EHRA/HRS/APHRS expert consensus on ventricular arrhythmias

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Introduction

This international consensus statement of the European Heart Rhythm Association (EHRA), Heart Rhythm Society (HRS), and Asia Pacific Heart Rhythm Society is intended to provide clinical guidance for the management of patients with ventricular arrhythmias (VAs). It summarizes the consensus of the international writing group members and is based on a systematic review of the medical literature regarding VAs.

The spectrum of VAs ranges from those that are benign and asymptomatic to those that produce severe symptoms including sudden cardiac death (SCD). In addition, many patients exhibit multiple forms of VAs over time. Thus, clinicians who encounter patients with VAs face important questions regarding which diagnostic tests are needed and which treatments, if any, should be offered. The Writing Committee recognizes that the manner in which patients present with VAs varies greatly. The electrocardiographic recording of a VA may be the first and only manifestation of a cardiac abnormality; alternatively, patients with a prior diagnosis of cardiac disease may later develop these arrhythmias. Thus, the specific arrhythmia and the underlying structural heart disease (SHD), if any, may have important prognostic and treatment implications.

This document addresses the indications for diagnostic testing, the present state of prognostic risk stratification, and the treatment strategies that have been demonstrated to improve the clinical outcome of patients with VAs. In addition, this document includes recommendations for referral of patients to centres with specialized expertise in the management of arrhythmias. Wherever appropriate, the reader is referred to other publications regarding the indications for implantable cardioverter-defibrillator (ICD) implantation, catheter ablation, inherited arrhythmia syndromes, congenital heart disease (CHD), the use of amiodarone, and the management of patient with ICD shocks, syncope, or those nearing end of life. The consensus recommendations in this document use the standard Class I, IIa, IIb, and III classification and the corresponding language: ‘is recommended’ for Class I consensus recommendation; ‘can be useful’ for a Class IIa consensus recommendation; ‘may be considered’ to signify a Class IIb consensus recommendation; ‘should not’ or ‘is not recommended’ for a Class III consensus recommendation (failure to provide any additional benefit and/or may be harmful). The level of evidence supporting these recommendations is defined as ‘A’, ‘B’, or ‘C’ depending on the number of populations studied, whether data are derived from randomized clinical trials, non-randomized...
Expert consensus recommendations on ventricular arrhythmias

For treatment of patients with non-sustained VAs, we propose the following consensus recommendations.

Premature ventricular complexes

Premature ventricular complexes (PVCs) are common both in patients with and without SHD and may be asymptomatic even for patients with high frequency of these beats. Other patients may be highly symptomatic with relatively few ectopic beats. Although a recent meta-analysis of patients without clinically apparent SHD demonstrated an increased incidence of adverse events in patients with frequent PVCs, only one of the included studies used echocardiography to establish structural disease. The independent prognostic importance of PVCs in the presence of structural disease is not clear. Early studies demonstrated an association...
with increased cardiovascular mortality after MI and with increased total mortality in patients with LV hypertrophy (LVH). However, these studies were observational and performed in an era prior to modern management. In a study of patients with congestive heart failure [ejection fraction (EF) < 35%], PVC frequency did not predict the risk of sudden death and did not provide prognostic information beyond other clinical variables.

**Premature ventricular complex-induced cardiomyopathy**

Several studies have demonstrated an association between frequent PVCs and a potentially reversible cardiomyopathy, which in selected patients resolves after catheter ablation. The number of PVCs/24 h that is associated with impaired LV function has generally been reported at burdens above 15–25% of the total cardiac beats, though this may be as low as 10% (Table 1). However, since PVCs may be the result of an underlying cardiomyopathy, it may be difficult to prospectively determine which of these sequences is operative in a given patient. Importantly, the vast majority of patients with frequent PVCs will not go on to develop cardiomyopathy but currently available data do not allow for accurate risk prediction. A recent longitudinal study followed 239 patients with frequent PVCs (>1000 per day) and no SHD [echo and magnetic resonance imaging (MRI)] for 5.6 years with no adverse cardiac events and no decline in overall LV ejection fraction (LVEF).

**Diagnostic evaluation**

**Electrocardiogram and ambulatory monitoring**

The presence of at least some PVCs during 24 h ambulatory monitoring is extremely common and may be considered normal. Because the finding of PVCs during 24 h ambulatory monitoring is very likely, any conclusion that they are related to symptoms requires careful correlation. In two studies in which SHD was rigorously excluded, only 2 and 4% had >50 or >100 PVCs/24 h, respectively. The vast majority of patients without SHD who have PVCs have a benign prognosis. An exception may be a very small subset of patients with PVCs that have a short coupling interval (<300 ms) between the premature and the preceding beats, a finding which suggests the short QT syndrome and increases the risk of malignant VAs. It should be emphasized that this is a very small minority of patients with PVCs. As with other VAs, the first step in the evaluation of a patient with PVCs is to determine the presence or absence of SHD (Figures 1 and 2). For patients with arrhythmic or other cardiac symptoms, a resting 12-lead ECG is very helpful to evaluate the presence of myocardial scar (Q-waves or fractionated QRS complexes), the QT interval, ventricular hypertrophy, and other evidence of SHD. An echocardiogram provides assessment of right ventricular (RV) and LV structure and function, valvular abnormalities, and pulmonary artery systolic pressure and is recommended for patients with symptomatic PVCs, a high frequency of PVCs (>10% burden), or when the presence of SHD is suspected.

**Exercise testing**

For selected patients, especially when there is a suggestion of symptoms associated with exercise, exercise stress testing should be considered to determine whether PVCs are potenti-

**Imaging investigations**

Although the majority of patients with PVCs can be accurately assessed with a 12-lead ECG and echocardiography, contrast-enhanced MRI may provide additional diagnostic and prognostic data when the presence or absence of SHD remains in doubt. While there are no large-scale studies investigating which patients should undergo MRI, the management of several forms of SHD associated with PVCs may be guided by MRI, including dilated cardiomyopathy, hypertrophic cardiomyopathy (HCM), sarcoidosis, amyloidosis, and arrhythmogenic right ventricular cardiomyopathy (ARVC). In these conditions, the presence of ventricular wall

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**Table 1 PVC burden associated with LV dysfunction**

<table>
<thead>
<tr>
<th>n</th>
<th>%Lvd</th>
<th>%VEs Lvd</th>
<th>%VEs normal LV</th>
<th>P</th>
<th>Predictive PVC burden</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ban et al.</td>
<td>127</td>
<td>22%</td>
<td>31 ± 11%</td>
<td>22 ± 10%</td>
<td>0.001 26%</td>
</tr>
<tr>
<td>Deyell et al.</td>
<td>90</td>
<td>24%</td>
<td>32 ± 12%</td>
<td>27 ± 12%</td>
<td>0.077</td>
</tr>
<tr>
<td>Munoz et al.</td>
<td>70</td>
<td>24%</td>
<td>29 ± 15%</td>
<td>17 ± 14%</td>
<td>0.004 10% RV; 20% LV</td>
</tr>
<tr>
<td>Olgin et al.</td>
<td>51</td>
<td>24%</td>
<td>30 ± 11%</td>
<td>14 ± 15%</td>
<td>0.0001</td>
</tr>
<tr>
<td>Hasdemir et al.</td>
<td>249</td>
<td>7%</td>
<td>29 ± 9%</td>
<td>8 ± 7%</td>
<td>0.001 16%</td>
</tr>
<tr>
<td>Baman et al.</td>
<td>174</td>
<td>33%</td>
<td>33 ± 13%</td>
<td>13 ± 12%</td>
<td>0.0001 24%</td>
</tr>
<tr>
<td>Kanei et al.</td>
<td>108</td>
<td>19%</td>
<td>13 ± 11%</td>
<td>7 ± 9%</td>
<td>0.004</td>
</tr>
</tbody>
</table>

*Assuming 100 000 beats/24 h. Lowest PVC count associated with LV dysfunction was 10% (Baman). LV, left ventricle; LVD, left ventricular dysfunction; PVC, premature ventricular complexes; VE, ventricular ectopic.
Medical treatment

Catheter ablation
Consider if:
Failed/intolerant/declined medical Rx,
Single/dominant PVC morphology,
Probable reversible LV dysfunction

Continued PVC symptoms
- Ongoing PVC symptoms
- PVC interference with CRT

Reassurance
Reassurance re PVCs
Rx for SHD b
Possible reversible LV dysfunction (consider MRI-DE c)

No SHD

Figure 1  Management of PVCs. *See table for definitions of structural heart disease; bMedical therapy + ICD; cAbsence of high scar burden suggests reversibility. CRT, cardiac resynchronisation therapy; ICD, implantable cardioverter-defibrillator; LV, left ventricular; MRI-DE, magnetic resonance imaging with delayed enhancement; PE, physical examination; PVC, premature ventricular complexes; Rx, therapy; SHD, structural heart disease; VAs, ventricular arrhythmias.

