

## ECHOCARDIOGRAPHIC FINDINGS IN HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY (HOCM)

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**Contribution**

All the authors contributed significantly to the research that resulted in the submitted manuscript.

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### ABSTRACT

**Objective:** To find out various Echocardiographic findings in patients with HOCM.

**Methodology:** This was a retrospective cross-sectional study performed in Cardiology department Lady Reading Hospital, Peshawar. Data collected from the database computer section of echocardiography department from February 2009 to November 2011. The data was analyzed using SPSS version 14.

**Results:** Total study population was 28. Male were 14 (50%). Mean age was  $52.5 \pm 15.9$  years. Mean left ventricular end diastolic dimension was 3.84cm; inter-ventricular septal thickness 2cm (1.1-3.1) and posterior wall thickness was 1.17cm (0.6-1.6). Mean Left atrial (LA) diameter was 3.86cm (0.8-5.6). Mean peak gradient across LVOT was 48.43 mmHg. Mitral regurgitation (MR) was found in 19 (67.9%) patients. MR was mild in 57.1%, moderate in 7.1% and severe in 3.6% of patients. Aortic regurgitation (AR) was found in 13 (46.4 %) patients. AR was mild in 35.7%, moderate in 10.7% of patients. Left atrial size was increased in 14 (50%) patients. Mean LA diameter was 4.6cm, 5cm and 5.6cm in patients with mild, moderate, and severe MR respectively. So there was direct relation between severity of MR and LA diameter. The respective mean gradient across LVOT in patients mild, moderate and severe MR was 31mmHg, 43.5mmHg and 140mmHg. So higher the gradient across LVOT, more will be the MR and hence the LA size and the patient will be more symptomatic.

**Conclusion:** HOCM is significantly associated with both MR and AR and there is direct relation between severity of MR with LA diameter and LVOT gradient.

**Key Words:** HOCM, MR, AR, LVOT gradient, LA diameter, Interventricular Septum

## INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a heterogeneous cardiac disease with a diverse clinical presentation and course, presenting in all age groups from infancy to the very elderly.<sup>1,2,3</sup> HCM is a disease state characterized by unexplained LV hypertrophy associated with non dilated ventricular chambers in the absence of another cardiac or systemic disease.<sup>1,2,4,5</sup>

HCM is a global disease, the prevalence is about 0.2% (i.e., 1:500) in the general population, which is equivalent to at least 600,000 people affected in the United States.<sup>6,7,8</sup> Most affected individuals probably achieve a normal life expectancy without disability or the necessity for major therapeutic interventions.<sup>9-12</sup> On the other hand, in some patients, HCM is associated with disease complications that may be profound, with the potential to result in disease progression or premature death.<sup>1,2,3,13,14,15</sup> In fact, realistic mortality rates for HCM are only about 1% per year.<sup>6</sup>

HCM is caused by an autosomal dominant mutation in genes that encode sarcomere proteins or sarcomere-associated proteins.<sup>4,5,8</sup> HCM is caused by mutations in any one of 10 genes and appears in 50% of individuals in each generation.<sup>6</sup> First-degree family members should undergo periodic screening with echocardiography every five years for this autosomal dominant disorder.<sup>8</sup>

The clinical diagnosis of HCM is conventionally made with cardiac imaging, at present most commonly with 2-dimensional echocardiography and increasingly with Cardiovascular Magnetic Resonance (CMR). Clinically, HCM is usually recognized by maximal LV wall thickness  $\geq 15$  mm.<sup>7</sup> In the case of children, increased LV wall thickness is defined as wall thickness  $\geq 2$  standard deviations above the mean (z score  $\geq 2$ ) for age, sex, or body size.<sup>7</sup> There is a relatively linear association between maximal wall thickness and sudden death, with highest risk in patients with wall thickness  $\geq 30$  mm.<sup>16</sup>

