

Original Article

Dimensional Assessment of the Heart in Hypertrophic Cardiomyopathy Patients in Tertiary Care Hospitals

Rayhan Shahrear^{1*}, Mohiuddin Masum²¹Anatomy, Assistant Professor, Ibrahim Medical College, 1/1 Ibrahim Sharani, Shegunbagicha, Dhaka²Anatomy, Assistant Professor, Anwer Khan Modern Medical College, Dhanmondi R/A. Dhaka**Corresponding Author:** Dr. Rayhan Shahrear, E-mail: dr.rayhan.shahrear@gmail.com

ARTICLE INFO

Article history

Received: December 05, 2020

Accepted: March 30, 2021

Published: April 30, 2021

Volume: 9 Issue: 2

Conflicts of interest: None

Funding: None.

Key words:

Hypertrophic Cardiomyopathy,
Cardiac Morphology,
Concentric HCM

ABSTRACT

Hypertrophic cardiomyopathy (HCM) is one of the most prevalent disorders responsible for sudden cardiac death. Presentation of the symptoms varies due to the degree of thickening, and functional ability of the cardiomyocytes. The aims of the current study were to assess the clinical features, and cardiac morphology. This was a descriptive study with some analytical components. Thirty-four adult HCM patients were included within a duration of four months by patient selection checklist. After informed written consent, relevant information was noted and analyzed. Frequency distribution of phenotypes were, 56% asymmetric septal hypertrophy, 29% concentric hypertrophy, and 15% apical hypertrophy. Breathlessness and chest discomfort were present in 56% and 62% patients respectively, and higher in asymmetric septal HCM. Palpitation was very frequent in concentric HCM (90%). ECG revealed left ventricular hypertrophy in 85% of patients, and 79% of them had ST change. The interventricular septal thickness was narrower in apical type (14.80mm). The posterior wall thickness was higher in concentric HCM (19.20mm). The left atrial size was smaller in Concentric type(34.60mm).

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is one of the most common genetically predisposed cardiomyopathies [1, 2]. HCM may remain without symptom, or after a long time become symptomatic with commonly presented features like breathlessness, chest discomfort, and exertion [3, 4]. Sudden cardiac death (SCD) in young adults [3, 5, 6] may occur without other causes of hypertrophy of cardiac muscle [7, 8]. The prevalence of HCM is about 0.2% of the world population [1, 2, 6, 9, 10]. The phenotypic classification is widely used by clinicians which is based on the distribution of the hypertrophied cardiac muscles is like the following: (a) asymmetrical septal HCM is the commonest to be seen and it is diagnosed when the interventricular septum is predominantly involved, (b) the next common pattern is concentric hypertrophy, where the cardiac wall is diffusely hypertrophied, and (c) the apical type occurs when the apical cardiac muscle is involved [11-16]. Echocardiography, along with clinical assessment is the most convenient way for diagnosing the disease. The clinical profile of HCM received more focus from the researchers, but data relating with morphological change is scarce.

MATERIALS AND METHOD

The study was carried out in the Department of Anatomy, Bangabandhu Sheikh Mujib Medical University (BSMMU). Thirty-four Patients of HCM (Echocardiographic LV wall thickness ≥ 15 mm in any segment of the myocardium, eighteen years or above, without myocardial infarction, poorly-treated uncontrolled hypertension, infiltrative cardiac diseases, valvular and other diseases which can mimic features of HCM) diagnosed by cardiologists by echocardiography were collected from various hospitals, clinics, and from private practices by convenient sampling technique from October 2017 to January 2018. Patients having other predisposing causes were excluded. Informed written consent was taken from all patients. Clinical symptoms like breathlessness, palpitation and chest discomfort were noted down through extensive history taking from the patients. Height and weight of the patients were measured and BMI was calculated by the formula, $BMI = \frac{\text{Weight in KG}}{(\text{Height in m}^2)}$ [17]. The frequencies of the clinical cardiac phenotypes were measured from echocardiographic data. The clinical and echocardiographic data were analyzed, and the difference among the

types was measured, and One-way ANOVA was performed to determine the significance using SPSS version 23.0.

