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Original Research Article

Different Clinical Manifestations of Dilated Cardiomyopathy and Echocardiographic Findings: A Hospital Based Study

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Abstract

Objective: To investigate the various clinical manifestations of dilated cardiomyopathy (DCM), correlate echocardiographic findings with different clinical manifestations, and identify the variables that predict poor outcomes.

Objectives: The present study was carried out with following aims-

- a) To study the different clinical presentations of dilated Cardiomyopathy.
- b) To link Echocardiography findings to various clinical presentations.

c) To identify the variables that contribute to poor outcomes.

Methods: This prospective study will be carried out in patients admitted with symptoms and signs of heart failure in MKCG Medical college and Hospital.

Results: There were 35 (55%) males and 29 (45% females) out of 64 cases, for a male to female ratio of 1.27:1. Out of 64 cases 52 (81 percent) were Idiopathic. In 56 (88 percent) of the cases, dyspnea was the most common presenting clinical feature. In the Echo study, the majority of cases (39%) had an Ejection fraction (EF) in the 36-40% range, and the majority of cases (55%) had severe fractional shortening (FS).

Conclusion: DCM is one of the most common causes of heart failure, as well as the most common type of cardiomyopathy in the middle-aged and elderly population. Males are more prone to it. Biventricular failure is the most common clinical presentation, followed by left ventricular failure. It is critical to identify patients early and begin treatment as soon as possible.

Keywords: Dilated cardiomyopathy, Ejection fraction, heart failure, elderly population

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Introduction

Cardiomyopathy is a primary disease of the heart muscle that causes abnormal myocardial performance and is not the result of disease or dysfunction of other cardiac structures. The predominant feature is direct involvement of the heart muscle itself. They are distinctive in that they are not the result of pericardial, valvular, hypertensive, or congenital disease. [1] The prevalence of heart failure is approximately 1 to 1.5% of the adult population. Mortality and morbidity remain high (median survival of 1.7 years for males and 3.2 years for females. It is three times more common in males than females. It is also more common in black. [2]

Cardiomyopathy defined as a heterogeneous group of disorders of the myocardium associated with mechanical and/or electrical dysfunction, usually (but not always) presenting with inappropriate ventricular hypertrophy or dilatation, arising from a variety of causes and often being genetic. Cardiomyopathies are either confined to the heart or part of a generalized systemic disease that often leads to cardiovascular death or progressive heart failure-related disability. (American Association) Heart BRAUNWALD'S 11th EDITION) [3]. As information expands, this classification triad based on phenotype is becoming increasingly inadequate to define a disease or therapy. The identification of the genetic determinants of cardiomyopathy has suggested a four-way classification scheme: (etiology as being primarily cardiac) and secondary to other systemic diseases. (Harrison 20th edition) [4].

Dilated Cardiomyopathy (DCM)

An enlarged left ventricle with reduced systolic function, as measured by left ventricular ejection fraction, characterizes DCM. It represents the final common pathway generated by a variety of ischemic, toxic, metabolic, and immunological mechanisms that damage cardiac muscle. Although the initial injury to the myocardium can vary. Pathophysiology and clinical presentation are similar in all cultivars. The most common clinical presentation is congestive heart failure, usually left ventricular failure. The patient may also present with symptoms secondary to arrhythmias, stroke (embolic infarction), or sudden death. Systolic failure is more pronounced than diastolic dysfunction. Although DCM syndrome has many different etiologies, many converge on common pathways of secondary response and disease progression. (BRAUNWALD'S 11th EDITION) [3]. The prevalence and incidence of heart failure due to dilated cardiomyopathy in this part of Orissa is quite significant. Despite such a large number of patients with heart failure due to dilated cardiomyopathy coming for treatment to physicians in this part of Orissa, very few studies have been conducted on the clinical profile and echocardiographic abnormalities of DCM.

Objectives

The present study was conducted with the following objectives.

- Examination of the different clinical presentations of dilated cardiomyopathy.
- Correlating echocardiographic findings with different clinical presentations.
- To find out the variables that determine the bad results.

