ESC European Society of Cardiology

European Heart Journal - Case Reports (2023) **7**, 1–6
https://doi.org/10.1093/ehjcr/ytad010

CASE REPORT
Other

Electrical storm in hypertrophic cardiomyopathy: Cardiac magnetic resonance and sudden cardiac death risk stratification: a case report

Pedro Garcia Brás *, Sílvia Aguiar Rosa, Guilherme Portugal, and Mário Martins Oliveira

Department of Cardiology, Hospital de Santa Marta, Centro Hospitalar Universitário de Lisboa Central, Lisbon, Portugal

Received 29 May 2022; first decision 5 July 2022; accepted 5 January 2023; online publish-ahead-of-print 7 January 2023

Background Risk stratification for sudden cardiac death (SCD) is a key factor in the management of patients with hypertrophic cardiomyopathy (HCM). Cardiac magnetic resonance (CMR) has a unique role in the evaluation of HCM and offers superior diagnostic and prognostic information to assess the indication for a prophylactic implantable cardioverter-defibrillator (ICD).

Case summary A 39-year-old patient with non-obstructive HCM with a low ESC HCM Risk-SCD score underwent a CMR revealing a left ventricular apical aneurysm and extensive late gadolinium enhancement; a prophylactic ICD was thus implanted. A month later, the patient was admitted in refractory electrical storm with over 50 appropriate ICD shocks due to sustained ventricular tachycardia. Despite anti-arrhythmic therapy and mechanical ventilation, the evolution was unfavourable with haemodynamic instability; veno-arterial extracorporeal membrane oxygenation was implanted. The patient was submitted to CMR-guided epicardial VT catheter ablation with complications of LV thrombus and severe pericardial effusion.

Discussion This case details the complex risk stratification for SCD in patients with HCM, highlighting the important role of CMR in the integrated approach to risk stratification.

Keywords Hypertrophic cardiomyopathy • Cardiac magnetic resonance • Sudden cardiac death • Ventricular tachycardia • Ventricular tachycardia ablation • Implantable cardioverter-defibrillator • case report

ESC Curriculum 2.3 Cardiac magnetic resonance • 5.10 Implantable cardioverter defibrillators • 6.5 Cardiomyopathy

Learning points

- Review of the integrated approach in risk stratification of sudden cardiac death in hypertrophic cardiomyopathy. Looking beyond the ESC HCM SCD risk score, in-depth look at the role of the cardiac magnetic resonance imaging, 12-lead ECG and genetic testing in indication for ICD implantation in primary prevention.
- Review of the management of electrical storm in the ICU, including veno-arterial extracorporeal membrane oxygenation and CMR-guided ventricular tachycardia catheter ablation.

* Corresponding author. Tel: +351 913367020. Email: pedrobras3@gmail.com
Handling Editor: Sadaf Raza
Peer-reviewers: Elizabeth Paratz; Giacomo Tini Melato
Compliance Editor: Oliver Ian Brown
Supplementary Material Editor: Nikesh Jathanna

Introduction

Risk stratification for sudden cardiac death (SCD) is a key factor in the management of patients with hypertrophic cardiomyopathy (HCM) because the implantation of a prophylactic implantable cardioverter-defibrillator (ICD) can prevent the dismal complication that is SCD.

Current recommendations including the *European Society of Cardiology* (ESC) HCM Risk-SCD score consider clinical characteristics, family history, and echocardiographic findings.[1] Cardiac magnetic resonance (CMR) has a unique role in the evaluation of HCM—it provides superior diagnostic and prognostic information such as the presence and extension of late gadolinium enhancement (LGE), left ventricular (LV) apical aneurysm, or systolic dysfunction. These metrics can be important in assessing the need for prophylactic ICD.[2,3]

We describe here a clinical case that illustrates the current recommendations for risk stratification of SCD in HCM including the role of the 12-lead ECG, echocardiography, genetic testing, and the importance of CMR. The complex management of a patient with HCM presenting with refractory electrical storm is also addressed.

