Hypertrophic Cardiomyopathy: The Importance of Arrhythmic Events in Patients at Risk for Sudden Cardiac Death

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Objective: To evaluate, in patients with hypertrophic cardiomyopathy and risk for SCD who underwent implantable cardioverter-defibrillator (ICD) implantation: a- the occurrence of arrhythmic events; b- the occurrence of clinical events and their correlation with arrhythmic events; c- the occurrence of ICD shock therapy and clinical and functional correlations; d- clinical and functional predictors of prognosis.

Methods: Twenty six patients with hypertrophic cardiomyopathy and risk factors for SCD undergoing ICD implantation from May, 2000 to January, 2004 (mean follow-up = 20 months) were studied. Fourteen patients (53.8%) were females and the mean age was 42.7 years. ICD was indicated for primary prevention of sudden cardiac death in 16 patients (61.5%), and for secondary prevention in 10 patients (38.5%). Twenty patients (76.9%) presented syncope prior to ICD implantation; half of them were related to ventricular fibrillation or sustained ventricular tachycardia, 15 (57.7%) had a history of familial sudden death, 12 patients (46.2%) had nonsustained ventricular tachycardia on the 24-hour Holter monitoring, and five (19.2%) had an interventricular septal thickness greater than 30 mm.

Results: Four shock therapies were recorded by the ICD in potentially lethal arrhythmias (three patients with sustained ventricular tachycardia and one patient with ventricular fibrillation) during the follow-up. One death occurred, probably due to a thromboembolic stroke. Four patients had recurrence of syncope with no arrhythmic event recorded by the ICD. The statistical analysis showed a significant difference in early ICD shock therapy in patients whose interventricular septal thickness was greater than 30mm.

Conclusion: 1- occurrence of arrhythmic events in 50% of the patients; most of them (62%) were ventricular tachycardia, whether sustained (31%) or nonsustained (31%); in the remaining patients paroxysmal supraventricular tachycardia was observed. 2- recurrent syncope in the minority of the patients (16%), however not associated with the presence of arrhythmic events. 3- the presence of an interventricular septal thickness greater than 30mm in the echocardiogram was associated with early shock therapy (p = 0.003). 4- absence of clinical or functional predictors.

Key words: Hypertrophic cardiomyopathy, arrhythmia, implantable cardioverter-defibrillator, risk factors, sudden cardiac death.

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant genetic disease[1] characterized by left ventricular hypertrophy in the absence of other causes of myocardial mass enlargement. HCM is considered the leading cause of sudden cardiac death (SCD) among young individuals, including competitive athletes[2]. Because it is a polygenic disease with clinical heterogeneity and known mutations, interest has grown in the search of new mutations through clinical studies and laboratory tests that could determine the risk of sudden death (SD)[3,4].

Syncope and SCD, which are the most dramatic clinical manifestations of the disease[5,6], usually result from ventricular dysfunction.

The fundamental objectives of the treatment of HCM are the relief of symptoms and prevention of SCD. SCD frequently affects young individuals previously asymptomatic or with mild symptoms. The incidence of SCD is of 1% per year among adults with HCM, and of 2 to 4% among children and adolescents[7,8,9,10]. Risk stratification is very difficult and the definition of an efficient prophylactic treatment hasproven a significant clinical challenge[6,11,12,13]. In a recent publication on SD causes in 387 young athletes, the major cause, occurring in 102 cases (26.4%), was HCM; in 29 other deaths (7.5%) there was left ventricular hypertrophia of undetermined cause[2]; the doubt whether HCM could be the cause remained unclarified.

Some studies, however, have demonstrated evidences of

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predictive factors of a higher risk for SCD in HCM[11,13,14,15,16]. Among these findings, VT seems to be the main mechanism of SCD of the disease[15], and is related to the degree of fibrosis between the myocardial fibers. The presence of NSVT is associated with an unfavorable course only in the presence of symptoms of low systemic or cerebral blood flow. In this case, amiodarone is the antiarrhythmic drug of choice, although its impact on the survival of these patients is controversial[17]. Some reports show that patients with NSVT have a higher risk for SCD than those without NSVT[14].

The presence of recurrent syncopes, even in situations where VT is not documented, seems to be an important risk marker of sudden death, but there are no conclusive studies on this respect.

