reassessment showed arterial pressure of 110/50 mmHg, elevated jugular venous pressure to mid neck, mild peripheral edema, and cardiomegaly. Due to the relative urgency, potential risks of spinal hematoma associated with low-molecular weight heparin, and the patient's wishes, general anesthesia was elected.

Premedication was with 50 mg ranitidine and 1.2 g amoxycillin/clavulanic acid *iv*, and 0.3 M sodium citrate 30 ml *po*. Standard monitoring was used and radial artery and pulmonary artery catheters were inserted under local anesthesia. Baseline hemodynamic parameters are shown in the Table. After pre-oxygenation, general anesthesia with cricoid pressure was induced using 0.6 mg alfentanil and 14 mg etomidate. The trachea was intubated after 80 mg succinylcholine and neuromuscular block was continued using vecuronium. Anesthesia was maintained using isoflurane, at end-tidal concentration of 0.5%, in nitrous oxide 50% and oxygen 50%. A 1.62 kg girl was delivered with Apgar scores of 6 (1 min) and 10 (5 min). After delivery, 5 mg morphine, and 10 IU oxytocin were given over five minutes. Total blood loss was estimated at 500 ml and intravenous fluid replacement of 1000 ml lactated Ringer's solution was given.

The hemodynamic changes at phases of the operation are summarized in the Table. There were no major hemodynamic changes after induction of anesthesia and tracheal intubation. After delivery, cardiac output decreased to $2.0 \text{ L}\cdot\text{min}^{-1}$ and dobutamine infusion was started at $5 \text{ }\mu\text{g}\cdot\text{kg}^{-1}\cdot\text{min}^{-1}$. At the end of the operation, neuromuscular block was reversed using 2.5 mg neostigmine and 1.2 mg atropine, given over five minutes. After tracheal extubation, the patient was observed for 30 min in the operating theatre before transfer to the intensive care unit (ICU) for overnight observation. Analgesia was provided by patient-controlled analgesia using morphine.

TABLE Perioperative hemodynamic measurements

	BP (mm Hg)	PAP (mm Hg)	PCWP (mm Hg)	CVP (mm Hg)	CO ($\text{l}\cdot\text{min}^{-1}$)	HR (min^{-1})
Pre-induction	110/60	70/47	36	25	3.7	95
Post-induction	108/55	68/43	35	24	3.6	88
Delivery	114/70	67/45	37	25	2	90
Skin Closure*	112/68	60/46	38	27	2.6	87

*after dobutamine

BP Arterial blood pressure, PAP Pulmonary arterial pressure PCVVT Pulmonary capillary wedge pressure
CVP Central venous pressure CO Cardiac output HR Heart rate

In the ICU, the patient was hemodynamically stable. Dobutamine was stopped and she was transferred back to the labour ward observation area the next day. However, because of oliguria she was transferred to the coronary care unit where she stayed for 18 days while her clinical condition was optimized using dobutamine, furosemide, digoxin, potassium and warfarin. Captopril was started but replaced with losartan after two days because of intractable cough. Echocardiography showed ejection fraction to be 30%. Warfarin was started on the second day after surgery. The low molecular weight heparin, which had been continued since 18 wk gestation, was stopped on the fifth day after surgery after the international normalized ratio (INR) was confirmed to be greater than 2.0. Three weeks after surgery, the patient was discharged. Warfarin was continued after discharge and intermittent monitoring showed the INR to be consistently greater than 2.0. However, two weeks after discharge, the patient suffered a severe embolic stroke. Her cardiac function continued to deteriorate and she died five months later in a convalescent hospital.

Discussion

Our patient was diagnosed as having idiopathic dilated cardiomyopathy as she became symptomatic during early pregnancy and there was no identifiable cause for her heart disease. She did not conform to the accepted diagnostic criteria for the better known entity of peripartum cardiomyopathy; these criteria were established by Demarkis in 1971² and include: (1) development of heart failure in the last month of pregnancy or within the first five postpartum months, (2) absence of a determinable etiology, and (3) absence of demonstrable heart disease before the last month of pregnancy. More recently, it has been suggested that echocardiographically demonstrable impairment in left ventricular function should be added as a fourth criterion.³

The obstetric, anesthetic and medical management of peripartum cardiomyopathy has been described previously, but dilated cardiomyopathy first presenting in early pregnancy is unusual. Although the clinical features of our patient resembled those of peripartum cardiomyopathy, the early presentation gave rise to a number of additional clinical considerations. It was anticipated that the patient's condition might deteriorate as the magnitude of the hemodynamic changes of pregnancy increased as the pregnancy progressed.⁴ Therefore, termination of pregnancy was offered, but this was declined by the patient and her husband who requested to continue the pregnancy until the fetus had a good chance of viability. Management therefore focussed on medical treatment to optimize her heart function until planned Cesarean delivery. In a previous

report, Mazor *et al.* described a 34 yr old patient with idiopathic dilated cardiomyopathy who first presented at 21 wk gestation with dyspnea and tachycardia.¹ Similar to our case, their patient also refused termination of pregnancy and was managed conservatively with medical treatment. Gestation was allowed to continue to 36 wk when labour was induced. She eventually proceeded to Cesarean section because of slow progress of labour but, in contrast to our case, regional anesthesia was used. Both the neonate and the parturient survived the operation without reported morbidity.

The optimum anesthetic technique for Cesarean section patients with peripartum or other forms of congestive cardiomyopathy is controversial and both general anesthesia^{5,6} and regional anesthesia^{7,8} have been described. Brown *et al.* described the use of general anesthesia because they feared catastrophic effects of reduction in systemic vascular resistance caused by epidural blockade⁵. Slowly titrated epidural anesthesia has been recommended as it avoids the use of cardiodepressant drugs and may improve myocardial performance by reducing left ventricular afterload.^{7,8} Mellor and Bodenham considered both methods dangerous and described the use of infiltration anesthesia supplemented with bilateral ilio-inguinal nerve block and intravenous fentanyl.⁹ We believe that close intra-operative and postoperative monitoring may be more important than the choice of anesthesia.

Patients with idiopathic dilated cardiomyopathy are at risk of thromboembolism¹⁰ and, therefore, the use of thromboprophylaxis has been recommended.¹¹ Unfortunately, our patient suffered a severe embolic stroke at home despite the use of preoperative low-molecular weight heparin and postoperative warfarin which highlights the importance of this complication. Use of thromboprophylaxis has important implications for the anesthesiologist. Epidural and spinal anesthesia are contraindicated during full anticoagulation and there has been recent concern about the risk of spinal hematoma after epidural analgesia in patients receiving low molecular weight heparin.^{12,13} This was a consideration in our decision to use general anesthesia in our patient.

In summary, idiopathic dilated cardiomyopathy may rarely present in early pregnancy. Important aspects in management include a multidisciplinary approach and consideration of termination of pregnancy. Close peripartum monitoring is required and thromboembolic complications are a major risk.

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