

Cardiompatia ipertrofica: Stratificazione del rischio di morte improvvisa ed indicazione all'ICD



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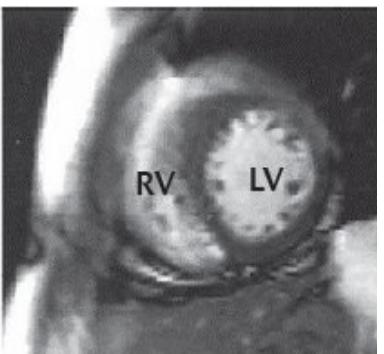
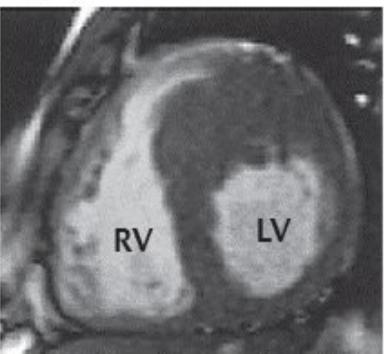
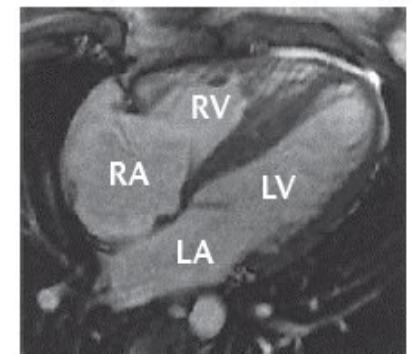
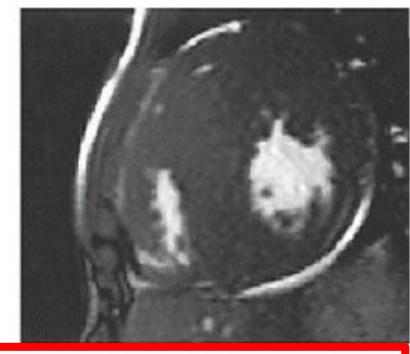
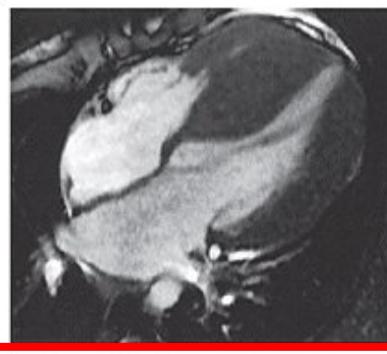
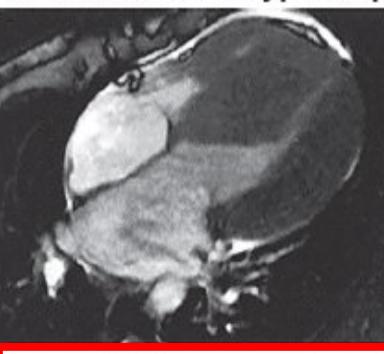
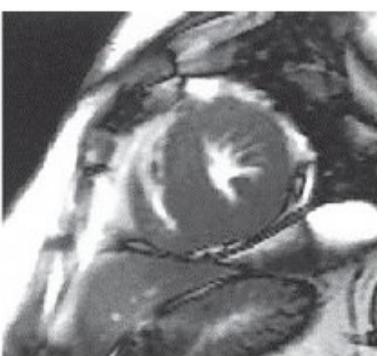
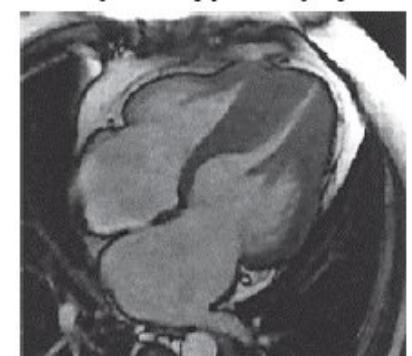
