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DILATED CARDIOMYOPATHY IN YOUNG ADULTS – CHALLENGES AND OUTCOME

Cardiology

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ABSTRACT

Background: Dilated cardiomyopathy (DCM) refers to a heterogenous group of disorders affecting the myocardium characterized by progressive ventricular dilation and dysfunction. Clinical profile, echocardiographic profile, prognosis and outcomes were assessed in 61 patients with DCM between 18 and 40 years of age and were periodically reviewed and followed up at 1 month, 3 months, 6 months and 1 year.

Resuts: Mean age was 30.9 ± 6.87 years. Among the 61 patients, there was a dominance of males (68.8%) over females (31.2%). On follow up, 7 patients (11.5%) had died; 5 (8.2%) had undergone heart transplantation; 31 patients (50.8%) had persistent left ventricular (LV) dysfunction; 6 (9.8%) had complete recovery and 12 (19.7%) had partial recovery of left ventricular systolic function. Overall survival at 1 year was 88.5%.

Conclusion: DCM has variable clinical course. Early diagnosis and optimal therapy improves symptoms and survival.

KEYWORDS

Dilated Cardiomyopathy, LV Dysfunction, Echocardiography

INTRODUCTION

Dilated cardiomyopathy (DCM) is characterized by progressive ventricular dilation and depressed myocardial contractility in the absence of abnormal pressure or volume overload or significant coronary artery disease [1]. Annual incidence varies between 5 and 8 cases per 100,000 population[2]. It represents one of the most common causes of heart transplantation[2]. Patients with DCM can present with a highly variable clinical course. Patients may be asymptomatic or present with dyspnea New York Heart Association (NYHA) class I to IV, decompensated heart failure, arrhythmias and sudden cardiac death. DCM can affect any age group. Most cases of DCM are idiopathic. DCM may be due to genetic and non genetic causes like hypertension, ischemic heart disease, valvular disease, toxins, alcohol, arrhythmias, stress induced, systemic disorders like amyloidosis or hemochromatosis, metabolic, chemotherapy and myocarditis. Even these nongenetic forms are influenced by individual genetic predisposition. Our study aimed to analyse the various clinical presentations and outcomes of DCM in young adults.

METHODOLOGY

This prospective study was conducted at a Tertiary Care Hospital in South India from January 2017 to August 2018. 61 newly diagnosed patients with DCM aged > 18 years and < 40 years were included. DCM was diagnosed by LV dilatation and LV ejection fraction (EF) < 40% on echocardiography. Patients with ischemic heart disease, valvular heart disease, congenital heart disease, chronic kidney disease, cor pulmonale, diabetes mellitus, hypertension, peripheral arterial disease, sepsis, pregnancy or peripartum and malignancy were excluded. History, clinical examination and echocardiography was done. LV dimensions were noted and ejection fraction was measured by Modified Simpsons method. LV diastolic function was assessed. Follow up echo was done after 1, 3 and 6 months and after 1 year to assess improvement after optimal medication such as angiotensin converting enzyme inhibitors, beta blockers and diuretics like furosemide and spironolactone. Recovery of LV systolic function, persistent LV dysfunction, death and heart transplantation were the primary outcomes.

RESULTS

Mean age of study subjects was 31.3 ± 6.97 years. There were 42 males (68.8%) and 19 females (31.2%) with ratio of 2.2:1. Two patients (3.2%) had dyspnea NYHA class I, 42 patients (68.8%) in class II, 11 (18%) in class III and 6 (9.8%) in class IV dyspnea. Two patients presented with recurrent ventricular tachycardia. Echocardiography showed dilated left ventricle associated with reduced fractional shortening (mean 16.15 ± 4.7%) and reduced ejection fraction (mean 31 ± 5.7%). Our study observed that mean LV end diastolic dimension was 59.17 ± 8.5 mm and LV end systolic dimension was 49.75 ± 8.2 mm. LV thrombus was present in 5 patients. Arrythmias were seen in 4 cases (6.6%) out of which 2 had recurrent ventricular tachycardia (VT), 1 had atrial flutter and 1 had atrial fibrillation. Optimal medical management improved symptoms and EF. Follow-up at 3 months, 6 months and 1 year showed that 7 patients (11.5%) had died. The cause of death was refractory heart failure in 4, sepsis and acute renal failure after cardiac transplantation in 1 and sudden cardiac death in two cases. Recurrent hospitalisation for heart failure was seen in 4 patients (6.6%). 5 patients (8.2%) had undergone heart transplantation and 1 patient had Implantable Cardioverter Defibrillator (ICD) inserted. Serial echo showed persistent severe LV dysfunction in 31 patients (50.8%). Normalisation of LV EF was seen in 6 patients (9.8%) and partial recovery of LV function (EF > 45%) was seen in 12 patients (19.7%). Overall survival at 1 year was 88.5%.

