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Study of Different Clinical Presentations of Dilated Cardiomyopathy (DCM) and to Correlate Echocardiographic Findings with Different Clinical Presentations and to Find Out the Variables Which Determine Poor Outcomes

Bibhujit Padhy

Assistant Professor, MKCG Medical college

ABSTRACT:

Objective: To study the different clinical presentations of dilated Cardiomyopathy(DCM) and to correlate echocardiographic findings with different clinical presentations and to find out the variables which determine poor outcomes.

OBJECTIVES:

Keeping this in view; the present study was undertaken with following aims.

a) To study the different clinical presentations of dilated Cardiomyopathy.

b) To correlate Echocardiography findings with different clinical presentations.

c) To find out the variables which determine the 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 from November 2019 to November 2021.

Results: out of 50 cases there were 28(56%) males and 22(44%) females giving male to female ratio 1.27:1. Out of 50 cases 40(80%) were Idiopathic. Dyspnea was most common presenting clinical feature in 43(86%) cases.In Echo study maximum number of cases 21(42%) were having Ejection fraction(EF) in the range of 36-40% and maximum number of cases 26(52%) had severe fractional shortening(FS).

Conclusion: DCM is one of the commonest causes of heart failure and it is most common type of cardiomyopathy and is common in middle aged and elderly population .More common in males. Most common clinical presentation is biventricular failure followed by left ventricular failure.Early identification and immediate starting of treatment is very important.

Key words -DCM,EF,FS

INTRODUCTION:

Cardiomyopathy is a primary disorder of the heart muscle that causes abnormal myocardial performance and is not the result of disease or dysfunction of other cardiac structures. The dominant feature is a direct involvement of heart muscle itself are distinctive because they are not the result of pericardial, valvular, hypertensive or congenital disease.[1]

The prevalence of heart failure is about 1 to 1.5 %of adult population. The mortality and morbidity remain high (median survival of 1.7 years for men and 3.2 years for women. It occurs 3 times more frequently in males as compared to females. It is also more common in black.[2]

Cardiomyopathy defined as "a heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilation and are due to a variety of causes and frequently are genetic. Cardiomyopathies either are confined to the heart or are part of a generalized systemic disorder often leading to cardiovascular death or progressive heart failure–related disability." **(American Heart Association) – BRAUNWALD'S 11th EDITION)** [3]

Expanding information renders this classification triad based on phenotype increasingly inadequate to define disease or therapy. Identification of genetic determinants of cardiomyopathy has suggested a four-way classification scheme of (etiology as primary affecting primarily the heart) and **secondary** to other systemic disease. **(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 produced by a variety of ischemic, toxic, metabolic and immunological mechanisms damaging the heart muscles. Though the initial insult to the myocardium may vary. Pathophysiology and clinical presentation are similar in all varieties. The most common clinical presentation is congestive heart failure, usually left ventricular failure. The patient can also present with symptoms secondary to arrhythmias, stroke (embolic infraction) or sudden death. *Systolic failure* is more prominent than diastolic dysfunction. Although the syndrome of DCM has many disparate etiologies many converge to 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. In spite of 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 regarding the clinical profile and echocardiographic abnormalities of DCM.

Hence I have taken interest to take this A Study of clinical and Echocardiographic profile of DILATED CARDIOMYOPATHY.

OBJECTIVES :

Keeping this in view; the present study was undertaken with following aims.

d) To study the different clinical presentations of dilated Cardiomyopathy.

e) To correlate Echocardiography findings with different clinical presentations.

To find out the variables which determine the poor outcomes.

METHODS :

Selection of cases of DCM

Patients both males and females, admitted in cardiology and medicine wards of MKCG.M.C.H, Berhampur, during the period of November 2019- November 2021, randomly taken and diagnosed as cases of DCM on clinical and echocardiography 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

These were the group of patients from which all possible non familial causes of DCM were excluded.

