

# Symptomatology and Clinical Features of Cardiomyopathy in Adults: A Prospective Study.

Pravin B. Vanjari<sup>1</sup>, Abhaysinh Bhosale<sup>2</sup>, Santosh S. Mali<sup>3</sup>

<sup>1</sup>Assistant professor, Department Of Medicine, Government Medical College Miraj and PVPGH Sangli -Maharashtra, India.

<sup>2</sup>Assistant professor, Department Of Medicine, Government Medical College Miraj and PVPGH Sangli -Maharashtra, India.

<sup>3</sup>Associate Professor, Department Of Medicine, Government Medical College Miraj and PVPGH Sangli -Maharashtra, India.

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## ABSTRACT

**Background:** Cardiomyopathies are diseases of heart muscle that result from myriad of insults such as genetic defects, cardiac myocyte injury, or infiltration of myocardial tissues. Cardiomyopathies result from insults to both cellular elements of the heart, notably the cardiac myocyte and processes that are external to cells such as deposition of abnormal substances into the extracellular matrix. Cardiomyopathies are traditionally defined on the basis of structural and functional phenotypes like dilated characterized primarily by an enlarged ventricular chamber and reduced cardiac performance, hypertrophic characterized by thickened hypertrophic ventricular walls and enhanced cardiac performance and restrictive characterized by thickened stiff ventricular walls that impede diastolic filling of the ventricle. The symptoms vary depending upon the type and severity of cardiomyopathy and may include fatigue, dyspnea, orthopnea and edema. The clinical features may include tachycardia, tachypnea, Raised JVP, pulmonary edema and hepatomegaly amongst others. We conducted this study to know the symptomatology and clinical features in adult patients diagnosed with cardiomyopathy. **Aims and Objectives:** To study the symptomatology and clinical Features of cardiomyopathy in adults. **Methods:** This was a prospective study of 50 patients of cardiomyopathy admitted in the medicine department of a tertiary care medical college situated in an urban area. The study was conducted after due approval from institutional ethical committee and obtaining informed consent from the patients. The patients were included in the study on the basis of predefined inclusion criteria. Patients having any exclusion criteria were excluded from the study. Symptomatology and clinical features of the patients were studied. The data was tabulated and analyzed using SPSS 16.0 version software. **Results:** The study consisted of 50 patients of cardiomyopathy out of which 26 (52%) were males and 24 (48%) were females with a M: F ratio of 1:0.92. Commonest types of cardiomyopathy was found to be dilated cardiomyopathy (46%) followed by ischemic cardiomyopathy (44%). More than 50% of the cases were found to be between the age group of 51 to 70 years. The most common symptoms in these patients were dyspnea (100%), chest pain (30%), Pedal Edema (30%) and palpitation (22%). Class III dyspnea was seen in 21 (42%) patients and class II dyspnea was seen in 18 (36%) patients. The most common Clinical feature seen in patients was found to be raised JVP (34%) followed by murmurs (32 %), basal crepts (32 %) and pedal edema (26 %). **Conclusion:** Cardiomyopathy is one of the important causes of morbidity and mortality in adult population. Early recognition and prompt management of cardiomyopathy will retard the progression of the disease and reduce morbidity and mortality in patients with cardiomyopathy.

**Keywords:** Cardiomyopathy, Signs and Symptoms, Early Recognition, Morbidity and Mortality.

## INTRODUCTION

Cardiomyopathies are diseases of heart muscle that result from myriad of insults such as genetic defects, cardiac myocyte injury, or infiltration of myocardial tissues.<sup>[1]</sup> Cardiomyopathies result from insults to both cellular elements of the heart, notably the cardiac myocyte and processes that are external to cells such as deposition of abnormal substances into the extracellular matrix.<sup>[2]</sup>

Although cardiomyopathy is easily defined classification of its various forms is difficult because majority of cases are associated with generalized cardiac dilation and ventricular systolic dysfunction in which etiology is unknown.<sup>[3]</sup> Knowledge about Cardiomyopathies falls into several categories like etiology, gross anatomy, histology, genetics, biochemistry, immunology, hemodynamic function, prognosis, treatment, and others.<sup>[4]</sup> Several classifications of Cardiomyopathies have been proposed based on extensive research but no single classification utilizes all areas of knowledge with much overlap between them. The only currently used clinical classification of cardiomyopathy developed was that of WHO and international society and federation of cardiology.<sup>[5]</sup>

