



## Case Report

## Peripartum Cardiomyopathy (PPCM): How to Diagnose and Deal with?

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## ABSTRACT

**Background:** Peripartum cardiomyopathy (PPCM), a diagnosis of exclusion, is identified with the presentation of heart failure (HF) secondary to left ventricular (LV) systolic dysfunction without any other cause of HF recognized in the last month of pregnancy or within first five months after delivery, abortion, or miscarriage. It is a life-threatening condition that is frequently underdiagnosed and inadequately treated, whereas the morbidity and mortality rates range between 7% and 50%. Therefore, it is necessary to report the case related to this condition.

**Objective:** This case report aims to describe the importance of early diagnosis and treatment in PPCM

**Case Illustration:** A 34-year-old woman was referred to RSSA with worsening shortness of breath (SOB). She has given birth approximately 2.5 months before admission. History taking, clinical and ancillary findings revealed the diagnosis of PPCM. She was treated with a diuretic, aldosterone antagonist, ACE-I, beta-blocker, anticoagulant, and bromocriptine. The symptoms were improved in the following days. The patient was discharged in good condition and educated to comply with her medications.

**Conclusion:** Swift clinical judgment and related objective examinations are compulsory to establish a diagnosis and benefit the patient.

### 1. Introduction

PPCM is defined as idiopathic cardiomyopathy developing in the last month of pregnancy or within the first five months after delivery, abortion, or miscarriage. It should be under the condition that no other causes for heart failure are identified, with a left ventricular (LV) ejection fraction (EF) <45%.<sup>1,2</sup> There are several predisposing factors of PPCM including multiparity and multiple pregnancies, ethnicity, older age of mother, pre-eclampsia, extended utilization of tocolytic beta-agonists, smoking, diabetes, hypertension, malnutrition, and family history.<sup>1-3</sup>

Diagnosis establishment of PPCM obligates supporting echocardiographic findings of left ventricular dysfunction with LVEF <45% and oftentimes left ventricular dilatation in the absence of an alternative explanation, though the later is not always present. Other findings in echocardiography may also include left atrial or biatrial enlargement, right ventricular dilatation and dysfunction, pulmonary hypertension, functional mitral and tricuspid regurgitation, and intracardiac thrombus.<sup>2,3,7</sup>

A patient with PPCM was reported in this case. She has suffered from shortness of breath (SOB) while doing heavy activities since 2.5 months ago. She did not take any medical care. Because of worsening SOB, she brought to RSSA and diagnosed with PPCM. Her condition improved during care and discharged with appropriate condition.

### 2. Case Illustration

A 35-year-old primiparous woman referred to RSSA with aggravating SOB at rest since a day before admission. The SOB was followed by palpitation, cold sweating, and leg swelling. She complained that the SOB became worse in the last seven days. There were not any chest pain nor syncope. Antenatal care was routinely performed at the midwife. There was no history of hypertension before pregnancy, diabetes, heart disease, nor a family history of heart disease. The patient was obese, but she did not have a serious condition before admission. In the last month of pregnancy, a pre-eclampsia was developing. She gave birth through cesarean section 2.5 months ago due to pre-eclampsia. History of dyspnea on effort (DOE) while doing

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moderate to heavy activities, orthopnea (OE), and paroxysmal nocturnal dyspnea (PND) were found since two months ago. She did not ever seek any medical consultation at that time.

Physical examination revealed compos-mentis, blood pressure (BP) 130/85 mmHg, heart rate (HR) 125 beats per minute (bpm), respiration rate (RR) 24 times per minute (tpm), temperature (T) 36.3 °C, and saturation (SpO<sub>2</sub>) 99% on oxygen (O<sub>2</sub>) nasal cannula (NC) 4 litre per minute (lpm). Anaemia was not found. Jugular venous pressure (JVP) was R + 5 cmH<sub>2</sub>O. There were heart enlargement findings on auscultation with palpable apex cordis at Intercostalis (ICS) VI midclavicular (MCL) sinistra and found systolic murmur grade III/VI at the apex. There was also rales on the mediobasal bilateral lung.

White cell blood count was 10100/μL, Hb 10.3 g/dl, C reactive protein (CRP) 6.8 mg/dl (normal value < 0.3), Nt pro BNP 3164 pg/ml (normal value < 100 pg/ml). The electrocardiography (ECG) examination result showed sinus tachycardia with poor R wave progression, as seen in Figure 1. Chest x-ray (CXR) examination revealed cardiomegaly with congestive pulmonum (Figure 2). Echocardiography findings showed the presence of a reduced ejection fraction of left ventricular systolic function (EF 38%), fractional shortening (FS) of 11% along with mild mitral regurgitation and mild tricuspidal regurgitation. Left atrial and ventricle appeared enlarged and diffusely hypokinetic. End-diastolic volume of the left ventricle was 103 ml/m<sup>2</sup> and end-diastolic dimension 5.0 cm with an end-diastolic dimension index of 2.9 cm/m<sup>2</sup>.