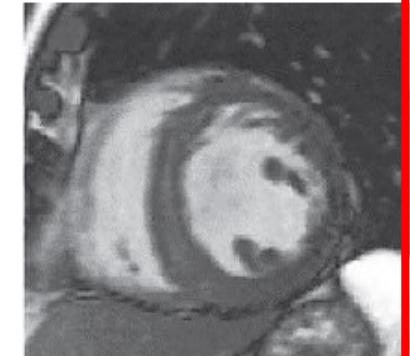
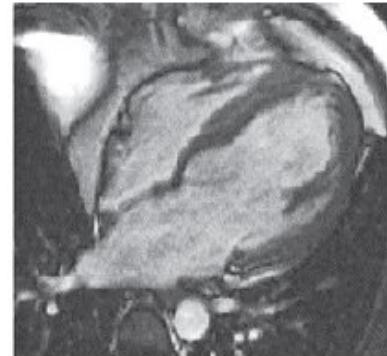
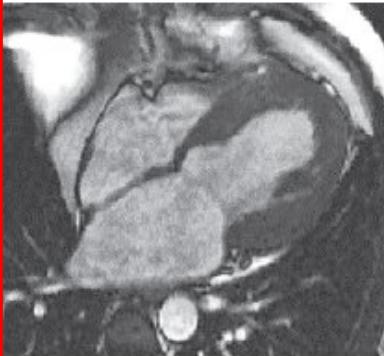
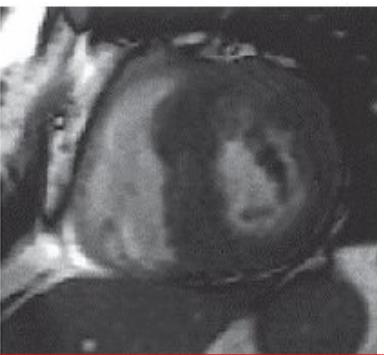
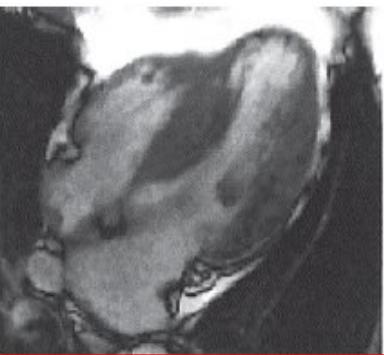
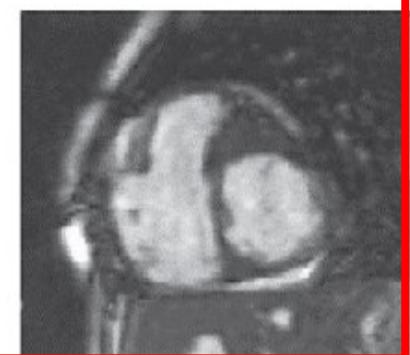
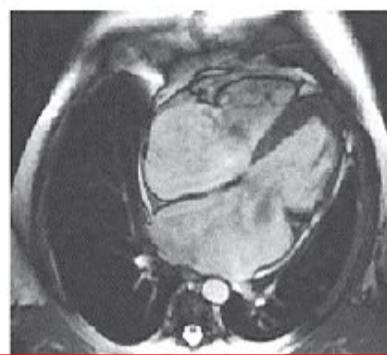
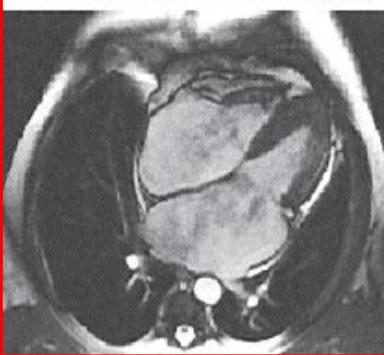
Cardiomielopatia ipertrofica: Stratificazione del rischio di morte improvvisa Quantificazione del rischio ed indicazione all'ICD



