

# Hypertrophic Cardiomyopathies at the Sylvanus Olympio Teaching Hospital of Lome: Epidemiological, Diagnostic, Therapeutic and Evolutive Aspects

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## Abstract

**Background:** Hypertrophic cardiomyopathy (HCM) is a form of cardiomyopathy that may involve several aspects. The aim of our study is to describe the epidemiological, diagnostic, therapeutic and short-term prognostic aspects of this form of cardiomyopathy at the Sylvanus Olympio Teaching Hospital of Lome. **Materials and Methods:** This was a cross-sectional study that was carried out over a four-year period from January 1, 2016 to December 31, 2019. We included in this study, patients admitted to the Cardiology Department of the Sylvanus Olympio Teaching Hospital of Lome, in whom the diagnosis of hypertrophic cardiomyopathy was retained at echocardiography in the absence of any other cause that could explain the significant hypertrophy of the walls. **Results:** The prevalence of hypertrophic cardiomyopathies in our study was 0.31%. The mean age of patients was  $51.35 \pm 15.28$  years with a male predominance (sex ratio M/F of 1.22). The majority of patients (60%) were between 45 and 74 years old. The clinical presentation was dominated by congestive heart failure in 15 patients (75%). Half of the patients (50%) had type III hypertrophic cardiomyopathy according to Maron's classification. Seven patients (35%) had obstructive HCM and the mean thickness of the interventricular septum in diastole was 15.88. Left ventricular systolic function was impaired in 40% of patients. No patient was able to do a genetic test. The combination of beta-blocker (95%), an inhibitor of the renin-angiotensin-aldosterone system (90%) and furosemide (85%) constituted the essential part of the treatment combined with Lifestyle changes. No patients have benefited from implantable cardioverter defibrillators. The yearly mor-

tality rate at the end of our study was 70%. **Conclusion:** Hypertrophic cardiomyopathy remains a relatively rare pathology. It is often a late diagnosis in the context of heart failure with limited therapeutic means, explaining its heavy morbidity and mortality.

## Keywords

Hypertrophic Cardiomyopathy, Echocardiography, Togo

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## 1. Introduction

Hypertrophic cardiomyopathy (HCM) is a primary myocardial disease that can have several clinical and anatomical aspects, often of familial genetic origin, autosomal dominant, genotypic and phenotypic expression very variable and whose idiopathic character is retained after excluding other causes of myocardial hypertrophy such as high blood pressure (hypertension), aortic narrowing and amyloidosis [1]. Cardiovascular disease (CVD), including cardiomyopathies, has become the leading cause of death worldwide over the past decade [2]. HCM is part of the group of cardiomyopathies, which includes, in addition to HCM, dilated cardiomyopathies (CMD), restrictive cardiomyopathies, arrhythmogenic cardiomyopathies, non-compaction of the left ventricle and Tako-Tsubo disease [3]. This is a major cause of sudden death of young people, particularly athletes under the age of 35 [4]. The prevalence of HCM is estimated to be 0.07% in Germany according to Husser [5], and 0.08% in China according to Zou [6]. In Africa, some studies have shown that HCM represented 34% of all cardiomyopathies diagnosed on echocardiography in Ethiopia [7], and in Ghana, HCM is the third most common form of cardiomyopathy [8]. Although less common than dilated cardiomyopathy, HCM has especially a prognostic problem [8]. No studies have yet been carried out in Togo on HCM. The objective of our study was to determine the epidemiological, diagnostic, therapeutic, evolutionary and prognostic characteristics of HCM.

