

Implantable Cardioverter Defibrillator and Hypertrophic Cardiomyopathy. Experience at Three Centers

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Introduction and objectives. Although implantable cardioverter-defibrillators (ICDs) are recommended for high-risk patients with hypertrophic cardiomyopathy (HCM), there is no agreement on their general use. Moreover, little information is available on ICD use in this setting in Spain. Our aims were to describe the characteristics of HCM patients who received ICDs at three hospitals in Spain, and to study indications for device implantation and the results of follow-up in device users.

Methods. We evaluated risk factors for sudden death in HCM patients with ICDs, including family history of sudden death, recurrent syncope, maximum wall thickness ≥ 30 mm, left ventricular outflow pressure gradient >30 mm Hg, abnormal blood pressure response to exercise, and nonsustained ventricular tachycardia. During regular follow-up, appropriate and inappropriate administration of ICD therapy was recorded.

Results. Of 726 HCM patients, 45 (6.2%) had an ICD (mean age 43 [20] years). The proportion of patients with ICDs at the three centers studied was highly variable despite patients' clinical characteristics being similar. The indication for implantation was primary prevention in 27 patients and secondary prevention in 18. During follow-up (median 32 months), ICD therapy was administered appropriately in 10 (22.0%) patients (in 9, as secondary prevention and, in 1, as primary prevention). The annual appropriate ICD therapy rate was 11.1% for secondary prevention and 1.6% for primary prevention. Two patients

received an ICD to treat ventricular fibrillation and 8, to treat sustained ventricular tachycardia. The only significant predictor of appropriate ICD therapy was a history of sustained ventricular tachycardia or ventricular fibrillation (hazard ratio = 13.3, $P=0.014$).

Conclusions. The percentage of HCM patients undergoing ICD implantation at Spanish hospitals was highly variable, possibly due to different selection criteria. When used as secondary prevention, ICD therapy was administered appropriately in a high proportion of cases (50% in 3 years).

Key words: Hypertrophic cardiomyopathy. Implantable cardioverter-defibrillator. Sudden death.

Desfibrilador automático en la miocardiopatía hipertrófica. Experiencia de 3 centros

Introducción y objetivos. El desfibrilador automático implantable (DAI) es el tratamiento recomendado en la miocardiopatía hipertrófica (MCH) de alto riesgo, aunque no hay acuerdo en sus indicaciones. Hay pocos datos sobre su utilización en nuestro país. El objetivo es describir las características de los pacientes con MCH a los que se les implantó un DAI y analizar los resultados de esta terapia.

Métodos. Se analizaron los factores de riesgo de muerte súbita en los pacientes portadores de DAI de 3 centros con consultas dedicadas a la MCH (antecedentes personales y familiares de muerte súbita, síncope recurrente, grosor ≥ 30 mm y gradiente subaórtico > 30 mmHg, respuesta anormal de la presión al esfuerzo y taquicardia ventricular no sostenida) y la indicación del implante. Se realizó un seguimiento periódico y se registraron las terapias adecuadas e inadecuadas.

Resultados. De 726 pacientes, 45 (6.2%) eran portadores de DAI (edad de 43 ± 20 años). La proporción de pacientes con DAI en los 3 centros fue muy variable, a pesar de que las características de los pacientes eran similares. La indicación fue prevención primaria en 27 pacientes y secundaria en 18. Con un seguimiento de 32 meses, 10 pacientes (22%) recibieron tratamiento adecuado (9 de prevención secundaria y uno de prevención primaria). La tasa anual de tratamientos adecuados fue del 11,1% en prevención secundaria y del 1,6% en pre-

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Lorenzo Monserrat receives research support from Fundación Sanofi-Aventis. Lorenzo Monserrat and Xusto Fernández are funded by Red Cardiovascular RECAVA from Instituto de Salud Carlos III (C03/01). Dr Francisco Marín and Dr Juan G. Martínez declare that they have received a research grant from Medtronic Ibérica.

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Manuscript received July 26, 2005.
Accepted for publication January 23, 2006.

ABBREVIATIONS

ICD: implantable cardioverter defibrillator.

HCM: hypertrophic cardiomyopathy.

SD: sudden death.

RR: relative risk.

CI: confidence interval.

VT: ventricular tachycardia.

vención primaria. El único factor asociado con el tratamiento adecuado fue el antecedente de taquicardia ventricular sostenida o fibrilación ventricular (riesgo relativo [RR] = 13,3; $p = 0,014$).

