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AHA/ACC/HRS GUIDELINE

2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death: Executive Summary

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society

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PREAMBLE

Since 1980, the American College of Cardiology (ACC) and American Heart Association (AHA) have translated scientific evidence into clinical practice guidelines with recommendations to improve cardiovascular health. These guidelines, which are based on systematic methods to evaluate and classify evidence, provide a cornerstone for quality cardiovascular care. The ACC and AHA sponsor the development and publication of guidelines without commercial support, and members of each organization volunteer their time to the writing and review efforts. Guidelines are official policy of the ACC and AHA.

Intended Use

Practice guidelines provide recommendations applicable to patients with or at risk of developing cardiovascular disease. The focus is on medical practice in the United States, but guidelines developed in collaboration with other organizations may have a global impact. Although guidelines may be used to inform regulatory or payer decisions, their intent is to improve patients' quality of care and align with patients' interests. Guidelines are intended to define practices meeting the needs of patients in most, but not all, circumstances and should not replace clinical judgment.

Clinical Implementation

Guideline-recommended management is effective only when followed by healthcare providers and patients. Adherence to recommendations can be enhanced by shared decision-making between healthcare providers and patients, with patient engagement in selecting interventions based on individual values, preferences, and associated conditions and comorbidities.

Methodology and Modernization

The ACC/AHA Task Force on Clinical Practice Guidelines (Task Force) continuously reviews, updates, and modifies guideline methodology on the basis of published standards from organizations including the Institute of Medicine[P-1,P-2] and on the basis of internal reevaluation. Similarly, the presentation and delivery of guidelines are reevaluated and modified on the basis of evolving technologies and other factors to facilitate optimal dissemination of information at the point of care to healthcare professionals.

Toward this goal, this guideline heralds the evolved format of presenting guideline recommendations and associated text called "modular knowledge chunk format." Each modular "chunk" includes a table of related recommendations, a brief synopsis, recommendation-specific supportive text, and when appropriate, flow diagrams or additional tables. References are provided within the modular chunk itself to facilitate quick review. This format also will facilitate seamless updating of guidelines with focused updates as new evidence is published, and content tagging for rapid electronic retrieval of related recommendations on a topic of interest. This evolved format was instituted when this guideline was near completion; therefore the current document represents a transitional formatting that best suits the text as written. Future guidelines will fully implement this format, including provisions for limiting the amount of text in a guideline.

Recognizing the importance of cost–value considerations in certain guidelines, when appropriate and feasible, an analysis of the value of a medication, device, or intervention may be performed in accordance with the ACC/AHA methodology.[P-3]

To ensure that guideline recommendations remain current, new data are reviewed on an ongoing basis, with full guideline revisions commissioned in approximately 6-year cycles. Publication of new, potentially practice-changing study results that are relevant to an existing or new medication, device, or management strategy will prompt evaluation by the Task Force, in consultation with the relevant guideline writing committee, to determine whether a focused update should be commissioned. For additional information and policies regarding guideline development, we encourage readers to consult the ACC/AHA guideline methodology manual[P-4] and other methodology articles.[P-5–P-8]

Selection of Writing Committee Members

The Task Force strives to avoid bias by selecting experts from a broad array of backgrounds. Writing committee members represent different geographic regions, sexes, ethnicities, races, intellectual perspectives/biases, and scopes of clinical practice. The Task Force may also invite organizations and professional societies with related interests and expertise to participate as partners, collaborators, or endorsers.

Relationships With Industry and Other Entities

The ACC and AHA have rigorous policies and methods to ensure that guidelines are developed without bias or improper influence. The complete relationships with industry and other entities (RWI) policy can be found online. Appendix 1 of the current document lists writing committee members' relevant RWI. For the purposes of full transparency, writing committee members' comprehensive disclosure information is available online, as is the comprehensive disclosure information for the Task Force.

Evidence Review and Evidence Review Committees

When developing recommendations, the writing committee uses evidence-based methodologies that are based on all available data.[P-4–P-7] Literature searches focus on randomized controlled trials (RCTs) but also include registries, nonrandomized comparative and descriptive studies, case series, cohort studies, systematic reviews, and expert opinion. Only key references are cited.

An independent evidence review committee (ERC) is commissioned when there are ≥1 questions deemed of utmost clinical importance that merit formal systematic

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review. This systematic review will strive to determine which patients are most likely to benefit from a test, medication, device, or treatment strategy and to what degree. Criteria for commissioning an ERC and formal systematic review include: a) the absence of a current authoritative systematic review; b) the feasibility of defining the benefit and risk in a time frame consistent with the writing of a guideline; c) the relevance to a substantial number of patients; and d) the likelihood that the findings can be translated into actionable recommendations. ERC members may include methodologists, epidemiologists, healthcare providers, and biostatisticians. When a formal systematic review has been commissioned, the recommendations developed by the writing committee on the basis of the systematic review are marked with "SR."

Guideline-Directed Management and Therapy

The term *guideline-directed management and therapy* (GDMT) encompasses clinical evaluation, diagnostic testing, and pharmacological and procedural treatments. For these and all recommended medication treatment regimens, the reader should confirm the dosage by reviewing product insert material and evaluate the treatment regimen for contraindications and interactions. The recommendations are limited to medications, devices, and treatments approved for clinical use in the United States.

Class of Recommendation and Level of Evidence

The Class of Recommendation (COR) indicates the strength of the recommendation, encompassing the estimated magnitude and certainty of benefit in proportion to risk. The Level of Evidence (LOE) rates the quality of scientific evidence that supports the intervention on the basis of the type, quantity, and consistency of data from clinical trials and other sources (Table 1).[P-4,P-6,P-8]

The reader is encouraged to consult the full-text guideline[P-9] for additional guidance and details about the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. The executive summary contains mainly the recommendations.

Glenn N. Levine, MD, FACC, FAHA
Chair, ACC/AHA Task Force on Clinical Practice Guidelines

1. INTRODUCTION

1.1. Methodology and Evidence Review

The recommendations listed in this clinical practice guideline are, whenever possible, evidence-based. An initial extensive evidence review, which included literature derived from research involving human subjects, published in English, and indexed in MEDLINE (through PubMed), EMBASE, the Cochrane Library, the Agency for Healthcare Research and Quality, and other selected databases relevant to this guideline, was conducted from April 2016 to September 2016. Key search words included, but were not limited, to the following: sudden cardiac death, ventricular tachycardia, ventricular fibrillation, premature ventricular contractions, implantable cardioverter-defibrillator, subcutaneous implantable cardioverter-defibrillator, wearable cardioverter-defibrillator, and catheter ablation. Additional relevant studies published through March 2017, during the guideline writing process, were also considered by the writing committee, and added to the evidence tables when appropriate. The final evidence tables are included in the Online Data Supplement and summarize the evidence used by the writing committee to formulate recommendations. Additionally, the writing committee reviewed documents related to ventricular arrhythmias (VA) and sudden cardiac death (SCD) previously published by the ACC, AHA, and the Heart Rhythm Society (HRS). References selected and published in this document are representative and not all-inclusive.

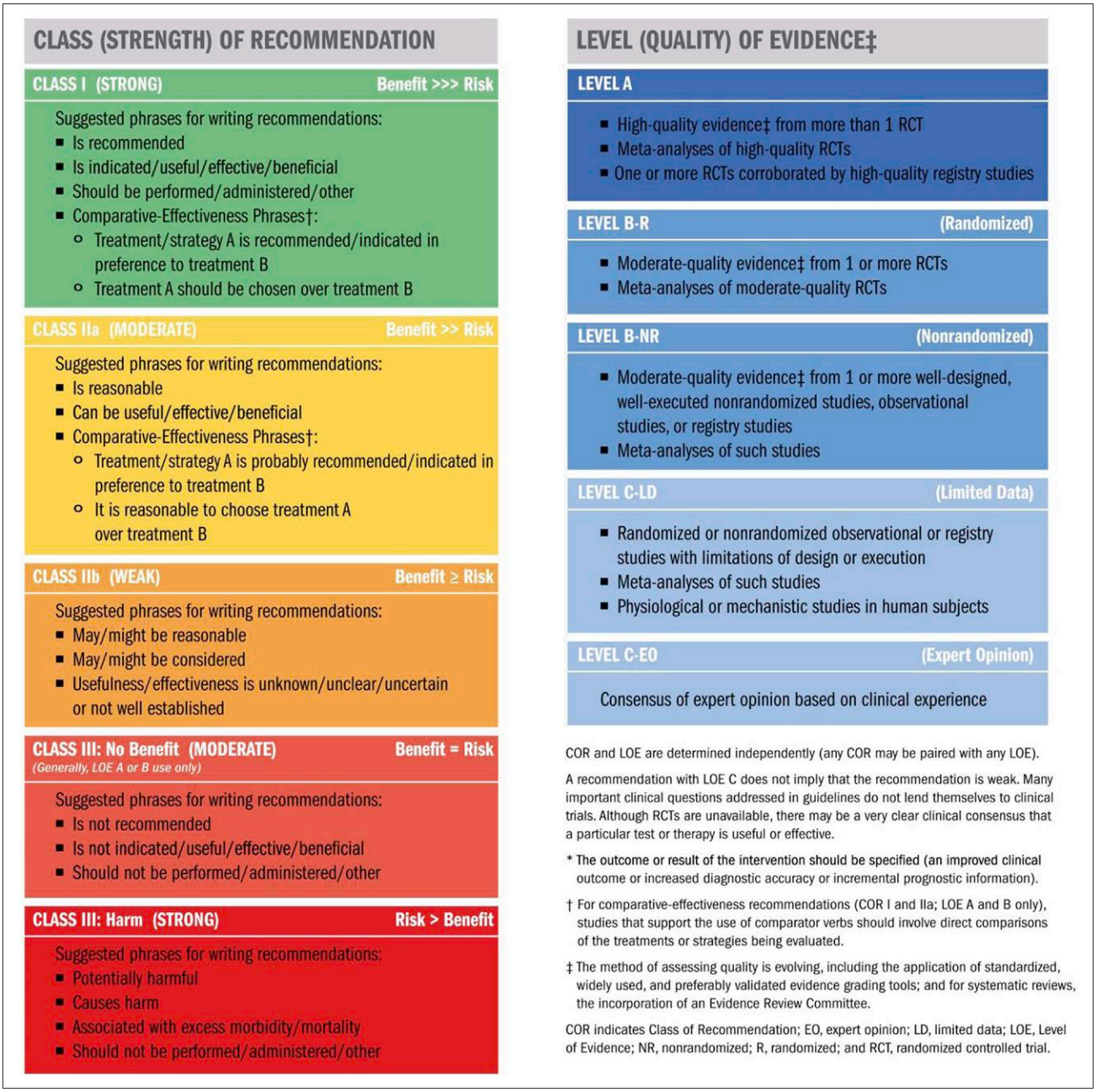
As noted in the Preamble, an independent ERC was commissioned to perform a formal systematic review of 2 important clinical questions for which clear literature and prior guideline consensus were felt to be lacking or limited (Table 2). The results of the ERC review were considered by the writing committee for incorporation into this guideline. Concurrent with this process, writing committee members evaluated other published data relevant to the guideline. The findings of the ERC and the writing committee members were formally presented and discussed, then guideline recommendations were developed. The "Systematic Review for the 2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death" is published in conjunction with this guideline.[S1.4-1]