The patient was diagnosed with moderate PPCM. She was medicated with loop diuretic therapy intravenously with adjusting dose according to congestion state, angiotensin-converting enzyme inhibitor (ACE-i), mineralocorticoid receptor antagonist (MRA) agent, and bromocriptine. Then, after congestion was improved, beta-blocker was initiated. In the following days, she received optimization of heart failure treatment. She routinely took controlled medication. After six months of medication, echocardiography examination was performed and showed the recovery of LV function.



Figure 1. Thoracic X-ray showed cardiomegaly with congestive pulmonum deviation and T waves inversion in precordial leads

### 3. Discussion

PPCM typically manifests itself as heart failure in the last month of pregnancy or several months after delivery or abortion where other underlying factors of heart failure is not identified. The condition is potentially fatal. The cause is largely not comprehended although it is often linked to various risk factors, such as multiparities, pre-eclampsia, maternal age  $\geq 30$  years old, and black race [6,11,12]. As found in this patient, a history of pre-eclampsia and older maternal age were found.

PPCM is frequently missed because of the prominent symptom, dyspnea, which is a common complaint in normal pregnancy, particularly when the patient does not fit the typical epidemiology [6,13]. According to this case, the patient had suffered from intermittent DOE, PND, OE, leg swelling, and palpitation ten days after the labour. Because there is no marked limitation on her daily activities, she did not seek any medical help. About two months later, the symptoms became worsen. It was accompanied by palpitation and leg swelling. Because of worsening symptoms, she was brought to the hospital and referred to RSSA. She was diagnosed with PPCM. Physical examination supported to history taking of the patient, including tachycardia, tachypnea, increased JVP, basal bilateral rales from lung auscultation, and shifted apex palpable at ICS VI MCL confirming the cardiomegaly, and leg swelling. Other supporting findings of PPCM in this patient were increased NT-pro BNP, cardiomegaly and congestive pulmonum from CXR, and systolic dysfunction of LV with decreased fractional shortening found from echocardiography. The course of symptoms and supporting patient findings were correlated with a diagnostic pathway in patients with moderate PPCM (Figure 2).<sup>6,9,12</sup>

The treatment adheres to the standard protocols for other types of systolic HF. Although the therapeutic plans for heart failure are well-entrenched, antepartum prescriptions need adjustments to avoid fetal toxicity. Moreover, the implications of breastfeeding infants also should be considered. The aims are to improve the hemodynamic status, minimize HF symptoms, and optimize long-term outcomes.<sup>9,12,13</sup> Related to this patient, she received intravenously diuretic during the first day of care until the congestion improving. She also got additional bromocriptine. According to the guideline, the BOARD regiment was given to this patient, including bromocriptine, oral HF drugs, anticoagulant, vasodilators, and diuretic agents. The role of these regiments was discussed in the following paragraphs.

Diuretics are useful to decrease the preload and relieve pulmonary congestion or peripheral oedema. Some of the most frequently-used diuretics such as furosemide and hydrochlorothiazide, are generally safe during pregnancy and subsequent lactation. The patient received furosemide to decongest the congestion state. Spironolactone, a potassium-sparing diuretic, has been used as one of heart failure treatment; however, the lack of sufficient data regarding its appliance in pregnancy urge for cautious use in that condition. Other drugs categories are contraindicated during pregnancy because of its teratogenic effect, such as angiotensin-converting enzyme inhibitors and angiotensin receptor blockers. Beta-blockers, administered along with ACE inhibitors, are the first-line heart failure option, reducing mortality through modulation of neurohormonal activation. To optimize heart failure therapy in this patient, she received those regiments, including MRA, ACE inhibitor, and beta-blocker agents<sup>6,9,12</sup>.

Based on the aforementioned pathophysiological process of 16kD-prolactin-mediated PPCM and supported by randomized studies, the addition of bromocriptine, a prolactin-blocker, to the standard regiment of heart failure treatment exerts favourable effect on LVEF