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1. DIAGNOSI Spessore di parete in qualsiasi segmento del VS

Adulti ≥ 15 mmfamiliari ≥ 13 mmBambini ≥ 2 SD**a** Asymmetrical septal hypertrophy**d** Biventricular hypertrophic**b** Apical hypertrophy**e** 'End-stage' dilatation**c** Midcavity obstruction**f** Restrictive cardiomyopathy

Late Sodium Current Inhibition Reverses Electromechanical Dysfunction in Human Hypertrophic Cardiomyopathy

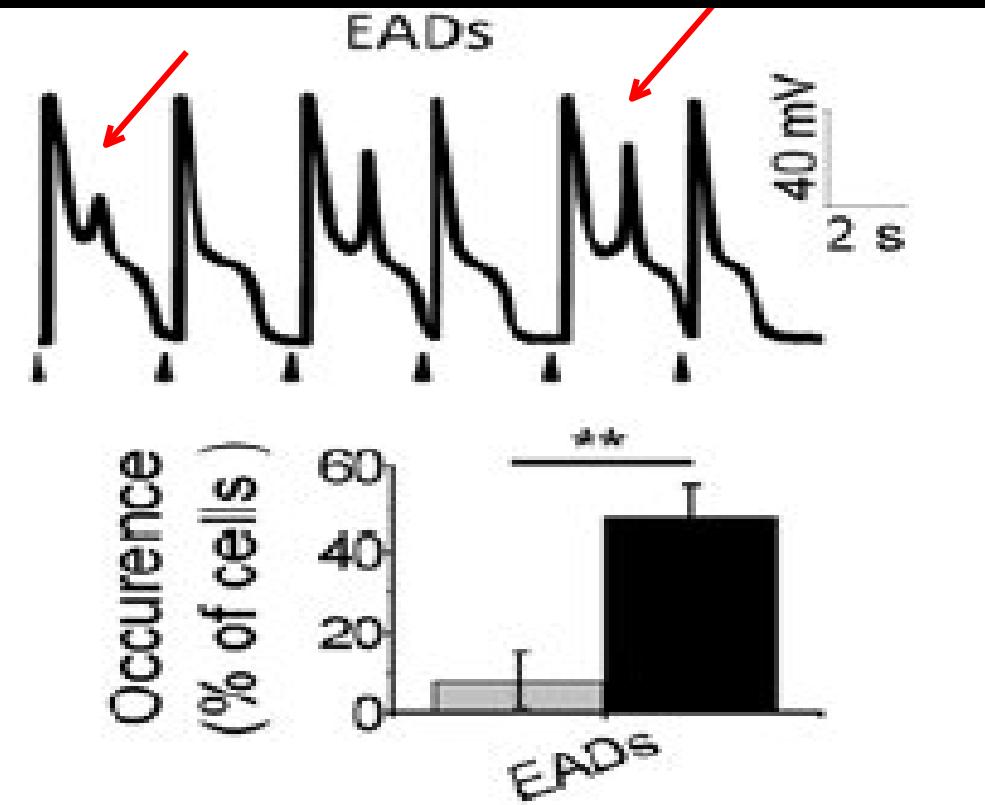
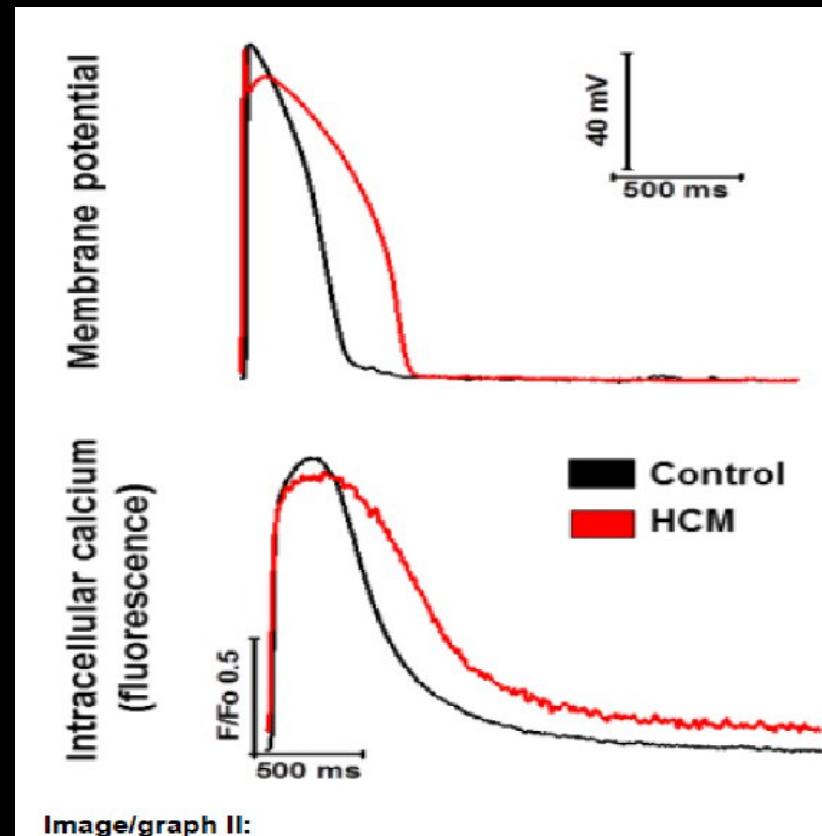
Raffaele Coppini, MD, PhD*; Cecilia Ferrantini, MD, PhD*; Lina Yao, PhD; Peidong Fan, PhD; Martina Del Lungo, PhD; Francesca Stillitano, PhD; Laura Sartiani, PhD; Benedetta Tosi, MD; Silvia Suffredini, PhD; Chiara Tesi, PhD; Magdi Yacoub, MD; Iacopo Olivotto, MD; Luiz Belardinelli, MD; Corrado Poggesi, MD; Elisabetta Cerbai, PhD; Alessandro Mugelli, MD

Circulation, 2012

ELETTROFISIOLOGIA CELLULARE

cardomiociti prelevati dopo miectomia

- la ripolarizzazione
- le postdepolarizzazioni precoci (EAD) sono molto frequenti



