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Peripartum Cardiomyopathy: Facts and Figures

Nissar Shaikh, Firdos Ummunnisa, Arshad Chanda, Mohammad A. Imran, Adel Ganaw, Umm-e-Amara, Zia Mahmood, M.A. Rahman, Mohammad Nayeemuddin, Moad Ehfeda, Muhammad Zubair, Ahmed Atef Shible, Ranjan Matthias, Muhammad Shakeel Riaz, Hafiz Hamid Habib, Masood Khattak and A.R. Raju Vegesna

Abstract

Peripartum cardiomyopathy (PPCM) is a rare clinical entity during pregnancy. PPCM is a diagnosis of exclusion. These patients do not have prior history of heart disease, and there are no other known possible causes of heart failure. It is more common in African countries, may be related to the consumption of kanwa, in the postpartum period. The multiparity, African descent and pregnancy-induced hypertension are a few risk factors for PPCM. The exact etiology of PPCM is not known; possible theories range from myocarditis to the maladaptation to the changes of pregnancy. The clinical manifestation varies from shortness of breath to thromboembolic phenomenon. Echocardiography is essential for diagnosis as well as differential diagnosis of PPCM. These patients preferably are managed in tertiary healthcare facilities. Anticoagulation and antiarrhythmic medications are pillars for the management of PPCM patients. If required, mechanical devices should be used temporarily. PPCM patients may need heart transplant. The beneficial role of bromocriptine and immunosuppression is not clear in PPCM patients. Subsequent pregnancies should be avoided to prevent the PPCM occurrence.

Keywords: echocardiography, left ventricular ejection fraction, peripartum cardiomyopathy, multiparity, heart failure, anticoagulation and antiarrhythmic medications

1. Introduction

Peripartum cardiomyopathy (PPCM) is the heart muscle weakness that begins during the late pregnancy and extending into 5 months in puerperium without any other known etiology. PPCM is a dilated cardiomyopathy and involves systolic dysfunction, decreased left ventricular ejection fraction (EF), and congestive heart failure. There is an increased risk of atrial and ventricular arrhythmias and thromboembolism. PPCM is a diagnosis of exclusion, patients have no prior history

of heart disease, and there are no other known possible causes of heart failure (HF). Echocardiogram is essentially used for both diagnosis and monitoring purposes in the progress of PPCM. Although initially described in the 18th century, it was recognized as distinct clinical in 1930, and Demakis et al. described and defined PPCM [1]. PPCM was initially termed as pregnancy-associated cardiomyopathy, toxic postpartum heart failure, Meadows' syndrome, postpartum myocarditis, and Zaria syndrome.

2. Definition

There are varieties of definitions for PPCM; the more simplified one is by the working group on peripartum cardiomyopathy from the European Society of Cardiology in 2010. It avoids under diagnosis by adopting a broader definition. It contains the following characteristics:

1. The development of heart failure in last month of pregnancy or up to 5 months in postpartum period
2. Absence of another known or identifiable cause for heart failure
3. Systolic dysfunction of left ventricle (LV) and LV ejection fraction of less than 0.45 with or without LV dilatation [2]

3. Epidemiology

PPCM constitutes less than 1% of all cardiovascular events related to pregnancy. It is more common in Africa, 1:300 pregnancies. This may be due to the consumption of kanwa, a tradition, for 40 postpartum days. Kanwa is a dry salt and causes hypervolemia and hypertension. Ninety percent of PPCM occurs within 2 months of delivery [3]. In a recently concluded study about the incidence of PPCM from 43 countries, affecting females from all ethnicity and all continents, the incidence widely differs depending on geographic location. PPCM was common in Nigeria (1100), Haiti (1300), South Africa (11,000), Canada (12,400), and Denmark (110,149) and was lowest in Japan (1,20,000 live births). There are case reports from other countries [4]. Hence the information is not complete as many countries are not having a registry. The higher incidence in Nigeria may be related to above-mentioned postpartum high salt intake or may be a genetic factor superimposing the geographic and dietary variabilities [5].