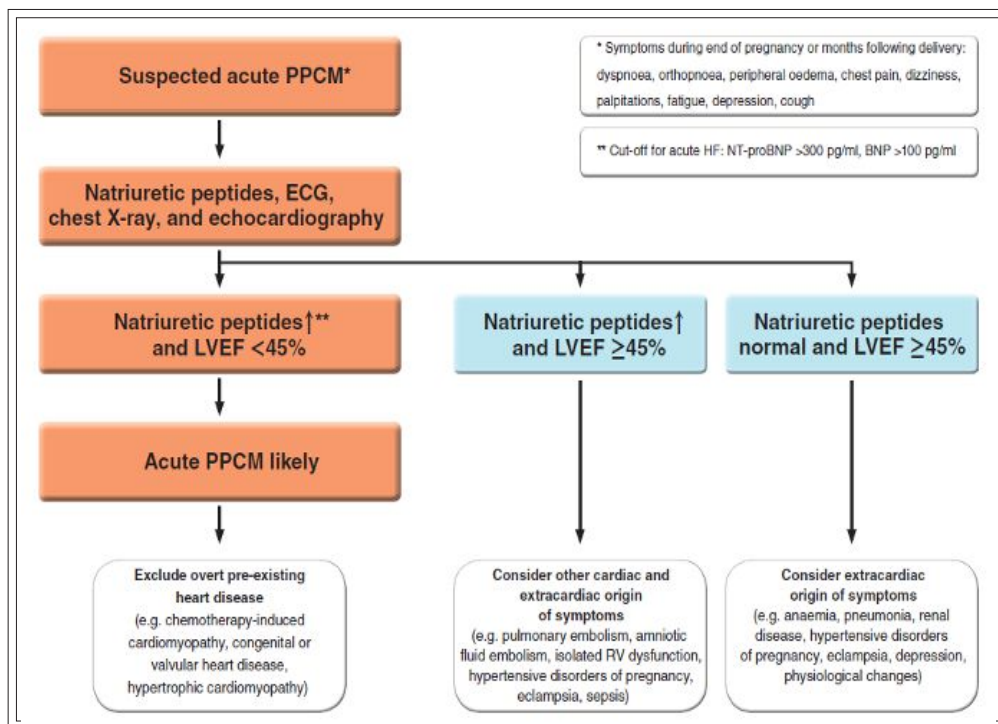


Figure 2. Diagnostic pathway in patients with suspected peripartum cardiomyopathy (PPCM). BNP, B-type natriuretic peptide; ECG, electrocardiogram; HF, heart failure; LVEF, left ventricular ejection fraction; NT-proBNP, N-terminal pro-B-type natriuretic peptide; RV, right ventricular.<sup>1</sup>

and mortality rate in severe acute PPCM cases. However, it is important to note that bromocriptine will effectively disrupt lactation and necessitate a discussion with the mother if this option is needed. Nonetheless, the evidence for bromocriptine efficacy in PPCM was derived from relatively small studies with many confounding variables. More data and better insight on the role of bromocriptine in PPCM may be provided from the on-going EURObservational Programme.<sup>9,12,13</sup> Anticoagulant is endorsed by the American Heart Association if EF < 30% during late pregnancy and up to 8 weeks post-partum. If bromocriptine is used, therapeutic anticoagulation should be considered.<sup>12,14</sup> Class recommendation of bromocriptine administration along with prophylactic (therapeutic) anticoagulation is IIa C.<sup>15</sup> Even though bromocriptine and anticoagulant agents, are included in the class IIa C recommendation from the guideline, she still received those regimens because of PPCM condition.

According to this case, the symptoms were improved in the following days with those regimens. She was discharged with the appropriate condition. PPCM patients and their partners should be carefully counselled and informed (class I recommendation) regarding the longer-term prognosis. In the patient with PPCM, subsequent pregnancy is not recommended if LVEF does not normalize (class III C recommendation). Information Education Communication (IEC) was given to the patient, including consuming a low-salt diet, eating a well-balanced diet, taking medicine, and controlling the doctor regularly.

A 6-month visit with echocardiography examination in PPCM is needed until the LV function is recovered to an LVEF >50%. In women with LV recovery who remain stable after tapering of heart failure drug therapy, an annual visit is recommended for up to 10 years. There is no consensus about whether heart failure medication can be stopped in women with a recovered LV function or subclinical dysfunction. Some of the PPCM Study Group members recommend life-long heart failure therapy at the highest tolerated dose based on the fact that

deterioration of LV function has been observed in women with normalized cardiac function. If patients display incomplete recovery signs despite recovered LVEF such as persistent LV dilatation or reduced myocardial strain, a continuation of heart failure drugs (ACE inhibitor, beta-blocker, MRA) should be considered. IEC was important to PPCM patient making good adherence and improving the survival rate subsequently.<sup>12,14,15</sup>

#### 4. Conclusion

We reported a case of a 34-year-old woman with moderate PPCM presentation. She received BOARD regimens treatment. The symptoms were improved the following day. She was discharged in good condition. She was advised to take regular control and consume the medication routinely. Echocardiography follows up in the sixth month after hospital discharge was advised to evaluate the LV function and direct future therapy.

#### 5. Declarations

##### 5.1. Ethics Approval and Consent to participate

Patient has provided informed consent prior to involve in the study.