2) Non Familial dilated cardiomyopathies

a) Peripartum cardiomyopathy (AHA – pub – Apr 2016)

- Development of cardiac failure in the last month of pregnancy or within 5 months of delivery.
- Absence of identifiable causes of cardiac failure.
- Absence of recognizable heart disease before the last month of pregnancy.

b) Alcoholic cardiomyopathy (AHA)

DCM developing in patients taking 80 gram per day for males and 40 gram per day for females for more than 5 years.

c) Anthracycline Cardiomyopathy

DCM in a patients (Susceptible individuals) on cancer treatment receiving a cumulative dose of > 550mg/m^2 of doxorubicin.

Echocardiographic criteria

1) Internal dimension of ventricles at end diastole are increased while septal and free wall thickness reminds normal or reduced.
2) Abnormal ventricular contractility is the sine qua non of IDC and EF< 45% is generally required for diagnosis.
3) Global hypokinesia.
4) Intracavitary thrombi most frequently seen at the left ventricular apex.
5) Mitral regurgitation and tricuspid regurgitation due to dilation of annulus.

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 normal ranges 2-4 cm.

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

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

Normal ranges 30 - 45%

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

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

Wall thickness can be measured.

Normal ranges 6-12 mm

Thin - <6 mm as dilated cardiomyopathy

Think - >12 mm as LV hypertrophy.

Diastolic Function

2D echo does not help to make care direct assessment of LV diastolic dysfunction. 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. AT is the time from onset of diastolic flow to the peak of E Wave. The deceleration time is the time from peak to the point where the deceleration slop hits the baseline.

If the LV is stiffer than usual then

Diminished AMVL excursion (E wave)

Increase in A wave size.

Reduced E: A ratio

The isovolumic relaxation time is usually 48 - 65 ms. The IVRT often increases the diastolic dysfunction.

Two abnormal mitral flow patterns or recognized.

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.

Intracardiac thrombus:

2D imaging is the best technique to identify thrombus which is usually echo bright, the following favor the diagnosis of thrombus.

Mural thrombus does not thicken during systole while myocardium thickens.

PWall motion near a thrombus is nearly always abnormal whereas it is often normal near other pathology (eg., tumor)

Thrombus usually has clear identifiable edge which distinguishes it from hazy stagnant blood.

Color floor mapping can distinguish thrombus from stagnant flow.

Study design

The cases of dilated cardiomyopathy thus selected were asked thoroughly about the history of their diseases as per the proforma. All cases were asked in detail about, the present history, past history, personal history, family history and drugs history, which were relevant for evolution of DCM. Detailed history is also asked to rule out secondary causes of DCM like amount and duration of intake of alcohol, any history of drug treatment like doxorubicin for cancers, history of ischemic heart disease, hypertension, diabetes, thyrotoxicosis, hyperthyroidism, also detailed history is taken regarding development of symptoms of DCM during the peripartum period. Also history of sickling is asked in every patient, as sickle Cell disease is highly frequent in this part of Orissa. Family history specifically asked to rule out familial origin.

Then detailed investigations were done as per the proforma and consists of routine blood examination (including FBS, PPBS, Serum Urea, Creatinine, Na^+, K^+) sickling 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 conclusion drawn about clinical features, electrocardiography and echocardiographic correlation of dilated cardiomyopathy.

The present study included 50 cases who fulfilled clinical and echocardiographic criteria for dilated cardiomyopathy.

TABLE – 1

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

Age	**No of Cases in Percentage**					**TOTAL**
	Female		**Male**			
	No	**%**	**No**	**%**		
11-20	1	2%	0	0	1	2%

21-30	2	4%	0	0	2	4%
31-40	1	2%	2	4%	3	6%
41-50	3	6%	4	8%	7	14%
51-60	7	14%	9	18%	16	32%
61-70	8	16%	10	20%	18	36%
71 & above	0	0	3	6%	3	6%
Total	**22**	**44%**	**28**	**56%**	**50**	**100%**

