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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Recurrent and Life-Threatening Peripartum Cardiomyopathy

Diagnosis, Delivery Considerations, and Management

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ABSTRACT

Peripartum cardiomyopathy is an idiopathic reduction in left ventricular systolic function (ejection fraction <45%) toward the end of pregnancy or in the months after delivery. A multidisciplinary approach to management with shock team support is key to identifying and adequately treating patients with refractory heart failure and peripartum cardiomyopathy. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:681–4) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 38-year-old African-American woman, gravida 6 para 3, with a history of peripartum cardiomyopathy (PPCM) diagnosed 10 years earlier during her second pregnancy, severe mitral regurgitation (MR), and hypertension presented to the heart failure clinic at 23 weeks' gestation. Since her last episode of PPCM, the patient was advised against further pregnancies but did not undergo permanent sterilization. In the clinic, she was asymptomatic and appeared euvolemic. A transthoracic echocardiogram (TTE) from 3 years earlier showed a dilated, hypokinetic left ventricle (LV) with an ejection fraction (EF) of 20% and severe MR. A repeat TTE was recommended but not completed. At 36 weeks, she presented with worsening orthopnea, dyspnea, weight gain, and bilateral lower extremity edema acutely over 2 weeks. Vital signs on presentation included blood pressure of 145/81 mm Hg, tachycardia, tachypnea, and hypoxia requiring 4 L of oxygen via nasal canula. Admission electrocardiogram showed sinus tachycardia, biatrial enlargement, and LV hypertrophy.

LEARNING OBJECTIVES

- To review the epidemiology, outcomes, and risk factors for recurrence of peripartum cardiomyopathy.
- To provide a summary of the latest guidelines regarding the medical management, delivery considerations, and hemodynamic support of patients with PPCM.

DIFFERENTIAL DIAGNOSIS

The most likely diagnosis was decompensated systolic/diastolic heart failure due to PPCM. Other diagnoses included other forms of cardiomyopathy (viral, stress, and ischemic), pre-eclampsia (PEC) causing pulmonary edema, valvular heart disease, and myocardial infarction.

INVESTIGATIONS

Initial laboratory test results showed troponin I level of 0.14 ng/ml, brain natriuretic peptide level of 379

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, or patient consent where appropriate. For more information, visit the *JACC: Case Reports* author instructions page.

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ABBREVIATIONS AND ACRONYMS

CO = cardiac output

EF = ejection fraction

LV = left ventricle

MR = mitral regurgitation

PA = pulmonary artery

PEC = pre-eclampsia

PPCM = peripartum cardiomyopathy

TTE = transthoracic echocardiogram

pg/ml, and an elevated urine spot protein/creatinine ratio. Results of the remaining laboratory tests, including liver function test, were normal. Chest x-ray film showed cardiomegaly and pulmonary vascular congestion (Figure 1). TTE showed a severely dilated LV (LV internal dimension in diastole, 7.4 cm) with LVEF of 10%, dilated right ventricle, severe eccentric MR, estimated pulmonary artery systolic pressure of 73 mm Hg, and biatrial enlargement (Figure 2).

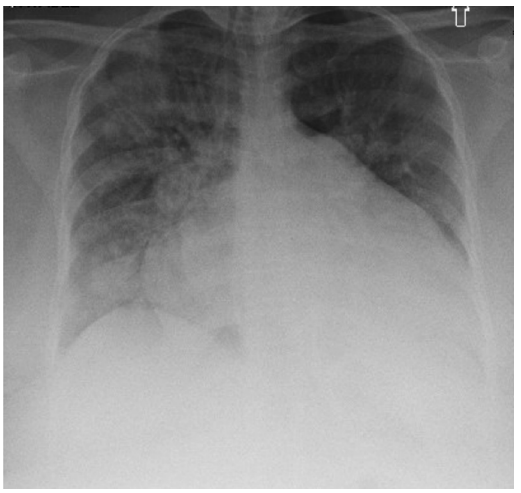
MANAGEMENT

The patient was admitted to the cardiac critical care unit and diuresed. To aid in delivery planning, the heart failure team recommended placement of a pulmonary artery (PA) catheter, which showed right atrial pressure of 13 mm Hg, right ventricle pressure of 65/28 mm Hg, pulmonary capillary wedge pressure of 30 mm Hg, PA pressure of 60/28/39 mm Hg with PA saturation of 76%, systemic vascular resistance of 1300 dynes/s/cm^{-5}, and Fick cardiac output (CO) and cardiac index of 5.23 l/min and 3.24 l/min/m^2, respectively. After multidisciplinary discussions, the maternal-fetal medicine team recommended a cesarean birth as the mode of delivery, and the heart failure team advised pre-surgery shock prophylaxis. The patient proceeded with a cesarean and bilateral tubal ligation after placement of femoral sheaths to prepare for emergent extracorporeal membrane oxygenation. Intraoperatively, the patient became hypotensive, requiring intravenous vasopressor support with norepinephrine and epinephrine. Post-operatively, she became acutely hypertensive, with systolic blood pressure of 200 mm Hg. Vasopressors were weaned, and she was started on nitroprusside, furosemide, and epoprostenol with stabilization of her hemodynamics.