Timeline

1 year prior to the first observation	Diagnosis of non-obstructive HCM after embolic stroke due to LV thrombus
Day 1	Cardiomyopathies Unit outpatient clinic first observation 12-lead ECG showing Q waves in leads V4-V6 and QRS fragmentation in the limb leads CMR study: maximum wall thickness of 30 mm, LV apical aneurysm and extensive LGE (29% of LV mass)
Day 5	Prophylactic ICD implantation
Day 32	Patient admitted in the ICU with refractory electrical storm, over 50 appropriate ICD shocks due to sustained VT. Anti-arrhythmic therapy, sedation, mechanical ventilation. VA-ECMO implantation
Day 36	VA-ECMO decannulation: acute thrombosis of the superficial and common right femoral arteries Emergent surgical thromboembolectomy
Day 38	Epicardial VT catheter ablation, guided by CMR imaging
Day 41	Severe pericardial effusion Limited response to anti-inflammatory drugs Started on corticosteroid therapy
Day 42	Echocardiography: two LV apical thrombi Anticoagulation with direct oral anticoagulant
Day 54	Discharged in sinus rhythm, with no visible pericardial effusion or LV thrombi
24-month follow-up	No VT recurrence, ICD therapies or pericardial effusion

Case presentation

We report a 39-year-old male patient with non-obstructive HCM with previous history of an embolic stroke due to LV thrombus under oral anticoagulation with rivaroxaban (refused therapy with a vitamin K antagonist). He had a remarkable family history (see Supplementary material online, *Figure S1*) of one brother with HCM diagnosis and two cousins with SCD at the age of 40. Genetic testing revealed no pathogenic mutations but a heterozygous mutation of the TTN gene and a homozygous mutation of the TRIM63 gene, both variants of uncertain significance.

Given the patient's family history, he was referred to the Cardiomyopathies Unit Outpatient clinic. He reported no previous episodes of syncope or other cardiovascular symptoms. The 12-lead electrocardiogram (*Figure 1*) showed QRS fragmentation in the limb leads and Q waves with ST-segment elevation in the inferior and lateral leads: the latter is suggestive of an LV apical aneurysm in patients with a previous LV thrombus and embolic stroke. CMR showed a maximum wall thickness (MWT) of 30 mm, an LV apical aneurysm, and extensive LGE (*Figure 2*, Supplementary material online, *Videos S1–S3*) totalling 29% of the LV mass. Holter monitoring for 24 h showed no premature ventricular contractions or non-sustained ventricular tachycardia (VT). The patient did not present LV outflow tract obstruction (peak gradient of 15 mmHg), and there was mild left atrium dilation (46 mm).

According to the ESC HCM Risk-SCD score,[1] the risk for SCD was low (risk at 5 years of 3%, i.e. ICD generally not indicated). While there was no family history of SCD in first-degree relatives, there was a family history of two cousins with SCD at the age of 40. Moreover, the patient's ECG revealed pathological Q waves with notched QS complexes and 2 mm ST-segment elevation in several leads. The CMR showed an apical aneurysm and extensive LGE indicating the presence of considerable myocardial fibrosis. After multidisciplinary discussion with the electrophysiology team—and considering the aforementioned CMR prognostic high-risk markers—the patient, understanding the risks and benefits of an ICD and its role in the prevention of SCD, consented to an urgent prophylactic ICD.

A month after ICD implantation, the patient was urgently transferred to the Intensive Care Unit with haemodynamic instability in refractory electrical storm with over 50 appropriate ICD shocks due to sustained VT. The patient's admission rhythm strips are presented in *Figure 3A* and show a sustained VT at 192 b.p.m. A chest X-ray was performed (*Figure 3B*) revealing no ICD generator or lead displacement. Admission transthoracic echocardiogram (see Supplementary material online, *Figure S2*, Supplementary material online, *Videos S4–S6*) showed LV hypertrophy with septal predominance (32 mm) consistent with the patient's background diagnosis. Electrolyte imbalance was ruled out and immediate anti-arrhythmic therapy was started with amiodarone (600 mg/50 mL at 4 cc/h) followed by esmolol (500 mg/50 mL at 250 µg/kg/min) and lidocaine (1 g/50 mL at 4 µg/min) perfusion, but without success. Patient sedation, tracheal intubation, and ICD-mediated overdrive pacing therapies (ramp and burst strategies with different cycle lengths) were also unsuccessful. The integrated effect of anti-arrhythmic therapy, sedation, and ICD internal cardioversion was successful in the initial control of the electrical storm, with conversion to sinus rhythm.