4. Risk factors

The following are the risk factors for PCCM, even though the etiology of PPCM is still not clear.

1. Age over 30 years, pregnancy with multiple fetuses [6].
2. Multiparty, African descent [7].
3. Maternal cocaine abuse [8].

4. Long-term tocolytic therapy [9].
5. History of preeclampsia, eclampsia, or peripartum hypertension, the hypertensive disorders of pregnancy are significant predispose for PPCM, and the preeclampsia patient is four times more susceptible for developing PPCM [10].

5. Etiopathology

In the pathology of peripartum cardiomyopathy, the macroscopic view is a pale myocardium, dilated heart often with intramural thrombus in the ventricles. There will be endocardial thickening and pericardial effusion. The myocardial cellular hypertrophy and myofibril degeneration with areas of fibrosis with interstitial edema are the nonspecific findings. In a small group of patients, there are features of myocarditis with the presence of inflammatory cell infiltration of myocardium, focal necrosis, variable hypertrophy, and fibrosis of the myocardium [11].

Although PPCM is a separate clinical entity, the exact etiology of PPCM is still not known. Following hypothesis had been proposed for the etiology.

5.1 Familial

The familial clustering of PPCM is well known; it could be due to genetic or environmental factors. Ancestry with African genomics may be a risk factor and explains the higher prevalence of PPCM in Haiti, Africa, and black women in other countries. The guanine nucleotide-binding protein beta-3 (GNB3) subunit has a polymorphism called C825T. This polymorphism is associated with an increased risk of hypertension, low plasma renin, and cardiac remodeling. GNB3 has a prevalence of 50% in black individuals compared with 10% in white individuals [12].

5.2 Myocarditis

First time Melvin et al. proposed myocarditis as a cause for PPCM. Myocarditis could be viral or autoimmune. With pregnancy there is increased in susceptibility to both. In another study, endomyocardial biopsies in five patients showed features of myocarditis. The reason for lesser positive biopsy report among the studies may be related to small sample size and timing of biopsy with relation to the onset of symptoms. The incidence of inflammation is greater in patients who are biopsied soon after presentation [13, 14]. The proinflammatory-inflammatory cytokines may play a role in the pathogenesis and progression of PPCM, as the cytokines that are elevated in PPCM compared with controls include tumor necrosis factor (TNF)-alpha and interleukin-6 [15].

5.3 Abnormal immune response

The entrance of fetal cells into the maternal circulation remains in the circulation without rejection due to weak immunogenic paternal haplotype of chorionic cell. If these cells lodge into the cardiac tissue, immune response may be triggered. The raised titres of immunoglobulins and other autoantibodies in patients with PPCM are suggestive of abnormal immune response, whereas other studies found no significant difference in levels of immunoglobulins and other autoantibodies in PPCM and control group of patients [12, 16].

5.4 Maladaptation to stress of pregnancy

The hyperdynamic circulation during pregnancy causes remodeling and transient hypertrophy of the left ventricle. The exaggerated reduction in left ventricular systolic function with stress of gestational hypertension may contribute to heart failure in PPCM patients. In pregnancy 40–50% increase in blood volume and cardiac output occurs which results in transient LV hypertrophy. The hemodynamic stress of gestational hypertension, may contribute to the development of PPCM and heart failure, with an angiogenic imbalance may better explain the association between preeclampsia and PPCM [17].

5.5 Angiogenic imbalance and balance

Human and animal studies suggest that PPCM is caused by systemic angiogenic imbalance. Animals that lack cardiac PGC-1 α , a regulator of proangiogenic factors such as VEGF, develop severe PPCM. These data may also explain why preeclampsia and multiple gestations are risk factors for PPCM. During late gestation, the human placenta secretes VEGF inhibitors such as soluble fms-like tyrosine kinase 1 (sFlt1), which damages the vasculature, with higher levels seen with multiple gestations or preeclampsia. Preeclampsia and multiple gestations are risk factors for PPCM as in these patients subclinical cardiac dysfunction correlates with sFlt1 levels [18].