Patient history
Prior diagnosis of SHD?
Risk factors for SHD?
Symptoms of SHD?
Non-cardiac disorders that may affect the heart?
Syncpe?
Family Hx SCD?
Family Hx of SHD?
Physical examination evidence for SHD?
Electrocardiogram baseline 12-lead ECG
12-lead ECG during VA
ECG monitoring
Echocardiogram
Exercise testing / Imaging coronary arteriography if indicated

Abnormal basic evaluation
Normal basic evaluation
Abnormal advanced imaging
Inconclusive basic evaluation
Normal advanced imaging

Structural heart disease
Advanced imaging
MRI-DE
CT coronary arteriography

No structural heart disease

Figure 2  Evaluation for the presence or absence of structural heart disease. CT, computed tomography; MRI-DE, magnetic resonance imaging with delayed enhancement; VA, ventricular arrhythmia.
motion abnormalities or myocardial scar detected by delayed gado-
linium enhancement may provide useful prognostic information. In
selected patients for whom the diagnosis of ARVC is suspected, the
signal-averaged ECG (SAECG) may provide useful information and
forms a minor diagnostic criterion for this disorder.

**Treatment**

**Indications for treatment in patients without structural heart disease**

In the absence of SHD, the most common indication for treating
PVCs remains the presence of symptoms that are not improved by
explanation of their benign nature and reassurance from the
physician. In addition, some patients may require treatment for
frequent asymptomatic PVCs if longitudinal imaging surveil-
ance reveals an interval decline in LV systolic function or an
increase in chamber volume. For patients with >10,000 PVCs/
24 h, follow-up with repeat echocardiography and Holter
monitoring should be considered. In patients with fewer PVCs,
further investigation is only necessary should symptoms in-
crease. It should also be recognized that PVC burden often
fluctuates over time.

**Indications for treatment in patients with structural heart disease**

In patients with SHD, symptoms form the primary grounds for
considering whether treatment is indicated. Elimination of high
burden PVCs (>10%) in patients with impaired LV function can be
associated with significant improvement of LV function, even
when significant scarring is present. Catheter ablation may also
be helpful when frequent PVCs interfere with cardiac resynchroniza-
tion therapy.

**Management of premature ventricular complexes (options)**

**Medical therapy**

For patients without SHD and mild symptoms, the first step in treat-
ment of patients with PVCs is education of the benign nature of this
arrhythmia and reassurance. No large-scale randomized trials of drug
treatment for PVCs in the absence of heart disease have been per-
formed. For patients whose symptoms are not effectively managed
in this manner, a trial of beta-blockers or non-dihydropyridine
calcium antagonists may be considered though the efficacy of these
agents is quite limited with only 10–15% of patients achieving
>90% PVC suppression, similar to placebo. It should also be
recognized that the data supporting the use of calcium blockers are
less than for beta-blockers and that these agents may themselves
produce significant symptoms. While membrane-active AADs are
more effective to suppress PVCs, the risk–benefit ratio has not
been carefully evaluated in patients without SHD. Nevertheless,
these agents are highly effective and may significantly improve symp-
toms in markedly symptomatic patients. Because these agents may in-
crease the risk of mortality in patients with significant SHD, perhaps
with the exception of amiodarone, caution is advised before using
them for PVC suppression.

**Catheter ablation**

Randomized trials of PVC suppression with catheter ablation have not been performed. However, multiple studies indicate
high efficacy of ablation with PVC elimination in 74–100% of patients. However, these studies have typically included
highly symptomatic patients typically with a very high burden of
PVCs. Thus, catheter ablation should only be considered for
patients who are markedly symptomatic with very frequent
PVCs. In addition, procedural success may be dependent on site
of origin with lower efficacy reported for coronary venous and
epicardial foci than for other sites. Although complete PVC
elimination is the goal of ablation, it should be noted that partial
success may still be associated with significant improvement in
LV systolic function. The efficacy of catheter ablation may be
reduced for patients with multiple morphologies of PVCs or
those for whom the clinical PVC morphology cannot be
induced at the time of the procedure. The published complication
rates of catheter ablation for PVC suppression are generally low
(∼1%). Catheter ablation of PVCs is recommended for highly
selected patients who remain very symptomatic despite conserva-
tive treatment or for those with very high PVC burdens associated
with a decline in LV systolic function.

**Non-sustained ventricular tachycardia**

Although several different definitions have been used, NSVT is
defined as runs of beats arising from the ventricles with duration
between 3 beats and 30 s and with cycle length of <600 ms
(100 b.p.m.). Similar to PVCs, NSVT is a relatively common
finding in patients with either structurally normal or abnormal
hearts. Non-sustained ventricular tachycardia is found in
nearly 6% of patients evaluated for palpitations. Diagnostic and
therapeutic considerations for NSVT are included in several
recent guideline and consensus documents. In general,
therapy for the underlying cardiac disease is indicated rather than
for the arrhythmia itself. However, the finding of NSVT should
always trigger further evaluation of the patient and a practical ap-
proach can be usefully divided into a general approach (Table 2),
patients with an apparently normal heart (Table 3) and those with
SHD (Table 4).

**Non-sustained ventricular tachycardia in the structurally normal heart**

Exercise-related NSVT is relatively common and appears to be
associated with a worse prognosis when it occurs during recov-
ery. Polymorphic NSVT requires extensive evaluation in both
symptomatic and asymptomatic patients with careful assess-
ment for the presence of coronary ischaemia. An important inher-
ited arrhythmia which may present as exercise-induced NSVT is
CPVT. This condition is typically manifested by polymorphic
or bidirectional VT which are triggered by sympathetic stimulation
and exercise (commonly occurring at an exercise level of
120–130 b.p.m.) and is associated with an increased risk of
sudden death. The underlying mechanism of CPVT is calcium
overload leading to delayed afterdepolarizations as a result of
mutations in the genes coding for ryanodine receptor or calsequestrin proteins. Non-sustained ventricular tachycardia is a relatively common finding among athletes.67,68 Other causes of NSVT in the absence of SHD include QT interval prolongation caused by mutations in proteins regulating repolarizing currents drugs (LQTS) or electrolyte abnormalities. Athletes with NSVT should be evaluated for the presence of HCM, a diagnosis which may overlap with some degree of LVH as an adaptation to exercise. Because of this challenging distinction, expert consultation should be obtained if this diagnosis is suspected. Although only limited data are available regarding the significance of NSVT in athletes without a structural cardiac disease, discontinuation of training is not generally recommended.

**Non-sustained ventricular tachycardia in structural heart disease**

Non-sustained ventricular tachycardia is common in ischaemic heart disease and can be recorded in 30–80% of patients during long-term ECG monitoring where it is usually asymptomatic.69,70 No studies have demonstrated a mortality benefit of suppressing NSVT with either AADs or catheter ablation and treatment is usually not indicated in asymptomatic patients. A range of studies have demonstrated that NSVT occurring during the first few days after an acute coronary event has no adverse long-term prognostic significance. However, when NSVT occurs 48 h or more after MI, there is an increased mortality and morbidity even when asymptomatic.74 For a patient with non-ischaemic dilated cardiomyopathy, the prognostic significance of NSVT is uncertain and no studies have provided precise guidance for treatment in this group of patients.75

The occurrence of NSVT in patients with an implanted ICD is associated with an increased frequency of shocks and all-cause mortality.76 For these patients, programming the ICD to a long VT detection time and a high ventricular fibrillation (VF) detection rate may be especially important.77,78

**Diagnostic evaluation**

For patients with an apparently normal heart, the 12-lead ECG should be scrutinized for evidence of typical outflow tract VT,53,54,56,62 (Figure 3) polymorphic VT (PMVT), including torsades de pointes (Tdp), or an inherited arrhythmia syndrome, such as the long QT, short QT, Brugada, or early repolarization syndromes (ERS)4,4a (Figure 4). Outflow tract VAs typically have an inferior axis with either RV or LV origin. When the precordial transition is \(<V3 and the ratio of the R- and S-waves in lead V2 during PVCs or VT divided by this ratio during sinus rhythm exceeds 0.6, a LV outflow tract origin is strongly suggested. In addition to the ECG, an echocardiogram to assess the presence or absence of SHD should also be considered for all patients with NSVT. For cases where SHD is suspected but cannot be definitively diagnosed with echocardiography, cardiac MRI may be especially useful to confirm the presence or absence of myocardial scar or wall motion abnormalities. Classification of NSVT should be attempted using a scheme similar to Tables 3 and 4. Evaluation in CHD is described in a separate section.