In fact, one third of patients have no obstruction either at rest or with physiologic provocation, one will have obstruction under basal (resting) conditions (defined as gradients  $\geq 30$  mm Hg) and one third or more of patients will have labile, physiologically provoked gradients ( $< 30$  mm Hg at rest and  $\geq 30$  mm Hg with physiologic provocation).<sup>17</sup> Outflow obstruction usually occurs in HCM by virtue of mitral valve systolic anterior motion (SAM) and mitral-septal contact.<sup>18-23</sup> It is clinically important to distinguish between the obstructive and non obstructive forms of HCM because management strategies are largely dependent on the presence or absence of symptoms caused by obstruction.<sup>7</sup>

Mitral regurgitation is common in patients with left ventricular outflow obstruction (LVOT) obstruction.<sup>18, 19, 24</sup>

Evaluation of the presence and degree of mitral regurgitation is performed by color Doppler echocardiography. Mitral regurgitation occurs in almost all patients with obstructive HCM as a consequence of systolic anterior motion (SAM).<sup>26</sup> There is direct relation between the pressure gradient and the severity of MR.<sup>25,26</sup> In one study MR was found in 90% of cases of HCM, MR jet was posteriorly directed in 97% and centrally directed in 3%.<sup>26</sup> In other study MR was found in all cases of SAM and 50% of cases without SAM.<sup>27</sup>

Aortic regurgitation (AR) has been sparsely reported in Hypertrophic Cardiomyopathy.<sup>28</sup> Color Doppler echocardiography showed an aortic regurgitant signal in 23% of the patients.<sup>29</sup>

LA dimension is a novel and independent marker of prognosis in HCM, particularly relevant to the identification of patients at risk for death related to heart failure. Increase in LA dimension was predictive of long-term outcome, independent of co-existent atrial fibrillation or outflow obstruction.<sup>30</sup> The prevalence of atrial fibrillation increases progressively with age and LA size, which in turn is related to the degree of hypertrophy, severity of MR, and diastolic dysfunction.<sup>31</sup>

The aim of this study was to find out various Echocardiographic finding in patients with HOCM.

## METHODOLOGY

This was a retrospective cross sectional study performed in Cardiology department Lady Reading Hospital, Peshawar. Data was collected from the data base computer section of echocardiography department from February 2009 to November 2011. The data was analyzed using SPSS version 14.

## RESULTS

Total patients were 28 in the study. 14 (50%) patients were male and 14 (50%) patients were female. Mean age was 52.5 years (range 5-90). Mean left ventricular end diastolic dimension (LVEDD) was 3.84 cm. Mean inter ventricular septal thickness (IVS) was 2 cm (1.1-3.1). Mean posterior wall thickness was 1.17cm (0.6-1.6). All patients with HOCM were having systolic anterior motion (SAM). Mean Left atrial (LA) diameter was 3.86cm (0.8-5.6). Mean peak gradient across Left ventricular outflow tract (LVOT) was 48.43 mmHg (Table 1).

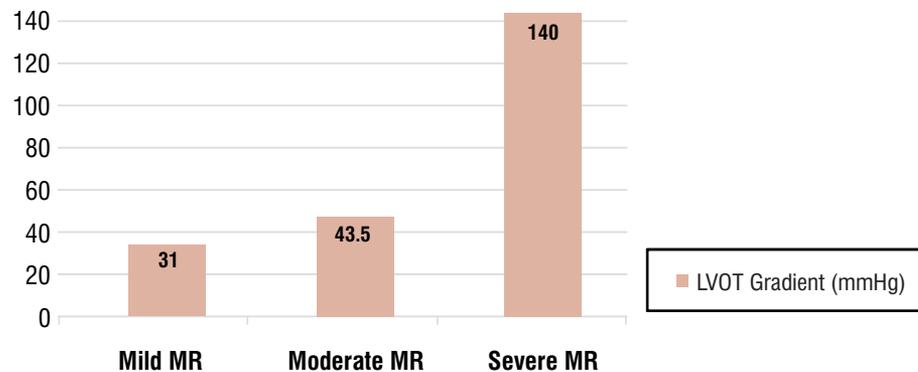
Mitral regurgitation (MR) was found in 19 (67.9%) patients (Fig 1). MR was mild in 57.1%, moderate in 7.1% and severe in 3.6% of total patients. Aortic regurgitation (AR) was found in 13 (46.4 %) patients. AR was mild in 35.7%, moderate in 10.7% of patients. Left atrial size was increased in 14 (50%) of patients. Moreover there was a direct relation between severity of MR and LA diameter. Mean LA diameter was