RESULTS

Frequency distribution of cardiac phenotypes is shown in Figure I. Among the thirty-four HCM patients, 56% of patients had asymmetric septal hypertrophy (ASH), 29% had concentric hypertrophy, and 15% had the apical type of HCM. The concentric HCM patients were younger and leaner than the other two groups (Table I) and showed higher frequency in the aspect of chest discomfort and changes in ECG (Table II). The mean interventricular septal thickness was 19.59 mm. However, in the apical type it was also considerably low (14.80 mm). The mean posterior wall thickness was 13.68 mm, and it was notably significantly higher in concentric HCM (19.20 mm) (Table III).

DISCUSSION

HCM is characterized by the thickening of the cardiac wall with or without any functional changes. The muscular thickening is occurring in HCM has been classified on several bases [18]. The phenotypic pattern is the commonest of all the classifications used by the clinicians.

Singhal, Dastidar [16] found asymmetric septal hypertrophy was observed in the highest prevalence (65%) by using cardiac magnetic resonance (CMR) scan. The apical and concentric types followed. Chun, Choi [13] stated

that asymmetrical hypertrophy was prevalent in 60-70%. Concentric HCM was 42%, and apical HCM had a 25% prevalence.

The present study showed, around 90% of the patients were male. The average age was 50.26 years. In Bangladesh a study by Guha, Islam [19] showed, 90% of patients were male, had an average age of 52 years. Another study by Ahsan, Kabir [20] showed, 95% of patients were male; the average age was 46 years. Spirito and Maron [21] indicated the frequency of the male population was 66% and had an average age of 41 years. Masri, Pierson [22] found that the rate of the male patients was

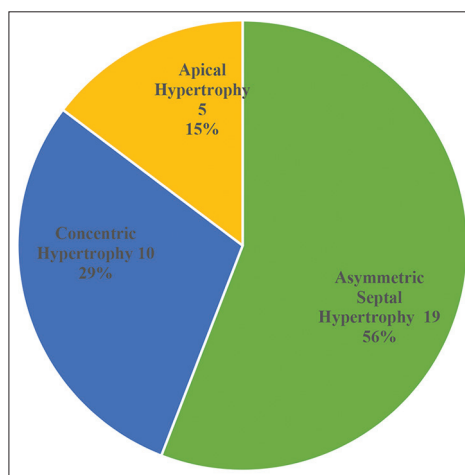


Figure I. Percentage frequencies of cardiac phenotypes of HCM

Table I. Demographic characteristics

Demographic characteristics	ASH (N=19)	Concentric (N=10)	Apical (N=5)	Total (N=34)	p-value
Age (year)	50.84±14.66	44.40±12.64	59.80±11.74	50.26±14.21	0.136
Male %	89% (17)	90% (9)	100% (5)	91% (31)	0.752
Female %	11% (2)	10% (1)	0% (0)	9% (3)	
BMI (kg/m ²)	26.60±3.09	24.53±3.98	27.50±5.37	26.12±3.77	0.259

*p-value is significant <0.05

Table II. Clinical characteristics

Clinical characteristics	ASH % (N=19)	Concentric % (N=10)	Apical % (N=5)	Total % (N=34)	p-value
Chest Discomfort	68% (13)	60% (6)	40% (2)	62% (21)	0.528
Breathlessness	68% (13)	40% (4)	40% (2)	56% (19)	0.271
Palpitation	79% (15)	90% (9)	80% (4)	82% (28)	0.768
LVH in ECG	79% (15)	100% (10)	80% (4)	85% (29)	0.314

*p-value is significant <0.05

Table III. Echocardiographic characteristics of different cardiac phenotypes of HCM

