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Letter to the Editor

Transatlantic differences in assessment of risk of sudden cardiac death in patients with hypertrophic cardiomyopathy

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Hypertrophic cardiomyopathy (HCM) is defined by the presence of increased ventricular wall thickness or mass in the absence of abnormal loading conditions (hypertension, valve disease) sufficient to cause the observed abnormality [1,2]. Most HCM patients are asymptomatic or mildly symptomatic, but some develop symptoms such as palpitations, dyspnoea, angina or syncope [3]. An infrequent presentation of the disease is sudden cardiac death (SCD), with an annual incidence of approximately 1% [1,2]. The only effective therapy of this most dangerous complication is an appropriate discharge of an implantable cardioverter-defibrillator (ICD). Based on two decades of clinical investigation, risk stratification criteria have been developed [1,2]. However, according to recently published guidelines, there are marked differences between the assessment of risk in primary prevention of SCD in Europe and North America.

The primary prevention of SCD, published in 2011 in the American (American College of Cardiology/American Heart Association; ACCF/AHA) guidelines for the diagnosis and treatment of HCM, is based on the identification of five binary risk factors: an unexplained syncope (Class IIa), family history of premature sudden death (Class IIa), left ventricular hypertrophy ≥30 mm (Class IIa), documented non-sustained ventricular tachycardia on ambulatory ECG monitoring (Class IIb) and an increase of the systolic blood pressure by ≤20 mm Hg during exercise stress testing (Class IIb). When ≥1 risk factors are present, ICD implantation is recommended [1].

On the contrary, identification of high-risk patients in the recently published European Society of Cardiology (ESC) guidelines is based on individualized assessment of the probability of SCD over the next five years and ICD implantation is indicated if the risk is ≥6% (Class IIa) or ≥4 and <6% (Class IIb). Several new risk factors were included in the ESC risk stratification (age of the patient, left atrial diameter and outflow pressure gradient), while one risk factor (blood pressure response during exercise) was excluded [2].

We have retrospectively evaluated 133 consecutive HCM patients (48.1 ± 15.8 years, 45% females) diagnosed in our center in whom all potential risk factors according to ACCF/AHA (Table 1) and ESC risk stratifications were available. Patients with ICD implanted in secondary prevention of SCD (Class I) have not been included. Mean duration of follow-up was 5.5 ± 2.2 years (median 5.5 years; range 2–100 months, 677 patient-years). Nine patients (6.8%) underwent ICD implantation according to the National HCM Guidelines [4] during the study period. Five patients (3.8%) died (two of pulmonary carcinoma, two of stroke and one soon after surgery). A total of three patients (2.3%) experienced one appropriate ICD discharge (the annual risk of sudden mortality event was 0.55%). A comparison of risk stratification according to ESC and ACCF/AHA guidelines in these three patients is presented in Table 2.

Based on risk stratification, 8 vs. 29 patients (6% vs. 22%; $p < 0.01$) would undergo ICD implantation according to ESC and ACCF/AHA guidelines, respectively, when the class of recommendation IIa was considered. Similarly, a significant difference – 26 vs. 56 patients (19.7% vs. 42.4%; $p < 0.01$) – was found when we included patients in the classes of recommendations IIa and IIb.

The existence of the two different prediction models of SCD is somewhat confusing. However, the issue is not only the mere existence of the two different stratification systems; more importantly, the models have markedly different results when applied to the same HCM cohort. The American system, which has been used for many years, recommends ICD implantation for a wider spectrum of HCM patients. On the contrary, the European prediction model offers a more individual prognostic evaluation, including the assessment of the annual risk of SCD. However, we have to bear in mind that this model needs to be independently validated in the future.

In summary, this is the first analysis dealing with the long-term outcome of HCM patients, considering both existing systems of prediction of SCD. The risk of SCD in an HCM tertiary center registry is less than 1% per year. Two systems evaluating the risk of SCD and determining ICD implantations led to extremely different results.

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Table 1
Incidence of major risk factors according to ACCF/AHA guidelines.

Risk factor	Patients
ACCF/AHA guidelines	%
FH	9.1
Syncope	10.6
IVS >30 mm	4.5
NsVT	9.9
Inadequate BP increase	15.9

FH — family history of sudden death, IVS — interventricular septum, NsVT — non-sustained ventricular tachycardia, inadequate BP increase — an increase of the systolic blood pressure by ≤ 20 mm Hg during exercise stress testing.

Table 2
Risk factors according to ACCF/AHA guidelines and estimated 5-year risk according to ESC guidelines in patients with appropriate ICD discharge.

	ESC	ACCF/AHA
	5-year risk of sudden death (%)	Number of risk factors
1.	9.64	2
2.	13.7	3
3.	4.46	2

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

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