

# Acute pulmonary edema as clinical presentation of a peripartum cardiomyopathy in a very young patient

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## ABSTRACT

We present a case of a 20-year-old woman at 34th week of pregnancy, hospitalized because of the risk of premature delivery. Because of a progressive worsening of clinical conditions with dyspnoea and desaturation, an emergency caesarean section with healthy fetal extraction was necessary. In the immediate post-operative period there was a sudden worsening of dyspnea with hypoxic-hypercapnic respiratory failure and severe pulmonary edema, with subsequent endotracheal intubation and mechanical ventilation. Echocardiography showed left atrial and ventricular dilatation, global hypokinesia with reduced systolic function (EF 30%), moderate mitral regurgitation with symmetrical tethering of the mitral leaflets and pericardial effusion. Cardiac magnetic resonance showed a non-inflammatory pathway and was helpful to provide information in the differential diagnosis, so after a massive therapy we progressively observed

a complete stabilization of the hemodynamic clinical picture, normalization of cardiac troponin and NT-proBNP levels and final full cardiac recovery. The early signs and symptoms of heart failure in peripartum cardiomyopathy may mimic physiological changes occurring during/after pregnancy, delayed diagnosis may occur. Although most patients with peripartum cardiomyopathy improve with medical therapy, nearly a third of all patients develop a worsening heart failure, and may also be associated with severe and lasting complications including cardiogenic shock and death.

**Keywords:** Acute pulmonary edema, Heart failure, Peripartum cardiomyopathy, Pregnancy

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## INTRODUCTION

Peripartum cardiomyopathy (PPCM) is defined as a non-familial form of peripartum heart failure characterized as an idiopathic cardiomyopathy presenting with heart failure secondary to left-ventricular systolic

dysfunction towards the end of pregnancy or in the months following delivery, where no other cause of heart failure is found. The left ventricle (LV) may not be dilated but the ejection fraction (EF) is nearly always reduced below 45%. PPCM is considered an independent disease, whose diagnosis relies on its relation to pregnancy and the exclusion of other cardiomyopathies [1].

The early signs and symptoms of heart failure in PPCM patients may mimic physiological changes occurring during/after pregnancy, it might result in delayed diagnosis. The differential diagnosis of acute PPCM includes myocarditis, pre-existing cardiomyopathy, valve disease, or congenital heart disease [2].

The etiology of PPCM is still unknown, and many potential causes have been proposed but not proven. These include viral myocarditis, abnormal immune response to pregnancy, abnormal response to increased hemodynamic burden of pregnancy, hormonal abnormalities, excessive prolactin production, malnutrition, low selenium levels, stress-activated cytokines, inflammation and apoptosis, in addition to genetic factors [3-4].

Strong associations have been shown between PPCM and older maternal age, history of hypertension, multiple pregnancies, and African American background.

**CASE REPORT**

A 20-year-old Italian woman at the 34th week of pregnancy was hospitalized at the gynecology department of our hospital, because of the risk of premature delivery. Until hospitalization was a pregnancy with regular course (negative virological examination, hematochemical routine tests and morphological ultrasound). She had a history of partial epilepsy and two previous pregnancies (the first one without any complications and two years later a spontaneous abortion in the third month).

On admission she had a severe Sideropenic anemia (RBC 2.31 x10<sup>6</sup>/mL, Hgb 6.1 g/dL, sideraemia 16 mg/dL, ferritin 6.8 ng/dL) and low levels of C-reactive protein (4.7 mg/L), blood pressure was 105/70 mmHg, arterial oxygen saturation was 98% and 12-lead ECG showed sinus tachycardia with diffuse alterations of repolarization. In the following two days there was a progressive worsening of clinical conditions, with dyspnoea and desaturation, and on the third day an emergency caesarean section was performed in spinal anesthesia without intraoperative complications and with healthy fetal extraction.