Despite an initial short period of sinus rhythm, the patient maintained long salvos of NSVT associated with hypotension, signs of impaired peripheral perfusion, and hyperlactatemia. There was progressive worsening of haemodynamic instability (further exacerbated by the sedative therapy) despite the aforementioned VT management strategies. There was a decision for veno-arterial extracorporeal membrane oxygenation (VA-ECMO) after discussion in a multidisciplinary Heart Team. The patient's evolution under ECMO support was favourable albeit with intermittent episodes of NSVT. Upon ECMO decannulation there was acute thrombosis of the superficial

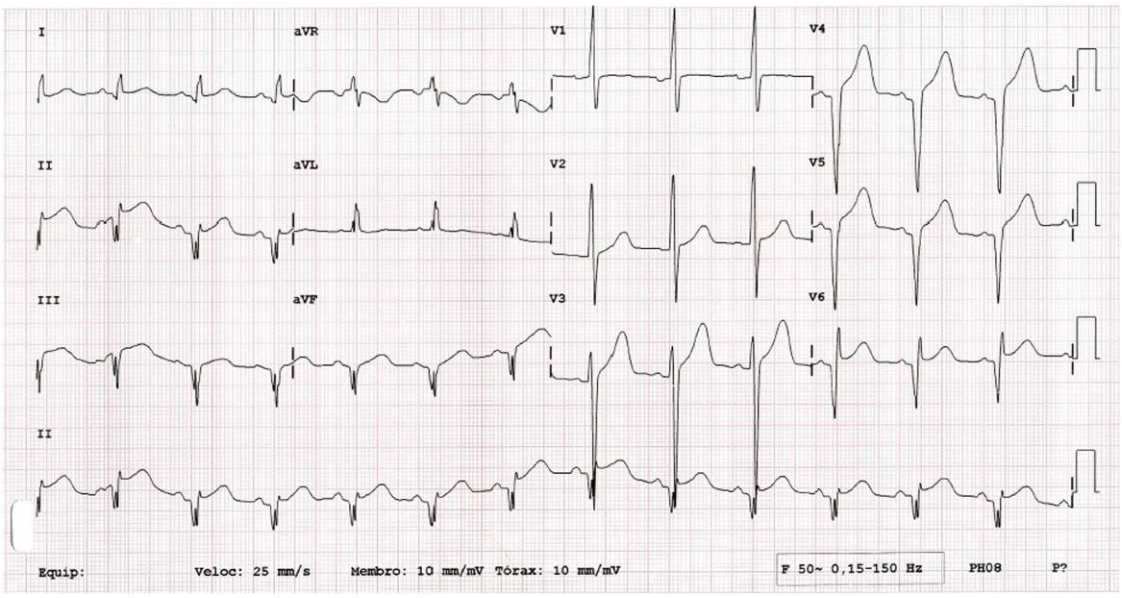

Figure 1 Outpatient 12-lead electrocardiogram. Sinus rhythm, 80 b.p.m., left axis deviation, Q waves and 2 mm ST-segment elevation in the inferior and lateral leads, and notched QS complexes in the lead limbs. The association of Q waves and ST-segment elevation is suggestive of LV apical aneurysm. The presence of fragmented QRS complexes reflects inhomogeneous activation of the ventricles and has a high predictive value for myocardial scar.

and common right femoral arteries, and an emergent surgical thromboembolectomy was performed.