##### 5.2. Consent for publication

Not applicable.

##### 5.3. Availability of data and materials

Data used in our study were presented in the main text.

##### 5.4. Competing interests

Not applicable.

##### 5.5. Funding source

Not applicable.

### 5.6. Authors contributions

Idea/concept: MS. Design: MS. Control/supervision: CT, HM, NK. Data collection/processing: MS. Extraction/Analysis/interpretation: MS. Literature review: CT, HM, NK. Writing the article: MS. Critical review: CT, HM, NK. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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## References

- Bauersachs, J., König, T., van der Meer, P., Petrie, M. C., Hilfiker-Kleiner, D., Mbakwem, A., & Mueller, C. 2019. Pathophysiology, diagnosis and management of peripartum cardiomyopathy: a position statement from the Heart Failure Association of the European Society of Cardiology Study Group on peripartum cardiomyopathy. *European journal of heart failure*, 21(7), 827-843.
- Hilfiker-Kleiner D, Sliwa K. 2014. Pathophysiology and epidemiology of peripartum cardiomyopathy. *Nat Rev Cardiol*. 11:364–370.
- Honigberg, M. C., & Givertz, M. M. 2019. Peripartum cardiomyopathy. *Bmj*, 364, k5287.
- Huang, Y., Chen, T., Zhang, M., Yang, X., Ding, G., & Yang, L. 2018. Successful management of fatal peripartum cardiomyopathy in a young pregnant woman: A case report. *Medicine*, 97(15).
- Isogai, T., & Kamiya, C. A. 2019. Worldwide incidence of peripartum cardiomyopathy and overall maternal mortality. *International heart journal*, 60(3), 503-511.
- Karafiatova, L., Lazarova, M., & Taborsky, M. 2017. Peripartum cardiomyopathy—A case report and concise review. *Cor et Vasa*, 59(3), e272-e276.
- Kearney, L., Wright, P., Fhadil, S., & Thomas, M. 2018. Postpartum cardiomyopathy and considerations for breastfeeding. *Cardiac failure review*, 4(2), 112.
- Khurana R, Bin Jardan YA, Wilkie J, Brocks DR. 2014. Breast milk concentrations of amiodarone, desethylamiodarone, and bisoprolol following short-term drug exposure: two case reports. *J Clin Pharmacol*. 54(7): 828–31. <https://doi.org/10.1002/jcph.272>. PMID:24482268.
- Kim, M. J., & Shin, M. S. 2017. Practical management of peripartum cardiomyopathy. *The Korean journal of internal medicine*, 32(3), 393.
- Patten IS, Rana S, Shahul S, Rowe GC, Jang C, Liu L, Hacker MR, Rhee JS, Mitchell J, Mahmood F, Hess P, Farrell C, Koulisis N, Khankin EV, Burke SD, Tudorache I, Bauersachs J, del Monte F, Hilfiker-Kleiner D, Karumanchi SA, Arany Z. 2012. Cardiac angiogenic imbalance leads to peripartum cardiomyopathy. *Nature*. 485:333–338.
- Ponikowski P, Voors AA, Anker SD, et al. 2016. 2016 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure: The Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC). *Eur Heart J*. 37(27):2129–200. <https://doi.org/10.1093/eurheartj/ehw128>. PMID:27206819.
- Prameswari, H. P., Dewi, T. I., Hasan, M., Martanto, E., & Aprami, T. M. 2018. Hypertension in pregnancy as the most influential risk factor for PPCM. *Br J Cardiol*, 25, 111-4.
- Sliwa K, Petrie MC, Hilfiker-Kleiner D, et al. 2018. Long-term prognosis, subsequent pregnancy, contraception and overall management of peripartum cardiomyopathy: practical guidance paper from the Heart Failure Association of the European Society of Cardiology Study Group on Peripartum Cardiomyopathy. *Eur J Heart Fail*. 20:951–62. <https://doi.org/10.1002/ejhf.1178>. PMID:29578284.
- Regitz-Zagrosek, V., Roos-Hesselink, J.W., Bauersachs, J., Blomström-Lundqvist, C., Cifkova, R., De Bonis, M., Iung, B., Johnson, M.R., Kintscher, U., Kranke, P. and Lang, I.M., 2018. 2018 ESC guidelines for the management of cardiovascular diseases during pregnancy: the task force for the management of cardiovascular diseases during pregnancy of the European Society of Cardiology (ESC). *European heart journal*, 39(34), pp.3165-3241.
- Davis, M.B., Arany, Z., McNamara, D.M., Goland, S. and Elkayam, U., 2020. Peripartum Cardiomyopathy: JACC State-of-the-Art Review. *Journal of the American College of Cardiology*, 75(2), pp.207-221.