## 2. Patients and Methods

This is a cross-sectional study that took place in the Cardiology Department of the Sylvanus Olympio Teaching Hospital, from January 1, 2016 to December 31, 2019, a period of 4 years. We included patients of both sexes and of any age hospitalized or followed in the Cardiology Department in whom the diagnosis of HCM was made at echocardiography in the absence of any other cause that could explain the significant hypertrophy of the walls. The following causes: hypertension, aortic stenosis amyloidosis and athlete's heart have been excluded on clinical and echocardiographic arguments. The diagnosis of HCM was retained if the thickness of the left ventricle was greater than or equal to 15 mm in one or more myocardial segment at echocardiography in the absence of differential diagnosis, or thickness of the left ventricle greater than or equal to 13 mm

in case of familial form. Patients suspected of HCM haven't documented by echocardiography were not included in this study. The course of our study included for each patient, an interrogation, a clinical examination, paraclinical examinations (chest X-ray, ECG, echocardiography and biology). The transthoracic echocardiography was performed with vivid4 and/or Esaote devices. The collection of clinical, paraclinical, therapeutic and evaluationary data was done using a pre-established survey sheet. Checks were carried out at 3, 6 and 12 months. At each appointment, patients were questioned and examined for adherence, signs of heart failure, the occurrence of complications, relapses with or without re-hospitalization. The circumstances, dates and places of death of patients who died outside the cardiology department were provided by the parents. We asked the parents (father, mother, sisters and brothers), if there were any to confirm the presence of familial cases of HCM.

To carry out this study, we had the authorization of the management of the hospital and the approval of the ethics committee. Statistical analysis was performed with EPI info software version 3.5.4.

### 3. Results

#### 1) Epidemiologic aspects

During the study period, we identified 6305 patients admitted to the cardiology department of the Sylvanus Olympio Teaching Hospital for cardiovascular pathology, including 1651 with cardiomyopathies of all forms (20%), and 20 cases of HCM. The prevalence of HCM was therefore 0.31% of all cardiovascular pathologies and 1.6% according to cardiomyopathies. The average age of patients was  $51.35 \pm 15.28$  years. Patients between 45 and 75 years old were 60%. There was a male predominance with a sex ratio M/F of 1.22. Menopause, smoking and Hypercholesterolemia were the most common cardiovascular risk factors. Some patients had several associated risk factors (**Table 1**).

#### 2) Clinical data

In this study, 15 patients or 75% of patients had congestive heart failure, 8 (40%) had a systolic murmur of mitral insufficiency and only one (5%) had a systolic murmur of tricuspid insufficiency. The average heart rate was  $103.58 \pm$

**Table 1.** Distribution of patients according to cardiovascular risk factors.

	Effective	Percentage
Menopause	5	25
Smoking	5	25
Hypercholesterolemia	5	25
Sedentary lifestyle	4	20
Familial form of HCM	3	15
Hypertriglyceridemia	2	10
Diabetes	1	5

16.74 beats per minute with extremes of 72 and 142 beats per minute.

### 3) Paraclinical data

Twelve (60%) patients had left ventricular hypertrophy. Heart rhythm disorders were common (ventricular and atrial extrasystoles, atrial fibrillation, ventricular tachycardia) on electrocardiogram (Table 2).

All patients had cardiomegaly on chest x-rays. The mean cardiothoracic index was  $0.60 \pm 0.05$ . Alveolar-interstitial syndrome was found in 10% of patients.

Echocardiography found in seven patients (35%) with obstructive HCM and the average thickness of the interventricular septum in diastole was 15.88 (Table 3). Eleven patients (55%) had a left atrial dilation and three patients (15%) had a dilated left ventricle. Left ventricular systolic function was impaired in 40% of patients. A mitral regurgitation was found in 40% of cases. Left ventricular filling pressures were higher in 11 patients (55%). Pulmonary arterial hypertension was present in 8 patients (40%), dilation of the right cavities in 3 patients (15%). Pericardial effusion was found in 6 cases (30%).

Half of the patients had HCM type III according to the Maron classification (Table 4).

Biologically, 11 patients (55%) had renal failure and anemia was found in 8 patients or 40%.

