Idiopathic hypertrophic cardiomyopathy in diyala: echocardiographic analysis of 12000 adolescents and adults

DR. Adel Hessen AL-Husseiny– Lecturer-Department of medicine- College of Medicine-University Of Diyala

الخلاصة

في هذه الدراسة قد أجرينا مسح بفحص صدى القلب لأثنى عشر ألف شخص لتحديد الخصائص الوبائية والسر يريع لهذا المرض من 1999-2005.ستة آلاف شخص كانوا أصحاء و ستة آلاف مريض تقدموا بأعراض قلبيه . المدى العمري كان 20-75 عاما. المرضى الذين لديهم أمراض قلبيه أو يتناولون أدويه قلبيه قد استبعدوا من الدراسة. تم استبيان التاريخ المرضي الشامل و الفحص ألسريري و تخطيط القلب الكهربائي وفحص أشعة الصدر و فحوص صدى القلب(الحركي ،ثنائي الأبعاد و الدوبلر الطيفي والملون) ،إضافة إلى الفحوص ألمختبريه.أظهرت الدراسة انه من بين 12000 شخص تم فحصهم كان هنالك 18 حالة(0,15%) تضخم العضله ألقلبيه ألاعتلالي ،عشرة ذكور(0,083)وثمانية إناث(0,06%). ستة حالات(33,3%)، ثلالة ذكور و ثلالة إناث، كانوا أصحاء و اثنا عشر حاله (سبعة ذكور وخمسة إناث) كان لديهم أعراض قلبيه .أربعة عشر مريضا كانت أعمارهم اقل من 30،حاله واحده اقل من 20 و ثلاثة حالات كانوا فوق 40 سنه. 77.7% من المرضى كانوا في الصف الاول حسب تصنيف جمعية القلب في نيويورك. خمسة مرضى كانت لديهم نتائج موجبه في المسح العائلي للمرض. أظهرت فحوص صدى القلب أن تضخم الحاجز ألبطيني غير المتناظر هو النوع الأكثر شيوعا للمرض ،حيث وجد في خمسة عشر مريضا (83.3%).فحوص الدوبلراظهرت وجود انسداد (اعاقه) لمجرى البطين الايسرالخارج في سبعة مرضى (38.8%):تضخم العضله القلبية ألاعتلالي ألانسدادي.وجد تخطيط القلب الكهربائي غير طبيعي في ثلاثة عشر مريض(72.2%) والارتجاف الأذيني في حاله واحده (5,5%)

ABSTRACT

This study was conducted in diyala governorate, with echocardiographic analysis of 12000 subjects to determine the clinical and epidemiological aspects of HCM from 1999-2005. Six thousands person were healthy and Six thousands patients who presented with cardiac complaints. Age range of the sample was 15-70 year, patients with known cardiac diseases or taking cardiac drugs were excluded. Thorough clinical history, physical examination, electrocardiography (ECG), chest radiographs, echocardiography (M-mode, two dimensional B-mode and continues wave (CW) Doppler mode) and standardized laboratory tests were performed .Our study showed that among 12000 persons screened 18 cases of proved HCM were diagnosed (0.15%),10 males(0.0833%) and 8 females(0.0666%),six cases 33.3% (3 males(50%) and 3 females (50%) were asymptomatic at diagnosis. Among six thousands patients presented with cardiovascular complaints (chest pain, dyspnea, palpitation, features of heart failure, pre-syncope and syncope) twelve (12) cases (0.01%) were diagnosed, seven males (58.3%) and five females (41.6%). Most of cases were in young age group: Fourteen patients (77.7%) were (20-39) years of age, and only 1<20 and 3 >40, male/female ratio was 1.25:1. Fourteen patients 77.7% were in NYHA -1 functional class and 5 patients (27.7%) had positive results on family screening for HCM. Echocardiographic study showed that fifteen patients (83.3) had asymmetrical septal hypertrophy (ASH) and 3 patients (16.6%) had concentric LVH. Doppler echocardiography showed substantial obstruction to left ventricular outflow tract (LVOT) in 7 (38.8%) patients (the range of the mean peak systolic pressure gradient was from 30 to 71 mm Hg: Hypertrophic obstructive cardiomyopathy. Thirteen subjects (72.2%) had abnormal ECG,atrial fibrillation reported in one patient (5.5%) .The estimated total prevalence of HCM in diyala governorate is 0.15% (150/100000).

