Peripartum Cardiomyopathy

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Abstract

Peripartum cardiomyopathy (PPCM) is a rare type of cardiomyopathy of unknown etiology associated with significant mortality and morbidity and characterized by heart failure in late pregnancy or puerperium. Recently PPCM workshop committee has recommended inclusion of echocardiographic features of LV dysfunction to redefine PPCM. Subsequent pregnancies are associated with a very high mortality in these patients and hence should be avoided. Women with PPCM continue to have significant mortality despite the use of conventional drugs for managing heart failure. Use of newer drugs such as immunoglobulin, pentoxifylline, bromocriptine, and cabergoline along with newer interventions such as plasmapheresis, immunoadsorption, ventricular assist devices and last but not the least the heart transplantation hold promise for future.

Introduction

Peripartum cardiomyopathy (PPCM) is a rare but critical disorder causing heart failure in women in late pregnancy or puerperium. It was first described in the 18th century but was recognized as a separate clinical entity in 1930. In 1971 Demakis et al described 27 patients who presented during the puerperium with cardiomegaly, abnormal electrocardiographic findings, and congestive heart failure, and named the syndrome “The peripartum cardiomyopathy”. Following the recommendations of workshop committee on PPCM that echocardiographic features of the left ventricular dysfunction should be incorporated to diagnose PPCM. The definition of PPCM was modified which now includes following four criteria, three clinical and one echocardiographic –

1. Development of heart failure during last trimester of pregnancy or first six months post partum.
2. Absence of any identifiable cause for cardiac failure.
3. Absence of any recognizable heart disease prior to last trimester of pregnancy.
4. Echocardiographic criteria- Demonstrable echocardiographic proof of left ventricular systolic dysfunction. Ejection fraction less than 45%, left ventricular fractional shortening less than 30% or left ventricular end-diastolic dimension>2.7cm/m square of body surface area.

Epidemiology

Peripartum Cardiomyopathy is relatively rare condition. The precise incidence in India is not known, an incidence of one case per 1374 live births has been reported from a tertiary care hospital from South India. The National Hospital Discharge Survey (1990–2002) estimated that it occurs in one in every 2,289 live births in the United States. The prevalence is reported to be one case per 6000 live births in Japan. The disease appears to be more common in African American women. The rate varies in other populations; in South Africa reported incidence is higher in 1 in 1000 live birth. A much higher incidence of I in 300 live births has been reported from Haiti, and an extremely high rate of 1% has been reported from Nigeria. The higher prevalence in developing countries may be attributed to environmental, ecological, and cultural puerperal and post puerperal practices besides diagnostic criteria and reporting standards.

Risk Factors

Incidence of peripartum cardiomyopathy has been found to be greater in multiparous women and in those with advanced maternal age. Multifetal pregnancy (twin and triplets), preeclampsia, and gestational hypertension appear to have a higher incidence. African American race, Obesity, Maternal cocaine and alcohol abuse, smoking and long term tocolytic therapy have been attributed as risk factors for PPCM. Role of Selenium and Zinc deficiency in PPCM is controversial.

Etiopathogenesis

Peripartum cardiomyopathy is generally considered a form of idiopathic primary myocardial disease associated with the pregnant state. Although several plausible etiologic mechanisms have been suggested, none of them is definite.

Nutritional: Many nutritional disorders have been suggested as causes, but other than salt overload, none has been validated by epidemiological studies. Higher incidence in African countries has been attributed to the consumption of kanwa, a tradition for 40 post-partum days. Kanwa is a dry salt and causes hypervolemia and hypertension. Ninety percent of PPCM occurs within two months of delivery.