Cardiomyopathies are traditionally defined on the basis of structural and functional phenotypes like

### Name & Address of Corresponding Author

Dr.Pravin B. Vanjari ,  
Assistant Professor,  
Department of Medicine,  
Government Medical College,  
Miraj and PVPGH Sangli- ,  
Maharashtra,  
India. 416416

dilated characterized primarily by an enlarged ventricular chamber and reduced cardiac performance, hypertrophic characterized by thickened hypertrophic ventricular walls and impaired cardiac performance and restrictive characterized by thickened stiff ventricular walls that impede diastolic filling of the ventricle.<sup>[6]</sup> A recently appreciated structural and functional phenotype is arrhythmogenic right ventricular dysplasia /cardiomyopathy. The dilated cardiomyopathy phenotype is often viewed as a final common pathway of numerous cardiac injuries and is the most common cardiomyopathic phenotype.<sup>[7]</sup> This classification is based upon basic disturbances in function. Importance of this scheme is that all cardiomyopathies are readily placed in one of the three categories & therapeutic approach to each category are distinctly different.

The common symptoms associated with cardiomyopathy may depend upon the type and severity of cardiomyopathy. The usual symptoms which may be seen in any type of cardiomyopathy may include fatigue, exertional dyspnea, paroxysmal nocturnal dyspnea and orthopnea. The signs which may be seen in cardiomyopathy may include pedal edema, increased abdominal girth due to ascites, cyanosis, clubbing, jugular venous distension, basal crepts, S3 gallop and congestive hepatomegaly.<sup>[8]</sup>

There can be history of long standing systemic illnesses like diabetes, hypertension, angina or family history of cardiomyopathy or sudden death in any of the family members. Auscultation may reveal tachycardia, S3 Gallop, congestive hepatosplenomegaly and signs of raised JVP.<sup>[9]</sup> The differential diagnoses which must be considered include acute coronary syndrome, pericarditis, cardiac tamponade and drug toxicity. The workup usually consists of imaging (CXR, thallium scan), Electrocardiography, echocardiography, Cardiac MRI, endocardial biopsy and cardiac catheterization in selected cases.<sup>[10]</sup> The management of cardiomyopathy is similar to treatment of congestive cardiac failure. Drug used to treat cardiomyopathy may include ACE inhibitors, Beta blockers, aldosterone antagonists, cardiac glycosides, diuretics and vasodilators. In some patients having arrhythmias antiarrhythmic drugs may be required.<sup>[11]</sup> Some patients may also need anticoagulation. In some selected cases left ventricular assist devices, cardiac resynchronization therapy and ventricular restoration surgery may be done. Lastly in some patients cardiac transplantation may be the only hope.<sup>[12]</sup>

We conducted this prospective study to know the symptomatology and clinical features of patients having cardiomyopathy. Our emphasis was on early detection so as to be able to intervene at an appropriate time which may be helpful in reducing morbidity and mortality.

## MATERIALS AND METHODS

This was a prospective cohort study of the 50 patients of cardiomyopathy who were admitted in medicine department of a tertiary care institute situated in an urban area. The institutional ethical committee approved the study and informed consent was obtained from all the patients. The patients were included in this study on the basis of a predefined inclusion criteria, any patient having any exclusion criteria was excluded from the study. A detailed history was taken in all the patients with a special emphasis on presence of diabetes, hypertension or ischemic heart disease. If present the duration was also noted down. History suggestive of cardiomyopathy in any of the family member or sudden death of any family member in past was enquired into. Demographic details like age, sex and address were noted down. Presenting complaints and past history was also noted down. General and systemic examination including cardiovascular system examination was done. Pulse rate and rhythm was noted. JVP was obtained. Cardiac auscultation was done to find out presence of abnormal heart sounds like S3 Gallop or murmur. Biochemical investigations like hepatic and renal function tests, electrolytes and blood sugar levels were done in all cases. Chest X ray, ECG and Echocardiography was done in all cases and findings were noted down. The diagnosis of cardiomyopathy was done on the basis of imaging, ECG and 2D-Echo. . The data was tabulated and analyzed using SPSS 16.0 version software.