INTRODUCTION

There is limited data about the clinical and epidemiological characters of hypertrophic cardiomyopathy (HCM) in diyala governorate where more one million of the Iraqi's population live,

Hypertrophic cardiomyopathy (HCM) is a fascinating disease of marked heterogeneity. HCM was originally characterized by massive myocardial hypertrophy of non dilated left ventricle (LV) in the absence of known cause, a dynamic left ventricular outflow obstruction, and increased risk of sudden death (1). HCM has range of clinical and morphological features and can occur from infancy to old age. It is now well accepted that multiple mutations in genes encoding for the cardiac sarcomere are responsible for the disease . HCM clinical diagnosis is by 2-dimensional echocardiographic identification of otherwise unexplained left ventricular wall thickening in the presence of a nondilated cavity (2). Age of onset of LVH ranges from early childhood to late adulthood and depends, in part, on the underlying genetic cause (3, 4). Although HCM gene mutation is present at birth, it may be decades before LVH becomes clinically detectable (5). Because left ventricular hypertrophy usually develops between the ages of 5 and 15 years, a normal echocardiogram in a child does not exclude the presence of HCM (6). HCM is increasingly recognized with wide spread application of echocardiography. Though it is relatively rare disease, it is important to diagnose for a number of reasons:

1. Hypertrophic cardiomyopathy (HCM) caused by sarcomere mutations is the most common genetic cardiovascular disorder (7, 8, 9), so screening of relatives is important.

2. It is the most important cause of sudden cardiac death in young age group especially when the disease is asymptomatic (very common form); annual incidence is about 6% in children and young adults and 1% in adults 45 to 60 years of age (10,11). Hypertrophic cardiomyopathy accounts for 36% of deaths in athletes younger than 35, making it the most common cause of sudden death in this age-group (12). Causes of syncope and sudden death include ventricular arrhythmias, atrial fibrillation (AF), bradyarrhythmias, ischemia, and left ventricular outflow obstruction. Data suggest that ventricular tachyarrhythmias are the cause of sudden death in most patients (6, 13).

3. Accurate diagnosis of HCM is important for appropriate management of major HCM comorbidities, including, AF, stroke, heart failure and sudden cardiac death (14, 15)

4. Clinical symptoms and signs are similar to more common cardiac diseases (16, 17)), and even misdiagnosed as psychological disorder

5.Wrong diagnosis leads not only to non useful but potentially harmful treatments, as the nitrates used for coronary artery disease, diuretics and ACE-inhibitors used for HF are poorly tolerated by HCM patients

The main stay of diagnosis of HCM is echocardiography (Echo.) which is both easily accessible, non invasive, very sensitive and specific in diagnosing the disease, its subtypes, for assessment of homodynamic effects (obstructive ,non-obstructive) and for screening of relatives.

The most common subtypes of hypertrophic cardiomyopathy is diffuse hypertrophy of the ventricular septum (70% to 75% of cases) followed by basal septal hypertrophy (10% to 15%), concentric hypertrophy (<5%), hypertrophy of the lateral wall (1% to 2%), and apical hypertrophy <1% (18).

Aim of this study

Using non invasive tests with echocardiography being the gold standard test in screening patients and asymptomatic persons for the presence of HCM, keeping the suspicion of the HCM will increase the

chance of diagnosis to avoid missing the potentially lethal disease and to avoid mismanagement results from misdiagnosis.

PATIENTS AND METHODS

To study the prevalence and clinical manifestations of HCM in diyala governorate peoples,a cross sectional prospective study was conducted in baquba general hospital and private clinic with a total of 12000 persons evaluated from May 1999-May 2005 with random sampling design. The subjects were grouped according to age (15–19,20-29, 30–39, 40–49, 50–59, 60–70 years). Enrollment of subjects from the same family was avoided. Half of sample numbers were healthy asymptomatic persons and half of them were symptomatic and seek medical advice because of cardiac complaints.

All participants were asked to complete questionnaires to determine if they had been evaluated previously for cardiovascular disease; none reported a history of hypertrophic cardiomyopathy or similar problems. A detailed pedigree analysis and clinical history were obtained from all patients. Patients with hypertension (by history, measurement, taking antihypertensive medications), aortic valve disease, highly trained athletes, heavy manual workers, those with known or diagnosed diabetes mellitus, renal impairment and patients taking cardiac drugs, all were excluded from the study . A history of syncope was defined as one or more episodes of unexplained loss of consciousness within the 12 months. Chest pain was classified as exertional or atypical if it lasted >30 min at rest in the absence of myocardial infarction. Dyspnea was coded according to the New York Heart Association (NYHA) classification (19). A family history of sudden death was defined as sudden cardiac death in two or more first-degree relatives <40 years old. Physical examination included the measurement of height, body weight, blood pressure, and chest and abdominal examination. Standard 12-lead electrocardiograms were recorded for each subject, and the diagnosis confirmed and specified with echocardiography. In subjects suspected of having hypertrophic cardiomyopathy, we performed additional laboratory studies, including hematocrit, routine serum biochemistry tests, urinalysis, and chest radiography, to exclude other causes of left ventricular hypertrophy.