The ACC and AHA have acknowledged the importance of value in health care and have called for eventual development of a Level of Value for clinical practice recommendations.[S1.4-2] Available cost-effectiveness data were determined to be sufficient to support 2 specific recommendations in this guideline (see Sections 7.1.1 and 7.1.2). As a result, a Level of Value was assigned to those 2 recommendations on the basis of the "ACC/AHA Statement on Cost/Value Methodology in Clinical Practice Guidelines and Performance Measures," as shown in Table 3.[S1.4-2] Available quality of life (QoL) data were deemed to be insufficient to support specific recommendations in this guideline.

1.2. Organization of the Writing Committee

The writing committee consisted of cardiac electrophysiologists (including those specialized in pediatrics), general

Table 1. Applying Class of Recommendation and Level of Evidence to Clinical Strategies, Interventions, Treatments, or Diagnostic Testing in Patient Care* (Updated August 2015)



adult and pediatric cardiologists (including those specialized in critical care and acute coronary syndromes [ACS], genetic cardiology, heart failure, and cost-effectiveness analyses), a geriatrician with expertise in terminal care and shared decision-making, and a lay representative, in addition to representatives from the ACC, AHA, HRS, and the Heart Failure Society of America (HFSA).

1.3. Document Review and Approval

This document was reviewed by 2 official reviewers nominated by the ACC, AHA, and HRS; 1 official lay reviewer nominated by the AHA; 1 organizational reviewer nominated by the HFSA; and 28 individual content reviewers. Reviewers' RWI information was distributed to the writing committee and is published in this document (Appendix 2).This document was approved for publication by the governing bodies of the ACC, the AHA, and the HRS; and endorsed by the HFSA.

1.4. Scope of the Guideline

The purpose of this AHA/ACC/HRS document is to provide a contemporary guideline for the management of

Table 2. Systematic Review Questions on SCD Prevention

Question Number	Question	Section Number
1	For asymptomatic patients with Brugada syndrome, what is the association between an abnormal programmed ventricular stimulation study and SCD and other arrhythmia endpoints?	6.9.1.3.
2	What is the impact of ICD implantation for primary prevention in older patients and patients with significant comorbidities?	9.2.

ICD indicates implantable cardioverter-defibrillator; and SCD, sudden cardiac death.

adults who have VA or who are at risk for SCD, including diseases and syndromes associated with a risk of SCD from VA. This guideline supersedes the "ACC/AHA/ESC 2006 Guidelines for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death."[S1.4-4] It also supersedes some sections of the "ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities,"[S1.4-5] specifically those sections on indications for the implantable cardioverter-defibrillator (ICD); and, it updates the SCD prevention recommendations in the "2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy."[S1.4-6] Some recommendations from the earlier guidelines have been updated as warranted by new evidence or a better understanding of existing evidence, and irrelevant or overlapping recommendations were deleted or modified.

In the current guideline, sudden cardiac arrest (SCA) is defined as the "sudden cessation of cardiac activity so that the victim becomes unresponsive, with no normal breathing and no signs of circulation."[S1.4-7] If corrective measures are not taken rapidly, this condition progresses to SCD. Cardiac arrest is used to signify an event that can be reversed, usually by cardiopulmonary resuscitation (CPR), administration of medications and/or defibrillation or cardioversion. SCA and SCD can result from causes other than VA, such as bradyarrhythmias, electromechanical dissociation, pulmonary embolism, intracranial hemorrhage, and aortic dissection; however, the scope of this document includes only SCA and SCD due to VA.

Table 3. Proposed Integration of Level of Value Into Clinical Practice Guideline Recommendations*

Level of Value
High value: Better outcomes at lower cost or ICER <$50 000 per QALY gained
Intermediate value: $50 000 to <$150 000 per QALY gained
Low value: ≥$150 000 per QALY gained
Uncertain value: Value examined but data are insufficient to draw a conclusion because of no studies, low-quality studies, conflicting studies, or prior studies that are no longer relevant
Not assessed: Value not assessed by the writing committee
Proposed abbreviations for each value recommendation: *Level of Value: H to indicate high value; I, intermediate value; L, low value; U, uncertain value; and NA, value not assessed*

*Dollar amounts used in this table are based on US GDP data from 2012 and were obtained from WHO-CHOICE Cost-Effectiveness Thresholds.[S1.4-3]

GDP indicates gross domestic product; ICER, incremental cost-effectiveness ratio; QALY, quality-adjusted life-years; and WHO-CHOICE, World Health Organization Choosing Interventions that are Cost-Effective.

Reproduced from Anderson, et al.[S1.4-2]

This guideline includes indications for ICDs for the treatment of VA and prevention of SCD, but it does not delve into details on individual device selection and programming, including considerations relevant to cardiac resynchronization therapy (CRT), bradycardia pacing, and hemodynamic monitoring. These important aspects of ICD management have been covered in an HRS expert consensus statement.[S1.4-8] An AHA science advisory discusses the use of wearable cardioverter-defibrillators.[S1.4-9] The findings of that document were reviewed; however, recommendations on this topic were developed independently of that document. This guideline includes indications for catheter ablation of VA, but does not provide recommendations on specific techniques or ablation technologies, which were beyond the scope of this document.

Recommendations for interventional therapies, including ablation and the implantation of devices, apply only if these therapies can be implemented by qualified clinicians, such that outcomes consistent with published literature are a reasonable expectation. The writing committee agreed that a high degree of expertise was particularly important for performance of catheter ablation of VA, and this point is further emphasized in relevant sections. In addition, all recommendations related to ICDs require that meaningful survival of >1 year is expected; meaningful survival means that a patient has a reasonable quality of life and functional status.

Although this document is aimed at the adult population (≥18 years of age) and offers no specific recommendations for pediatric patients, some of the literature on pediatric patients was examined. In some cases, the data from pediatric patients beyond infancy helped to inform this guideline.

The writing committee recognized the importance of shared decision-making and patient-centered care and, when possible, it endeavored to formulate recommendations relevant to these important concepts. The importance of a shared decision-making process in which the patient, family, and clinicians discuss risks and benefits of diagnostic and treatment options and consider the patients' personal preferences is emphasized (see Section 15).

In developing this guideline, the writing committee reviewed previously published guidelines and related statements. Table 4 contains a list of guidelines and statements deemed pertinent to this writing effort and is intended for use as a resource, obviating repetition of existing guideline recommendations.

During final production review of the guidelines, several recommendations were refined to better reflect the data and current recommended medical practice. These refinements were reviewed and approved by the writing committee, the Task Force, and ACC, AHA, and HRS organizational leadership. These recommendations were:

- Section 6.1.1., recommendation 1
- Section 6.1.3., recommendation 2
- Section 6.2.1., recommendation 1
- Section 6.9.1.4., recommendation 2
- Section 9.4., recommendation 6

Readers should refer to these sections for the updated text.