### 4) Therapeutic aspects

Lifestyle changes were routinely prescribed to all patients, which included rest, a low-sodium diet, cessation of smoking intoxication, a low-sugar diet in case of diabetes and a lipid-lowering diet in case of dyslipidemia. Treatment was essentially symptomatic. Beta-blockers, rennin angiotensin aldosterone system inhibitors and loop diuretics were the most prescribed (Table 5). No implantable

**Table 2.** Distribution of patients according to electrocardiographic signs.

Effective		Percentage
Left ventricular hypertrophy	12	60
Left atrial hypertrophy	10	50
Conduction disorders	10	50
Repolarization disorders	9	45
Ventricular extrasystoles	5	20
Right atrial hypertrophy	3	15
Auricular extrasystoles	2	10
Ventricular tachycardia	3	15
Atrial fibrillation	2	10
Left anterior fascicular block	2	10
Complete right bundle branch block	2	10
Myocardial infarction aspect	2	10
Incomplete right bundle branch block	1	5

**Table 3.** Echocardiographic parameters.

	Average value	extreme value
Interventricular septum thickness end-diastolic (mm)	15.88	13.2 - 23.8
Interventricular septum thickness end-systolic (mm)	17.61	13.8 - 24
Posterior wall thickness in diastole (mm)	14.42	8.1 - 20.7
Posterior wall thickness in systole (mm)	17.95	13.2 - 22.2
IVSd/LVPWd*	1.64	1.45 - 1.98
Left ventricular end-diastolic diameter (mm)	43.36	20 - 76.5
Left ventricular end-systolic diameter (mm)	32.75	15 - 62.7
Left auricle area (cm <sup>2</sup> )	24.80	12 - 35
Right auricle area (cm <sup>2</sup> )	24.11	14 - 31
Left auricle diameter (mm)	38.43	28 - 52.9
Left ventricular ejection fraction (%)	43.35	27 - 86
Left ventricular fractional shortening (%)	30.3	9 - 54

\*Interventricular septum thickness end-diastolic/Posterior wall thickness in diastole.

**Table 4.** Distribution of patients according to Maron's classification.

	Effective	Percentage
Type I	1	5
Type II	6	30
Type III	10	50
Type IV	3	15
<b>Total</b>	<b>20</b>	<b>100,00</b>

**Table 5.** Distribution of patients by drug prescription.

	Effective	Percentage
Beta-blockers	19	95
Renin-angiotensin system inhibitors	18	90
Loop diuretics	17	85
Calcium channel blockers	14	70
Low molecular weight heparin	13	65
Aldosterone inhibitor	4	20
Nitrate derivatives	3	15
Statin	3	15

cardioverter defibrillator therapy has been instituted. No patients have undergone septal alcoholism or myectomy.

### 6) Evolutive aspects

Of the 20 patients, 11 (55%) were hospitalized and 9 (45%) were followed on an outpatient basis. The average duration of hospitalization was 11.73 days with extremes of 7 to 20 days. Two deaths were recorded during hospitalization. The remaining patients were followed for at least 12 months. At 12 months we recorded 14 patients who died, an overall yearly mortality of 70%. The leading cause of death was sudden death in 10 patients (71%).

## 4. Discussion

### 1) Main results

Our work has therefore regained a prevalence of HCM of 0.31%. The average age of patients was  $51.35 \pm 15.28$  years with a male predominance. Seventy-five percent of patients were in congestive heart failure. Echocardiography found in seven patients (35%) with obstructive HCM and the average thickness of the interventricular septum in diastole was 15.88. Left ventricular systolic function was impaired in 40% of patients. Half of the patients had HCM type III according to Maron's classification. Beta-blockers, inhibitors of the renin-angiotensin-aldosterone system and loop diuretics were the most prescribed. No implantable cardioverter defibrillator therapy has been instituted. No patients have undergone septal alcoholism or myectomy. The overall mortality rate in 1 year was 70% with sudden death as the main cause.

### 2) Epidemiologic aspects

In our series, HCM has proven to be a rare condition representing 0.31% of cardiovascular pathologies in cardiology at Chu so. HCM remain a rare condition around the world. In the West according to Husser *et al.* [5] in Germany, HCM has hospital prevalence between 0.03% and 0.2%. In France, according to Veselka *et al.* [9] the prevalence is 0.2%.