**Treatment**

**Non-sustained ventricular tachycardia in the absence of structural heart disease**

Most short-lasting monomorphic NSVTs originate from the RV or LV outflow tracts (Table 3, Figure 3). These arrhythmias only require treatment if they are symptomatic, incessant, or produce LV dysfunction. Sudden death is very rare in patients with outflow tract VT. The treatment of these arrhythmias is either medical with a beta-blocker, a non-hydropyridine calcium blocker, class IC drugs, or with catheter ablation.80 Non-sustained ventricular tachycardia with a focal mechanism may also occur from the papillary muscles and...
Table 3  Non-sustained ventricular tachycardia with apparent normal heart

<table>
<thead>
<tr>
<th>NSVT clinical presentation</th>
<th>ECG</th>
<th>Risk of sudden cardiac death</th>
<th>Diagnostic evaluation</th>
<th>Alternative diagnostic considerations</th>
<th>Treatment</th>
<th>Treatment to be considered</th>
<th>Key references</th>
</tr>
</thead>
<tbody>
<tr>
<td>Typical RVOT</td>
<td>LBBB, inf. axis, axis transition V3–V4</td>
<td>Very rare</td>
<td>Standard</td>
<td>Differentiate from ARVC</td>
<td>Beta-blocker, verapamil, IC drugs with symptoms</td>
<td>Catheter ablation</td>
<td>Latif et al.62</td>
</tr>
<tr>
<td>Typical LVOT</td>
<td>Inferior axis, transition &lt; V3</td>
<td>Very rare</td>
<td>Standard</td>
<td>RVOT VT</td>
<td>Beta-blocker, verapamil, IC drugs with symptoms</td>
<td>Catheter ablation</td>
<td>Latif et al.62</td>
</tr>
<tr>
<td>Idiopathic reentrant LV tachycardia</td>
<td>RBBB, LS axis</td>
<td>Very rare</td>
<td>Standard EP testing</td>
<td>Ischaemic heart disease, CM</td>
<td>Verapamil if symptomatic</td>
<td>Catheter ablation</td>
<td>Latif et al.62</td>
</tr>
<tr>
<td>Other focal VT</td>
<td>Multiple morphologies, monomorphic</td>
<td>Uncommon</td>
<td>Exercise testing or catecholamine stimulation</td>
<td>Ischaemic heart disease, CM</td>
<td>Beta-blocker for the arrhythmia</td>
<td>Catheter ablation</td>
<td>Latif et al.62</td>
</tr>
<tr>
<td>Exercise</td>
<td>Multiple</td>
<td>Increased risk when NSVT in recovery</td>
<td>Ischaemic heart disease, cardiomyopathy</td>
<td>CPVT</td>
<td>Underlying disease</td>
<td>Beta-blockers, flecainide</td>
<td>Jouven et al.65, Frolkis et al.66</td>
</tr>
<tr>
<td>Athlete</td>
<td>Multiple</td>
<td>If it disappears with increased exercise low risk</td>
<td>Evaluate for latent HCM or ischaemic heart disease</td>
<td>HCM</td>
<td>No treatment training can continue</td>
<td>None</td>
<td>Biffi et al.67,68</td>
</tr>
<tr>
<td>Hypertension valvular disease</td>
<td>Multiple morphology</td>
<td>As without arrhythmia</td>
<td>Consider ischaemic heart disease</td>
<td>Ischaemic heart disease, CM</td>
<td>Treat HTN</td>
<td>Beta-blocker</td>
<td></td>
</tr>
<tr>
<td>Polymorphic VT</td>
<td>Polymorphic</td>
<td>High</td>
<td>Evaluated for CAD, CPVT, inherited arrhythmia syndromes</td>
<td>Purkinje fibre triggering focus</td>
<td>Underlying disease</td>
<td>Revascularization, ICD, beta-blocker, catheter ablation</td>
<td>Zipes et al.60</td>
</tr>
<tr>
<td>TdP VT</td>
<td>Long QT, TdP</td>
<td>High</td>
<td>Medications, congenital LQTS</td>
<td>Medications, K^+, Mg^{++}, Ca^{++}</td>
<td>Stop medications, correct electrolytes</td>
<td>ICD, beta-blocker</td>
<td>Sauer and Newton-Cheh69</td>
</tr>
</tbody>
</table>

ARVC, arrhythmogenic right ventricular cardiomyopathy; CAD, coronary artery disease; CM, cardiomyopathy; CPVT, catecholaminergic polymorphic ventricular tachycardia; HCM, hypertrophic cardiomyopathy; HTN, hypertension; ICD, implantable cardioverter-defibrillator; LS, left-superior; LV, left ventricular; LVOT, left ventricular outflow tract; NSVT, non-sustained ventricular tachycardia; RBBB, right bundle branch block; RVOT, right ventricular outflow tract; TdP, torsade pointes; VT, ventricular tachycardia.
respond to beta-blockers or catheter ablation. In addition, reentrant LV VT utilizing false tendons can be treated with verapamil, though with a relatively high recurrence risk on oral therapy. Catheter ablation is effective for idiopathic reentrant LV VT and should be considered even when this sustained arrhythmia is terminated by intravenous verapamil. Catheter ablation can be recommended for patients with idiopathic NSVT that is highly symptomatic and drug refractory, especially if it is exercise-induced.

### Table 4 Non-sustained ventricular tachycardia in structural heart disease

<table>
<thead>
<tr>
<th>Clinical setting</th>
<th>Risk of sudden cardiac death</th>
<th>Arrhythmia specialist evaluation</th>
<th>Diagnostic evaluation</th>
<th>Diagnostics to be considered</th>
<th>Treatment</th>
<th>Treatment to be considered</th>
<th>Key references</th>
</tr>
</thead>
<tbody>
<tr>
<td>ACS within 48 h</td>
<td>No increased risk</td>
<td>No</td>
<td>Coronary artery disease</td>
<td>Monitoring</td>
<td>Beta-blockers</td>
<td></td>
<td>Hohnloser et al.</td>
</tr>
<tr>
<td>ACS after 48 h</td>
<td>Risk increased</td>
<td>Yes</td>
<td>Consider EPS if moderate LV dysfunction</td>
<td>Continued evaluation for repetitive arrhythmias</td>
<td>Beta-blockers</td>
<td>ICD</td>
<td>Zipes et al.</td>
</tr>
<tr>
<td>Previous MI, EF 31–40</td>
<td>Increased risk</td>
<td>Yes</td>
<td>EPS</td>
<td>ICD with inducible VT/VF</td>
<td>ICD, see relevant guidelines</td>
<td>Zipes et al.</td>
<td></td>
</tr>
<tr>
<td>Previous MI, EF ≤ 30 Chronic heart failure, EF ≤ 30</td>
<td>Increased risk</td>
<td>Yes</td>
<td>Non-driven by arrhythmia</td>
<td>ICD</td>
<td>Antiarrhythmic medical therapy or ablation with symptoms</td>
<td>Zipes et al.</td>
<td></td>
</tr>
<tr>
<td>Syncope with chronic CAD, EF &gt; 40</td>
<td>Increased risk</td>
<td>Yes</td>
<td>EP testing, ischaemia testing</td>
<td>ICD with inducible VT/VF</td>
<td>Additional antiarrhythmic therapy or ablation with symptoms</td>
<td>Zipes et al.</td>
<td></td>
</tr>
<tr>
<td>Non-ischaemic dilated CM</td>
<td>Uncertain</td>
<td>Yes</td>
<td>Uncertain</td>
<td>EP testing</td>
<td>Uncertain</td>
<td>ICD, see relevant guidelines</td>
<td>Zipes et al.</td>
</tr>
<tr>
<td>HCM</td>
<td>Increased risk</td>
<td>Yes</td>
<td>Echo, MRI</td>
<td>Beta-blocker, ICD</td>
<td>ICD</td>
<td>Zipes et al.</td>
<td></td>
</tr>
<tr>
<td>LQTS</td>
<td>Increased risk</td>
<td>Yes</td>
<td>Genetic screening</td>
<td>Beta-blocker</td>
<td>ICD</td>
<td>Zipes et al.</td>
<td></td>
</tr>
<tr>
<td>Short QT syndrome</td>
<td>Increased risk</td>
<td>Yes</td>
<td>Provocative testing</td>
<td>With syncope or cardiac arrest: ICD</td>
<td>Quinidine</td>
<td>Aliot et al.</td>
<td></td>
</tr>
<tr>
<td>Brugada syndrome</td>
<td>Increased risk</td>
<td>Yes</td>
<td>Genetic screening</td>
<td>With syncope or cardiac arrest: ICD</td>
<td>Quinidine</td>
<td>Aliot et al.</td>
<td></td>
</tr>
<tr>
<td>ER syndrome</td>
<td>Increased risk</td>
<td>Yes</td>
<td>Provocative testing</td>
<td>Genetic screening</td>
<td>With syncope or cardiac arrest: ICD</td>
<td>Quinidine</td>
<td>Aliot et al.</td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>Yes</td>
<td>Genetic screening</td>
<td>With syncope or cardiac arrest: ICD</td>
<td>Quinidine</td>
<td>Aliot et al.</td>
<td></td>
</tr>
</tbody>
</table>

CAD, coronary artery disease; CM, cardiomyopathy; EF, ejection fraction; EP, electrophysiology; EPS, electrophysiological study; ER, early repolarisation; HCM, hypertrophic cardiomyopathy; ICD, implantable cardioverter-defibrillator; LV, left ventricular; MI, myocardial infarction; VF, ventricular fibrillation; VT, ventricular tachycardia.