In the immediate post-operative period, there was a sudden worsening of dyspnea with hypoxic-hypercapnic respiratory failure and clinical picture of severe pulmonary edema (with extensive pleural effusion and rales up to the lungs apices). She was therefore transferred to the intensive care unit, was performed endotracheal intubation and connected to mechanical ventilation in assisted/controlled mode.

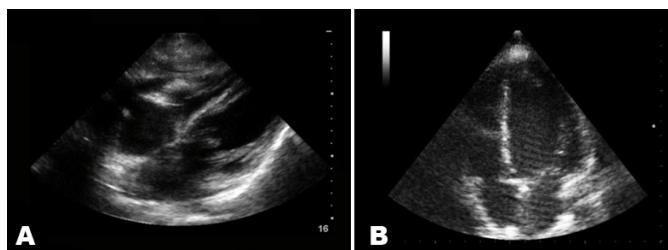
Echocardiography showed a slight dilated LV with

normal wall thickness; global hypokinesia with reduced systolic function (EF 30%); left atrial dilatation; moderate mitral regurgitation with symmetrical tethering of the mitral leaflets, slight anterior pericardial effusion (Figures 1A, 1B).

It was set diuretic therapy with endovenous infusion of furosemide, antibiotic therapy with levofloxacin and piperacillin/tazobactam, ionic correction of acidosis (sodium bicarbonate) and hydro-electrolytic balance, antithrombotic prophylaxis with enoxaparin and low dose of ramipril and aldactone. The investigation for common pathogens responsible for myocardial injury (legionella, chlamydia, mycoplasma, pneumococcus, coxsackievirus) was negative.

Following the progressive improvement in clinical conditions, our patient was extubated successfully and transferred to cardiology unit with an initial improvement of systolic cardiac function (EF 45%) and a pathological ECG with Q-waves in V1-V2, high-voltage positive T-waves in inferior leads and V3-V6 (Figure 2). Blood test demonstrated elevated levels of cardiac troponin I (225 ng/L) and NT-proBNP (759 pg/mL), normal creatinine (0.4 mg/dL), increase in hemoglobin levels (10.8 g/dL) and sideraemia (48 µg/dL).

Cerebral and thoracic computed tomography (CT) excluded intra and extra-axial hemorrhages, recent ischemic lesions and pulmonary embolism, confirming instead a severe acute pulmonary edema (Figures 3(A and B)).



Figures 1: (A) 2-D transthoracic echocardiogram in subcostal view with pericardial effusion during acute phases, (B) apical four-chamber view.

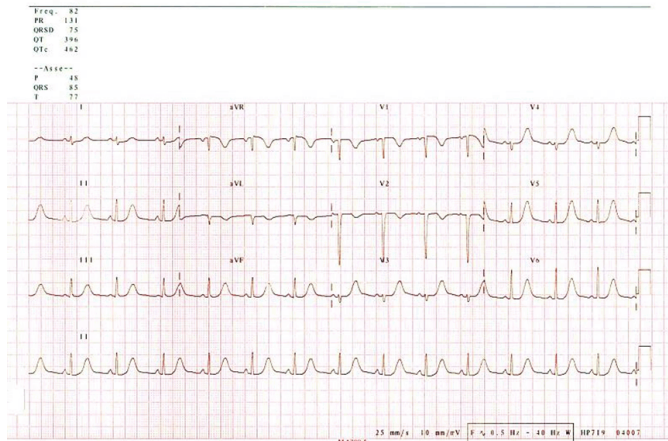


Figure 2: Pathological ECG with Q-waves and high-voltage positive T-waves.

Cardiac magnetic resonance imaging (MRI) showed a global hypokinesia with mild reduction in left ventricular systolic function (EF 46%), slight pericardial and pleural effusion, mitral regurgitation. In the T2-weighted sequences, no areas of hyperintensity of the signal, referable to myocardial edema. In the sequences acquired immediately after the administration of contrast media ev, no areas of hypointensity of the signal with subendocardial extension, referable to defects of perfusion at rest. In late sequences no areas of intra-myocardial hyperintensity, referable to fibrosis (Figure 4A–C).