Methods

Selection of cases of DCM

Both male and female patients admitted to cardiology and medical wards of MKCG.M.C.H, Berhampur, during the period from November 2019 to November 2021, were randomly selected and diagnosed as cases of DCM based on clinical and echocardiographic criteria.

Exclusion Criteria: The following patients were excluded from the study.

- 1. Patients with essential hypertension
- 2. Patients with congenital heart disease
- 3. Patients with valvular heart disease
- 4. Patients with coronary artery disease
- 5. Patients with pericardial disease

Inclusion Criteria:

A) Clinical criteria

1) DCM- This was the patient group from which all possible non-familial causes of DCM were excluded.

- 2) Non Familial dilated cardiomyopathies
- a) Peripartum cardiomyopathy-
- Development of heart failure in the last month of pregnancy or within 5 months of delivery.
- Absence of identifiable causes of heart failure.
- Absence of discernible heart disease before the last month of pregnancy.

b) Alcoholic cardiomyopathy (AHA)

• DCM develops in patients who take 80 grams per day for men and 40 grams per day for women for more than 5 years.

c) Anthracycline Cardiomyopathy

• DCM in patients (susceptible individuals) on cancer treatment receiving a cumulative dose of > 550 mg/m2 doxorubicin.

d) Echocardiographic criteria

- The inner dimension of the ventricles at the end of diastole is increased, while the septal and free wall thickness are normal or reduced.
- Abnormal ventricular contractility is the prerequisite for IDC and EF < 45% is generally required for diagnosis.
- Global hypokinesia.
- Intracavitary thrombi are most commonly observed at the left ventricular apex.
- Mitral regurgitation and tricuspid regurgitation due to the dilatation of the ring.

Assessment of LV systolic function

LVEDD is the end of diastole. The normal ranges 3.5. – 5.6cm. LVESD is at the end of systole, which occurs at the peak downward motion of the IVS. The normalranges 2-4 cm.

Fractional shortening is the % change in LV internal dimensions between systole and diastole.

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$$FS = \frac{LVEDD - LVESD}{LVEDD} \ge 100\%$$

Normal ranges 30 - 45%

The ejection of fraction (EF) is the % change in LV volume between systole and diastole and is calculated by

$$EF = \frac{LVEDV - LVESV}{LVEDV} \ge 100\%$$

Wall thickness can be measured. Normal ranges 6-12 mm

Thin - <6 mm as dilated cardiomyopathyThink - >12 mm as LV hypertrophy.

Diastolic Function

Using M mode, motion of anterior mitral value leaflet (AMVL) during diastole has a characteristic M shaped (EA) pattern. In the normal heart, there is characteristic mitral flow pattern. The E wave is the result of passive early diastolic LV filling. The A wave represents active late diastolic LV filling due to LA contraction. The acceleration time (AT) and deceleration time (DT) of the E-wave can be measured.

Slow relaxation pattern - E wave small, A wave large, AT is prolonged, IVRT prolonged. Decrease LV relaxation due to diastolic dysfunction associated with LV hypertrophy or myocardial ischemia.

Restrictive pattern –E wave very tall, A wave is small, DT short, IVRT short. Reduced LV filling may be caused by restrictive cardiomyopathy or constrictive pericarditis (conditions causing rapid use of LV diastolic pressure).

RV function:

Using M mode and 2D echo, estimates can be made of RV internal dimension, wall thickness and ejection fraction.

Study design

The cases of dilated cardiomyopathy selected in this way were questioned in detail about their medical history according to the form. All cases were asked in detail about current history, past history, personal history, family history, and drug history relevant to the development of DCM. A detailed medical history is also taken to rule out secondary causes of DCM such as amount and duration of alcohol consumption, any drug treatment such as doxorubicin for cancer, history of ischemic heart disease, hypertension, diabetes, thyrotoxicosis, hyperthyroidism, as well as a detailed medical history is taken regarding development of DCM symptoms during the peripartum period. The history of sickle cell anemia is also queried on each patient as sickle cell anemia is very common in this part of Orissa. Family history specifically requested that a family origin be ruled out.