Echocardiography:Two-dimensional and M-mode echocardiography were performed using conventional methods (20, 21, 22) with an Siemens versa plus and Toshiba systems , With Doppler facilities (spectral and color mode).The transducer frequency was 2.5-3.5 MHz. Hypertrophic cardiomyopathy was diagnosed on the basis of two-dimensional echocardiographic demonstration of a hypertrophied (wall thickness ≥15 mm) and non-dilated LV in the absence of another cardiac or systemic disease capable of producing the magnitude of hypertrophy evident (2,23) . In summary, end-diastolic left ventricular wall thickness (LVWT) was recorded at the mitral valve and papillary muscle level in the anterior and posterior septum, as well as in the lateral and posterior left ventricular wall using short-axis two-dimensional images. Anterior and posterior septal thickness at the apex was assessed from the apical four-chamber and the parasternal short-axis views. The severity of hypertrophy was graded as mild-moderate if septal or PW thickness is between 15-20 mm, and as severe if more. Markers for LVOT-obstruction are:-SAM-AML &-Mid-systolic closure of AV-cusps Cardiac dimensions were measured from the M-mode echocardiograms directly from the screen, according to the recommendations of the American Society of Echocardiography (24, 25, 26). In the parasternal short-axis view, the left ventricle was divided into four regions that identified the anterior and posterior ventricular septa, and the lateral and posterior free walls. Wall thickness was assessed directly from the television monitor with the aid of calipers and the calibration scale produced by the instrument. Continuous-wave Doppler was used to measure LV outflow gradient (27). Left ventricular outflow obstruction was defined as a peak instantaneous pressure gradient between LV-cavity and ascending aorta during systole under basal (resting) conditions ≥30 mm Hg], there is now widespread recognition that the subaortic gradient (30 mm Hg or more) and associated elevations in intra-cavity LV pressure reflect true mechanical impedance to outflow and are of pathophysiologic and prognostic importance to patients with HCM (28,29).Statistical data are expressed by mean value ±SD.

Results:

Baseline clinical Characteristics of the study sample and HCM patients are shown in table 1

Table 1. The distribution of clinical characteristics of HCM patients at presentation

Clinical symptoms and signs	No.	%
No. of screened population Male/female ratio	12000 1/1	50
Number of patients/Total No. of screened sample	18/12000	0.015
Mean age at initial evaluation (yrs)	32±6.8	
Range of age/yr	15-70	
Male gender (%)	10	55.5
Asymptomatic patients at diagnosis	6	33.3
Mean duration of symptoms (yrs) in symptomatic group	5.2±2.1	
Presyncope and syncope	5	27.7
NYHA= New York Heart Association. Class I	14	77.7
Class II	03	16.6
Class III/IV	1	5.5
Mean systolic/diastolic BP (blood pressure)	120/80	
Chest pain	10	55.5
Exertional	6	33.3
Atypical	4	22.2
Family history of sudden death	0	0
Bifid carotid pulse	3	16.6
Lifting apex beat	4	22.2
Systolic murmur	5	27.7
Fourth sound	9	50

Basal crepitation	2	11.1
Heart failure	1	5.5

Among 12000 selected participants with ages ranged from15 to 70 years (mean 32±6.8) and sex ratio 1:1eighteen subjects were identified as having definite HCM , including 10 men and 8 women.Most of cases were in young age group: 14 (77.7%) patients were (20-39) years of age, and only1<20 and 3 >40, , 20 and three more 40 year. Male/female ratio was 1.25:1. (Table 1 , 2) The prevalence of HCM was estimated to be about 0.15% (150 per 100,000). Six of these 18 subjects were free of cardiac symptoms at the time of the Echo. study and had no previous suspicion of cardiovascular disease. Chest pain was the commonest symptom, notified in 10 cases (55.5%).NYHA functional class 1 (currently had normal exercise tolerance) was observed in 14(77.7%), 3(16.6%) were mildly symptomatic in class II, and 1(5.5%) were severely symptomatic in classes III and IV. syncope or pre-syncope were occurring in 5 (27.7%) of the patients. 5 patients (27.7%) had positive results on family screening for HCM (Table 3), no patient reported family history of sudden death. Systolic murmur and fourth heart sound noticed in (27.7%) and (50%) respectively

Table 2-The distribution of HCM patients according to the age and gender

Age groups of HCM patients	No. of cases	Males	females
15-19	1	0	1
20-29	9	5	4
30-39	5	3	2
40-49	2	2	0
50-59	0	0	0

60-70	1	0	1
Total	18	10	8

Table-3-The distribution of HCM among the families of affected patients

The disease among families	No.of patients	%
Positive results*	5	27.7
Negative results	13	72.3
Total	18	100

Notes: 1.All screened first degree relatives were older than 15 year

2. Three patient's families defaulted from screening

3.the family screening done utilizing 2D-Echocardiography

***One or more 1st degree relative with HCM**

The echocardiogaphic features of HCM patients are represented in table 4.