Due to the aggressive arrhythmic presentation with a high risk of recurrence the patient was submitted to epicardial VT catheter ablation guided by CMR imaging. As previously described in patients with HCM and apical aneurysm,[4] electroanatomical mapping showed an extensive zone of apical necrosis in both ventricles that extended to the posterior region of the left ventricle (*Figure 4*). This area had very late potentials in sinus rhythm, and monomorphic VT was induced, with diastolic potentials observed in the scar region during the tachycardia. Radiofrequency (RF) energy application in this region interrupted the VT (see Supplementary material online, *Figure S3*). After additional RF applications, no more local abnormal ventricular activation electrograms were identified, with no late potentials and no local capture with 8 V. VT was non-inducible after ablation.

The patient developed severe pericardial effusion after VT catheter ablation (maximum of 27.6 mm diameter) with limited response to anti-inflammatory drugs and moderate pleural effusion (see Supplementary material online, *Figure S4*, Supplementary material online, *Videos S7* and *S8*). Corticosteroid therapy was then started. Bedside echocardiography re-evaluation indicated two LV apical thrombi (diameter of 10 × 7 and 14 × 7 mm—Supplementary material online, *Figure S4C*, Supplementary material online, *Video S9*). These were attributed to anticoagulation interruption due to the VT ablation procedure. The patient was started on direct oral anticoagulant and was discharged two weeks later with pericardial effusion and LV thrombi regression, on rivaroxaban, bisoprolol, diltiazem, amiodarone, furosemide, and prednisolone (tapering). In a 24-month follow-up there was no VT recurrence, ICD therapies, or pericardial effusion.

Discussion

SCD remains a major consequence of HCM, and the identification of the patients with HCM who are most likely to benefit from a prophylactic ICD remains challenging. There are several characteristics to consider in HCM such as the low positive predictive value for appropriate therapies and the possible complications including lead fracture, infection, and inappropriate therapies.[5]

The ESC HCM Risk-SCD score for stratification of SCD risk is a valuable tool in making the decision to implant a prophylactic ICD in HCM patients, who are often young and asymptomatic; however, this score may be falsely reassuring in selected cases of patients with low-risk scores—particularly in the setting of adverse LV remodelling. While originally shown to have a good discriminatory power,[6] several trials have reported a lower sensitivity of this model in different regions. Consequently, there is a risk of recognizing fewer high-risk patients.[3,7] Notably, in one trial, 64% of SCDs occurred in patients initially deemed low risk.[8] Another potential pitfall of this score is the fact that some variables such as NSVT, LV outflow tract gradient, and left atrial diameter are dynamic and may vary with the natural history of HCM. As previously described, the patient's ESC HCM SCD risk score[1] was low. In this case, a prophylactic ICD implantation would not generally be recommended (*Figure 5*).

Since 2011, the North American guidelines[9] have recommended prophylactic ICD implantation in HCM patients with a previous episode of SCD, ventricular fibrillation, or sustained VT. A prophylactic ICD is reasonable in the presence of major risk factors for SCD such as family history of SCD, history of suspected cardiac syncope, or massive LV hypertrophy (≥30 mm). In accordance with the North American guidelines, a prophylactic ICD should be considered (IIa recommendation) in our patient described here with massive LV hypertrophy (MWT 30 mm).

There is increasing evidence of the underperformance of traditional risk factors for prediction of SCD. Concurrently, there is a pressing need for evaluation of novel risk markers that assess different mechanisms of HCM leading to SCD risk.[10] The presence of areas of disorganized architecture and myocardial fibrosis are a hallmark of HCM and predispose to re-entry circuits that may lead to life-threatening arrhythmias. Tissue characterization with cardiovascular magnetic resonance (CMR) can assess the location and extension of these pathological