Although an ICD should be considered for all patients with a significantly reduced LVEF (<0.35), there may be a role for programmed electrical stimulation in selected patients with NSVT and ischaemic heart disease who have less severe LV dysfunction (LVEF < 0.40). Implantable cardioverter-defibrillator implantation is recommended in this group of patients if VF or sustained VT is inducible with programmed electrical stimulation. Similarly, if NSVT is observed in a patient with a prior MI, a history of syncope, and LVEF ≤ 40%, EPS is generally recommended to guide treatment, usually with ICD implantation, should sustained VT be inducible. Non-sustained ventricular tachycardia in an asymptomatic patient with a LVEF > 40% does not usually require specific antiarrhythmic therapy, and the goal is optimized treatment of the underlying heart disease. In the setting of HCM, ICD therapy is an appropriate consideration if NSVT is present with or without other major risk factors. In general, AAD therapy may be considered for patients with SHD who experience symptomatic, recurrent NSVT not resolved by revascularization, optimization of medical therapy, or treatment of reversible factors.

**Non-sustained ventricular tachycardia in patients with structural heart disease**

The recording of polymorphic NSVT should prompt a thorough evaluation for the presence of coronary ischaemia as the primary therapy for this arrhythmia should be directed to improving coronary perfusion. If non-sustained PMVT can be classified as a CPVT, the risk of life-threatening arrhythmia is high and beta-blockade therapy with potential placement of an ICD is recommended. In cases of TdP VT, any medication or electrolyte disturbance that prolongs repolarization should be addressed.
Figure 3  (A) Right ventricular (RV) outflow tract VT. (B) Left coronary cusp VT.
Sustained monomorphic ventricular tachycardia

Expert consensus recommendations on SMVT

1. A 12-lead ECG should be recorded during sustained VTs whenever possible and practical. (I) LOE B
2. For patients with newly diagnosed sustained monomorphic VT (SMVT) and no evidence of SHD on resting ECG or echocardiography
   a. cardiac MRI may provide additional information (IIb), LOE B
   b. signal-averaged ECG may provide additional information (IIb), LOE C
   c. exercise testing may provide additional information (IIb), LOE B
3. For patients with a wide QRS complex tachycardia in whom the diagnosis is uncertain, an invasive EPS should be considered to identify the tachycardia mechanism. (IIa) LOE C
4. For patients with SHD and SMVT, an ICD is recommended in the absence of contraindications. (I) LOE A
5. For patients with SHD and recurrent SMVT, specific treatment of VAs with AADs (amiodarone, mexiletine, or sotalol), catheter ablation, and/or antitachycardia pacing (ATP) from an ICD should be considered in addition to an ICD. Treatment of the underlying SHD or ischaemia will in most cases not be sufficient to prevent monomorphic VT (MMVT) recurrences. (IIa) LOE B
6. For patients with an ICD as primary prophylaxis, programming to a long VT detection interval and a high VF detection rate should be considered. (IIa) LOE A

Monomorphic VT is defined as sustained when lasting longer than 30 s or requires earlier intervention due to haemodynamic instability. Most commonly, sustained MMVT occurs in the setting of diseased myocardium, but may also be idiopathic, occurring in patients with no detectable myocardial disease.

Importance and prognosis

No structural disease—idiopathic ventricular tachycardia

In the absence of SHD, SMVT is generally associated with an excellent prognosis. The presence of syncope or PMVT is unusual in the absence of SHD or an inherited arrhythmia syndrome. Rarely, idiopathic VT can have a malignant clinical course, usually with a very rapid rate or a short initiating coupling interval.

Sustained monomorphic ventricular tachycardia in patients with structural heart disease

The large majority of patients with SMVT who present for therapy have significant SHD. The most frequent aetiology is ischaemic heart disease, comprising 54–59% of patients for whom an ICD is implanted or who are referred for catheter ablation. Sustained VT is associated with increased mortality risk in the setting of reduced ventricular systolic function. The mortality risk attributable to VT in patients with preserved ventricular function is less well defined. Implantable cardioverter-defibrillator shocks are also associated with inherent risk and multiple studies have demonstrated that defibrillator shocks, both appropriate and inappropriate, are associated with increased mortality and reduced quality of life. The association of ICD shocks and total mortality appears mainly to be a function of worsening cardiac disease rather than a specific consequence of shocks. Programming of ICDs with long VT detection times prior to the delivery of antitachycardia therapies and rapid VF detection rates reduces shocks and improves mortality in patients receiving an ICD for primary prophylaxis. The value of programming a long VT detection time in patients with a history of sustained MMVT or VF is less certain. Although it has not been determined whether suppression of VT by either pharmacological means or catheter ablation improves survival in patients with sustained MMVT, treatment to avoid recurrent symptoms is appropriate and these therapies may improve survival in patients presenting with recurrent VT storm.
Diagnostic evaluation

Electrocardiogram

The key distinction to make in the investigation of SMVT is to discern the presence or absence of SHD, see Table 2. A 12-lead ECG helps to confirm the diagnosis of VT,106–110 provide important insight into the underlying mechanism (Tables 3 and 4), identify the presence of SHD,111 and suggest the site of origin. This is especially important when catheter ablation is planned.112,113 A resting ECG should be performed in all patients with sustained VT. The presence of Q-waves or fragmentation of the QRS complex suggests underlying structural disease (Figure 5).114,115

Cardiac imaging

The presence of myocardial scar is more likely to be associated with poorly tolerated VT, haemodynamic collapse, degeneration to VF, and sudden death. In most cases, echocardiography can adequately demonstrate myocardial structure and function. If echocardiography is normal, more detailed imaging using cardiac MRI can exclude less clearly evident myocardial scar, arrhythmogenic RV cardiomyopathy, non-ischaemic cardiomyopathy with preserved EF, HCM, or cardiac sarcoidosis.116 It may also be helpful to reevaluate ventricular function when a patient with previously known SHD presents with SMVT.

Signal-averaged electrocardiogram

An SAECG, recorded during the baseline rhythm may permit the identification of slow myocardial conduction by recording low-amplitude potentials but does not help in scar localization. A negative test has been associated with better prognosis117 but with only modest positive predictive value.118 The SAECG may be most useful in identifying ARVC where a positive test forms a minor criterion in the diagnostic component for this disorder.39,119

Invasive electrophysiological study

Patients presenting with syncope or sustained palpitations who have evidence of myocardial scar as well as those with a wide-complex tachycardia for whom the diagnosis of VT is not certain may benefit from a provocative EPS. Although the standalone negative and positive predictive values of this testing are limited,88,120 inducible SMVT is highly associated with recurrent VT and may provide clues to the cause of syncope or other symptoms suggestive of a VA. Electroanatomical mapping of the RV has been used to identify otherwise unapparent RV scar.121,122

Testing for ischaemia

Transient myocardial ischaemia is an uncommon sole cause of recurrent sustained VT that is monomorphic. Most patients with coronary artery disease who develop sustained MMVT have a fixed region of myocardial scar that is a sequela of prior MI, often occurring many years earlier.95–97 Patients with a new presentation of sustained MMVT should have a thorough evaluation to define the presence or absence of underlying heart disease, which includes echocardiography, exercise testing, and stress/perfusion imaging. For most patients where the coronary artery disease is suspected as the underlying diagnosis, coronary angiography should be considered.123–126 However, treatment of ischaemia alone is unlikely to prevent
recurrences of MMVT. Cardiac MRI and positron emission tomography - computed tomography may provide evidence for myocardial scar that is not evident with other imaging modalities and may be especially useful to differential occult SHD from idiopathic VT.127

Treatment
Acute therapy for sustained ventricular tachycardia
Ventricular fibrillation should be immediately defibrillated using a non-synchronized mode. The use of intravenous amiodarone has been associated with a higher survival probability than when lidocaine is administered to patients resuscitated from VF.128 The acute treatment of sustained VT is largely based on the patient’s symptoms and haemodynamic tolerance of the arrhythmia. For patients with sustained MMVT who are unconscious or who have experienced haemodynamic collapse, direct current cardioversion synchronized to the QRS on the surface ECG should be immediately performed. Patients who are conscious but have marked hypotension or profound symptoms from VT should be given prompt intravenous sedation and then cardioverted. A trial of intravenous lidocaine (1 mg/kg) may be given as preparation is made for sedation, though the efficacy for termination of sustained VT is only ~15%.129 For patients with sustained VT who are haemodynamically stable or have only mild symptoms, a 12-lead ECG should be recorded and carefully analysed before therapy is initiated. For patients without SHD and a QRS morphology suggesting an idiopathic outflow tract VT, a trial of a short-acting intravenous beta-blocker may be useful to terminate VT. However, for patients with SHD with sustained VT, the most efficacious pharmacological agent is intravenous amiodarone.129 This agent may be associated with hypotension if administered rapidly, usually via a central venous catheter. These patients must be continuously observed and intravenous sedation and cardioversion should be readily available and applied if symptoms worsen or haemodynamic deterioration occurs. Patients with TdP VT should be cardioverted if the arrhythmia is sustained. For those with recurrent non-sustained TdP VT, atrial pacing at a rate of at least 90 b.p.m. is highly effective to prevent recurrences. Intravenous isoproterenol can be useful to suppress recurrent TdP or VF in patients with the Brugada syndrome (BrS).