5.6 Prolactin role

Alteration in prolactin processing is involved in the pathogenesis of peripartum cardiomyopathy; animal with a knockout in the cardiac tissue-specific signal

Figure 1. Summary of etiopathogenesis of PPCM (from [19]).

transduction and activator of transcription 3 (STAT3) developed PPCM. Reduction in STAT3 leads to increased cleavage of prolactin into an anti-angiogenic and proapoptotic 16 kDa isoform by cathepsin D. The 16 kDa prolactin fragment (16K PRL) causes endothelial damage leading to myocardial dysfunction; 16K PRL induces microRNA-146a expression in endothelial cells, which leads to most of the anti-angiogenic effects of 16K PRL. Females with PPCM have elevated levels of microRNA-146a compared with healthy postpartum women or women with other cardiomyopathies (**Figure 1**) [20].

6. Clinical presentations

PPCM is common in first postpartum month; it is also rare before 36 weeks of gestation (**Figure 2**). The common presentations of PPCM are dyspnea, cough, orthopnea, and paroxysmal nocturnal dyspnea, which may be confused with the physiological changes of pregnancy. Initial diagnosis may be delayed since symptoms such as nonspecific fatigue, shortness of breath, and pedal edema are similar to those observed in normal pregnancy. The patient may have arrhythmia and even cardiac arrest rarely [4]; PPCM can present with thromboembolic manifestations; patients with left ventricular ejection fraction (LVEF) < 35% are at risk for developing left ventricular thrombus. PPCM is manifested after 38 weeks of pregnancy, whereas pregnant patients with chronic heart disease develop sign and symptoms of heart failure in the second trimester of pregnancy due to stress of hemodynamic overload.

Figure 2.
Pregnancy and occurrence of PPCM [19].

7. Signs and symptoms

Signs and symptoms are variable and similar to that in other forms of systolic HF due to cardiomyopathy. Signs include an elevated jugular venous pressure, displaced apical impulse, a third heart sound, and a murmur of mitral regurgitation. Signs and symptoms of systemic or pulmonary thromboembolism may be present. Various studies have reported varying rates of thromboembolism, and further data are required to quantify the risk of this complication. Patients with PPCM left ventricular thrombus were identified by echocardiography in 16 of 100 patients with PPCM (with mean LVEF of 26%) in a case series [2, 21].

8. Diagnosis

As mentioned initially diagnosis of PPCM based on three clinical criteria, development of heart failure in last month of pregnancy or in the initial 5 months following delivery with the absence of another identifiable cause of HF and left ventricular (LV) systolic dysfunction with an left ventricular ejection fraction (LVEF) of $< 45\%$. The last criteria will exclude patients with accelerated hypertension, diastolic dysfunction, systemic infection, pulmonary embolism, or preeclampsia or amniotic fluid embolus. Chest X-ray, electrocardiogram (ECG), and echocardiogram should be performed in patients who are clinically suspected of having PPCM.

Studies showed brain natriuretic peptide (BNP) levels, cardiac magnetic resonance (CMR) imaging, cardiac catheterization, and endomyocardial biopsy (EMB) will be helpful in these patients. Bacterial cultures and viral titers are usually not indicated, as these tests are nonspecific and without proven value in patients with myocarditis. The novel markers, plasma concentrations of proangiogenic and anti-angiogenic factors, placenta growth factor, and fms-like tyrosine kinase 1 receptor, have been proposed to be used to distinguish patients with PPCM [22].

8.1 Electrocardiogram (ECG)

It is not specific in PPCM patients and may show sinus tachycardia rarely arrhythmias and nonspecific ST segment and T wave changes. PR and QRS intervals may be prolonged. An ECG is helpful in identifying conditions in the differential diagnosis such as myocardial infarction and pulmonary embolism [1].