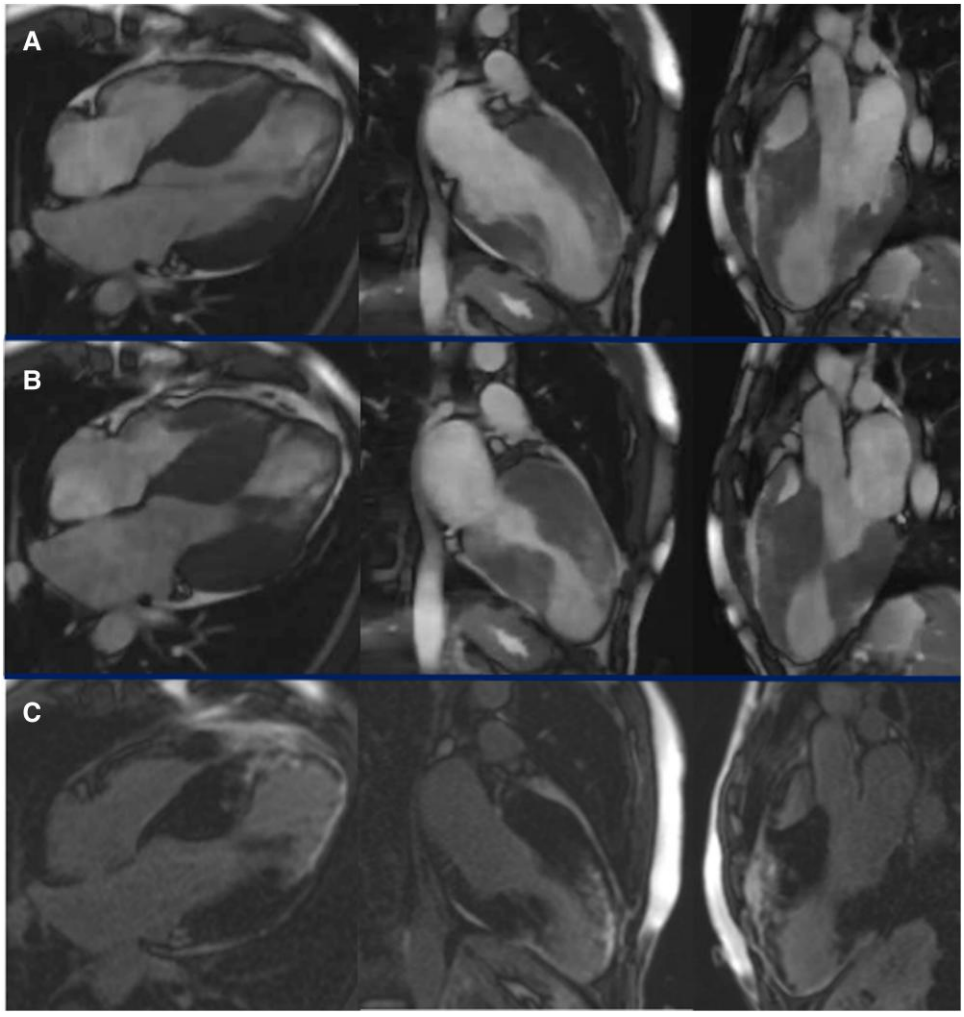

Figure 2 CMR imaging. End-diastole (*A*) and end-systole (*B*) images detailing a MWT of 30 mm and an apical aneurysm. With contrast injection, note the presence of diffuse and extensive LGE in the apical region of the left ventricle (C).