The treatment of ventricular tachyarrhythmias with an implantable cardioverter defibrillator (ICD) demonstrates that patients with HCM resuscitated from cardiac arrest, as well as those with syncopal SVT, show an important improvement of prognosis and life expectancy after ICD implantation[18,19,20]. This can be an excellent supporting role to its antitachycardia function (through strategies of programming) allowing continuous monitoring of the cardiac electrical activity by means of arrhythmic event storage.

Strong predictors of SCD in HCM, such as syncope of non-identified etiology, history of familial SCD, echocardiographic measurement of the septum or of the LV posterior wall greater than 30mm, onset of symptoms in childhood, presence of paroxysmal or persistent atrial fibrillation, and other little-studied predictors such as genetic mutations, proved to be properly assessed using ICD implantation which, above all, would obviously provide these patients with therapeutic safety.

Although HCM is an infrequently found disease, rarely responsible for SCD, the clinical evidences reported above and the fact that ICD has recently become available in our setting encouraged us to conduct this study.

Objectives

To evaluate, in patients with HMC and risk for SCD undergoing ICD implantation: a) the occurrence of arrhythmic events; b) the occurrence of clinical events and their correlation with arrhythmic events; c) the occurrence of ICD shock therapy and clinical and functional correlations; d) clinical and functional predictors of prognosis.

Methods

Twenty six consecutive patients at risk for SCD were prospectively selected among 476 HCM patients of a cohort from May, 2000 to January, 2004. All patients signed the Informed Consent Form.

Patients of any age or gender with HCM (obstructive or non-obstructive form) were included and classified according to the indication of ICD into two groups: a) group of secondary prevention of SCD, those resuscitated from cardiopulmonary arrest (CPA) or syncopal SVT; b) group of primary prevention, patients without potentially documented arrhythmia, but with two or more factors predictive of risk for SCD associated: history of SCD in first-degree relatives (parents and siblings younger than 40 years of age); recurrent syncope of non-identified etiology; LV wall thickness > 30mm and with NSVT on the 24-hour Holter monitoring.

Patients with systolic heart failure, valve disease, systemic hypertension, as well as pregnant women were excluded.

Thus, 26 patients participated in the study; 14 of them (53.8%) were females, and the mean age 42.7 years. All had a normal ejection fraction on the echocardiogram, and most (88.5%) had a heart failure functional class I or II, according to the New York Heart Association (NYHA). Other clinical and demographic characteristics of the patients are shown in Table 1.

Tests Performed:

Clinical Evaluation - The clinical evaluation was focused on the occurrence of familial SCD (parents and siblings younger than 40 years of age), presence of syncope, dizziness, palpitations, dyspnea and chest pain. Functional classification according to the NYHA criteria, medication used, and optimized daily dose.

Echo-doppler - Two-dimensional echocardiographic study, M-mode and Doppler (pulsatile, continuous and color flow mapping) using an Acuson model Sequoia instrument (Mountain View, CA), obtaining parasternal and apical views. M-mode and Doppler echocardiographic records were guided by the two-dimensional study and followed the recommendations of the American Society of Echocardiography[21].

Criteria for the echocardiographic diagnosis of HCM[22] - (measurements taken in 3 consecutive cardiac cycles, and values calculated using the simple mean):

1) Basic measurements of the M-mode echocardiogram:

- left atrial diameter;
- LV posterior wall diastolic thickness;
- interventricular septal diastolic thickness;
- LV end-systolic and diastolic diameter.

2) Values calculated from the basic M-mode measurements:

- ejection fraction using the Teicholz formula[23] : $dV = D^3 \times (7/2.4 + D)$;
- $sV = S^3 \times (7/2.4 + S)$ $EF = dV - sV / dV$

3) Basic measurements on two-dimensional echocardiography:

- Diastolic thickness of the wall with the greatest thickness.

4) Basic measurements on Doppler:

- E wave = peak transmitral protodiastolic flow velocity;
- A wave = peak transmitral telediastolic flow velocity;
- E/A ratio = ratio between E- wave and A- wave velocities;
- LV isovolumetric relaxation time;
- E wave deceleration time.

