

Workup and management of rhythm disorders in myocarditis and inflammatory cardiomyopathy: a clinical consensus statement of the European Heart Rhythm Association and the Heart Failure Association of the ESC, the ESC Working Group on Myocardial & Pericardial Diseases, the European Association of Preventive Cardiology of the ESC, the Heart Rhythm Society, the Asian Pacific Heart Rhythm Society, and the Latin American Heart Rhythm Society

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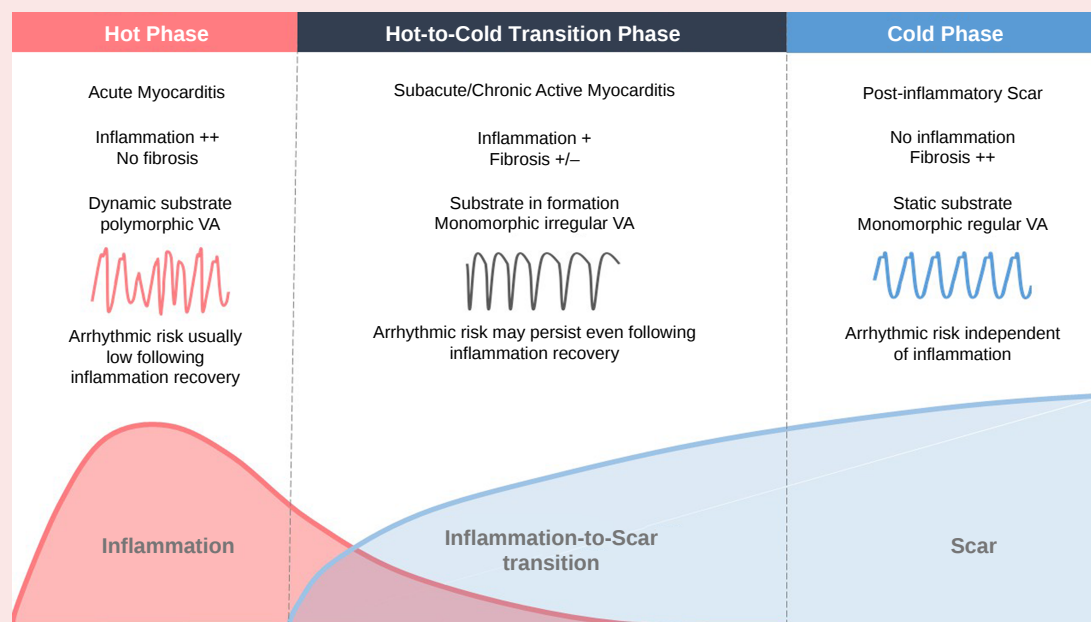
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Myocarditis is an important and frequently underrecognized cause of atrioventricular conduction disease and ventricular arrhythmias and may also be associated with atrial arrhythmias and sinus node dysfunction. Arrhythmic risk evolves over time as a result of the interplay between myocardial inflammation, fibrosis, and genetic predisposition. Despite its clinical relevance, guidance for the diagnosis, treatment, and follow-up of arrhythmias in myocarditis has remained limited. This consensus statement provides a structured framework for the management of arrhythmias in myocarditis and inflammatory cardiomyopathy (Infl-CMP), integrating disease phase and genetic background into clinical decision-making. It offers practical guidance across the disease spectrum, including diagnostic evaluation, therapeutic strategies, and longitudinal follow-up. The document was jointly developed by the European Heart Rhythm Association of the European Society of Cardiology (ESC) in collaboration with the Heart Failure Association of the ESC, the ESC Working Group on Myocardial & Pericardial Diseases, and the European Association of Preventive Cardiology of the ESC, together with the Heart Rhythm Society, the Asia Pacific Heart Rhythm Society, and the Latin American Heart Rhythm Society. Consensus advice is organized using a phase-aware framework distinguishing hot, hot-to-cold, and cold phases, with clinical consensus statements graded according to opinion-, observational-, or randomized trial-based evidence and supported by formal author voting. Arrhythmia management is stratified by disease phase. During active inflammation, antiarrhythmic therapy is combined with aetiology-targeted treatment or immunosuppression when indicated. In later stages, substrate-based strategies include pharmacological therapy, device implantation, or ablation. Emphasis is placed on hot-to-cold transition as an arrhythmogenic window and on risk stratification.

Graphical Abstract



Keywords

Myocarditis • Inflammatory cardiomyopathy • Arrhythmias • Rhythm disorders • Sudden cardiac death • Heart failure • Hot • Cold • Hot-to-cold

Table of contents

Introduction	4	Functional testing	14
Arrhythmic myocarditis: a clinical interface between		Programmed ventricular stimulation	14
inflammation and electrophysiology	4	Other provocative testing	14
Methods	5	Summary	14
Chapter 1: risk stratification and patient profiling	5	Chapter 5: device strategies in arrhythmic myocarditis	14
Concepts and definitions: inflammation meets		Risk stratification for device therapy	16
electrophysiology	5	Choosing the right device	16
Aetiological spectrum and arrhythmia patterns	6	Temporary protection and device deferral	16
Mechanisms and substrates of arrhythmia in myocarditis	6	Genetic substrates and ICD timing	16
Summary	7	Programming, follow-up, and reassessment	16
Chapter 2: substrate-oriented diagnostics in myocarditis and		Summary	18
inflammatory cardiomyopathy	8	Chapter 6: electrophysiological mapping and ablation in	
Diagnostic rationale	8	arrhythmic myocarditis	18
ECG and rhythm monitoring	8	Electroanatomical mapping: applications and techniques	18
Echocardiography	8	Catheter ablation of ventricular arrhythmias	18
Cardiac magnetic resonance	9	VT ablation in specific myocarditis subtypes	18
PET and cardiac CT	9	Supportive strategies in ablation context	18
Electroanatomic mapping as a diagnostic test	9	Ablation of supraventricular arrhythmias	18
Endomyocardial biopsy	10	Summary	21
Genetic and autoimmune substrates	10	Chapter 7: surveillance—follow-up and long-term	
Multidisciplinary and specialized care	10	monitoring	21
Summary	10	Structured follow-up and rhythm monitoring	21
Chapter 3: pharmacological management of arrhythmias in		Monitoring antiarrhythmic therapy	23
myocarditis	10	Activity guidance, return to sport, and pregnancy	23
Antiarrhythmic therapy in the hot phase	10	Long-term management: device and pharmacotherapy	
Rhythm and substrate control in the cold phase	13	reassessment	24
Immunomodulatory therapy	13	Summary	24
Special clinical scenarios	14	Chapter 8: application of clinical case scenarios	26
Summary	14	Case 8.1: High-degree AV block in a young patient:	
Chapter 4: provocative tests in arrhythmic myocarditis	14	unmasking inflammatory cardiomyopathy	26
		Case 8.2: Atrial fibrillation in systemic rheumatologic	
		disease with chronic myocarditis	28

Case 8.3: Polymorphic ventricular tachycardia in active myocarditis	28
Case 8.4: Scar-related ventricular tachycardia in healed myocarditis	28
Case 8.5: Recurrent arrhythmic myocarditis and hot-phase genetic cardiomyopathy	29
Case 8.6: Paediatric myocarditis with Brugada-like ECG pattern	31
Case 8.7: Arrhythmic myocarditis in the postpartum period	31
Case 8.8: Arrhythmic myocarditis in a competitive athlete	32
Closing remarks	32
Chapter 9: perspectives, limitations, and conclusive remarks	33
Hot-to-cold transition: a new frontier in arrhythmic risk stratification	33
Arrhythmic risk stratification and the role of genetics	33
Artificial intelligence and predictive modelling	33
Novel immunomodulatory therapies	34
Arrhythmia-specific clinical trials and registries	34
Inflammation resolution and risk persistence	34
Refinement of classification systems	34
Conclusion	34
Supplementary material	35
Acknowledgements	35
Funding	35
References	35

Introduction

Arrhythmic myocarditis: a clinical interface between inflammation and electrophysiology

Arrhythmic myocarditis represents a critical overlap between myocardial inflammation and electrical instability. The condition frequently manifests with ventricular arrhythmias (VA), conduction disturbances, or electrical storm—even in the absence of overt heart failure or left ventricular (LV) dysfunction.^{1–3} Supraventricular tachyarrhythmias may occur in the whole spectrum of inflammatory myopericardial syndromes (IMPS).¹ Inflammatory and fibrotic substrates evolve over time, and the clinical trajectory can shift rapidly from acute, reversible inflammation to chronic, scar-driven arrhythmogenicity. While the 2025 European Society of Cardiology (ESC) guidelines¹ classify myocarditis as acute, subacute or chronic based on clinical criteria (duration of symptoms), arrhythmic risk often evolves through stages that do not perfectly match with the time of symptom onset. We therefore introduce the phases ‘hot’, ‘hot-to-cold’, and ‘cold’, corresponding to active inflammation, transition to early scar formation, and stable post-inflammatory scar. This model complements the ESC framework and provides the additional granularity required for arrhythmic risk assessment and management.

We hereby define the phases (or stages) of arrhythmic myocarditis as follows:

- **Hot phase:** Characterized by active myocardial inflammation with electrically unstable myocardium, typically presenting during acute myocarditis with polymorphic/pleomorphic ventricular arrhythmias, new-onset conduction disturbances, supraventricular tachyarrhythmias, or electrical storm, often irrespective of left ventricular function.

Example: a patient presenting with ventricular fibrillation and

newly diagnosed acute myocarditis with myocardial oedema and necrosis on imaging/histology, without evidence of tissue scarring.

- **Hot-to-cold transition:** a distinct electrically high-risk window occurring after the acute inflammatory phase, in which inflammatory activity progressively declines while early fibrotic remodelling promotes the development of re-entrant arrhythmogenic substrates. During this stage, ventricular arrhythmias increasingly reflect organized re-entry mechanisms despite apparent normalization of biomarkers and partial recovery on cardiac imaging.

Example: a patient presenting with monomorphic ventricular tachycardia and persistent myocardial inflammation on imaging/histology, associated with evolving tissue scarring.

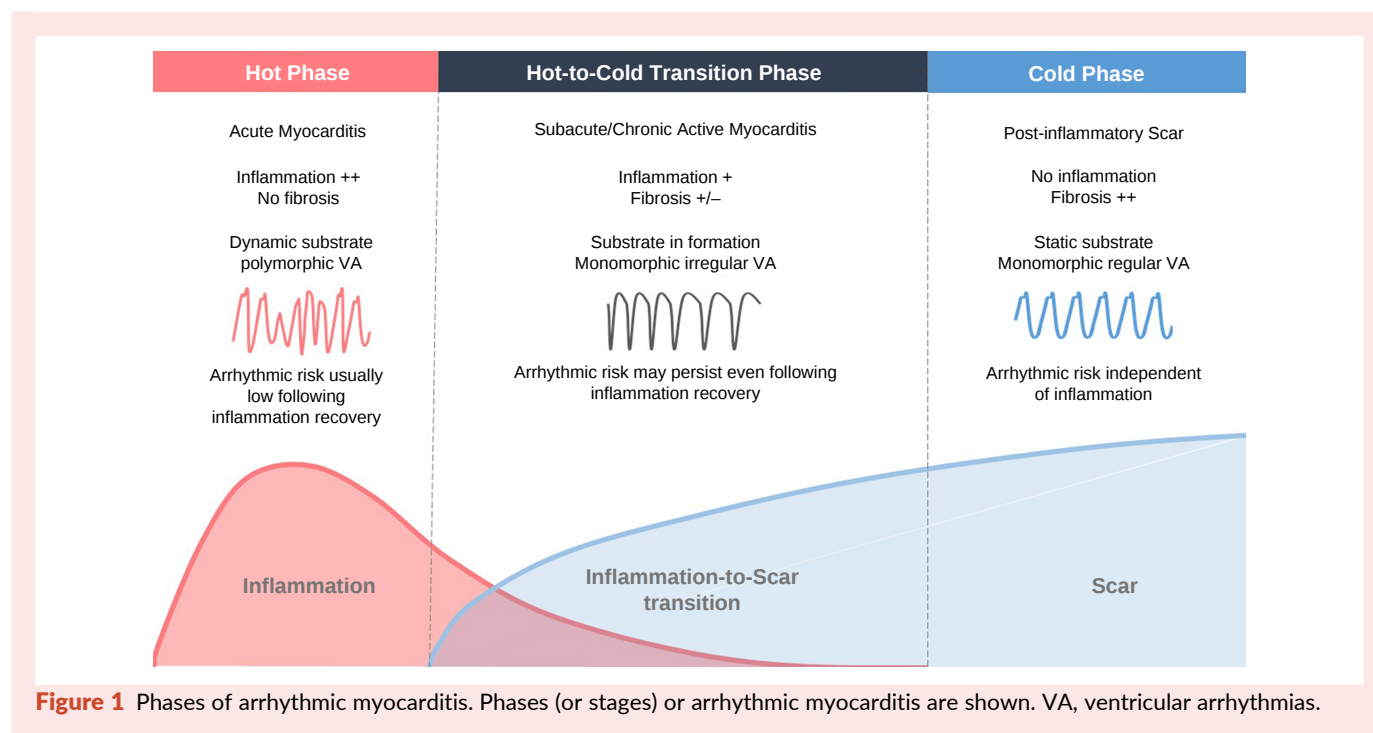
- **Cold phase:** Defined by chronic post-inflammatory scar-related arrhythmogenicity, in which ventricular tachycardia and conduction disease are predominantly sustained by stable fibrotic substrate rather than active inflammation.

Example: a patient presenting with monomorphic ventricular tachycardia late after healed myocarditis, with isolated tissue scarring on imaging/histology and no residual inflammation.

The hot phase is marked by active inflammation, polymorphic/pleomorphic VA, new-onset bradyarrhythmias and supraventricular tachyarrhythmias.^{4,5} During the cold phase tachyarrhythmias and conduction disease emerge from residual myocardial scarring.^{4,5} In the hot-to-cold transition phase re-entrant VA may manifest despite progressive normalization of imaging and biomarkers.^{6,7} Acknowledging this phase is essential for improving risk stratification and guiding treatment.^{6,8} Therapeutic decisions—whether anti-inflammatory, antiarrhythmic, interventional, or device-based—must therefore be tailored to the underlying phase and rhythm phenotype.⁸ Figure 1 shows the phases of arrhythmic myocarditis.

Not all patients are at equal risk. Genetic predisposition plays a pivotal role in shaping both the inflammatory response and the arrhythmic profile.^{9–11} Variants in genes such as *LMNA*, *DSP*, or *FLNC* may predispose to early VA, atrioventricular (AV) block, or rapid fibrotic remodelling—even in the setting of preserved ejection fraction or mild inflammatory triggers.^{12,13} In particular, the concept of *hot-phase genetic cardiomyopathy* (HPGC) has emerged as a unifying framework for genetically primed myocarditis-like presentations with disproportionate electrical instability.^{10,14} We also acknowledge the ESC 2025 terminology for the IMPS, distinguishing myopericarditis (pericarditis-predominant with limited myocardial injury and preserved systolic function) from perimyocarditis (myocarditis-predominant with pericardial involvement).¹ Since arrhythmic risk is driven by myocardial substrate (scar burden/topology, VA), our phase model (hot, cold, transition phase) applies mainly to perimyocarditis.

In this clinical consensus statement, we provide a structured, phase-specific, and rhythm-guided framework for managing arrhythmic myocarditis and inflammatory cardiomyopathy (Infl-CMP). By integrating electrophysiology, imaging, immunology, and genetics, this document aims to support decision-making in this complex clinical field. Arrhythmic myocarditis is not a fixed entity—it is a moving target. Clinical vigilance, interdisciplinary collaboration, and phase-adapted action are essential for improving patient outcomes.¹



Methods

This clinical consensus statement is not intended as a guideline but as a support for clinical management. The definitions of the category of advice and associated areas of uncertainty are based on updated systematic review of the literature, as detailed in [Supplementary material online, Table S0.1](#). The type of supporting evidence and strength of evidence are listed in [Supplementary material online, Table S0.2](#). Throughout, ESC 2025 Guidelines recommendations are cited as context,¹ while our statements are issued as consensus advice with expert opinion (OPN), observational study (OBS), and randomized clinical trials (RCT) grading as detailed in [Supplementary material online, Table S0.2](#).

Each advice statement underwent independent voting by all co-authors, regardless of evidence level. In detail, $\geq 70\%$ agreement was required for inclusion; $\geq 90\%$ defined strong consensus. We report our grading (RCT/OBS/OPN) and voting percentage for each statement; where helpful, we add the ESC Guidelines recommendation as contextual information.

Chapter 1: risk stratification and patient profiling

Who is at risk?

Risk stratification in myocarditis and Infl-CMP requires a nuanced, phase-oriented understanding of the interaction between inflammatory activity, structural remodelling, and arrhythmic phenotype. Both patient-specific and disease-specific factors—genetic background, inflammation severity, substrate distribution—contribute to arrhythmic risk, which evolves over time. This chapter summarizes definitions, aetiologies, clinical phases, and risk enhancers that shape the arrhythmic trajectory in myocarditis, with supporting tables and structured references.