Myocarditis: Association between myocarditis and PPCM was suggested by some investigators who reported a high incidence of myocarditis on endomyocardial biopsy in patients with peripartum cardiomyopathy, later reports however found a comparable incidence in age and sex matched nonpregnant group with idiopathic dilated cardiomyopathy. The prevalence of myocarditis in patients with peripartum cardiomyopathy ranged from 8.8% to 78% in different studies. On the other hand, the presence or absence of myocarditis alone does not predict the outcome of peripartum cardiomyopathy. According to this hypothesis after a cardio tropic viral infections pathologic immune response occurs that is probably inappropriately directed against native cardiac tissue proteins, leading to ventricular dysfunction. Bultmann et al found parvovirus B19, human herpes virus, Epstein-Barr virus and cytomegalovirus DNA in endomyocardial biopsy specimens from 8 (31%) of 26 patients with peripartum cardiomyopathy that was associated immunohisto logically with interstitial inflammation. Kühl et al found, that in patients with viral infection confirmed by endomyocardial biopsy, the median left ventricular ejection fraction improved in those in whom the virus was cleared, whereas it was found to be decreased in those in where the virus persisted.

Chimerism: In a phenomenon called Chimerism, there is mixture of genotype, sometimes provoking an immune response.
The serum from patients with peripartum cardiomyopathy has been found to contain autoantibodies in high titers, which are not present in serum from patients with idiopathic cardiomyopathy. Most of these antibodies are against normal human cardiac tissue proteins of 37, 33, and 25 kD. Multiparity is a risk factor for the development of this disorder, suggesting that previous exposure to foetal or paternal antigen may elicit an abnormal myocardial inflammatory response. The timing of presentation in the immediate postpartum period supports an autoimmune pathogenesis. Adaptive changes in the maternal immune system allow the foetal tolerance necessary for successful pregnancy and include the induction of suppressor cells. These adaptive changes likely underlie the clinical observation that autoimmune disorders such as multiple sclerosis which affect women during their reproductive years typically have lower relapse rates during pregnancy itself, with a marked increase in the immediate postpartum period. The majority of cases of peripartum cardiomyopathy present in early postpartum period of the same pregnancy during which the restoration of the maternal immune system results in an increase in autoimmune exacerbations in other disorders.

Apoptosis and inflammation: Apoptosis (programmed cell death) of cardiac myocytes occurs in heart failure and may contribute to progressive myocardial dysfunction. Experiments in mice suggest that apoptosis of cardiac myocytes has a role in peripartum cardiomyopathy. Plasma levels of C-reactive protein and tumour necrosis factor alpha (markers of inflammation) were found to be elevated and correlated with higher left ventricular dimensions and lower left ventricular ejection fractions at presentation. Recent studies have indicated that increased proteolytic cathepsin D activity in cardiomyocyte result in 16kDa prolactin fragment with angiogenic and apoptotic properties which may contribute to development of this disease. Pharmacological blockade of prolactin might emerge as a novel disease specific therapeutic option in near future. Another observation that there is elevation in the concentration of the inflammatory cytokines such as tumour necrosis factor-alpha in post partum period has been attributed as a causative factor by some workers.

An abnormal hemodynamic response: during pregnancy blood volume and cardiac output increases. In addition the after load decreases because of the relaxation of vascular smooth muscle. The increase in volume and cardiac output causes transient and reversible hypertrophy of the left ventricle to meet the needs of the mother and foetus. The transient left ventricular systolic dysfunction during the third trimester and early postpartum period returns to baseline once the cardiac output decreases, given the hemodynamic stress decompenation usually occurs during pregnancy in women with previous subclinical ischemic or myopathic heart disease in last trimester of pregnancy but in cases of PPCM because of some unknown familial, genetic or environmental factors this decompenation starts late in gestation and in more than 75% cases it is diagnosed only during post partum period, but the same argument stands against this theory as well.