24-hour dynamic electrocardiography using the Holter system - Use of Marquette 8000 portable recorders with calibration set at 1mV: 10mm and amplitude-modulated (AM) wave record, containing a cassette tape for continuous

	N (%)
Gender	
Male	12 (46.2)
Female	14 (53.8)
Age	
17 to 40 years	11 (42.3)
40 to 70 years	15 (57.7)
Mean	42.7 years
Indication of ICD	
Primary prevention of SCD	16 (61.5%)
Secondary prevention of SCD	10 (38.5%)
Familial SCD	
Yes	15 (57.7)
No	11 (42.3)
Syncope	
Present	20 (76.9)
Absent	6 (23.1)
Functional class (NYHA)	
I-II	23 (88.5)
III	3 (11.5)
Medication used	
β-blocker	10 (38.5)
Verapamil	3 (11.5)
Amiodarone	3 (11.5)
Amiodarone + Verapamil	1 (3.8)
Echocardiogram	
IV septum < 30mm	21 (80.8)
IV septum > 30mm	5 (19.2)

IV - Interventricular

Table 1 - Patient distribution according to clinical characteristics

recording for further analysis in a GE-Marquette, MARS – version 4.0 Holter monitoring instrument revised by a technician and a responsible physician. The variables analyzed were: mean heart rate, number of supraventricular extrasystoles per hour, presence or absence of sustained and non-sustained PSVT and occurrence of atrial fibrillation; number and type of ventricular extrasystoles, and presence or absence of NSVT (3 or more consecutive beats with a heart rate higher than 100 bpm).

Electrophysiological study - Indicated in 16 patients as a supporting test to clarify the symptoms (syncopes), using a programmed ventricular pacing protocol consisting of: induction of ventricular arrhythmia with two pacing cycles and three extra stimuli in the apex and right ventricular outflow tract; programmed atrial pacing with two cycles, and two extra stimuli to assess atrial arrhythmias, in addition to measurement of basic intervals of the intracardiac conduction system.

ICD implantation - The type of indication defined the group (primary or secondary prevention) and corresponded to the inclusion criteria of the study, which are described in the Guidelines of the Departments of Arrhythmia and Clinical Electrophysiology of the Brazilian Society of Cardiology and Artificial Cardiac Pacing of the Brazilian Society of Cardiovascular Surgery[24].

The pacing systems used were of the double-chamber type, complying with the conventional technique of endocardial pacemaker (PM) implantation using radioscopy. Two bipolar endocardial leads were implanted, one positioned in the right atrial appendix region, and the other in the right ventricular apex.

ICD programming followed a protocol: pacing frequency of 40bpm for the bradycardia zone (PM function), with three detection zones of tachycardia: Zone 1- frequency between 140 and 160 bpm; Zone 2- frequency between 160 and 180 bpm; Zone 3- frequency higher than 180 bpm.

For the two first zones, therapy functions were deactivated and the ICD operated as a recording monitor for all events occurring between these frequency zones, which were called Holter Zones.

In Zone 3, the shock therapy function was activated (cardioversion and/or defibrillation). The shock energy was programmed according to the defibrillation threshold obtained in the surgery.

Follow-up - The follow-up was comprised of clinical and electronic assessment of the ICD every 90 days, or whenever the patient had a syncope or reported ICD shock. The parameters assessed were: pacing threshold, sensitivity threshold, lead impedance and battery condition. The events recorded and stored by the ICD were retrieved in the form of intracardiac electrogram, including the mark channel recording. Shock therapy was considered appropriated when delivered in response to a potentially lethal arrhythmia (SVT or VF detected by the ICD), which was reverted by a cardioversion or defibrillation shock therapy.

The sequence of these procedures and the periodicity of ICD post-implantation assessments are shown in Figure 1.

Statistical methodology - Relative (percentage) and absolute (N) class frequencies of each qualitative variable were used. For quantitative variables, means and medians were used to summarize the information, followed by minimum and maximum standard deviations to indicate data variability.

The Pearson chi square test (VIEIRA, 1998)[25] was used to compare frequency distributions of qualitative variables. Fisher's exact test was used in situations where the values expected were lower than 5. The association between variables was considered significant for p values <0.05.

The survival analysis approach was also used, considering ICD shock as the endpoint of interest. Survival probabilities for the total of patients were calculated using the Kaplan-Meier method (Armitage, 1987)[25]. To assess the influence of variables such as familial SD, syncope, and interventricular septal thickness, Kaplan-Meier curves for the categories of each variable were plotted. The Log-Rank test was used to compare the curves (Armitage, 1987)[25].

Findings from the clinical assessment (recurrence of syncope), non-invasive laboratory tests, and EPS underwent statistical treatment to compare them with the information stored by the ICD.