Standard heart failure therapy (ramipril, bisoprolol, furosemide and spironolactone) and antibiotic therapy was continued with a complete stabilization of the hemodynamic clinical picture, normalization of cardiac troponin and NT-proBNP levels, improvement of left ventricular function to serialized echocardiographic controls (EF 52%) and discharge at the 13th day of hospitalization.

The young patient was visited a month after discharge into perfect clinical conditions and a full cardiac recovery by echocardiography: left sides with normal sizes; normal systolic function (EF 65%); slight mitral regurgitation (reduced), absence of pericardial effusion.

For this reason we opted for a gradual discontinuation of therapy with a frequent monitoring of LV function every 3–6 months for the follow years.

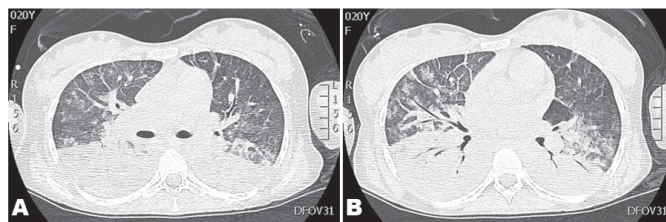
## DISCUSSION

The clinical presentation of patients with PPCM is similar to those with other forms of systolic HF secondary to cardiomyopathy, but may be highly variable. Early signs and symptoms of PPCM may often mimic normal physiological findings of pregnancy and include pedal oedema, weight gain, physical discomfort, dyspnoea on exertion, orthopnoea, paroxysmal nocturnal dyspnoea, and persistent cough. Additional signs and symptoms experienced in PPCM include abdominal discomfort secondary to hepatic congestion, dizziness, praecordial pain, palpitations, and in the later stages postural hypotension, third heart sound and pulmonary rales [1].

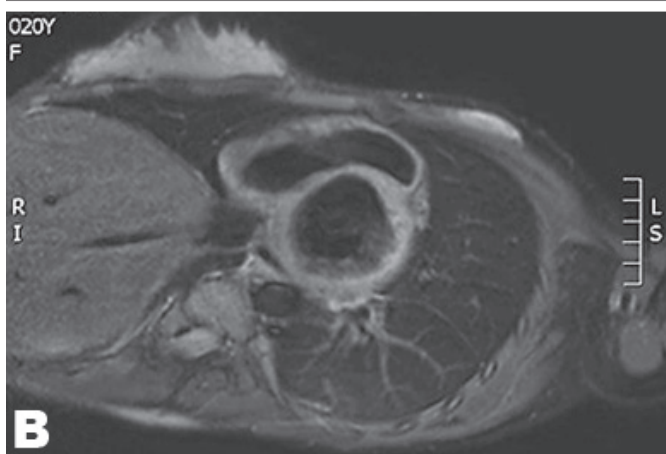
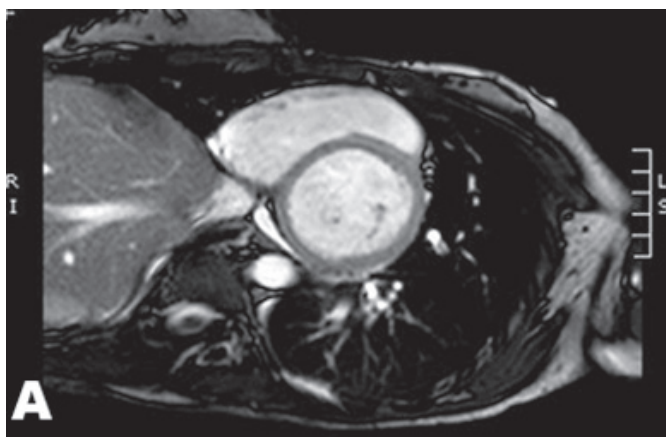
For this reason, and because of the low incidence of this condition, the diagnosis of PPCM is often missed or delayed, allowing the development of preventable complications.