Pharmacological therapy for idiopathic ventricular tachycardia
The indication for treatment of idiopathic VT derives largely from the symptom burden. Beta-blockade and non-hydropyridine calcium channel blockade are low-risk therapies which have modest effectiveness.130,131 Antiarrhythmic drug therapy using sotalol, flecainide, mexiletine, propafenone, or amiodarone is more effective, but carries the potential for pro-arrhythmic risk, and a greater side-effect profile.132

Catheter ablation for idiopathic ventricular tachycardia
For focal VT with an ECG pattern highly suggestive of right ventricular outflow tract (RVOT) VT,113 catheter ablation is highly successful and carries low procedural risk. The most frequent limitation is the lack of VT inducibility during the procedure. The success rate of ablation of outflow tract VTs arising from non-RVOT sites may be somewhat lower and may involve greater procedural complexity but should be considered. Fascicular VT and focal VTs from non-outflow tract sites such as the LV or RV papillary muscles may also be amenable to catheter ablation, with the principal limitation being inducibility of the arrhythmia and achieving adequate mapping and catheter contact to abolish the VT.3,44–58,71,79,132–134 In addition, it should be appreciated that papillary muscle VT has a significant risk of recurrence after initially apparent successful catheter ablation.

Pharmacological therapy for ventricular tachycardia with structural heart disease
The presence of SHD increases the risk of pro-arrhythmia from membrane-active AADs, such that they are generally used only with the added protection afforded by an ICD.33 There is no evidence that antiarrhythmic therapy alone improves survival in patients with sustained MMVT.135–137 Sotalol has been demonstrated to reduce the frequency of sustained MMVT recurrences138,139 in patients with SHD. In the OPTIC trial, sotalol reduced all-cause ICD shocks at 1 year from 38.5 to 24.3% [hazard ratio (HR) 0.61, P = 0.055].138 A smaller study suggested that sotalol was inferior to metoprolol.140 In these trials, sotalol was associated with a safety profile which was similar to that of beta-blockers alone. In the presence of a normal or near-normal QT interval at baseline, and normal or near-normal renal function, sotalol is a reasonable first pharmacological therapy to suppress recurrences of sustained MMVT. In comparison with beta-blocker therapy alone, amiodarone has been demonstrated to markedly reduce recurrent appropriate ICD therapy during 1-year follow-up when used for secondary prophylaxis (HR 0.30, P < 0.001).138 However, longer term use of amiodarone for secondary prophylaxis is associated with high rates of VT recurrence and serious adverse effects and may increase mortality compared with placebo.141,142 Other AADs that have been used to reduce recurrences of sustained MMVT include dofetilide143 and the combination of mexiletine and amiodarone.144,145 Dofetilide is not Food and Drug Administration approved for use in VAs and is not available in many parts of the world. Limited experience is also present with combinations of sotalol and either quinidine or procainamide,145 or amiodarone plus mexiletine plus either quinidine or procainamide.146

Implantable cardioverter-defibrillator implantation and programming
An ICD is indicated for most patients with SHD and sustained VT.64 Implantable cardioverter-defibrillator implantation has been demonstrated to improve survival in patients with VT and reduced systolic function.95–97 Implantable cardioverter-defibrillator implantation is indicated for patients with sustained MMVT and myocardial scar, even when systolic function is normal or near-normal1 based upon extrapolation from randomized trials including patients with low EF. Although evidence of mortality benefit is scant in the absence of severe systolic dysfunction, ICD implantation may simplify management and follow-up of these patients.

Catheter ablation
Catheter ablation for VT is an important non-pharmacological alternative or adjunct to AAD therapy.2 Catheter ablation has been demonstrated to reduce appropriate ICD shocks for patients with ischaemic cardiomyopathy when utilized after a first presentation with VA.147 In patients with prior MI, reduced EF, and haemodynamically stable VT, catheter ablation significantly reduces recurrences of VT, with the greatest benefit in patients with EF > 30%.148 Cooled tip catheter ablation was superior to AAD therapy for reducing recurrences of sustained MMVT in patients with ischaemic heart disease.
who had failed amiodarone. Although catheter ablation reduces recurrences of sustained MMVT in patients with ischaemic cardiomyopathy, a reduction in mortality has yet to be demonstrated. Catheter ablation has also been successfully used in patients with non-ischaemic cardiomyopathy where the ablation target is often on the epicardial surface of the ventricles and the procedure may be more complex. The long-term effectiveness of catheter ablation for non-ischaemic cardiomyopathies has been less well studied than for ischaemic cardiomyopathies.

While either catheter ablation or AAD therapy may be used as a first-line therapy for VT in the setting of prior MI, catheter ablation is the preferred therapy for patients presenting with incessant sustained MMVT. While encouraging, the long-term success of catheter ablation for sustained MMVT in patients with non-ischaemic cardiomyopathy is less well defined than in patients with ischaemic heart disease. Thus, AAD therapy is often used as first-line therapy with catheter ablation reserved for those with recurrent VT while receiving medications. Procedural complications with catheter ablation of sustained MMVT in the presence of SHD have generally been reported in <5% of patients and may include atrioventricular (AV) conduction block, cardiac perforation, stroke/transient ischaemic attack, heart failure, or death, usually in <3% of patients.

Sustained polymorphic ventricular tachycardia/ventricular fibrillation

Expert consensus recommendations on sustained polymorphic VT/VF

1. Patients with polymorphic VT or VF should be thoroughly evaluated for the presence of SHD, inherited arrhythmia syndromes, early repolarization, coronary artery spasm, and pro-arrhythmic effects of medications using:
   a. Twelve-lead ECG during the arrhythmia (when feasible) and during normal rhythm. (I) LOE C
   b. Echocardiography. (I) LOE B
   c. Coronary arteriography. (I) LOE B

2. Specific antiarrhythmic therapies, e.g. quinidine in patients with idiopathic VF, sodium channel blocker therapy in patients with long QT syndrome (LQTS) III, intensive autonomic inhibition in patients with catecholaminergic VTs, or quinidine in BrS, should be considered in close cooperation with a specialist in these diseases to reduce the risk of recurrence as an adjunct to—and rarely as an alternative to—defibrillator therapy in survivors of polymorphic VTs. Detailed guidance can be found in the APHRS/EHRA/HRS document on inherited arrhythmia syndromes. (IIa) LOE B

3. For patients with VT/VF storm, reversible factors such as electrolyte abnormalities, pro-arrhythmic drugs, ischaemia, and decompensated chronic heart failure should be corrected. (I) LOE C

4. Pharmacological suppression of VT/VF storm with beta-adrenergic blockers, amiodarone, and/or lidocaine should be considered in all patients. (IIa) LOE C

5. For patients with VT/VF storm in whom pharmacological suppression has not been effective and who are unstable, neuroaxial modulation, mechanical ventilation, catheter ablation, and/or anaesthesia may be considered. (IIb) LOE C

6. Catheter ablation of VTs or a triggering focus of VF should be considered in patients with VT/VF storm when adequate experience is available. (IIb) LOE C

7. For patients with VT/VF storm and significant SHD, implantation of a LV assist device (LVAD) or heart transplant evaluation should be considered and discussed early after the initial event. (IIa) LOE C

Polymorphic ventricular tachycardia is defined as a ventricular rhythm faster than 100 b.p.m., with clearly defined QRS complexes that change continuously from beat to beat indicating a changing ventricular activation sequence. The QT interval can either be normal or prolonged during intervening sinus rhythm in patients with PMVT. When PMVT occurs in the setting of a prolonged QT interval and has a distinctive pattern where the QRS complexes appear to be twisting around the isoelectric baseline, the arrhythmia is referred to as TdP. In cases of TdP, a long—short ventricular cycle length typically characterizes the initiating sequence, and the QT interval is almost always prolonged during sinus rhythm. Torsades de pointes VT is strongly associated with drugs or electrolyte abnormalities that delay repolarization. Thus, the occurrence of this arrhythmia should always prompt a search for precipitating factors that should be corrected.

Polymorphic VT has more than one morphologically distinct QRS complex occurring during the same episode of VT, but the QRS is not continuously changing. Ventricular fibrillation differs from PMVT in that VF is a chaotic tachycardia without consistently identifiable QRS complexes. It is important to distinguish PMVT, TdP, and VF because the mechanisms and the ultimate therapies for each may differ.

Importance and prognosis

Following cardiopulmonary resuscitation and protection of cerebral function in a patient with VF or sustained PMVT, the initial diagnostic step is to exclude an acute coronary syndrome (ACS) or MI. An ischaemic cause of these arrhythmias is very common and emergent coronary angiography and revascularization may significantly improve prognosis. In the absence of evidence for myocardial ischaemia, the structure and function of the ventricles should be assessed with echocardiography. A scheme for a diagnostic workup is shown in Figure 6. Patients who have impaired LV systolic function after MI (LVEF < 0.35) are at higher risk of sudden death in the first 3 months and may benefit from a wearable defibrillator. The LV function should be reassessed 40 days after MI to determine whether there is an indication for an ICD. Patients who are treated with coronary revascularization after MI are also at risk, especially if the LVEF is < 0.35. These patients may also benefit from a wearable defibrillator with reassessment of LV function and the indication for an ICD at 90 days post-revascularization.