Student's t tests were used for quantitative variables, and Mann Whitney non-parametric tests were used for independent samples, when data variability did not occur. The chi square test and Fisher's exact test were used for qualitative variables.

Results were considered statistically significant when their descriptive levels (p values) were lower than 0.05. For statistical calculations, the SPSS (Statistical Package for the Social Sciences) software version 10.0 for Windows was used.

Results

The mean follow-up period was 19 ± 11.1 months, ranging from 2.6 to 42.1 months.

In 14 (53.8%) patients, arrhythmias were recorded by the 24-hour Holter monitoring prior to ICD implantation. Twelve patients (46.2%) presented NSVT, six of which were from the secondary prevention group and six from the primary prevention group; PSVT occurred in 6 patients (23%). Four patients (15.4%) presented both types of arrhythmia and no cases of sustained tachyarrhythmia were observed.

The EPS documented hemodinamically unstable ventricular arrhythmia in 9 (56.2%) patients; SVT occurred in 4 of them (25%), VF in 4 (25%), and ventricular flutter in 1 (6.2%).

Indication of ICD for secondary prevention occurred in 10 (38.5%) patients: 5 (19.2%) survivors of cardiac arrest, and 5 (19.2%) with documented syncopal SVT; it was also indicated in 16 (61.5%) for primary prevention (association of two predictive factors of SCD); 5 (19.2%) with familial SCD and syncope; 4 (15.4%) with familial SCD and interventricular septal thickness > 30mm; 2 (7.7%) with familial SCD and NSVT on the 24-hour Holter monitoring; and 5 (19.2%) with syncope and NSVT on the 24-hour Holter monitoring. Among the indications of ICD for primary prevention, the most frequent criterion was familial SCD, which occurred in 11 (42.3%) patients.

The time elapsed between patient selection (after clinical assessment, echocardiographic study, 24-hour Holter monitoring and EPS) and ICD implantation was in average approximately 15 days.

During follow-up, the intracardiac electrogram records of the ICD did not document any arrhythmia in 13 (50%) patients. In five (19.2%) patients and in 4 (15.4%) PSVT and episodes of NSVT were respectively recorded. In four patients (15.4%), potentially lethal arrhythmias (3 SVT and 1 VF) were reverted with ICD shock therapy; in three of them the ICD had been indicated for secondary prevention and in only 1 patient for primary prevention (Tab. 2).

Recurrence of syncope without intracardiac electrocardiographic record of arrhythmia in the ICD occurred in 4 (15.4%) patients (Tab. 2). These patients underwent a tilt table test with a prolonged passive protocol[24], whose result was negative.

Only one death was recorded during the whole patient follow-up 24 months post-ICD implantation, due to CPA (Table 2). ICD shock therapy for reversion of VF had been documented in this patient three months prior to death. Additionally, the patient had a history of ischemic stroke with mild neurological sequela and was taking an oral anticoagulation medication for permanent atrial fibrillation.

The ICD-shock-free survival curve using the Kaplan-Meier method demonstrated that by the end of 30.9 months ICD