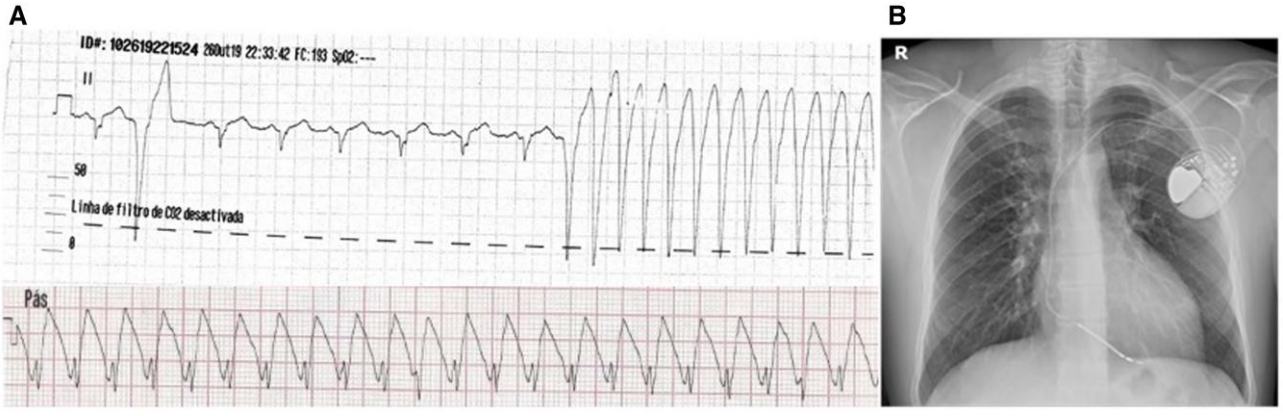

Figure 3 Admission rhythm strips and chest x-ray. Emergency medical service rhythm strips (*A*) revealing sustained VT at 192 b.p.m. The admission chest x-ray (*B*) shows no ICD generator or lead displacement.

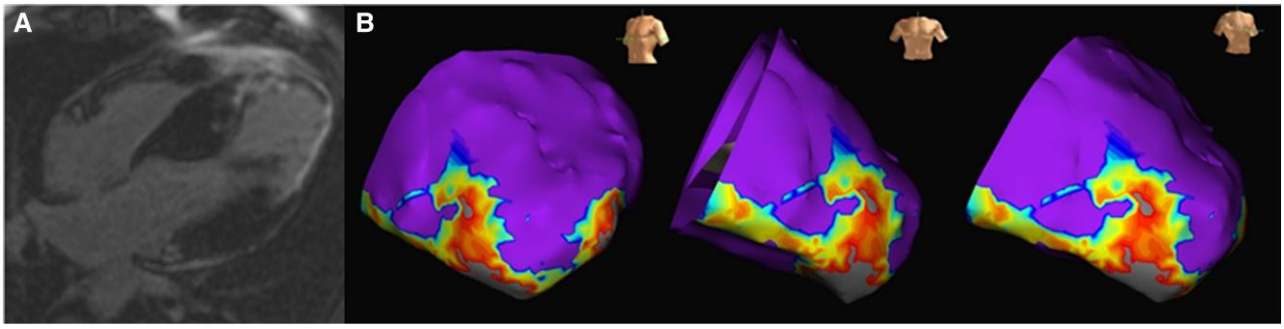

Figure 4 CMR-guided epicardial VT ablation. LGE region in CMR (*A*) revealing apical fibrosis, compatible with the arrhythmic substrate documented in the electrophysiological study voltage map (*B*).