Table 4. Associated Guidelines and Statements

Title	Organization	Publication Year (Reference)
Guidelines		
Syncope	ACC/AHA/HRS	2017[S1.4-10]
Heart failure	ACCF/AHA	2017[S1.4-11] 2016,[S1.4-12] and 2013[S1.4-13]
Valvular heart disease	AHA/ACC	2017[S1.4-14] and 2014[S1.4-15]
Supraventricular tachycardia	ACC/AHA/HRS	2015[S1.4-16]
Ventricular arrhythmias and the prevention of sudden cardiac death	ESC	2015[S1.4-17]
Guidelines for cardiopulmonary resuscitation and emergency cardiovascular care	AHA	2015[S1.4-18]
Atrial fibrillation	AHA/ACC/HRS	2014[S1.4-19]
Non–ST-elevation acute coronary syndromes	AHA/ACC	2014[S1.4-20]
Assessment of cardiovascular risk	ACC/AHA	2013[S1.4-21]
ST-elevation myocardial infarction	ACCF/AHA	2013[S1.4-22]
Acute myocardial infarction in patients presenting with ST-segment elevation	ESC	2012[S1.4-23]
Device-based therapies for cardiac rhythm abnormalities	ACCF/AHA/HRS	2012[S1.4-24]
Coronary artery bypass graft surgery	ACCF/AHA	2011[S1.4-25]
Hypertrophic cardiomyopathy	ACCF/AHA	2011[S1.4-6]
Percutaneous coronary intervention	ACCF/AHA/SCAI	2011[S1.4-26]
Secondary prevention and risk reduction therapy for patients with coronary and other atherosclerotic vascular disease	AHA/ACCF	2011[S1.4-27]
Scientific Statements		
Wearable cardioverter-defibrillator therapy for the prevention of sudden cardiac death	AHA	2016[S1.4-9]
Optimal implantable cardioverter defibrillator programming and testing	HRS/EHRA/APHRS/SOLAECE	2016[S1.4-8]
Treatment of cardiac arrest: current status and future directions: strategies to improve cardiac arrest survival	IOM	2015[S1.4-28]
Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities	ACC/AHA	2015[S1.4-29]
Ventricular arrhythmias	EHRA/HRS/APHRS	2014[S1.4-30]
Arrhythmias in adult congenital heart disease	PACES/HRS	2014[S1.4-31]
Implantable cardioverter-defibrillator therapy in patients who are not included or not well represented in clinical trials	HRS/ACC/AHA	2014[S1.4-32]
Cardiac sarcoidosis	HRS	2014[S1.4-33]
Inherited primary arrhythmia syndromes	HRS/EHRA/APHRS	2013[S1.4-34]

ACC indicates American College of Cardiology; ACCF, American College of Cardiology Foundation; AHA, American Heart Association; APHRS, Asia Pacific Heart Rhythm Society; EHRA, European Heart Rhythm Association; ESC, European Society of Cardiology; HRS, Heart Rhythm Society; PACES, Pediatric and Congenital Electrophysiology Society; SCAI, Society for Cardiovascular Angiography and Interventions; and, SOLAECE, Sociedad Latinoamericana de Estimulacion Cardiaca y Electrofisiologia.

1.5. Abbreviations

Abbreviation	Meaning/Phrase
ACS	acute coronary syndrome
CPR	cardiopulmonary resuscitation
CRT	cardiac resynchronization therapy
ECG	electrocardiogram
ERC	evidence review committee
GDMT	guideline-directed management and therapy
HCM	hypertrophic cardiomyopathy
HF	heart failure
HF*p*EF	heart failure with preserved ejection fraction
HF*r*EF	heart failure with reduced ejection fraction
ICD	implantable cardioverter-defibrillator
LV	left ventricular
LVAD	left ventricular assist device
LVEF	left ventricular ejection fraction
MI	myocardial infarction
NICM	nonischemic cardiomyopathy
NSVT	nonsustained ventricular tachycardia
PCI	percutaneous coronary intervention
PVC	premature ventricular complex
QoL	quality of life
RCT	randomized controlled trial
RVOT	right ventricular outflow tract
SCA	sudden cardiac arrest
SCD	sudden cardiac death
VA	ventricular arrhythmia
VT	ventricular tachycardia

2. EPIDEMIOLOGY

2.1. General Concepts

VA include a spectrum that ranges from premature ventricular complex (PVC) to ventricular fibrillation (VF), with a clinical presentation that ranges from a total lack of symptoms to cardiac arrest. Most life-threatening VA are associated with ischemic heart disease, particularly in older patients.[S2.2.2-1] The risks of VA and SCD vary in specific populations with different underlying cardiac conditions, and with specific family history and genetic variants, and this variation has important implications for studying and applying therapies.

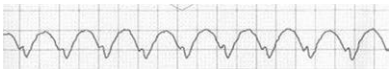
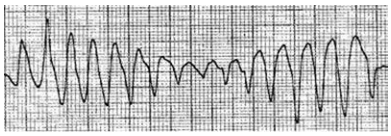
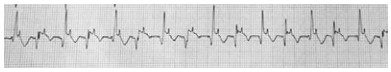
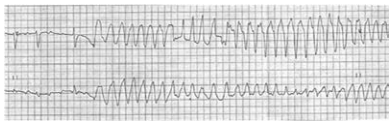
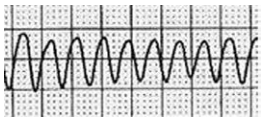
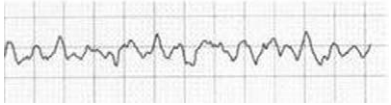
2.1.1. Premature Ventricular Complexes and Nonsustained VT

PVCs are common and increase in frequency with age. Although PVCs were found in a healthy military population in only 0.6% of those <20 years of age and 2.7% of those >50 years of age[S2.2.2-5] on 12-lead ECGs, longer term monitoring shows PVCs in about 50% of all people with or without heart disease.[S2.2.2-6] The presence of PVCs on 2 minutes of monitoring of middle-aged patients in the ARIC (Atherosclerosis Risk In Communities) study was associated with increased risk of both ischemic heart disease events and mortality, with or without prevalent ischemic heart disease.[S2.2.2-7,S2.2.2-8] In the general population, frequent PVCs, which are defined as the presence of at least 1 PVC on a 12-lead ECG or >30 PVCs per hour, are associated with increased cardiovascular risk and increased mortality.[S2.2.2-9] In a study from Taiwan of patients without sustained VT or structural heart disease who had 24-hour Holter monitoring for clinical evaluation, multifocal PVCs were associated with increased risk of death and nonfatal cardiovascular adverse outcomes.[S2.2.2-10] In the same population, nonsustained ventricular tachycardia (NSVT) was independently associated with increased risk of death and other cardiovascular adverse outcomes, including stroke.[S2.2.2-11] An association of PVCs with increased risk of stroke was also seen in the ARIC population.[S2.2.2-8]

Because some studies have shown an association of PVCs with adverse outcomes, the detection of PVCs, particularly if multifocal and frequent, is generally considered a risk factor for adverse cardiovascular outcomes, and such patients are generally evaluated to ensure they do not have underlying conditions (eg, ischemic heart disease, left ventricular [LV] dysfunction) that warrant further treatment to reduce risk. PVC and NSVT in patients with cardiovascular disease are common and have been associated with adverse outcomes.[S2.2.2-12,S2.2.2-13] In CAST (Cardiac Arrhythmia Suppression Trials), treatment of patients with post-myocardial infarction (MI) who took antiarrhythmic medications (eg, flecainide, encainide, moricizine) increased the risk of death despite suppression of VA.[S2.2.2-14,S2.2.2-15] Treatment of PVCs with antiarrhythmic medications has not been shown to reduce mortality and, in the post-MI population, treatment with class I sodium channel–blocking medications (eg, quinidine, flecainide) increases the risk of death.[S2.2.2-15,S2.2.2-16] Likewise, in patients with a reduced LVEF class I, sodium channel–blocking medications and d-sotalol increase the risk of death.[S2.2.2-16,S2.2.2-17] Beta blockers, nondihydropyridines calcium channel blockers, and some antiarrhythmic medications may relieve symptoms of palpitations.[S2.2.2-18]

PVCs that occur during an exercise test are associated with a higher risk of death.[S2.2.2-19] In 1 study, PVCs that occur during recovery are a stronger predictor of death than PVCs occurring only during exercise.[S2.2.2-20] However, PVCs are common in trained athletes who have palpitations, in whom there does not appear to be increased risk of death based on studies of small numbers of athletes, at least in those without other cardiovascular abnormalities.[S2.2.2-21,S2.2.2-22] Complex PVCs may not represent a benign finding in endurance athletes. An electrophysiological study may be needed to assess patients' arrhythmogenic risk.[S2.2.2-22] Very frequent PVCs, >10 000 to 20 000 a day, can be associated with depressed LV