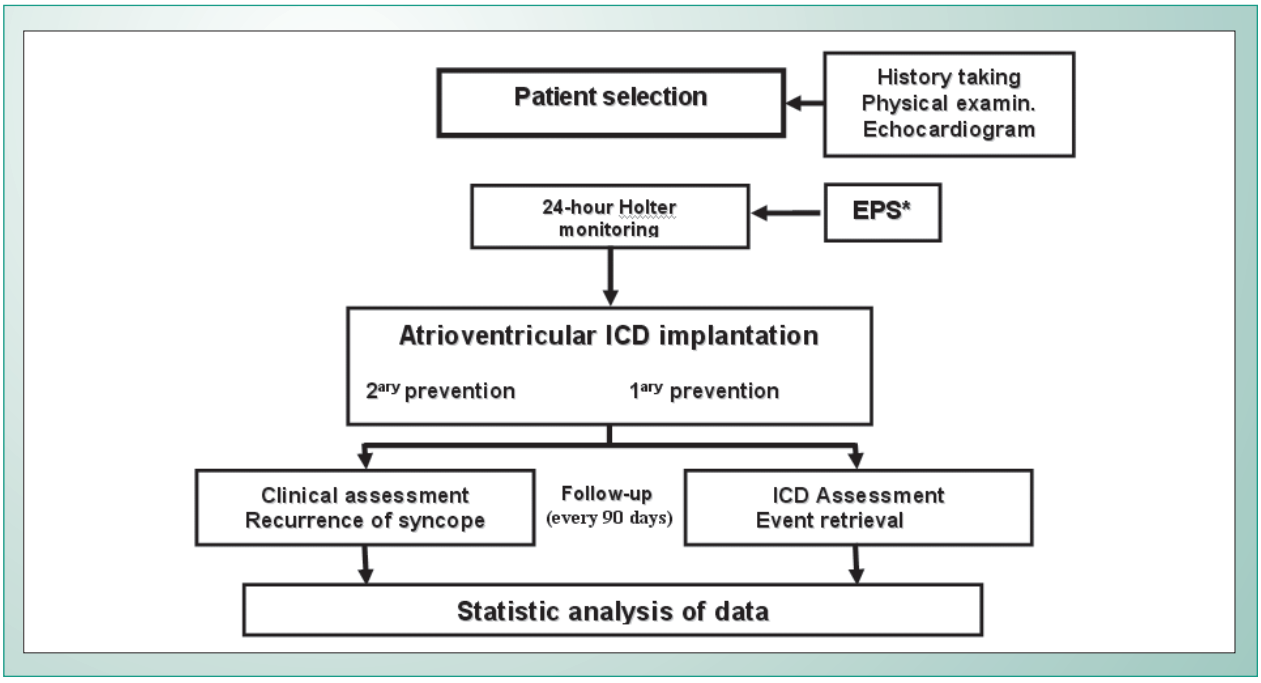

*Fig. 1 - Diagram of the study design sequence. *to clarify the symptoms.*

shock therapy had not occurred in 22 patients (Fig. 2A). The occurrence of shock therapies was not different among the patients with primary or secondary indications for ICD (p=0.517).

The comparative analysis of events according to the presence or absence of history of familial SD showed (Log-Rank test) that there was no difference between the groups (Fig. 2B).

When the occurrence of events (ICD shock) was compared with the occurrence of syncope prior to ICD implantation, no significant difference was observed either, as shown in Figure 2C.

In relation to the event-free survival curve (ICD shock) compared with the echocardiographic interventricular septal thickness, ICD shocks were demonstrated to occur earlier in patients who had an interventricular septal thickness greater than 30mm (p=0.03), as shown in Figure 2D.

No significant relation was observed between the arrhythmic events (PSVT, NSVT, SVT and VF) retrieved from the ICD electrograms and the clinical variables (gender, age, history of familial SCD, syncope prior to ICD implantation), echocardiographic variables (interventricular septal measurement greater or smaller than 30mm), presence of NSVT on the 24-hour Holter monitoring and presence of arrhythmias on the EPS (PSVT, NSVT, SVT and VF). Likewise, no relation was observed between recurrent syncope post-implantation and these variables.

Age under or above 40 years, as considered for statistical analysis, also did not discriminate a higher or lower incidence of either arrhythmic events recorded on the ICD electrograms, or of recurrence of syncope. The comparative analysis of arrhythmic events recorded by the ICD and recurrence of syncope in patients who presented syncope prior to implantation revealed that the four patients who had recurrence of syncope after ICD implantation presented previous syncope (Tab. 3).

Comparing the patients with history of familial SCD in relation to the arrhythmic events recorded by the ICD and the recurrence of post-implantation syncope, no statistically significant difference was observed. Also, the analysis of arrhythmic events retrieved from the electrograms recorded by the ICD or recurrence of syncope in relation to induction of potentially lethal arrhythmia such as SVT or VF on the EPS did not discriminate a higher incidence of occurrences (Tab. 3).

Interventricular septal thickness greater or smaller than 30mm (echocardiogram) and the presence of NSVT on the 24-hour Holter monitoring did not discriminate a higher or lower incidence of arrhythmic events recorded by the ICD or a recurrence of syncope (Tab. 3).

Discussion

Currently, the major focus of interest for the prognostic investigation of HCM is aimed at the subgroup of patients at high risk for sudden death. This prospective study is the first to be conducted in our country in patients with HCM and ICD indicated for risk of SCD. The population was highly selected, with a low prevalence of the disease, so the interpretation of the findings should be cautiously made.