DISCUSSION

PPCM is an idiopathic reduction in LVEF (EF <45%) during pregnancy or in the postpartum period in the absence of other etiologies (1). The incidence of PPCM in the United States is estimated to be approximately 1 in 2,500 to 4,000 live births (1). PPCM has serious risks for both morbidity and mortality, with mortality rates ranging from 5% to 25% (1–3). In the IPAC (Investigations of Pregnancy-Associated Cardiomyopathy) study, 13% of women developed major events (i.e., death, mechanical support, heart transplant) or failed to recover their LVEF (2). Notably, 5% of heart transplants in women in the United States are due to PPCM (4). Additionally, LVEF recovery does not eliminate the risk of recurrence: 20% of women, despite EF recovery, will have a re-exacerbation in subsequent pregnancies compared with 54% of women with persistent LV dysfunction (3).

FIGURE 1 Admission Chest X-Ray

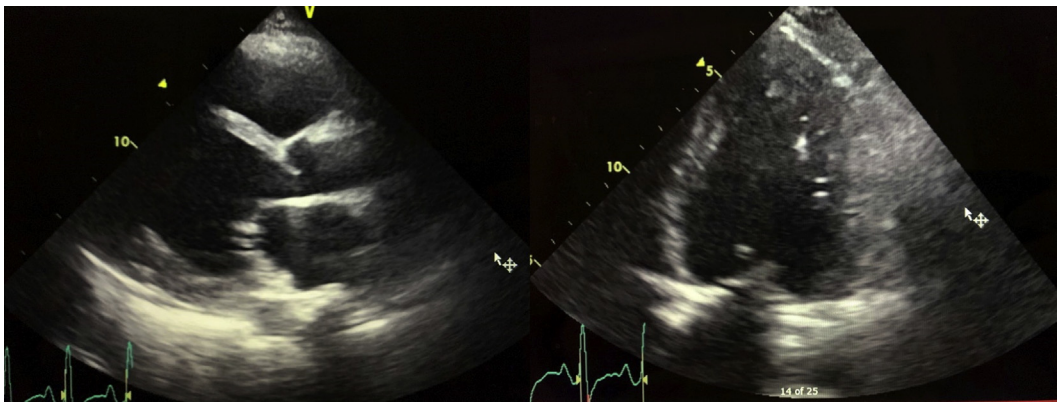


Admission chest x-ray film showing pulmonary vascular congestion and cardiomegaly.

The demands of pregnancy lead to significant changes in cardiovascular physiology. Through pregnancy, coupled with a decrease in systemic vascular resistance, CO increases 30% to 50% above pre-pregnancy levels by the second trimester and rises further with contractions and postpartum autotransfusion (5). In women with LV dysfunction, abnormal contractility prevents adaptation to the increased preload and CO associated with pregnancy, leading to elevated filling pressures and pulmonary edema (6). The pathogenesis of PPCM remains largely unknown; however, multiple known risk factors exist, including PEC, increased maternal age (>30 years), African American race, multiple gestations, and maternal hypertension (1,7). There is also a strong association between PEC and PPCM, with a shared pathophysiology that includes upregulation of placental antivascular factors such as soluble vascular endothelial growth factor receptor (sFlt-1) (7).

The management of PPCM relies on a multidisciplinary team approach focused on the hemodynamic stability of mother and fetus (Figure 3). Multiple factors require added attention when caring for patients with PPCM, including optimal timing, mode of delivery, and availability of mechanical support. Many cardiac regimens require adjustments during pregnancy because of teratogenicity. Angiotensin-

