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Peripartum Dilated Cardiomyopathy with End-Stage Left Ventricular Dysfunction Refractory to Medical and Electrical Therapy: A Case Report and Comprehensive Literature Review

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Abstract | **Case Report**

Background: Peripartum cardiomyopathy (PPCM) is a rare and potentially life-threatening form of dilated cardiomyopathy presenting in late pregnancy or early postpartum. Although many patients recover left ventricular function, a subset progress to end-stage heart failure refractory to guideline-directed medical and device therapy, requiring heart transplantation. ***Case presentation:*** We report the case of a 47-year-old multiparous woman with PPCM diagnosed during late pregnancy, who developed severe left ventricular dysfunction complicated by thromboembolic stroke and malignant arrhythmias. Despite optimal guideline-directed medical therapy and cardiac resynchronization therapy with defibrillator (CRT-D), she progressed to refractory NYHA class IV heart failure and was listed for heart transplantation. ***Conclusion:*** PPCM may evolve toward advanced heart failure despite optimized treatment. Early recognition, multidisciplinary care, and timely referral for advanced therapies including mechanical circulatory support and heart transplantation are essential in refractory cases.

Keywords: Peripartum cardiomyopathy; dilated cardiomyopathy; heart failure; transplantation; cardiac resynchronization therapy; thromboembolism.

INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a rare idiopathic cardiomyopathy defined by the development of heart failure secondary to left ventricular (LV) systolic dysfunction toward the end of pregnancy or within five to six months postpartum in women without preexisting structural heart disease or identifiable alternative causes of cardiomyopathy [1–4]. Diagnostic criteria include an LV ejection fraction (LVEF) <45%, often associated with LV dilatation, although nondilated phenotypes have also been reported [3,5].

The global incidence of PPCM varies widely, ranging from approximately 1 in 1,000 to 1 in 15,000 live births, with higher prevalence reported in Africa and parts of Asia [6–9]. Risk factors include multiparity, advanced maternal age, multiple gestation, preeclampsia, hypertensive disorders of pregnancy, African ancestry, and genetic susceptibility [2,6,10].

The etiopathogenesis of PPCM is multifactorial and incompletely understood. Proposed mechanisms include oxidative stress–mediated cleavage of prolactin into a cardiotoxic 16-kDa fragment, inflammatory and autoimmune responses, viral myocarditis, endothelial dysfunction, angiogenic imbalance, and genetic predisposition involving mutations in dilated cardiomyopathy-associated genes such as *TTN* [11–16].

Clinically, PPCM presents with nonspecific symptoms of heart failure with reduced ejection fraction (HFrEF), including dyspnea, orthopnea, peripheral edema, and fatigue [3,7]. Major complications include ventricular arrhythmias, thromboembolism, cardiogenic shock, and progressive end-stage heart failure [4,17]. Although many patients experience partial or complete recovery of LV function, approximately 10–20% progress to advanced heart failure requiring mechanical circulatory support or heart transplantation [6,18–20].

We report a case of severe PPCM evolving toward refractory end-stage heart failure despite optimal medical and device therapy, highlighting the role of transplantation as a definitive therapeutic option and providing a comprehensive review of the literature.

CASE PRESENTATION

A 47-year-old woman with no cardiovascular risk factors and no significant medical history was admitted in 2018 for progressive dyspnea and peripheral edema occurring during the final month of pregnancy. Her obstetric history included eight pregnancies with one live birth and seven prior intrauterine fetal demises. There was no history of hypertension, diabetes, alcohol consumption, or cardiotoxic drug exposure.

Transthoracic echocardiography revealed a dilated left ventricle with severely reduced systolic function (LVEF approximately 25%) and global hypokinesis. Coronary angiography showed normal coronary arteries. Cardiac magnetic resonance imaging confirmed dilated cardiomyopathy without late gadolinium enhancement suggestive of infiltrative or ischemic disease. A diagnosis of PPCM was established.

She was initiated on guideline-directed medical therapy (GDMT) for HFrEF, including beta-blockers, sacubitril/valsartan, mineralocorticoid receptor antagonists, and later sodium-glucose cotransporter-2 (SGLT2) inhibitors. Sacubitril/valsartan was discontinued due to intolerance.