Begley[18] documented the importance of ICD in survivors of SCD with HCM, however with reservations about the indication for primary prevention. Our study included: a) patients of secondary prevention of SCD, in three (12%) of whom appropriate shock therapy (VT/VF) occurred in a short period of time, all with potentially lethal arrhythmias prior to ICD implantation; b) patients of primary prevention, of whom only one presented appropriate shock six months after implantation. The low number of occurrence of shock in the group of primary prevention is likely due to the short follow-up period of the patients studied.

Corroborating Maron's findings[13,14], in our series, history of SCD among parents and siblings was not an isolated predictor of a higher occurrence of ICD shock therapy or record of arrhythmic events. Elliott and McKenna[7], however, reported a higher incidence of these events among patients with familial SD, which can be related to genotypic / phenotypic characteristics of the HCM.

Unlike some studies with a longer follow-up period, in which ventricular tachyarhythmia was considered responsible for syncopal features[18,19,20], in our study syncope previous to ICD implantation, which was present in 20 of our patients, was not associated with a higher incidence of arrhythmic events. Recurrence of syncope after ICD implantation with no arrhythmic event recorded, as occurred in 4 patients of our case series, should not reduce the value of this clinical manifestation, although isolated, in the indication of ICD. These four patients underwent tilt table test, whose result was negative (prolonged passive protocol)[24], thus ruling out a neurocardiogenic mechanism as the cause of syncope. Therefore, for these cases, the elucidation of the etiology of syncopes seems to depend on the follow-up period and on a judicious observation of further diagnostic approaches.

Among the anatomical and functional findings that characterize HCM, the most significant in the present study was the association between an interventricular septal thickness greater than 30mm, on the echocardiogram, with the early occurrence (6 or 7 months) of ICD shock therapy (p = 0.003).

In our study, NSVT (24-hour Holter monitoring) was the most prevalent arrhythmic event (46.2%), but was not a marker of poor prognosis. However, the absence of arrhythmias was not associated with a favorable progression of the disease. Maron[14] reported the importance of NSVT as a predictor of a higher incidence of SCD (risk of 8% in 3 years); he also reported that the absence of this arrhythmia provides a reduction of these rates by approximately 1%. In the present study, the ICD electrogram storage system was fundamental to document arrhythmic events: the highest prevalence was of ventricular tachyarrhythmias (SVT and VF), which is consistent with the findings in the literature.

Favale[27] reported the role of atrial fibrillation (incidence of 20%) as a precursor of fatal tachycardia in HCM. Two other non-randomized controlled trials[18,19] studied the documentation of arrhythmic events by intracardiac electrograms (most of them with single-ventricular chamber devices). Although supraventricular arrhythmias did not represent a threat to our patients, all received double-chamber (atrioventricular) devices. This is an important recommendation in this disease,

because, unlike single-chamber devices, atrioventricular devices have an advanced functional capacity for algorithmic discrimination and storage of arrhythmic events, thus significantly reducing the rate of inappropriate shocks.

EPS in the HCM risk stratification is controversial in the literature[28,29], and the findings in our study were also inconclusive. Two patients with induced SVT did not present clinical arrhythmia in 2 years of follow-up, and in one patient with SVT reverted by ICD shock therapy no arrhythmia was induced in the EPS.

Begley[18] estimated the survival rate of patients with ICD and HCM at risk for SCD at approximately 90% in a 10-year follow-up. One single death occurred in our case series (24 months post-ICD implantation): a female patient with permanent atrial fibrillation and occurrence of appropriate shock (VF) three months prior to death.

The occurrence of arrhythmic events and documented appropriate shock therapies in our study demonstrated that the indication of ICD in patients resuscitated from CPA or with syncopal SVT (secondary prevention) is unquestionable. On the other hand, in patients with potential risk of SCD without documented lethal arrhythmias (primary prevention) after a mean 20-month follow-up, the role of ICD could not be defined. For these situations, compliance with the guidelines

