

Effectiveness of contemporary risk stratification in patients with hypertrophic cardiomyopathy: a pilot study

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Objectives

- To determine the incidence of sudden cardiac death (SCD) in hypertrophic cardiomyopathy (HCM) patients
- To determine the effectiveness of contemporary stratification systems (ESC HCM SCD risk score, ACC/AHA risk factors)

Methods and characteristics

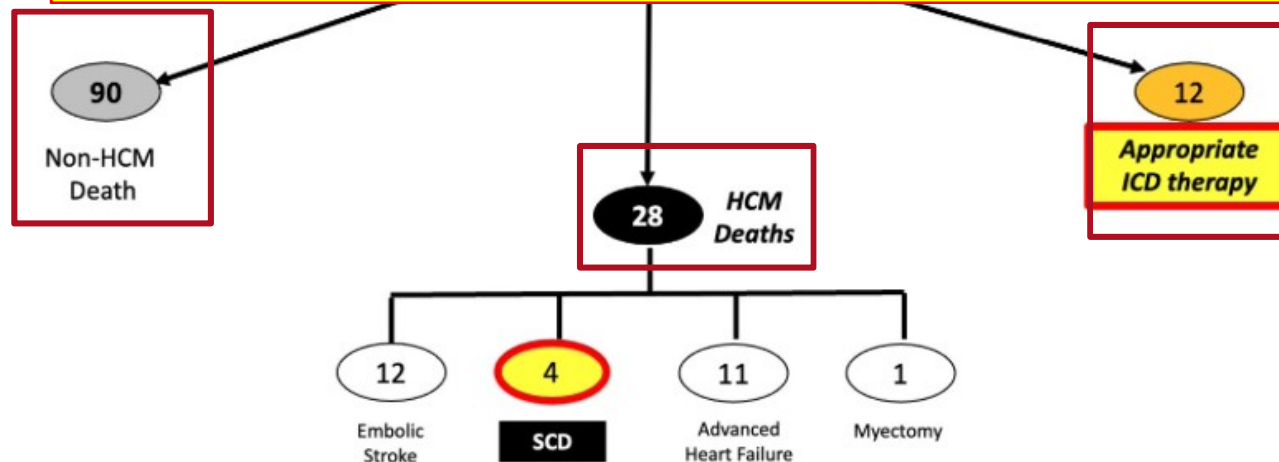
- **505 patients risk-stratified, mean follow-up 10 ± 8 years**
- **71 (14%) implanted with an ICD**
 - Risk-stratified aged 39 ± 17 years, 26 (35%) women
 - Maximal left ventricular wall thickness (MLWT) 23 ± 7 mm
 - Left atrial (LA) dimension 46 ± 7 mm

AEs within long-term follow-up (10 ± 8 years)

76 % died from comorbidities

All-cause mortality $\approx 2 \%$ /year

HCM-related mortality $\approx 0,5 \%$ /year



Appropriate ICD therapy

- Average age at ICD therapy 46 ± 19 years
- **≈ 7 years to first ICD therapy**
- MLWT 25 ± 6 mm
- LA 47 ± 7 mm
- **No patients with ICDs died from SCD**

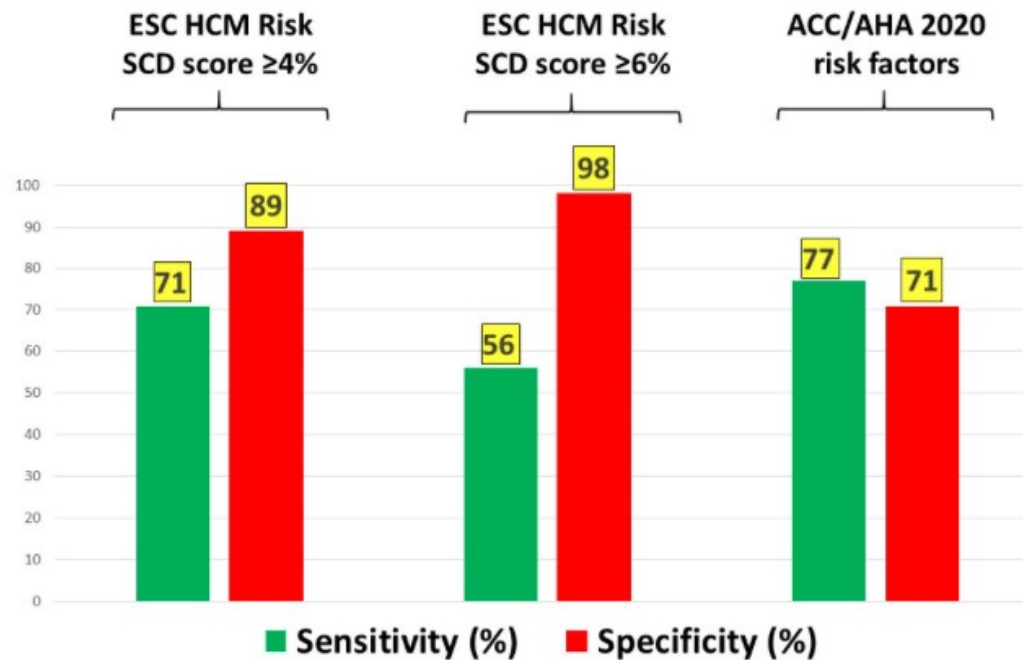
ICD complications

- Five (7%) patients suffered from ICD complications
 - 4 (6%) inappropriate shock
 - one patient experienced infective endocarditis requiring system extraction

SCD patients

- Mean age 59 ± 14 years (time of risk stratification)
- **Mean age at death 68 ± 10 years**
- All males
- **HCM-SCD mortality $\approx 0,07$ %/year**
- LA 49 ± 2 mm
- **Mean ESC score $2.0 \pm 0.9\%$**
- **3 patients (75%) had no risk factors present according to ACC/AHA**

Comparison of risk stratification methods



HCM Risk-SCD score $\geq 6\%$

- sensitivity 56%
(95% CI 21-86)
- specificity 98%
(95% CI 96-99)

ACC/AHA individual RF

- sensitivity 77%
(95% CI 46-95)
- specificity 71%
(95% CI 67-75)

Conclusions

- SCD is rare in HCM patients managed at an expert center with current risk stratification.
- The ESC HCM Risk-SCD score is less sensitive but more specific than the ACC/AHA approach.
- The majority of contemporary treated HCM patients die from comorbidities and not HCM-related causes.