Latent idiopathic Cardiomyopathy: According to this theory unmasking of latent idiopathic dilated cardiomyopathy is brought to fruition by the hemodynamic stresses of pregnancy. Hypertension during pregnancy, and pre-eclampsia, are both reported in a higher frequency in women with peripartum cardiomyopathy and support that hemodynamic stresses may play a role. However keeping the normal hemodynamic changes of pregnancy in mind, it is well known that most women with compromised cardiac function such as valvular heart disease present with symptoms of heart failure by the end of the second trimester. Absence of cardiac symptoms in PPCM until the postpartum period argues against the latent cardiomyopathy theory.

Pathological Changes

In a patient of PPCM after post mortem the heart specimen appear pale, soft, dilated and heavier in comparison to normal heart. Cardiac chambers quite often show mural thrombi, Gray white patches of endocardial thickening are usually seen at the site of mural thrombi. Heart valves and coronary arteries appear normal, pericardial effusion is occasionally seen. On histopathological examination evidence of degeneration, fibrosis, interstitial oedema, and fatty and mononuclear cell infiltration along with sparse or abundant collection of eosinophils in myocardium is seen, myocardial hypertrophy is also found in some patients. Electron microscopy reveals varying degree of enlargement, destruction or fragmentation of myofibrils, increase in size and number of mitochondria, along with the deposition of glycogen and some abnormal proteinacious material. On histochemical examination of myocardial cells, the most important observation is the presence of sarcoplasmic fat vacuoles containing triglyceride without any increase of the lipofuscin or amyloid. Besides these, the low levels of plasma albumin and pre albumin, selenium and zinc have also been observed as mentioned earlier.

Clinical Features

Symptoms of PPCM are the same as in patients with systolic dysfunction who are not pregnant. New or rapid onset of the following symptoms require prompt evaluation: cough, orthopnea, paroxysmal nocturnal dyspnoea, fatigue, palpitations, weight gain, haemoptysis, chest pain, and unexplained abdominal pain. Physical examination often reveals enlarged heart, tachycardia, and decreased pulse oximetry. Blood pressure may be normal, elevated jugular venous pressure, third heart sound, and loud pulmonic component of the second heart sound are usually found, mitral and/or tricuspid regurgitation, and pulmonary rales are also noted. Worsening of peripheral oedema, ascites and hepatomegaly are quite often seen. Arrhythmias are commonly found which may be responsible for embolic phenomenon peripheral or pulmonary. In some patients small to moderate pericardial effusion may be found. Presence of elevated blood pressures (systolic>140 mm Hg and/or diastolic>90 mm Hg) and proteinuria suggests preeclampsia. Overall clinical presentation and hemodynamic changes are indistinguishable from those found in other forms of dilated cardiomyopathy. Few patients with high output heart failure have also been reported.

Diagnosis

Diagnosis of PPCM requires a very high level of suspicion by treating physicians and obstetrician. They should consider peripartum cardiomyopathy in any peripartum patient with unexplained symptoms of heart failure. The greatest dilemma is the lack of specific clinical criteria allowing distinction between PPCM presenting with the new onset heart failure and other causes of systolic dysfunction. Therefore all other possible causes of dilated heart with heart failure must be thoroughly excluded before accepting clinical diagnosis of PPCM.

Lab Investigations

X-ray chest: may reveal cardiomegaly and pulmonary venous
congestion with interstitial or alveolar oedema. Occasionally, pleural effusion may be found. As x-ray chest is not essential to diagnose PPCM, its routine use during pregnancy should be discouraged and if needed may be done using an abdominal shield.

ECG: may be normal or might show sinus tachycardia, atrial fibrillation, non-specific ST segment changes, conduction abnormalities and other types of arrhythmias.

