FULL LENGTH ARTICLE

Hypertrophic cardiomyopathy in Saudi Arabian population: Clinical and echocardiographic characteristics and outcome analysis

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Background: **Current published literature on hypertrophic cardiomyopathy (HCM) comes primarily from Western populations. There is no published data on clinical and echocardiographic characteristics and long-term outcome of HCM in an Arab population.**

Methods: **We conducted a retrospective analysis of all patients 16 years or older diagnosed with HCM at our institution. Detailed clinical and echocardiographic data were collected and outcome was analyzed.**

Results: **A total of 69 patients were identified as having HCM. The mean age was 42 ± 16 years with 71% male patients. All patients were Saudi citizens with Arab ancestry. Details about family history and presenting symptoms were available for 44 and 48 patients consecutively. Nine (18%) patients were asymptomatic and were diagnosed based on abnormal cardiac auscultation. The commonest presenting symptoms were dyspnea with or without chest pain and palpitations occurring in 40 (81%) patients. Only 4 (9%) of 44 patients had a family history of HCM and /or sudden cardiac death (SCD). The most common ECG abnormality was left ventricular hypertrophy (LVH) present in 60 (86%) patients. The commonest septal hypertrophy morphology was mid-septal (catenoid) in 30 (43%) followed by neutral in 23 (33%), basal septal (sigmoid) in 3 (4%) and apical in 6 (8%) patients. Twenty (28%) patients had evidence of resting left ventricular cavity gradient of ⩾30 mmHg. Eleven (16%) patients had evidence of biventricular hypertrophy. Left ventricular ejection fraction was normal in 65 (94%) patients. Over a median (25–75 percentile) follow-up of 7 years (4.5–10), only 3 patients died, all of non-cardiac causes. There were no cases of SCD during the follow-up period. Six patients required an implantable cardioverter-defibrillator (ICD); five for primary prevention and one for secondary prevention. Only 1 patient progressed to end stage dilated cardiomyopathy.**

Conclusion: **The natural history of hypertrophic cardiomyopathy in the Saudi population appears to be benign with catenoid morphology being the most common septal hypertrophy pattern. Risk of SCD appears to be quite low in this population.**

Keywords: Hypertrophic cardiomyopathy, Echocardiography, Septal morphology, Outcome

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Introduction

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant familial disease due to mutations of the myocardial sarcomeric contractile proteins and occurs all over the world across all races [1,2]. Hypertrophic cardiomyopathy is defined by the presence of significant left ventricular hypertrophy (LVH) in the absence of secondary factors like systemic hypertension and aortic stenosis [1]. The reported prevalence in the West is about 0.2% or 1:500 in the general population [3]. Hypertrophic cardiomyopathy is a diverse disease with variable phenotypic expression (i.e. LVH pattern) with a substantial percentage of patients living a normal life without any significant limitation and minimal risk of sudden cardiac death (SCD) [4–8]. However, some patients will have significant symptoms including dyspnea, chest pain, syncope leading to substantial morbidity as well as mortality reported to be as high as 3–6% [6]. The most feared complication of HCM is SCD due to ventricular arrhythmias though it has been reported to occur in only 0.1–1% of patients [5]. This apparent discrepancy between incidence of SCD in different studies has been attributed to selection bias in tertiary care centers [9]. The pattern of left ventricular hypertrophy is quite variable in HCM and is associated with difference in morbidity and mortality. For instance, it has been reported that apical HCM in the Japanese population as well as those in North America is associated with mostly benign outcome [10,11]. The most common pattern of LVH is asymmetric septal hypertrophy involving the basal anterior septum as well as anterior free wall [12]. Despite initial reports of a close relationship between septal morphology and hemodynamics in HCM, there has been no reported association between the patterns of hypertrophy and survival [13]. Recently several groups have reported an association of the septal morphology and sarcomeric mutation by genotypes that may help in better risk assessment and management of patients with HCM [14,15]. Resting left ventricular outflow tract obstruction (LVOTO) has been shown in some studies to be an independent predictor of subsequent heart failure and death [16]. Nearly all of the current published data on HCM comes from Western populations. A PUBMED search failed to show any published clinical or epidemiological study of HCM in the Arab/Middle Eastern population. We sought to look at the clinical and echocardiographic characteristics and outcome of HCM in the Saudi population.

Abbreviations

HCM	hypertrophic cardiomyopathy
SCD	sudden cardiac death
ICD	implantable cardioverter-defibrillator
LVH	left ventricular hypertrophy
LVOT	left ventricular outflow tract
A4	apical four-chamber
PLAX	parasternal long axis
DT	deceleration time
Sa	lateral and septal mitral annular systolic velocity
Ea	lateral and septal mitral annular early diastolic velocity
Aa	lateral and septal mitral annular late diastolic velocity
IVRT	isovolumetric relaxation time
Ar-A	difference (in milliseconds) between atrial systolic flow reversal in pulmonary veins and mitral A-wave duration
SAM	systolic anterior motion of mitral leaflet
MR	mitral regurgitation
ECG	electrocardiogram
VT	ventricular tachycardia
LVEF	left ventricular ejection fraction
LVOTO	left ventricular outflow tract obstruction
CMR	cardiac magnetic resonance

Materials and methods

We retrospectively identified all patients between the ages of 16–80 years who were diagnosed with hypertrophic cardiomyopathy from an echocardiographic database between January 1997 and January 2007. HCM was diagnosed based on echocardiographic evidence of LVH $\geqslant$ 15 mm or between 12 and 15 mm in presence of family history of HCM. Patients with secondary causes of LVH like systemic hypertension and aortic stenosis or diagnosed infiltrative cardiomyopathy were excluded. Detailed baseline clinical characteristics including family history of HCM, presenting symptoms, ECG abnormalities and echocardiographic parameters were recorded. Patients were followed up for a median (25–75 percentile) of 7 (4.5–10) years in out-patient clinic or via telephonic contact in case of missed appointments. Primary end point was cardiac mortality as a result of SCD or progressive heart failure. Secondary outcome was all cause mortality. The study was approved by the institutional review board.

Echocardiographic data

Standard views were obtained using Philips iE-33 (Phillips Healthcare, Andover, MA (USA) and GE vivid E9 (GE Healthcare, USA). Two dimensional (2-D), M-Mode, color Doppler echocardiog-