8.2 Brain natriuretic peptide (BNP)

Plasma BNP or pro-BNP (proBNP) measurement is helpful in the evaluation and diagnosis of patients with heart failure. Patients with PPCM have increased BNP and NT-proBNP levels, higher than seen in healthy women during pregnancy or postpartum.

8.3 Chest X-rays

Commonly it will show enlargement of the cardiac silhouette with pulmonary congestion and/or interstitial edema and pleural effusions. However, a chest radiograph is not necessary to make a diagnosis of HF or PPCM and exposes the patient to ionizing radiation.

8.4 Echocardiography (echo)

Echocardiogram is essential in the diagnosis of PPCM; it reveals a global reduction in LV systolic function with LVEF of $< 45\%$. Left ventricle is commonly but not always dilated. Assessment of right ventricular systolic pressures can be performed (**Figure 3**). Echo may show presence of left atrial thrombus, dilated right ventricle, abnormal ventricular wall motion, mitral and tricuspid regurgitation, and pericardial effusion.

8.5 Cardiac magnetic resonance imaging (CMR)

If echocardiography is technically suboptimal, CMR can help. It helps in assessing LV function and LV volumes. Experience with CMR in PPCM is limited and its role is still being evaluated.

Figure 3.
Four chamber view of PPCM A: Echocardiographic four chamber view of PPCM. B: Echocardiographic Mitral regurgitation view of PPCM.

8.6 Cardiac catheterization

It may be helpful in critically ill patients, requiring a complete assessment or ongoing evaluation of their hemodynamic state.

8.7 Left heart catheterization and coronary angiography

It is required in selected patients in whom it is necessary to evaluate coronary artery disease as a potential cause for the cardiomyopathy. The coronary angiography exposes the patient to ionizing radiation and it is commonly not required in patients with suspected PPCM.

8.8 Endomyocardial biopsy (EMB)

EMB is recommended in clinical scenarios in which a biopsy is anticipated to yield a diagnosis of a specific condition with treatment implications. These scenarios include heart failure with hemodynamic compromise of less than 2 weeks duration or heart failure of less than 3 months duration if associated with heart block, new ventricular arrhythmias, or refractory heart failure. EMB is not recommended for the routine evaluation of heart failure, as there are no pathognomonic findings in PPCM. The risk of a serious acute complication is less than 1% using flexible biptomes.

9. Differential diagnosis

PPCM should be differentiated from other forms of cardiomyopathy (**Figure 4**), heart failure, pulmonary thromboembolism, severe eclampsia, and pneumonia. From history, in physical examination and investigations, one must exclude myocardial infarction, idiopathic dilated cardiomyopathy, and valvular heart disease [9].

Figure 4.
Four chamber echocardiographic view showing thrombus in PPCM.

10. Management

The therapeutic approach of PPCM is the same as for other types of HF with left ventricular systolic dysfunction. Precautions should be taken to ensure the safety of the mother and the unborn or breastfeeding child; these patients may need antiarrhythmic drugs, anticoagulation therapy, mechanical support, and the use of investigational medications (bromocriptine).

The cornerstone in the management of PPCM is to reduce preload and after load and increase the cardiac contractility. Heart failure during pregnancy may be acute or acute on chronic. The pregnant patient with known cardiac disease can present in stable condition during early stages of pregnancy. Careful physical examination should be done. Their management is mainly adjustment of their medication and monitoring for cardiac failure. The initial New York Heart Association functional class status should be documented. Serial ECG and echocardiogram should be performed.