Cause di Morte improvvisa nella CMI

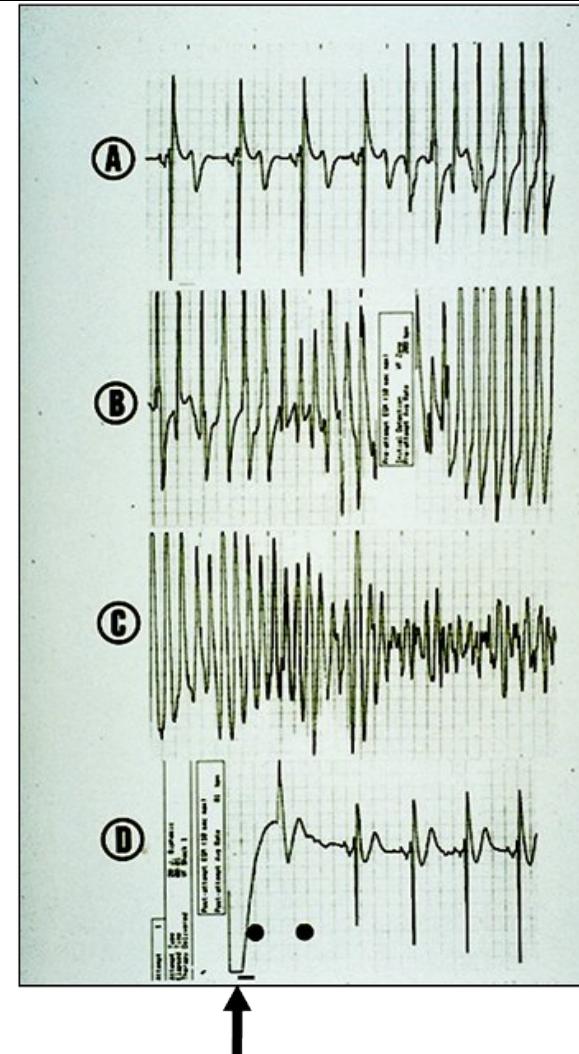
- FV
- TVS polimorfa → FV
preceduta da:
 - TVS monomorfa
 - Tachicardia sinusale
 - Fibrillazione Atriale
 - BSA/BAV eccezionalmente

35 y/o – Brother
(Sudden death)

5 y: {
36 y/o – ICD
40 y/o – Generator replaced

9 y: {
41 y/o – Appropriate shock #1
50 y/o – Appropriate shock #2

52 y/o – Present

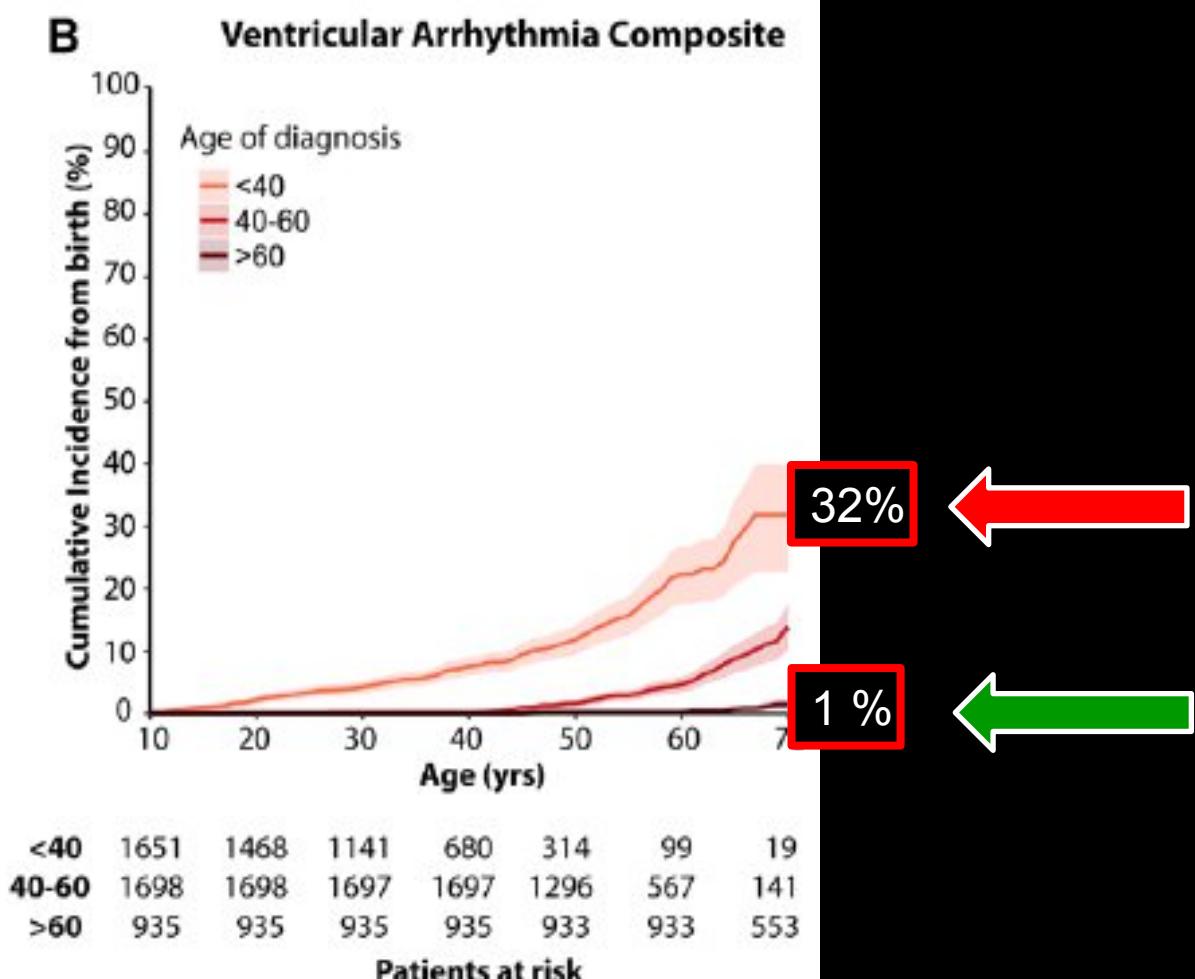


SOTTOGRUPPI DI PZ CON CMI E RISCHIO DI MORTE IMPROVVISA RIDOTTO

- ECG NORMALE
- POSTMIECTOMIA TEVS
- ETA' > 60 a.