Echocardiographic characteristics	ASH (N=19)	Concentric (N=10)	Apical (N=5)	Total (N=34)	p-value
	Mean±SD	Mean±SD	Mean±SD	Mean±SD	
Interventricular Septal Thickness (mm)	20.63±3.547	20.20±4.917	14.80±4.438**	19.65±4.48	0.026*
Posterior wall thickness (mm)	11.53±2.170	19.20±3.615***	10.80±1.789	13.68±4.44	<0.001*
Left Atrial Size (mm)	41.26±7.759	34.60±5.502	42.00±2.550	38.50±6.36	0.036****
End Diastolic Diameter (mm)	42.00±7.303	41.10±7.141	43.00±3.391	41.88±6.69	0.875
End Systolic Diameter (mm)	26.37±5.134	23.40±6.450	25.40±3.286	25.35±5.37	0.378

*p-value is significant <0.05

64% and the average was 50 years. From these studies, the frequency of male patients seemed to be higher in the study population. Vlassoff [23] reviewed several studies in Bangladesh and concluded that gender determines the consequence of poor health.

Patients in the current study had an average BMI of 26 kg/m². BMI was 26 kg per sq-meter in the French population [24]. In the American community, the average BMI was 29 kg per sq-meter [25]. This variation reflects the stature difference of patients from country to country. HCM is found in patients with higher BMI in compare to the general population. Like all other cardiovascular diseases [26], increased BMI could be a causative factor for triggering symptoms in HCM patients.

HCM is a life-threatening disease, but it can remain asymptomatic for the whole life. Some develop symptoms when the thickening becomes marked. Cardinal symptoms of HCM include breathlessness, angina or chest discomfort, and palpitation. In severe cases, patients may suffer from a syncopal attack, cardiac arrest, and even death. However, palpitation was the most commonly occurring symptom among patients (82%). Breathlessness and chest discomfort was prevalent in 56% and 62% of the patients respectively. Sultana, Haque [27] found chest pain was the most prominent symptom (33%), followed by breathlessness (23%) and palpitation (20%). Kubo, Gimeno [28] also observed palpitation was more prominent (37%). Breathlessness was the most frequent symptom among the patients [24, 29]. After observing these frequencies of symptoms, no symptoms cannot be identified as a single diagnostic feature of HCM. Instead, they altogether provide a reliable indication of cardiovascular disease which is required a further investigation to be confirmed.

Even after finding substantial evidence from the symptoms, HCM patients have to go under specific diagnostic modalities to confirm the diagnosis. ECG is the first-line investigation for diagnosing HCM [5, 8]. The key feature that is found in the ECG of HCM patients is a tall QRS complex with widespread Q-wave [30]. ST-segment depression and T-wave inversion in the left-sided leads reveals ischemic strain [31]. However, these features ECG might not be available and have to proceed to further evaluation, especially in asymptomatic patients. Here in the present study, 85% of the HCM patients showed LVH pattern in the ECG, and 79% of them had developed the strain pattern.

In the present study, measurements were taken from the echocardiography reports. The interventricular septum is usually found to be the most thickened part of the ventricular wall. In this study, the mean value of the interventricular septal thickness was found at 19.65 mm. Espinola-Zavaleta, Vega [32] found the interventricular septal thickness in concentric HCM was lower than asymmetric septal HCM. However, we found that in our study population interventricular septal thickness these two groups were almost similar. The overall value observed in this study was similar to that of the other reviews. A comparison with some studies is shown in Figure II. Another critical measurement of the ventricular is posterior wall

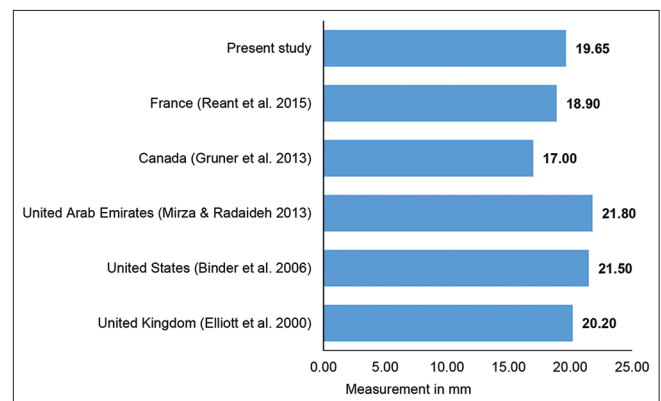


Figure II. Mean values of interventricular septal (IVS) thickness found in various studies in comparison to the present study