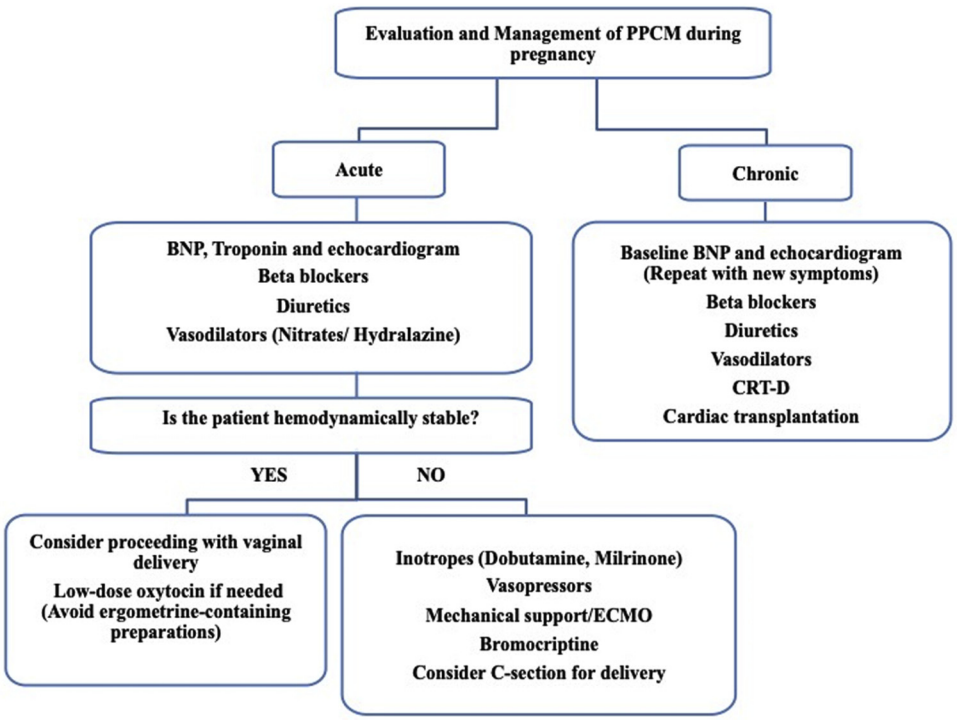
FIGURE 2 Transthoracic Echocardiogram Images



Transthoracic echocardiogram images showing dilated left ventricle.

converting enzyme inhibitors and angiotensin receptor blockers are both contraindicated during pregnancy because of teratogenic effects, and aldosterone antagonists are relatively contraindicated because of their anti-androgenic effect on male fetuses (5). Beta-blockers, especially metoprolol, are generally safe for use, and hydralazine and nitrates are utilized for afterload reduction during pregnancy (5). Diuretics are generally continued during pregnancy in the setting of acute or chronic heart failure,

FIGURE 3 Peripartum Cardiomyopathy Management



Summary of peripartum cardiomyopathy management. BNP = B-type natriuretic peptide; CRT-D = cardiac resynchronization therapy defibrillator; ECMO = extracorporeal membrane oxygenation.

pulmonary edema, or cardiogenic shock (5). Bromocriptine has been proposed to have beneficial effects on LV recovery, although clinical use remains limited because of the small size of these studies (1,3).

Currently, there are limited and conflicting data regarding the long-term management of patients with PPCM and recovered LV function (3). One study showed no deterioration in LV function during a 2-year follow-up period in 15 PPCM patients after discontinuation of beta-blockers and angiotensin-converting enzyme inhibitors/angiotensin receptor blockers (8). Conversely, the recently published TRED-HF (Withdrawal of pharmacological treatment for heart failure in patients with recovered dilated cardiomyopathy) trial showed that patients with dilated cardiomyopathy more frequently relapsed 6 months after discontinuing heart failure medications compared with those continuing treatment (44% vs. 0%, $p = 0.0001$) (9). Only 2 of the patients stopping treatment had PPCM, and 1 of the 2 relapsed (9). A suggested approach is to gradually discontinue heart failure medications in patients with PPCM after recovery, stabilization of LVEF, and normalization of LV size over several months, along with close serial echocardiographic monitoring during the discontinuation period (3).

In critically ill pregnant women, norepinephrine is a first-line agent for vasopressor support, although dopamine and dobutamine are alternatives in a low output state (5,10). In terms of mechanical support, intra-aortic balloon pump, LV assist devices, and extracorporeal membrane oxygenation have been used successfully in patients with low-output cardiogenic shock as a bridge to either recovery or transplant (2,3,10).

Decisions regarding the timing and mode of delivery require a multidisciplinary approach with accurate hemodynamic measurements. Although cesarean delivery is not required in all patients with PPCM, approximately 43% PPCM deliveries are via cesarean section (10). Vaginal delivery is preferred unless there is an obstetric emergency or hemodynamic contraindications indicating cardiac inadequacy or instability when attempting to meet the increase in preload/CO (1,10).

FOLLOW-UP

The patient was successfully extubated and discharged on furosemide, lisinopril, and metoprolol succinate based on the recommendations of the heart failure team. She did not plan to breastfeed. TTE at 6 months postpartum showed a severely reduced LV systolic function (EF 20% to 25%), dilated LV (LV internal dimension in diastole, 8.1 cm), with a diffusely hypokinetic LV. Given the persistent LV dysfunction, she is being evaluated for a heart transplant.

CONCLUSIONS

PPCM is a rare but serious condition associated with significant morbidity and mortality that remains poorly understood in terms of etiology and pathogenesis. A multidisciplinary approach is key to identifying and adequately treating patients with PPCM. Medical management should focus on controlling symptoms and preventing complications during pregnancy, with mechanical support reserved for patients with a low-output state and as additional support during pregnancy or the early postpartum period.

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KEY WORDS cardiovascular disease in women, delivery considerations, mechanical support, pregnancy, peripartum cardiomyopathy