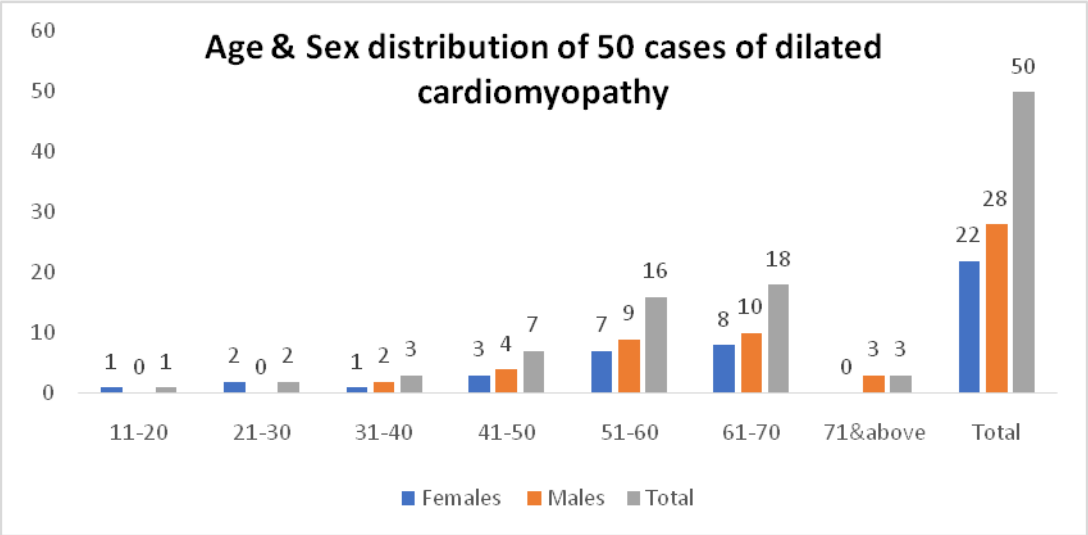


Chart-1

TABLE-2

Showing the distribution of IDCM and DCM due to other causes

Types of DCM	No. of Cases	Percentage
IDIOPATHIC	40	80%
Alcohol induced Cardiomyopathy	4	8%
Hypothyroidism	2	4%
Thyrotoxicosis	2	4%
Peripartum Cardiomyopathy	2	4%
TOTAL	**50**	**100%**