We regained a male predominance with a sex ratio M/F equal to 1.22. Male predominance is thought to be explained by a higher penetrance of the disease in men than in women and earlier expression of HCM in men than in women. Charron regained a penetrance of the disease by 58% in women and 77% in men [10]. He also found an earlier expression of the pathology in men than in women.

The average age of patients was  $51.35 \pm 15.28$  years with extremes between 26 and 75 years. Husser *et al.* [5] in Germany had found an average age of  $63 \pm 17$ . Saar *et al.* [11] In Dakar had found an average age of 53.25 years with extremes of 27 and 79 years.

These epidemiological data by age show that the pathology affects both young and elderly subjects. In addition, patients in sub-Saharan Africa are relatively younger than those in Europe.

### 3) Clinical data

Congestive heart failure was found in 75% of cases. This situation is explained by the late arrival in hospitals for patients with HCM.

### 4) Paraclinical data

One of the electrical abnormalities found in our series were rhythm disorders

including ventricular extrasystoles (20%), atrial extrasystoles (10%), atrial fibrillation (10%) and ventricular tachycardia (15%). It is equivalent to saying that ventricular and atrial hyperexcitability accounted for 30% of cases hence its frequency during HCM. These data are similar to the result of Charron *et al.* [10] who found a frequency of 31.2% of hyperexcitability in 2015. Atrial fibrillation is thought to be the cause of ischemic strokes according to data from the literature [10] [12].

Left ventricular hypertrophy was found in 12 patients (60%) with electrocardiogram. Charron *et al.* [10] found left ventricular hypertrophy in about 55% of cases, which seemed to be associated with a more pronounced degree of ultrasound hypertrophy.

In our series, intraventricular conduction disorder was present in 5 patients (25%) and there was no atrioventricular conduction disorder. Charron and Desnos [10] [13] had also found cases of conduction disorders in the HCM.

The maximum ultrasound septal hypertrophy found in our work was on average 15.88 mm which is similar to the result of Niamkey [14] who had found 17.5 mm. Maron's type III predominated (50%). Our proportion is lower than that of Niamkey (83.3%) [14]. According to Maron [4], several studies found a maximum septal hypertrophy on average between 21 and 23 mm and they found 52% type III.

#### **5) Therapeutic aspects**

All patients were put on medical treatment. Beta-blockers were the most prescribed. According to Charron in 2005 [10], propranolol was the most prescribed. Their prescription would be explained by their effect on the reduction of morbidity-mortality and on the slowing of the heart rate.

In our series no patient had received an implantable automatic defibrillator for lack of technical platform and financial problems.

#### **6) Evolutive and prognostic aspects**

The evolution of HCM is marked by the occurrence of episodes of cardiac decompensation and complications leading to myocardial decay. The intra-hospital mortality rate recorded in our series was high (25%). The evolution was marked by a mortality rate in 12 months of 70%. This would explain the severe prognosis of HCM in our series. The cause of death was sudden death, the main circumstance of death described by several authors [12] [14].

#### **7) Limitations and constraints of the study**

The strengths of our study lie in the fact that it is the first in Togo and it was prospective. Moreover despite the rarity of this pathology, we had registered 20 patients. Our study has some short comings including:

- an insufficient technical platform making diagnosis difficult etiological;
- the modest income of our populations limits the realization of certain additional examinations.

Nevertheless, we believe that the objectives we had set ourselves at the beginning of our work have been achieved.

## 5. Conclusions

Hypertrophic cardiomyopathies are rare and serious conditions. They reach relatively young subjects and are clinically manifested by congestive heart failure. The most common electrical signs were left ventricular hypertrophy. The most common rhythm disorders were ventricular extrasystoles. Cardiac Doppler ultrasound confirms the diagnosis and specifies the degree of myocardial hypertrophy. HCM had a serious problem with therapeutic management, the inaccessibility of implantable cardioverter defibrillators and other interventional management techniques. Nevertheless, beta-blockers were widely prescribed in order to reduce the risk of rhythmic complications. Mortality of HCM is high both during hospitalization and during follow-up.

The relative scarcity of HCM makes it necessary to focus on screening during medical consultations with systematic family surveys.

## Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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