Genotype and Lifetime Burden of Disease in Hypertrophic Cardiomyopathy

Insights From the Sarcomeric Human Cardiomyopathy Registry
(SHaRe) Circulation 2018



SOTTOGRUPPI DI PZ CON CMI

E RISCHIO DI MORTE IMPROVVISA AUMENTATO

E

QUANTIFICAZIONE DEL RISCHIO

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC)

<http://doc2do.com/hcm/webHCM.html>

An International External Validation Study of the 2014 European Society of Cardiology Guideline on Sudden Cardiac Death Prevention in Hypertrophic Cardiomyopathy (Evidence from HCM)

Constantinos O'Mahony, Fatima Jichi, Steve R. Ommen, Imke Christiaans, Eloisa Arbustini, Pablo Garcia-Pavia, Franco Cecchi, Iacopo Olivotto, Hiroaki Kitaoka, Israel Gotsman, Gerald Carr-White, Jens Møgensen, Loizos Antoniades, Saidi Mohiddin, Mathew S. Maurer, Hak Chiaw Tang, Jeffrey B. Geske, Konstantinos C. Siontis, Karim Mahmoud, Alexa Vermeer, Arthur Wilde, Valentina Favalli, Oliver Guttmann, María Gallego-Delgado, Fernando Domínguez, Ilaria Tanini, Toru Kubo, Andre Keren, Teofila Bueser, Sarah Waters, Issa F. Issa, James Malcolmson, Thomas Burns, Neha Sekhri, Christopher W. Hoeger, Rumana Z. Omar and Perry M. Elliott

HCM Risk-SCD Calculator

Age Years

Maximum LV wall thickness mm Transthoracic Echocardiographic measurement

Left atrial size mm Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation

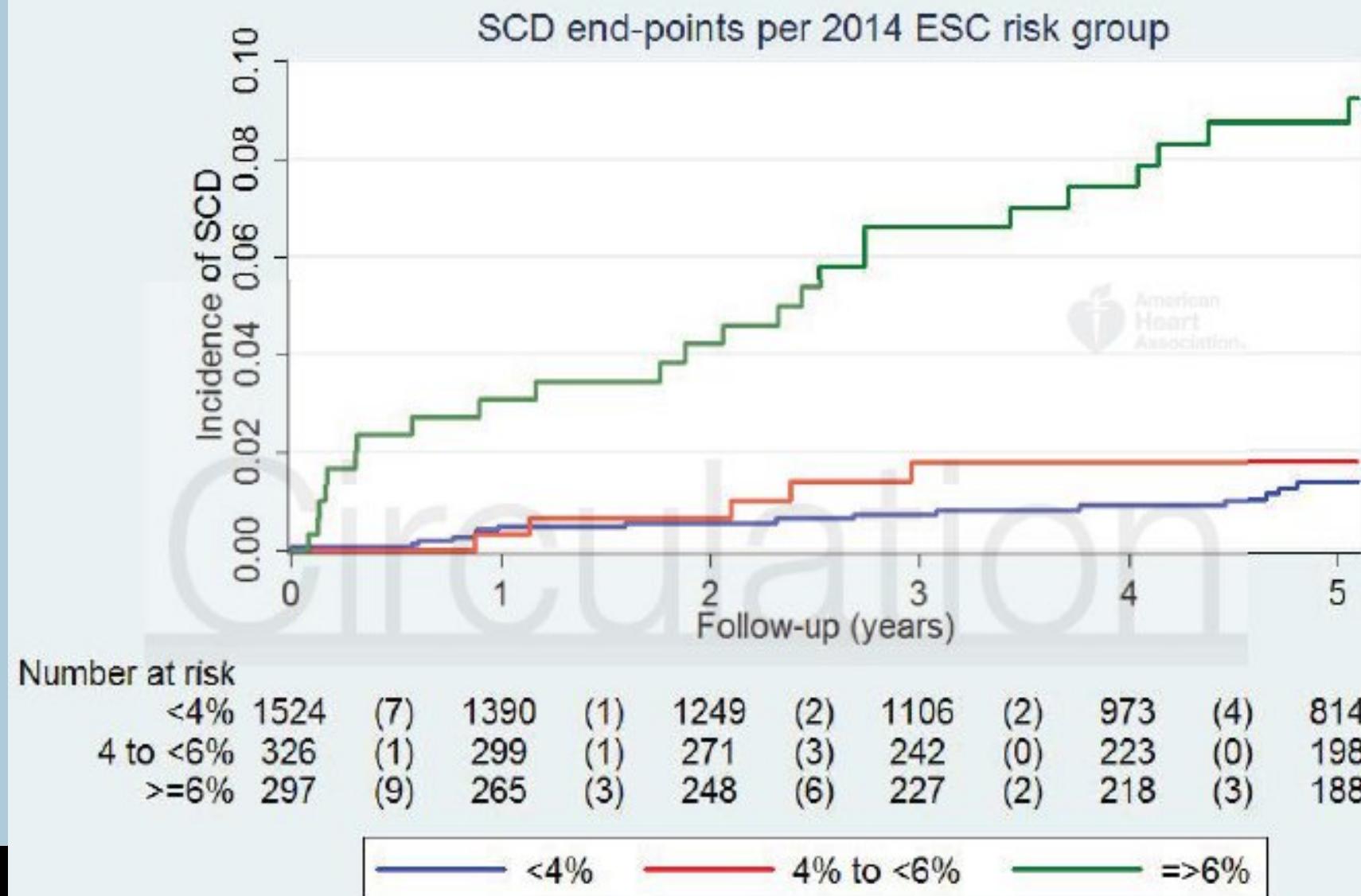
Max LVOT gradient mmHg The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernoulli equation: Gradient = $4V^2$, where V is the peak aortic outflow velocity