Echocardiography in PPCM shows variable degrees of LV dilatation with moderate to severe depression of systolic function, right ventricular and bi-atrial dilatation and moderate to severe mitral and tricuspid regurgitation with increased pulmonary pressures.

Cardiac magnetic resonance imaging (MRI) has an important role in prognostic stratification, by the assessment of LV size, function and contractile reserve, the detection of mural thrombi or myocardial fibrosis and finally the presence or absence of myocardial late gadolinium enhancement [5]. By distinguishing inflammatory (late



Figures 3 (A and B): Chest CT with pleural effusion and parenchymal consolidation, interstitial edema, alveolar edema (ground glass), enlargement of pulmonary segmental arteries.



Figures 4: (A) cardiac MRI obtained at end diastole showing basal short-axis slices, (B) normal T2-weighted signal, (C) absence of delayed gadolinium enhancement.

gadolinium enhancement in T1-weighted sequences; hypersignal in T2-weighted sequences) and non-inflammatory (absence of enhancement and hypersignal) forms of PPCM, cardiac MRI can be helpful at initial presentation to consider appropriate physiopathologic hypothesis and to guide further etiologic investigations. It can also guide therapeutic options, being in favor of using immunosuppressor agents in inflammatory forms of PPCM and provide critical information in the differential diagnosis of myocarditis, Takotsubo or ischemic cardiomyopathy [6–7].

Although the prognosis is more favorable than in other types of cardiomyopathies, PPCM may be associated with severe and lasting complications including severe HF, pulmonary edema, cardiogenic shock, cardiopulmonary arrest secondary to HF or arrhythmias, thromboembolic complications, and death. Predictors of complications are left ventricular ejection fraction (LVEF) < 25%, non-Caucasian ethnic background, and delay of diagnosis. [8]

A number of factors have been shown to be associated with a higher likelihood of recovery, including LV diastolic dimension (< 55 to 60 mm) and systolic function (LVEF < 30% to 35% and fractional shortening < 20%) at the time of diagnosis, lack of troponin elevation, a lower level of plasma BNP, absence of LV thrombus, breast-feeding, diagnosis after the delivery, and non-African American ethnicity [2].

Currently, PPCM is treated according to the ESC guidelines for heart failure in pregnancy (e.g. hydralazine and nitrates). After delivery, standard therapy for heart failure is recommended in PPCM including beta-blockers, ACE-inhibitors/AT1-blockers, intravenous inotropes, intravenous and oral vasodilators, mineralocorticoid receptor antagonists (MRA), and diuretics [9–10].

Although most patients with PPCM improve with medical therapy, nearly a third of all patients develop worsening heart failure. A general agreement among experts suggests continued therapy with standard heart failure medications for a minimum of 12 months [11].

## CONCLUSION

Pregnancy, delivery, and the peripartum period provide a challenge to the entire female organism, therefore understanding normal pregnancy is important for the timely recognition of cardiovascular pathologies, to distinguish the peripartum discomfort in healthy women from signs of cardiovascular disease. This is a major challenge for physicians and patients, and may explain why diagnosis of PPCM is frequently delayed and still underestimated.

Gradual discontinuation with frequent monitoring of LV function is reasonable in patients with complete recovery of LV systolic function (LVEF > 55%) and normal LV size, contrarily standard heart failure therapy should be continued in patients with persistently reduced LVEF for several years or even lifelong.

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## Author Contributions

Riccardo Bentivegna – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Egle Corrado – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising

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Giuseppe Coppola – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Emanuele Grassettonio – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Salvatore Novo – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

### **Guarantor of Submission**

The corresponding author is the guarantor of submission.

### **Source of Support**

None

### **Consent Statement**

Written informed consent was obtained from the patient for publication of this report.

### **Conflict of Interest**

Authors declare no conflict of interest.

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