Concepts and definitions: inflammation meets electrophysiology

According to World Health Organization/ESC consensus, myocarditis is diagnosed histologically by myocardial inflammatory

infiltrates with non-ischaemic myocyte necrosis,^{1,3,15,16} and immunohistologically by ≥ 14 leukocytes/mm² including ≥ 7 CD3+ T cells.^{1,3,17–20} Refined diagnostic criteria for lymphocytic myocarditis with a scoring system (so-called Seaport criteria) have been recently published,²¹ and may help to rule out differential diagnoses. Infl-CMP describes chronic myocarditis in association with cardiac dysfunction and ventricular remodelling with clinical phenotype of hypokinetic, either dilated or non-dilated cardiomyopathy with/without arrhythmogenic substrate.¹ These entities form a dynamic continuum ranging from self-limiting inflammation to fibrotic remodelling and heart failure. EMB defines both myocarditis stage and histotype ([Figure 2](#)).^{3,22} The 2025 ESC guidelines on Myocarditis and Pericarditis highlight the value of cardiac magnetic resonance (CMR) as the first-choice gold standard diagnostic test in uncomplicated scenarios.^{1,23} Instead, in patients haemodynamically unstable, as well as in those with other arrhythmia-related limitations to CMR (implanted cardiac devices, fast/irregular heartbeat), EMB is still strongly advised.^{1–3,24} In addition to troponin and natriuretic peptides, C-reactive protein (CRP) is useful to monitor inflammatory activity in suspected IMPS.^{1,25} The combination of EMB, multimodality imaging and biomarkers may help to appropriately stage arrhythmic myocarditis. [Figure 3](#) shows relevant diagnostic tests and the main applications of EMB and CMR.

In turn, identification of the hot-to-cold phase requires patient-tailored, guideline-directed multi-parameter assessment: (i) EMB (when indicated): partial regression of inflammation with early fibrosis; (ii) CMR: residual or near-normal T2 values with persistent or modifying late gadolinium enhancement (LGE); (iii) Rhythm monitoring: persistence of VA despite clinical improvement; and (iv) Clinical/biomarkers: improving symptoms with declining troponin/CRP. This phase is frequently subacute/chronic by ESC Guidelines criteria,¹ yet carries distinct arrhythmic implications that may influence timing of ablation or device therapy.

A relapsing pattern may be associated with genetically primed disease, often linked to variants in *DSP* and other desmosomal

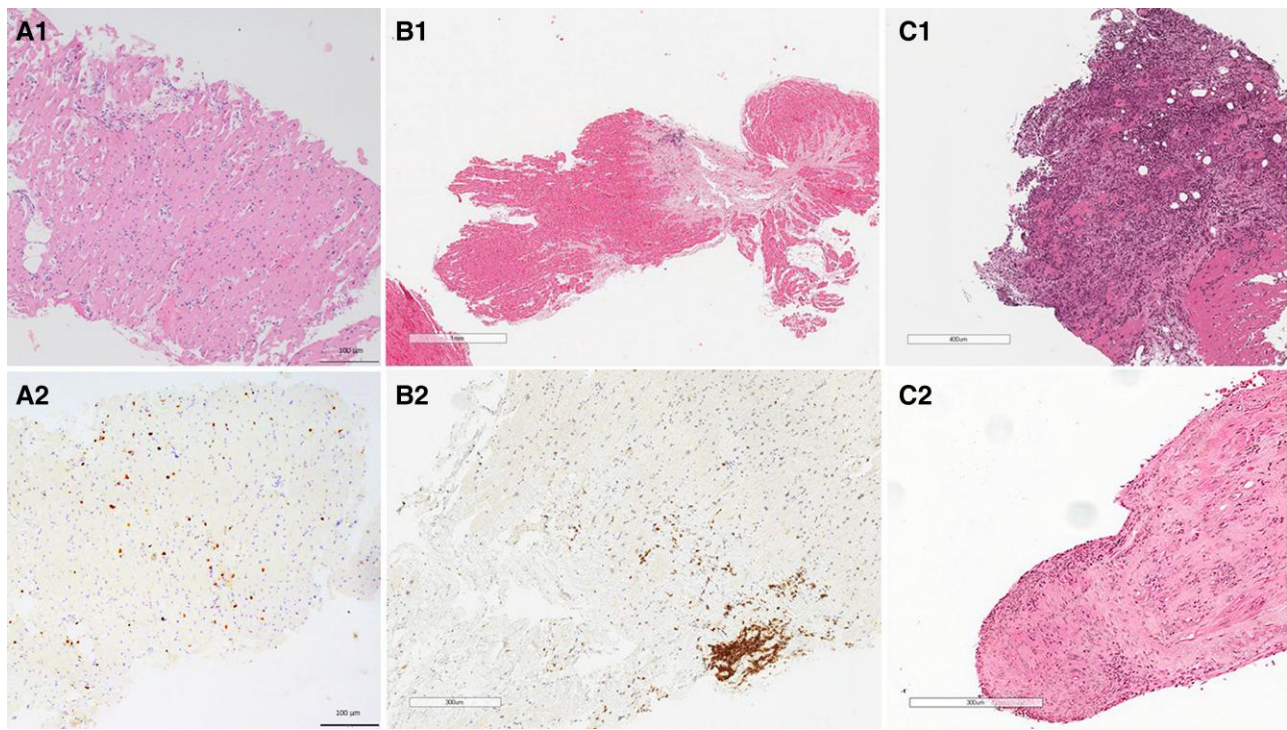


Figure 2 EMB-defined stages and histotypes of arrhythmic myocarditis. EMB-defined stages and histotypes of arrhythmic myocarditis. (A) Lymphocytic myocarditis, hot phase (acute): inflammatory infiltrates, oedema, non-ischaemic necrosis (A1, HE), CD3+ T lymphocytes $> 7/\text{mm}^2$ and $>15/\text{high power field}$ (mild myocarditis according to 2025 Seaport criteria) (A2, IHC). (B) Lymphocytic myocarditis, hot-to-cold phase (chronically active): inflammatory infiltrates, replacement fibrosis (B1, HE), >2 clusters of CD3+ T lymphocytes (moderate myocarditis according to 2025 Seaport criteria) (B2, IHC). (C) Special histotypes with strong arrhythmic associations, namely: giant cell myocarditis (C1, HE) and cardiac sarcoidosis with noncaseating granulomas (C2, HE). EMB, endomyocardial biopsy; HE, haematoxylin eosin; IHC, immunohistochemistry.

genes.^{9–11,26–30} These entities, grouped under the umbrella of HPGC,^{1,14} challenge classical distinctions and require dedicated stratification strategies. [Supplementary material online, Table S1.1](#) summarizes working definitions of arrhythmic myocarditis and overlap conditions, including subtypes with recurring inflammation, scar formation, or genetic triggers. The temporal and mechanistic evolution of inflammation is further illustrated in [Supplementary material online, Table S1.2](#), which aligns phases with clinical and electrophysiological implications.

Aetiological spectrum and arrhythmia patterns

The arrhythmogenic potential of myocarditis varies by aetiology (see [Supplementary material online, Table S1.3](#)). Viral pathogens—such as parvovirus B19, human herpesvirus 6 (HHV6), and adenovirus—may drive both acute VA and delayed scar-related ventricular tachycardia (VT).^{1,31} In some cases (e.g. HHV6), latency and reactivation may contribute to relapses.²

Bacterial and parasitic infections add specific patterns: Lyme myocarditis causes mostly reversible AV block;^{32,33} *Trypanosoma cruzi* (Chagas disease) leads to sustained VT and conduction disease due to diffuse fibrosis.³⁴ Tuberculosis is more frequently associated with pericardial diseases but may also cause isolated myocarditis.¹ A range of tropical infections (e.g. Dengue, malaria, leptospirosis, rickettsioses, West Nile

virus) may present with arrhythmic complications in the febrile or subacute phase.³⁵ These are summarized in [Supplementary material online, Table S1.4](#), highlighting the importance of travel history in unexplained arrhythmias.

Immune-mediated forms (e.g. sarcoidosis, giant cell, eosinophilic myocarditis) may affect the conduction system or provoke sustained VA.^{4,36} Giant cell myocarditis (GCM) in particular shows aggressive course and demands early immunosuppression.^{37,38} Arrhythmias frequently complicate myocarditis even in the context of systemic rheumatologic diseases.³⁹ Drug-induced or immune-mediated myocarditis (e.g. immune checkpoint inhibitors) increasingly contributes to malignant arrhythmias.⁴⁰

Finally, pathogenic/likely pathogenic variants in cardiomyopathy-associated genes, including but not limited to desmoplakin (*DSP*), filamin C (*FLNC*), and lamin A/C, (*LMNA*) encoding genes, may predispose to electrical instability, even under mild inflammatory triggers.^{10,29} Inflammatory events may unmask these genetic substrates, initiating a progressive course. [Supplementary material online, Table S1.5](#) consolidates risk-enhancing features and clinical red flags⁴¹ that require early recognition.

Mechanisms and substrates of arrhythmia in myocarditis

Arrhythmias in myocarditis range from polymorphic VT in the hot phase to re-entrant scar-related VT in the cold stage,

Table of Advice 1 Concepts, definitions, and phase labelling

Advice	Phase	Category of advice	Strength of evidence	Vote (%)	References
Label arrhythmic myocarditis at every patient contact with phase (hot; hot-to-cold; cold).	All			85	• Peretto et al. 2020⁴ • Peretto, et al. 2020⁵ • Peretto et al. 2025⁸
Define myocarditis stage by CMR and, when appropriate, EMB.	All			85	• Ferreira et al. 2018²³ • Schulz-Menger et al. 2025¹ • Caforio et al. 2013³
Identify the hot-to-cold transition phase by CMR (persistent/evolving LGE and residual T2-imaging) and/or EMB (fibrosis and persistent inflammation), and treat it as an evolving and proarrhythmic window.	Hot-to-Cold			85	• Schulz-Menger et al. 2025¹ - Rav-Acha et al.⁶
Refer to multidisciplinary teams when uncertainty exists in diagnosis or treatment decisions.	All			100	• Peretto et al. 2020⁵¹

CMR, cardiac magnetic resonance; EMB, endomyocardial biopsy; ESC, European Society of Cardiology; LGE, late gadolinium enhancement; OBS, observational study-based evidence; OPN, expert opinion-based evidence.

Chapter 2: substrate-oriented diagnostics in myocarditis and inflammatory cardiomyopathy

How to detect and define the substrate?

This chapter outlines how to detect and define the arrhythmogenic substrate in myocarditis and inflammatory cardiomyopathy. The diagnostic tools presented here support the structured algorithms discussed in Chapter 3. [Supplementary material online, Tables S2.1 and S2.3](#) provide a concise overview of diagnostic red flags, immune-genetic markers, and substrate-related arrhythmogenic findings. [Figure 4](#) shows representative examples of imaging findings in arrhythmic myocarditis.

Diagnostic rationale

Accurate identification of the arrhythmogenic substrate is central to managing rhythm disorders in myocarditis. Diagnostic strategies must distinguish inflammation-driven arrhythmias in the hot phase from scar- or genetically mediated mechanisms in the cold phase. This requires a multiparametric approach combining biomarkers, ECG, imaging, rhythm monitoring, and—when appropriate—EMB or genetic testing. In IMPS, early separation of myopericarditis from perimyocarditis matters because risk pathways diverge. Myopericarditis follows a pericarditis-led pathway; myocardial ‘flags’—meaningful scar or VA—prompt closer rhythm surveillance.¹

ECG and rhythm monitoring

Standard ECG remains a key initial tool and may reveal conduction delays, QRS fragmentation, or repolarization

abnormalities.⁵² Distinct ECG features, rather than VA types (e.g. sustained VT, NSVT, PVCs) are suggestive for different stages of myocarditis. In the hot phase, polymorphic/pleomorphic VA or electrical instability predominate, while monomorphic VT with fixed QRS morphology suggests scar-related re-entry in the cold phase.^{4,5} However, clinical onset with sustained monomorphic VT may be observed also in the hot-to-cold transition phase.^{6,53} In this setting, analysis of VT cycle stability may help differentiate transition (high variability) vs. post-inflammatory stage (low variability).^{4,5} To enable appropriate characterization of VA, 12-lead recording is always advised.^{4,54} [Supplementary material online, Table S2.1](#) summarizes key arrhythmia types, ECG clues, and substrate correlations. Holter and extended rhythm monitoring are essential for risk stratification, especially in patients with unexplained symptoms, persistent LGE, or reduced left ventricular ejection fraction (LVEF).⁵⁵ Implantable loop recorders (ILRs) allow sensitive detection of arrhythmias, and may help monitoring selected patients who do not meet the criteria for implantable cardioverter defibrillator (ICD).⁵⁶ The suggested criteria for candidacy to ILR are listed in Table of Advice 2. For device carriers, high-sensitivity programming and remote monitoring help quantify arrhythmia burden over time.

Echocardiography

Echocardiography is valuable for initial and serial assessment.^{57,58} In acute myocarditis, it may reveal global systolic dysfunction or wall motion abnormalities, oedema-related pseudohypertrophy, or pericardial effusion.³ In chronic stages, findings include wall thinning with regional akinesia or

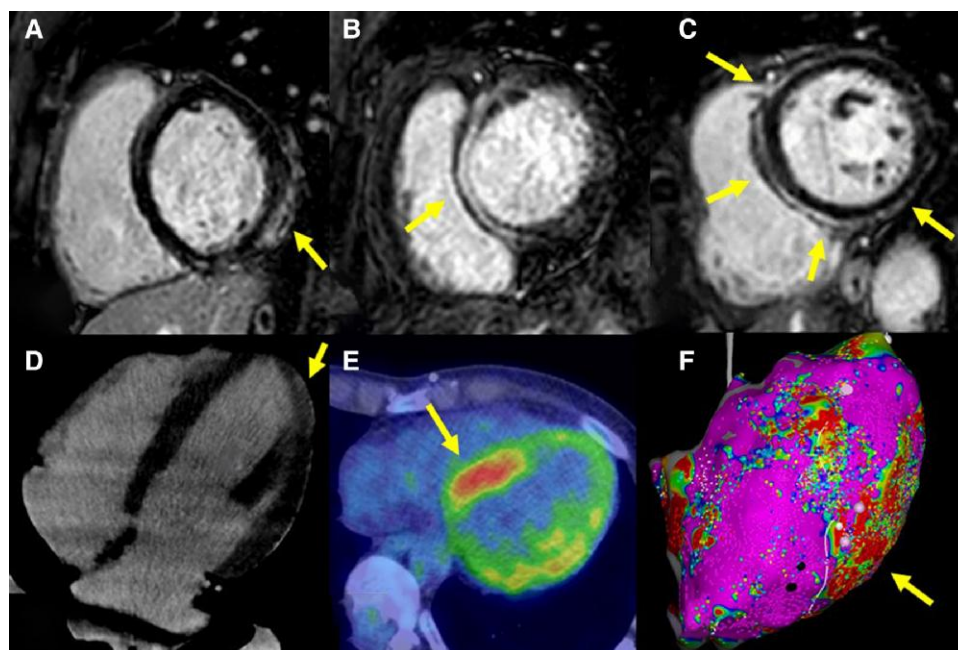


Figure 4 Imaging findings in arrhythmic myocarditis. Imaging findings are shown for patients with arrhythmic myocarditis. (A–C). CMR showing nonischaemic LGE in a patient with: (A) autoimmune myocarditis (inferolateral distribution, arrow); (B) *FLNC* gene variant (septal distribution, arrow); (C) *DSP* gene variant (ring-like distribution, arrows). (D) CT scan showing nonischaemic late iodine enhancement (lateral apical distribution, arrow) in a patient with acute myocarditis and normal coronary arteries. (E) FDG-PET scan consistent with hot-phase cardiomyopathy in a patient with *LMNA* gene variant (septal distribution, arrow). (F) Endocardial bipolar EAVM (left anterior oblique view; cut-off values 0.5 mV for dense scar—red; 1.5 mV for normal voltage—purple) showing low-voltage areas (dominant inferolateral distribution, arrow) in a patient with chronic myocarditis (cold phase). CMR, cardiac magnetic resonance; *DSP*, desmoplakin gene; EAVM, electroanatomic voltage mapping; FDG-PET, 18F-fluorodeoxyglucose positron emission tomography; *FLNC*, filamin C gene; LGE, late gadolinium enhancement; *LMNA*, lamin A/C gene.

dyskinesia. Global longitudinal strain enhances detection of systolic dysfunction even in patients with preserved LVEF.¹

Cardiac magnetic resonance

CMR with contrast remains the gold standard for non-invasive tissue characterization.^{1,2} As deemed by the updated Lake Louise criteria,²³ T2-weighted imaging and T₁-mapping detect oedema in the hot phase. As a marker of myocardial injury, LGE may revert after the acute phase of myocarditis⁵⁸; as opposed, striae of LGE persisting in the chronic stage commonly indicate fibrosis,^{1,4} and are linked to arrhythmia recurrences following presentation with sustained VT.⁵⁹ Recent data⁶ suggest that the diagnostic consistency of CMR in arrhythmic myocarditis is higher as compared to prior studies.⁵⁸ Moreover, striae of LGE displaying either septal/anteroseptal or ring-like patterns (see [Supplementary material online, Table S2.1](#)) are linked to sustained VA and worse outcomes in most^{13,60} but not all studies.⁶ Extensive LGE (although this definition lacks a universally accepted quantitative cut-off) provides prognostic insight and may prompt ICD consideration in borderline cases, especially during the cold phase of myocarditis.⁶¹ Finally, delayed-enhanced CMR plays a growing role in planning and guiding electrophysiological procedures.⁶²

