

Congestive heart failure in pregnancy: a case of peripartum cardiomyopathy

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ABSTRACT

A healthy 28-year-old woman developed full-blown pulmonary oedema in the 36th week of gestation. Echocardiography revealed a globally enlarged heart with reduced systolic function. A remarkable clinical response with regain of normal ventricular function was noted with early medical intervention. This case report illustrates peripartum cardiomyopathy, a unique form of dilated cardiomyopathy affecting women during/following gestation. Clinician familiarity with this entity increases the probability of prompt appropriate treatment, offering patients the best possible prognosis.

INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a rare and potentially life-threatening complication of pregnancy whose underlying cause remains unknown. An uncommon form of dilated cardiomyopathy, this disorder ultimately results in congestive heart failure late in pregnancy or in the early puerperium. Its natural history is extremely variable, ranging from the spontaneous recovery of ventricular function to refractory disease often necessitating cardiac transplantation. In recent studies, the reported incidence of death or cardiac transplantation were in the range of 12 to 18%, compared with a mortality rate of up to 50% reported in the 1980s. As early intervention is believed to improve prognosis, clinician familiarity with PPCM is essential, thus ensuring timely and optimal treatment to women stricken with PPCM.

CASE REPORT

A 28-year-old primigravid Ghanese woman with an unremarkable previous medical history presented in the 36th week of gestation with respiratory failure. She was mechanically ventilated and an emergency caesarean section was performed. Following surgery the patient was haemodynamically stable and was admitted to the intensive care unit. Physical findings included distension of neck veins, rapid heart sounds, an S3 gallop, a grade 2/6 blowing apical systolic murmur radiating to the axilla, bilateral pulmonary rales, and bilateral pitting oedema of the lower limbs. Laboratory tests including a complete blood count, chemistry profile, coagulation tests and D-Dimers level, erythrocyte sedimentation rate (ESR), thyroid function tests and urinalysis were all in the normal range. The electrocardiogram was interpreted as normal sinus rhythm with no signs of acute ischaemia. The chest X-ray revealed an enlarged cardiac silhouette and pulmonary congestion (*figure 1*). Haemodynamic characteristics are shown in *table 1*.

Once stabilised, the patient was transferred to the department of internal medicine for further evaluation and treatment. Echocardiography revealed an enlarged left ventricular end-diastolic diameter (LVEDD) of 59 mm (normal 46 mm +/- 4), left ventricular end-systolic diameter (LVESD) of 44 mm (normal 30 mm +/- 4), and poor global contraction with a shortening fraction (SF) of 26% (normal 34 to 44%). No valvular abnormalities were seen and regional systolic dysfunction was not detected. A ventilation perfusion scan, duplex imaging of the lower limbs and fundoscopy were interpreted as normal. Conventional treatment for heart failure with sodium restriction, digoxin, diuretics, and vasodilator agents (angiotensin-converting

Table 1
Measured haemodynamic characteristics

	MEASURED VALUE	NORMAL MEAN VALUE REFERENCE (NORMAL RANGE IN BRACKETS)*
Mean pulmonary capillary wedge pressure	30 mmHg	9 mmHg (4-12 mmHg)
Mean pulmonary artery pressure	37 mmHg	9-18 mmHg (systolic 15-30 mmHg, diastolic 4-12 mmHg)
Cardiac index	2.4 L/min/m ²	2.8-4.2 L/min/m ²
Systemic vascular resistance	790 dyne/sec ⁻¹ /cm ⁵	1100 dyne/sec ⁻¹ /cm ⁵ (700-1600 dyne/sec ⁻¹ /cm ⁵)
Pulmonary vascular resistance	176 dyne/sec ⁻¹ /cm ⁵	70 dyne/sec ⁻¹ /cm ⁵ (20-130 dyne/sec ⁻¹ /cm ⁵)

* Normal value references are based on: Armstrong WF, Feigenbaum H. Echocardiography. In: Braunwald E, Zipes DP, Libby P, eds. *Heart Disease: a Textbook of Cardiovascular Medicine*. 6th edn. Philadelphia, Pennsylvania: WB Saunders Company, 2001:160-236.¹³

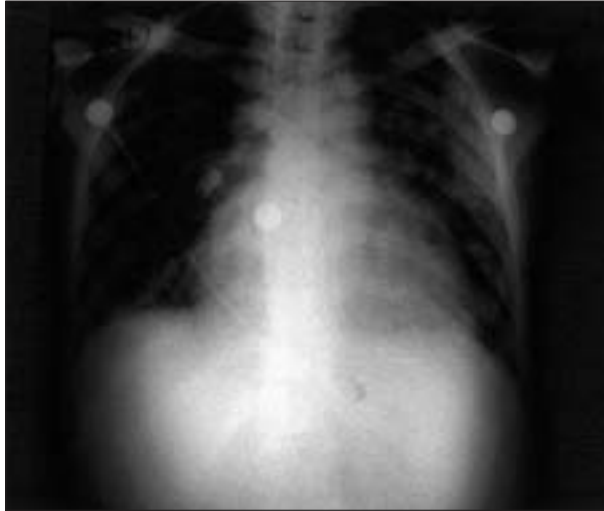


Figure 1
Chest X-ray on admission demonstrating an enlarged cardiac silhouette and pulmonary congestion



Figure 2
Chest X-ray after three months demonstrating resolution of cardiac enlargement after treatment

enzyme inhibitor) was initiated, with a dramatic improvement clinically, from NYHA functional class 4 to NYHA functional class 2 at the time of discharge. At a follow-up visit three months later, the patient was asymptomatic, with a normal chest X-ray (*figure 2*) and normalisation of the former echocardiography abnormalities (LVEDD 50 mm, SF 42%).