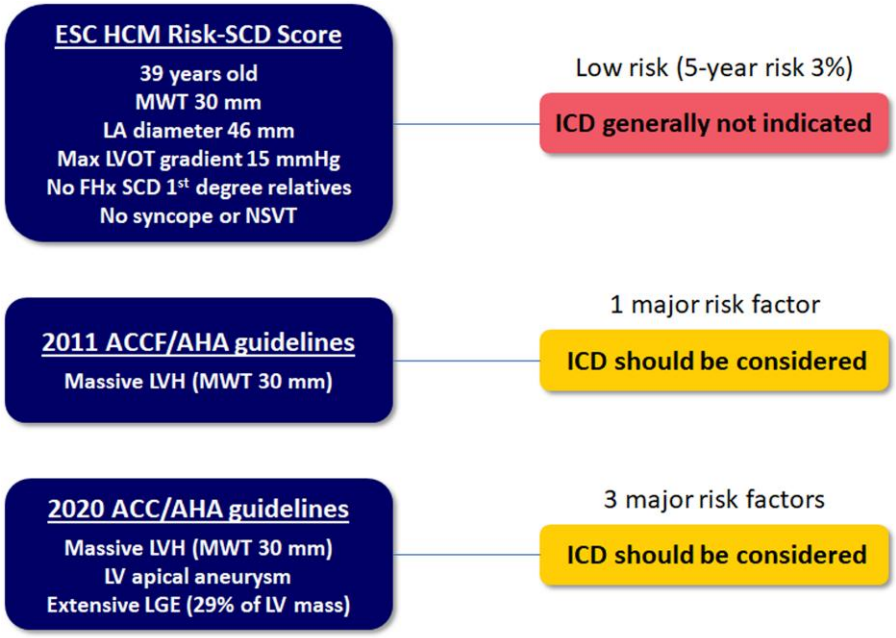

Figure 5 Predicted risk for SCD and indication for prophylactic ICD according to the different available scoring systems.

features with LGE. The presence of extensive LGE (quantified as ≥15% of LV mass) is a proven marker of SCD risk.[2] LV apical aneurysms also predispose to re-entry circuits in the junction of the scarred aneurysm and adjacent myocardium. These aneurysms are associated with monomorphic VT.[10] LV systolic dysfunction, defined as LV ejection fraction (EF) of <50%, is also a novel predictor of SCD risk.[11]

The 2020 ACC/AHA guidelines[9] include these findings in the integrated approach to stratify SCD risk. A prophylactic ICD is reasonable in the presence of at least one of the following risk factors for ICD (apical aneurysm, LVEF <50% or extensive LGE on CMR). In keeping with these recommendations, our patient presented three major risk factors for SCD: massive LVH (MWT 30 mm), LV apical aneurysm, and extensive LGE (29% of LV mass). Thus, a prophylactic ICD should be considered (IIa recommendation).

CMR can also detect myocardial scar that is increasingly relevant in VT catheter ablation as a CMR-guided approach has been associated with lower fluoroscopy and RF times. It can also potentially increase non-inducibility rates and lower VT recurrence after substrate ablation.[12]

The presence of fragmented QRS complexes on the ECG reflects conduction delay due to inhomogeneous ventricular activation. It has a high predictive value for myocardial scar, which entails risk of

arrhythmic events in patients with non-ischaemic cardiomyopathy.[13] However, studies regarding the use of the ECG in risk stratification of HCM patients have shown conflicting results. No ECG pattern can currently be used for clinical decision-making.[14] Regarding genetic testing in HCM, the detection of compound mutations may predispose patients to adverse disease progression, and multiple sarcomere mutations have been associated with a risk of SCD even in the absence of conventional risk factors.[15] Other pathophysiological features of HCM such as microvascular or autonomic dysfunction, exercise-induced ischaemia, and myocardial disarray are also implicated in SCD. Future models could potentially study their role in risk stratification of SCD or employ machine learning neural networks—a promising tool undergoing validation in the identification of patients with HCM.[10,16]

Conclusion

This case details the highly complex process of SCD prevention and management in HCM patients. It highlights the need for an accurate evaluation of who may benefit from prophylactic ICD therapy. Systematic application of state-of-the-art risk stratification criteria, in which CMR plays an important role, is of paramount importance as timely implantation of a prophylactic ICD can prevent lethal arrhythmic events in HCM patients.

Lead author biography

Pedro Garcia Brás is a Cardiology fellow in Hospital de Santa Marta, Centro Hospitalar Universitário de Lisboa Central, currently undertaking an International Fellowship in St Bartholomew's Hospital, London, specializing in Cardiomyopathies and Inherited Cardiac Conditions.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

Acknowledgements

The authors would like to thank the Cardiomyopathies and Electrophysiology Unit of the Santa Marta Hospital for their invaluable input in improving the manuscript.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The patient has provided written informed consent for the submission of this manuscript, in accordance with COPE guidelines.

Conflict of interest: None declared.

Funding: None declared.