PET and cardiac CT

¹⁸F-fluorodeoxyglucose positron emission tomography (FDG-PET) is particularly helpful in suspected sarcoidosis or

GCM.¹ Special patient preparation lacking carbohydrates and using a high fat diet with prolonged fasting is needed. It detects metabolically active lesions, even in preserved myocardium.⁶³ Anteroseptal FDG uptake correlates with arrhythmic risk.⁶⁴ In selected cases, gallium-67 scintigraphy plays as an alternative technique to monitor myocardial inflammation.⁶⁵ Coronary CT angiography helps to exclude significant coronary artery disease;³ delayed iodine enhancement CT may visualize fibrosis when CMR is contraindicated or altered by device-related artifacts.⁶⁶ Both FDG-PET and CT scan provide relevant information during the pre-procedural workup for patients undergoing VA catheter ablation.⁶²

Electroanatomic mapping as a diagnostic test

Electroanatomic mapping allows identification of arrhythmogenic scar substrates, especially in the cold phase of myocarditis.⁴ In detail, electroanatomical voltage mapping (EAVM) identifies low-voltage areas that correspond to fibrotic substrate. Bipolar mapping detects dense scar (<0.5 mV), while unipolar mapping captures deeper or epicardial involvement (<8.3 mV LV, <5.5 mV RV) with higher sensitivity.^{67–70} In most patients with myocarditis, substrate abnormalities are located in inferolateral LV wall (either epicardial or endocardial unipolar mapping), whereas septal localization is more common in sarcoidosis, GCM, and HPGC as *LMNA* or *FLNC*.^{65,71–73}

In the cold phase, EAVM guides ablation strategies;⁷⁰ in the hot phase, it may help distinguishing transient oedema from

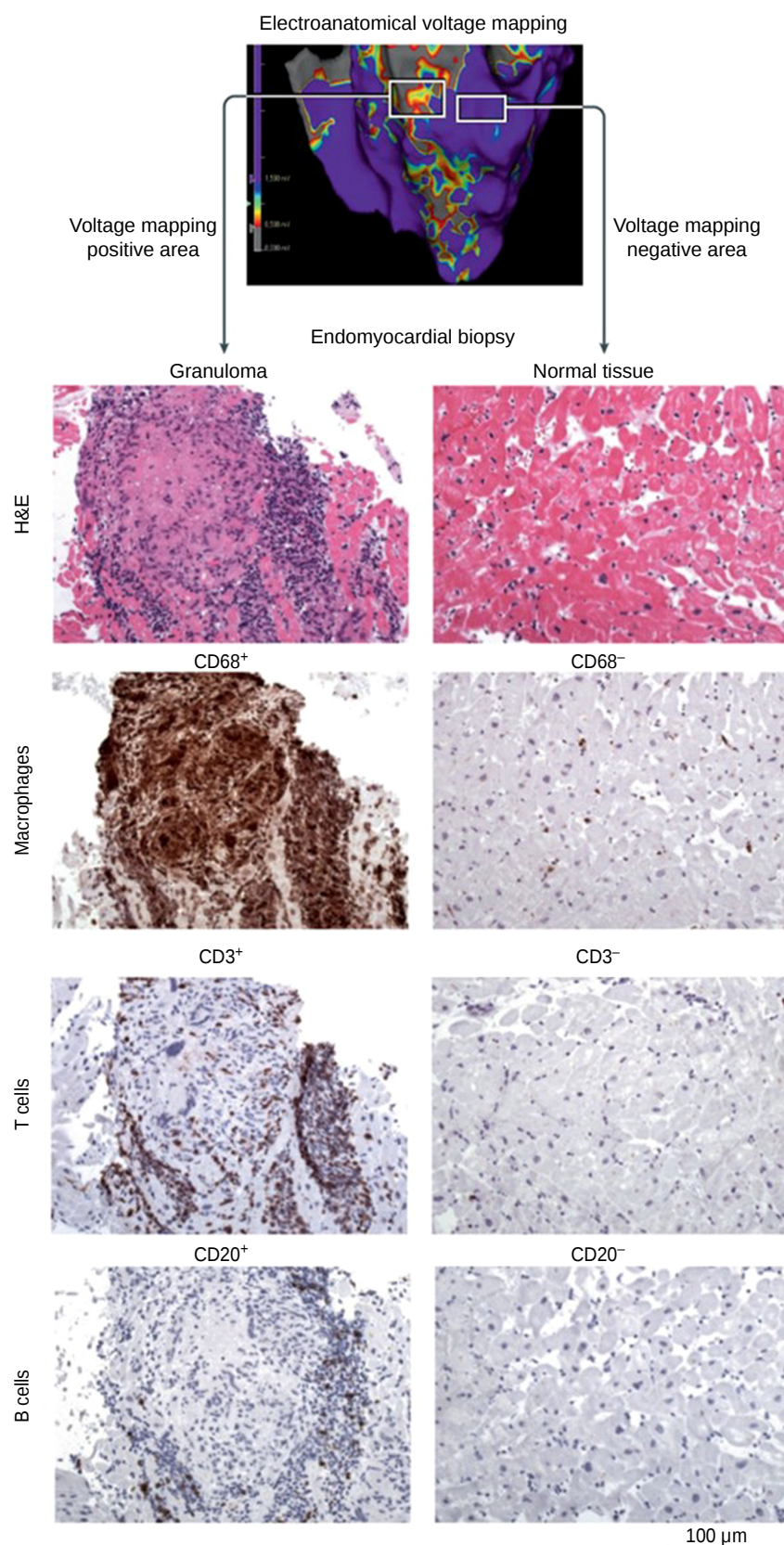


Figure 5 EAVM-guided EMB. Usefulness of EAVM in guiding EMB in a patient with cardiac sarcoidosis and patchy distribution of substrate abnormalities. Low-voltage areas (endocardial bipolar map, left anterior oblique view; cut-off values 0.5 mV for dense scar—red; 1.5 mV for normal voltage—purple) reveal inflammation and noncaseating granulomas, whereas normal voltage points to healthy tissue. Permissions for Figure obtained by Tschöpe *et al.* 2021.² EAVM, electroanatomic voltage mapping; EMB, endomyocardial biopsy.

Table of Advice 2 Diagnostic work-up

Advice	Phase	Category of advice	Strength of evidence	Vote (%)	References
Use CMR with contrast at baseline (T2 and T1, including LGE topology and burden) and schedule repeat CMR at ~ 6 months.	Hot, Hot-to-cold			95	• Schulz-Menger et al. 2025 ¹
Obtain continuous ECG telemetry (possibly 12-lead monitoring) in all patients with myocarditis admitted to the hospital, irrespectively of their clinical presentation.	All			85	• Schulz-Menger et al. 2025 ¹ • Peretto et al. 2020 ⁵ • Peretto et al. 2021 ⁵⁵
Consider ILR in patients with no indication to ICD/WCD, particularly in the following scenarios: • sporadic/rare symptoms; • asymptomatic arrhythmias; • isolated risk features as NSVT, LVEF <50% at discharge, atypical LGE (extensive, septal/anteroseptal, ring-like), high-risk genotypes.	All			85	• Peretto et al. 2021 ⁵⁵
Obtain EMB when results will change management (aetiology definition, immunosuppressive therapy, device/ablation planning) and/or when CMR is inconclusive.	All			100	• Schulz-Menger et al. 2025 ¹ • Caforio et al. 2013 ³ • Halushka et al. 2025 ²¹ • Peretto et al. 2020 ⁸
Consider EAVM-guided EMB in patients with patchy disease (i.e. sarcoidosis) or focal myocarditis.	All			90	• Tschoepe et al. 2021 ² • Casella et al. 2020 ⁷⁶
Obtain EAVM to characterize arrhythmic substrate (LVA topology/burden) in patients with myocarditis requiring invasive EP evaluation.	All			95	• Casella et al. 2021 ⁶⁷ • Peretto et al. 2020 ⁶⁹
Consider FDG-PET in patients with suspected sarcoidosis; or with suspected myocarditis and limitations to CMR (i.e. implantable device carriers), or with inconclusive results from CMR and EMB.	Hot, Hot-to-cold			100	• Peretto et al. 2022 ⁶⁴
Perform genetic testing in patients with: • family history of myocarditis, cardiomyopathy or sudden death • recurrent myocarditis; • severe/exertional arrhythmias; • persistent systolic dysfunction; • atypical LGE (extensive, septal/anteroseptal, ring-like).	All			95	• Schulz-Menger et al. 2025 ¹ • Lota et al. 2022 ⁹ • Ammirati et al. 2022 ²⁸ • Peretto et al. 2023 ²⁶ • Caforio et al. 2025 ⁸⁹

CMR, cardiac magnetic resonance; EAVM, electroanatomic voltage map; ECG, electrocardiogram; EMB, endomyocardial biopsy; EP, electrophysiological; ESC, European Society of Cardiology; FDG-PET, 18F-fluorodeoxyglucose positron emission tomography; ICD, implantable cardioverter defibrillator; ILR, implantable loop recorder; LGE, late gadolinium enhancement; LVA, low-voltage areas; LVEF, left ventricular ejection fraction; NSVT, non-sustained ventricular tachycardia; OBS, observational study-based evidence; OPN, expert opinion-based evidence; PV/LPVs, pathogenic/likely-pathogenic variants; RCT, randomized clinical trial-based evidence; VA, ventricular arrhythmias; WCD, wearable cardioverter defibrillator.

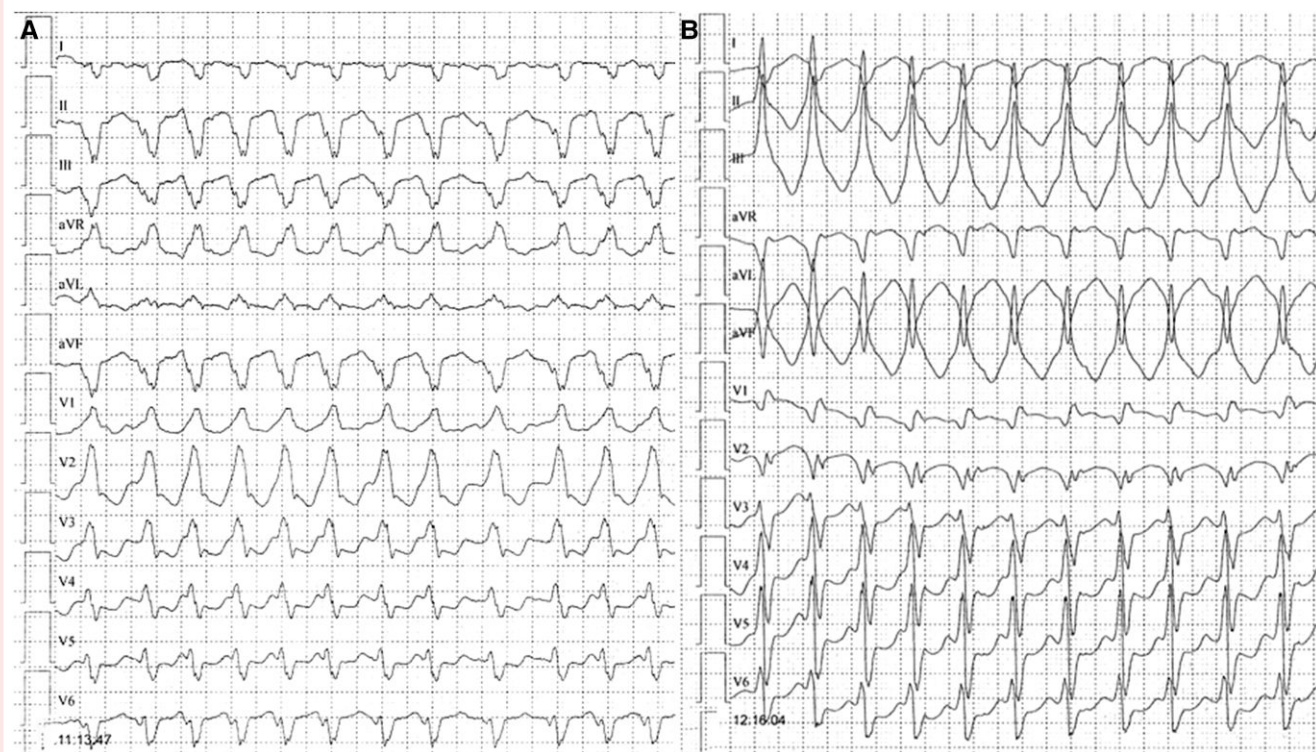


Figure 6 ECG-guided treatment in arrhythmic myocarditis. ECG features of sustained monomorphic VTs are shown in two patients with myocarditis. (A) Right bundle branch block, superior right axis (negative in lead DI) morphology suggests inferolateral LV VT exit, as commonly observed in the cold phase of myocarditis; variability in cycle length suggests hot-to-cold transition phase in a patient with acute presentation (VT) but evolving inflammatory process: immunomodulatory treatment is suggested to target VT before considering ablation ('cool down, then ablate' strategy). (B) Regular VT with qR in V1 and RS V4-V6 and inferior axis, suggesting cold-phase and septal origin in a patient with history of cardiac sarcoidosis: immunomodulatory therapy may not help, and referral to an experienced centre for VT ablation is advised. In all cases, ECG provides only partial information. Treatment strategies are driven by the clinical context and gold standard diagnostic techniques for myocarditis (cardiac magnetic resonance, endomyocardial biopsy), LV, left ventricular; VT, ventricular tachycardia.

chronic myocarditis with preserved or mildly reduced LVEF has been proven safe and effective in controlling NSVT/PVC burden.⁹⁵ The management of VA in acute heart failure should follow updated expert consensus documents.^{77,91,94} For a complete overview of drug profiles, see [Supplementary material online, Table S3.1](#). For the use of immunomodulatory therapy during hot/hot-to-cold phases, please see Chapter 3.3.

Rhythm and substrate control in the cold phase

In the chronic, post-inflammatory stage, monomorphic VTs arising from scar regions become predominant. Beta-blockers remain foundational.¹ Sotalol may be used cautiously in patients with preserved or non-severely reduced LVEF, provided QTc is monitored closely.^{5,77} In selected cases—such as post-inflammatory myocarditis with normal or near-normal function and in the absence of extensive LGE—Class IC agents may be considered.^{5,77,94}

Importantly, heart failure pharmacotherapy complements arrhythmic management by stabilizing myocardial substrate.^{1,25} As recommended by the ESC Guidelines on Heart Failure,²⁵ early initiation of renin-angiotensin-aldosterone system

inhibition, angiotensin receptor-neprilysin inhibitors, mineralocorticoid receptor antagonists, and sodium-glucose cotransporter 2 inhibitors may improve outcomes even in the underinvestigated field of myocarditis/Infl-CMP.¹ See [Supplementary material online, Table S3.2](#) for phase-adapted heart failure strategies. Monitoring results should be revisited after 3–6 months¹ to reassess the need for device therapy or further procedural interventions.

Immunomodulatory therapy

To date, the use of immunomodulatory therapy is reserved to patients with EMB-proven diagnosis of myocarditis following a demanding clinical presentation, which often involves heart failure and arrhythmias.¹ In autoimmune, virus-negative myocarditis, immunosuppression is essential to mitigate inflammatory substrate and reduce arrhythmia burden.³ Steroids associated with azathioprine or mycophenolate mofetil form the standard first-line approach (usually minimum 6 for up to 12–24 months);^{96,97} in GCM, triple immunosuppressive therapy is indicated. Immunosuppressive therapy may indirectly stabilize the arrhythmogenic substrate during the hot/hot-to-cold phases, and contribute to lower VA burden in the long term.^{98–100} The use of anakinra and other biologicals in arrhythmic myocarditis

is still investigational.^{26,99,101} Therapeutic agents and indications are summarized in [Supplementary material online, Table S3.3](#). Indications should follow existing myocarditis and Infl-CMP guidelines.¹ Electrical storm may require i.v. immunosuppressive therapy in fulminant myocarditis,⁹³ provided ongoing infections are excluded. Consistent with ESC 2025 guidelines,¹ immunosuppression in virus-negative disease is biopsy-guided and managed by dedicated multidisciplinary teams.⁵¹ Pericarditis-directed therapy in IMPS include non-steroidal anti-inflammatory drugs and colchicine according to the current ESC recommendations.¹

Special clinical scenarios

In pregnancy, arrhythmia treatment must account for foetal safety.¹ Among beta-blockers, propranolol and labetalol are safe; however, to improve the control of arrhythmias the cautious use of other agents (e.g. metoprolol, sotalol) is allowed.^{94,102,103} Amiodarone should be avoided during the first trimester.^{91,98} In life-threatening arrhythmias, lidocaine or procainamide may be used.⁹⁴ Multidisciplinary counselling is advised, especially in cases mimicking peripartum cardiomyopathy,¹⁰⁴ to guide both therapy and genetic evaluation.¹² Genetic predisposition, Chagas cardiomyopathy or ICI-associated myocarditis require tailored strategies.¹ In Chagas disease, ablation is frequently needed because of drug-refractory VA.³³ In ICI myocarditis, drug withdrawal is part of the treatment and should be discussed within a multidisciplinary team.¹ In HPGC preliminary data support the use of immunosuppressive therapy^{26,105}; however, treatment type and duration are still to be defined. When diagnostic resources or aetiology-specific treatments are unavailable, referral to specialized tertiary centres is strongly advised.¹

Summary

Pharmacological therapy in myocarditis must be stratified by disease phase and arrhythmia mechanism. Antiarrhythmic drugs, heart failure agents, and immunomodulatory therapies work synergistically to control rhythm, modulate substrate, and stabilize long-term risk. [Supplementary material online, Tables S3.1 through S3.3](#) provide a structured overview for phase-appropriate therapy selection. Device and procedural strategies are addressed in detail in the following chapters.