Patients without structural heart disease

Polymorphic VT or VF in the absence of SHD suggests the presence of an inherited arrhythmia syndrome such as CPVT, the long QT, short QT, Brugada, or ERS (see Table 5). A resting 12-lead ECG should be recorded as close as possible to the VA episode as the chances of making the correct diagnosis is highest at this time. Recording of all 12 ECG leads over a longer period may be very useful to identify the morphology and location of PVCs that trigger PMVT or VF. Use of the Valsalva manoeuvre or high precordial leads may improve the sensitivity of the 12-lead ECG for detecting such triggers. In addition, the QRS and QT changes occurring
Figure 6  Sustained polymorphic ventricular tachycardia/ventricular fibrillation. aLV function should be reassessed at 40 days after MI or 90 days after revascularization. ACS, acute coronary syndrome; ACLS, advance cardiovascular life support; CAD, coronary artery disease; ICD, implantable cardioverter-defibrillator; LVEF, left ventricular ejection fraction; SHD, structural heart disease.

Table 5  Conditions that can cause PVT/VF in the absence of SHD and potential therapies

<table>
<thead>
<tr>
<th>Clues</th>
<th>Tests to Consider</th>
<th>Diagnoses</th>
<th>Therapies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Long QT/T-wave alternans</td>
<td>ECG/Monitor</td>
<td>Congenital LQTS</td>
<td>Beta-blockers/stellatectomy</td>
</tr>
<tr>
<td>TdP pattern</td>
<td></td>
<td></td>
<td>Avoid QT prolonging drugs</td>
</tr>
<tr>
<td>History of seizures</td>
<td>Epinephrine challenge</td>
<td></td>
<td>Mexilirine/flecainide (LQT3)</td>
</tr>
<tr>
<td>Specific trigger (loud noise)</td>
<td>Genetic testing</td>
<td></td>
<td>Pacemaker/ICD</td>
</tr>
<tr>
<td>Long QT/T-wave alternans</td>
<td>ECG/Monitor</td>
<td>Acquired LQTS</td>
<td>Mg²⁺⁺⁺⁺K⁺⁺⁺⁺</td>
</tr>
<tr>
<td>TdP pattern</td>
<td></td>
<td></td>
<td>Stop offending drug</td>
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<tr>
<td>Renal failure</td>
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<td></td>
<td>Temporary pacing</td>
</tr>
<tr>
<td>New medication or drug abuse</td>
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</tr>
<tr>
<td>AV block</td>
<td>ECG/Monitor</td>
<td>Bradycardia</td>
<td>Pacemaker</td>
</tr>
<tr>
<td>Incomplete RBBB with STE in leads V1–V2</td>
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<td>BrS</td>
<td>Isoproterenol/quinidine</td>
</tr>
<tr>
<td>Fever</td>
<td>ECG Drug challenge</td>
<td></td>
<td>Anipyretic</td>
</tr>
<tr>
<td>Drug challenge</td>
<td>Genetic testing</td>
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<td>Ablation</td>
</tr>
<tr>
<td>Monomorhic PVC trigger</td>
<td>ECG/Monitor</td>
<td>Focal PVC origin</td>
<td>Ablation/ICD</td>
</tr>
<tr>
<td>J-point elevation</td>
<td>ECG</td>
<td>Early repolarization</td>
<td>ICD</td>
</tr>
<tr>
<td>Ventricular pre-excitation</td>
<td>ECG</td>
<td>WPW</td>
<td>Ablation</td>
</tr>
<tr>
<td>Short QT interval</td>
<td>ECG</td>
<td>Short QTS</td>
<td>ICD</td>
</tr>
<tr>
<td>Bidirectional VT pattern exercise-induced</td>
<td>Digoxin level</td>
<td>CPVT</td>
<td>Stop digoxin</td>
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<tr>
<td></td>
<td>Exercise test</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>Genetic testing</td>
<td>Andersen-Tawil syndrome</td>
<td>Beta-blockers/CCBs/flecainide</td>
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<td></td>
<td></td>
<td>Digoxin toxicity</td>
<td>ICD</td>
</tr>
<tr>
<td>STE and chest pain</td>
<td>Proactive testing</td>
<td>Coronary spasm</td>
<td>Vasodilators/coronary stent</td>
</tr>
<tr>
<td>Short-coupled PVC trigger</td>
<td>ECG/Monitor</td>
<td>Idiopathic</td>
<td>ICD</td>
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</table>

BrS, Brugada syndrome; CCBs, calcium channel blockers; CPVT, catecholaminergic polymorphic ventricular tachycardia; ICD, implantable cardioverter-defibrillator; LQTS, long QT syndrome; PVC, premature ventricular complex; RBBB, right bundle branch block; short QTS, short QT syndrome; STE, ST elevation; TdP, torsade de pointes; VT, ventricular tachycardia; WPW, Wolff-Parkinson-White (syndrome).
after extrasystoles as well as during standing may help to identify J-wave abnormalities or abnormalities of the QT interval. Ambulatory monitoring may help identifying QTc prolongation during sleep. The role of genetic testing has been recently reviewed and plays an important part in the evaluation of patients in whom an inherited arrhythmia syndrome is suspected and for the family members of patients with these syndromes.

**Exercise testing**

In the setting of a normal resting 12-lead ECG, the occurrence of polymorphic PVCs and bidirectional VT during exercise suggests the diagnosis of CPVT. Exercise testing is also useful in the diagnosis of LQTS when the QT is of borderline duration at rest. The absence of QTc shortening at higher heart rate favours the diagnosis of long QT. The recovery phase of exercise testing may unmask BrS or LQTS patients with a normal ECG at baseline.

**Pharmacological testing**

Different tests have been proposed to evaluate polymorphic VT/VF in the absence of SHD. Intravenous sodium channel blocker challenge may unmask the BrS. Epinephrine challenge may help unmasking the LQTS, especially LQTS Types 1 and 2. Iso-proterenol challenge has been proposed to identify the early stages of ARVC, though this is rarely used in modern practice. It can also be an option for familial screening of CPVT when stress testing is negative. Adenosine may be used to unmask pre-excitation in patients with the Wolff-Parkinson-White (WPW) syndrome where the diagnosis is unclear during baseline electrocardiographic recordings.

**Patients with structural heart disease**

The resting ECG evidence of ischaemia, injury, or infarction - see Table 6.

Acute coronary syndromes and old Q-wave MI are the principal causes of PMVT/VF associated with a normal QTc interval. In addition, transient myocardial ischaemia may induce PMVT or VF, especially during conditions of stress or exercise. The presence of ST depression, elevation, or Q-waves in a patient with PMVT or VF should lead to prompt coronary angiography. In the absence of acute ECG evidence of ischaemia or injury, an invasive or non-invasive evaluation of coronary artery perfusion is indicated. It should be noted that LV and RV function may be depressed immediately after cardiac arrest and may markedly improve over a period of days to weeks. A prolonged or fragmented QRS (fQRS) is a predictor of SCD, appropriate ICD shocks, and all-cause mortality in patients with ischaemic cardiomyopathy. The presence of fQRS in patients with left bundle branch block (LBBB) is of particular prognostic significance.

The resting ECG may strongly suggest the diagnosis of a dilated cardiomyopathy when the QRS is prolonged or ARVC when epsilon waves or localized QRS duration \(\geq 110\) ms are recorded in surface leads V1, V2, or V3, with inverted T-waves in V2 and V3. The

<table>
<thead>
<tr>
<th>Clues</th>
<th>Tests to consider</th>
<th>Diagnoses</th>
<th>Therapies</th>
</tr>
</thead>
<tbody>
<tr>
<td>ECG evidence of ischaemia, injury, or infarction</td>
<td>Stress test, Coronary angiography, Echo/MRI</td>
<td>Coronary artery disease, Post-myocardial infarction</td>
<td>Coronary revascularization, Beta-blockers, Sotalol/amiodarone, Intra-aortic balloon pump, ICD</td>
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<tr>
<td>Angina/heart failure</td>
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<tr>
<td>Prior coronary revascularization</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Heart failure</td>
<td>Echo/MRI, Coronary angiography</td>
<td>Dilated non-ischaemic cardiomyopathy</td>
<td>Avoid cardiotoxins, Beta-blockers, Sotalol/amiodarone/ICD</td>
</tr>
<tr>
<td>Alcoholism</td>
<td>Echo/MRI, Coronary angiography</td>
<td>Hypertrophic cardiomyopathy</td>
<td>Beta-blockers, Sotalol/amiodarone/ICD</td>
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<tr>
<td>Systolic murmur</td>
<td>Echo/MRI, Genetic testing</td>
<td>Arrhythmogenic cardiomyopathy</td>
<td>Beta-blockers, Sotalol/amiodarone/ICD</td>
</tr>
<tr>
<td>Syncope</td>
<td>Echo/MRI, Genetic testing</td>
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<tr>
<td>Family history of sudden death</td>
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<tr>
<td>Left ventricular hypertrophy</td>
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<td></td>
<td></td>
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<tr>
<td>Family history of sudden death</td>
<td>Echo/MRI, Genetic testing</td>
<td></td>
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<tr>
<td>Epsilon wave</td>
<td>Echo/MRI, Genetic testing</td>
<td></td>
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<td>Pulmonary symptoms</td>
<td>Echo/MRI, Chest CT, Tissue biopsy</td>
<td>Sarcoïdosis</td>
<td>Immunosuppresion, Beta-blockers, Sotalol/amiodarone/ICD</td>
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<td>Dermatitis</td>
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<tr>
<td>Recent flu-like illness</td>
<td>Serology, Cardiac biopsy, Echo/MRI</td>
<td>Myocartitis</td>
<td>Beta-blockers, Sotalol/amiodarone/ICD</td>
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<td>Mid-systolic click</td>
<td>Echo/MRI, Genetic testing</td>
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<tr>
<td>Systolic murmur</td>
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<tr>
<td>Marfanoid body habitus</td>
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</table>

CT, computed tomography; ICD, implantable cardioverter-defibrillator; MRI, magnetic resonance imaging.
presence of PVCs with a LBBB morphology and QRS axis of $-90^\circ$ to $+110^\circ$ also suggests ARVC. In HCM patients, LVH may be associated with pathological Q-waves, giant ($\geq 10$ mm) T-negative wave, or ST depression.