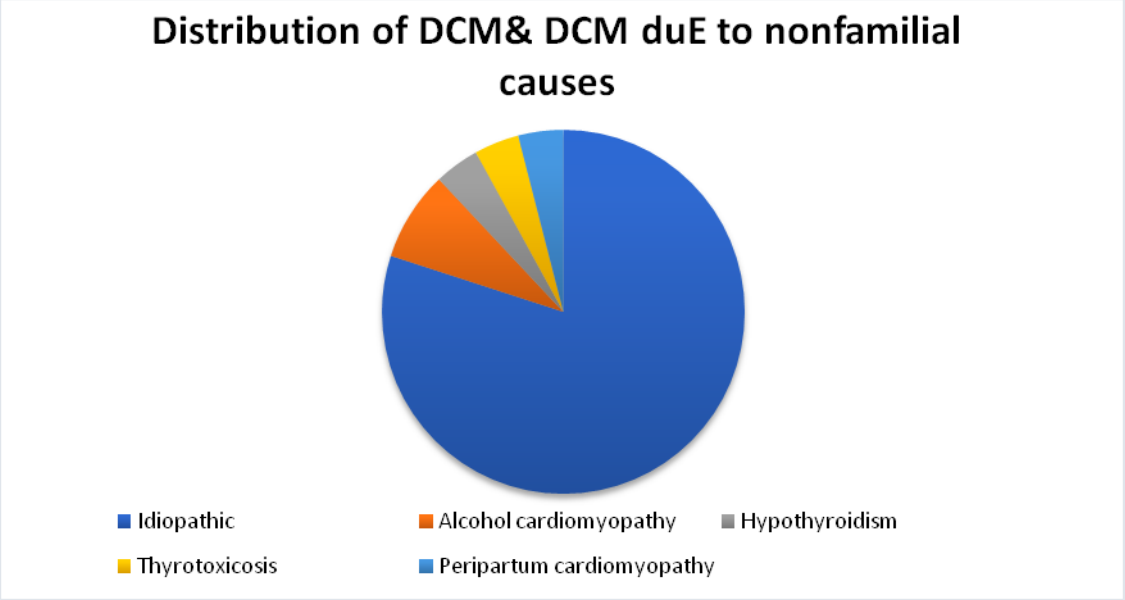


Chart-2

TABLE – 3

Showing the presenting clinical features at the time of admission

Presenting Features	No. of cases	Percentage
Dyspnea	43	86%
Palpitation	14	28%
Peripheral edema	10	20%
Fatigue	4	8%
Syncope	8	16%
Abdominal distension	3	6%
Hemiparesis	2	4%
Fever cough	5	10%
Chest pain	3	6%

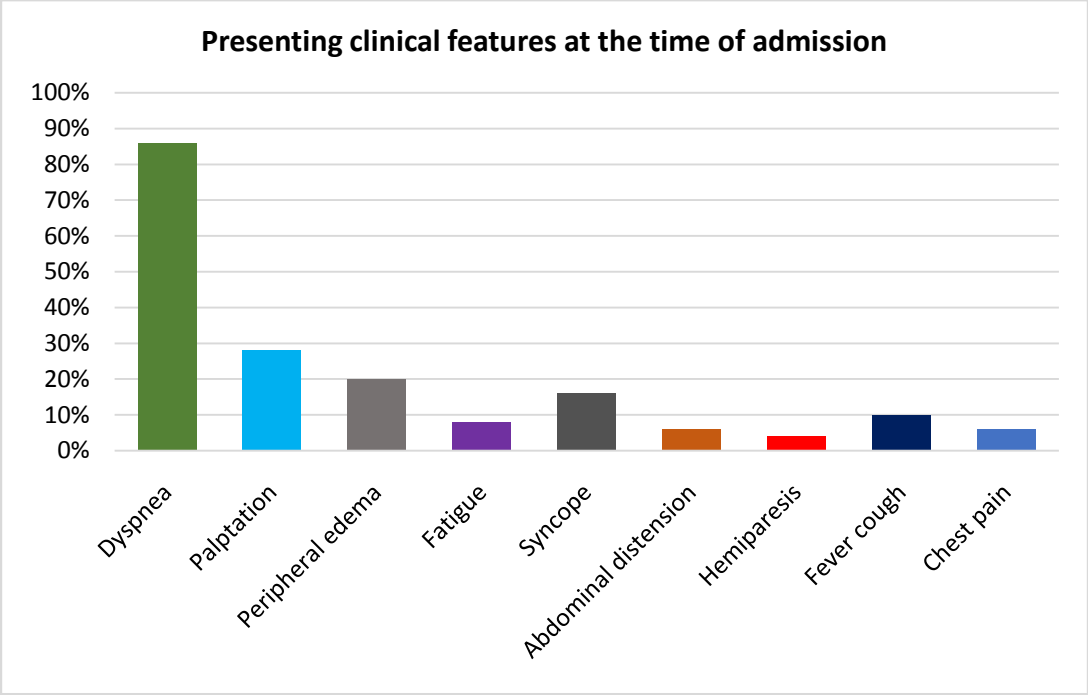


Chart-3

TABLE – 4

Distribution of patients presenting with dyspnea according to NYHA Classification

NYHA	Number of cases	Percentage
I	0	0
II	15	34.88%
III	11	25.58%
IV	17	39.53%
Total	**43**	**100%**

Out of 43 cases presenting with dyspnea, 17 cases (39.53%) belonged to NYHA class – IV, 15 cases to NYHA class – II (34.88%) and 11 cases to NYHA III (25.58%)

TABLE – 5

Observations - Ejection fraction

Ejection Fraction in %	No. of cases	Percentage
15-20	1	2%
21-25	2	4%
26-30	6	12%
31-35	16	32%
36-40	21	42%
41 & above	4	8
Total	**50**	**100%**

Maximum number of cases **(n=21)** was having EF in the range **(36-40)** followed by 16 cases in the range (31-35).