Echocardiography: Echocardiography remains an important tool for evaluation and follow up for women with postpartum cardiomyopathy. The finding of a decrease in myocardial systolic function, as manifested by a decrease in left ventricular ejection fraction or fractional shortening is essential to the diagnosis. Left ventricular dilatation is also frequently evident, particularly in those women presenting late. Mild compensatory left ventricular hypertrophy can be seen, however, marked increases in LV wall thickness may suggest primary hypertrophic cardiomyopathy, an entity with a very distinct natural history and prognosis. A small pericardial effusion may be seen in the early and immediate postpartum period. Valvular morphology is generally normal; however, with marked LV enlargement mitral regurgitation secondary to annular dilatation may be seen, tricuspid and pulmonary regurgitation are also seen some times. Overall echocardiographic features of postpartum cardiomyopathy are indistinguishable from those of primary non-ischemic dilated cardiomyopathy.23

Endomyocardial biopsy: The endomyocardial biopsy may show features of myocarditis, as many cases of PPCM seem to be related to myocarditis, but the decision for biopsy should be taken after thorough discussion between patient and treating physicians. Biopsies in patients with peripartum cardiomyopathy have the highest yield when performed early after the onset of symptoms; however there is paucity of literature on this aspect of diagnostic modality.

Viral and bacterial titer and cultures: such as coxsackie B virus antibody titer should be considered in selected cases, these have more research implications rather than real diagnostic one.

MRI: Magnetic resonance imaging (MRI) may be used as a complementary tool to diagnose peripartum cardiomyopathy, and it may prove to be important in identifying the mechanisms involved. It can measure global and segmental myocardial contraction, and it can characterize the myocardium. Delayed contrast enhancement (with gadolinium) can help to differentiate the type of myocyte necrosis, i.e. myocarditis vs. ischemia. Myocarditis has a nonvascular distribution in the subepicardium with a nodular or band-like pattern, whereas ischemia has a vascular distribution in a subendocardial or transmural location. Cardiac MRI can be used to guide biopsy to the abnormal area, which may be much more useful than the blind biopsy. Recently Cardiac Magnetic Resonance Imaging (CMRI) has been used in prognostication in PPCM.24

Right heart catheterization: In women with persistent heart failure, hemodynamic instability or evidence of an organ dysfunction, right heart catheterization to assess the filling pressures and cardiac output should be considered. It will also demonstrate enlargement of all chambers of heart predominantly the left ventricle.

Biochemical evaluation: Usually reveals little or no elevation in creatinine kinase, or cardiac troponin. In women with acute heart failure and hemodynamic compromise, assessment of liver function tests and renal function provides an assessment of organ perfusion.

Differential Diagnosis

PPCM should be differentiated from other forms of Cardiomyopathy. The most common and confusing being Idiopathic Dilated Cardiomyopathy (IDCM), though PPCM is identical to IDCM in many ways, most researchers now agree that it is a distinct clinical entity.25 PPCM occurs at a younger age and is generally associated with better prognosis, it occur mostly post partum (78-93%) whereas IDCM usually manifests by the second trimester. Higher incidence of myocarditis is found in PPCM along with unique sets of antigen and antibodies against myocardium which is not seen in IDCM. Heart size returns to normal after delivery in more number of PPCM patients as compared to IDCM and contrary to IDCM, PPCM may lead to rapid worsening of clinical course and poor outcome.26 Other common causes of heart failure, such as valvular heart diseases, coronary artery disease including acute myocardial infarction, pulmonary thromboembolism, severe eclampsia and pneumonia should be excluded on the basis of history, physical examination and investigations.

The most common alternative diagnosis is the occult valvular heart disease which can be effectively ruled out by transthoracic echocardiography. The finding of normal systolic function excludes postpartum cardiomyopathy and should lead to an evaluation for forms of high output failure such as anaemia and thyrotoxicosis. Although ischemic heart disease is uncommon in this population, women with significant risk factors such as Type I diabetes should have at least a non-invasive assessment for coronary ischemia, and if questions persist should undergo angiography. In women with persistent heart failure, hemodynamic instability or evidence of an organ dysfunction, right heart catheterization to assess filling pressures and cardiac output should be considered.