Chapter 4: provocative tests in arrhythmic myocarditis

When should we go invasive

The decision to perform a provocative test in arrhythmic myocarditis depends on disease phase, symptom severity, and diagnostic uncertainty. This chapter discusses indications for conventional and cardiopulmonary exercise stress test, and invasive electrophysiological studies. [Supplementary material online, Table S4.1](#) summarizes procedural indications by clinical context.

Functional testing

Functional testing supports longitudinal risk stratification in clinically stable patients. Cardiopulmonary exercise test (CPET) may reveal reduced functional reserve or delayed oxygen kinetics, indicating subclinical inflammation or early heart failure. In clinically stable patients, exercise stress testing (EST) may be useful

≥3–6 months after active inflammation to detect exertional arrhythmias or electrical instability.¹⁰⁶ EST has a relevant role in allowing safe readmission to sport in athletes.^{1,107} See [Supplementary material online, Table S4.1](#) for test modalities, indicative findings, and clinical relevance.

Programmed ventricular stimulation

Invasive programmed ventricular stimulation (PVS) may have a role in stratifying arrhythmic risk of myocarditis and guiding treatment decisions.¹ Indeed, PVS can identify individuals at higher risk of life-threatening VA, particularly when indication to ICD for primary prevention is uncertain. The evidence in support of PVS is stronger in cardiac sarcoidosis¹⁰⁸ than in unspecified myocarditis. Per expert consensus, PVS may find application in selected patients with myocarditis, as in those with either intermediate risk of SCD, unexplained syncope, or indication to pacemakers in the presence of concurrent risk factors for VA. Overall, the technique was shown to be more reliable when used in the cold stage of myocarditis.¹⁰⁴ Instead, the use of early PVS during active myocarditis is discouraged.^{1,109} Finally, either invasive or non-invasive PVS can be used in candidates to ablation to capture the morphology of the clinical VT by 12-lead ECG recording.⁶⁸

Other provocative testing

Provocative pharmacologic testing—such as ajmaline, flecainide, or procainamide challenge—may unmask latent Brugada syndrome or conduction defects in ambiguous cases.^{46,77,110} Tilt-table or autonomic stress testing may clarify syncope mechanism and help avoid unnecessary device therapy in patients with vagal prodromes.⁷⁷ However, results should be interpreted with caution, as arrhythmias remain a major cause of syncope in patients with myocarditis.^{1,4} In general, these tests are reserved for selected patients and should only be performed when the result will directly inform management decisions. Such findings may inform decisions on ICD indication, ablation candidacy, or activity counselling.

Summary

Non-invasive functional tests such as CPET and EST are informative in clinically stable patients late after the acute phase of myocarditis (cold phase). Invasive PVS may be useful to guide arrhythmic risk prediction and ablation strategy, especially in patients with cold phase myocarditis. Comprehensive risk stratification should integrate imaging, histological findings, genetic information, and clinical parameters.¹

Chapter 5: device strategies in arrhythmic myocarditis

When is a device indicated

Device therapy in myocarditis and Infl-CMP must be personalized, based on disease phase, arrhythmia type, LVEF, genetic predisposition, and the extent of myocardial scar. The challenge lies in balancing early protection against sudden cardiac death with the risks of overtreatment in a potentially reversible condition. This chapter provides advice on ICD, cardiac resynchronization therapy (CRT), pacemaker, wearable cardioverter-defibrillator (WCD), and conduction system pacing (CSP), integrating substrate, rhythm burden, and recovery trajectory. Indications by disease phase and substrate type are summarized

Table of Advice 3 Immunomodulation and antiarrhythmic drug therapy

Advice	Phase	Category of advice	Strength of evidence	Vote (%)	References
In patients with non-infectious myocarditis, initiate EMB-guided immunosuppression in demanding clinical presentations (mainly heart failure and arrhythmias)	Hot, hot-to-cold			100	- De Luca <i>et al.</i> 2020⁹⁶ - Frustaci <i>et al.</i> 2009⁹⁷ - Caforio <i>et al.</i> 2024⁹⁸ - Peretto <i>et al.</i> 2020⁹⁹ - Lakkireddy <i>et al.</i> 2019¹⁰⁰
In EMB-proven non-infectious myocarditis, favour steroid-sparing agents (AZA/MMF; MTX in sarcoidosis), ensure regular multidisciplinary surveillance, and tailor treatment duration (6–24 months) to clinical endpoints.	Hot, hot-to-cold			100	- Caforio <i>et al.</i> 201³³ - Caforio <i>et al.</i> 2024⁹⁸ - Peretto <i>et al.</i> 2020⁹⁹
Manage non-lymphocytic myocarditis (eosinophilic, giant cell, sarcoidosis) according to updated guidelines. Consult an infectious disease specialist for viral/bacterial/protozoal myocarditis. Withdraw immune checkpoint inhibitor (ICI) or other toxic agents upon multidisciplinary risk/benefit evaluation, and start specific therapy if needed.	Hot, hot-to-cold			100	- Schulz-Menger <i>et al.</i> 202⁵¹ - Caforio <i>et al.</i> 2024⁹⁸
Manage acute-phase electrical storms according to the updated protocols, and consider using i.v. immunosuppressive agents after excluding infections.	Hot			95	Lenarczyk <i>et al.</i> 2024⁹³
Use beta-blockers in patients with any stage of myocarditis to treat or prevent arrhythmias, unless contraindicated.	All			90	- McDonagh <i>et al.</i> 2021²⁵ - Merino <i>et al.</i> 2025⁹⁴
In patients with arrhythmic myocarditis and limitations/unsatisfactory response to betablockers, use: - amiodarone (or sotalol outside the acute phase and in the absence of decompensated heart failure) if LVEF is < 40%; - class IC agents (i.e. flecainide, propafenone) if LVEF is ≥40% (in selected cases, outside the acute phase and in the absence of decompensated heart failure).	All			100	- McDonagh <i>et al.</i> 2021²⁵ - Zeppenfeld <i>et al.</i> ⁷⁴ - Connolly <i>et al.</i> 2006⁹² - Merino <i>et al.</i> 2025⁹⁴ - Peretto <i>et al.</i> 2026⁹⁵
Use renin–angiotensin–aldosterone system inhibitors, angiotensin receptor–neprilysin inhibitors (ARNIs), mineralocorticoid receptor antagonists, and sodium–glucose cotransporter 2 inhibitors, according to the recommendations of the ESC Guidelines on Heart Failure.	All			100	- McDonagh <i>et al.</i> 2021²⁵ - Merino <i>et al.</i> 2025⁹⁴

AZA, azathioprine; EMB, endomyocardial biopsy; ESC, European Society of Cardiology; LVEF, left ventricular ejection fraction; MMF, mofetil mycophenolate; MTX, methotrexate; OBS, observational study-based evidence; OPN, expert opinion-based evidence; RCT, randomized clinical trial-based evidence; VA, ventricular arrhythmias.

Advice	Phase	Category of advice	Strength of evidence	Vote (%)	References
Perform symptom-limited exercise testing (± CPET) before return-to-play to exclude exertion-triggered VA.	Hot-to-cold, cold			90	- Schulz-Menger et al. 2025¹ - Peretto et al. 2022¹⁰⁶
Use PVS in cold phase for risk refinement or ablation planning.	Cold			80	- Cronin et al. 2019⁶⁸ - Zeppenfeld et al. 2022⁷⁷ - Peretto et al. 2020¹⁰⁹
Avoid EST and PVS during the acute phase of myocarditis.	Hot			95	- Schulz-Menger et al. 2025¹ - Peretto et al. 2020¹⁰⁹

CPET, cardiopulmonary exercise test; EST, exercise stress test; OBS, observational study-based evidence; OPN, expert opinion-based evidence; PVS, programmed ventricular stimulation; VA, ventricular arrhythmias.

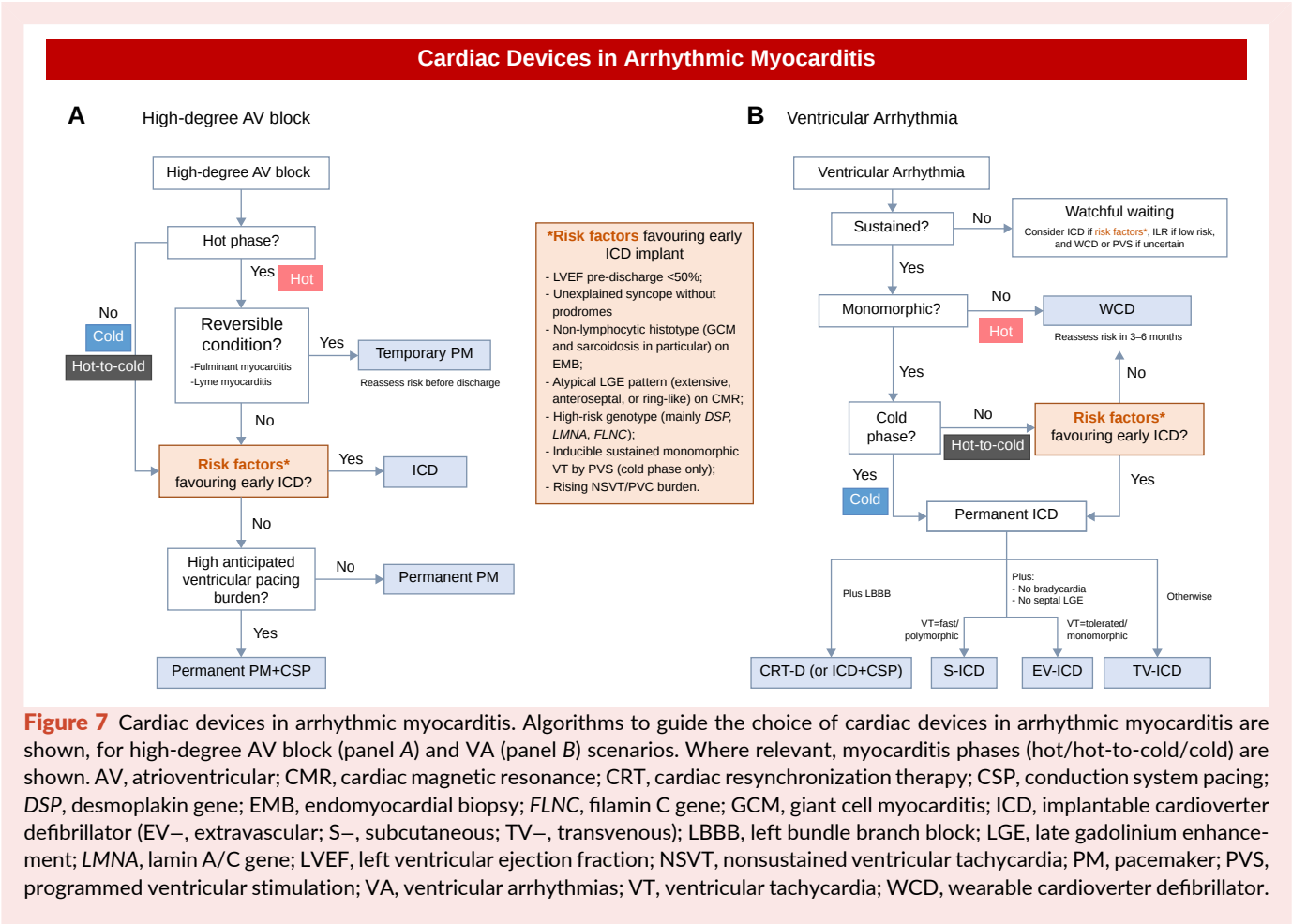


Table of Advice 5 Device therapy

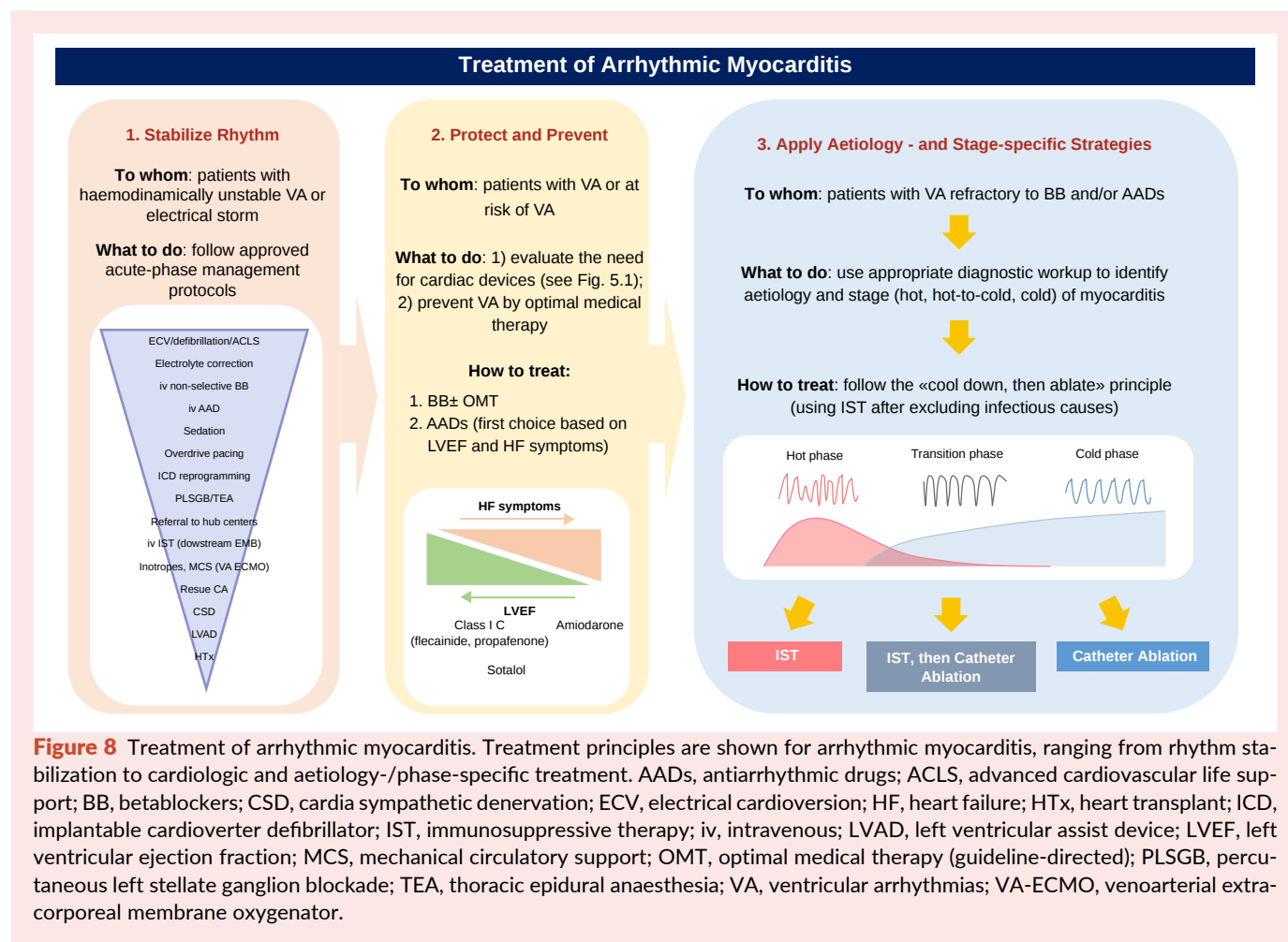
Advice	Phase	Category of advice	Strength of evidence	Vote (%)	References
Device indication					
In acute myocarditis, use temporary pacing (preferentially transvenous) to treat high-degree AV block.	Hot			100	• Schulz-Menger <i>et al.</i> 2025 ¹
In acute myocarditis, prefer WCD (life vest) bridging and reassess indication to ICD after 3–6 months.	Hot, Hot-to-Cold			95	• Schulz-Menger <i>et al.</i> 2025 ¹
In cold phase myocarditis, implant ICD in patients with sustained monomorphic VT, especially if haemodynamically not-tolerated (secondary prevention).	Cold			100	• Schulz-Menger <i>et al.</i> 2025 ¹
In cold phase myocarditis, implant ICD in patients with LVEF<35% in spite of optimal treatment for 3 months (primary prevention).	Cold			100	• Arbelo <i>et al.</i> 2023 ¹²
In uncertain cases, evaluate the following risk factors favouring early ICD implant over WCD, especially if presenting in combination. Secondary prevention	All			95	• Schulz-Menger <i>et al.</i> 2025 ¹ • Rav-Acha <i>et al.</i> 2024 ⁶ • Peretto <i>et al.</i> 2025 ¹³ • Ammirati <i>et al.</i> 2019 ⁵⁰ • Gentile <i>et al.</i> 2021 ⁵³ • Rosier <i>et al.</i> 2020 ¹²²
Both primary and secondary prevention					
• Sustained monomorphic VT.					
• LVEF pre-discharge<50%;					
• Unexplained syncope without prodromes					
• Non-lymphocytic histotype (GCM and sarcoidosis in particular);					
• Atypical and quantitatively significant LGE pattern (extensive, septal/anteroseptal, or ring-like);					
• High-risk genotype (mainly DSP, LMNA, FLNC);					
• Inducible sustained monomorphic VT by PVS (cold phase only);					
• Rising NSVT/PVC burden.					
Special aetiologies					
In Lyme myocarditis, avoid a permanent pacemaker unless high-degree AV block is persistent (despite antibiotic treatment for 2–3 weeks) or late-onset (>3–6 months).	Hot, Hot-to-Cold			100	• Schulz-Menger <i>et al.</i> 202 ⁵¹