Table 5. Table of Definitions of Commonly Used Terms in this Document

Term	Definition or Description
Ventricular tachycardia[S2.2.2-2]	Cardiac arrhythmia of ≥3 consecutive complexes originating in the ventricles at a rate >100 bpm (cycle length: <600 ms). Types of VT: Sustained: VT >30 s or requiring termination due to hemodynamic compromise in <30 s. Nonsustained/unsustained: ≥3 beats, terminating spontaneously. Monomorphic: Stable single QRS morphology from beat to beat. Polymorphic: Changing or multiform QRS morphology from beat to beat. Bidirectional: VT with a beat-to-beat alternation in the QRS frontal plane axis, often seen in the setting of digitalis toxicity or catecholaminergic polymorphic VT Monomorphic VT  Polymorphic VT  Bidirectional VT 
Torsades de pointes[S2.2.2-2]	Torsades de pointes is polymorphic VT that occurs in the setting of a long QT interval and is characterized by a waxing and waning QRS amplitude. It often has a long-short initiating sequence with a long coupling interval to the first VT beat and may present with salvos of NSVT. The twisting of the points, although characteristic, may not always be seen, especially if the episode is nonsustained or if only a limited number of leads are available. Torsades de pointes can result from bradycardia including high-grade AV block that leads to a long-short sequence initiating torsades de pointes. 
Ventricular flutter[S2.2.2-2]	A regular VA ≈300 bpm (cycle length: 200 ms) with a sinusoidal, monomorphic appearance; no isoelectric interval between successive QRS complexes. 
Ventricular fibrillation[S2.2.2-2]	Rapid, grossly irregular electrical activity with marked variability in electrocardiographic waveform, ventricular rate usually >300 bpm (cycle length: <200 ms). 
Sudden cardiac arrest[S2.2.2-2]	SCA is the sudden cessation of cardiac activity such that the victim becomes unresponsive, with either persisting gasping respirations or absence of any respiratory movements, and no signs of circulation as manifest by the absence of a perceptible pulse. An arrest is presumed to be of cardiac etiology unless it is known or likely to have been caused by trauma, drowning, respiratory failure or asphyxia, electrocution, drug overdose, or any other noncardiac cause.
Sudden cardiac death[S2.2.2-2]	Sudden and unexpected death occurring within an hour of the onset of symptoms, or occurring in patients found dead within 24 h of being asymptomatic and presumably due to a cardiac arrhythmia or hemodynamic catastrophe.
VT/VF storm[S2.2.2-3]	VT/VF storm (electrical storm or arrhythmic storm) refers to a state of cardiac electrical instability that is defined by ≥3 episodes of sustained VT, VF, or appropriate shocks from an ICD within 24 h.
Primary prevention ICD[S2.2.2-2]	ICD placement with the intention of preventing SCD in a patient who has not had sustained VT or SCA but who is at an increased risk for these events.
Secondary prevention ICD[S2.2.2-2]	ICD placement in a patient with prior SCA, sustained VT, or syncope caused by VA.
Structural heart disease*	This term encompasses IHD, all types of cardiomyopathy, valvular heart disease, and adult congenital heart disease.
Cardiac channelopathy[S2.2.2-4]	Arrhythmogenic disease due to a genetic abnormality that results in dysfunction of a cardiac ion channel (eg, long QT syndrome, catecholaminergic polymorphic VT).

*The definition of this term may differ across publications. Refer to the entry for the definition used in this document.

AV indicates atrioventricular; ICD, implantable cardioverter-defibrillator; IHD, ischemic heart disease; NSVT, nonsustained ventricular tachycardia; SCA, sudden cardiac arrest; SCD, sudden cardiac death; VA, ventricular arrhythmia; VF, ventricular fibrillation; and VT, ventricular tachycardia.

function in some patients that is reversible with control of the PVCs, and has been referred to as PVC-induced cardiomyopathy.[S2.2.2-23,S2.2.2-24] (See also Section 8.5. PVC-Induced Cardiomyopathy.) Very rarely, idiopathic PVCs from the outflow tract may trigger malignant VA in patients without structural heart disease.[S2.2.2-25,S2.2.2-26]

2.1.2. VT and VF During ACS

Approximately half of patients with out-of-hospital cardiac arrest with the first rhythm identified as VF and who survive to hospital admission have evidence of acute MI (AMI).[S2.2.2-27] Of all out-of-hospital cardiac arrests, >50% will have significant coronary artery lesions on acute coronary angiography.[S2.2.2-27] Of patients hospitalized with AMI, 5% to 10% have VF or sustained VT prior to hospital presentation, and another 5% will have VF or sustained VT after hospital arrival, most within 48 hours of admission. A study of patients with non–ST-elevation ACS who underwent cardiac catheterization within 48 hours found VT/VF in 7.6% of patients, with 60% of those events within 48 hours of admission.[S2.2.2-28] Accelerated idioventricular rhythm is a common arrhythmia in patients with acute MI, including patients with ST-segment elevation MI undergoing primary percutaneous coronary intervention (PCI). Accelerated idioventricular rhythm is more closely related to the extent of infarction than to reperfusion itself.[S2.2.2-29]

Sustained VA that occurs in the setting of an ACS is more often polymorphic VT or VF than monomorphic VT. Risk factors for VT/VF include prior history of hypertension, prior MI, ST-segment changes at presentation, and chronic obstructive pulmonary disease.[S2.2.2-30] A nationwide Danish study found that 11.6% of patients with ST-segment elevation MI who underwent PCI had VF prior to the PCI, and that VF was associated with alcohol consumption, preinfarction angina, anterior infarct location, and complete coronary occlusion at the time of coronary angiography.[S2.2.2-31] In a select group of patients undergoing primary PCI in a clinical trial, 5.7% developed sustained VT or VF, with two thirds of these events occurring prior to the end of the catheterization, and 90% within 48 hours from the procedure. VT or VF after primary PCI was associated with lower blood pressure, higher heart rate, poor coronary flow at the end of the procedure, and incomplete resolution of ST elevation.[S2.2.2-32] Importantly, and in contrast to some earlier studies, VT or VF at any time was associated with a substantially higher risk of death within 90 days. Late VT or VF (after 48 hours of hospital presentation) was associated with a higher risk of death than early VT or VF (within 48 hours of hospital presentation).[S2.2.2-33]

2.1.3. Sustained VT and VF Not Associated With ACS

Patients with structural heart disease are at an increased risk for sustained VT and VF. Sustained VT that is not associated with an ACS is often monomorphic as it is usually due to scar-related reentry, but it may degenerate to VF.[S2.2.2-34] The risk and predictors of VT in patients with structural heart disease depend on the type, severity, and duration of structural heart disease, increasing with the severity of ventricular dysfunction and the presence of symptomatic HF. Monomorphic VT occurring in the absence of structural heart disease is commonly referred to as idiopathic VT and is often due to an automatic focus in a characteristic location, giving rise to typical electrocardiographic appearances. Polymorphic VT and VF occurring in the absence of structural heart disease are rare and may be due to a cardiac channelopathy,[S2.2.2-35,S2.2.2-36] medication-induced long QT syndrome,[S2.2.2-36] or they may be idiopathic.[S2.2.2-37,S2.2.2-38]

2.2. Sudden Cardiac Death

2.2.1. Incidence of SCD

SCA and its most common consequence, SCD, constitute major public health problems, accounting for approximately 50% of all cardiovascular deaths,[S2.2.2-1,S2.2.2-39] with at least 25% being first symptomatic cardiac events.[S2.2.2-1,S2.2.2-40,S2.2.2-41] In addition, analyses of the magnitude of SCD are limited, in part because of the broad range of estimates of the risk based on different epidemiological methods.[S2.2.2-42] During the past 20 to 30 years, SCD accounted for approximately 230 000 to 350 000 deaths per year in the United States, with a range of <170 000 to >450 000, depending on epidemiological methods, data sources, and inclusion criteria.[S2.2.2-41,S2.2.2-43] The lowest of these extremes came from national extrapolation of data from specific local programs, while the highest rates included noncardiac causes of sudden death such as pulmonary embolism or intracranial bleeding. The mid-range numbers were largely based on death certificate studies that required a code inclusive of ischemic heart disease.

The 2017 update of cardiovascular statistics from the AHA estimated the total annual burden of out-of-hospital cardiac arrest at 356 500.[S2.2.2-44] An additional 209 000 in-hospital cardiac arrests occur annually.[S2.2.2-45] Among the out-of-hospital cardiac arrest group, approximately 357 000 events trigger emergency rescue response, with 97% occurring in adults >18 years of age.

The survival statistics for out-of-hospital cardiac arrest remain disappointing, with an estimated 10% overall survival rate.[S2.2.2-44] Among the subgroup of 70% of out-of-hospital cardiac arrests that occur in the home, survival is 6%. The best reported outcomes are from locations with highly developed and publicly visible emergency rescue response, along with the combination of public location of cardiac arrest,

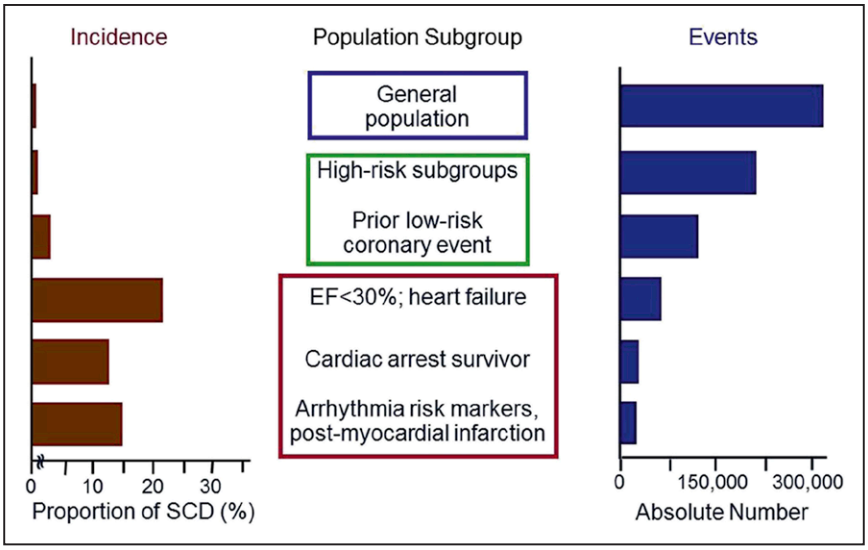


Figure 1A. SCD incidence and total events.[S2.2.2-1]
EF indicates ejection fraction; and SCD, sudden cardiac death.

bystander witnesses willing to provide CPR, first responders arriving quickly, shockable rhythm at initial contact, availability of automated external defibrillators (AEDs), and possibly a benefit from telecommunication-directed CPR.[S2.2.2-46,S2.2.2-47] Survival to hospital discharge after in-hospital cardiac arrests is estimated to be 24%.[S2.2.2-48] In all settings, survival statistics appear to be better when rhythms recorded by responders are shockable (VF, pulseless VT), compared with pulseless electrical activity or asystole.[S2.2.2-49] Although the apparent increase in the incidence of pulseless electrical activity or asystole could be due to the later arrival of medical care, the decrease in the incidence of shockable rhythm has also been attributed, in part, to improvements in diagnosis and treatment of structural heart disease.[S2.2.2-40]

2.2.2. Population Subgroups and Risk Prediction

Risk prediction for SCA and SCD is complex. Risk analysis is divided into 2 general categories: population risk prediction and individual risk prediction.[S2.2.2-41,S2.2.2-50] Conventional epidemiological markers provide insight into probabilities for the development of ischemic heart disease within a general class of subjects, but adequately tested and validated profiles for SCA risk stratification of individuals in the general population do not presently exist. The challenge of defining SCA risk in individuals derives from a population model characterized by large numbers of events diluted into a very large denominator (Figure 1). The overall population can be subgrouped into categories based on integration of age, presence and extent of disease, and identification of small, high-risk subgroups within the large denominator general population.