Then detailed examinations were performed according to the form and consists of routine blood test (including FBS, PPBS, serum urea, creatinine, Na+, K+), sickle test, Hb electrophoresis, thyroid function test for relevant cases, ECG, chest X-ray and finally echocardiography (2D and Doppler). The results thus obtained were documented, analyzed and conclusions were drawn about clinical features, electrocardiography and echocardiographic correlation of dilated cardiomyopathy.

The present study included 64 cases who fulfilled clinical and echocardiographic criteria for dilated cardiomyopathy.

	No of Cases in Percentage		TOTAL
Age	Female	Male	Number (0/)
	Number (%)	Number (%)	Number (%)
11-20 years	2 (3%)	1 (2%)	3 (5%)
21-30 years	3 (5%)	1 (2%)	4 (6%)
31-40 years	2 (3%)	3 (5%)	5 (8%)
41-50 years	4 (6%)	5 (8%)	9 (14%)
51-60 years	8 (13%)	10 (16%)	18 (28%)
61-70 years	9 (14%)	11 (17%)	20 (31%)
71 and above years	1 (2%)	4 (6%)	5 (8%)
Total	29 (45%)	35 (55%)	64 (100%)

 Table 1: Showing the incidence of age and sex in cases subjected to study



Chart 1

Types of DCM	No. of Cases	Percentage
Idiopathic	52	81%
Alcohol induced Cardiomyopathy	4	6%
Hypothyroidism	3	5%
Thyrotoxicosis	3	5%
Peripartum Cardiomyopathy	2	3%
TOTAL	64	100%

 Table 2: Showing the distribution of DCM due to other causes





Presenting Features	No. of cases	Percentage
Dyspnea	56	88%
Palpitation	17	27%
Peripheral edema	13	20%
Fatigue	5	8%
Syncope	9	14%
Abdominal distension	4	6%
Hemiparesis	3	5%
Fever cough	6	9%
Chest pain	5	8%

 Table 3: Showing the presenting clinical features at the time of admission



Table 4: Distribution of patients presenting with dyspnea according to NYHA Classification

NYHA	Number of cases	Percentage
Ι	0	0
II	19	34.55%
III	14	25.45%
IV	22	40.00%
Total	55	100%

Out of 55 cases presenting with dyspnea, 22 cases (40%) belonged to NYHA class – IV, 19 cases to NYHA class – II (34.55%) and 14 cases to NYHA III (25.45%)

Ejection Fraction in %	No. of cases	Percentage
15-20	2	3%
21-25	3	5%
26-30	7	11%
31-35	19	30%
36-40	25	39%
41 & above	8	13%
Total	64	100%

Table 5: Obse	rvations - E	jection fraction
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Maximum number of cases (n=25) was having EF in the range (36-40) followed by 19 cases in the range (31-35).



Chart 4	ĺ
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Table 6: Observation – Fractional Shortening			
Fractional Shortening	No. of Cases	Percentage	
Mild (20-25%)	6	9%	

9		0
Mild (20-25%)	6	9%
Moderate (15 – 20%)	23	36%
Severe (<15%)	35	55%
Total	64	100%

Chart 5

Out of 64 patients, 55% patients had severe fractional shortening, 36% had moderate, 9% had mild fractional shortening.

Findings	No. of cases	Percentage
Mitral regurgitation	23	36%
Tricuspid regurgitation	17	27%
Aortic regurgitation	8	13%
Left ventricular clot	4	6%
Left atrial enlargement	32	50%
Right ventricular dilatation	23	36%
Pericardial effusion	2	3%
Diastolic dysfunction	6	9%
Pulmonary artery hypertension	4	6%

Table 7: Other associated findings in Echocardiography

Chart 6

Analysis of clinical and echocardiographic findings with final outcome

Out of 64 cases, 9 patients died, 17 patients were in NYHA grade – IV, 15 were in NYHA Grade – II and 11 were in NYHA Grade –III.

Clinical Feature EF (<35%) EF (>35%) Total				
Syncope	9	1	10	
Without syncope	23	31	54	

 Table 8: NYHA Grade

This table shows that out of 64 cases, 10 cases presented with syncope, 9 of which were associated with severe LV dysfunction. Using Fisher's exact test to test the association of syncope with LV function severity, the value with probability of significance (0.05) is 0.0488 and the p-value is < 0.05. Therefore, the association of syncope and LV function severity is statistically significant, and therefore syncope can be considered a poor prognostic factor.

