

Prognosis of Hypertrophic Cardiomyopathy

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ABSTRACT

Background: The actual prognosis of hypertrophic cardiomyopathy, a disorder previously thought of to be notorious for an increased risk of untimely death is poorly defined.

Objective: The present study describes the findings of patients with Hypertrophic Cardiomyopathy followed for more than nineteen years in a large clinic population.

Materials and Methods: A clinic population of 113 patients with Hypertrophic Cardiomyopathy was prospectively studied to assess cardiac mortality in the overall groups and in selected subgroups commonly thought to be at high risk for sudden death. Diagnosis of hypertrophic cardiomyopathy was based on the presence of left ventricular hypertrophy without a known cause. Left ventricular hypertrophy was determined by echocardiography.

Results: During follow-up there were 11 cardiac and 2 non cardiac deaths. The annual cardiac mortality was 1% (95% confidence interval 0.2-1.8%). Relative risk for cardiac death was not significantly different in the presence of young age (< 30 years), family history for Hypertrophic Cardiomyopathy (HCM) and sudden death, history for syncope or previous cardiac arrest or both, ventricular tachycardia on 24-hour, holter monitoring or operation for refractory symptoms and outflowtract obstruction.

Conclusion: HCM has a relatively benign prognosis (1% cardiac annual mortality) that is 2-4 times less than previously thought. These findings might have important consequences for risk assessment in individual patients. Echocardiography is obligatory to determine the presence, and extent of myocardial hypertrophy. In addition, the technique allows differentiation between Hypertrophic Cardiomyopathy and Athlete's Heart.

INTRODUCTION

Hypertrophic cardiomyopathy is a primary cardiac disease often genetically transmitted with a diverse clinical and morphologic expression and characterized by unexplained left ventricular hypertrophy¹.

Most studies in the past are based on populations from large tertiary referral centers of highly selected patients. These studies have reported a severe prognosis with annual mortality due to sudden cardiac death of 2-4%^{2,3}. Recent studies however have suggested a more benign prognosis^{4,5}. The latter findings have important implications for treatment strategies and risk stratification for premature cardiac death. In the present study we describe our findings in a large clinic population of Hypertrophic Cardiomyopathy (HCM) and show that prognosis is significantly more benign than previously thought.

In addition we give some clues which can be found by echocardiography to differentiate between Hypertrophic Cardiomyopathy and Athlete's Heart.

Materials and Methods

Patients

Between 1970 and 1990, 113 patients with HCM were examined initially and at yearly intervals. Diagnosis was based on the presence of left ventricular hypertrophy of unexplained cause. Before 1979, diagnosis was based on clinical parameters alone and after 1979, diagnosis was based on echocardiography.

Echocardiographic diagnosis were a non dilated left ventricle with muscular hypertrophy (any wall thickness > 15 mm) in the absence of known causes for left ventricular hypertrophy such as valvular aortic stenosis or systemic hypertension. For children the degree of left ventricular hypertrophy was diagnosed in relation to body surface area. Figure 1 shows typical echocardiographic findings in a patient with Hypertrophic Cardiomyopathy.

All patients were followed on a yearly basis or more frequently as indicated. All patients were followed by one physician only. Sudden death was assumed to be of cardiac origin as a witnessed death within one hour after the onset of symptoms or an unwitnessed death in a subject known to be alive and functioning normally 24 hours before.

Follow up data were available for all patients, thus no patients were lost to follow up. Because of the Dutch Registration system patients who moved out of town could be traced in every case.

Statistics

Survival estimates and 95% confidence intervals were calculated according to the Kaplan Meier method. Yearly mortality rate was based on all available follow-up time.

The Cox regression model was used to assess individual risk factors for sudden death. relative risk and 95% confidence intervals are reported for each indicator (Figure 2).

RESULTS

The baseline characteristics of the 113 study patients (60 male and 53 female) at the time of the initial visit are shown in Table 1.

Seven patients reported multiple sudden death in the family. Twenty four patients had syncope at presentation.

Fifty seven patients were asymptomatic or had trivial symptoms (New York Heart Association I), 40 were mildly symptomatic (class II) and 16 were moderately symptomatic (class III). No patients had New York Heart Association class IV.

Follow up data

No patients were lost to follow-up. Mean follow up was 7 ± 6 years (range 1-19 years). During the follow up period 12 patients died and 1 was successfully resuscitated from cardiac arrest. Two patients died of non-cardiac causes. The remaining 11 deaths were cardiac of which 9 were sudden by defined criteria.

Mean age at cardiac death was 47 ± 18 years (range 26-80). The annual cardiac mortality in this population was 1% (95% confidence interval (0.2 - 1.8%)) (See also figure 2).

DISCUSSION

As is shown from the results of our study⁴ prognosis in Hypertrophic Cardiomyopathy is more benign than previously thought. Although overall mortality rates of 2-4% have been found earlier, sometimes up to annual mortality rates of 8.6%, the findings of the present study are in contrast to those previous reports (1-3).

Table 1

Baseline characteristics of patients with Hypertrophic CM

	Number %
Patients 113	
Male/female 60/53	
mean age (yrs) first visit	38 ± 15 (range 12-76)
NYHA functional class	
I	57 (50%)
II	40 (36%)
III	16 (14%)
IV	0 (0%)
LVOT gradient	
(at rest and/or provocation)	
≥ 50 mmHg	+ 38 (34%) - 68 (60%) 0 7 (6%)

Abbreviations

LVOT = left ventricular outflow tract;
 NYHA = New York Heart Association
 = + present 0 = absent - unknown
 CM = cardiomyopathy

Our study is both prospective and all-inclusive since the analysis has not excluded patients who are asymptomatic or have had cardiac surgery.