Over the following years, the patient experienced multiple episodes of acute decompensated heart failure requiring hospitalization. In 2019, she developed a left middle cerebral artery ischemic stroke secondary to a left ventricular thrombus, successfully treated with thrombolysis and long-term anticoagulation with vitamin K antagonists. She subsequently developed recurrent supraventricular tachycardia and complete left bundle branch block with QRS duration of 150 ms, prompting implantation of a cardiac resynchronization therapy device with defibrillator (CRT-D) in 2021.

In December 2022, she presented with NYHA class IV dyspnea, orthopnea, and severe peripheral edema. Physical examination revealed signs of biventricular failure, including elevated jugular venous pressure, hepatomegaly, and massive lower limb edema. Pulmonary auscultation demonstrated diffuse crackles up to the apices. Electrocardiography showed a paced rhythm at 86 bpm with persistent conduction abnormalities. Chest radiography demonstrated cardiomegaly with pulmonary congestion and CRT-D leads in situ. (figure 1)

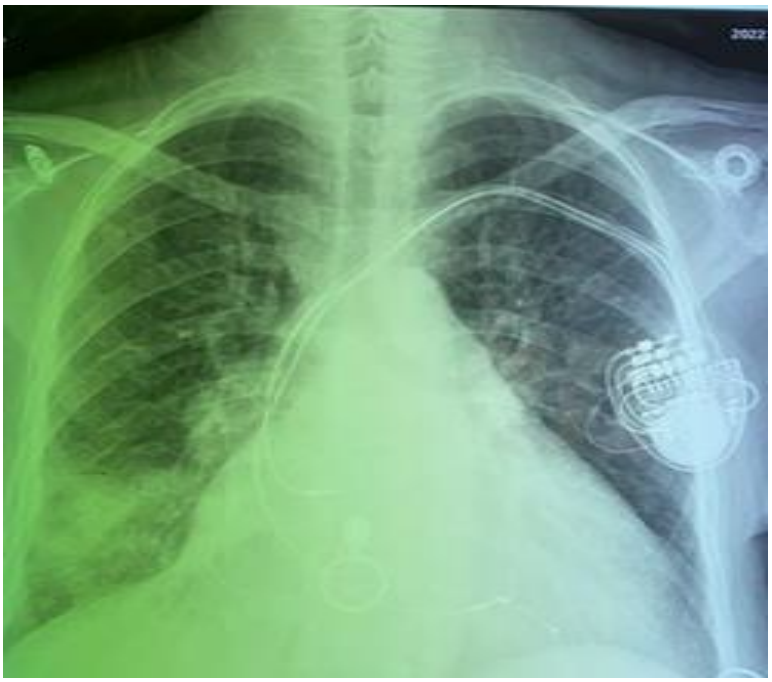

Figure 1: Chest X-ray showing cardiomegaly with an apex extending below the diaphragm, CRT-D leads in situ, and a right basal pulmonary opacity

Laboratory testing was unremarkable, including normal renal and hepatic function, and negative immunologic work-up. Echocardiography revealed a markedly dilated left ventricle (end-diastolic diameter 64 mm), severe global hypokinesis, LVEF 15–20% by Simpson biplane method, spontaneous intracavitary contrast, and severe left atrial enlargement.(figure 2+ 3)