Patient	Gender	Age (years)	Syncope	Spontaneous arrhythmic event	Familial SCD	Iv septum (mm)	24-h Holter monitoring	EPS	ICD indication (prevention)	Follow-up period (months)	Post-ICD Syncope	Shock (SVT / VF)	Death
1	F	31	YES	-	+	29	NSVT	NP	1ary	23.7			
2	M	48	NO	-	+	25	NSVT	NP	1ary	40.1			
3	F	56	YES	SVT	-	27		NP	2ary	27.6			
4	F	23	YES	SVT	-	23		NAD	2ary	29.9		YES (SVT)	
5	M	17	YES	CPA	-	22	NSVT	NAD	2ary	36.5			
6	F	61	YES	CPA	-	28	NSVT	NP	2ary	23.7		YES (VF)	YES
7	F	60	YES	SVT	+	17		NP	2ary	25.0	YES		
8	M	21	YES	-	-	25		SVT	1ary	42.1			
9	F	59	YES	-	-	25	NSVT	SVT+VF	1ary	13.1	YES		
10	M	20	NO	-	+	37		NAD	1ary	6.6			
11	F	45	NO	-	+	23		NAD	1ary	15.9			
12	M	57	YES	-	-	22	NSVT	SVT+VF	1ary	16.0	YES		
13	F	37	YES	SVT	-	21		SVT	2ary	17.6			
14	F	38	YES	-	+	29	NSVT	NSVT	1ary	16.5			
15	F	70	YES	-	+	20	NSVT	Flutter V	1ary	11.2			
16	F	31	YES	CPA	+	23		NAD	2ary	8.3			
17	F	49	YES	SVT	-	24		NP	2ary	13.3	YES		
18	F	47	YES	-	+	36		NP	1ary	11.0			
19	M	51	YES	CPA	+	32		SVT	2ary	16.5		YES (SVT)	
20	M	54	YES	-	-	24	NSVT	SVT+VF	1ary	9.6			
21	M	42	YES	CPA	-	19		NAD	2ary	38.3			
22	M	54	NO	-	+	37	NSVT	NP	1ary	5.9		YES (SVT)	
23	F	38	NO	-	+	9	NSVT	NP	1ary	16.2			
24	M	49	YES	-	+	38	NSVT	VF	1ary	7.3			
25	M	29	NO	-	+	22		SVT	1ary	18.4			
26	M	24	YES	-	+	18		NP	1ary	2.6			

M – Male; F – Female; SVT – Sustained ventricular tachycardia; CPA – Cardiopulmonary arrest; Familial SCD –Familial sudden cardiac death; IV septum – interventricular septum; NSVT – Non-sustained ventricular tachycardia; EPS – electrophysiological study; VF – Ventricular fibrillation; V – Ventricular flutter; NP – Not performed; NAD – nothing abnormal detected; ICD – Implantable cardioverter defibrillator; 1ary – primary prevention; 2ary – secondary prevention.

Table 2 - Clinical and epidemiological characteristics, types of arrhythmic events and clinical and laboratory findings of each patient included in the study

Events N (%)		Age (years) N		Gender N		Syncope N		Familial SCD N		EPS N		IV septum N		24h NSVT N	
		<40	>40	Male	Fem	No	Yes	No	Yes	(-)	(+)	<30	>30	No	Yes
AT 5 (19.2)		2	3	3	2	1	4	2	3	3	2	4	1	3	2
NSVT 4 (15.4)		2	2	3	1	2	2	1	3	3	1	3	1	2	2
SVT or VF 4 (15.4)		1	3	2	2	1	3	2	2	3	1	2	2	2	2
Absent 13 (50)	*with syncope	0	4	1	3	0	4	3	1	2	2	4	0	2	2
	without syncope	6	3	3	6	2	7	3	6	6	3	8	1	4	5

*N – number of patients; AT – atrial tachyarrhythmia; NSVT – non-sustained ventricular tachycardia; EPS – electrophysiological study; SVT – sustained ventricular tachycardia; VF – ventricular fibrillation; *PVP – programmed ventricular pacing not performed in 2 patients.*

Table 3 - Types of arrhythmic events and rate of occurrence obtained from the electrograms recorded by the ICD according to epidemiological characteristics and laboratory findings