**Treatment**

**Implantable cardioverter-defibrillator therapy**
The ICD is the primary therapy for patients with sustained PMVT or VF when there is no completely reversible cause.\(^5\text{,}4\text{a}\text{,}189\)

**Antiarrhythmic drug therapy**

While beta-adrenergic blockers may help to stabilize patients during acute ischaemia, the primary therapy for ischaemia-induced PMVT or VF is coronary revascularization. Beta-blockers are recommended for patients with LQTS and CPVT.\(^4\text{,}4\text{a}\text{,}190\text{–}192\) In small case series, quinidine has been shown to be effective for preventing polymorphic VT/VF recurrence in idiopathic VF, BrS, short QT syndrome, and ERS.\(^193\text{–}196\) Although calcium channel blockers (verapamil) in combination with beta-blockers have been proposed for treatment of CPVT,\(^197\text{,}198\) their efficacy seems quite limited. Flecainide may be considered in association with beta-blockers in case of recurring polymorphic VT/VF in the setting of CPVT\(^199\) and LQT3.\(^200\)

**Catheter ablation**

Catheter ablation may be considered for patients with recurrent PMVT or VF when there is a consistent PVC morphology (or a limited number of morphologies) that trigger these arrhythmias.\(^201\text{–}208\) When a patient has polymorphic VT/VF induced by the same PVC morphology, the target of catheter ablation is usually a rapidly firing focus situated in the Purkinje network of either the RV or LV.\(^204\) These Purkinje fibres may induce PMVT or VF in patients without SHD or in patients with prior MI.\(^204\text{,}206\) Purkinje network triggers of polymorphic VT or VF are characterized by episodes of frequent arrhythmias usually with the same initiating QRS morphology that is relatively narrow. Patients presenting with this syndrome should be monitored, ideally with continuous 12-lead electrocardiography, to identify the triggering PVC morphology. If possible, catheter ablation should be performed during a period of increased arrhythmia frequency to maximize the chances of recording electrograms from the triggering focus. In cases of recurrent polymorphic VT/VF in patients with the BrS, an epicardial substrate involving the RVOT may be amenable to catheter ablation.\(^209\text{,}210\) Even when catheter ablation of foci triggering PMVT or VF has been successful, an ICD remains indicated.\(^204\)

**The resuscitated cardiac arrest survivor**

Patients who are resuscitated from cardiac arrest must be rapidly evaluated for the presence of SHD, an inherited arrhythmia syndrome, a triggering VA focus, or a non-cardiac cause (see Figure 6). Immediately following resuscitation, the clinical focus must be to minimize cerebral damage, often with the use of therapeutic hypothermia.\(^161\text{–}164\) Evidence of MI or ischaemia usually requires prompt coronary angiography and revascularization.\(^166\) In addition, the function of both ventricles should be evaluated with echocardiography. These considerations have been discussed in detail in the preceding sections.

**Ventricular tachycardia/ventricular fibrillation storm**

Ventricular tachycardia/ventricular fibrillation storm represents a true medical emergency that requires a multi-disciplinary approach to care (Table 7).\(^70\text{,}211\text{–}231\) Ventricular tachycardia/ventricular fibrillation storm is generally defined as the occurrence of three or more episodes of VT or VF within 24 h, requiring either ATP or cardioversion/defibrillation. Upon hospitalization, the patient’s risk should be stratified according to haemodynamic tolerance of the clinical VT and co-morbidities.\(^211\) High-risk patients should be admitted to an intensive care unit and evaluated for sedation, intubation, and mechanical haemodynamic support.

Acute treatment is aimed to reduce VA episodes and maximize the chances of survival. For patients with an ICD, the detection criteria and therapies should be reprogrammed to minimize inappropriate shocks,\(^77\) prevent shocks for potentially self-terminating VTs, and favour ATP therapies when feasible. Even though triggers of VT/VF storm are only rarely found,\(^70\) patients should be screened for such reversible causes as electrolyte imbalances, ischaemia, acute valvular disease, and pro-arrhythmic drugs.

Antiarrhythmic drugs should be used for the acute phase to stabilize the patient.\(^214\text{,}223\) Beta-blockers have improved short-term outcome.\(^212\) Short-acting drugs, such as esmolol, might be considered in severely compromised patients, when an acute hypotensive effect is potentially likely.\(^228\) Even in patients already on oral beta-blocker therapy, intravenous administration of beta-blockers may help to reduce further ES episodes.\(^212\) Beta-blockers can be combined with amiodarone to improve rhythm stability.\(^212\) Because intravenous lidocaine is relatively ineffective for termination of haemodynamically stable VTs and its prophylactic use has been associated with higher mortality,\(^214\) this agent is a third choice drug for short-term treatment. In patients with severely impaired LV systolic function, the use of AADs should be weighed against the risks of worsening congestive heart failure and pro-arrhythmia.

Catheter ablation should be considered early after hospitalization (within 48 h) in patients with recurrent shocks despite acute treatment after the correction of metabolic, respiratory, and circulatory...
imbalances and a trial of AADs. Catheter ablation has been demonstrated to restore stable sinus rhythm maintenance during 7 days of in-hospital monitoring. Complete elimination of VT inducibility during programmed electrical stimulation after ablation is associated with reduced VT recurrence during long-term follow-up; prevention of clinical VT inducibility has also been associated with a significant reduction of cardiac mortality. Beneficial effects of catheter ablation on VT recurrences and survival are evident both in low- and high-risk patients. For patients who cannot be stabilized pharmacologically, neuraxial modulation, such as left cardiac sympathetic denervation (CSD) and spinal cord stimulation, may significantly reduce arrhythmias burden. This may allow stabilization before catheter ablation or LVAD implantation. Since VT/VF storm may be an indicator of poor prognosis, especially in patients with advanced SHD, early consultation with heart failure specialists should be considered to evaluate the advisability of mechanical cardiac support or cardiac transplantation.

Ventricular arrhythmias in patients with a left ventricular assist device

Left ventricular assist devices can be used as a bridge to cardiac transplantation or as a permanent, destination therapy for congestive heart failure. Despite their effectiveness, VAs occur in 25–59% of patients after LVAD implantation and are associated with a markedly increased risk of ICD shocks and overall mortality. Pre-operative VAs are the strongest predictor of VT or VF after LVAD implantation (HR 13.7). For patients with pre-operative VAs, the risk of post-operative VT or VF is ~50% within the first 18 months after LVAD implantation. Although LVADs provide haemodynamic support during VAs, patients may experience palpitations, dyspnoea, and right heart failure. Post-operative VT and VF in LVAD recipients tend to be refractory to AAD therapy and may require catheter ablation. Since many forms of VT in patients with non-ischaemic dilated cardiomyopathy have critical zones of reentrant circuits in the subepicardial myocardium, the post-operative pericardial scar is a potential source for VT/VF. LVAD implantation may result in permanent scarring in the subepicardial myocardium, and post-operative pericardial scarring produced by LVAD implantation may enhance the risk of VT/VF. Patients with non-ischaemic dilated cardiomyopathy who have pre-operative VT may benefit from concomitant surgical ablation of VT at the time of LVAD implantation.

Ventricular arrhythmias in congenital heart disease

Indications for programmed ventricular stimulation in patients with congenital heart disease

Inducible VAs (VF, MMVT, or PMVT) predict increased risk of both arrhythmia event and overall mortality in CHD patients carefully selected for programmed ventricular stimulation. Selection to enhance risk may include arrhythmia symptoms (sustained palpitations/syncope) and/or combinations of less robust predictors, such as older age, QRS duration >180 ms, complex ventricular ectopy, RV or LV dysfunction, and depressed exercise tolerance. In older patients with CHD and in patients with tetralogy of Fallot, positive ventricular stimulation is associated with
increased risk of ICD use, sudden death, and poor haemodynamic outcome, resulting in a positive predictive value of 20–60%. In contrast, EP testing in unselected and younger patient groups with CHD appears to have less predictive value. Supraventricular arrhythmia, particularly atrial tachycardia, is common in patients with CHD and may contribute a substantial portion of inappropriate ICD therapies, suggesting that assessment for atrial tachycardia should be included when EPSs are performed in these patients.

