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Case report



# A Case of Perinatal Cardiomyopathy

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## Abstract

Background: Peripartum cardiomyopathy (PPCM) is rare heart disease that threatens the lives of pregnant women.
Patient: In the current report, we reported a case of a 40-week pregnant patient with PPCM.
Diagnoses: The patient was diagnosed with PPCM.
Intervention: The patient underwent emergency cesarean section and received other supportive treatments, mainly including cardiotonic, diuretic, bromocriptine, hemodialysis, improvement of cardiac function, blood-pressure-lowering, hepatic protection, supplementing albumin, delactation, promoting uterine contraction, the noninvasive ventilator-assisted ventilation improved oxygen delivery and CRRT reduced cardiac load.
Outcomes: The relative clinical symptoms have significantly become better after 58 days treatment and the patient has discharged.
Lessons: This case emphasized the significance of early diagnosis of PPCM in pregnant women.

## INTRODUCTION

Peripartum cardiomyopathy (PPCM) is idiopathic cardiomyopathy and its pathogenesis is little unknown. In 2010, the European Society of Cardiology (ECS) defined PPCM as a heart failure (HF) related disease secondary left ventricular (LV) systolic dysfunction towards the end of pregnancy or in the months following delivery. The diagnosis of PPCM needs to exclude other reasons of HF. The manifestations of PPCM include LV systolic dysfunction, LV ejection fraction (LVEF) less than 45%, accompanied with or without LV dilatation [1]. At present, the recognized risk factors of PPCM patients include advanced age pregnancy (more than 30 years old), polyembryony, Afro-Caribbean, malnutrition and hypertensive disorder complicating pregnancy [2]. The majority patients spontaneously recovered after delivery, while some patients may need cardiac transplantation [3]. Herein, a case of PPCM with evident improvement of symptoms following multidisciplinary collaborative and comprehensive supportive treatment in our center is reported.

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## CASE DESCRIPTION

### Characteristics of patient

The patient, a 37-year-old pregnant woman, without previous history of organic heart disease, was admitted on March 20, 2018 because of "34+6 weeks of menopause and 1 week of dyspnea after activity". Her childbearing history was 1-0-2-1. One week before admission, the patient presented dyspnea after activity, orthopnoea at night, accompanied with edema lower limbs, nausea and vomiting after eating, which were gastric contents. High blood pressure (BP) accompanied with renal dysfunction was found at 12 weeks of gestation. During gestation, the highest BP was 162/112 mmHg; the urine protein was 4+; the level of serum creatinine was 286 μmol/L. B-ultrasonography of urinary system indicated enhanced echo and multiple crystallization of renal parenchyma. The patient orally administrated labetalol regularly to reduce BP.

### Physical examination on entry

Patient was examined thoroughly after admission and following indexes were recorded on physical examination.
BP: 120/83 mmHg; shortness of lungs breath sounds;

without wet/dry vacuum noise; the cardiac percussion revealed that the border of cardiac dullness slightly extended to the left; heart rate: 106 beat/min with regular rhythm; no pathological noises were detected in each valve area; no touch between liver and spleen; no edema of lower limbs; NT proBNP: more than 25000 pg/mL; blood biochemistry: $K^+$-5.55 mmol/L, creatinine-11 μmol/L and albumin-25.3 g/L; chest radiograph showed a dilatation of the heart shadow; echocardiography (UCG): whole heart dilation, LV end-diastolic diameter (LVEDd): 60.6 mm, systolic function of LV: decreased, the rejection fraction (EF): 34.8%, moderate-severe mitral regurgitation, mitral ring dilatation, the middle of interatrial septum shunting from left to right and patent foramen ovale or reopening.

### Clinical diagnosis and intervention

The patient was diagnosed with suspected peripartum cardiomyopathy (PPCM; grade IV of Cardiac function), renal insufficiency, hyperpotassemia and hypoalbuminemia. The patient was given cardiotonic, diuretic and other supportive treatments. After the contraindication of operation was eliminated, the woman gave birth to a child by emergency caesarean section on the day after admission and then was conducted postoperative ICU monitoring. Examination showed high creatinine, high serum potassium and oliguria after ICU entry. The patient was given hemodialysis, improvement of cardiac function, blood-pressure-lowering, hepatic protection, supplementing albumin, delactation, promoting uterine contraction and other treatments. After medically stable, the patient was transferred to the Department of Nephrology for further treatment and maintained regular hemodialysis.