References

1. Elliott P, Anastasakis A, Borger MA, Borggrefe M, Cecchi F, Charron P, *et al.* 2014 ESC guidelines on diagnosis and management of hypertrophic cardiomyopathy. *Eur Heart J* 2015;**121**:7–57.
2. Chan R, Maron BJ, Olivotto I, Pencina MJ, Assenza GE, Haas T, *et al.* Prognostic value of quantitative contrast-enhanced cardiovascular magnetic resonance for the evaluation of sudden death risk in patients with hypertrophic cardiomyopathy. *Circulation* 2014;**130**:484–449.
3. Maron MS, Rowin EJ, Wessler BS, Mooney PJ, Fatima A, Patel P, *et al.* Enhanced American college of cardiology/American heart association strategy for prevention of sudden cardiac death in high-risk patients with hypertrophic cardiomyopathy. *JAMA* 2019;**4**:644–657.
4. Igarashi M, Nogami A, Kurosaki K, Hanaki Y, Komatsu Y, Fukamizu S, *et al.* Radiofrequency catheter ablation of ventricular tachycardia in patients with hypertrophic cardiomyopathy and apical aneurysm. *JACC: Clinical Electrophysiology* 2018;**4**:339–350.
5. Geske JB, Ommen SR, Gersh BJ. Hypertrophic cardiomyopathy: clinical update. *JACC: Heart Failure* 2018;**6**:364–375.
6. O'Mahony C, Jichi F, Pavlou M, Monserrat L, Anastasakis A, Rapezzi C, *et al.* A novel clinical risk prediction model for sudden cardiac death in hypertrophic cardiomyopathy (HCM risk-SCD). *Eur Heart J* 2014;**35**:2010–2020.
7. Wang J, Zhang Z, Li Y, Xu Y, Wan K, Chen Y. Variable and limited predictive value of the European society of cardiology hypertrophic cardiomyopathy sudden-death risk model: A meta-analysis. *Canadian Journal of Cardiology* 2019;**35**:1791–1799.
8. Choi YJ, Kim HK, Lee SC, Park JB, Moon I, Park J, *et al.* Validation of the hypertrophic cardiomyopathy risk-sudden cardiac death calculator in Asians. *Heart* 2019;**105**:1892–1897.
9. Ommen SR, Mital S, Burke MA, Day SM, Deswal A, Elliot P, *et al.* 2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy. *Circulation* 2020;**142**:e558–e631.
10. Pelliccia F, Gersh BJ, Camici PG. Gaps in evidence for risk stratification for sudden cardiac death in hypertrophic cardiomyopathy. *Circulation* 2021;**143**:101–103.
11. Maron BJ, Desai MY, Nishimura RA, Spirito P, Rakowski H, Towbin JA, *et al.* Management of hypertrophic cardiomyopathy: JACC state-of-the-art review. *J Am Coll Cardiol* 2022;**79**:390–414.
12. Soto-Iglesias D, Penela D, Jáuregui B, Acosta J, Fernández-Armenta J, Linhart M, *et al.* Cardiac magnetic resonance-guided ventricular tachycardia substrate ablation. *JACC: Clinical Electrophysiology* 2020;**6**:436–447.
13. Jain R, Singh R, Yamini S, Das M. Fragmented ECG as a risk marker in cardiovascular diseases. *Curr Cardiol Rev* 2014;**10**:277–286.
14. Finocchiaro G, Sheikh N, Biagini E, Papadakis M, Maurizi M, Sinagra G, *et al.* The electrocardiogram in the diagnosis and management of patients with hypertrophic cardiomyopathy. *Heart Rhythm* 2020;**17**:142–151.
15. Maron BJ, Maron MS, Semsarian C. Double or compound sarcomere mutations in hypertrophic cardiomyopathy: A potential link to sudden death in the absence of conventional risk factors. *Heart Rhythm* 2012;**9**:57–63.
16. Ko W, Siontis K, Attia Z, Carter R, Kapa S, Ommen SR, *et al.* Detection of hypertrophic cardiomyopathy using a convolutional neural network-enabled electrocardiogram. *J Am Coll Cardiol* 2020;**75**:722–733.