Multiple criteria should thus be used to determine which patients with VAs should undergo EPS, including symptoms, haemodynamic status, and surgical history (flowchart/Table 9). Deferring EPS in favour of clinical surveillance is warranted for patients with favourable risk profiles (good ventricular function, lesser grades of ectopy, minimal arrhythmia symptoms).

Management of ventricular tachycardia and premature ventricular complexes in patients with congenital heart disease

Clinical presentations that indicate the need for therapy of VAs in patients with CHD include arrhythmia-related symptoms and deterioration of ventricular function. In patients with CHD who have an increased risk of ICD use, sudden death, and poor haemodynamic outcome, resulting in a positive predictive value of 20–60%.

Table 8 Arrhythmia considerations in selected congenital heart disease

<table>
<thead>
<tr>
<th>Ventriculotomy</th>
<th>RV pressure/volume overload</th>
<th>SVT</th>
<th>VT</th>
<th>Bradycardia</th>
<th>Systemic ventricle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>+</td>
<td>LV</td>
</tr>
<tr>
<td>Atrial switch for d-TGA</td>
<td>+/-</td>
<td>+</td>
<td>++</td>
<td>+</td>
<td>RV</td>
</tr>
<tr>
<td>Ebstein’s disease</td>
<td>+/-</td>
<td>+</td>
<td>++</td>
<td>+</td>
<td>LV</td>
</tr>
<tr>
<td>Arterial switch for d-TGA</td>
<td>+/-</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>LV</td>
</tr>
<tr>
<td>Single ventricle</td>
<td>-</td>
<td>+</td>
<td>++</td>
<td>+</td>
<td>Either</td>
</tr>
<tr>
<td>Simple repairs</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>LV</td>
</tr>
</tbody>
</table>

d-TGA, transposition of the great vessels; LV, left ventricle; RV, right ventricle; SVT, supraventricular tachycardia; VT, ventricular tachycardia. SVT includes atrial reentry in addition to accessory pathway mediated and atrioventricular nodal reentrant tachycardia. Bradycardia includes relative frequency of heart block and significant sinus node dysfunction that may require pacing/limit drug therapy.

Figure 7 Management of VAs in CHD. ATP, antitachycardia pacing; CHD, congenital heart disease; d-TGA, transposition of the great vessels; ICD, implantable cardioverter-defibrillator; LVOTO, LV outflow tract obstruction; PMVT, polymorphic ventricular tachycardia; SMVT, sustained monomorphic ventricular tachycardia.
implanted ICD, frequent appropriate shocks often require prevention of recurrences. Strategies for management include suppressive antiarrhythmic therapy, ablation (catheter or surgical), and ATP. There are no prospective studies of sufficient power to be directive of therapy in this group of patients. Thus, recommendations regarding management of VA in this patient group are based on expert consensus.

Antiarrhythmic agents are often used to suppress PVCs and NSVT of lower grades. Efficacy is usually defined as symptomatic improvement or reduction in ectopic events, but suppression of PVCs is not known to be associated with change in mortality. Evidence for safety and efficacy of AAD therapy (e.g. mexiletine, propafenone, sotalol, and amiodarone)284–286 is derived from case series in populations with mixed arrhythmia mechanisms, or is inferred from studies performed in adult populations, although amiodarone has also been prospectively studied in an acute setting in a small number of paediatric patients with VT.287 These advanced antiarrhythmic agents are largely reserved for suppressing excessive arrhythmia in patients with ICD therapy or those where ICD therapy is deferred for anatomical reasons. Programming ICDs to high VF detection rates and long VT detection times is important to minimize unnecessary ATP or shocks for NSVT that would have been self-terminating without device intervention. While there are similarly no prospective data on beta-blockers in patients with CHD who have VAs, their broad safety profile results in them being a popular choice to suppress ectopy.

Several studies have documented the feasibility of using catheter ablation to treat MMVT in patients with CHD.288–291 Success rates in these studies have ranged from ~60 to >90% case series of 11–20 patients and in one case ablation has been proposed in conjunction with use of Class III antiarrhythmic agents.292 Access to the ventricular endocardium may be limited by some anatomical abnormalities or surgical corrections. Extrapolating from adult data with normal cardiac anatomy, in patients with frequent (>15% or more) monomorphic ventricular ectopy and newly recognized or progressive ventricular dysfunction, ablation of PVCs may be a useful adjunct if suppressive AAD therapy fails.23,293 The European Society of Cardiology guidelines for adults with CHD suggest the ablation may be reasonable as monotherapy for SMVT.

Recent mapping studies suggest predictable anatomical substrates for reentry in many patients with typical biventricular anatomy (i.e. that seen after repair of tetralogy of Fallot).294 These include the

Table 9 Clinical factors to consider when evaluating CHD patient for electrophysiological study

<table>
<thead>
<tr>
<th>Findings</th>
<th>Response</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Highest risk features</strong></td>
<td>Sustained MMVT Cardiac arrest</td>
</tr>
<tr>
<td><strong>High risk</strong></td>
<td>Syncope Sustained palpitations</td>
</tr>
<tr>
<td><strong>Intermediate risk</strong></td>
<td>Older age at initial repair (&gt;1 year) Older age (&gt;25–30 years) Prior palliative procedures</td>
</tr>
<tr>
<td><strong>RV haemodynamic burden</strong></td>
<td>LV dysfunction (&lt;55% for tetralogy) NSVT on monitoring</td>
</tr>
<tr>
<td>• RVp &gt; 50% systemic Moderate + pulmonary insufficiency RV end-diastolic volumes &gt; 150 mL/m² (Ref279) Increased heart size on CXR RV function &lt;45% QRSd &gt; 180 ms</td>
<td></td>
</tr>
<tr>
<td><strong>Low risk</strong></td>
<td>Simple ‘repairs’ without residual expected or less isolated ectopy on monitoring No symptoms</td>
</tr>
<tr>
<td><strong>Intermediate risk</strong></td>
<td>Older age (20–30 years) Prior palliative procedures Ventriculotomy</td>
</tr>
<tr>
<td><strong>Complicating Features</strong></td>
<td>Need for CRMD management Anticipated haemodynamic interventions</td>
</tr>
<tr>
<td>Pulmonary valve replacement Tricuspid valvuloplasty CRT</td>
<td></td>
</tr>
</tbody>
</table>

CHD, congenital heart disease; CRT, cardiac resynchronisation therapy; CXR, chest X-rays; EPS, electrophysiological study; ICD, implantable cardioverter-defibrillator; LV, left ventricular; MMVT, monomorphic ventricular tachycardia; NSVT, non-sustained ventricular tachycardia; RVp, right ventricular pressure; SVT, supraventricular tachycardia.
corridor between the tricuspid annulus and the RVOT, and the infundibular septum. While surgical ablation based on this anatomical template can be added to surgical repairs, the efficacy of this approach remains uncertain.

A recent, small study has suggested that ATP may be efficacious in paediatric and congenital patients with ICDS. The relative simplicity of this approach in combination with the more compelling evidence derived from adult studies suggest that this should be considered in all implanted patients.

Ventricular arrhythmias occur more frequently in patients with significant haemodynamic burden. New or increasing VAs should always trigger a careful evaluation of the underlying haemodynamic issues. However, correction of haemodynamic problems alone does not eliminate VAs.

Younger children and infants

The writing group has focused on the adult patient. These data are reasonably extrapolated to the older adolescent. For the younger adolescent, the school age child and particularly for infants both the natural history, the risk/benefit calculations for drugs, ablation, and device therapy may be different. Therefore, consultation with centres experienced in the management of children is appropriate. Recent guidelines focus on this particular population.

Evidence gaps and future research directions

The writing group acknowledges the lack of randomized controlled clinical trials to support many of the recommendations put forward in this document. While we acknowledge the historical reasons (see the Introduction section), the writing committee strongly supports controlled trials in adequately diagnosed contemporary patient cohorts. There is ample evidence to support the use of defibrillators in patients at highest risk for sudden death. While the initial approach to these patients was done in high-risk patients, the available data suggest that ECG parameters and/or cardiac imaging could better define those patients who will benefit from a defibrillator, and possibly delineate patient groups at lower risk who will not. The use of AADs for the suppression and symptomatic improvement of patients with VAs is in clear need of controlled trials. Such trials, possibly aiming for symptomatic improvement, e.g. measured by patient-reported outcomes in addition to monitoring of arrhythmia recurrence, seem feasible in relatively small patient cohorts. Ongoing studies are evaluating whether early interventional treatment of VAs by catheter ablation can improve outcomes in patients with an implanted defibrillator. Furthermore, long-term outcomes after catheter ablation in specific disease entities, e.g. RVOT tachycardia or fascicular tachycardia are available and support the use of ablation in these patients. The role of specific antiarrhythmic interventions (by AADs or catheter ablation) in addition to treatment of the underlying heart disease warrants further clinical research. Furthermore, a contemporary description and characterization of the causes and underlying cardiovascular diseases, including inherited arrhythmogenic syndromes, would allow better strategies to earlier identify patients at risk for sudden death, thus setting the stage for intervention trials aimed at reducing sudden death.

Conflict of interest: none declared.

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