Family History of SCD No Yes History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).

Non-sustained VT No Yes 3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.

Unexplained syncope No Yes History of unexplained syncope at or prior to evaluation.

Risk of SCD at 5 years (%):



AHA-ACC HCM Guidelines 2020 SCD Risk Assessment & ICD Recommendations

Figure 3. ICD Patient

Selection

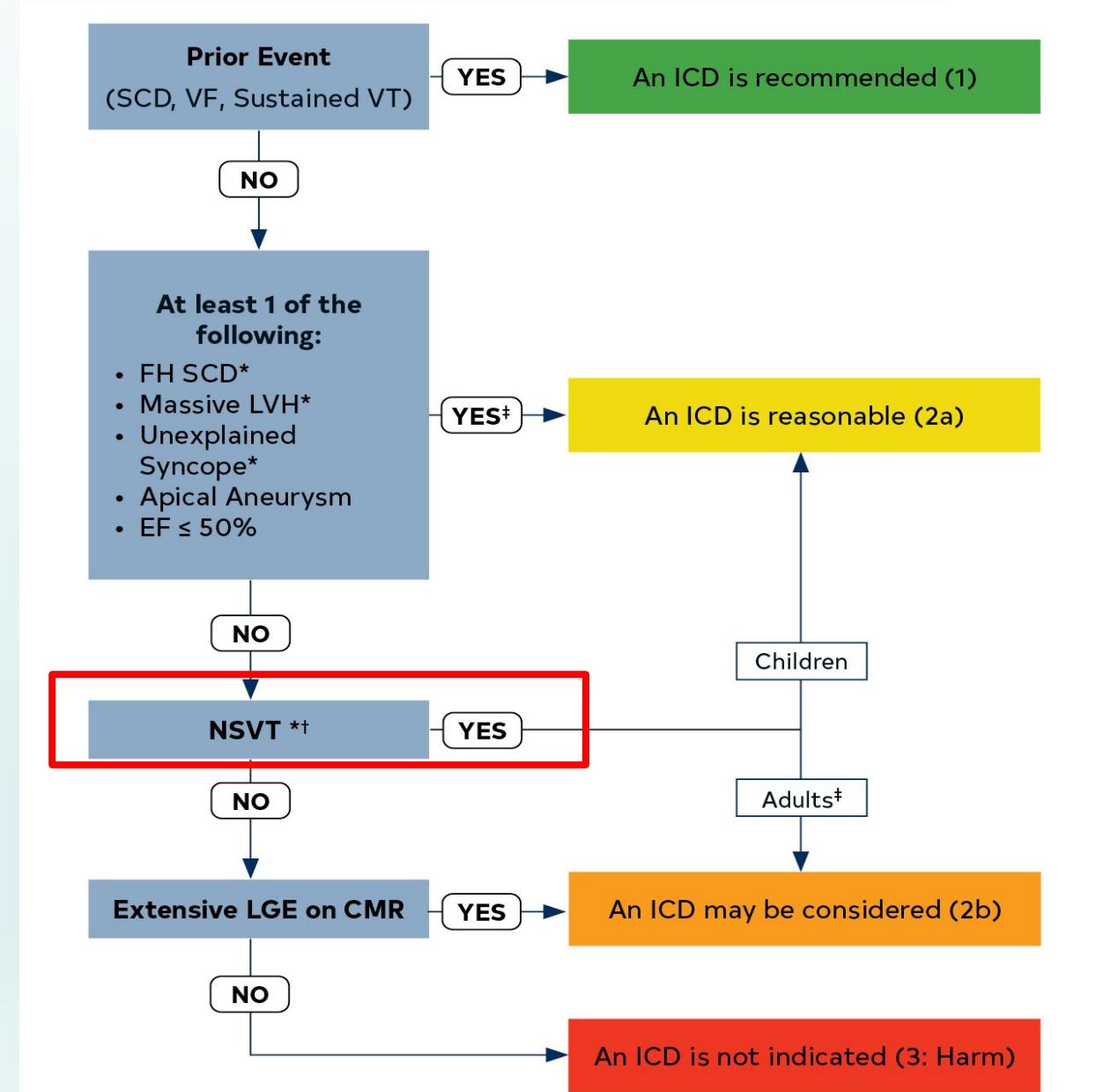
Colors correspond to the Class of Recommendation in Table 2.