Treatment

Management of Heart failure

A. During Pregnancy: Early diagnosis and prompt treatment are the keys to optimize pregnancy outcome. When considering diagnostic tests or treatment during pregnancy, the welfare of the foetus should always be considered along with that of the mother. Patients with severe forms of heart failure will require ICCU management, with monitoring of arterial blood pressure (ABP), central venous pressure (CVP), and sometimes pulmonary artery catheter (PAC). Coordinated management with specialist’s team is essential. Angiotensin converting enzyme (ACE) inhibitors and ARBs are contraindicated in pregnancy because these can cause birth defects,27 although remaining the main treatment option for postpartum women with heart failure. The teratogenic effects occur particularly in the second and third trimester, characterized by foetal hypotension, pulmonary hypoplasia, oligohydramnios, anuria, and renal tubular dysplasia. However a recent study suggested risk of malformations even after first trimester exposure to ACE inhibitors.

Digoxin, loop diuretics, sodium restriction and drugs that reduce afterload such as hydralazine and nitrates have been proven to be safe and are the mainstays of medical therapy of heart failure during pregnancy. Digoxin is effective due to its inotropic and rate reducing effect. Diuretics are useful because of the preload reduction along with salt restriction. They are relatively safe in pregnancy and lactation; however one should be cautious regarding volume depletion which may result in to dehydration causing uterine hypoperfusion.
...and foetal distress. Calcium channel blockers were not used earlier because of fear of negative contractile effects and potential risk of uterine hypoperfusion but recently Amlodipine has been found to improve survival in non ischemic Cardiomyopathy patients. Beta-blockers have strong evidence of efficacy in patients with heart failure, but they have not been tested in peripartum cardiomyopathy. Nevertheless, beta-blockers have long been used in pregnant women with hypertension without any known adverse effects on the foetus, and patients taking these agents prior to diagnosis can continue to use them safely.

**B. During Post Partum period** - Treatment is identical to that for nonpregnant women with dilated Cardiomyopathy. ACE inhibitors and ARBs are useful. The usual target dose is one half the maximum antihypertensive doses. Diuretics are given for symptomatic relief; spironolactone or digoxin is used in patients who have New York Heart Association class III or IV symptoms. The dose of spironolactone is 25 mg/day after dosing of other drugs is maximized. The goal with digoxin therapy is the lowest daily dose to obtain a detectable serum digoxin level, which should be kept at less than 1.0 ng/ml. Beta-blockers are recommended as they improve symptoms, ejection fraction, and survival. Non-selective beta-blockers such as carvedilol and selective ones such as metoprolol have shown benefit. The goal dosage is carvedilol 25 mg twice a day or metoprolol 100 mg once a day.

**Anticoagulant Therapy**

Due to high risk of venous and arterial thrombosis anticoagulation with subcutaneous heparin should be instituted in these patients more so in bedridden patients, those with LVEF <35%, presence of atrial fibrillation, mural thrombi, obese patients and those with history of thromboembolism. Hypercoagulable state of pregnancy coupled with stasis of blood due to ventricular dysfunction makes PPCM patients prone for thrombus formation. This situation may persist up to six weeks after postpartum, hence use of heparin is advocated in ante partum period and that of heparin or warfarin in the post partum period, as warfarin is contraindicated in pregnancy because of its teratogenic effect while use of both heparin and warfarin is safe in lactation. Patients with evidence of systemic embolism, with severe left ventricular dysfunction or documented cardiac thrombosis, should receive anticoagulation. Anticoagulation should be continued until return of normal left ventricular function is documented.