Continued

Table of Advice 5 Continued

Advice	Phase	Category of advice	Strength of evidence	Vote (%)	References
In cardiac sarcoidosis, GCM, or HPGC presenting with high-degree AV block, favour ICD over pacemaker implant.	Hot, Hot-to-Cold			90	- Schulz-Menger et al. 2025¹ - Arbelo et al. 2023¹²
In HPGC, estimate genotype (mainly <i>LMNA</i> , <i>DSP</i> , <i>FLNC</i>) as a risk modifier, not as a stand-alone indication to ICD.	All			100	- Arbelo et al. 2023¹² - Peretto et al. 2025¹³
Device choice					
Favour transvenous over subcutaneous ICD (S-ICD) in the following scenarios: - need for pacing/symptomatic bradycardia; - risk of AV block (GCM, sarcoidosis, high-risk genotype as <i>LMNA</i>, septal LGE); - non-fast, tolerated VT, responsive to ATP (case-by-case decision; here, extracardiac ICD is an alternative).	Hot-to-cold, cold			100	- Markman et al. 2025¹¹⁵ - Wilkoff et al. 2016¹²⁷
When transvenous ICD is indicated, prefer dual-chamber devices in the following scenarios: - bradycardia (to avoid unnecessary ventricular pacing and/or preserve AV synchrony); - supraventricular tachyarrhythmias (to improve VT/VF discrimination); - high-burden PVCs (to favour atrial overdrive pacing in selected refractory cases).	Hot-to-cold, cold			95	- Wilkoff et al. 2016¹²⁸ - Fadahunsi et al. 2015¹²⁹
Favour PM implant, only after carefully excluding the risk factors favouring early ICD (see above).	Hot-to-cold, cold			100	Wilkoff et al. 2016¹²⁸
Align with ESC Guidelines on Heart Failure for indications to CRT after the acute phase.	Hot-to-cold, cold			100	McDonagh et al. 2021²⁵
Use CSP in patients with an anticipated ventricular pacing burden >20%, as well as in cases who cannot benefit from CRT.	Hot-to-cold, cold			85	Glikson et al. 2025¹¹⁶

AV, atrioventricular; ATP, anti-tachycardia pacing; CSP, conduction system pacing; CRT, cardiac resynchronization therapy; *DSP*, desmoplakin gene; ESC, European Society of Cardiology; *FLNC*, filamin C gene; GCM, giant cell myocarditis; HPGC, hot-phase genetic cardiomyopathy; ICD, implantable cardioverter defibrillator; LGE, late gadolinium enhancement; *LMNA*, lamin A/C gene; LV, left ventricular; LVEF, left ventricular ejection fraction; OBS, observational study-based evidence; OPN, expert opinion-based evidence; PM, pacemaker; PVC, premature ventricular complex; PV/LPVs, pathogenic/likely-pathogenic variants; PVS, programmed ventricular stimulation; VA, ventricular arrhythmias; VF, ventricular fibrillation; VT, ventricular tachycardia; WCD, wearable cardioverter defibrillator.



inflammatory diseases (e.g. sarcoidosis), coordination with inflammation control is advised.¹⁴⁹

If AV node ablation is part of rate control, the 'cool down, then ablate' principle still applies; post-ablation pacing can follow the CSP consensus (LBBAP commonly preferred; His bundle pacing feasible with attention to thresholds and a possible back-up lead).¹¹⁶ Even in this setting, it is advised to carefully evaluate the risk factors favouring the choice of ICDs over pacemakers (Table of Advice 5).

Summary

Invasive strategies in arrhythmic myocarditis must be precisely timed and substrate-adapted. Catheter ablation is strongly advised in patients with scar-related VT during the cold phase, where outcomes are favourable and substrate clearly defined.^{8,69} Here, EAVM serves both interventional and diagnostic purposes, especially when imaging is inconclusive or EMB targeting is needed. Disease-specific patterns—as seen in sarcoidosis, Chagas cardiomyopathy, or DSP-associated inflammation—require tailored approaches. Life-threatening arrhythmias in the hot phase demand stabilization prior to ablation. Invasive procedures should be performed in experienced centres under multi-disciplinary guidance. The ESC 2025 guidelines are non-prescriptive for ablation in active disease¹; our 'cool

down, then ablate' approach is consensus-based and safety-oriented. See [Supplementary material online, Tables S6.1–S6.3](#) and Table of Advice 6 for detailed advices to support the clinical decision-making.

Chapter 7: surveillance—follow-up and long-term monitoring

How closely and how long to monitor effectively therapy

Long-term rhythm surveillance in myocarditis requires a structured, risk-adapted strategy that reflects the dynamic course of inflammation, scar evolution, and arrhythmic progression. This chapter provides phase-specific advices for follow-up intervals, remote monitoring, pharmacologic therapy reassessment, and device downgrading. [Supplementary material online, Tables S7.1–S7.4](#) summarize key indicators and interventions.

Structured follow-up and rhythm monitoring

According to the ESC Guidelines,^{1,6,12,25,77} longitudinal follow-up should include repeat assessment of rhythm status, mechanical function, and fibrosis burden. Follow-up intervals typically include 3, 6, and 12 months (see [Supplementary material online, Table S7.1](#)), depending on disease activity and clinical course.¹ In patients with recent myocarditis, restriction from

Advice	Phase	Category of evidence	Strength of evidence	Vote (%)	References
Patient selection and Timing					
Follow the 'cool down, then ablate' principle, and postpone VT ablation to the cold phase of myocarditis to target monomorphic VTs.	Cold			100	- Peretto <i>et al.</i> 2025⁸ - Peretto <i>et al.</i> 2020⁶⁹
Avoid VT ablation in acute/fulminant myocarditis, unless it is needed as a rescue procedure for refractory electrical storms.	Hot			100	- Peretto <i>et al.</i> 2025⁸ - Lenarczyk <i>et al.</i> 2024⁹³
Prefer ablation in patients with occurrence of VT during the cold phase, despite optimal medical treatment including betablockers and/or antiarrhythmics.	Cold			100	- Peretto <i>et al.</i> 2025⁸ - Peretto <i>et al.</i> 2020⁶⁹
The use of VT ablation in replacement for ICD is not advised as a standard practice.	Cold			100	- Peretto <i>et al.</i> 2025⁸ - Zeppenfeld <i>et al.</i> 2022⁷⁷
Prefer PVC ablation in patients with monomorphic high-burden ectopy causing symptoms or PVC-induced cardiomyopathy, when medical treatment is not effective, not tolerated, or not desired.	Cold			100	Cronin <i>et al.</i> 2019⁶⁸
Avoid ablation in patients with polymorphic PVCs during active myocarditis, and prefer immunosuppression as part of medical treatment instead.	Hot, hot-to-cold			100	Lakkireddy <i>et al.</i> 2019^{data-ode-nwrap=100}
Prefer ablation of atrial fibrillation in patients with symptomatic and drug-refractory arrhythmia during the chronic stage of myocarditis.	Cold			100	Weng <i>et al.</i> 2020¹⁴⁹
Avoid ablation in patients with acute-phase atrial fibrillation, and prefer immunosuppression as part of medical treatment instead.	Hot, hot-to-cold			95	- Peretto <i>et al.</i> 2025⁸ - Peretto <i>et al.</i> 2020⁹⁹
Techniques for VT ablation					
Use endo-epicardial approach in patients with myocarditis, especially when VT morphology and/or imaging identifies an epicardial or deep substrate.	Cold			95	Peretto <i>et al.</i> 2020⁶⁹

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Table of Advice 6 Continued

Advice	Phase	Category of evidence	Strength of evidence	Vote (%)	References
Use high-density mapping catheters to characterize border-zone circuits typical of inflammatory scars.	Cold			85	• Cronin et al. 2019 ⁶⁸
Define ablation endpoints as both: 1) non-inducibility of clinical VTs; and 2) substrate homogenization by abolition of LPs/LAVAs.	Cold			90	• Cronin et al. 2019 ⁶⁸
Perform VT ablation at experienced centres with available resources, including EP expertise and mechanical circulatory support (i.e. VA-ECMO) for unstable arrhythmias.	All			100	• Zeppenfeld et al. 2022 ⁷⁷ • Lenarczyk et al. 2024 ⁹³

ICD, implantable cardioverter defibrillator; LAVA, local abnormal ventricular activity; LP, late potential; OBS, observational study-based evidence; OPN, expert opinion-based evidence; PVCs, premature ventricular complexes; VA, ventricular arrhythmias; VA-ECMO, venoarterial extracorporeal membrane oxygenator; VF, ventricular fibrillation; VT, ventricular tachycardia.

competitive or high-intensity sports is generally advised for 3 to 6 months,¹ with individualized reassessment based on imaging and clinical stability. Patients with prior sustained VA,^{53,108} reduced LVEF at presentation^{150,151} or discharge,⁶ persistent atypical LGE (e.g. extensive, septal/anteroseptal, ring-like) on CMR,^{13,60,67,113} or high-risk genotypes (e.g. LMNA, DSP, FLNC)^{12,13,28} require intensive follow-up.

As recommended by the ESC Guidelines on Myocarditis and Pericarditis,¹ reassessment during follow-up should include cardiac biomarkers, ECG, echocardiogram, rhythm monitoring, functional tests, and restaging of myocarditis by gold standard techniques. Remission of myocarditis is suggested by symptom relief, normalization of cardiac biomarkers, absence of residual LV dilation or dysfunction, and clearance of arrhythmias.¹ Phase-adapted surveillance of rhythm disorders through serial ECGs, Holter or ILR monitoring is central. Polymorphic VAs may reflect residual inflammation, while monomorphic VT suggests either fixed substrate or hot-to-cold transition phase.⁴⁻⁶ ILR carriers benefit from remote telemetry, with alerts for bradycardia <40 bpm, pauses >6 s, or asymptomatic AF >30 s.¹⁵² CMR follow-up within 6 months evaluates the resolution or persistence of LGE and T2 abnormalities (hot-to-cold transition phase), which informs both arrhythmic risk and the safety of re-engaging in physical activity.¹⁰⁷ In the presence of ICDs or other limitations to CMR, FDG-PET scan and biomarker reassessment may help in disease monitoring.⁶⁴ EMB may be repeated in selected cases to guide critical therapeutic choices as immunosuppression.⁸⁹

Monitoring antiarrhythmic therapy

Antiarrhythmics use requires structured safety surveillance. [Supplementary material online, Table S7.2](#) outlines specific monitoring for agents such as amiodarone, sotalol, flecainide, and mexiletine.^{1,94} Indications should be re-evaluated regularly,

particularly in the setting of changing LVEF, arrhythmia morphology, or suspected drug toxicity. Antiarrhythmics tapering may be considered in the long term in patients with resolved inflammation, low-risk substrate, and absence of residual VA, but only after shared decision-making and re-evaluation via rhythm and imaging surveillance.

Activity guidance, return to sport, and pregnancy

Physical activity is strongly discouraged during the hot and hot-to-cold transition phases.^{1,107} Clearance requires: resolution of symptoms, troponin and inflammatory markers, normalization or near-complete LVEF recovery, absence of arrhythmias on extended monitoring, and remission of myocarditis with or possibly without residuals.¹ This phase-aware clearance pathway complements the 2025 ESC Guidelines on Myocarditis and Pericarditis emphasis on structured follow-up and individualized return to exercise by adding arrhythmia-specific gates (no symptoms, normalized troponin and natriuretic peptides, normal LVEF, no VT/NSVT on Holter and exercise testing, T2/FDG quiescence, stable/non-progressive LGE).¹ Persistent LGE alone is not an absolute contraindication, but mandates individualized discussion; atypical LGE (extensive, septal/anteroseptal, or ring-like), EAVM-confirmed low-voltage areas and lack of VA suppression with exercise argue against competitive sport until stability is confirmed on serial CMR and continuous rhythm monitoring by ILR in selected cases.^{13,55,153} Documenting the absence of high-risk genotype is not a current requirement, but may help the decision making when HPGC is suspected.⁴¹ The 2025 ESC Guidelines on Myocarditis and Pericarditis suggest a personalized approach for activity guidance, with athletic training and competitive sport generally withheld for 1–6 months.¹ In myopericarditis (pericarditis-predominant, preserved LVEF, minimal myocardial LGE, no VA), return to play is primarily

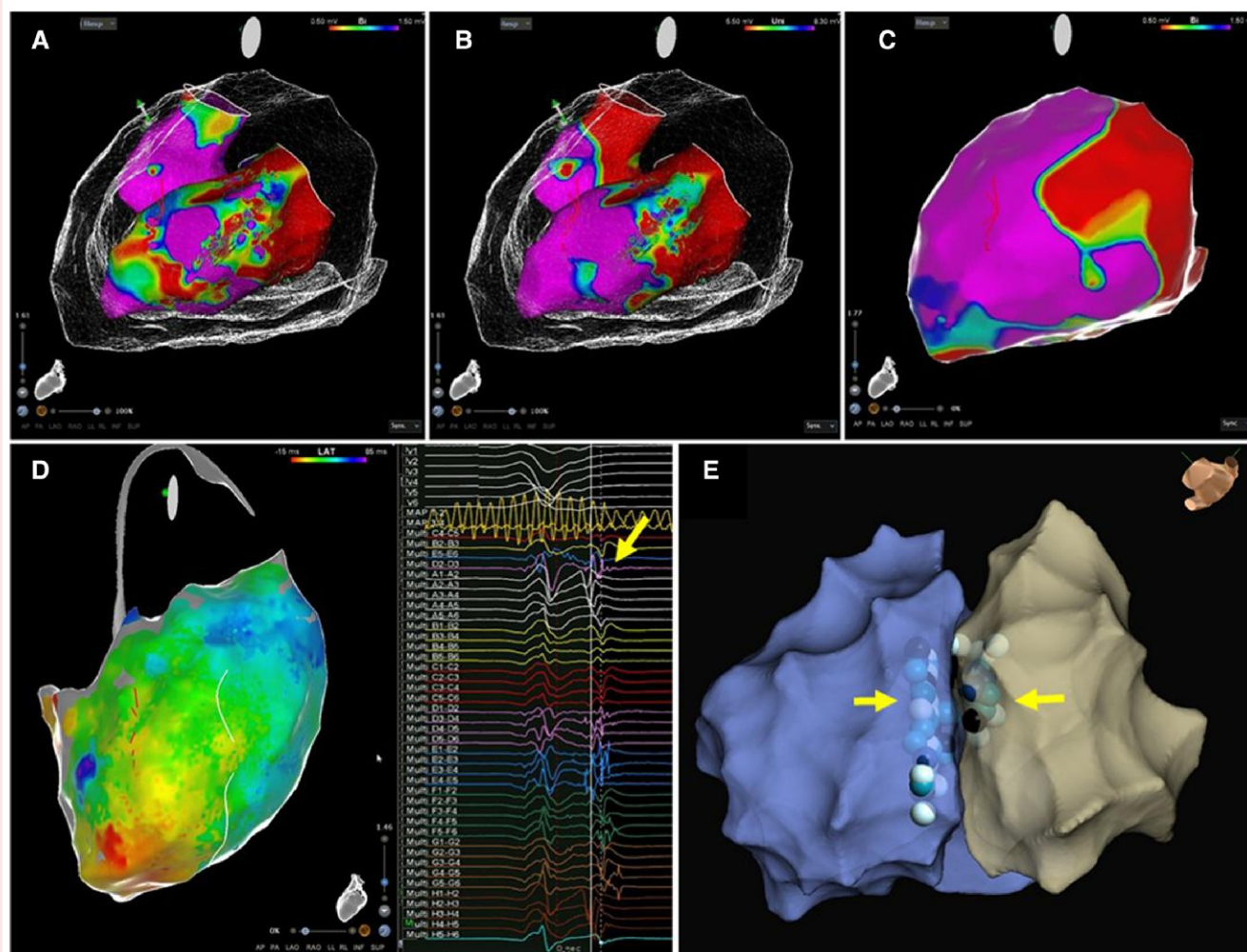


Figure 9 Findings during VT ablation in cold-phase myocarditis. Findings of VT ablation are shown for patients with myocarditis. A-C. EAVM in a patient with a history of myocarditis and extensive inferolateral LV scar. Dense scar (red) and borderzone (rainbow) are compared as recorded by endocardial bipolar (A), endocardial unipolar (B) and epicardial bipolar map (C). (D) Late potentials (arrow) in a patient with chronic myocarditis (cold phase) and inferolateral scar. (E) Ablation lesions (arrows) at both sides of interventricular septum in a patient with giant cell myocarditis and multiple septal VTs refractory to triple immunosuppression. EAVM, electroanatomic voltage mapping; VT, ventricular tachycardia.

pericarditis-led and may be shorter than arrhythmic myocarditis, where a more conservative strategy is generally advised. Women in childbearing age should receive appropriate counselling regarding allowed drugs, risks, and delivery mode.¹⁰² All patients should be educated on warning symptoms and be offered fast-track reassessment for symptom relapse.