Tenth day after admission, the patient reappeared unable to prostrate at night, orthopnoea, and presented stuffiness and dyspnea under quiescent condition. After multidisciplinary discussion in our hospital, the patient was diagnosed with PPCM after considering her heart dilatation and the grade IV cardiac function and excluding other causes of heart failure (HF). There were no significant improvements of cardiac function after cardiotonic and diuretic therapies. Thus, the patient was transferred to ICU on the same day and given treatment combined the noninvasive ventilator-assisted ventilation improved oxygen delivery+CRRT reduced cardiac load. Patient was treated with bromocriptine to black prolactin. Meanwhile, she received cardiotonic, diuretic, antiarrhythmic therapies and other comprehensive treatments. Two weeks after treatments, the symptoms of chest tightness and shortness of breath have been improved; the patient was able to prostrate at night; the CCRT treatment was stopped; the patient was transferred to nephrology department again and given regular hemodialysis. On April 28, 2018, the symptoms of chest tightness, shortness of breath and edema of lower limbs were markedly improved, and the patient was allowed to discharge from hospital. Re-examination of UCG showed that EF value was still low; the LV dilatation was still obvious and there were no notably improvement compared with before. Since discharged, the patient received regular hemodialysis in the outpatient department of nephrology and no dyspnea occurred again. On September 1, 2018, re-examination of UCG in outpatient follow-up revealed EF-32%, LV dilatation; left atrium size-56.6 mm×43.6 mm×40 mm, increase of LV intraventricular trabecular structure, mild regurgitation of mitral and tricuspid.

## DISCUSSION

### Incidence and risk factors

PPCM is a rare cause of HF in pregnant women. According to reports, the incidences of PPCM were both low at abroad and home. At abroad, the incidence of PPCM ranges from 1/15000 to 1/300 [1, 4], while the range of PPCM incidence at home was 2.1%-12.2% in the past 14 years between 1986-2000 [5]. As many studies reported, PPCM is a multifactorial disease and the advanced age pregnancy (more than 30 years old), polyembryony, multifetal pregnancy, African American race, malnutrition and hypertensive disorder complicating pregnancy are both the recognized risk factors for PPCM currently [2, 6, 7]. In addition, smoking, alcoholism and cocaine abuse may be also the risk factors for PPCM [8]. Although the epidemiology of PPCM has been reported in some literatures, more prospective and systematic studies of it are needed.

### Pathogenesis of PPCM

Although the exact pathogenesis of PPCM remains not fully elucidated, many relevant hypotheses have been proposed. For examples, studies revealed that virus infection and autoimmunity of heart could be a potential pathogenesis of PPCM [9, 10]; the occurrence in women with mothers or sisters might indicate the genetic roles in the pathogenesis of PPCM [11]. Additionally, many researchers found myocarditis in PPCM patients by endocardial myocardial biopsy (EMB) [12]. Studies also observed that many inflammatory factors were increased in PPCM patients [13, 14], that revealed that inflammation might involve in the pathogenesis of PPCM. Recently, plenty of convincing evidences indicated that the cascade reactions of increase of oxidative stress, prolactin splitting into N-terminal 16kDA prolactin fragment (16K PRL) and impairment of vascular endothelial growth factor signal are implicated in the development of PPCM [7, 13, 15]. Although there are many hypotheses about the

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pathogenesis of PPCM, the precise mechanisms are still unknown and more studies are required to provide more direct and convincing evidences.

## Clinical presentation

PPCM patients usually have no organic heart disease or lack of any signs of heart disease before pregnancy and mainly present clinical symptoms towards the late pregnancy or five months after delivery. The main clinical symptoms of PPCM are similar to those of the LV systolic HF caused by other reasons. The most common manifestation of patients is dyspnea and other common symptoms are typical symptoms and signs of LV congestive HF, including dyspnoea on exertion, orthopnoea, paroxysmal nocturnal dyspnea, pedal edema and coughing up pink frothy sputum. Physical examination found increased jugular venous pressure, cardiac dilatation, transverse or downward displacement of apex cordis, moist rales of lung, tachycardia, third heart sounds, hepatomegaly and edema of lower limbs in PPCM patients [16]. In the advanced stage of PPCM, patients may present the signs of right HF such as jugular vein distention, liver congestion and serosal effusion. In addition, PPCM patients whose LVEF is less than 35% may even present the signs and symptoms of arterial thromboembolism, such as cerebral embolism, pulmonary embolism, acute myocardial infarction, mesenteric arterial embolism, renal infarction and splenic infarction [11, 17, 18]. Hence, appropriate strategies should be taken to assess and prevent the risk of various thrombo-embolisms.