Table-4-The distribution of echcardiographic measurements among HCM patients

Echocardiographic parameter	No	%
ASH	15	83.3
Concentric LVH	3	16.6
VS Thickness, mm Ant Post	 21±4 20±5	
LV Free-Wall Thickness, mm AL Post	 11±1.3 10±2.4	
LVEDD (mm)	41±5.6	
LVESD (mm)	27±4.2	
LA (mm) LA>37 mm	37±2.5 8	 44.4
EF%	74±3	
SAM-AML Mid-systolic closure of AV	9 5	50 27.7

Pressure gradients>30mmHg	7	38.8

Data are presented as the mean value or number (%) of patients.

HCM indicates hypertrophic cardiomyopathy;ASH,,asymetrical septal hypertrophy, VS, ventricular septum; Ant, anterior; Post, posterior; LV, left ventricular; AL, anterolateral; LVED, LV end-diastolic dimension;LVESD,left ventricular end systolic dimension , LA, left atrium, EF.ejection fraction; SAM-AML, systolic anterior motion of anterior mitral valve leaflet; LVH, LV hypertrophy.AV,aortic valve .

Left ventricular wall thickness was between 15 and 29 mm in the 18 subjects with hypertrophiccardiomyopathy. . In the 11982 subjects without hypertrophic cardiomyopathy, the mean (± SD) interventricular septal thickness was 9.2 ± 2.6 mm (range, 6 to 14 mmul)

The location of the ventricular wall hypertrophy was confined to the interventricular septum in 15(83.3) subjects and 3 (16.6%) patients had concentric LVH. The left ventricle end-diastolic cavity dimension was between 44 and 52 mm. SAM-AML was observed in 50% of patients, while mid-systolic closure of AV in 27.7% , dilated LA in 44.4% and EF 74±3 was documented in 17 (94.4%) . Doppler echocardiography showed substantial obstruction to left ventricular outflow in 7 (38.8%) patients (the range of the mean peak systolic pressure gradient was from 30 to 71 mm Hg). The electrocardiographic (ECG) and chest -X-ray radiologic results are given in table 5.

Table-5-The distribution of patients according to electrocardiographic findings

ECG	No.	%
ST-T abnormalities	13	72.2
Definite LVH	11	61.1
P- mitralae	8	44.4
Q-wave	2	11.1
Arrhythmias		
BBB	1	5.5
Atrial fibrillation	1	5.5
Ventricular ectopics	3	16.6
Normal ECG	5	27.7
Total	13	72.2

Table -6-The distribution of patients according to radiologic findings

Chest X- ray	No.	%
Cardiomegaly	3	16.6
Pulmonary venous Congestion	2	11.1
Total	3	16.6

Thirteen subjects (72.2%) had abnormal electrocardiograms: 2(11.1%) with abnormal Q waves in more than two leads, 1(5.5%) with complete right bundle branch block, 13 (72.2%) with ST-segment and T-wave changes, 8 (44.4%) with left atrial enlargement; 1 (5.55) had AF. Radiologically 3 patients (16.6%) had cardiomegaly (cardiothoracic ratio>50%) and 2 (11.1%) had pulmonary venous congestion.

DISCUSSION

Although HCM is believed to be a relatively uncommon cardiac disease, the frequency with which it occurs in diyala governorate has not been defined. To address this issue, the patient population echocardiography was used to assess the clinico-epidemiologic manifestations of HCM. which was diagnosed in 18 patients(10 males and 8 females) from 12000 persons screened (0.15%), majority of them were <40 year old,third of them were asymptomatic at time of diagnosis, and 38.8% had obstructive features other studies also reported that the prevalence of unexplained left ventricular hypertrophy (LVH) in the general population is estimated to be 1 in 500. Zou et al ,(30) reported the results of cross-sectional epidemiologic survey that comprised more than 8000 residents used echocardiography with a random sampling recruitment design. In this community-based sample, the disease was identified in 0.16% of subjects. This prevalence is almost identical to that previously reported with echocardiography in the Coronary Artery Risk Development In Young Adults (CARDIA) study (20), which comprised a cohort of 4111 men and women, aged 23 to 35 years of age selected from the general population had echocardiographic evidence of HCM in 7 subjects (0.17%). Prevalence in men and women was 0.26:0.09%; 0.24:0.10%. Ventricular septal thickness was 15 to 21 mm (mean, 17 mm) in the 7 subjects. Only 1 of the 7 subjects had ever experienced important cardiac symptoms attributable to HCM,. ECGs were abnormal in 5 of the 7 subjects.: HCM was present in about 2 of 1000 young adults..These prevalence figures are also similar to that reported by Hada et al (31) who documented hypertrophic cardiomyopathy by echocardiography in 0.2% of Japanese workers
Marcel J. M. Kofflard,et al (32) found in large study included 221 patients that , 28% were younger than 30 years and 20 (9%) were older than 65 years. The mean age (±SD) at diagnosis of HCM was 37 ± 17 years(our result was 32±6.8). A positive family history of HCM was present in 110 patients (49%), of whom 52 patients (23%) also reported a sudden death in a first degree relative. Reported symptoms included angina in 58 patients (26%), dyspnea in 81 (36%), syncope in 43 (19%), and palpitations in 43 (19%). At presentation, 100 patients (44%) were asymptomatic or had trivial symptoms (NYHA functional class I), 101 patients (45%) had mild symptoms of exertional angina and/or dyspnea (class II), and 24 patients (11%) were moderately symptomatic (class III . On echocardiography, mean interventricular septum width was 21 ± 4 mm (16 to 40 mm), 30 patients (13%) had marked left ventricular hypertrophy (interventricular septal width ≥25 mm). Left ventricular outflow tract (LVOT) gradient, at rest or provocation was ≥30 mm Hg in 98 patients (44%), as determined by Doppler echocardiography or cardiac catheterization. At the initial visit, 7 patients (3%) presented with persistent atrial fibrillation
Maron BJ, et al (33)in study of Prevalence of hypertrophic cardiomyopathy in an outpatient population referred for echocardiographic study,found a prevalence of 0.5%. Ages were 50 to 69 years. 25% had evidence of obstruction to left ventricular outflow. Before echocardiographic study, each of the patients with HCM had signs or symptoms of cardiac disease, but the correct diagnosis had not been suspected.