Long-term management: device and pharmacotherapy reassessment

Beyond the acute phase, patients may enter a phase of clinical stability—raising questions about therapy duration and device permanence. In patients, arrhythmia-free for ≥ 5 years without LGE or high-risk genotype, downgrade/explantation may be considered as an expert consensus strategy beyond guidelines and following multidisciplinary assessment (see [Supplementary material online, Table S7.4](#)). Medical therapy may be cautiously tapered in recovered patients without fibrosis or genetic

predisposition. However, ongoing treatment remains the default in individuals with persistent LGE, incomplete LVEF recovery, or underlying HPGC. Patients receiving immunosuppressive therapy should undergo close multidisciplinary surveillance to review medications, evaluate toxicity, and ensure protection from infections.^{3,96} Treatment duration may be longer in patients with systemic rheumatologic diseases or HPGC.^{26,105}

Summary

A structured follow-up strategy—integrating rhythm surveillance, imaging, and functional assessment—is essential to ensure patient safety, prevent relapse, and guide lifestyle changes. [Supplementary material online, Tables S7.1–S7.4](#) offer practical frameworks for clinical follow-up, drug monitoring, return-to-activity decisions, and long-term device strategies. Intensive rhythm surveillance is essential in patients with

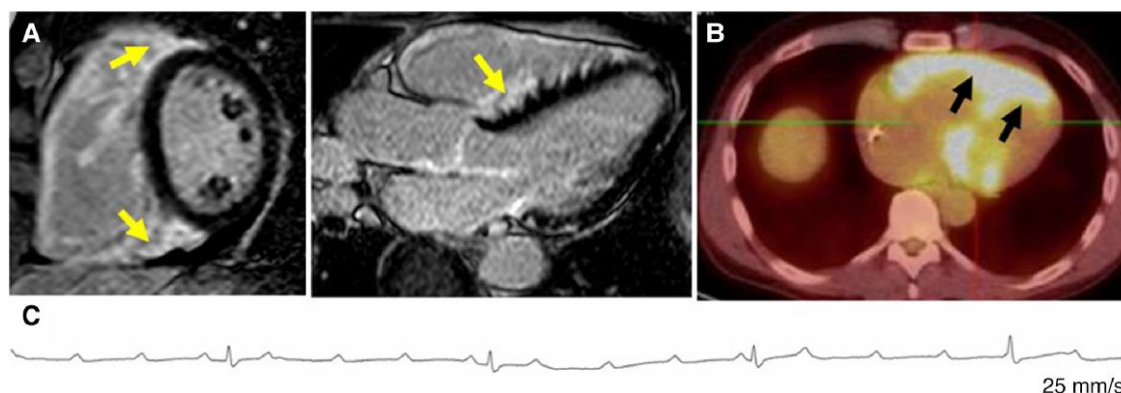


Figure 10 CMR (A) demonstrating septal and right ventricular late gadolinium enhancement (LGE); FDG-PET (B) showing extensive myocardial inflammation (arrows) and mediastinal/hilar lymph node uptake, consistent with cardiac sarcoidosis presenting with high-degree AV block (C).

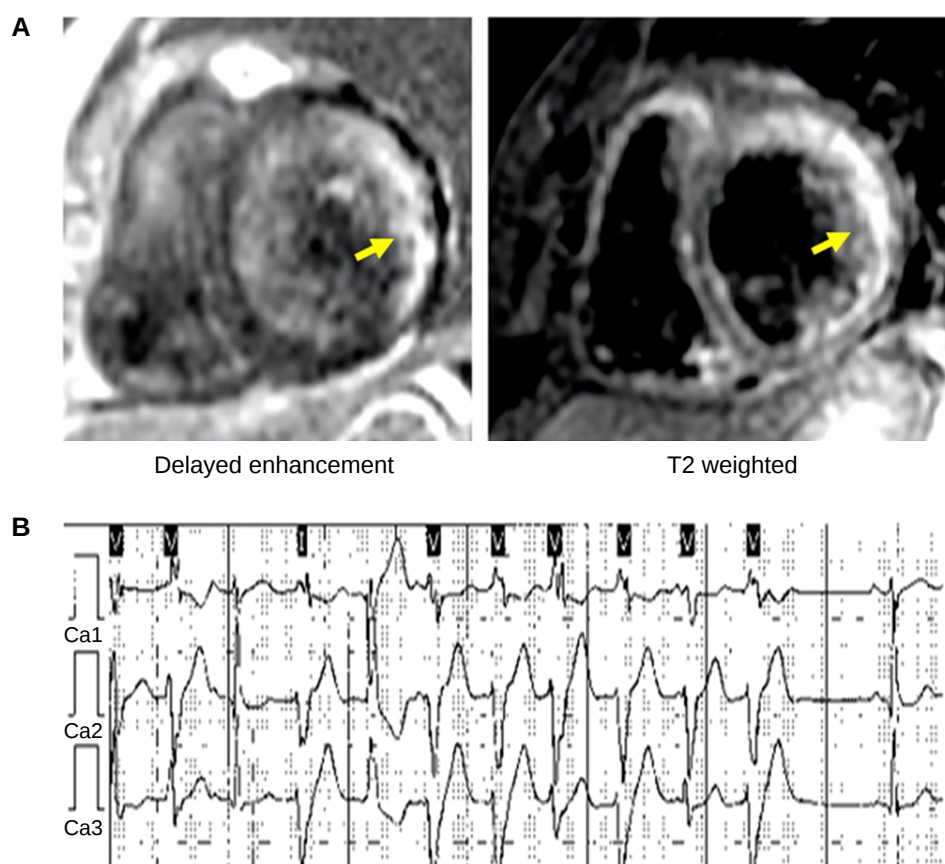


Figure 11 CMR (A) revealing extensive lateral wall LGE and myocardial oedema on T2-weighted imaging; acute inflammation identified as the underlying substrate for polymorphic non-sustained ventricular tachycardia (B).

persistent inflammation, fibrosis, or high-risk genotypes. The possibility of downgrading or explanting devices should be weighed carefully, guided by arrhythmic silence, imaging

stability, and genetic context. Reassessment of therapy duration—both pharmacologic and device-based—should be embedded in a personalized, multidisciplinary care model.

Table of Advice 7 Return-to-play, lifestyle, and follow-up

Advice	Phase	Category of evidence	Strength of evidence	Vote (%)	References
Plan patient multidisciplinary follow-up by in-person visits and/or remote telemedicine consultations, according to the ESC schedule (exams and timing).	All			100	- Schulz-Menger et al. 2025¹ - Peretto et al. 2020⁵¹
Evaluate arrhythmia burden and trends during follow-up using 12-lead ECG Holter, or ILR depending on phase and symptoms.	All			100	Peretto et al. 2021⁵⁵
Repeat CMR (or FDG-PET if CMR is limited) by 6 months to confirm transition to cold phase.	Hot-to-Cold			95	- Schulz-Menger et al. 2025¹ - Peretto et al. 2022⁶⁴
Favour return-to-play with an individualized approach, upon verification of all safety gates: - symptoms remission; - normalized biomarkers; - stable/normalized LVEF; - no documented VT/NSVT by EST and rhythm monitoring; - absence of inflammation (CMR T2/FDG); - stable/non-progressive LGE.	Hot-to-Cold, Cold			95	- Schulz-Menger et al. 2025¹ - Pelliccia et al. 2019¹⁰⁷
Periodically reassess sports/occupation eligibility and ICD indication as substrate stabilizes.	Cold			100	Pelliccia et al. 2019¹⁰⁷
Educate on warning symptoms (syncope, palpitation, chest pain recurrence, exercise intolerance) and favour rapid access pathway for evaluation.	All			100	

CMR, cardiac magnetic resonance; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; ESC, European Society of Cardiology; EST, exercise stress test; FDG-PET, 18F-fluorodeoxyglucose positron emission tomography; ICD, implantable cardioverter defibrillator; OBS, observational study-based evidence; OPN, expert opinion-based evidence.

Tailored return-to-activity guidance and periodic rhythm re-evaluation help prevent recurrence and support functional recovery. Multidisciplinary coordination between heart failure, electrophysiology, imaging, and immunology teams is critical for optimizing long-term outcomes.^{1,51}

Chapter 8: application of clinical case scenarios

How does it work in real life?

This chapter presents eight clinical vignettes that illustrate how rhythm disturbances in myocarditis and inflammatory cardiomyopathy are managed in real world setting. These vignettes focus on arrhythmia-predominant myocarditis where the ESC Guidelines¹ necessarily provide a framework but non-prescriptive steps. The clinical cases are meant to illustrate individualized, phase-adapted diagnostic and

treatment strategies, intentionally spanning complex scenarios where expert advice may help in clinical decisions. For challenging clinical questions (Q), the degree of concordance among experts is shown in ad-hoc consensus footers.

Case 8.1: High-degree AV block in a young patient: unmasking inflammatory cardiomyopathy

A 43-year-old man presented with fatigue and abdominal discomfort. ECG revealed a complete AV block (third-degree). Troponin T was mildly elevated; CMR showed oedema and sub-epicardial LGE in the septum and RV (see Figure 10) (Q.8.1.1). FDG-PET confirmed inflammation. EMB revealed multinucleated giant cells, consistent with GCM. An ICD was implanted (Q8.1.2), and immunosuppression initiated (Q8.1.3) with corticosteroids (prednisone 1 mg/kg/day, oral, then tapered),

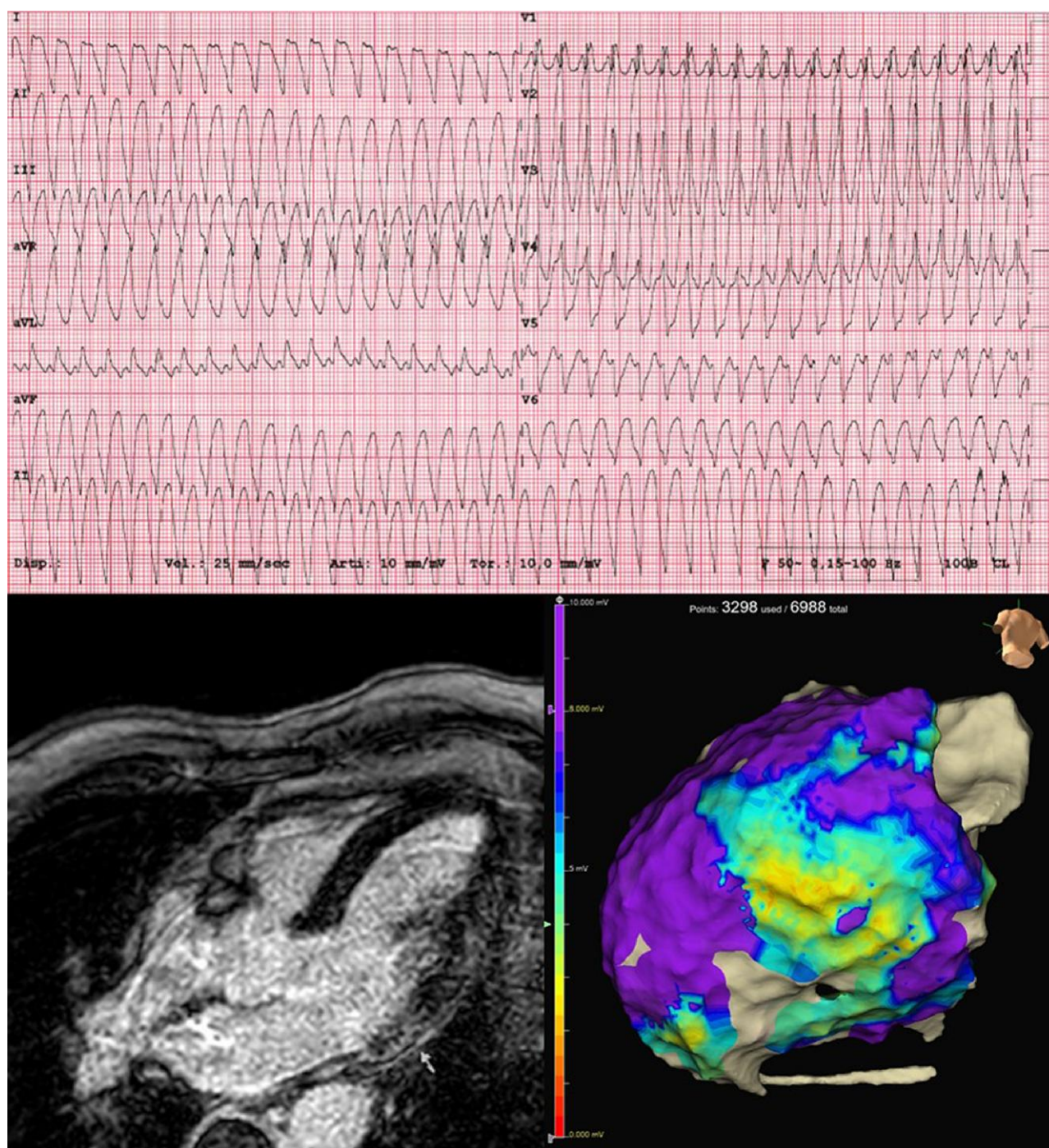


Figure 12 Monomorphic VT documented on 12-lead ECG (A). CMR without oedema but with subepicardial LGE (B), indicating a chronic scar-related re-entrant substrate. High-density epicardial voltage mapping of the lateral left ventricle (C) showing extensive low-voltage areas (<1.5 mV); Genetic testing was planned.

mycophenolate mofetil (2.5 g day, oral), and cyclosporine (300 mg day, oral) for 6 months.

Consensus footer:

Q8.1.1	Next exam after CMR?	EMB (47%), PET (41%), genetic test (12%)
Q8.1.2	Device type?	PM (6%), TV-ICD (75%), WCD (19%)
Q8.1.3	IST indicated?	Yes (100%), No (0%)

Rationale: This case illustrates the typical presentation of acute myocarditis with high-grade AV block. Inflammatory infiltration of the conduction system is frequently observed in GCM and sarcoidosis (see Chapters 1.2 and 1.3). The indication for permanent pacing or ICD therapy should be guided by the underlying disease subtype and associated arrhythmic risk (see [Supplementary material online, Table S5.1](#)). In GCM, even transient AV block may herald malignant arrhythmias and frequently requires permanent ICD implantation.

Key message: AV block in younger patients may reflect sarcoidosis or GCM. CMR, PET, and EMB are essential; early ICD and tailored immunosuppression are often required.

Case 8.2: Atrial fibrillation in systemic rheumatologic disease with chronic myocarditis

A 38-year-old woman with systemic lupus erythematosus (SLE) developed frequent atrial fibrillation episodes. EMB had previously confirmed lupus myocarditis, active stage. Successful rhythm control was initially obtained by corticosteroids, methotrexate, and metoprolol. A few years later, atrial fibrillation recurred. CMR showed no gross oedema or dense LGE (Q8.2.1). Catheter ablation was performed (Q8.2.2); voltage mapping showed preserved atrial substrate. In light of the persistence of sporadic palpitation an ILR was implanted (Q8.2.3); direct oral anticoagulant initiated.

Consensus footer:

Q8.2.1	EMB needed as well?	Yes (41%), No (59%)
Q8.2.2	Would have started antiarrhythmic drugs before ablation?	Yes (76%), No (24%)
Q8.2.3	Follow-up by Holter ECG instead?	Yes (53%), No (48%)

Rationale: Autoimmune myocarditis often affects the atria and may lead to atrial fibrillation or flutter, especially in systemic inflammatory diseases such as SLE (see Chapters 1.2 and 1.3). Rhythm

monitoring and recurrence risk should guide decisions on ILR implantation or ablation strategies (see [Supplementary material online, Table S6.3](#)). Structured follow-up with device-based monitoring is advised in patients with persistent inflammation or recurrent SVA (see [Supplementary material online, Table S7.3](#)).

Key message: Autoimmune-mediated atrial fibrillation may require ablation despite immunosuppressive therapy. ILRs support rhythm monitoring; therapy must address both systemic and cardiac inflammation.

Case 8.3: Polymorphic ventricular tachycardia in active myocarditis

A 28-year-old man presented with syncope and polymorphic VT (see [Figure 11](#)). Coronary arteries were normal. CMR showed oedema and inferolateral LGE. EMB confirmed acute, virus-negative lymphocytic myocarditis (Q8.3.1). He received corticosteroids (prednisone 1 mg/kg/day tapered down over 3 months) and mofetil mycophenolate (titrated up to 3 g day for 12 months), and a WCD was prescribed (Q8.3.2). At 6 months, CMR showed LGE without residual abnormality in T2-weighted sequences, systolic function was normal in both ventricles, and both exercise stress test and Holter ECG showed no arrhythmias. Genetic testing was negative. WCD was removed (Q8.3.3). No proven or suspected VA recurrences were observed in the long term.