There are other studies that have reported benign prognosis in specific subgroups of patients with HCM such as the elderly, the non-obstructed and these with apical hypertrophy.

The results of our study and those of Spirito⁶ who had similar results can be explained by patient selec-

Table 2

HCM	Athlete's Heart	
+	Unusual Pattern of LVH	-
+	LV cavity < 45mm	-
+	LA enlargement	-
+	Bizarre EG patterns	-
+	Family History of HCM	-
+	Abnormal LV filling	-

tion.

In other centers reporting a more malignant prognosis patients with more severe progression of the disease are probably over represented. In our study all patients representing the clinical spectrum of HCM were studied.

Because of the low prevalence of cardiac death in our population we were not able to detect so called riskfactors for sudden cardiac death.

We also believe that expensive electrophysiologic or genetic testing is not justified for the overall population of HCM patients since annual mortality is so low.

Our experience has given also some insight for the use of echocardiography to differentiate between HCM and the Athlete's Heart. Although these findings are no absolute criteria, they have shown to be very helpful.

CONCLUSION

HCM has a benign prognosis of 1% annual cardiac mortality/year.

Echocardiography is obligatory for the diagnosis, follow up and differentiation of the disease from other causes of left ventricular hypertrophy.

References

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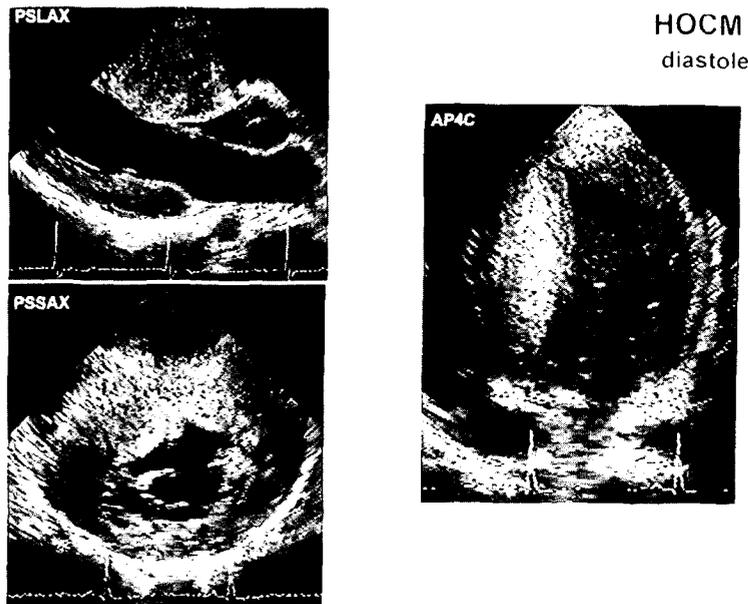


Figure 1.

Representative examples of a 2D Echocardiogram in a patient with Hypertrophic Obstructive Cardiomyopathy (HOCM). In PSLAX (parasternal long axis) the hypertrophied interventricular septal muscle is clearly seen. This is even better illustrated in the PSSAX (parasternal short axis) when the IVS (between 9 and 2 o'clock) is severely hypertrophied. The AP 4C (apical 4 chamber view) shows the IVS as the bright structure at the left side of the picture.

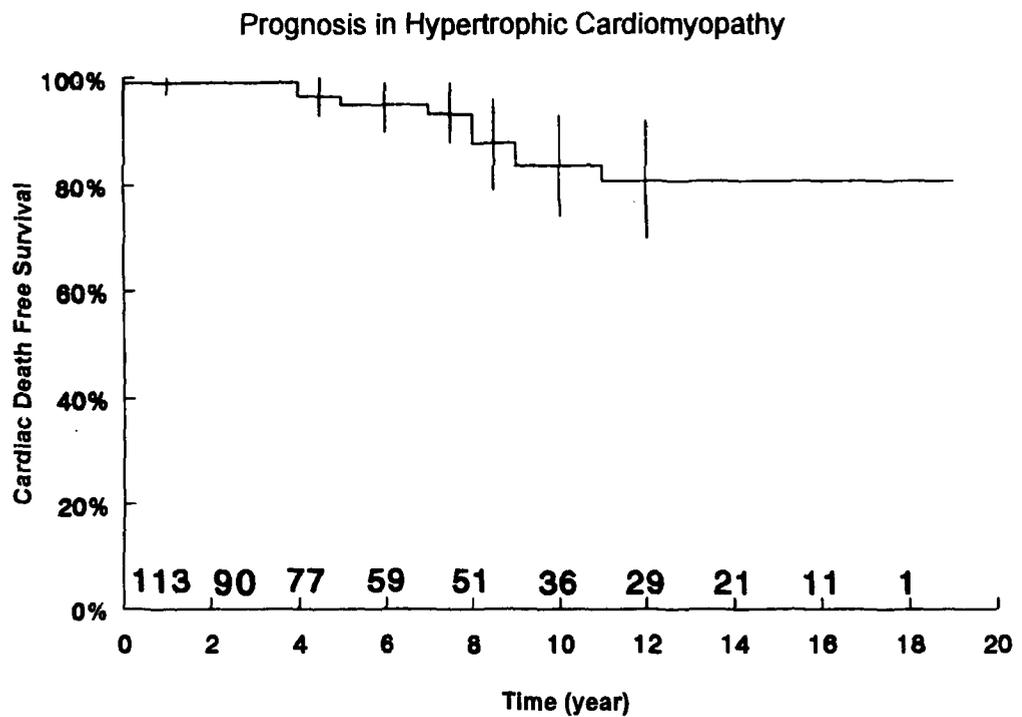


Figure 2.

Kaplan-Meyer survival for the patients described in the manuscript. Vertical lines indicates 95% confidence intervals. The number of patients at every year of the study is represented at the