DISCUSSION

A number of disorders may cause heart failure in gravid women with no underlying heart disease. The coincidence of pregnancy and heart failure raises the possibility of high output heart failure, yet echocardiography findings of poor left ventricular contractility minimised the probability of this diagnosis. Myocarditis, often responsible for heart failure in pregnancy, is a possible aetiology, and several studies have demonstrated its occurrence in >50% of women with PPCM who underwent endomyocardial biopsy.¹ However, in the presented case myocarditis was considered

unlikely due to the absence of a recent febrile illness history, the normal ESR and creatine phosphokinase values, and the negative serology for autoimmune markers and for infectious agents (viral and bacterial). However, in the current case endomyocardial biopsy was not performed, making definitive exclusion of myocarditis impossible. Given the clinical findings and the exclusion of other potential diagnoses, PPCM seemed the most probable diagnosis. In 1971, Demakis *et al.*² established the diagnostic criteria for peripartum cardiomyopathy which include:

1. Development of heart failure in the last month of pregnancy or within five months of delivery;
2. The absence of another identifiable cause for heart failure;
3. The absence of a recognisable heart disease prior to the final month of pregnancy.

Subsequently, a fourth echocardiography criterion was added: left ventricular dysfunction, as manifested by depressed shortening fraction or ejection fraction.³

The true incidence of PPCM is unknown. In the USA it is estimated that PPCM affects 1000 to 1300 women per year.³ Multiparity, advanced maternal age, multifoetal pregnancy,

toxaemia, and Afro-American descent have been identified as risk factors for PPCM. While the underlying pathophysiological process has yet to be elucidated, theories attempting to explain the pathogenesis of PPCM include abnormalities in the serum level of relaxin, deficiency of selenium, the presence of stress-induced proinflammatory cytokines, an abnormal immune response with high titres of autoantibodies reacting against cardiac tissue proteins,³ and underlying myocarditis.^{1,4} To date, the aetiology of this rare cardiomyopathy is unknown. In contrast to heart failure in gravid women with an underlying heart disease, patients with PPCM present toward the end of gestation or after delivery. Common symptoms include chest pain, dyspnoea, orthopnoea and cough. Echocardiography assessment provides the ultimate diagnosis, and the management of PPCM is based on conventional therapy for heart failure, including oxygen supplementation, sodium restriction, diuretics, digitalis and vasodilator agents. Angiotensin-converting enzyme inhibitors (ACE inhibitors), vasodilator agents commonly used in the treatment of heart failure, are absolutely contraindicated during pregnancy because of the potential of prenatal and postnatal developmental disorders. These disorders include oligohydramnios, intrauterine growth retardation, neonatal renal failure, congenital structural defects (i.e. skull, skeleton, lungs), and early postnatal death.⁵ Given the lack of data regarding the use of β -blocking agents in PPCM, these drugs should be considered second-line drugs, preferentially for use after delivery.³ The risk for thromboembolic events has been reported in PPCM patients. As this complication might occur in as many as 50% of patients,⁶ and in particular in those with severely depressed left ventricular ejection fraction,³ the consideration of anticoagulation treatment in adjunct to standard heart failure management is recommended. Endomyocardial biopsy is recommended only when such therapy fails to yield improvement.³ For women who fail maximal medical management, the remaining option is cardiac transplantation, with a 60% five-year survival rate.⁷ In contrast to earlier data estimating mortality rates ranging from 25 to 50%,⁸ Felker *et al.*⁹ reported a five year-survival rate of 94%. Despite these encouraging statistics, there remains a small subset of women whose disease follows a rapid and irreversible course with death resulting from an arrhythmia, thromboembolic complications, and ultimately pump failure occurring within three months of diagnosis. Although there is no consensus regarding the risk of relapse in future pregnancies, cardiac function is ultimately predictive of the patient's prognosis. The persistence of cardiac dysfunction beyond six months, seen in an estimated 50% of cases, usually indicates an irreversible disease process. Elkayam *et al.*¹⁰ found that 21% of the women who regained normal ventricular function suffered heart failure during subsequent pregnancies reinforcing

earlier findings of Lampert *et al.*¹¹ Hence, an event-free future pregnancy, even in women with recovered cardiac function, cannot be guaranteed. Decision-making is perhaps more clear-cut for the group of women suffering persistent heart failure. With a mortality rate of 8 to 17%,¹² subsequent pregnancies in this group should be discouraged. In conclusion, we present a patient who was diagnosed with peripartum cardiomyopathy, primarily by exclusion of other possible diagnoses. PPCM is a clearly defined entity of a yet unknown aetiology and a potentially lethal complication of pregnancy. Clinician familiarity increases the probability of prompt and appropriate treatment, offering patients the best possible prognosis.

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