Patients presenting heart failure during pregnancy or the peripartum period require a detail history and physical examination and the evaluation of severity of decompensation. An ECG may reveal deteriorating left ventricular functions, arrhythmia, LVH, or arterial abnormality. The therapeutic approach in these patients includes optimizing hemodynamics, reducing after load, optimizing preload, and cardiac contractility. These can be achieved by treatment of pulmonary congestion, control of hyper-/hypotension, treatment of cardiac arrhythmia, and prevention of thromboembolic events.

10.1 Antiarrhythmic therapy

Arrhythmias are common in patients hospitalized for PPCM. The occurrence of arterial and ventricular arrhythmias is variable. In one of the studies, 18.7% PPCM patients had an arrhythmia, and ventricular tachycardia was in 4.2% with cardiac arrest in 2.2%. In smaller studies, the reported incidence of ventricular tachycardia was 20 and 25%. The atrial fibrillation was reported to occur in 3.1–11.9% of patients with PPCM [23].

Ventricular arrhythmias should be treated aggressively in PPCM patients. Class III antiarrhythmic medications are the best option. Intravenous medications are needed in PPCM patients admitted to the intensive care therapy unit. Therapy with inotropes such as dobutamine, adrenaline, and milrinone should be directed by invasive cardiac monitoring. While interpreting the invasive hemodynamic monitoring, one should take into account the normal changes that occur during pregnancy. Digoxin is safe to use in pregnancy. Diuretics can be used if salt restriction is not sufficient. Beta-blocker improves left ventricular function in patients of PPCM, but ACE inhibitors are the drugs of choice in postpartum PPCM [24].

10.2 Anticoagulation therapy

The anticoagulation in PPCM is a must as pregnancy itself is a hypercoagulable state, in addition to PPCM, dilatation of heart, and turbulent flow of blood. For pregnant patients requiring anticoagulation, decisions and choosing anticoagulation therapy are challenging due to the risk of bleeding in all stages of pregnancy and the potential teratogenic effects of warfarin in the first trimester, dosage of various agents, and management during labor and delivery. PPCM patients receiving bromocriptine have an increased risk of thromboembolic events; hence, it is suggested to start anticoagulation therapy in patients with PPCM treated with bromocriptine. There is no clear data, but expert suggests anticoagulation for patients with PPCM with acute cardiac thrombus or evidence of systemic embolism. Before delivery unfraction or low-molecular-weight heparin is the choice, whereas in postpartum period warfarin is used.

10.3 Mechanical and device support and cardiac transplantation

The decisions to use implantable cardioverter defibrillator (ICD) or cardiac resynchronization therapy in PPCM patients should be taken after a detailed natural history and the potential of recovery of ventricular function. All these therapies should be deferred at least for 3 months and possibly for 6 months of presentation as 20–60% of PPCM patients have complete recovery of left ventricular ejection functions to normal by 6 months to 5 years [25].

Mechanical circulatory support (MCS) is considered early for PPCM patients who are hemodynamically unstable and unresponsive to medical therapy with maximal inotropic support. A device can be implanted in the acute phase either to work as a bridge-to-recovery and subsequent weaning when the ventricular function improves, or it can be a bridge-to-bridge for implantation of a longer durable device. The bridge-to-transplantation is rarely required as the initial approach as a high proportion of PPCM patients will have recovery of ventricular function. Hence initially a temporary device should be used if required in these patients.

Severely reduced LVEF alone should not be an indication for the use of aggressive therapies (MCS and cardiac transplantation) in PPCM patients. If MCS is indicated, various devices can be used including intra-aortic balloon counterpulsation (IABP), venoarterial extracorporeal membrane oxygenation (ECMO), and LV assist device (LVAD). The choice of device will depend on the hemodynamic status of the patient and local availability and expertise. Venoarterial ECMO has been associated with an increase in prolactin levels, which may be detrimental in PPCM patients [26].

Loyaga-Rendon et al. reported that the PPCM patients who received durable mechanical circulatory support had better survival rate than that of females without PPCM with a survival rate of 83% in PPCM. These may be related to PPCM

patients who were younger and had fewer comorbidities. The rates of myocardial function recovery were poor, 6% in the PPCM group and 2% in without PPCM [27].