Conclusiones. En consultas dedicadas a la MCH, el porcentaje de pacientes portadores de DAI varía en función del grado de selección de la población de origen. La incidencia de terapias adecuadas es elevada en prevención secundaria (el 50% en 3 años).

Palabras clave: Miocardiopatía hipertrófica. Desfibrilador automático implantable. Muerte súbita.

INTRODUCTION

The incidence of sudden death (SD), the most dramatic complication of hypertrophic cardiomyopathy (HCM), varies according to the patient series and the age of the affected individuals.¹ In recent years, the identification of various indicators associated with SD has allowed patients to be stratified according to their risk of developing this complication.^{2,3}

The use of implantable cardioverter defibrillators (ICD) has helped to reduce SD in various types of heart disease.⁴ However, in the case of HCM, the available data are less consistent, the patient series are small, and the length of follow-up short. In such studies, the main reason for implantation of an ICD is secondary prevention following aborted SD or sustained ventricular tachycardia (VT).^{5,6} Nevertheless, the results of a retrospective multicenter study containing a large number of patients,⁷ in which the benefit of ICD implantation was shown in both primary and secondary prevention of SD,⁸ suggest that the indications for ICD implantation should be expanded in HCM.

Despite the publication of treatment guidelines, the use of ICDs varies in different countries,⁹ and consequently, there is increasing interest in establishing registries of its use.¹⁰ These differences are more accentuated in the context of HCM, since

uniform criteria are not available for ICD implantation.¹¹

The aim of this study was to describe the characteristics of patients in Spain in whom ICDs were implanted for treatment of HCM, to analyze the incidence of ICD discharges, and to identify the variables associated with the requirement for appropriate ICD therapy.

PATIENTS AND METHODS

Patients

The study was performed in 3 Spanish hospitals with clinics dedicated to the treatment of patients with HCM. The study group included 726 patients (436 men) diagnosed with HCM who had a mean age of 50 ± 19 years and were in follow-up between January 2000 and November 2005 (Table 1). Of those patients, 45 (6.2%) had an ICD (28 men and 17 women; age at implantation, 43 ± 20 years). Similar diagnostic protocols and risk stratifications were used in all patients. The criterion for diagnosis of HCM was the presence of a left ventricular wall thickness of at least 15 mm without any other cause that could lead to ventricular hypertrophy, and in the case of first-degree relatives of affected individuals, current criteria were used.¹² Out of all the patients who attended follow-up appointments, 45 had received an ICD, either for secondary prevention (aborted SD or sustained VT) or primary prevention (in the presence of risk factors for SD).

Methods

The risk stratification protocol in the 3 hospitals involved specific questioning, along with 12-lead ECG, echocardiography, symptom-limited treadmill exercise testing, and Holter ECG. An assessment of the annual risk of SD was performed in all patients undergoing follow-up in the 3 hospitals. The following were defined as risk factors for SD: aborted SD, sustained VT, family history of SD, recurrent syncope without apparent cause, ventricular hypertrophy of at least 30 mm, left ventricular outflow tract gradient of more than 30 mm Hg at rest,¹³ abnormal blood pressure response during exercise in individuals under 45 years of age (abnormal blood pressure response was defined as the inability to increase systolic blood pressure by 25 mm Hg during the test¹⁴), and nonsustained VT in the Holter-ECG recording (3 or more ventricular ectopic beats at a rate of at least 120 beats per minute and a duration of less than 30 seconds).^{1-3,10}

Patients were assessed periodically in the subspecialty clinic of each hospital. Patients with ICDs, follow-up was undertaken at least every 3 to 6

TABLE 1. Number of Patients, Indications, and Appropriate and Inappropriate Discharges in Each Hospital*