thickness. The clinical diagnosis made by the physician with echocardiography is mostly based on the ratio of the interventricular septal thickness to the posterior wall thickness. The cut-off value for differentiating the type is 1.3 [15, 16]. In asymmetric septal HCM, the thickness of the interventricular septum increases in a higher amount than that of the posterior wall. Therefore, the ratio is larger than in concentric HCM, where the cardiac wall is thickened diffusely. In the apical type, the apex is thickened, and the interventricular wall and the posterior wall may or may not be thickened. A study showed that the posterior wall thickness in their study population was 13 mm and 11 mm respectively [29, 33]. In the present study, it was 13.68 mm, which relatively corresponds to the other reviews. The values found in this study were significant among the cardiac phenotypes of HCM.

End-diastolic and end-diastolic diameter in the current study was 41.88mm and 25.35mm. Reduction of the difference between these two values indicates the reduced left ventricular cavity size. That will mean, there will be less space for blood and subsequently diminished cardiac output. These values in this study were found within the normal range [34] and corresponded with other studies [22, 28, 29, 33, 35, 36]. Also, the left atrial size in the concentric HCM was found smaller than the other phenotypes.

CONCLUSION

This study has enlightened the frequency, demography, clinical feature, and cardiac morphology of HCM patients with different cardiac phenotypes. Asymmetric septal type HCM was found to have the highest prevalence in the study. The asymmetric septal type of HCM had a higher severity of symptoms and frequency. The concentric type showed more change in cardiac morphology.

ACKNOWLEDGMENTS

The authors would like to thank and express their gratitude to our supervisor Prof. Laila Anjuman Banu, Professor of Anatomy Department, BSMMU. We also like to show our gratitude to all the clinicians for their generous help,

especially Professor Dr. M. Nazrul Islam, Dr. SM Ahsan Habib, Associate Professor, Dr. Mohammad Faisal Ibne Kabir, Assistant Professor, Department of Cardiology, BSMMU and Dr. Abul Mokarram Badruddin Sardar, Associate Professor, Department of Cardiology, BIRDEM.