Consensus footer:

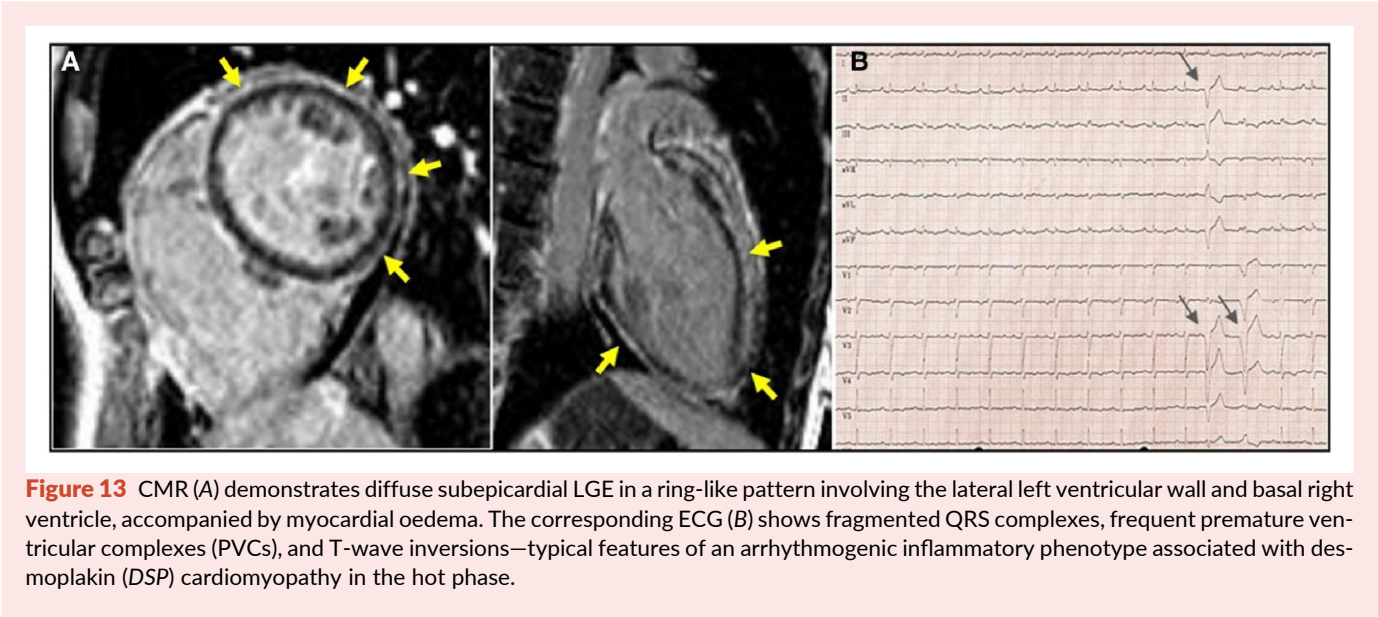
Q8.3.1	Would have performed EMB to enable safe use of IST?	Yes (71%), No would have started betablocker/antiarrhythmic therapy without EMB (29%)
Q8.3.2	Agree on choosing WCD?	Yes (94%), No, would have chosen ICD instead (6%)
Q8.3.3	Safe to remove WCD at month 6?	Yes (65%), No, would reassess risk by repeating EMB (6%), No, would reassess risk by PVS (29%)

Rationale: Polymorphic VT or electrical storm in acute myocarditis typically reflects active inflammation and substrate instability (see Chapters 1.3 and 3.1). In virus-negative cases, immunosuppressive therapy should be initiated alongside rhythm control (see [Supplementary material online, Table S3.1](#) and [S3.3](#)). Ablation is not advised in the hot phase unless life-threatening instability persists despite optimized medical therapy (see [Supplementary material online, Table S6.1](#)). Temporary protection with a WCD or mechanical support may be needed (see [Supplementary material online, Table S5.1](#)).

Key message: Inflammatory polymorphic VT requires early immunosuppression. WCDs mitigate transient SCD risk in reversible phases.

Case 8.4: Scar-related ventricular tachycardia in healed myocarditis

A 66-year-old man with prior EMB-proven lymphocytic myocarditis presented with monomorphic VT (see [Figure 12](#)) despite ongoing treatment with metoprolol 200 mg/day and



amiodarone 200 mg/day. CMR showed dense subepicardial scar without oedema; FDG-PET was negative. Mapping confirmed a stable re-entry circuit. Ablation and secondary prevention ICD were performed (Q8.4.1–8.4.3).

Consensus footer:

Q8.4.1	Right timing for VT ablation?	Yes (88%), No, would have repeated EMB to start immunosuppression instead (12%)
Q8.4.2	Agree with ablation first, ICD next?	Yes (82%), No, would have implanted ICD first (18%)
Q8.4.3	Would have suggested ICD even in the event of a prior successful VT ablation?	Yes (88%), No (12%)

Rationale: This patient with recurrent sustained VT and inferolateral scar represents a typical post-inflammatory phenotype with fixed substrate (see Chapters 1.1 and 1.3). Imaging (LGE) and electroanatomical mapping (LVA) guide ablation strategy and ICD indication (see [Supplementary material online, Tables S5.1 and S6.1 and](#)). In stable chronic myocarditis, ICD therapy and targeted ablation can prevent recurrence and improve quality of life. Extensive LVA suggest chronic cardiomyopathic process rather than focal post-inflammatory scarring, and called for ICD implant independently of VT ablation results.

Key message: Cold-phase scar-related VT requires ablation and ICD. Inactive myocarditis does not warrant immunosuppressive therapy.

Case 8.5: Recurrent arrhythmic myocarditis and hot-phase genetic cardiomyopathy

A 32-year-old woman presented with recurrent episodes of chest pain, palpitations, and exertional dyspnoea. Family history was unremarkable. ECG revealed fragmented QRS complexes with low voltages, frequent PVCs, and T-wave inversions. Echocardiogram showed LVEF 50%. CMR revealed subepicardial, ring-like LGE involving the lateral LV and RV basal segments (burden 15%), alongside myocardial oedema (see [Figure 13](#)). EMB confirmed virus-negative inflammatory cardiomyopathy (CD3+ T lymphocytes > 7/mm²). Genetic testing identified a pathogenic DSP variant (Q8.5.1).

During telemetry, frequent non-sustained VT, and frequent PVC (14% burden) were documented. An ICD was implanted for primary prevention of sudden cardiac death (Q8.5.2). Immunosuppressive therapy (Q8.5.3) included prednisone 1 mg/kg plus azathioprine 2 mg/kg per os. Sotalolol, a class III antiarrhythmic with beta-blocking effects, was initiated (80 mg tid, oral) to suppress ventricular ectopy and reduce arrhythmic burden, given the preserved LVEF and absence of contraindications. Given the autosomal dominant inheritance pattern of DSP-related disease, cascade family screening was initiated, including cardiogenetic counselling, ECG, echocardiography, and CMR in first-degree relatives.

Consensus footer:

Q8.5.1	Would have considered genetic testing even if family history was unremarkable and EMB concluded for myocarditis?	Yes, because of LGE pattern on CMR (82%), No, would have managed just as virus-negative myocarditis (18%)
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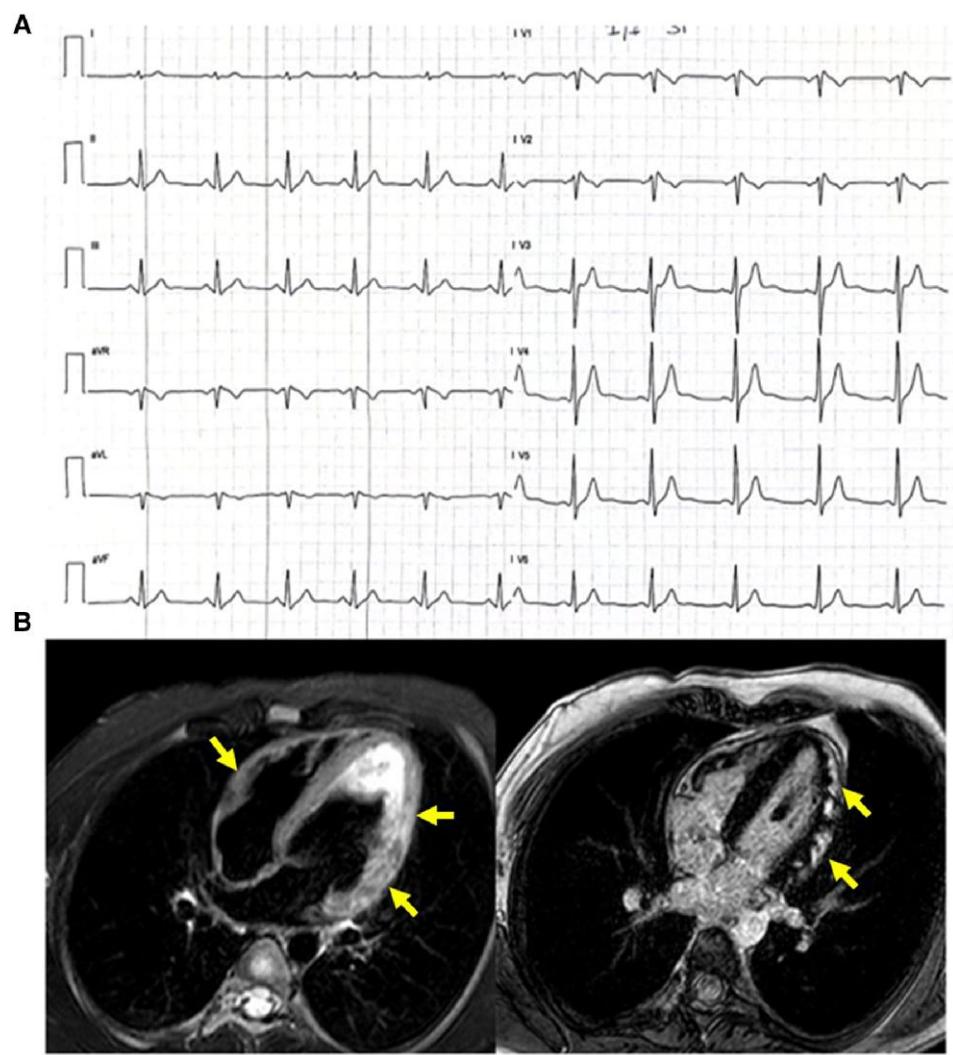


Figure 14 Electrocardiogram (A) showing fever-induced type 1 Brugada pattern; CMR (B) revealing diffuse myocardial oedema of both ventricles and non-ischaemic LGE in lateral LV wall—findings consistent with transient inflammatory modulation of RV conduction.

Continued

Q8.5.2	Would have preferred S-ICD instead of transvenous device?	Yes (41%), No (59%)
Q8.5.3	Would have started immunosuppression despite diagnosis of genetic cardiomyopathy?	Yes, because of EMB findings (58%), No, would have limited to cardiological therapy (42%)

Rationale: As detailed in Chapters 1.1 and 1.2, recurrent myocarditis with ring-like or biventricular LGE is highly suggestive of DSP-related HPGC. DSP variants often mimic ARVC on imaging, but up to one-quarter present with atypical or left-dominant patterns—including global ring-like enhancement and subepicardial

fibrosis—which may mask the underlying genetic aetiology unless specifically considered. This case reflects a high arrhythmic risk profile with troponin elevation, frequent PVCs, fragmented QRS, and subepicardial scar (see [Supplementary material online, Table S1.3](#)). Beta-blockers and amiodarone remain first-line pharmacotherapy (see [Supplementary material online, Table S3.1](#)); sotalol is a cold-phase alternative in patients with preserved function and low QT risk, to avoid long-term toxicity of amiodarone.⁹³ Given the documented NSVT and the DSP genotype, early ICD implantation was justified (see [Supplementary material online, Table S5.1](#)), and lack of risk features for bradyarrhythmias support the choice of S-ICD. Long-term rhythm surveillance and genetic counselling are essential and should follow the risk-adapted strategy outlined in Chapter 7 (see [Supplementary material online, Table S7.3](#)).

Key message: Recurrent myocarditis with ring-like or biventricular LGE may indicate DSP-related hot-phase cardiomyopathy—even in the absence of typical ARVC features. Recognizing these atypical patterns is critical for early risk stratification, ICD implantation, family screening, and individualized therapy.

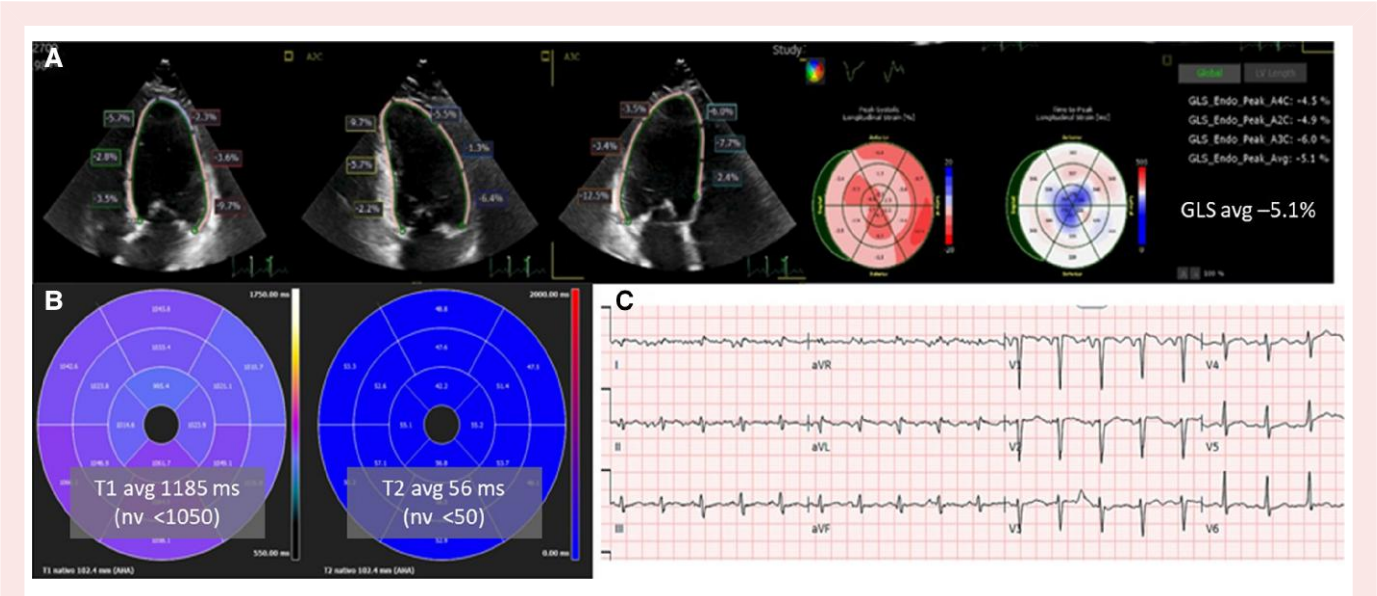


Figure 15 Transthoracic echocardiography (A) with tissue Doppler imaging (TDI) demonstrating severely reduced left ventricular systolic function and elevated filling pressures (E/e' ratio); CMR (B) showing T1 and T2 mapping abnormalities consistent with myocarditis; corresponding ECG (C) with sinus tachycardia. Findings support a diagnosis of inflammatory postpartum cardiomyopathy in the differential with peripartum cardiomyopathy.

Case 8.6: Paediatric myocarditis with Brugada-like ECG pattern

A 14-year-old boy presented with fever and Brugada-like ECG (see Figure 14). He had no family history of sudden death or cardiomyopathy. Biventricular function was normal. The patient had no history of syncope or palpitation. No ventricular arrhythmias occurred. CMR showed diffuse oedema and LV lateral LGE; elevated native T1 and T2 mapping supported the diagnosis of myocarditis (Q8.6.1). The ECG normalized after defervescence. Genetic testing for SCN5A was negative (Q8.6.2). He was discharged with indication to reassessment though Holter ECG and in 3 months (Q8.6.3). At 6 months, CMR showed no oedema and no residual LGE. The patient was readmitted to sport activity.

Consensus footer:

Q8.6.1	Would have performed EMB here?	Yes, to enable safe use of immunosuppression (29%), No, given the absence of treatment endpoints (71%)
Q8.6.2	Would have performed genetic testing?	Yes (35%), Yes, but not limited to SCN5A (47%), No (18%)
Q8.6.3	Agree with rhythm monitoring strategy (Holter ECG)?	Yes (76%), No, would have chosen WCD (12%), No, would have chosen ILR (12%)

Rationale: As discussed in Chapter 1.2, febrile illnesses can transiently unmask Brugada-like ECG patterns due to inflammatory modulation of cardiac sodium channels. This phenomenon is particularly relevant in paediatric myocarditis, where SCN5A mutations may coexist or be mimicked by inflammation. Chapter 1.3 supports a non-invasive, watchful follow-up strategy in children without sustained ventricular arrhythmias, preserved cardiac function, and negative genetic screening. Serial ECGs and CMR (see Supplementary material online, Table S2.1 and S7.1) are critical to confirm recovery. ICD implantation is generally not indicated in the absence of arrhythmic events, structural abnormalities, or genetic predisposition.