In other large study by Barry J. Maron,et al (34) about the clinical profile of hypertrophic cardiomyopathy identified de novo in rural communities, found that among a total of 15,137 echocardiograms were performed HCM was identified in 44 patients during the survey (0.29%). At

diagnosis, ages were 16 to 87 years (mean 57); 14 patients were >60 years of age, and only two were <30 years. 24 patients (83%) had either no or only mild symptoms; 5 (17%) evidenced severe functional limitation; Basal left ventricular outflow obstruction (gradients 30 to 82 mm Hg) was evident in 11 patients (38%). electrocardiograms that were frequently normal (about 25%) and showed evidence of LVH in (10%). Fifteen patients (51%) were female. At the time of diagnosis, 12 patients (41%) were judged to have no functional limitation, although four of these had experienced transient symptoms such as presyncope or syncope; 12 others (41%) had mild exertional symptoms (functional class II), and five (18%) had severe symptoms and functional limitation (class III). In eight of the latter 17 functionally limited patients, the onset of symptoms had been delayed until ≥70 years of age (with the oldest 83 years). Based on continuous wave Doppler examination, 11 of the 29 study patients (38%) had subaortic obstruction under basal conditions (peak instantaneous outflow gradients of 20 to 82 mm Hg), Maximum left ventricular wall thicknesses were 14 to 30 mm (mean 21) . Analysis of the patterns of left ventricular hypertrophy showed wall thickening confined to one segment of the wall (i.e., anterior ventricular septum) in 14 patients (48%), involving two segments (anterior and posterior septum or anterior septum and anterolateral free wall) in 13 patients (45%) and diffusely involving three or four segments (substantial portions of anterior and posterior septum as well as anterolateral free wall) in only two patients (7%)Left atrial dimension was 27 to 54 mm (mean 40), and cavity enlargement (>40 mm) was evident in 17 of the 29 patients (60%). Left ventricular end-diastolic dimension was 31 to 56 mm (mean 42), and >55 mm in only one patient. A variety of abnormal patterns and abnormalities were evident on the 12-lead ECG, either alone or in combination: 1) ST segment and T wave alterations (n = 14); 2) abnormally deep Q waves in inferior and lateral leads, (n = 9); 3) conduction abnormalities including left or right bundle branch block and left or right anterior hemiblock (n = 5); 4) reduced R wave in the right precordial leads (n = 3), atrial fibrillation was present in 4 patients, Increased precordial voltages consistent with a pattern of left ventricular hypertrophy (R or S wave ≥25 mm; range to 38 mm) were present in only 3 patients (10%). Of note, 7 patients (24%) had ECGs judged to be within normal limits; each of these had mild-to-moderate hypertrophy with maximum left ventricular wall thickness <20 mm, most prominent in the basal portion of the wall. The proportion of patients with the obstructive form of HCM (i.e., 27% with left ventricular outflow gradients ≥30 mm Hg) was similar to that generally reported by other authors (13 ,35) The same author in another study (36)to determine the prevalence of HCM in middle-aged and older adult american Indians ,HCM was present in 8 previously undiagnosed patients (0.23%; 2 of 1,000) based on a left ventricular (LV) wall thickness ± 15 mm and a nondilated cavity that was not associated with another cardiac disease. Four subjects were men and 4 were women, with prevalences by gender of 0.3% (3 of 1,000) and 0.18% (1.8 of 1,000), respectively. Previous echocardiographic screening studies in the United States and Japan have estimated that the prevalence of hypertrophic cardiomyopathy is between 17 and 170 per 100,000 persons (37, 38). The large variability is probably due to methodological differences in subject recruitment and echocardiographic interpretation. The Levine RA. (39) reported the prevalence of HCM in the general population is estimated to be from 0.2% to as high as 4.9% (very high) when echocardiographic screening is used . Third of our patients were asymptomatic at diagnosis,other studies also reported that patients may remain stable over long periods of time with up to 25% of a HCM cohort achieving normal longevity (75 years of age or older) (40,41,42,43).