Increasing age is a strong predictor of risk for SCA, but it is not linear. Risk in the general population, over time, beginning at 35 years of age has been estimated at 1 per 1000 population per year, increasing from a risk <1000 at the younger end of that spectrum to a higher risk in the elderly.[S2.2.2-41] However, an analysis of

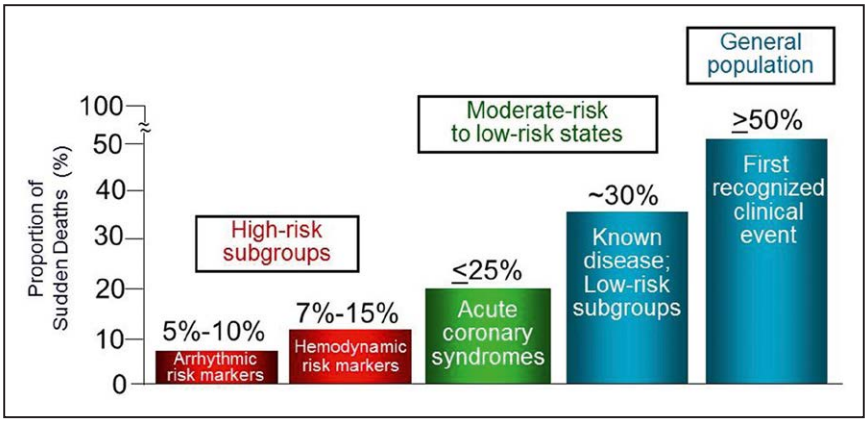


Figure 1B. SCD and clinical subsets.[S2.2.2-1]
SCD indicates sudden cardiac death.

lifetime risk of SCD, derived from the Framingham data, suggested that the incidence of SCD decreases in later years, especially in people >75 years of age.[S2.2.2-51] The data also suggested that SCD is uniformly more common in men than in women at all age groups. In contrast, the population of children, adolescents, and young adults has an overall annual risk of 1 per 100 000, and there is somewhat a higher risk of SCD at the younger end of that age range.[S2.2.2-41] An age-associated transition range, from the mid-20s to 35 to 40 years of age, is characterized by a steep increase in risk from that of the adolescent group to the middle-aged group, corresponding to the emergence of ischemic heart disease.

Although ischemic heart disease remains the most common underlying substrate associated with SCD, the incidence of ischemic heart disease-related SCD appears to be decreasing,[S2.2.2-52] with various forms of cardiomyopathy associated with myocardial fibrosis and LV hypertrophy increasing.[S2.2.2-53] In addition, a trend over time has suggested that out-of-hospital cardiac arrest patients who are admitted alive to a hospital are becoming more likely to have high-risk clinical profiles, as opposed to manifest disease.[S2.2.2-54] The younger population—children, adolescents, and young adults—is affected by a series of disorders that manifest earlier in life, including the genetic structural disorders and cardiac channelopathies, myocarditis, congenital heart disease, and other rare disorders.[S2.2.2-43] During the transition range, from the mid-20s to the mid-30s, causes of SCA and SCD include a lower proportion of inherited diseases and increasing proportion of ischemic heart disease (>40% of cases).[S2.2.2-43]

Despite the small progress that has been made in risk prediction of SCA and SCD, the greatest challenge is to identify the relatively small, high-risk subgroups concealed within the large general population who have no identified disease but are at risk of SCA as their first cardiac event (Figure 1).[S2.2.2-50]

3. GENERAL EVALUATION OF PATIENTS WITH DOCUMENTED OR SUSPECTED VA

3.1. History and Physical Examination

Recommendation for Syncope* Referenced studies that support the recommendation are summarized in Online Data Supplement 1.		
COR	**LOE**	**Recommendation**
I	B-NR	1. Patients presenting with syncope for which VA is documented, or thought to be a likely cause, should be hospitalized for evaluation, monitoring, and management.[S3.1-1–S3.1-4]

*This section covers practices that are well accepted, and a new recommendation was determined to only be warranted for syncope.

Table 6. Important Considerations in the Evaluation of Patients With Known or Suspected VA

Component	Assessment and Findings Relevant for VA and/or SCD Risk
History	1. Symptoms/events related to arrhythmia: Palpitations, lightheadedness, syncope, dyspnea, chest pain, cardiac arrest
	2. Symptoms related to underlying heart disease: Dyspnea at rest or on exertion, orthopnea, paroxysmal nocturnal dyspnea, chest pain, edema
	3. Precipitating factors: Exercise, emotional stress
	4. Known heart disease: Coronary, valvular (eg, mitral valve prolapse), congenital heart disease, other
	5. Risk factors for heart disease: Hypertension, diabetes mellitus, hyperlipidemia, and smoking
	6. Medications
	Antiarrhythmic medications
	Other medications with potential for QT prolongation and torsades de pointes
	Medications with potential to provoke or aggravate VA
	Stimulants including cocaine and amphetamines
	Supplements including anabolic steroids
	Medication-medication interaction that could cause QT prolongation and torsades de pointes
	7. Past medical history
	Thyroid disease
	Acute kidney injury, chronic kidney disease, or electrolyte abnormalities
	Stroke or embolic events
	Lung disease
	Epilepsy (arrhythmic syncope can be misdiagnosed as epilepsy)
	Alcohol or illicit drug use
	Use of over-the-counter medications that could cause QT prolongation and torsades de pointes
	Unexplained motor vehicle crashes
Family History	1. SCD, SCA, or unexplained drowning in a first-degree relative
	2. SIDS or repetitive spontaneous pregnancy losses given their potential association with cardiac channelopathies
	3. Heart disease
	IHD
	Cardiomyopathy: Hypertrophic, dilated, ARVC
	Congenital heart disease
	Cardiac channelopathies: Long QT, Brugada, Short QT, CPVT
	Arrhythmias
	Conduction disorders, pacemakers/ICDs
	4. Neuromuscular disease associated with cardiomyopathies
	Muscular dystrophy
	5. Epilepsy

(*Continued*)

Table 6. Continued

Component	Assessment and Findings Relevant for VA and/or SCD Risk
Examination	1. Heart rate and regularity, blood pressure
	2. Jugular venous pressure
	3. Murmurs
	4. Pulses and bruits
	5. Edema
	6. Sternotomy scars

ARVC indicates arrhythmogenic right ventricular cardiomyopathy; CPVT catecholaminergic polymorphic ventricular tachycardia; IHD, ischemic heart disease; SCA, sudden cardiac arrest; SCD, sudden cardiac death; SIDS, sudden infant death syndrome; and VA, ventricular arrhythmia.

3.2. Noninvasive Evaluation

3.2.1. 12-lead ECG and Exercise Testing

Recommendations for 12-lead ECG and Exercise Testing
Referenced studies that support the recommendations are summarized in Online Data Supplement 2.

COR	LOE	Recommendations
I	B-NR	1. In patients with sustained, hemodynamically stable, wide complex tachycardia, a 12-lead ECG during tachycardia should be obtained.[S3.2.1-1–S3.2.1-3]
I	B-NR	2. In patients with VA symptoms associated with exertion, suspected ischemic heart disease, or catecholaminergic polymorphic ventricular tachycardia, exercise treadmill testing is useful to assess for exercise-induced VA.[S3.2.1-4,S3.2.1-5]
I	B-NR	3. In patients with suspected or documented VA, a 12-lead ECG should be obtained in sinus rhythm to look for evidence of heart disease.[S3.2.1-6]

3.2.2. Ambulatory Electrocardiography

Recommendation for Ambulatory Electrocardiography
Referenced studies that support the recommendation are summarized in Online Data Supplement 3 and 4.

COR	LOE	Recommendation
I	B-NR	1. Ambulatory electrocardiographic monitoring is useful to evaluate whether symptoms, including palpitations, presyncope, or syncope, are caused by VA.[S3.2.2-1–S3.2.2-4]

3.2.3. Implanted Cardiac Monitors

Recommendation for Implanted Cardiac Monitors
Referenced studies that support the recommendation are summarized in Online Data Supplement 5.