### Inclusion criteria

Patients were selected from those presenting with

1. Signs and symptoms of congestive cardiac failure.
2. Abnormal ECG changes.
3. A symptomatic patients having unexplained cardiomegaly on chest X-ray.

### Exclusion criteria

1. Valvular heart disease, congenital heart disease and pericardial disease were not included.
2. Patients with history of acute MI were not included.
3. Those who refused consent to be part of this study were excluded from the study.

## RESULTS

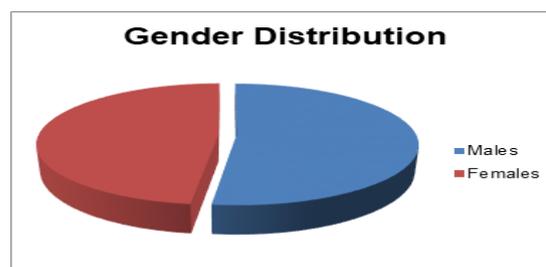


Figure 1: Gender Distribution of the studied cases.

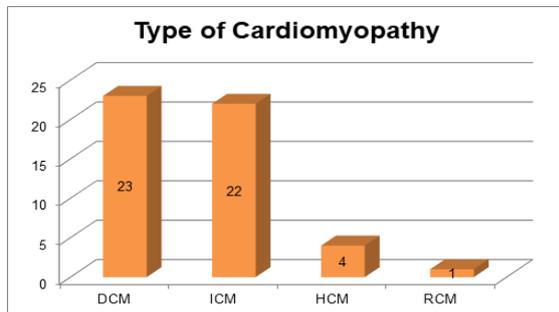
This was a prospective study comprising of 50 adult patients with cardiomyopathy. Out of these 50 patients there were 26 males and 24 females with a M: F ratio of 1:0.92. The difference was not found to be statistically significant.

The analysis of gender distribution on the basis of type of cardiomyopathy showed that in DCM Females were commonly affected (30%) while ICM was more common in males (30%) while in HCM males were predominantly affected (6%).

**Table 1: Gender Distribution and type of cardiomyopathy.**

| Gender | Type of CM |      |                 |                     |      | Total |
|--------|------------|------|-----------------|---------------------|------|-------|
|        | DC M       | IC M | Obstructive HCM | Non Obstructive HCM | RC M |       |
| Male   | 8          | 15   | 0               | 3                   | 0    | 26    |
| Female | 15         | 7    | 1               | 0                   | 1    | 24    |
| Total  | 23         | 22   | 1               | 3                   | 1    | 50    |

The most common type of cardiomyopathy was found to be dilated cardiomyopathy (46%) followed by Ischemic cardiomyopathy (44%). Dilated and Ischemic cardiomyopathy patients formed 90% of the total studied cases. Hypertrophic and restrictive cardiomyopathy was seen in 10% of the patients.



**Figure 2: Type of cardiomyopathy in the studied cases.**

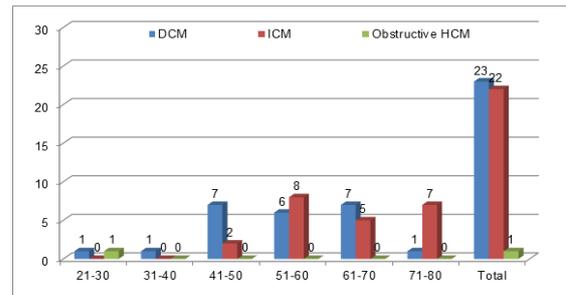
**Table 2: Age distribution of the patients with cardiomyopathy.**

| Age Group | Type of CM |      |                 |                     |      | Total |
|-----------|------------|------|-----------------|---------------------|------|-------|
|           | DC M       | IC M | Obstructive HCM | Non-obstructive HCM | RC M |       |
| 21-30     | 1          | 0    | 1               | 0                   | 0    | 2     |
| 31-40     | 1          | 0    | 0               | 1                   | 0    | 2     |
| 41-50     | 7          | 2    | 0               | 0                   | 0    | 9     |
| 51-60     | 6          | 8    | 0               | 1                   | 0    | 15    |
| 61-70     | 7          | 5    | 0               | 1                   | 1    | 14    |
| 71-80     | 1          | 7    | 0               | 0                   | 0    | 8     |
| Total     | 23         | 22   | 1               | 3                   | 1    | 50    |

The analysis of the patients on the basis of age group revealed that the most common age group to be affected by cardiomyopathy was 51-60 years (30%)

followed by 61-70 years (28%), 41-50 years (18%) and 71-80 years (16%).