Hospital	Alicante	Murcia	A Coruña	Total
Number of patients	143	223	360	726
Sex, men/women	73/70	139/84	229/131	441/285
Age, mean $\pm$ SD, y	58 $\pm$ 18	49 $\pm$ 18	50 $\pm$ 17	51 $\pm$ 18
Heart failure, NYHA $\geq$ 2	88 (61.5)	173 (77.6)	182 (50.6)	443 (61.0)
Maximum ventricular wall thickness, mean $\pm$ SD, mm	21 $\pm$ 5	20 $\pm$ 8	20 $\pm$ 6	20 $\pm$ 5
Left atrial diameter, mean $\pm$ SD, mm	40 $\pm$ 8	44 $\pm$ 4	43 $\pm$ 8	43 $\pm$ 7
Outflow tract gradient, n (%)	63 (44)	75 (34)	97 (27)	235 (32)
Number of patients with ICD	17 (11.9)	15 (6.7)	13 (3.6)	45 (6.2)
Primary prevention	12	9	6	27
Secondary prevention	5	6	7	18
Sustained VT	4	3	3	10
Ventricular fibrillation/aborted SD	1	3	4	8
Number of patients receiving appropriate discharges	5 (29.4)	2 (13.3)	3 (23.1)	10 (22.2)
Cause of appropriate discharge				
Sustained VT	3	2	3	8
Ventricular fibrillation	2	0	0	2
Number of patients receiving inappropriate discharges	4 (23.5)	3 (20.0)	6 (46.2)	13 (28.9)
Sinus tachycardia	3	2	3	8
Atrial flutter	1	1	1	3
Sensing problems	0	0	2	2
Infectious endocarditis	1 (5.9)	0	0	1 (2.2)

*Data shown as number of patients (%) unless otherwise indicated. NYHA indicates New York Heart Association; ICD, implantable cardioverter defibrillator; VT, ventricular tachycardia; SD, sudden death.

months or when considered necessary by the patient's doctor on the basis of suspicion or evidence of ICD discharges. The intracardiac ECG recordings stored in the devices were analyzed. Discharges were classified as appropriate when preceded by ventricular fibrillation or sustained VT, and inappropriate when preceded by sinus tachycardia or atrial fibrillation. Inappropriate discharges caused by device dysfunction or associated complications were also recorded.

Statistical Analysis

Continuous variables are expressed as mean $\pm$ SD and discrete variables as percentages. Analysis of variance (ANOVA) was used to assess the relationship between continuous and discrete variables. Discrete variables were compared using the χ^2 test. The influence of the different variables studied on the occurrence of ICD discharges was assessed by Cox regression analysis with calculation of the relative risk (RR) and 95% confidence interval (CI). Kaplan-Meier survival curves were also prepared and comparisons made between them using the log rank test. Results were considered significant when *P* was less than .05.

RESULTS

Out of 726 patients, 45 (6.2%) had received an ICD. The mean age at implantation was 42.8 $\pm$ 20.3 years and 28 (62.2%) of the patients were men. Obstructive

HCM was presented by 20 (44.4%) of the patients who received an ICD. The maximum ventricular wall thickness was 25.1 $\pm$ 6.8 mm. An electrophysiological study was performed prior to ICD implantation in 14 of the 45 patients (31.1%). Sustained VT was induced in 9 (64.3%) of those patients, while ventricular fibrillation was only induced in 1 (7.1%). The treatment was indicated for secondary prevention in 18 patients (40%): 8 following aborted SD (ventricular fibrillation having been identified in 7) and in the remaining 10 patients due to sustained VT. In 27 patients (60.0%), the ICD was indicated for primary prevention. Table 1 shows the number of implants, the indications, and the number and causes of appropriate and inappropriate therapies in patients from each hospital.

Assessment of risk indicators for SD revealed that 18 patients (40%) had a history of aborted SD or sustained VT, 14 patients (31.1%) had a family history of SD, 21 (46.7%) had recurrent syncope, 13 (28.9%) had severe hypertrophy, 20 (44.4%) had significant left ventricular outflow tract gradient, 25 (55.6%) presented an abnormal blood pressure response during exercise testing (although only 19 of those were younger than 45), and 29 patients (64.4%) presented nonsustained VT in Holter ECG. Table 2 shows the risk factors for SD in the patients studied. Of the 45 patients with ICDs, 2 (4.4%) presented 5 of the 7 recorded risk factors, 8 patients (17.8%) presented 4 factors, 23 patients (51.1%) presented 3 factors, 10 patients (22.2%) presented 2 risk factors, and only 2

TABLE 2. Risk Factors in Patients With Implantable Cardioverter Defibrillators*

Hospital	Alicante (n=17)	Murcia (n=15)	A Coruña (n=13)	Total (n=45)
History of SD or sustained VT	5	6	7	18
Family history of SD	8	4	2	14
Syncope	7	10	4	21
Nonsustained VT	12	8	9	29
Abnormal blood pressure response†	6	7	6	19
Wall thickness ≥ 30 mm	4	4	5	13
Gradient >30 mm Hg	10	4	6	20
Total number of risk factors/number of patients (mean)	52/17 (3.06)	43/15 (2.87)	39/13 (3.00)	134/45 (2.98)

*SD indicates sudden death; VT, ventricular tachycardia.