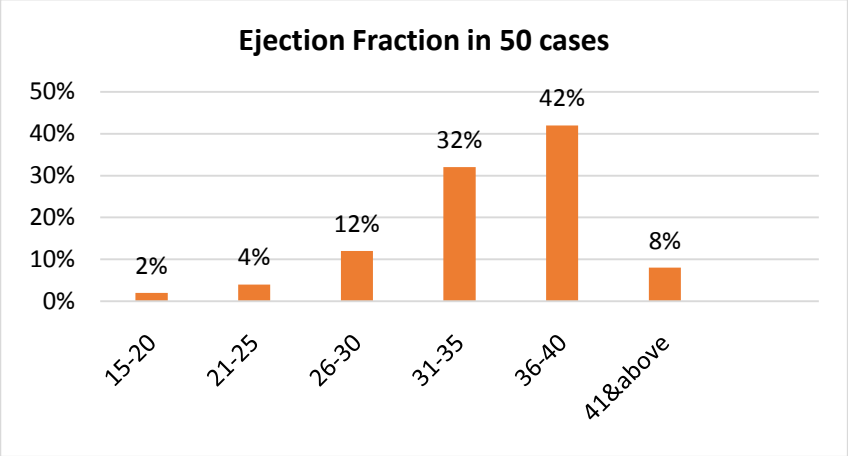


Chart-4

TABLE-6

Observation – Fractional Shortening

Fractional Shortening	**No. of Cases**	**Percentage**
Mild (20-25%)	4	8%
Moderate (15 –20%)	20	40%
Severe (<15%)	26	52%
Total	**50**	**100%**

Out of 50 patients, 52% patients had severe fractional shortening, 40% had moderate, 8% had mild fractional shortening

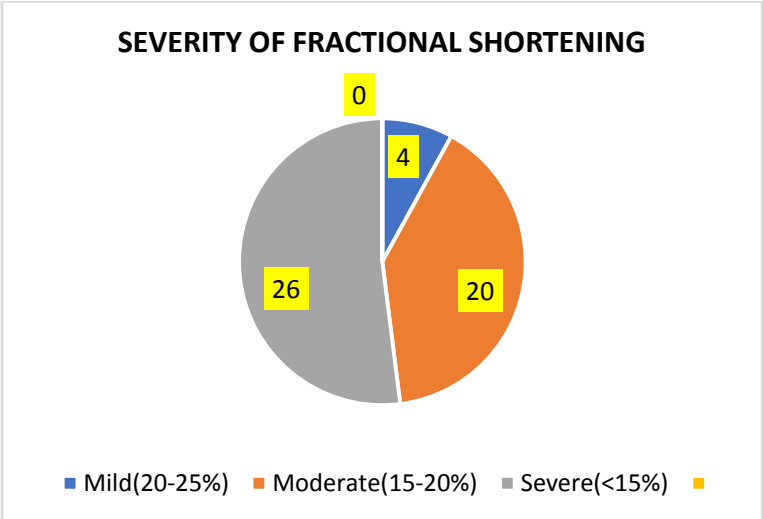


Chart-5

TABLE – 7

Other associated findings in Echocardiography

Findings	No. of cases	Percentage
Mitral regurgitation	18	36%
Tricuspid regurgitation	13	26%
Aortic regurgitation	6	12%
Left ventricular clot	3	6%
Left atrial enlargement	25	50%
Right ventricular dilatation	18	36%
Pericardial effusion	2	4%
Diastolic dysfunction	5	10%
Pulmonary artery hypertension	3	6%