REFERENCES

- Compton G, Nield L, Dragulescu A, Benson L, Grosse-Wortmann L. Echocardiography as a Screening Test for Myocardial Scarring in Children with Hypertrophic Cardiomyopathy. *Int J Pediatr*. 2016;2016:1980636.
- Voilliot D, Huttin O, Hammache N, Filippetti L, Vaugrenard T, Aliot E, et al. Impact of Global and Segmental Hypertrophy on Two-Dimensional Strain Derived from Three-Dimensional Echocardiography in Hypertrophic Cardiomyopathy: Comparison with Healthy Subjects. *J Am Soc Echocardiogr*. 2015;28(9):1093-102.
- Bashyam MD, Purushotham G, Chaudhary AK, Rao KM, Acharya V, Mohammad TA, et al. A low prevalence of MYH7/MYBPC3 mutations among familial hypertrophic cardiomyopathy patients in India. *Mol Cell Biochem*. 2012;360(1-2):373-82.
- Lind JM, Chiu C, Semsarian C. Genetic basis of hypertrophic cardiomyopathy. *Expert Rev Cardiovasc Ther*. 2006;4(6):927-34.
- Houston BA, Stevens GR. Hypertrophic cardiomyopathy: a review. *Clin Med Insights Cardiol*. 2014;8(Suppl 1):53-65.
- Maron BJ, Gardin JM, Flack JM, Gidding SS, Kurosaki TT, Bild DE. Prevalence of hypertrophic cardiomyopathy in a general population of young adults. Echocardiographic analysis of 4111 subjects in the CARDIA Study. Coronary Artery Risk Development in (Young) Adults. *Circ*. 1995;92(4):785-9.
- Binder J, Ommen SR, Gersh BJ, Van Driest SL, Tajik AJ, Nishimura RA, et al. Echocardiography-guided genetic testing in hypertrophic cardiomyopathy: septal morphological features predict the presence of myofibrillar mutations. *Mayo Clin Proc*. 2006;81(4):459-67.
- Maron BJ, McKenna WJ, Danielson GK, Kappenberger LJ, Kuhn HJ, Seidman CE, et al. American College of Cardiology/European Society of Cardiology clinical expert consensus document on hypertrophic cardiomyopathy. A report of the American College of Cardiology Foundation Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines. *J Am Coll Cardiol*. 2003;42(9):1687-713.
- Garcia-Castro M, Coto E, Reguero JR, Berrazueta JR, Alvarez V, Alonso B, et al. [Mutations in sarcomeric genes MYH7, MYBPC3, TNNT2, TNNI3, and TPM1 in patients with hypertrophic cardiomyopathy]. *Rev Esp Cardiol*. 2009;62(1):48-56.
- Jacoby DL, DePasquale EC, McKenna WJ. Hypertrophic cardiomyopathy: diagnosis, risk stratification and treatment. *CMAJ*. 2013;185(2):127-34.
- Arad M, Seidman JG, Seidman CE. Phenotypic diversity in hypertrophic cardiomyopathy. *Hum Mol Genet*. 2002;11(20):2499-506.
- Elliott P, McKenna WJ. Hypertrophic cardiomyopathy. *Lancet*. 2004;363(9424):1881-91.
- Chun EJ, Choi SI, Jin KN, Kwag HJ, Kim YJ, Choi BW, et al. Hypertrophic cardiomyopathy: assessment with MR imaging and multidetector CT. *Radiographics*. 2010;30(5):1309-28.
- Mozaffarian D, Caldwell JH. Right ventricular involvement in hypertrophic cardiomyopathy: a case report and literature review. *Clin Cardiol*. 2001;24(1):2-8.
- De Garate E, Dastidar AG, Singhal P, Venuti G, Amadu A, Baritussio A, et al. 6 Prevalence and CMR characteristics of apical HCM: Abstract 6 Table 1. *Heart*. 2016;102(Suppl 3):A4.2-A5.
- Singhal P, Dastidar AG, Giuseppe V, Amadu A, Baritussio A, Scatteia A, et al. 144 Apical Versus Non-apical Hypertrophic Cardiomyopathy (HCM): Insight from Cardiac Magnetic Resonance Imaging: Abstract 144 Table 1. *Heart*. 2016;102(Suppl 6):A104.1-A.
- Stensland SH, Margolis S. Simplifying the calculation of body mass index for quick reference. *J Am Diet Assoc*. 1990;90(6):856.
- Parato VM, Antoncicchi V, Sozzi F, Marazia S, Zito A, Maiello M, et al. Echocardiographic diagnosis of the different phenotypes of hypertrophic cardiomyopathy. *Cardiovasc Ultrasound*. 2016;14(1):30.
- Guha B, Islam AKMM, Choudhury AK, Chowdhury AHK, Mandal AK. Echocardiographic Evaluation of Left Ventricular Diastolic Function after Percutaneous Coronary Intervention in Patients with Coronary Artery Disease. *Cardiovascular Journal*. 2012;4(2):127-31.
- Ahsan SA, Kabir MFI, Khaled FI, Joarder AI, Mohammad N, Rahman MA, et al. A study of changes in various echocardiographic parameters in patients with chronic stable angina undergoing percutaneous coronary intervention (PCI). *University Heart Journal*. 2015;9(2):99-106.
- Spirito P, Maron BJ. Patterns of systolic anterior motion of the mitral valve in hypertrophic cardiomyopathy: assessment by two-dimensional echocardiography. *Am J Cardiol*. 1984;54(8):1039-46.
- Masri A, Pierson LM, Smedira NG, Agarwal S, Lytle BW, Naji P, et al. Predictors of long-term outcomes in patients with hypertrophic cardiomyopathy undergoing cardiopulmonary stress testing and echocardiography. *Am Heart J*. 2015;169(5):684-92 e1.
- Vlassoff C. Gender differences in determinants and consequences of health and illness. *J Health Popul Nutr*. 2007;25(1):47-61.
- Reant P, Donal E, Schnell F, Reynaud A, Daudin M, Pillois X, et al. Clinical and imaging description of the Maron subtypes of hypertrophic cardiomyopathy. *Int J Cardiovasc Imaging*. 2015;31(1):47-55.
- Larsen CM, Ball CA, Hebl VB, Ong KC, Siontis KC, Olson TP, et al. Effect of Body Mass Index on Exercise Capacity in Patients With Hypertrophic Cardiomyopathy. *Am J Cardiol*. 2018;121(1):100-6.