The study of type of cardiomyopathy and the affected age group showed that the most common age groups affected in patients with DCM were 41-50 years and 61-70 years while in cases of ICM the most common age group affected was found to be 51-60 years. There was only 1 patient of obstructive HCM belonging to age group of 21-30 years while restrictive cardiomyopathy was seen in 1 patient belonging to age group of 61-70 years.

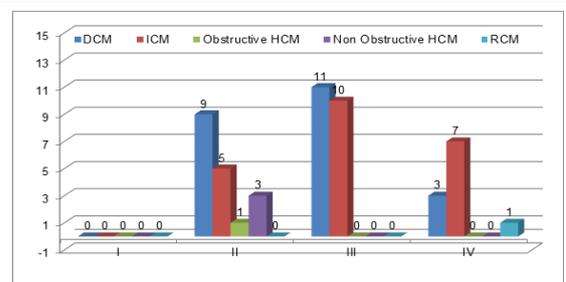


**Figure 3: Age distribution on the basis of type of cardiomyopathy.**

The analysis of symptoms of the patients with cardiomyopathy showed that the most common symptom was dyspnea which was seen in all the patients (100%) followed by chest pain (30%), Pedal edema (30%), palpitations (22%), cough (20%), fatigability (18%) and syncope (6%).

**Table 3: Type of cardiomyopathy and symptoms of the patients.**

| Symptoms Present | Type of CM present |      |                 |                     |      | Total |
|------------------|--------------------|------|-----------------|---------------------|------|-------|
|                  | DC M               | IC M | Obstructive HCM | Non-obstructive HCM | RC M |       |
| Dyspnea          | 23                 | 22   | 1               | 3                   | 1    | 50    |
| Fatigability     | 4                  | 5    | 0               | 0                   | 0    | 9     |
| Palpitation      | 5                  | 4    | 1               | 0                   | 1    | 11    |
| Chest Pain       | 4                  | 10   | 0               | 1                   | 0    | 15    |
| Pedal Edema      | 10                 | 4    | 0               | 0                   | 1    | 15    |
| Syncope          | 0                  | 2    | 0               | 1                   | 0    | 3     |
| Cough            | 6                  | 3    | 0               | 1                   | 0    | 10    |



**Figure 4: NYHA class of dyspnea in various types of Cardiomyopathies.**

Further study of NYHA class of dyspnea showed that the most common class of dyspnea seen was class III (42%) followed by class II (36%) and class IV dyspnea (22%).

The analysis of the clinical features of the patients with cardiomyopathy showed the most common clinical feature in this patient was raised JVP (34%) followed by murmur (32%), basal crepts (32%), edema feet (26%), S3 Gallop (16%) and Hepatomegaly (14%).

**Table 4: Type of cardiomyopathy and signs found in the patients.**

| Clinical Features | Type of CM present |     |                 |                     |     | Total |
|-------------------|--------------------|-----|-----------------|---------------------|-----|-------|
|                   | DCM                | ICM | obstructive HCM | Non-obstructive HCM | RCM |       |
| Raised JVP        | 10                 | 6   | 0               | 0                   | 1   | 17    |
| Edema feet        | 8                  | 4   | 0               | 0                   | 1   | 13    |
| Thrill            | 2                  | 1   | 1               | 2                   | 0   | 6     |
| Murmur            | 8                  | 5   | 1               | 2                   | 1   | 17    |
| S3 Gallop         | 6                  | 2   | 0               | 0                   | 0   | 8     |
| Basal Crepts      | 9                  | 5   | 0               | 1                   | 1   | 16    |
| Hepatomegaly      | 4                  | 3   | 0               | 0                   | 0   | 7     |
| Ascites           | 1                  | 2   | 0               | 0                   | 0   | 3     |

## DISCUSSION

In our prospective study of 50 patients with the commonest types of cardiomyopathy was found to be dilated cardiomyopathy (46%) followed by ischemic cardiomyopathy (44%) and non obstructive hypertrophic cardiomyopathy (6 %). Similar findings were seen in study conducted by Hollister R.M et al.<sup>[13]</sup> In their study of 52 patients with CM, 25 patients had DCM, 18 patients had HCM and 9 patients had RCM. Similar results were reported by Amash et al.<sup>[14]</sup>