**Table 1: Echocardiographic Findings in Patients with HOCM**

	Mean	Minimum	Maximum
LVEDD(cm)	3.84	2.5	6.10
IVS(cm)	2	1.1	3.1
PW(cm)	1.17	.6	1.8
LA Diameter(cm)	3.86	.8	5.6
LVOT peak gradient(mmHg)	48.43	10	140

Left ventricular end diastolic dimension (LVEDD), Inter ventricular septal thickne ss (IVS), Left ventricular outflow tract (LVOT), Posterior wall thickness (PW), Left atrial (LA)

**Figure 1: Relation Between Severity of MR and LVOT Gradient**



4.6cm with mild, 5cm with moderate and 5.6 cm with severe MR.

In our study we compared the mean gradient across LVOT with severity of MR. we found that in patients with mild MR, mean gradient across LVOT was 31 mmHg. In moderate MR, mean gradient across LVOT was 43.5 mmHg, and with severe MR it was 140 mmHg (Figure 1). So higher the gradient across LVOT, more will be the MR and hence the LA size and the patient will be more symptomatic.

**DISCUSSION**

We studied various Echocardiographic findings in patients with HOCM. All patients were having asymmetric septal hypertrophy. All patients with HOCM were having SAM. In previous studies it was described that SAM and asymmetric hypertrophy is important feature of HOCM and is present in almost all cases of HOCM.<sup>8,18-20</sup>

In our study we found equal prevalence of disease in both sexes. The disease was found in any age group. The age of patients ranged from 5 to 90 years in our study which strongly favors' the findings of studies done by Maron et al

and Towbin et al that HOCM is found in any age group.<sup>1,4</sup>

We found that almost all patients were having small LV. The mean LVEDD in our study was 3.84 which shows that these patients were having small LV cavities which are found in other studies as well.<sup>1,32-34</sup> In our study the mean IVS thickness was 2.00cm which is equal to the findings of Kiavar et al who found the mean IVS thickness in HOCM patients is 2±0.7cm.<sup>35</sup> The mean PW thickness in our study was 1.17cm which is favored in a study done by Michalak et al who found posterior wall thickness 1.2±0.4.<sup>36</sup>

In our study the mean LA diameter was 3.86 and 50% patients was having increase LA size. Previous studies also found increase in LA size in patients with HOCM.<sup>30</sup> LA size is important to find out because patients with increase LA size are having more prevalent AF and poor prognosis. Moreover we found that MR gradient is correlated with LA diameter. More the severity of MR, more is the mean LA size. This finding is favored by Rakowski et al that LA diameter is proportional to the severity of MR.<sup>31</sup>

We found in our study that MR and AR is very common in patients with HOCM. We found 67.9% frequency of MR and 46.4% frequency of AR in these patients. Rickards et al also

found 67% MR in patients with HOCM.<sup>37</sup> Shiota et al found 23% AR in their study, which is different from our study which shows frequency of AR up to 46%.<sup>29</sup> The difference may be due to the fact that we only studied patients of HCM with obstruction and Shiota et al studied all patients with HCM. It means that LVOT obstruction is associated with increase frequency of AR. The most interesting findings in our study is the relation between severity of MR with LVOT gradient. Higher the LVOT gradient more will be the MR. So we found a direct relation between the severity of MR and the LVOT gradient. This association between LVOT gradient and severity of MR has also described by Yu et al.<sup>26</sup>

The limitation of our study is that we have very small sample size of 28 cases only. With increasing data more interesting information can be obtained. The second limitation is that our study was a retrospective study obtained from computer record, so we couldn't get the desired information and detail echocardiography of these cases.

## CONCLUSION

HOCM is equally prevalent in both sexes and can be found at any age group. The disease is significantly associated with both MR and AR, and there is tendency towards increase left atrial diameter. Moreover there is direct relation between severity of MR and LVOT gradient.

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