†Abnormal blood pressure response only considered as a risk factor for SD in patients younger than 45.

TABLE 3. Characteristics of Patients Who Received Appropriate Discharges*

Patient	Prevention Type	Months of Follow-Up	Number of Episodes of Ventricular Fibrillation	Number Sustained VT	Number of Episodes Treated With Shocks	Number of Episodes With Syncope	Associated Disease
1	2	75	1	1	1	1	None
2	2	60	0	2	0	0	Apical aneurysm
3	2	28	0	124	0	0	Ischemic heart disease
4	1	28	1	2	1	1	None
5	2	25	0	35	1	0	Ischemic heart disease
6	2	57	2	0	2	0	None
7	2	6	0	3	0	0	Systolic dysfunction
8	2	11	0	1	1	0	Apical aneurysm
9	2	69	0	10	8	0	Ischemic heart disease
10	2	50	0	45	4	0	Systolic dysfunction

*VT indicates ventricular tachycardia.

patients (4.4%) presented 1 risk factor. An interesting finding was that patients who had presented aborted SD or sustained VT had a lower number of risk factors than those with an ICD implanted for the purpose of primary prevention (1.94 ± 1.11 vs 2.96 ± 0.84 risk factors; $P=.002$; without counting aborted SD or sustained VT as a risk factor in the analysis).

At the time of ICD implantation, 13 patients (28.9%) received treatment with amiodarone and 29 patients (64.4%) were treated with beta-blockers.

The median follow-up was 32 months (25-75 percentile, 19-55 months) and all received follow-up of at least 6 months. Length of follow-up was greater in patients treated for secondary prevention than those treated for primary prevention (22 [11-42] months vs 51 [29-84] months; $P=.004$). During the follow-up period, 14 patients (31.1%) were treated with amiodarone and 38 (84.4%) with beta-blockers. Two patients with prior heart failure died at 71 and 73 years, 69 and 50 months after ICD implantation, respectively. Both patients had received appropriate discharges for sustained VT in the first year following ICD implantation. In total, 10 out of 45 patients (22.2%) received at least 1 appropriate discharge. This occurred within 1 month of implantation in 3 patients, within the first year in 5, and at 22 and 57 months

following ICD implantation in the remaining 2 patients. The arrhythmia preceding ICD discharge was ventricular fibrillation in 2 patients and sustained VT in the remaining 8 patients. When discharges were analyzed in relation to the indication for ICD, they were observed in 9 out of 18 patients (50%) who received an ICD for the purpose of secondary prevention, in 3 of the 8 patients (37.5%) with a history of aborted SD, and in 6 of the 10 patients (60.0%) with a history of sustained VT, while appropriate discharges were only observed in 1 of the 27 patients (3.7%) with an ICD implanted for primary prevention of SD. The annual rate of appropriate discharges was 11.1% in secondary prevention and 1.6% in primary prevention. Table 3 shows the number of discharges in each patient.

Table 4 shows the relationship between the different risk factors and the occurrence of appropriate discharges. The incidence of appropriate discharges was higher in patients with a history of sustained VT or aborted SD (RR=13.3; 95% CI, 1.7-106.2; $P=.014$). Survival analysis did not reveal significant differences for the other risk factors. There were no significant differences in the number of risk factors between patients with or without a requirement for ICD therapy (3.4 ± 1.1 with ICD therapy vs 2.8 ± 0.8 without ICD

TABLE 4. Risk Factors for Sudden Death Associated With Appropriate Discharges. Frequencies of Each Risk Factor With Results of Univariate Analysis (Cox Regression) and Statistical Significance*