 Table 9: Comparison of Echocardiographic features with clinical outcomes and its statistical significance.

Variables	Severe Symptoms	Mild to mod symptoms	P value
EF < 35%	15	17	<0.05
EF > 35%	5	27	<0.03
LA Size $> 40 \text{ mm}$	17	15	<0.05
LA Size < 40mm	4	28	<0.03
RV Size > 26mm	15	8	<0.05
RV Size < 26mm	5	36	<0.03
LVEDD > 52mm	13	31	>0.05
LVEDD < 52mm	8	13	~0.03

Table 9 shows that severe symptoms (NYHA IV) have a statistically significant association with an ejection fraction < 35. For degrees of freedom, the Yates-corrected chi-square statistic is 4.5037. The p-value is 0.033822. Symptom severity also has a statistically significant association with enlarged left atrial size (> 40 mm) and right ventricular dilation (> 26 mm). The X2 value was 7.4449 and 13.1435, respectively, and the p-value is 0.006362 and 0.000289, respectively. But the extent of left ventricular end-diastolic interior dimension has no statistically significant association with symptom severity. The X2 value was 0.061 and the p-value is 0.804933.

Correlation between Ejection Fraction and Fractional shortening

Fractional shortening has moderate correlation with ejection fraction and is a good predictor of LV dysfunction

Discussion

Out of 64 cases there were 35 males (55%) and females 29 (45%), giving male female ratio of 1.27:1 the maximum number of cases (n=20) appeared in the age of 61-70. The mean age was 56 years. The mean age of males was 60 and that females was 53 years. In various epidemiological studies the *incidence* and prevalence of DCM were found to be morein males than in females. In 2021 by Orphanou *et al* [5] reported that prevalence seems to slightly higher in males with a female to male ratio between 1:1.3 and 1:1.5. H Mahmaljy *et al* [6] reported that most patients were seen between ages of 20 and 60 yrs. But the disorder may also affect children and the elderly.

Etiological Profile:

Table 10				
Etiology	Our Study(%)	Dudharejia <i>et al</i> [7] (%)	Srinivasan <i>et al</i> [8] (%)	
Idiopathic	81	74	12	
Alcohol	6	12	15	
Peripartum	3	2	9	
Thyrotoxicosis	5			
Hypothyroidism	5			
Diabetic	-	12	11	
Ischemic	-		47	

As per the table 2, our study matches with study conducted by Dudharejia*et al* [7], with idiopathic as the leading cause. Many studies have included ischemic cardiomyopathy as one of the cause of DCM. In a study by C.R.Srinivasan*et al* [8], only 12% were idiopathic but 47% were ischemic cardiomyopathy. According to the new definition of DCM, we excluded ischemic cardiomyopathy. Hence the variation in the etiological profile compared to other studies. Idiopathic disorders are the most common cause in most studies. Since no genetic studies could be performed in MKCG, familial causes of DCM could not be identified.

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Symptomatology

Dyspnea was the most common presenting clinical feature in 56 cases (88%) followed by palpitation 17 cases (27%) peripheral edema 13 cases (20%), syncope of 9 cases (14%) chest pain 5 cases (8%). 65.4% patients presented with NYHA grade III and IV. This was almost in accordance with Fuster*et al* [9], who reported that the most common presentation was dyspnea which occurred in 75.85% of patients; out of which 90% of patients had symptoms typical of NHYA class III & IV at the time of diagnosis. He also found palpitation in 30%, peripheral edema in 29%, chest pain on acceleration was present in 8 to 20% of patients, Systematic embolism in 1.5 to 4% of cases. In an Indian study by Routary*et al* [10], almost all cases presented with dyspnea.

Table 11				
Symptomatology	Our Study(%)	Ahmad <i>et al</i> (%) [11]	Sachin <i>et al</i> (%) [12]	
Dyspnea	88%	96.3	100	
Pedal edema	20%	56	70	
Cough	9%	56.3	60	
Palpitation	27%	65.4	56.6	
Abdominal pain	6%	41.8	33.3	
Syncope	14%	1.8	16.6	

As shown in table VIII, patients who presented with syncope, had severe left ventricular dysfunction. After applying X2test, the association of syncope and sudden death was statistical significant and syncope can be considered as a poor prognostic factor. Olshausen KV (AHJ)[13] in his study showed that syncope was associated with 19% of death.