**Antiarrhythmic Drugs**

As in other forms of non-ischemic dilated cardiomyopathy, ventricular arrhythmias can be an important clinical issue. No antiarrhythmic agent is completely safe during pregnancy quinidine and procainamide should be tried first because of their higher safety profile. Beta blockers may be useful, for atrial arrhythmias digoxin may be considered. Patients presenting with sudden death or ventricular tachycardia with hemodynamic compromise, strong consideration of an implantable cardioverter defibrillator (ICD) is warranted due to the potential for a fatal recurrence. For patients presenting with symptomatic ventricular tachyarrhythmia which are hemodynamically well tolerated, management can be tempered somewhat by the potential transient nature of the myopathy and amiodarone therapy at 200 to 400 mg orally every six hourly is an alternative. If left ventricular function recovers, the risk of serious arrhythmic event is markedly diminished and amiodarone therapy can be discontinued. For patients with asymptomatic non-sustained ventricular tachyarrhythmia we should not initiate amiodarone therapy, but focus on correction of metabolic abnormalities and consider the addition of a beta receptor antagonist if not already being utilized.

**Newer Treatment Modalities**

**Pentoxifylline** : Treatment with pentoxifylline, a xanthine derived agent known to inhibit the production of tumour necrosis factor alpha, has been shown to improve functional class and left ventricular function in patients with idiopathic dilated cardiomyopathy. Similar to other aetiologies of left ventricular dysfunction, elevated levels of TNF alpha has been found in these patients.

**Role of Bromocriptine and Cabergoline** : Most recently few studies have suggested the role of prolactin breakdown products in the aetiology of PPCM. Prolactin secretion can be reduced with bromocriptine which had beneficial effects in a small study. In one case study with use of cabergoline which is a long lasting antagonist of prolactin significant improvement in left ventricular functions were reported. It is premature to comment about usefulness of these drugs at present but it seems that they might become important treatment modality in management of PPCM in times to come.

**Immune Modulating Therapy** : Given the inflammatory nature of peripartum cardiomyopathy and the occasional appearance of myocarditis on endomyocardial biopsy, immunosuppressive and immune modulatory therapy has been utilized. Intravenous immunoglobulin improved the ejection fraction in several studies and also markedly reduced the levels of inflammatory cytokines. Plasmapheresis has also been utilized effectively for this purpose, and may be an alternative to immune globulin therapy in peripartum cardiomyopathy. Recently a small controlled study of immunoadsorption therapy in idiopathic dilated cardiomyopathy demonstrated significant improvements in left ventricular function, suggesting that therapies directed against humoral autoimmunity may have a significant role in the treatment of this disorders. Other proposed therapies which might be useful are calcium channel antagonists, statins, monoclonal antibodies and interferon beta.

**Other Interventions and Devices** : Because the disease is reversible in good number of patients, the temporary use of an intraaortic balloon pump or left ventricular assist devices may help in stabilizing critical patients. Extracorporeal membrane oxygenation has been tried successfully in some patients as a bridge to recovery. Ventricular tachycardia leading to cardiac arrest has been reported in PPCM patients, to avoid such situation increasing use of automated implantable cardioverter defibrillator (AICD) is being tried. In extreme situations multiorgan support systems such as ventilator therapy, continuous venovenous hemodialysis may be required besides circulatory assist devices. Plasmapheresis and immunoadsorption therapies are other newer techniques which have been tried for immunomodulation in these patients.

**Cardiac transplantation** : Patients with severe heart failure who does not respond despite maximal drug therapy may be considered for cardiac transplantation to survive because of high risk of mortality. Two reports comparing results of cardiac transplantation in age matched females with peripartum cardiomyopathy and idiopathic cardiomyopathy showed...
Follow-Up Management

Patients who show normal left ventricular function on echocardiographic evaluation at rest or with low-dose dobutamine stress test can be allowed to taper and then discontinue heart failure treatment in 6 to 12 months. They are encouraged to remain as active as their functional status allows, however aerobic activities and heavy lifting are discouraged for at least the first six months postpartum. Given the metabolic demands of lactation, breast feeding is strongly discouraged in more symptomatic patients. As pharmacologic therapy to the patient can be passed on to the child in breast milk discouraging the breast feeding in more functional patients. If breastfeeding is considered in these women, it has to be with careful monitoring of the baby. Echocardiogram should be repeated at 6 months post delivery. For those patients with persistent cardiomyopathy beta blockers may be added at this point if not already on therapy.