COR	LOE	Recommendation
IIa	B-R	1. In patients with sporadic symptoms (including syncope) suspected to be related to VA, implanted cardiac monitors can be useful.[S3.2.3-1–S3.2.3-4]

3.2.4. Noninvasive Cardiac Imaging

Recommendations for Noninvasive Cardiac Imaging
Referenced studies that support the recommendations are summarized in Online Data Supplement 6.

COR	LOE	Recommendations
I	B-NR	1. In patients with known or suspected VA that may be associated with underlying structural heart disease or a risk of SCA, echocardiography is recommended for evaluation of cardiac structure and function.[S3.2.4-1,S3.2.4-2]
IIa	C-EO	2. In patients presenting with VA who are suspected of having structural heart disease, cardiac magnetic resonance imaging (MRI) or computed tomography (CT) can be useful to detect and characterize underlying structural heart disease.

3.2.5. Biomarkers

Recommendation for Biomarkers
Referenced studies that support the recommendation are summarized in Online Data Supplement 7.

COR	LOE	Recommendation
IIa	B-NR	1. In patients with structural heart disease, measurement of natriuretic peptides (BNP or N-terminal pro-BNP) can be useful by adding prognostic information to standard risk factors for predicting SCD or SCA.[S3.2.5-1–S3.2.5-4]

3.2.6. Genetic Considerations in Arrhythmia Syndromes

Recommendation for Genetic Counselling*

COR	LOE	Recommendation
I	C-EO	1. In patients and family members in whom genetic testing for risk stratification for SCA or SCD is recommended, genetic counseling is beneficial.

*Please refer to section 7.9 in the full guideline for disease-specific recommendations.

3.3. Invasive Testing

3.3.1. Invasive Cardiac Imaging: Cardiac Catheterization or CT Angiography

Recommendation for Invasive Imaging: Cardiac Catheterization

COR	LOE	Recommendation
I	C-EO	1. In patients who have recovered from unexplained SCA, CT or invasive coronary angiography is useful to confirm the presence or absence of ischemic heart disease and guide decisions for myocardial revascularization.

3.3.2. Electrophysiological Study for VA

Recommendations for Electrophysiological Study References that support the recommendations are summarized in Online Data Supplement 8 and 9.		
COR	**LOE**	**Recommendations**
IIa	B-R	1. In patients with ischemic cardiomyopathy, NICM, or adult congenital heart disease who have syncope or other VA symptoms and who do not meet indications for a primary prevention ICD, an electrophysiological study can be useful for assessing the risk of sustained VT.[S3.3.2-1–S3.3.2-7]
III: No Benefit	B-R	2. In patients who meet criteria for ICD implantation, an electrophysiological study for the sole reason of inducing VA is not indicated for risk stratification.[S3.3.2-8–S3.3.2-11]
III: No Benefit	B-NR	3. An electrophysiological study is not recommended for risk stratification for VA in the setting of long QT syndrome, catecholaminergic polymorphic ventricular tachycardia, short QT syndrome, or early repolarization syndromes.[S3.3.2-12–S3.3.2-16]

4. THERAPIES FOR TREATMENT OR PREVENTION OF VA

4.1. Medication Therapy

Table 7. Pharmacological Characteristics of Available Antiarrhythmic Medications for Treating VA

Antiarrhythmic Medication (Class) and Dose	Uses in VA/SCA	Target	Electrophysiological Effects	Pharmacological Characteristics	Common Adverse Effects
Acebutolol PO 200–1200 mg daily or up to 600 mg bid	VT, PVCs	Beta 1, Mild intrinsic sympathomimetic activity	Sinus rate slowed AV nodal refractoriness increased	Active metabolite $t_{1/2}$: 8–13 h pProlonged with renal impairment) Metab: H Excr: F 60%, U 40%	Cardiac: Bradycardia, hypotension, HF, AVB Other: Dizziness, fatigue, anxiety, impotence, hyper/hypoesthesia
Amiodarone (III) IV: 300 mg bolus for VF/pulseless VT arrest; 150-mg bolus for stable VT; 1 mg/min x 6 h, then 0.5 mg/min x 18 h PO: 400 mg* q 8 to 12 h for 1–2 wk, then 300–400 mg daily; reduce dose to 200 mg daily if possible	VT, VF, PVC,	I_{Na}, I_{Ca}, I_{Kr}, I_{K1}, I_{Ks}, I_{to}, Beta receptor, Alpha receptor nuclear T3 receptor	Sinus rate slowed QRS prolonged QTc prolonged AV nodal refractoriness increased; increased DFT	$t_{1/2}$: 26–107 d Metab: H Excr: F	Cardiac: Hypotension, bradycardia, AVB, TdP, slows VT below programmed ICD detection rate, increases defibrillation threshold Other: Corneal microdeposits, thyroid abnormalities, ataxia, nausea, emesis, constipation, photosensitivity, skin discoloration, ataxia, dizziness, peripheral neuropathy, tremor, hepatitis, cirrhosis, pulmonary fibrosis or pneumonitis
Atenolol (II) PO: 25–100 mg qd or bid	VT, PVC, ARVC, LQTS	Beta 1	Sinus rate slowed AV nodal refractoriness increased	$t_{1/2}$: 6–7 h (prolonged with renal impairment) Metab: H Excr: F 50%, U 40%	Cardiac: Bradycardia, hypotension, HF, AVB Other: Dizziness, fatigue, depression, impotence
Bisoprolol (II) PO: 2.5–10 mg once daily	VT, PVC	Beta 1 receptor	Sinus rate slowed AV nodal refractoriness increased	$t_{1/2}$: 9–12 h Metab: H Excr: U	Cardiac: Chest pain, bradycardia, AVB Other: Fatigue, insomnia, diarrhea
Carvedilol (II) PO: 3.125–25 mg q 12 h	VT, PVC	Beta 1 and 2 receptors, Alpha	Sinus rate slowed AV nodal refractoriness increased	$t_{1/2}$: 7–10 h Metab: H Excr: F	Cardiac: Bradycardia, hypotension, AVB, edema, syncope Other: Hyperglycemia, dizziness, fatigue, diarrhea

(*Continued*)

Table 7. Continued

Antiarrhythmic Medication (Class) and Dose	Uses in VA/SCA	Target	Electrophysiological Effects	Pharmacological Characteristics	Common Adverse Effects
Diltiazem (IV) IV: 5–10 mg qd 15–30 min Extended release: PO: 120–360 mg/day	VT specifically RVOT, idiopathic LVT	$I_{Ca\text{-}L}$	Sinus rate slowed PR prolonged AV nodal conduction slowed	$t_{1/2}$: Injection 2–5 h, immediate release 4.5–12 h, extended release 12 h, and severe hepatic impairment 14–16 h Metab: H Excr: U	Cardiac: Hypotension, edema, HF, AVB, bradycardia, exacerbation of HFrEF Other: Headache, rash, constipation
Esmolol (II) IV: 0.5 mg/kg bolus, 0.05 mg/kg/min	VT	Beta 1 receptor	Sinus rate slowed AV nodal refractoriness increased	$t_{1/2}$: 9 min Metab: RBC esterases Excr: U	Cardiac: Bradycardia, hypotension, HF, AVB Other: Dizziness, nausea
Flecainide (IC) PO: 50–200 mg q 12 h	VT, PVC (in the absence of structural heart disease). Has a role in treating patients with CPVT	I_{Na}, I_{Kr}, I_{Kur}	PR prolonged QRS prolonged; increased DFT	$t_{1/2}$: 7–22 h Metab: H Excr: U	Cardiac: Sinus node dysfunction, AVB, drug-induced Brugada syndrome, monomorphic VT in patients with a myocardial scar, exacerbation of HFrEF Other: Dizziness, tremor, vision disturbance, dyspnea, nausea
Lidocaine (IB) IV: 1 mg/kg bolus, 1–3 mg/min 1–1.5 mg/kg. Repeat 0.5–0.75 mg/kg bolus every 5–10 min (max cumulative dose 3 mg/kg). Maintenance infusion is 1–4 mg/min although one could start at 0.5 mg/min	VT, VF	I_{Na}	No marked effect on most intervals; QTc can slightly shorten	Initial $t_{1/2}$ 7–30 min; terminal 90–120 min. Prolonged in HF, liver disease, shock, severe renal disease Metab: H Excr: U	Cardiac: Bradycardia, hemodynamic collapse, AVB, sinus arrest Other: Delirium, psychosis, seizure, nausea, tinnitus, dyspnea, bronchospasm
Metoprolol (II) IV: 5 mg q 5 min up to 3 doses PO: 25–100 mg Extended release qd or q 12 h	VT, PVC	Beta 1 receptor	Sinus rate slowed AV nodal refractoriness increased	$t_{1/2}$: 3–4 h Metab: H Excr: U	Cardiac: Bradycardia, hypotension, AVB Other: Dizziness, fatigue, diarrhea, depression, dyspnea
Mexiletine (IB) PO: 150–300 mg q 8 h or q 12 h	T, VF, PVC, has a role in patients with LQT3	I_{Na}	No marked effect on most intervals; QTc can slightly shorten	$t_{1/2}$: 10–14 h Metab: H Excr: U	Cardiac: HF, AVB Other: Lightheaded, tremor, ataxia, paresthesias, nausea, blood dyscrasias
Nadolol (II) PO: 40–320 mg daily	VT, PVC, LQTS, CPVT	Beta 1 and 2 receptors	Sinus rate slowed AV nodal refractoriness increased	$t_{1/2}$: 20–24 h Metab: none Excr: U	Cardiac: Bradycardia, hypotension, HF, AVB Other: Edema, dizziness, cold extremities, bronchospasm
Procainamide (IA) IV: loading dose 10–17 mg/kg at 20–50 mg/min Maintenance dose: 1–4 mg/min PO (SR preparation): 500–1250 mg q 6 h	VT	I_{Na}, I_{Kr}	QRS prolonged QTc prolonged; increased DFT	Metab: H $t_{1/2}$: 2–5 h; NAPA 6–8 h $t_{1/2}$ prolonged in renal dysfunction. Anephric: proc 11 h and NAPA 42 h Excr: U	Cardiac: TdP; AVB, hypotension and exacerbation of HFrEF Other: Lupus symptoms, diarrhea, nausea, blood dyscrasias
Propafenone (IC) PO: Immediate release 150–300 mg q 8 h Extended release 225–425 mg q 12 h	VT, PVC (in the absence of structural heart disease)	I_{Na}, I_{Kr}, I_{Kur}, Beta receptor, Alpha receptor	PR prolonged QRS prolonged; increased DFT	$t_{1/2}$: 2–10 h or 10–32 h $t_{1/2}$: extensive metabolizers 2–10 h; poor metabolizers 10–32 h. Metab: H Excr: U	Cardiac: HF, AVB, drug-induced Brugada syndrome Other: Dizziness, fatigue, nausea, diarrhea, xerostomia, tremor, blurred vision