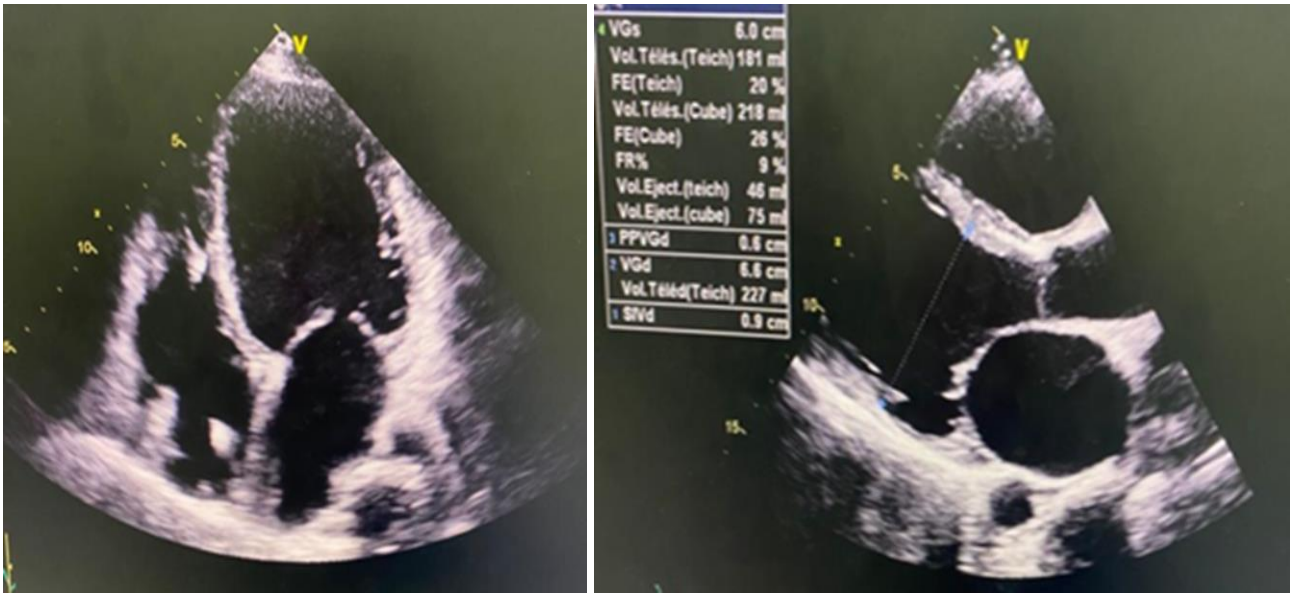

Figure 2+3: Transthoracic echocardiography (apical four-chamber view) showing a markedly dilated left ventricle with global hypokinesis and spontaneous intracavitary contrast

Despite maximal tolerated GDMT, optimization of CRT-D settings, and aggressive diuretic therapy, the patient remained symptomatic with refractory NYHA class IV heart failure. A multidisciplinary heart team evaluation was performed, and she was deemed eligible for advanced heart failure therapies. Following completion of pre-transplant assessment, she was listed for orthotopic heart transplantation and remains under close follow-up while awaiting donor allocation.

DISCUSSION

Epidemiology and Risk Factors

PPCM accounts for a small proportion of cardiomyopathies but represents a major cause of pregnancy-associated heart failure [6–9]. Geographic variability is striking, with higher incidences reported in Nigeria, South Africa, and Haiti, possibly reflecting genetic, environmental, nutritional, and socioeconomic factors [6,8,21]. Risk factors include multiparity, advanced maternal age, multiple gestation, hypertensive disorders of pregnancy, preeclampsia, prolonged tocolysis, and African ancestry [2,6,10,21].

Pathophysiology

The pathogenesis of PPCM is complex and multifactorial. Experimental and clinical evidence suggests that oxidative stress leads to proteolytic cleavage of prolactin into a 16-kDa fragment with antiangiogenic and proapoptotic effects on cardiomyocytes and endothelial cells [11–13]. This is supported by the observed therapeutic benefit of bromocriptine, a prolactin inhibitor, in selected patients [12,22].

Additional mechanisms include inflammatory activation, autoimmune responses against cardiac antigens, viral myocarditis, endothelial dysfunction, and impaired angiogenesis mediated by soluble fms-like tyrosine kinase-1 (sFlt-1) and placental growth factor imbalance [14–16,23]. Genetic studies have identified pathogenic variants in dilated cardiomyopathy-associated genes, particularly truncating variants in *TTN*, suggesting that PPCM may represent a pregnancy-triggered manifestation of genetic cardiomyopathy in some patients [15,24].

Clinical Presentation and Diagnosis

Patients typically present in late pregnancy or early postpartum with symptoms indistinguishable from acute heart failure syndromes, including dyspnea, orthopnea, paroxysmal nocturnal dyspnea, fatigue, and peripheral edema [3,4,7]. Physical findings may include tachycardia, pulmonary rales, elevated jugular venous pressure, and signs of low cardiac output.