## Diagnosis of PPCM

The key for the diagnosis of PPCM is the result of UCG which manifests as LV dilatation, decrease of contractile force and regurgitation of mitral and tricuspid [19]. Besides, other auxiliary examinations are also helpful in diagnosing PPCM, including laboratory examinations (electrolyte, creatinine, hemoglobin, hepatic enzyme and thyroid hormones), electrocardiography (ECG), chest radiography, cardiac magnetic resonance (MRI), cardiac catheterization, angiography and troponin T. For examples, ECG results of PPCM patients showed nodal tachycardia, ST-T wave abnormalities and prolonged interval between PR and QRS [20]. Chest radiography usually revealed heart shadow enlargement, pulmonary edema and congestion, and pleural effusion can be seen in severe patients. The shape and size of heart, LV systolic function and LV volume can be determined by MRI [21]. Taken together, diagnosis of PPCM should comprehensively consider the results of various examinations.

The difficulty of diagnosis is to exclude other reasons of cardiac dilatation and HF, such as ischemic cardiomyopathy, hypertensive cardiopathy, valvular heart disease, infection, intoxication, metabolic cardiomyopathy, pulmonary embolism and hyperthyroidism [1]. In the current case, the patient was diagnosed with gestational hypertension in the early stage. In fact, long-term continuous hypertension can result in myocardial remodeling and LV hypertrophy which includes interventricular septum thickness (IVST) or LV posterior wall thickness (LVPWT) ≥12 mm. However, the IVST and LVPWT of the patient were both normal. Considering the clinical manifestations and examination results, the patient was finally diagnosed with PPCM combined with gestational hypertension rather than HF solely caused by hypertension.

## Management of PPCM

The traditional drug therapies of PPCM include angiotensin-converting enzyme-inhibitors [22], b-Blockers [23], diuretics [24], aldosterone antagonists [25], bromocriptine [26] and antithrombotic. However, it should be noted that drugs need to be chosen more carefully and as far as possible to avoid drugs that affect the development of the fetus or babies because of the particularity of PPCM patients. The mechanical circulation supports such as intra-aortic balloon pump (IABP), extracorporeal membrane oxygenation (ECMO) or LV-assisted device (LVAD) can be considered when the traditional therapies don't work well [27]. In addition, extracorporeal membrane oxygenation is effective for refractory cardiogenic shock [28]. Also, anticoagulant therapy should be used when the PPCM patients present intracardiac thrombosis or systemic embolism. Finally, patients can undergo heart transplantation when the most treatments are ineffective, but the rejection rate and mortality of heart transplantation are relatively high [29]. Therefore, appropriate treatments should be taken according to the actual situation of patients.

Since most patients are pregnant women, the timing and manner of delivery during treatment is also important. PPCM patients who have delivered but not breastfeeding can be given standard treatments of HF. There would be no need for early childbirth if the the condition of the mother and the fetus were both stable [30]. On the contrary, for patients with advanced HF with hemodynamic instability, early delivery is required, and cesarean section is preferred [31]. Hence, clinical decision of timing and manner of delivery should be made jointly by multiple disciplines (cardiology, obstetrics, pediatrics and anesthesiology) according to the conditions of patients.

## Clinical outcomes

The clinical outcomes of PPCM patients are different. Most patients have a good prognosis and their heart function can recover 6 months after delivery at various degrees, while some patients develop to irreversible HF or even death in the advanced stage. Previous studies

have reported different mortality of PPCM. For instances, Elkayam et al found the mortality of PPCM was less than 10% [32]. Witlin et al showed a higher mortality (18%) [33], and Sliwa et al reported a much higher mortality (26%) [9]. A study including 100 PPCM patients revealed that 91% of patients with LVEF >30% and LVEDd < 6.0 cm recovered completely within 1 year after delivery, while those with LVEF < 30% and LVEDd > 6.0 cm had a recovery rate of 0% [34]. another study demonstrated that the patients with persistent LV dysfunction or LVEF ≤ 25% have a high risk of recurrent PPCM [1]. That indicated that the prognosis of PPCM is closely related to the recovery of ventricular function.

## CONCLUSION

PPCM is an idiopathic cardiomyopathy associated with pregnancy and its diagnosis needs to exclude other causes of cardiac dilatation and HF. The etiology of the disease is still unknown. The incidence is relatively low and clinical outcomes also vary widely. Since the similar symptoms to those of the normal third trimester, delay of seeing doctors and difficult for clinicians to identify them in the early stage, it requires a high level of diagnosis and treatment of clinicians. At present, the total number of domestic reports of the disease is not much. The current study reported a case of PPCM and concluded the epidemiology, pathogenesis, clinical manifestation, diagnostic approach, research progress of therapeutic method and clinical prognosis, in the hope of improving clinicians' understanding of the disease and early diagnosis and treatment in clinical work.

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