(*Continued*)

Table 7. Continued

Antiarrhythmic Medication (Class) and Dose	Uses in VA/SCA	Target	Electrophysiological Effects	Pharmacological Characteristics	Common Adverse Effects
Propranolol (II) IV: 1–3 mg q 5 min to a total of 5 mg PO: Immediate release 10–40 mg q 6 h; Extended release 60–160 mg q 12 h	VT, PVC, LQTS	Beta 1 and 2 receptors, I_{Na}	Sinus rate slowed AV nodal refractoriness increased	$t_{1/2}$: Immediate release 3–6 h Extended release 8–10 h Metab: H Excr: U	Cardiac: Bradycardia, hypotension, HF, AVB Other: Sleep disorder, dizziness, nightmares, hyperglycemia, diarrhea, bronchospasm
Quinidine (IA) PO: sulfate salt 200–600 mg q 6 h to q 12 h Gluconate salt 324–648 mg q 8 h to q 12 h IV: loading dose: 800 mg in 50 mL infused at 50 mg/min	T, VF, (including short QT syndrome, Brugada)	I_{Na}, I_{to}, I_{Kr}, M, Alpha receptor	QRS prolonged QTc prolonged; increased DFT	$t_{1/2}$: 6–8 h longer in HF, liver cirrhosis, and with older age Metab: H Excr: U	Cardiac: Syncope, TdP, AVB Other: Dizziness, diarrhea, nausea, esophagitis, emesis, tinnitus, blurred vision, rash, weakness, tremor; blood dyscrasias
Ranolazine (not classified) PO: 500–1000 mg q 12 h	VT	I_{Na}, I_{Kr}	Sinus rate slowed Tc prolonged	$t_{1/2}$: 7 h Metab: H Excr: U 75%, F 25%	Cardiac: Bradycardia, hypotension Other: Headache, dizziness, syncope, nausea, dyspnea
Sotalol (III) IV: 75 mg q 12 h PO: 80–120 mg q 12 h, may increase dose every 3 d; max 320 mg/d	VT, VF, PVC	I_{Kr}, Beta 1 and 2 receptor	Sinus rate slowed QTc prolonged AV nodal refractoriness increased; decreased DFT	$t_{1/2}$: 12 h Metab: none Excr: U	Cardiac: Bradycardia, hypotension, HF, syncope, TdP Other: Fatigue, dizziness, weakness, dyspnea, bronchitis, depression, nausea, diarrhea
Verapamil (IV) IV: 2.5–5 mg q 15–30 min Sustained release PO: 240–480 mg/d	VT (specifically RVOT, verapamil-sensitive idiopathic LVT)	$I_{Ca\text{-}L}$	Sinus rate slowed PR prolonged AV nodal conduction slowed	$t_{1/2}$: 3–7 h Metab: H Excr: U	Cardiac: Hypotension, edema, HF, AVB, bradycardia, exacerbation of HFrEF Other: Headache, rash, gingival hyperplasia, constipation, dyspepsia

* Although up to 800 mg every 8 h might be used, higher doses of amiodarone are associated with a higher risk of adverse events. Modified from Shleifer JW, et al.[S4.1-1]

Alpha indicates alpha-adrenergic receptor; ARVC, arrhythmogenic right ventricular cardiomyopathy; AV, atrioventricular; AVB, atrioventricular block; Beta, beta-adrenergic receptor; HF, heart failure; CPVT, catecholaminergic polymorphic ventricular tachycardia; DFT, defibrillation threshold; F, feces; H, hepatic; I_{Ca}, L-type calcium channel current; I_{K1}, inward rectifier potassium channel; I_{KACh}, muscarinic receptor-gated potassium channel; I_{KATP}, adenosine-activated potassium channel; I_{Kr}, rapid delayed rectifier potassium current; I_{Ks}, slow delayed rectifier potassium current; I_{Kur}, ultra-rapid delayed rectifier potassium current; I_{Na}, fast inward sodium current; I_{to}, transient outward potassium current; LQTS, long QT syndrome; LVT, left ventricular tachycardia; M, muscarinic; Metab, metabolism; NAPA, n-acetyl procainamide; PVC, premature ventricular complex; QTc, corrected QT interval; $t_{1/2}$, half-life; RVOT, right ventricular outflow tract; T3, triiodothyronine; TdP, torsades de pointes; U, urine; VT, ventricular tachycardia; and VF, ventricular fibrillation.

4.2. Preventing SCD With HF Medications

Recommendation for Pharmacological Prevention of SCD

References that support the recommendation are summarized in Online Data Supplement 10.

COR	LOE	Recommendation
I	A	1. In patients with HF*r*EF (LVEF ≤40%), treatment with a beta blocker, a mineralocorticoid receptor antagonist and either an angiotensin-converting enzyme inhibitor, an angiotensin-receptor blocker, or an angiotensin receptor-neprilysin inhibitor is recommended to reduce SCD and all-cause mortality.[S4.2-1–S4.2-8]

4.3. Surgery and Revascularization Procedures in Patients With Ischemic Heart Disease

Recommendations for Surgery and Revascularization Procedures in Patients With Ischemic Heart Disease

References that support the recommendations are summarized in Online Data Supplement 11.

COR	LOE	Recommendations
I	B-NR	1. Patients with sustained VA and survivors of SCA should be evaluated for ischemic heart disease, and should be revascularized as appropriate.[S4.3-1–S4.3-4]
I	C-EO	2. In patients with anomalous origin of a coronary artery suspected to be the cause of SCA, repair or revascularization is recommended.

4.3.1. Surgery for Arrhythmia Management

Recommendation for Surgery for Arrhythmia Management References that support the recommendation are summarized in Online Data Supplement 12.		
COR	**LOE**	**Recommendation**
IIb	C-LD	1. In patients with monomorphic VT refractory to antiarrhythmic medications and attempts at catheter ablation, surgical ablation may be reasonable.[S4.3.1-1–S4.3.1-7]

4.4. Autonomic Modulation

Recommendations for Autonomic Modulation References that support the recommendations are summarized in Online Data Supplement 13 and 14.		
COR	**LOE**	**Recommendations**
IIa	C-LD	1. In patients with symptomatic, non–life-threatening VA, treatment with a beta blocker is reasonable.[S4.4-1]
IIb	C-LD	2. In patients with VT/VF storm in whom a beta blocker, other antiarrhythmic medications, and catheter ablation are ineffective, not tolerated, or not possible, cardiac sympathetic denervation may be reasonable.[S4.4-2–S4.4-4]

5. ACUTE MANAGEMENT OF SPECIFIC VA

Recommendations for Management of Cardiac Arrest References that support the recommendations are summarized in Online Data Supplement 15 and 16.		
COR	**LOE**	**Recommendations**
I	A	1. CPR should be performed in patients in cardiac arrest according to published basic and advanced cardiovascular life support algorithms.[S5-1–S5-3]
I	A	2. In patients with hemodynamically unstable VA that persist or recur after a maximal energy shock, intravenous amiodarone should be administered to attempt to achieve a stable rhythm after further defibrillation.[S5-1,S5-4–S5-6]
I	A	3. Patients presenting with VA with hemodynamic instability should undergo direct current cardioversion.[S5-1–S5-3]
I	B-NR	4. In patients with polymorphic VT or VF with ST-elevation MI, angiography with emergency revascularization is recommended.[S5-7–S5-10]
I	C-EO	5. Patients with a wide-QRS tachycardia should be presumed to have VT if the diagnosis is unclear.
IIa	A	6. In patients with hemodynamically stable VT, administration of intravenous procainamide can be useful to attempt to terminate VT.[S5-11–S5-13]
IIa	B-R	7. In patients with a witnessed cardiac arrest due to VF or polymorphic VT that is unresponsive to CPR, defibrillation, and vasopressor therapy, intravenous lidocaine can be beneficial.[S5-1,S5-4,S5-5,S5-14,S5-15]
IIa	B-R	8. In patients with polymorphic VT due to myocardial ischemia, intravenous beta blockers can be useful.[S5-16,S5-17]
IIa	B-NR	9. In patients with a recent MI who have VT/VF that repeatedly recurs despite direct current cardioversion and antiarrhythmic medications (VT/VF storm), an intravenous beta blocker can be useful.[S5-17,S5-18]
IIb	A	10. In patients in cardiac arrest, administration of epinephrine (1 mg every 3 to 5 minutes) during CPR may be reasonable.[S5-1,S5-19–S5-24]
IIb	B-R	11. In patients with hemodynamically stable VT, administration of intravenous amiodarone or sotalol may be considered to attempt to terminate VT.[S5-5,S5-13,S5-25,S5-26]
III: No Benefit	A	12. In patients with cardiac arrest, administration of high-dose epinephrine (>1 mg boluses) compared with standard doses is not beneficial.[S5-19,S5-21]
III: No Benefit	A	13. In patients with refractory VF not related to torsades de pointes, administration of intravenous magnesium is not beneficial.[S5-27,S5-28]
III: Harm	B-R	14. In patients with suspected AMI, prophylactic administration of lidocaine or high-dose amiodarone for the prevention of VT is potentially harmful.[S5-16,S5-29]
III: Harm	C-LD	15. In patients with a wide QRS complex tachycardia of unknown origin, calcium channel blockers (eg, verapamil and diltiazem) are potentially harmful.[S5-30,S5-31]