As mentioned earlier 38.8%had obstructive features ,other studies stated that In roughly 25% of cases, there is associated obstruction to left ventricular outflow (hypertrophic obstructive cardiomyopathy) producing dynamic pressure gradient at the left ventricular outflow tract as a consequence of midsystolic apposition of the anterior mitral leaflet against the hypertrophic interventricular septum (44,45,46,47).

AF is found in only 5.5% of our patients, the report of Ho et al (48) is notable for certain clinical features to be particularly common in their cohort, such as atrial fibrillation (35% of their patients vs.

about 20%-25%) in other studies (49, 50).The relative rarity of AF in our study might be due to that the majority of our cases were in younger age group.

27.7% of our study patients had positive familial evidence of HCM; Charles Berul (51) reported that morphologic evidence of disease is found by echocardiography in approximately 25% of first-degree relatives of patients with HCM. Barry J. Maron reported 28 %(34).Diffuse involvement of IVS (anterior and posterior segment) is the predominant subtype found in our study, same reported by other studies (11, 30, 52).

COCLUSIONS:

From this study we concluded the followings:-

1. The disease is not very rare with the estimated total prevalence of HCM is 0.15% (150/100000).

2. Male/female ratio was 1.25:1.

3. Most of cases were in young age group: 77.7% were (20-39) years of age, and only 1<20 and 3 >40.

4. Third of diagnosed cases were asymptomatic at diagnosis.

5. About fourth of patients had positive results on family screening for HCM.

6. More than three-fourth of patients were NYHA -1 functional class (currently had normal exercise tolerance).

7. Echocardiographic analysis showed that the asymmetrical septal hypertrophy (ASH) was by far the commonest subtype of HCM followed concentric LVH .

8. Doppler echocardiography showed substantial obstruction to LVOT in 38.8% patients (the range of the mean peak systolic pressure gradient was from 30 to 71 mm Hg: Hypertrophic obstructive cardiomyopathy).

9. The abnormal electrocardiograms were found in 72.2% with AF being relatively rare finding at initial diagnosis.

REFERENCES

1. **Ommen SR**, **Nishimura RA**Hypertrophic cardiomyopathy. **: Curr Probl Cardiol. 2004 May;29 (5):239-91**

2. Barry J. Maron, MD **Hypertrophic Cardiomyopathy A Systematic Review *JAMA.*** 2002;287:1308-1320.

3.Niimura H, Patton KK, McKenna WJ, Soults J, Maron BJ, Seidman JG, Seidman CE. Sarcomere protein gene mutations in hypertrophic cardiomyopathy of the elderly. *Circulation.* 2002; 105: 446–451.

4.Rosenzweig A, Watkins H, Hwang DS, Miri M, McKenna W, Traill TA, Seidman JG, Seidman CE. Preclinical diagnosis of familial hypertrophic cardiomyopathy by genetic analysis of blood lymphocytes. *N Engl J Med.* 1991; 325: 1753–1760.

***5.* Carolyn Y. Ho, MD; Christine E. Seidman, MD A Contemporary Approach to Hypertrophic Cardiomyopathy *Circulation.* 2006;113:e858-e862.)**

6.Fananapazir L, McAreavey D. Hypertrophic cardiomyopathy: evaluation and treatment of patients at high risk for sudden death. Pacing Clin Electrophysiol 1997;20(2 Pt 2):478-501

7.Van Driest SL, Ellsworth EG, Ommen SR, Tajik AJ, Gersh BJ, Ackerman MJ. Prevalence and spectrum of thin filament mutations in an outpatient referral population with hypertrophic cardiomyopathy. *Circulation.* 2003; 108: 445–451.

8.Richard P, Charron P, Carrier L, Ledeuil C, Cheav T, Pichereau C, Benaiche A, Isnard R, Dubourg O, Burban M, Gueffet JP, Millaire A, Desnos M, Schwartz K, Hainque B, Komajda M. Hypertrophic cardiomyopathy: distribution of disease genes, spectrum of mutations, and implications for a molecular diagnosis strategy. *Circulation.* 2003; 107: 2227–2232.

9.Arad M, Maron BJ, Gorham JM, Johnson WH Jr, Saul JP, Perez-Atayde AR, Spirito P, Wright GB, Kanter RJ, Seidman CE, Seidman JG. Glycogen storage diseases presenting as hypertrophic cardiomyopathy. *N Engl J Med.* 2005; 352: 362–372.