26. van der Heijden DJ, van Leeuwen MAH, Janssens GN, Lenzen MJ, van de Ven PM, Eringa EC, et al. Body Mass Index Is Associated With Microvascular Endothelial Dysfunction in Patients With Treated Metabolic Risk Factors and Suspected Coronary Artery Disease. *J Am Heart Assoc.* 2017;6(9).
27. Sultana SA, Haque A, Khan Z, Iqbal SA, Malek MA. Clinical Profile of Hypertrophic Cardiomyopathy in a Tertiary Level Hospital. *Cardiovascular Journal.* 2014;7(1):31-7.
28. Kubo T, Gimeno JR, Bahl A, Steffensen U, Steffensen M, Osman E, et al. Prevalence, clinical significance, and genetic basis of hypertrophic cardiomyopathy with restrictive phenotype. *J Am Coll Cardiol.* 2007;49(25):2419-26.
29. Gruner C, Ivanov J, Care M, Williams L, Moravsky G, Yang H, et al. Toronto hypertrophic cardiomyopathy genotype score for prediction of a positive genotype in hypertrophic cardiomyopathy. *Circ Cardiovasc Genet.* 2013;6(1):19-26.
30. McLeod CJ, Ackerman MJ, Nishimura RA, Tajik AJ, Gersh BJ, Ommen SR. Outcome of patients with hypertrophic cardiomyopathy and a normal electrocardiogram. *J Am Coll Cardiol.* 2009;54(3):229-33.
31. Pollehn T, Brady WJ, Perron AD, Morris F. The electrocardiographic differential diagnosis of ST segment depression. *Emerg Med J.* 2002;19(2):129-35.
32. Espinola-Zavaleta N, Vega A, Basto DM, Alcantar-Fernandez AC, Guarner Lans V, Soto ME. Survival and clinical behavior of hypertrophic cardiomyopathy in a latin american cohort in contrast to cohorts from the developed world. *J Cardiovasc Ultrasound.* 2015;23(1):20-6.
33. Mirza SJ, Radaideh GA. Pattern of left ventricular hypertrophy seen on transthoracic echo in patients with hypertensive cardiomyopathy when compared with idiopathic hypertrophic cardiomyopathy. *J Pak Med Assoc.* 2013;63(1):16-9.
34. ECHOpedia. Normal Values of TTE 2017 [18th July 2017]. Available from: http://www.echopedia.org/wiki/Normal_Values_of_TTE.
35. Lopes LR, Syrris P, Guttmann OP, O'Mahony C, Tang HC, Dalageorgou C, et al. Novel genotype-phenotype associations demonstrated by high-throughput sequencing in patients with hypertrophic cardiomyopathy. *Heart.* 2015;101(4):294-301.
36. Elliott PM, Poloniecki J, Dickie S, Sharma S, Monserrat L, Varnava A, et al. Sudden death in hypertrophic cardiomyopathy: identification of high risk patients. *J Am Coll Cardiol.* 2000;36(7):2212-8.