	With Appropriate Discharges (n=10)	Without Appropriate Discharges (n=35)	RR (95% CI)	P
History of SD or sustained VT	9/10 (90%)	9/35 (26%)	13.3 (1.7-106.0)	0.02
Family history of SD	3/10 (30%)	11/35 (31%)	0.8 (0.2-3.1)	0.71
Syncope	7/10 (70%)	14/35 (40%)	2.6 (0.7-10.2)	0.16
Wall thickness ≥ 30 mm	2/10 (20%)	11/35 (31%)	0.6 (0.1-2.9)	0.52
Gradient >30 mm Hg	4/10 (40%)	16/35 (46%)	0.9 (0.3-3.2)	0.85
Nonsustained VT in Holter ECG	8/10 (80%)	21/35 (60%)	2.7 (0.6-12.8)	0.21
Abnormal blood pressure response	1/6 (17%)	24/31 (77%)	0.1 (0.0-1.1)	0.06
Number of risk factors	3.4 $\pm$ 1.1	2.8 $\pm$ 0.8	1.7 (0.9-3.3)	0.13

*RR indicates relative risk; CI, confidence interval; SD, sudden death; VT, ventricular tachycardia.

therapy; RR=1.68; 95% CI, 0.86-3.28; $P=.133$). The figure shows the Kaplan-Meier survival curves for patients with ICDs indicated for primary and secondary prevention (log rank test; $P=.004$).

Inappropriate discharges were observed in 13 patients (26.6%) and were due to either sinus tachycardia (8 cases), rapid atrial fibrillation (3 cases), or T-wave oversensing (2 cases). One patient presented endocarditis caused by infection of the ICD that necessitated removal of the device (Table 1).

DISCUSSION

This study is the first in which a large patient series has been used to analyze the indications and results of ICD implantation in a Spanish population with HCM. It represents one of the largest studies of the use of ICDs in HCM published to date.^{7,15-17}

No controlled studies have demonstrated the effectiveness of ICD implantation in patients with HCM and experience is based on results from small patient series, mainly treated for the purpose of secondary prevention. Despite the existence of a complete risk stratification based on known risk indicators, the precise identification of at-risk individuals and indications for ICD implantation are sometimes complicated.¹⁸ In the present study, the proportion of patients with an ICD is low (6.2%) but nevertheless similar to the majority of published studies.^{15,16,19,20} However, the percentage varies somewhat (3.6% to 11.9%) between the 3 hospitals involved in the study. Although this variation may indicate a lack of agreement regarding criteria for implantation, it could also reflect differences between the populations associated with each hospital. An inverse relationship was observed between the percentage of patients who received an ICD and the number of patients assessed in each hospital. It is possible that initially those patients referred to a subspecialty clinic are at higher risk or present greater complexity and that as the number increases the degree of selection of the population

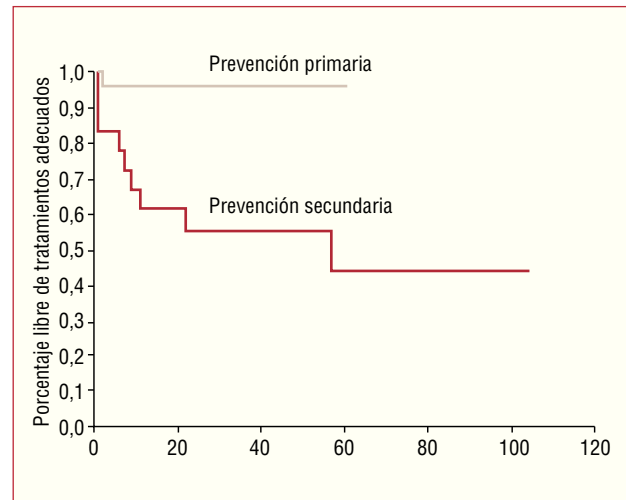


Figure. Curve for survival free from a first appropriate ICD discharge. Differences between primary and secondary prevention (log rank test; $P=.004$).

decreases. This possibility is supported by the observation that the mean number of risk factors associated with patients with an ICD was similar in all 3 hospitals. Various studies have demonstrated that the overall prognosis of HCM in unselected populations is benign.²¹⁻²³

In the present study, the indication for ICD implantation was secondary prevention in 40% of cases and primary prevention in 60%. Table 2 shows that the number of risk factors per patient was essentially the same in all 3 hospitals (a mean of 3 factors per patient). Various authors have recommended ICD implantation for primary prevention in patients with HCM and 2 or more risk factors for SD, and even in some patients with only a single risk factor.^{3,24} According to those recommendations, the criteria used in this study were correct but could be considered slightly restrictive. Some authors favor a slightly less restrictive position regarding the use of ICDs.⁸