DISCUSSION

DCM is a progressive myocardial disease with varied etiologies and presentations. DCM has been defined by the presence of fractional shortening < 25% (>2 SD) or ejection fraction < 45% (>2 SD), and LV end-diastolic diameter >117% (>2 SD of the predicted value or 112% corrected for age and body surface area), excluding any known cause of myocardial disease[2]. Familial DCM is defined by the presence of ≥2 affected relatives with DCM or a relative with unexplained sudden death before the age of 35 years. Randomized clinical trials show 30% to 40% incidence of nonischemic DCM compared with ischemic DCM[8]. Pressure and volume overload along with neurohormonal hyperactivation are involved in cardiac remodelling. Increased deposition of extracellular matrix, increased fibrils diameter and collagen cross-linking, increased ratio of type I to type III collagen and decreased elastin, cardiomyocyte apoptosis and necrosis, low rate of myocardial renewal contribute to this process[3]. Studies have reported recovery of cardiac function in 21–37% of patients[4]. Early treatment can reverse cardiac remodelling within the first 6 months of optimal medication[5]. Studies demonstrate that even though there is initial normalization of LVEF by medical or resynchronization therapy, long term follow up is mandatory because of risk of arrhythmic events and worsening of LV function[6], thereby prioritizing continued medication and follow up.

Our study showed male preponderance (M:F = 2.2:1). Reports show men have 2.5 times increased risk than women[4]. This is probably due to cardio protective nature of oestrogens. However, we also noted that the incidence of death before transplantation was higher among women (3 out of 19 [15.8%]) than men (3 out of 42 [7.1%]).

In our study, the most common presentation was dyspnea NYHA class

II (68.8%) whereas 6 patients (9.8%) had class IV dyspnea on admission. 2 patients presented with VT. 6 patients had symptoms of a nonspecific febrile illness suggestive of viral myocarditis 2-3 months prior to diagnosis. In our sub group, 2 patients were diagnosed with cytomegalovirus (CMV) myocarditis were critically ill requiring ventilatory support. Their LV function recovered dramatically within a month. Such cases of "fulminant myocarditis" due to viral etiology if diagnosed and treated early tend to have better outcomes despite being critically ill at the time of presentation.

The incidence of arrythmias in our study population was 6.6%. Increased risk of arrhythmias indicates pathological involvement of the cardiac conduction system. Kumar et al reported life threatening ventricular arrhythmia rates of 3% to 7% per year[7].

Overall survival at 1 year in our study was 88.5% which was in comparison to a study by Kumar et al[7]. Mortality was higher in patients with class IV dyspnea (50% mortality at 1 year).

The various etiologies and outcomes of young DCM patients in our study population are depicted in Figures 1 and 2 respectively.

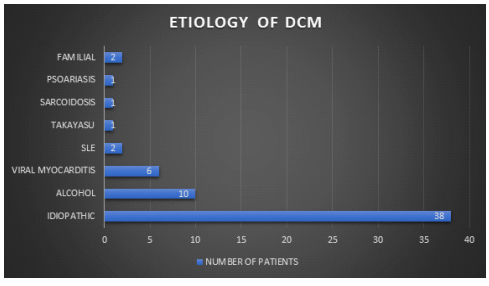

Figure 1: Etiology of DCM in our study population

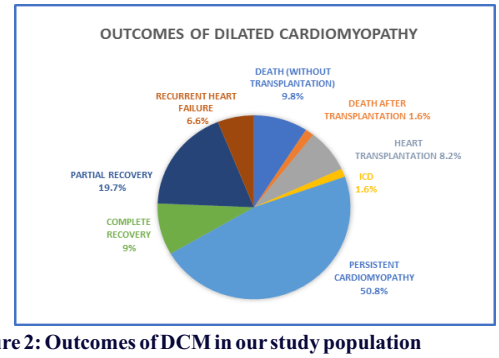

Figure 2: Outcomes of DCM in our study population

Management of DCM is focused on improving LV dimension and function, arrhythmia management and improving quality of life by reducing symptoms. Angiotensin Converting Enzyme Inhibitors (ACE-I), Beta blockers and mineralocorticoid receptor antagonists prevent disease progression and improve survival. Studies have noted that ACE-I inhibit progression of LV dilation and dysfunction whereas beta blockers can improve LV dysfunction in DCM[8]. Patients with DCM with complete left bundle branch block may benefit from cardiac resynchronization. Refractory heart failure can be managed using newer medications like angiotensin receptor-neprylisin inhibitor (valsartan/sacubitril) and the sinoatrial modulator, ivabradine or LV assist devices or cardiac transplantation. Optimal medical management can bring about significant improvement in EF and can lead to lesser number of ICD implantations and heart transplantations and thereby reduce cost and procedural complications.

Study limitations:
Small sample size that limits identification of risk factors for outcome. Long term outcomes > 1 year are not addressed in our subgroup.

CONCLUSION

Outcomes in DCM patients are variable depending upon the time of presentation, risk factors, LV ejection fraction at the time of diagnosis, LV end diastolic dimensions, LV diastolic dysfunction and treatment protocols. Dilated cardiomyopathy in the young is a real challenge and close monitoring and regular follow up with optimal medications play a pivotal role in the outcome and survival of young adults with dilated cardiomyopathy.

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