According to the older literature, the transplantation was performed in up to one-third of PPCM patients, whereas the contemporary reports demonstrate that transplantation rates vary from 4 to 23% of patients [28]. Hence PPCM patients with significant LV systolic dysfunction should be managed at a tertiary center with transplant facilities.

In a large study, it is found that the long-term survival in transplanted PPCM patients was worse compared with all others undergoing transplantation. PPCM patient who received a cardiac transplant had higher mortality, higher incidence of rejection, poorer graft survival, and higher retransplantation rates. Younger patient age, higher allosensitization, higher pretransplant acuity, and increased rejection rates are all important factors for poorer outcomes in these PPCM patients [28].

10.3.1 Wearable cardioverter-defibrillators

Further multicenter trial would be valuable to establish if these devices are beneficial; in a single smaller study, it was found to be useful.

10.4 Medical investigational therapy

Following therapies are not recommended for PPCM patients as its efficacy and safety have not been established.

10.4.1 Bromocriptine

The use of bromocriptine therapy in PPCM patients is controversial. The preliminary data have shown a benefit of bromocriptine in PPCM patients, but further studies are needed to establish safety and efficacy; it is suggested not to routinely use bromocriptine in PPCM patients. Its use is based upon experimental animal studies of prevention of PPCM by prolactin blockade with bromocriptine. Smaller and observational reports showed the beneficial response to bromocriptine therapy in patients with PPCM [29].

A multicenter study showed that the rate of full recovery (LVEF $\geq$ 50%) was not significantly higher in the 8-week group compared with the 1-week group. The patients in this trial had better outcomes than observed in prior series, but a placebo control group was not included in the study [30]. One should start anticoagulation in PPCM patients treated with bromocriptine, as the risk of thromboembolic complications.

10.4.2 Immunosuppressive therapy

The advantage of immunosuppressive therapy has been found to be useful in PPCM patients with biopsy-proven myocarditis, but its efficacy is unclear, and empiric immunosuppression in the absence of evidence of a responsive form of myocarditis is not recommended [31]. These medications have significant side effects.

10.4.3 Intravenous immunoglobulin

This therapy is tried in patients with myocarditis or newly onset dilated cardiomyopathy with no clear evidence of clinical benefit, and the efficacy of this approach has not been confirmed in any type of myocarditis.

11. Delivery

In PPCM patient the risks and benefits of early delivery should be considered and discussed. The 2010 European Society of Cardiology working group statement advised that early delivery is not required if the maternal and fetal conditions are stable. But the patient-related factors, gestational age, cervical status, fetal status, and the potential cardiovascular impact of continuing pregnancy must be considered in timing delivery. The decisions regarding timing and mode of delivery should be based on combined multidisciplinary meeting. In PPCM patients with advanced heart failure, prompt delivery of fetus is indicated for maternal cardiovascular indications and hemodynamic instability. The elective cesarean delivery is preferred for PPCM patients with advanced heart failure requiring inotropic therapy or mechanical circulatory support [12].

12. Breastfeeding

According to the expert group, breastfeeding is to be avoided because of the potential effects of prolactin subfragments, but in a study where PPCM patients chose to breastfeed, none had adverse maternal effects, and that rate of recovery of left ventricular function was significantly higher in lactating women, and accordingly given the benefits of breastfeeding, it is recommended that women who are stable should continue breastfeeding as long as it is compatible with their heart failure medications [32].

13. Complications

The most common complication is thromboembolism. A premature delivery rate of 25% has been reported in cases with PPCM. PPCM cases had increased incidence of cesarean section up to 40% [33].