The analysis of the cases with respect to gender and type of cardiomyopathy showed that in DCM group 15 out of 23 patients (65.52%) were females and 8 (34.48%) were males. In ICM group 15 out of 22 patients (68.18%) were males and 7 (31.92%) were females. As seen together DCM and ICM 23 out of 45 patients (51.11%) were males and 22 (48.89%) were females. Thus there was a slight male preponderance in our study. Similar male preponderance in the patients with cardiomyopathy was found in the studies conducted by Fuster et al and Rihal et al.<sup>[15,16]</sup>

The analysis of patients with hypertrophic cardiomyopathy showed that the mean age of the patients was 44 years and the M: F ratio was found to be 3:1. These findings were consistent with the

findings as reported by Braunwald et al in their study of 64 patients with cardiomyopathy. In our study of patients with hypertrophic cardiomyopathy there was no patient with family history of cardiomyopathy as well as history of sudden death in any of the family members. This was in contrast to the studies conducted by many authors who studied large number of patients with hypertrophic cardiomyopathy. Maron et al in their study of 70 families of HCM patients found that in 39 families, other members showed evidence of HCM of these 30 families had autosomal dominant transmission.<sup>[17]</sup> This variation may be due to small number of patients with HCM in our study. The analysis of symptomatology of the patients with HCM revealed that Dyspnea was invariable and was present in all 4 patients (100%). All patients had NYHA class 2 dyspnea. Palpitation was present in 1 patient while chest pain and syncope was present in 1 patient each. Goodwin et al found that out of 29 patients the common symptoms were dyspnea (69%), chest pain (38%), palpitation (13.8%) and syncope (41%).<sup>[18]</sup> The analysis of clinical features of the patient in the patients with HCM showed that 2 out of 3 patients of non-obstructive HCM had systolic murmur and precordial thrill and 1 patient had respiratory rales. Goodwin et al found that out of 29 cases 26 (90%) had murmur, 23 (79%) had LV apex, 10 (34%) had thrill and 20 (69%) had third or fourth heart sound. Braunwald et al reported that out of 64 cases all had murmur (100%), 33 (51%) had LV apex, 45 (73%) had thrill and 38 (59%) had third or fourth heart sound.<sup>[19]</sup>

The analysis of dilated cardiomyopathy showed that in DCM group 15 out of 23 patients (65.52%) were females and 8 (34.48%) were males. In ICM group 15 out of 22 patients (68.18%) were males and 7 (31.92%) were females. As seen together DCM and ICM 23 out of 45 patients (51.11%) were males and 22 (48.89%) were females. Thus there was slight male preponderance in our study. These results were similar to the study conducted by parole et al who also found a male preponderance in the patients with DCM. In the present study, dyspnea was invariable and is present in all 23 patients (100%). Dyspnea was class2 in 9 (40%), class3 in 11 (47%) and class4 in 3(13%) of patients. Fatigability (17%), palpitation (21%), chest pain (17%), pedal edema (43%), cough (25%) were other predominant symptoms. Abdominal pain, syncope were uncommon symptoms. None of the patients were asymptomatic. In patients with ICM dyspnea was present in all 22 patients (100%). Dyspnea was class2 in 5 (22%), class 3 in 10 (46%) and class 4 in 7 (32%) patients. Fatigability (22%), palpitation (18%), chest pain (46%), swelling of feet (18%), syncope (9%) were other common symptoms seen in these patients. In India, Parale et al (2001) have reported dyspnea in 100% patients. The finding of this study is consistent with our study.<sup>[20]</sup> In the present study most patients

with ICM and DCM (69%) had higher NYHA class of dyspnea (class3-4) as compared with other studies. This may be because of the fact that in our setup patients usually present late.

## CONCLUSION

Cardiomyopathy is one of the important causes of morbidity and mortality of cardiac origin. Symptoms and clinical features mostly depend upon the type and severity of cardiomyopathy. Early diagnosis and appropriate intervention will reduce the morbidity and mortality associated with cardiomyopathy. Medical management, ventricular assist devices, cardiac resynchronization therapy and ventricular restoration surgery are some of the management strategies depending upon the severity of the disease and general condition of the patient. Cardiac transplantation may be needed if all above measures fail.

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