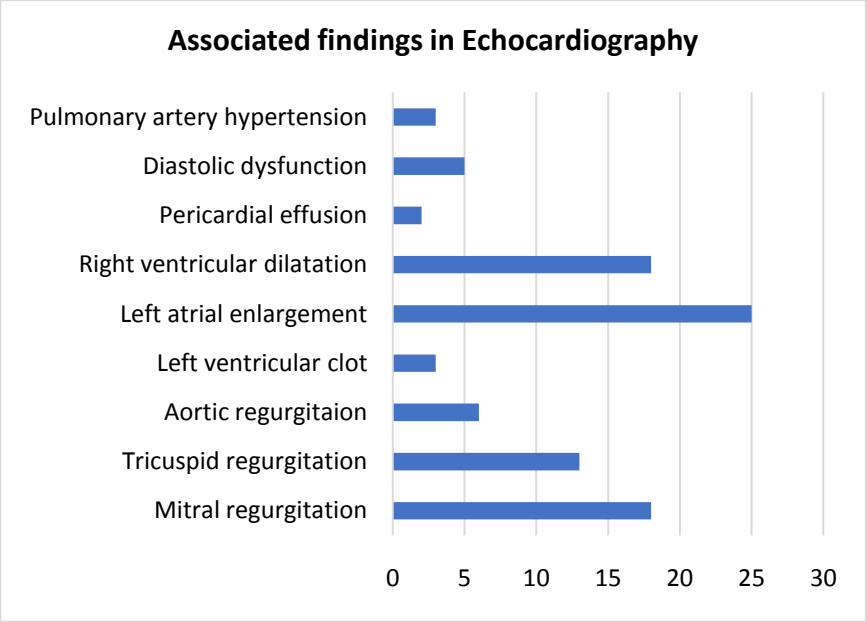


Chart-6

Analysis of clinical and echocardiographic findings with final outcome

Out of 50 cases, 9 patients died, 17 patients were in NYHA grade – IV, 15 were in NYHA Grade – II and 11 were in NYHA Grade –III.

TABLE – 8

Clinical Feature	EF (<35%)	EF (>35%)	Total
Syncope	7	1	8
Without syncope	18	24	42

Total	**25**	**25**	**50**

This table shows that out of 50 cases, 8 cases presented with syncope of which 7 were associated with severe LV dysfunction. On applying the Fisher exact test to test the association of syncope with severity of LV function, with probability of significance (0.05) the value is 0.0488 and p value is <0.05. So the association of syncope &severity of LV function is statistically significant and so syncope can be considered as poor prognostic factor.

TABLE-9

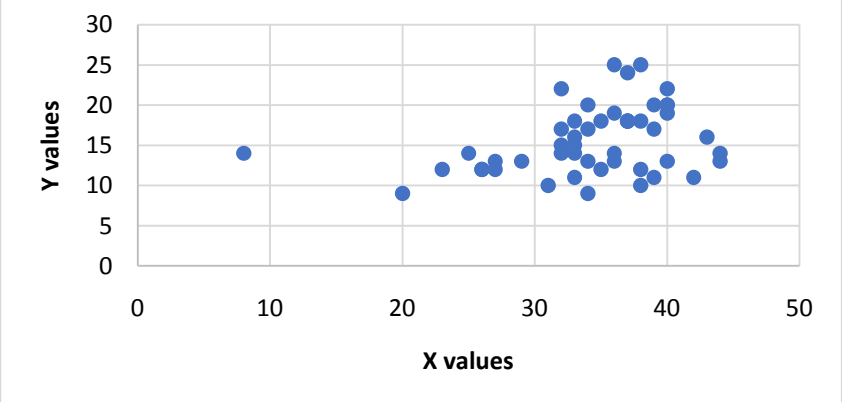
Comparison of Echocardiographic features with clinical outcomes and its statistical significance.