14. Prognosis

14.1 Maternal morbidity and mortality

The overall reported mortality rate in PPCM is better than other cardiomyopathy (**Figure 5**) patients approximately 10% in 2 years and 11–29% in 3 years [15]. Cardiac transplantation rates of less than 1–2% per year [15]. The mortality in PPCM patient is commonly caused by progressive pump failure, sudden death, or thromboembolic events. The following factors have been shown to increase the mortality in PPCCM patients (**Table 1**).

PPCM is associated with a significant morbidity including brain injury, cardio-pulmonary arrest, fulminant pulmonary edema, thromboembolic complications, and defibrillator or pacemaker implantation [36].

14.2 Neonatal and obstetric outcomes

Lower section cesarean delivery was performed in 40% of patients, largely for obstetric indications. Preterm birth was noted in 25 and 5.9% of infants were small for date [37].

Figure 5.
Comparative mortality of cardiomyopathies [34].

Worse New York Heart Association functional class
Left ventricular ejection fraction (LVEF) ≤ 25%
Black race
Multiparity
Age more than 35 years [35]

Table 1.
Risk factors for increased mortality in PPCM.

14.3 Subsequent pregnancy

In patients who have recovered or not recovered from left ventricular failure due to PPCM, there is a high risk of PPCM in subsequent pregnancy. Termination of pregnancy may not prevent relapse.

15. Prevention

In patients who have recovered from left ventricular failure due to PPCM, there is a high risk of PPCM in subsequent pregnancy, and so the best way to avoid PPCM is avoid subsequent pregnancy. The literature suggests that the patient or her partner undergo a sterilization procedure or the patient use a highly effective non-estrogen method of contraception. Though the risk of recurrence appears to be less in PPCM patient with recovered LV function and LVEF > 25%, still these patients should receive counseling, including the option of avoidance of subsequent pregnancy due to the risk of relapse of PPCM, heart failure, and death. There is not much evidence on the safety of contraceptives in PPCM patients; it is advised that the estrogen-progestin contraceptives should be avoided in PPCM patients with persistent LV dysfunction because of their potential to increase the risk of thromboembolism [38].

16. Conclusion

PPCM is a rare but potentially life-threatening disease of pregnancy. PPCM is common in postpartum period. PPCM patient has various risk factor including

hypertensive disorders of pregnancy. PPCM patients may present with shortness of breath, arrhythmias, pulmonary edema, and thromboembolic signs and symptoms. Echocardiography will essentially diagnose the PPCM, and it also helps in differential diagnosis. The management of PPCM patients is the management of heart failure with special consideration of fetus. Therapy may involve the use of mechanical devices or even cardiac transplant, but the anticoagulation and antiarrhythmic medication plays an important role in the management. The medical management with bromocriptine, immunosuppression, and immunoglobulin is controversial. Commonly the left ventricular function recovers, but in PCCM the maternal mortality is around 10%, and there is increased incidence of cesarean section. In subsequent pregnancy in patient with PCCM, heart failure may be more severe and may cause death. Family counseling is essential in these patients.

Author details

Nissar Shaikh^{1*}, Firdos Ummunnisa², Arshad Chanda¹,
Mohammad A. Imran¹, Adel Ganaw¹, Umm-e-Amara³, Zia Mahmood¹,
M.A. Rahman¹, Mohammad Nayeemuddin¹, Moad Ehfeda¹, Muhammad Zubair¹,
Ahmed Atef Shible⁴, Ranjan Matthias¹, Muhammad Shakeel Riaz¹,
Hafiz Hamid Habib⁵, Masood Khattak⁵ and A.R. Raju Vegesna¹

1 Surgical Intensive Care, Hamad Medical Corporation, Doha, Qatar

2 Dr. Halima Al Tamimi, OBGY Center, Doha, Qatar

3 Apollo Medical College and Research Center, Hyderabad, India

4 Clinical Pharmacist, SICU, Hamad Medical Corporation, Doha, Qatar

5 Cardiology Department, Heart Hospital Hamad Medical Corporation, Doha, Qatar

*Address all correspondence to: nissatfirdous99@gmail.com

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