10.Marian AJ, Roberts R. Recent advances in the molecular genetics of hypertrophic cardiomyopathy. Circulation 1995;92(5):1336-47

11. Michael DeLuca, MD; Tahir Tak, MD, PhD Hypertrophic cardiomyopathy Tools for identifying risk and alleviating symptoms VOL 107 / NO 7 / JUNE 2000 / POSTGRADUATE MEDICINE

12.Maron BJ, Thompson PD, Puffer JC, et al. Cardiovascular preparticipation screening of competitive athletes: a statement for health professionals from the Sudden Death Committee (clinical cardiology) and Congenital Cardiac Defects Committee (cardiovascular disease in the young), American Heart Association. Circulation 1996;94(4):850-6

13.Spirito P, Seidman CD, McKenna WJ, et al. The management of hypertrophic cardiomyopathy. N Engl J Med 1997;336(11):775-85

14..Maron BJ, Mitten MJ, Quandt EF, Zipes DP. Competitive athletes with cardiovascular disease: the case of Nicholas Knapp. *N Engl J Med.* 1998; 339: 1632–1635.

15.Maron BJ. Sudden death in young athletes. *N Engl J Med.* 2003; 349: 1064–1075.

16.Braunwald E. Valvular heart disease. In: Braunwald E. Heart disease: a text book of cardiovascular medicine. 5th ed. Philadelphia: Saunders, 1997:1007-77

17.Nishimura RA, Giuliani ER, Brandenburg RO, et al. Hypertrophic cardiomyopathy. In: Giuliani ER, Gersh BJ, McGoon MG, et al. Mayo Clinic practice of cardiology. 3d ed 2001.
18.Oh JK, Seward JB, Tajik AJ. Cardiomyopathies. In: Oh JK, Seward JB, Tajik AJ. The echo manual. 2d ed. Philadelphia: Lippincott-Raven, 1999:156-63

19.Criteria Committee of the New York Heart Association. Nomenclature and Criteria for Diagnosis. 9th ed. Boston: Little, Brown & Company; 1994.

20. B.J. Maron, J.M. Gardin, J.M. Flack *et al.*, 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. Circulation. 1996 Aug 1;94(3):588-9.

21.Shapiro LM, McKenna WJ. Distribution of left ventricular hypertrophy in hypertrophic cardiomyopathy: a two-dimensional echocardiographic study. *J Am Coll Cardiol*. 1983;2:437–444.

22.Maron BJ, Gottdiener JS, Epstein SE. Patterns and significance of distribution of left ventricular hypertrophy in hypertrophic cardiomyopathy: a wide-angle, two-dimensional echocardiographic study of 125 patients. *Am J Cardiol*. 1981;48:418–428

23. Maron BJ, McKenna WJ, Danielson GK, et al. American College of Cardiology/European Society of Cardiology clinical expert consensus document on hypertrophic cardiomyopathy J Am Coll Cardiol 2003;42:1687-1713.

24. S.H. Park, C. Shub, T.P. Nobrega *et al.*, Two-dimensional echocardiographic calculation of left ventricular mass as recommended by the American Society of Echocardiography: correlation with autopsy and M-mode echocardiography. *J Am Soc Echocardiogr* 9 (1996), pp. 119–128.

25. D.J. Sahn, A. DeMaria, J. Kisslo and A. Weyman, Recommendations regarding quantitation in M-mode echocardiography: results of a survey of echocardiographic measurements. *Circulation* 58 (1978), pp. 1072–1083.

26. L.E. Teichholtz, T. Kreulen, M.V. Herman and R. Gorlin, Problems in echocardiographic volume determinations: echocardiographic-angiographic correlations in the presence or absence of asynergy. *Am J Cardiol* 37 (1976), pp. 7–11.

27.Sasson Z, Yock PG, Hatle LK, et al. Doppler echocardiographic determination of the pressure gradient in hypertrophic cardiomyopathy J Am Coll Cardiol 1988;11:752-75

28. M.S. Maron, I. Olivotto, S. Betocchi *et al.*, Effect of left ventricular outflow tract obstruction on clinical outcome in hypertrophic cardiomyopathy. *N Engl J Med* 348 (2003), pp. 295–303.

29. M.J. Kofflard, F.J. ten Cate, C. van der Lee and R.T. van Domburg, Hypertrophic cardiomyopathy in a large community-based population: clinical outcome and identification of risk factors for sudden cardiac death and clinical deterioration. *J Am Coll Cardiol* 41 (2003), pp. 987–993

30.Zou Y, Song L, Wang Z, Ma A, Liu T, Gu H, Lu S, Wu P, Zhang Y, Shen L, Cai Y, Zhen Y, Liu Y, Hui R. Prevalence of idiopathic hypertrophic cardiomyopathy in China: a population-based echocardiographic analysis of 8080 adults. *Am J Med.* 2004; 116: 14–18

31. Y. Hada, T. Sakamoto, K. Amano *et al.*, Prevalence of hypertrophic cardiomyopathy in a population of adult Japanese workers as detected by echocardiographic screening. *Am J Cardiol* 59 (1987), pp. 183–184.