Echocardiography revealed mitral regurgitation in 36%, tricuspid regurgitation in 27%, LV clot in 6%, pericardial effusion in 3%, pulmonary HTN in 6%, left atrial enlargement, and Dilatation of the right ventricle detected in 50% and 36% of cases. As seen in Table 5 and Table 9, EF < 35% has a significant association with statistically symptom severity but no significant association with left ventricular end-diastolic dimension. Symptom severity was also related to left atrial size (> 40 mm) and right ventricular dilation.

Rossi A *et al* [14] describes functional mitral regurgitation was strongly associated with the outcome of patients with HF independently of

LV systolic function. PuwanantS *et al* [15] demonstrated that Right ventricular dilatation and dysfunction have prognostic significance and are correlated with a worse functional status and advanced LV failure.

Rabbani MU *et al* (JAPI 2002) [16], gives a picture of LV diastolic dysfunction in 27.8% of DCM patients. Left ventricular clot was present in 3 cases, one of which 2 cases presented with the left side hemiparesis. This makes about 6% of total cases. Following findings were found in these studies G. Singh SB [17] 2.9% of cases, Felker *et al* [18] 1-12% and Gracia Fernandez *et al* (2000) [19] 19%.

The ejection fraction & left atrial diameter of these 4 patients were 27%,39%,40% and 32mm,38mm,46mm respectively. This was agreeing with Purohit BV *et al* (2002) who showed that presence of LVclot in patients with DCM was associated with advanced age, low ejection fraction and large left atrial size. Vasan RS *et al* (NEJM) [20] increase in echocardiographic left ventricular internal dimensions was a risk factor for the development of congestive heart failure in

DCM. In studies elsewhere investigators were divided over the significance of left ventricular end diastolic dimension in assessing the severity. Unverferth DV (AMJ) [21] factors such as duration of symptoms, presence of mitral regurgitation & end-diastolic diameter were not significant predictors. Hagar et al [22] IDCM patients from 2009 to 2016 and stated that - following patient category had an improved prognosis: patients with LVEF \geq 40%, with device therapy, and those admitted to a cardiology ward. Thapa RK et al [23] conducted echocardiographic evaluation from 1st of February to 31st July 2018 in Kathmandu -Majority had reduced Left ventricular systolic function with an average (EF) of 39.6%.

As shown in chart-7, Fractional shortening has moderate correlation with ejection fraction and is a good predictor of LV dysfunction. This is in accordance with study conducted by Carmine Zoccaliet *et al* [24] where systolic function was evaluated by endoFS(fractional shortening), midwall FS& ejection fraction and there was moderate correlation between LV systolic function and Fractional shortening, but failed to predict all-cause mortality.

Conclusion

The prevalence and incidence of dilated cardiomyopathy in this part of our state is quite significant and it is one of the commonest causes of cardiac failure. Dilated cardiomyopathy is the most common type of cardiomyopathy and an important cause of congestive heart failure. Dilated cardiomyopathy is common in the middle aged and elderly population. It is more common in males. The most common clinical presentation is biventricular failure followed by left ventricularfailure. The most common type is idiopathic/familial followed bv cardiomyopathy, alcoholic peripartum cardiomyopathy, thyrotoxicosis and hypothyroidism. Chest radiograph showed cardiomegaly in most patients. Pulmonary plethora was seen in significant number of patients. Pleural effusion was seen less frequently. Arrhythmias should be monitored by holter monitoring as there is risk of sudden death associated with DCM. Echocardiography revealed reduced ejection fraction and global hypokinesia universally. Mitral regurgitation was present in significant number of patients. Ejection fraction correlated well with NYHA class. In summary, early detection and prompt initiation of treatment are very important as some patients may develop reverse remodeling and decrease the adverse events. Proper identification of family members of patients idiopathic/familial dilated with cardiomyopathy is necessary.

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