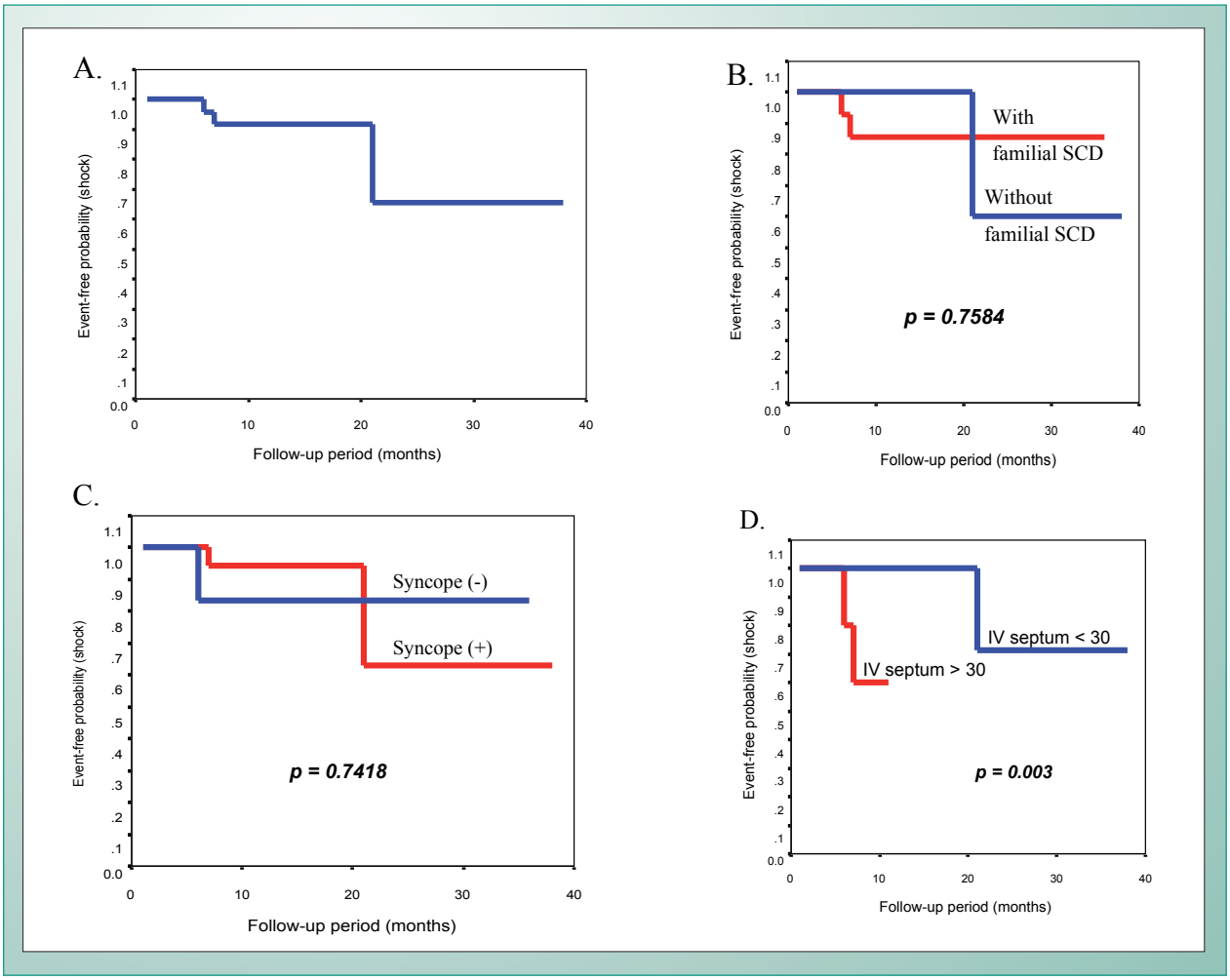

Fig. 2 - *Shock-free survival curves (Kaplan-Meier): A – Total case series. B – Presence or absence of history of familial SCD; C – Presence or absence of syncope prior to ICD implantation; D – Presence of interventricular septal thickness > 30mm (echocardiogram).*

of the Brazilian Society of Cardiac Arrhythmias – SBC is recommended.

Unfortunately, the present paper did not assess the genotypic characteristics of the patients studied. These characteristics will likely represent, in the near future, the decisive tools for the establishment of concrete predictors of prognosis in HCM.

Conclusions

The following findings were observed in patients with HCM and risk for SCD undergoing ICD implantation after a mean 20-month follow-up: 1- Occurrence of arrhythmic events in 50% of the patients (VT in 62%, of which SVT in 31% and NSVT in 31%); 2- Recurrent syncopes in the minority of the patients (16%), not associated with the presence of arrhythmic events; 3- Presence of interventricular septal thickness greater than 30mm, on the echocardiogram, associated with the occurrence of early shock therapy (p=0.003); 4- Absence of clinical or functional predictors.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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