The main risk factor that was associated with the occurrence of appropriate discharges was an individual history of aborted SD or sustained VT. Therefore, these results confirm the effectiveness of ICD implantation for secondary prevention in patients with HCM.^{7,25} The annual rate of appropriate discharges associated with secondary prevention was 11.1% and for primary prevention it was 1.6%, figures that are similar to those published previously for secondary prevention⁷ but are probably low for primary prevention.^{7,15,16} One of the limitations of studies addressing the benefit of ICD implantation for primary prevention of SD in HCM is the limited number of patients and the short follow-up times. It must be taken into account that although the annual rate of appropriate discharges is low, most patients who receive an ICD for HCM are young and have a prolonged life expectancy if SD does not occur. Nevertheless, it is important to correctly identify candidates, since the treatment is not without complications.

Paradoxically, the patients who had presented aborted SD or sustained VT displayed fewer risk factors than those with an ICD implanted for primary prevention (1.94 ± 1.18 vs 2.96 ± 0.84 ; $P = .002$). In fact, the main risk factor that was associated with the occurrence of appropriate discharges was an individual history of aborted SD or sustained VT. This finding reflects the difficulty of correctly identifying at-risk patients and undertaking effective primary prevention in HCM. The frequency and severity of the risk factors varies according to the population, age group, and probably the mutation responsible for the disease.^{2,3,26-30} Occasionally, the search for additional risk factors is not considered necessary or appropriate when a symptomatic episode of ventricular fibrillation or sustained VT has been presented. This is especially true in relation to exercise testing. An exercise test was performed in 11 out of 18 patients treated for the purpose of secondary prevention compared with 26 out of 27 patients in whom ICDs were implanted for primary prevention.

As shown in Table 3, a large proportion of the patients who required appropriate ICD discharges had some form of associated disease, particularly ventricular aneurysm, ischemic heart disease, or systolic dysfunction. It may be that the presence of those conditions affects the requirement for ICD discharges in patients with HCM. On the other hand, the presence of monomorphic sustained VT is an infrequent finding in HCM and is often associated with the presence of apical aneurysms in patients with midventricular obstruction.^{22,26,31} In our study, sustained VT was the most common indication for ICD implantation in the secondary prevention group and was also the most common trigger for appropriate ICD discharges. In 8 out of 10

appropriate discharges, the trigger was an episode of sustained VT. This finding complicates the assumption that appropriate ICD discharge can be equated with an episode of aborted SD, given that some patients can present well-tolerated episodes of sustained VT.

We observed a high incidence of inappropriate discharges, mainly triggered by sinus tachycardia and supraventricular arrhythmias. This finding supports the use of drug treatments for the control of heart rate (beta-blockers) and the prevention of arrhythmias (beta-blockers and amiodarone) in patients with an ICD. Beta-blockers are useful in young patients to prevent increased sinus rate, which could lead to inappropriate discharges. It is also important to control contributing conditions such as hyperthyroidism that can appear as a consequence of prior use of amiodarone, as has occurred in some of our patients. Although discharge triggered by recently initiated atrial fibrillation is, in theory, inappropriate, it can have beneficial effects. Atrial fibrillation has been shown to be associated with morbidity and mortality in patients with HCM.^{32,33} Inappropriate discharges caused by ICD dysfunction were also relatively frequent. For these reasons, when deciding to implant an ICD we must take into account that the rate of inappropriate discharges can be high, even with optimal adjustment of ICD parameters.

Further studies in a larger number of patients and with a longer follow-up period will be necessary to assess the benefit of using ICDs for primary prevention in patients with HCM. Continued evaluation of new risk indicators for SD is therefore necessary. The development of echocardiography and, particularly, cardiac magnetic resonance imaging with contrast agents could provide useful information regarding certain tissue characteristics that appear to be associated with clinical deterioration.^{34,35} The use of molecular biological techniques and the discovery of mutations associated with high risk can be expected to improve the accuracy of identification of patients at high risk for SD.^{18,36,37}

CONCLUSIONS

In Spain, the percentage of HCM patients who have an ICD is small (6.2%) but similar to that reported by other groups. However, there is a marked variation in the use of ICDs in different hospitals within Spain. The percentage of patients with an ICD varies essentially according to the degree of selection in the corresponding population. The incidence of appropriate discharges associated with patients receiving an ICD for secondary prevention is high (50% in 3 years). Longer follow-up will be necessary to assess the impact of this treatment in primary prevention.

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