Variables	Severe Symptoms	Mild to mod symptoms	P value
EF < 35%	12	13	<0.05
EF > 35%	4	21	
LA Size > 40 mm	13	12	<0.05
LA Size < 40mm	3	22	
RV Size > 26mm	12	6	<0.05
RV Size < 26mm	4	28	
LVEDD > 52mm	10	24	>0.05
LVEDD < 52mm	6	10	

From table – 9, it is evident that severe symptoms (NYHA – IV) has statistically significant association with ejection fraction <35. At degree of freedom the chi-square statistic with yates correction is 4.5037 p-value is 0.033822. Also the severity of symptoms has statistically significant association with enlarged left atrial size (>40 mm) and right ventricular dilatation (>26 mm). The X^2 value was 7.4449 and 13.1435 respectively and p-value is 0.006362 and 0.000289 respectively. But the extent of left ventricular end diastolic internal dimension has no statistically significant association with severity of symptoms. The X^2 value was 0.061 and p-value is 0.804933.

Chart-7

Correlation between Ejection Fraction and Fractional shortening



R factor – 0.3

P value is <0.05

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

DISCUSSION :

Out of 50 cases there were 28 males (56%) and females 22 (44%), giving male female ratio of 1.27 : 1 the maximum number of cases (n=18) 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 more in males than in females**.**

- In 2021 by **Orphanouet 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 Mahmaljyet 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:

Etiology	**Our Study(%)**	**Dudharejia et al**[7] **(%)**	**Srinivasan et al**[8] **(%)**
Idiopathic	80	74	12
Alcohol	8	12	15
Peripartum	4	2	9
Thyrotoxicosis	4		
Hypothyroidism	4		
Diabetic	-	12	11
Ischemic	-		47

As per the table 2, our study matches with study conducted by **Dudharejiaet 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.Srinivasanet al**[8], only 12% were idiopathic but 47% were ischemic cardiomyopathy. According to new definition of DCM, we have excluded ischemic cardiomyopathy. Hence the variation in etiological profile compared to other studies. Idiopathic is the most common cause in most of the studies. As genetic studies could not be done in MKCG, familial causes DCM could not be identified.

Symptomatology:

Dyspnea was the most common presenting clinical feature in 43 cases (86%) followed by palpitation 14 cases (28%) peripheral edema 10 cases (20%), syncope of 8 cases (16%) chest pain 3 cases (6%), embolism 2 cases (4%). 65.2% patients presented with NYHA grade III and IV.

This was almost in accordance with **Fusteret 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 **Routaryet al**[10], almost all cases presented with dyspnea.

Symptomatology	Our Study(%)	Ahmad et al (%)[11]	Sachin et al (%)[12]
Dyspnea	86%	96.3	100
Pedal edema	20%	56	70
Cough	10%	56.3	60
Palpitation	28%	65.4	56.6
Abdominal pain	6%	41.8	33.3
Syncope	16%	1.8	16.6

As shown in table VIII, patients who presented with syncope, had severe left ventricular dysfunction. After applying X^2test, the association of syncope and sudden death was statistical significantand 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.

Echocardiogram

In echocardiography mitral regurgitation was noted in 36% of cases, tricuspid regurgitation in 26% of cases, LV clot in 6%, pericardial effusion 4%, pulmonary HTN in 6% cases, left atrial enlargement and right ventricular dilatation in 50% and 36% cases respectively.

As seen in table 5 & table 9 symptoms EF<35% has statistical significant association with severity of symptoms but does not have significant association with left ventricular end diastolic dimension. Also the severity of symptoms had association with left atrial size (>40 mm) and the right ventricle dilatation.

Associated findings in echocardiography

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.

- **G. Singh SB**[17] 2.9% of cases
- **Felker et al**[18]1-12%
- **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 ≥ 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 – following were the results –

- Presented mostly with congestive heart failure
- Echocardiographic evaluation showed with mildly dilated Left ventricle
- Majority had reduced Left ventricular systolic function with an average (EF) of 39.6%
- No significant difference between male and female
- Here was no significant relation between age and average EF% (P=0.091).

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), midwallFS& 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 ventricular failure.
- The most common type is idiopathic/familial followed by alcoholic cardiomyopathy, 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 conclusion, early identification and immediate starting of treatment is very important, as some patients can develop reverse remodeling & to decrease the adverse events. Proper identification of family members of patients with idiopathic/familial dilated cardiomyopathy is necessary.

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