*ICD decisions in pediatric patients with HCM are based on ≥ 1 of these major risk factors: family history of HCM SCD, NSVT on ambulatory monitor, massive LVH, and unexplained syncope.

†In patients >16 years of age, 5-year risk estimates can be considered to fully inform patients during shared decision-making discussions.

‡It would seem most appropriate to place greater weight on frequent, longer, and faster runs of NSVT.

CMR indicates cardiovascular magnetic resonance; EF, ejection fraction; FH, family history; HCM, hypertrophic cardiomyopathy; ICD, implantable cardioverter-defibrillator; LGE, late gadolinium enhancement; LVH, left ventricular hypertrophy; NSVT, nonsustained ventricular tachycardia; SCD, sudden cardiac death; VF, ventricular fibrillation; and VT, ventricular tachycardia.



2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Developed by the task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC)

European Heart Journal (2022) 00, 1–130

Diagnostic evaluation and general recommendations

CMR with LGE is recommended in HCM patients for diagnostic work-up.^{716–718}

I	B
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Genetic counselling and testing are recommended in HCM patients.^{721–725}

I	B
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Participation in high-intensity exercise may be considered for asymptomatic adult HCM patients without risk markers.⁷³³

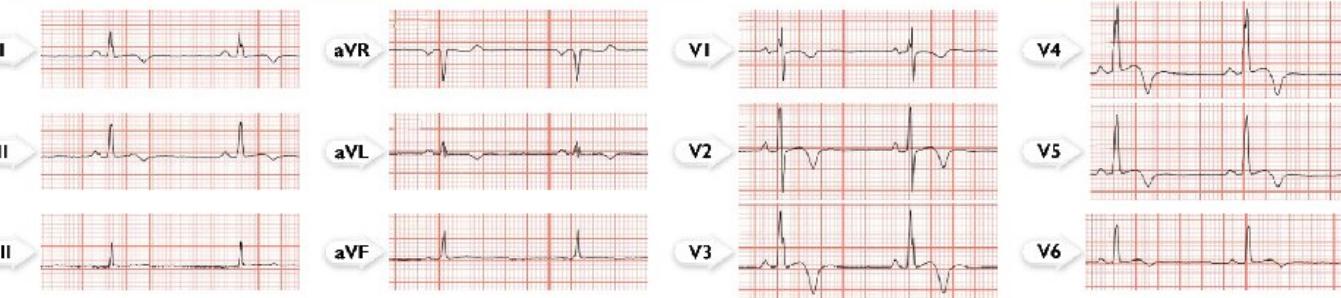
IIb	C
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2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

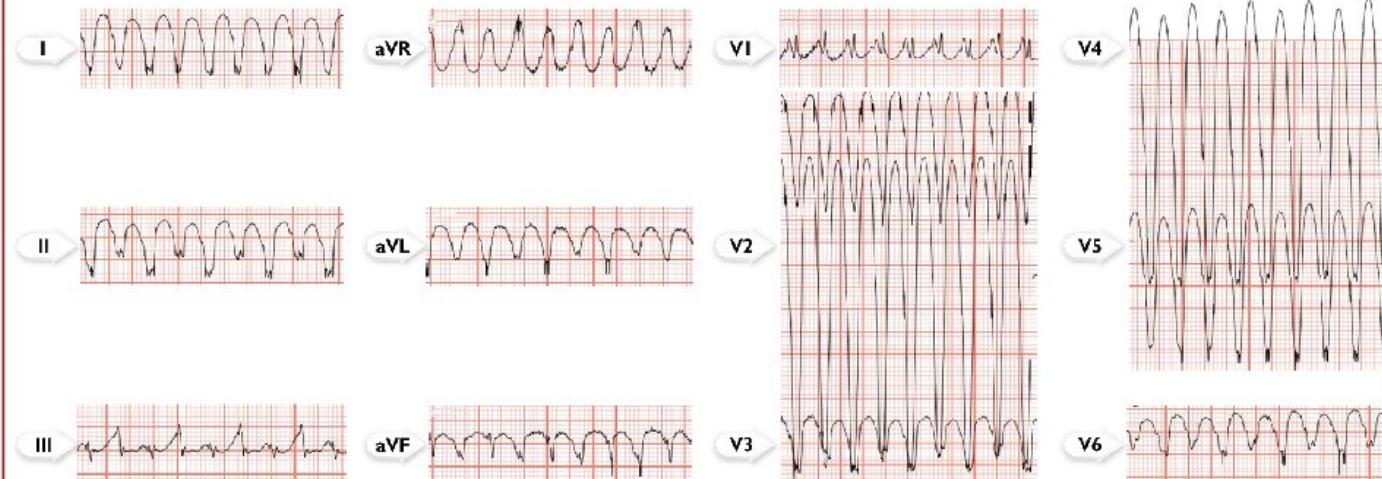
Developed by the task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC)