Prognosis

Although peripartum cardiomyopathy shares many features of other forms of dilated cardiomyopathy, an important distinction is that women with this disorder have a much higher rate of spontaneous recovery of left ventricular function on echocardiography in post partum period; nearly half of the women will normalize their ejection fraction during follow-up within six months. Prognosis is directly correlated to recovery of left ventricular function. For those women whose LVEF normalizes during follow-up, the prognosis is excellent as without the stimulus of a subsequent pregnancy the chance of development of heart failure or future LV dysfunction is minimal. For those women whose left ventricular function does not recover, prognosis remains guarded and mortality rates as high as 10-50% has been reported. LV size is an important predictor, as women presenting without significant LV dilatation appeared to have a greater chance of spontaneous recovery during follow-up. In contrast, women with marked LV dilatation at presentation appeared to have a greater likelihood of developing into a chronic cardiomyopathy. Initial NYHA class or hemodynamic status does not seem to predict the likelihood of subsequent recovery. A fractional shortening on echocardiogram less than 20% and a LV end diastolic dimension greater than or equal to 6 cm was associated with a threefold increase in persistent LV dysfunction.

Risks of Subsequent Pregnancies

In patients whose left ventricular function fails to normalize during follow-up, subsequent pregnancies carry a high risk of left ventricular deterioration and progressive heart failure, and hence pregnancy is strongly discouraged given the possible risk to the life of the mother. Mortality was reported to be in the range of 8 to 17 percent in a large study in this group in comparison to 0 to 2% in patients with normal left ventricular ejection fraction before the subsequent pregnancy. Real problem lies in the issue of how to advice 70 to 80% of women who seem to have a complete recovery of left ventricular size and function following PPCM. In an attempt to clarify the prognosis in these cases, women with a history of PPCM underwent a dobutamine stress echocardiography test to assess their left ventricular contractile reserve (Lampert et al 1997). The results of this study indicated that women with recovered PPCM has significantly less left ventricular contractility reserve than a group of normal matched controls. These finding are of concern and indicate a possible though small risk in women with apparently recovered left ventricular function. They should be explained duly during counselling. A recent large retrospective survey suggests that in women with normal ejection fractions at the time of subsequent pregnancy, there was an approximate 21% risk of the development of heart failure, and a drop in the mean ejection fraction from 0.56 to 0.49. However no serious complications were noted and the majority of these women had a successful delivery at term with close and careful monitoring. This was in marked contrast to the women with persistent LV dysfunction prior to pregnancy in which 19% mortality was reported. Overall recommendation clearly being that pregnancy is avoided in women with persistent poor left ventricular function. Women whose LV function normalizes should still be made aware of the risk of possible recurrence, dobutamine stress test should be performed and due counselling should be performed though in the majority of these women successful pregnancy can be accomplished with appropriate monitoring.

Prognosis

Peripartum cardiomyopathy is an uncommon but potential life threatening cardiac failure of unknown aetiology, encountered late in pregnancy or in the post partum period. Diagnosis of PPCM should essentially include echocardiographic substantiation of left ventricular dysfunction. Role of endomyocardial biopsy in day to day diagnosis is controversial. Usefulness of diuretics, vasodilators, digoxin, beta blockers and anticoagulant in medical management is well established. ACE inhibitors and ARB blockers should be avoided during pregnancy but should be started in post partum period. In resistant cases pentoxifylline, immunoglobulin and immunosuppressive drugs may be used. Bromocriptine and cabergoline hold promise for the future. Severe cases might require advanced life support systems and even heart transplantation. Prognosis is linked to recovery of left ventricular functions. Subsequent pregnancies are associated with a very high mortality more so in those whose LV functions do not improve even six month after puerperium hence pregnancy should be avoided in this group. In patients with normal cardiac function on echo evaluation subsequent pregnancy may be considered under close multidisciplinary supervision.

References