32. Marcel J. M. Kofflard,et al: HYPERTROPHIC CARDIOMYOPATHY. Hypertrophic cardiomyopathy in a large community-based population: clinical outcome and identification of risk factors for sudden cardiac death and clinical deterioration J Am Coll Cardiol, 2003; 41:987-993, doi:10.1016/S0735-1097(02)03004-8.

33. Maron BJ, et al Prevalence of hypertrophic cardiomyopathy in an outpatient population referred for echocardiographic study. 1: Am J Cardiol. 1994 Mar 15;73(8):577-80.

34. Barry J. Maron,et al Clinical profile of hypertrophic cardiomyopathy identified de novo in rural communities. J Am Coll Cardiol, 1999; 33:1590-1595

35. B.J. Maron, Hypertrophic cardiomyopathy. *Lancet* 350 (1997), pp. 127–133

36. Maron BJ,et al : Prevalence of hypertrophic cardiomyopathy in a population-based sample of American Indians aged 51 to 77 years (the Strong Heart Study). Am J Cardiol. 2004 Jun 15;93(12):1510-4.

37. K. Miura, H. Nakagawa, Y. Morikawa *et al.*, Epidemiology of idiopathic cardiomyopathy in Japan: results from a nationwide survey. *Heart* 87 (2002), pp. 126–130.

38. M.B. Codd, D.D. Sugrue, B.J. Gersh and L.J. Melton, III, Epidemiology of idiopathic dilated and hypertrophic cardiomyopathy: a population-based study in Olmsted County, Minnesota, 1975-1984. *Circulation* 80 (1989), pp. 564–572.

39. Levine RA. Echocardiographic assessment of the cardiomyopathies. In: Weyman AE. Principles and practice of echocardiography. 2d ed. Philadelphia: Lea & Febinger, 1994:781-804

40. B.J. Maron, S.A. Casey, L.C. Poliac, T.E. Gohman, A.K. Almquist and D.M. Aeppli, Clinical course of hypertrophic cardiomyopathy in a regional United States cohort. *JAMA* 281 (1999), pp. 650–655.

41. Maron BJ, Casey SA, Hauser RG, Aeppli DM. Clinical course of hypertrophic cardiomyopathy with survival to advanced age. J Am Coll Cardiol 2003;42:882–8

42. W.P. Fay, C.P. Taliercio, D.M. Ilstrup, A.J. Tajik and B.J. Gersh, Natural history of hypertrophic cardiomyopathy in the elderly. *J Am Coll Cardiol* 16 (1990), pp. 821–826.

43. E. Takagi, T. Yamakado and T. Nakano, Prognosis of completely asymptomatic adult patients with hypertrophic cardiomyopathy. *J Am Coll Cardiol* 33 (1999), pp. 206–211.

44.Wigle D, Sasson Z, Henderson MA, et al. Hypertrophic cardiomyopathy. The importance of the site and the extent of hypertrophy. A review Prog Cardiovasc Dis 1985;28:1-83.

45.Wigle ED, Rakowski H, Kimball BP, Williams WG. Hypertrophic cardiomyopathy—clinical spectrum and treatment Circulation 1995;92:1680-1692.

46.Sherrid M, Gunsburg DZ, Moldenhauer S, Pearle G. Systolic anterior motion begins at low left ventricular outflow tract velocity in obstructive hypertrophic cardiomyopathy J Am Coll Cardiol 2000;36:1344-1354.

47.Yoerger DM, Weyman AE Hypertrophic obstructive cardiomyopathy: mechanism of obstruction and response to therapy. Rev Cardiovasc Med. 2003 Fall;4(4):199-215

48. H.-H. Ho, K.L.F. Lee, C.-P. Lau and H.-F. Tse, Clinical characteristics of and long-term outcome in Chinese patients with hypertrophic cardiomyopathy. *Am J Med* 116 (2004), pp. 19–23.

49. I. Olivotto, F. Cecchi, S.A. Casey, A. Dolara, J.H. Traverse and B.J. Maron, Impact of atrial fibrillation on the clinical course of hypertrophic cardiomyopathy. *Circulation* 104 (2001), pp. 2517–2524

50. F. Cecchi, I. Olivotto, A. Montereggi, G.Santoro, A. Dolara and B.J. Maron, Hypertrophic cardiomyopathy in Tuscany: clinical course and outcome in an unselected regional population. *J Am Coll Cardiol* 26 (1995), pp.152-66.

51.Charles Berul: Hypertrophic, Cardiomyopathy, Health Sciences, The Chicago Medical School; Consulting Staff, Private Practice November 21, 2004.

52. H.G. Klues, A. Schiffers and B.J. Maron, Phenotypic spectrum and patterns of left ventricular hypertrophy in hypertrophic cardiomyopathy: Morphologic observations and significance as assessed by two-dimensional echocardiography in 600 patients. *J Am Coll Cardiol* 26 (1995), pp. 1699–1708.