European Heart Journal (2022) 00, 1–130

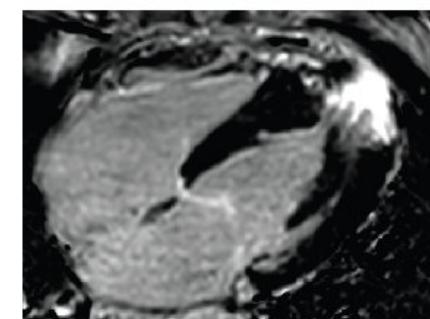
ECG sinus rhythm – Negative T waves V2-V6 and inferior leads



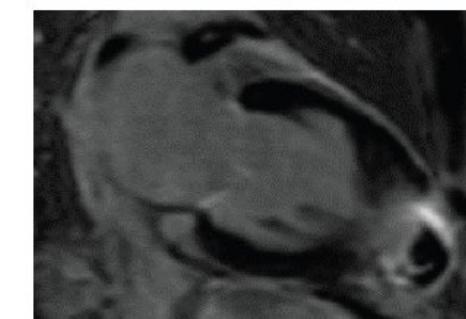
ECG VT – RBBB-like, superior axis (apical LV origin)



CMR – Apical aneurysm with LGE



4-chamber



2-chamber

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

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European Heart Journal (2022) 00, 1–130

Risk stratification and primary prevention of SCD

It is recommended that the 5-year risk of SCD is assessed at first evaluation and at 1–3-year intervals, or when there is a change in clinical status.

I

C

ICD implantation should be considered in patients aged 16 years or more with an estimated 5-year risk of SD $\geq 6\%$.^{c,85,728,729}

IIa

B

ICD implantation should be considered in HCM patients aged 16 years or more with an intermediate 5-year risk of SCD (≥ 4 to $< 6\%$)^c and with (a) significant LGE at CMR (usually $\geq 15\%$ of LV mass); or (b) LVEF $< 50\%$; or (c) abnormal blood pressure response during exercise test^d; or (d) LV apical aneurysm; or (e) presence of sarcomeric pathogenic mutation.^{716,717,722,736–739}

IIa

B

In children less than 16 years of age with HCM and an estimated 5-year risk of SD $\geq 6\%$ (based on HCM Risk-Kids score^e), ICD implantation should be considered.^{84,742}

IIa

B

ICD implantation may be considered in HCM patients aged 16 years or more with an estimated 5-year risk of SCD of ≥ 4 to $< 6\%$.^{c,85,728,729}

IIb

B

ICD implantation may be considered in HCM patients aged 16 years or more with a low estimated 5-year risk of SCD ($< 4\%$)^c and with (a) significant LGE at CMR (usually $\geq 15\%$ of LV mass); or (b) LVEF $< 50\%$; or (c) LV apical aneurysm.^{716,717,722,736–739}

IIb

B

AHA-ACC HCM Guidelines 2020 SCD Risk Assessment & ICD Recommendations

Referral to multidisciplinary HCM centers

Although the primary cardiology team can initiate evaluation, treatment, and longitudinal care, **referral to multidisciplinary HCM centers** with graduated levels of expertise **can be important to optimizing care** for patients with HCM.

Challenging treatment decisions —where reasonable alternatives exist, where the strength of recommendation is weak (e.g., **any Class 2b decision**) or is particularly nuanced, and for invasive procedures that are specific to patients with HCM—**represent crucial opportunities to refer patients** to these HCM centers.

AHA-ACC HCM Guidelines 2020 SCD Risk Assessment & ICD Recommendations

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4. SHARED DECISION-MAKING

Recommendation for Shared Decision-Making

Referenced studies that support the recommendation are summarized in [Online Data Supplement 1](#).

COR	LOE	Recommendation
1	B-NR	<ol style="list-style-type: none">1. For patients with HCM or at risk for HCM, shared decision-making is recommended in developing a plan of care (including but not limited to decisions regarding genetic evaluation, activity, lifestyle, and therapy choices) that includes a full disclosure of the risks, benefits, and anticipated outcomes of all options, as well the opportunity for the patient to express their goals and concerns.¹⁻⁶



CONCLUSIONI

La Stratificazione del rischio (valutazione individuale del rischio)
NON PUÒ ESSERE precisa:
...non può dirci quale paziente avrà un arresto cardiaco !!

Non possiamo basarci SOLO su un singolo fattore di rischio
o su un calcolo probabilistico (che comunque aiuta) !!

**La valutazione del rischio individuale (pur imprecisa)
aiuta a prendere decisioni INSIEME al paziente**



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GRAZIE !



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