Key message: Febrile Brugada-like patterns may reflect transient inflammation. Genetic testing and follow-up help avoid unnecessary ICD in children.

Case 8.7: Arrhythmic myocarditis in the postpartum period

A 36-year-old woman presented 10 days postpartum with heart failure symptoms. Echocardiogram showed severe biventricular dysfunction (LVEF 18%) (see Figure 15). CMR showed no LGE but elevated T1/T2 values (Q8.7.1). EMB revealed lymphocytic inflammatory infiltrates, no fibrosis and no necrosis (borderline myocarditis). Peripartum cardiomyopathy (PPCM) was considered in overlap with myocarditis. Genetic testing excluded cardiomyopathy-associated gene variants. She received heart failure therapy and WCD monitoring (Q8.7.2). At 6 months, LVEF normalized and WCD was withdrawn (Q8.7.3).

Consensus footer:

Q8.7.1	Given CMR results agree on performing EMB and genetic testing?	No, findings were already consistent with PPCM (12%), Yes, only EMB (12%), Yes, only genetic test (23%), Yes, both (53%)
Q8.7.2	Agree on bridging with WCD for 6 months?	Yes (82%), No, would have implanted ICD (6%), No would have follow-up non-invasively by Holter ECG (12%)
Q8.7.3	What would have proposed in the event of failure to normalize LVEF at month 6?	EMB-driven immunosuppression and further reassessment (35%), primary prevention ICD (65%)

Rationale: Postpartum myocarditis may mimic PPCM, but advanced tissue characterization (see Chapter 1.3 and [Supplementary material online, Table S3.4](#)) support differentiation. As outlined in Chapter 3.1 and 3.3, immunosuppression is not routinely indicated in PPCM but may benefit biopsy-proven Infl-CMP. Chapter 5.3 and [Supplementary material online, Table S5.1](#) support WCD use as a bridge in patients with severely reduced LVEF and elevated arrhythmic risk during the hot phase. Serial reassessment (see [Supplementary material online, Table S7.1](#)) ensures safe decision-making regarding defibrillator therapy, especially in reversible cases.

Key message: Postpartum myocarditis may mimic PPCM. Tissue mapping and WCDs support diagnosis and risk management.

Case 8.8: Arrhythmic myocarditis in a competitive athlete

A 29-year-old athlete had syncope after recent SARS-CoV-2 infection. Troponin was mildly elevated, but coronary CT scan was normal. Echocardiogram showed mild biventricular enlargement with normal function, overall consistent with athlete’s heart. CMR showed multiple LGE striae (septal and inferolateral, non-ischaemic pattern) and mild diffuse oedema. After 6 months, oedema resolved, LGE persisted. Genetic testing was negative (Q8.8.1). ILR showed multiple asymptomatic NSVT and the patient refused betablockers (see [Figure 16](#)); PVS induced sustained VT (Q8.8.2). He received ICD. Return to competitive sport was not advised (Q8.8.3).

Consensus footer:

Q8.8.1	At this point (persistent multifocal LGE, history of syncope, normal function, no documented VA, no high-risk genotype) would have chosen ILR?	Yes (69%), No, just Holter ECG by 6 months (0%), No, WCD for at least 3 months (12%), No, ICD implant right now (19%)
Q8.8.2	With multiple but asymptomatic NSVT in cold-phase would have performed PVS?	Yes (69%), No, would have implanted ICD independently of PVS (6%), No would have continue to follow-up (25%)
Q8.8.3	Agree on prohibiting competitive sport?	Yes in any case (56%), No with ICD and yes without (44%), No in any case (0%)

Rationale: As discussed in Chapters 1.2 and 1.3, athletes with myocarditis are at elevated arrhythmic risk, particularly when LGE persists beyond resolution of oedema. In this case, extensive LGE with septal involvement warranted advanced risk stratification with genetic testing, high-sensitivity detection of NSVT by ILR, and programmed ventricular stimulation (see [Supplementary material online, Table S4.1](#) and [Table S5.1](#)). The induction of sustained VT justified ICD implantation for primary prevention. Chapter 7.3 and [Supplementary material online, Table S7.2](#) emphasize that return to competitive sports is contraindicated in the presence of persistent scar, symptomatic arrhythmias, or device therapy due to the risk of sudden cardiac death.

Key message: In athletes with prior myocarditis, persistent/extensive/septal LGE, NSVT, and inducible VT indicate high arrhythmic risk. ICD implantation is required, and return to competitive sports is contraindicated due to the risk of sudden cardiac death.

Closing remarks

Across the clinical cases, the voting results revealed a generally high level of agreement among experts. However, some variation in case-specific responses reflected the inherent complexity of translating theoretical or guideline-based recommendations into individualized patient management. This diversity of expert opinion underscores the need for nuanced decision-making in real-world myocarditis care.

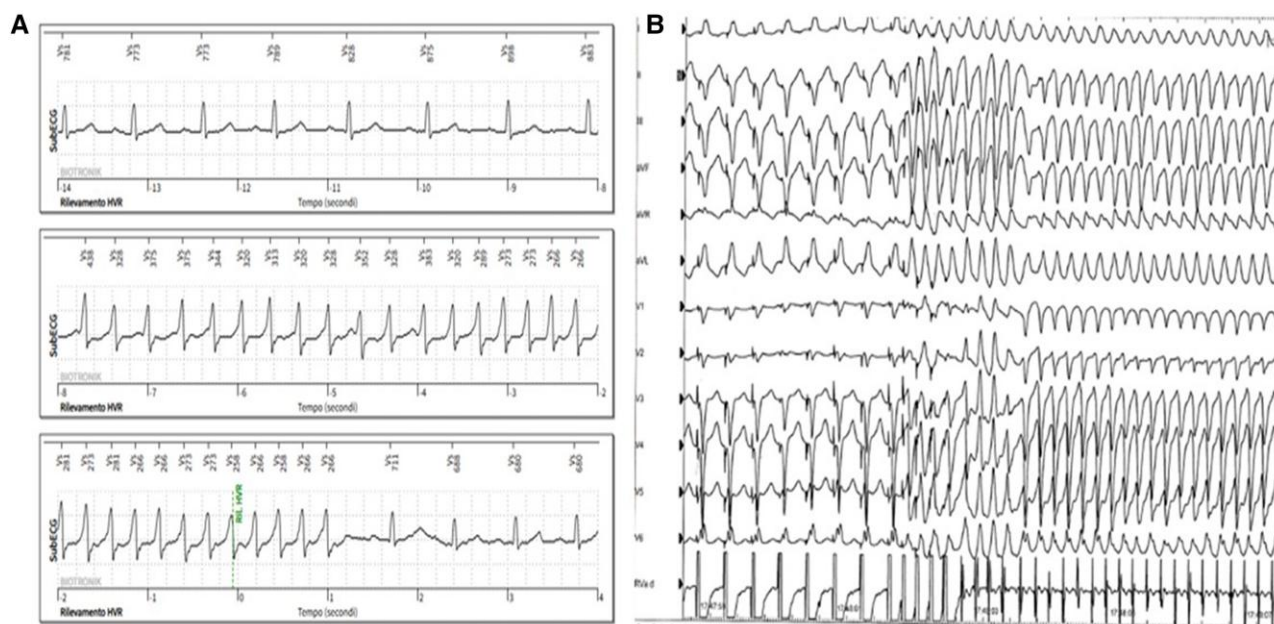


Figure 16 Spontaneous NSVT detected by ILR (A). Programmed ventricular stimulation (B) induced a sustained monomorphic VT with LBBB morphology (atypical for classic inferolateral myocarditis)—supporting chronic cardiomyopathic substrate in the athletic heart.

Chapter 9: perspectives, limitations, and conclusive remarks

Where do we go next

Rhythm disorders in myocarditis and Infl-CMP remain a dynamic and incompletely understood field. This chapter outlines unmet needs, emerging research directions, and future strategies for improving diagnostics, risk stratification, and individualized therapy. These proposals are hypothesis-generating and ESC-concordant,¹ designed to deepen areas where the guideline is concise.

Hot-to-cold transition: a new frontier in arrhythmic risk stratification

The recognition of a distinct hot-to-cold transition phase^{6,8} provides a novel and actionable framework for stratifying arrhythmic risk beyond the ESC acute/non-active dichotomy.¹ Observational data already support the clinical relevance of this phase, particularly in patients discharged with sustained monomorphic VT or reduced LVEF^{6,53}; vulnerability appears substrate-driven (fibrosis/scar) and is not fully mitigated by the resolution of inflammation. In practice, this translates into considering early ICD implant in high-risk cases (Table of Advice 5). In the remaining patients, WCD bridging and deferral of definitive ICD decisions to the cold/non-acute stage is advised—consistent with the ESC Guidelines on Myocarditis and Pericarditis in the setting of acute myocarditis¹—and reflects our view that permanent devices are safer and more informative once the substrate has stabilized. Prospective work should now refine the temporal boundaries of the transition, develop biomarkers of substrate evolution (including LGE topology and burden), and test intervention timing (duration of WCD, thresholds

for ICD, and the role of PVS and ablation after cool-down) in phase-stratified cohorts.

Arrhythmic risk stratification and the role of genetics

The integration of genetic profiling into risk stratification is essential. HPGC—particularly variants in *DSP*, *LMNA*, *FLNC*, other genes listed in [Supplementary material online, Table S2.2](#)—frequently manifest with myocarditis-like flares and VA. In keeping with the 2025 ESC Guidelines on Myocarditis and Pericarditis framework,¹ genetic evaluation is advised when clinical, imaging, or family cues raise suspicion (e.g. recurrent myocarditis, family history of cardiomyopathy/sudden death, exertion-triggered VA, or atypical scar patterns).^{9–11,26–30} In clinical decision-making, genotype functions as a risk modifier: it lowers thresholds for intensified surveillance through the hot-to-cold phase, may justify earlier inherited cardiomyopathy referral and family screening, and can influence indication and timing of interventions. Importantly, genotype is not a stand-alone indication for ICD outside established frameworks. Based on available preliminary evidence,^{26,105} this Task Force prompts the need for further investigation of immunosuppressive therapy in HPGC through dedicated prospective registries and clinical trials. Future studies should disentangle isolated myocarditis from genetically primed inflammatory expression and quantify genotype–substrate interactions across disease phases to refine phase-specific management.

Artificial intelligence and predictive modelling

In parallel with classical risk models, academic initiatives are developing AI tools such as ARHITHMOS, RACE-MYO, and

PROTECT-VT (ARHITHMOS (Academic Risk-based Health Intelligence Tool for Myocarditis and Sudden death) is an AI-driven risk modelling platform developed within a European academic consortium (coordinated by Utrecht Medical Center). RACE-MYO (Risk Assessment in Cardiomyopathy and Myocarditis) and PROTECT-VT (Prediction and Outcome Tools for Early Cardiac Threat—Ventricular Tachycardia) are research networks based in Germany and Israel, respectively, focusing on integrating imaging, ECG, and biomarker data to stratify arrhythmogenic risk in inflammatory cardiomyopathies, that integrate ECG features, cardiac imaging (including LGE extent and topology), serum biomarkers, and genomics into individualized risk trajectories. These tools remain investigational and lack large-scale external validation; clinical use is still exploratory. Priorities include open-access validation, transparent calibration and drift monitoring, standardized inputs (e.g. ECG formats, LGE quantification), and integration into audited decision-support with human oversight. Of particular relevance is reliable detection of phase transitions—notably entry into the hot-to-cold stage—together with early warning for VT/VF recurrence and prediction of therapy response. Consistent with the 2025 ESC Guidelines on Myocarditis and Pericarditis,¹ which emphasize established diagnostic and follow-up pathways and do not issue specific recommendations for AI-based risk prediction, such tools should enter routine care only after rigorous prospective evaluation

Novel immunomodulatory therapies

Within the ESC 2025 Guidelines on Myocarditis and Pericarditis context¹—biopsy-guided immunosuppression in virus-negative disease and no arrhythmia-directed recommendations—there remains a critical unmet need to determine whether targeted suppression of active myocardial inflammation can terminate malignant ventricular arrhythmias even with preserved LVEF, and prevent substrate maturation during the hot/hot-to-cold phases. In arrhythmia-predominant phenotypes, immunosuppression should be selective and management-changing (e.g. biopsy-proven autoimmune disease, relapsing inflammatory VT), and undertaken under multidisciplinary oversight in experienced centres.^{39,96,99} Priority questions are who should be treated (virus-negative patients with objectively or strongly suspected activity by CMR T2/FDG and/or recent histology), when (early, phase-aware initiation with WCD bridging and ICD deferral unless secondary prevention is mandatory), and what to use (short steroid pulse with structured taper vs. standard care; addition of steroid-sparing agents such as azathioprine/mycophenolate—or methotrexate in sarcoidosis; biologics targeting IL-1/JAK strictly investigational). In acute relapsing VT/VT-storm, where immediate EMB/CMR may be impractical, a time-limited, closely monitored trial in expert centres—after exclusion of infection and with pre-specified arrhythmic and imaging endpoints—may be justified. The current evidence for arrhythmia reduction is largely observational and requires prospective confirmation; hence prospective, phase-stratified studies are needed to define efficacy, safety, and the role of specific drug classes in arrhythmogenic myocarditis.

Arrhythmia-specific clinical trials and registries

A critical gap is the paucity of trials with arrhythmia-focused endpoints. Future studies should mandate rhythm outcomes (VT/VF recurrence, appropriate device therapies, sudden cardiac death), use adjudicated core endpoints, and stratify by

phase (hot vs. hot-to-cold vs. cold) to reflect the evolving substrate. Phase-stratified registries should also capture myocarditis cases requiring pacing to compare CSP vs. RVP/BiV-CRT in inflammatory conduction disease (patient-centred outcomes, LV function, VA burden). Harmonized definitions and multicentre registries linked to biobanked genotype, imaging (including LGE topology/burden), and histology will enable phenotype-specific analyses and robust meta-analyses across centres. This strategy is consistent with the 2025 ESC Guidelines' emphasis on standardized pathways and collaborative data infrastructures,¹ while adding the phase-aware, arrhythmia-specific detail not prescriptively defined in the guideline text.

Inflammation resolution and risk persistence

Long-term arrhythmic risk may persist despite apparent resolution of inflammation. This is especially true in patients with persistent LGE, non-recovery of LVEF, or genetic predisposition.^{1,12} Prospective, phase-stratified studies should explicitly evaluate IMPS, testing whether myopericarditis without myocardial 'flags' can safely follow shorter, pericarditis-led surveillance, while perimyocarditis adheres to myocarditis pathways. ILRs, periodic imaging, and advanced rhythm analytics (e.g. frequency-domain analysis, AI telemetry) should guide follow-up in high-risk individuals. A re-evaluation of ICD indications and sport eligibility over time is also warranted.

Refinement of classification systems

Current classifications do not fully capture the breadth of arrhythmic myocarditis nor its overlap with inflammatory cardiomyopathies. A phase-aware framework that maps the ESC acute/non-acute scheme onto hot/hot-to-cold/cold would sharpen clinical decision-making and improve trial eligibility in arrhythmia-predominant phenotypes across IMPS (myopericarditis to perimyocarditis). Future frameworks should distinguish:

- Purely inflammatory forms vs. genetically primed inflammation
- Arrhythmia-predominant vs. heart failure-dominant phenotypes
- Scar-mediated vs. inflammation-triggered arrhythmias

Such granularity is essential for rational therapy selection and meaningful study enrolment.

Conclusion

This consensus sits alongside and operationalizes the 2025 ESC Myocarditis Guidelines. Where the guidelines provide clear direction, we align and cite ESC recommendations as context; where arrhythmia-specific gaps persist—particularly around risk evolution and substrate dynamics—we offer expert consensus advice, transparently graded (OPN/OBS/RCT) and supported by author voting (see Tables of Advice). To make guideline principles actionable in rhythm care, we overlay a phase-aware lens—hot, hot-to-cold, cold—on the ESC acute/non-acute categorization, providing the additional granularity needed for device timing, return-to-play, ablation strategy, rhythm surveillance, and selective EMB decisions.

Looking ahead, a translational agenda is required to convert this framework into definitive practice. Priorities include validating the hot-to-cold phase (temporal boundaries, LGE topology/burden, biomarkers of substrate evolution); testing WCD-bridging vs. earlier ICD, the timing and yield of PVS in non-acute disease, and the 'cool-down, then ablate' strategy; evaluating whether targeted, biopsy/imaging-guided immunosuppression can reduce the burden of malignant VA in virus-negative patients with documented activity (including

'hot-phase' HPGC cohorts); and externally validating phase-aware AI that detects transitions and forecasts VT/VF using standardized inputs and audited decision support. Multicentre, phase-stratified registries and trials with adjudicated arrhythmia endpoints—linked to biobanked imaging, histology, and genotype—should underpin this work. We envisage periodic updates of this position as far as new evidence emerges.

Supplementary material

Supplementary material is available at *European Journal of Heart Failure* online.

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