Echocardiography remains the cornerstone of diagnosis, demonstrating reduced LVEF, LV dilatation, functional mitral regurgitation, and sometimes right ventricular dysfunction or intracardiac thrombus [3,4]. Cardiac MRI provides complementary information by assessing ventricular volumes, myocardial edema, fibrosis, and excluding alternative etiologies such as myocarditis or infiltrative cardiomyopathies [25].

Complications

Thromboembolic events occur in up to 10–20% of patients, reflecting severe LV dysfunction, blood stasis, hypercoagulability of pregnancy, and endothelial dysfunction [17,26]. Arrhythmias, including atrial fibrillation, ventricular tachycardia, and conduction abnormalities, contribute significantly to morbidity and mortality [27]. Cardiogenic shock and multiorgan failure may develop in fulminant cases [18,20].

Management

Management of PPCM is based on standard therapy for HFrEF, modified according to pregnancy and breastfeeding status [3,4,7,28]. During pregnancy, treatment options are limited due to fetal safety considerations, with beta-blockers, hydralazine-nitrates, and diuretics forming the therapeutic backbone [4,28]. Postpartum, full GDMT should be instituted promptly, including:

- Beta-blockers
- Renin–angiotensin system inhibitors or angiotensin receptor-neprilysin inhibitors (ARNI)
- Mineralocorticoid receptor antagonists
- SGLT2 inhibitors

These agents have demonstrated significant reductions in mortality and heart failure hospitalization in HFrEF populations and are increasingly extrapolated to PPCM patients [29–32]. Bromocriptine has shown benefit in selected cohorts by inhibiting prolactin secretion and improving LV recovery, although its routine use remains debated and requires concomitant anticoagulation due to thrombotic risk [12,22].

Anticoagulation is recommended in patients with LVEF ≤35%, intracardiac thrombus, atrial fibrillation, or previous thromboembolism [4,17]. Antiarrhythmic therapy and implantable cardioverter-defibrillators may be required in patients with malignant ventricular arrhythmias or persistent severe LV dysfunction [27].

Device Therapy and Advanced Heart Failure Management

Cardiac resynchronization therapy (CRT) is indicated in patients with persistent LVEF ≤35%, NYHA class II–IV symptoms despite optimal GDMT, and evidence of electrical dyssynchrony (QRS ≥130 ms, particularly with left bundle branch block morphology) [33,34]. CRT has been associated with improved functional class, ventricular remodeling, and survival in selected PPCM patients [35].

Mechanical circulatory support (MCS), including extracorporeal membrane oxygenation (ECMO), intra-aortic balloon pump, and durable left ventricular assist devices (LVADs), may be employed as bridge-to-recovery, bridge-to-decision, or bridge-to-transplant in patients with cardiogenic shock or refractory end-stage heart failure [18,20,36].

Orthotopic heart transplantation remains the definitive therapy for patients with irreversible myocardial dysfunction despite maximal medical and device therapy [19,20,37]. Outcomes after transplantation for PPCM are comparable to those for other indications, with reported 1- and 5-year survival rates exceeding 85% and 70%, respectively [19,37,38].

Prognosis and Long-Term Outcomes

Approximately 50–70% of PPCM patients experience partial or complete recovery of LV function within 6–12 months [6,18,39]. Predictors of favorable recovery include higher baseline LVEF, smaller LV dimensions, absence of right ventricular dysfunction, and early initiation of GDMT [18,39,40]. Conversely, LVEF <30% at diagnosis, LV end-diastolic diameter >60 mm, delayed diagnosis, and persistent symptoms beyond 6 months are associated with poor recovery and progression to chronic heart failure [6,18,39].

Future pregnancies carry a substantial risk of relapse, particularly in women with persistent LV dysfunction, and are generally contraindicated when LVEF remains below normal [41,42]. Even in patients with recovered LV function, recurrence rates of PPCM range from 20–30%, underscoring the importance of preconception counseling and close cardiologic surveillance [41–43].

CONCLUSION

Peripartum cardiomyopathy is a rare but potentially devastating disease that can progress to advanced heart failure despite optimal guideline-directed medical and device therapy. Early diagnosis, aggressive multidisciplinary management, and close follow-up are essential to improve outcomes. In refractory cases, advanced therapies including mechanical circulatory support and heart transplantation represent lifesaving options, as illustrated by the present case.

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