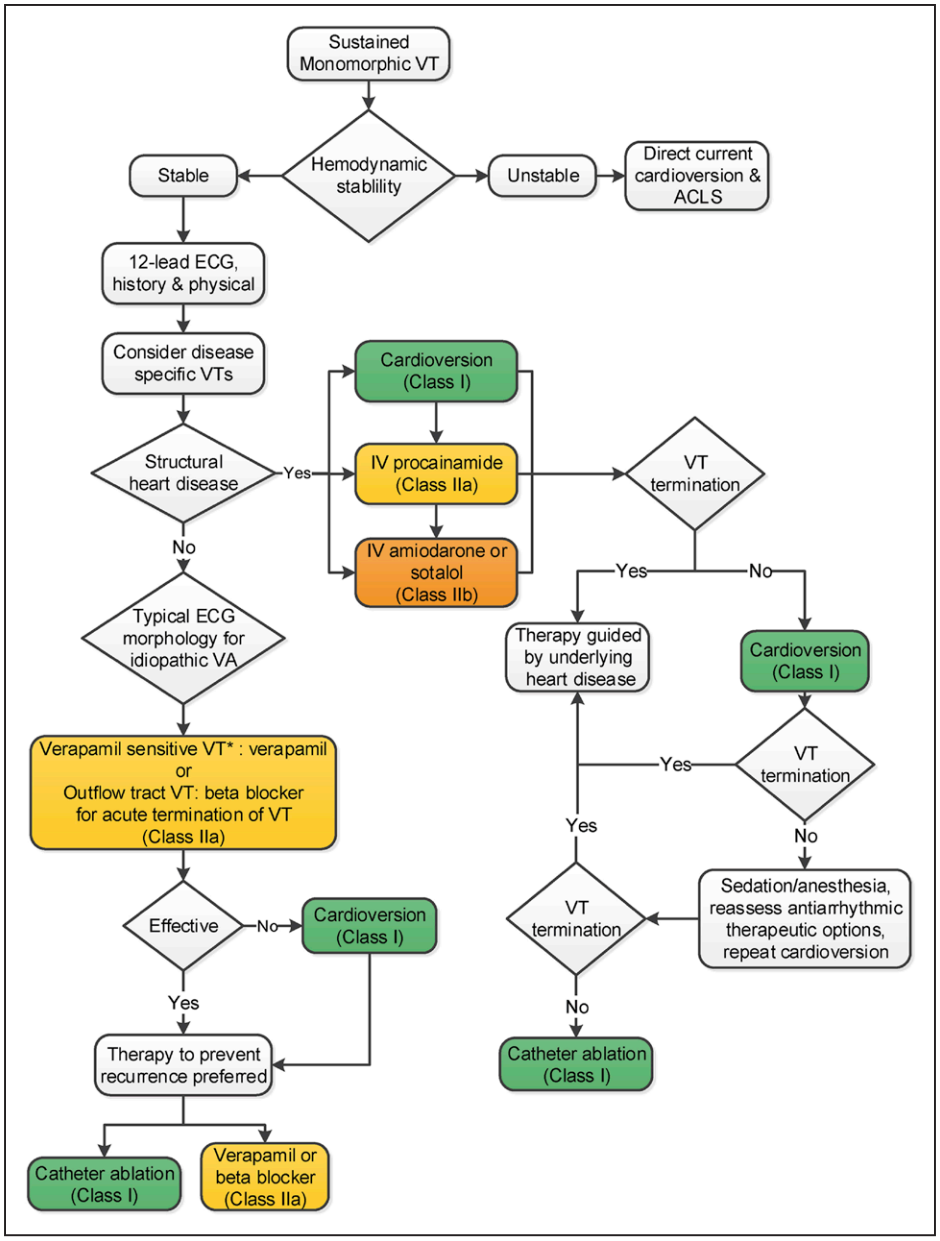


Figure 2. Management of sustained monomorphic VT.
Colors correspond to Class of Recommendation in Table 1. See Sections 7, 8.1.3, 8.2.3, and 10 in the full-text guideline for discussion. *Known history of verapamil sensitive or classical electrocardiographic presentation. ACLS indicates advanced cardiovascular life support; ECG, electrocardiogram; VA, ventricular arrhythmia; and VT, ventricular tachycardia.

6. ONGOING MANAGEMENT OF VA AND SCD RISK RELATED TO SPECIFIC DISEASE STATES

6.1. Ischemic Heart Disease

6.1.1. Secondary Prevention of SCD in Patients With Ischemic Heart Disease

Recommendations for Secondary Prevention of SCD in Patients With Ischemic Heart Disease

References that support the recommendations are summarized in Online Data Supplement 17 and 18.

COR	LOE	Recommendations
I	B-R	1. In patients with ischemic heart disease, who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R)[S6.1.1-1–S6.1.1-4] or stable sustained VT (LOE: B-NR)[S6.1.1-5] not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.
	B-NR	
Value Statement: Intermediate Value (LOE: B-R)		2. A transvenous ICD provides intermediate value in the secondary prevention of SCD particularly when the patient's risk of death due to a VA is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient's burden of comorbidities and functional status.[S6.1.1-6]
I	B-NR	3. In patients with ischemic heart disease and unexplained syncope who have inducible sustained monomorphic VT on electrophysiological study, an ICD is recommended if meaningful survival of greater than 1 year is expected.[S6.1.1-7]

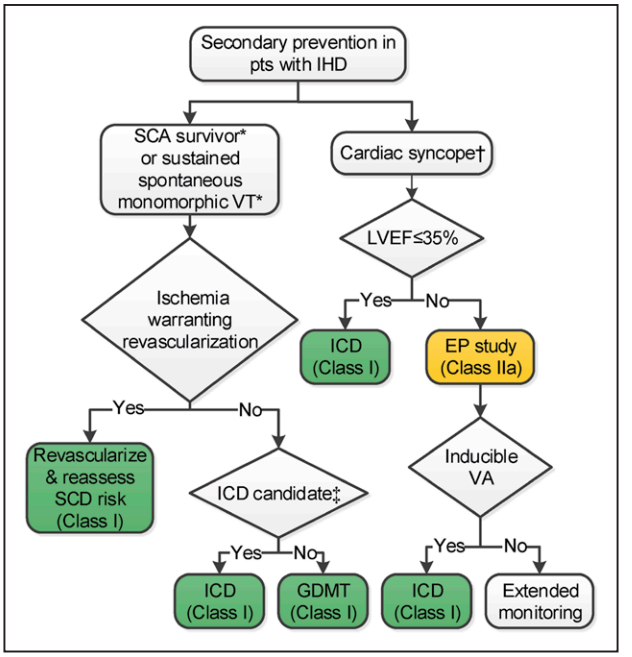


Figure 3. Secondary prevention patients with ischemic heart disease. Colors correspond to Class of Recommendation in Table 1. See Sections 4.3.1 and 7.1.1 in the full-text guideline for discussion. *Exclude reversible causes. †History consistent with an arrhythmic etiology for syncope. ‡ICD candidacy as determined by functional status, life expectancy, or patient preference. EP indicates electrophysiological; GDMT, guideline-directed management and therapy; ICD, implantable cardioverter-defibrillator; IHD, ischemic heart disease; LVEF, left ventricular ejection fraction; pts, patients; SCA, sudden cardiac arrest; SCD, sudden cardiac death; and VT, ventricular tachycardia.

6.1.1.1. Coronary Artery Spasm

Recommendations for Patients With Coronary Artery Spasm

References that support the recommendations are summarized in Online Data Supplement 20.

COR	LOE	Recommendations
I	B-NR	1. In patients with VA due to coronary artery spasm, treatment with maximally tolerated doses of a calcium channel blocker and smoking cessation are indicated to reduce recurrent ischemia and VA.[S6.1.1.1-1,S6.1.1.1-2]
IIa	B-NR	2. In patients resuscitated from SCA due to coronary artery spasm in whom medical therapy is ineffective or not tolerated, an ICD is reasonable if meaningful survival of greater than 1 year is expected.[S6.1.1.1-3–S6.1.1.1-6]
IIb	B-NR	3. In patients resuscitated from SCA due to coronary artery spasm, an ICD in addition to medical therapy may be reasonable if meaningful survival of greater than 1 year is expected.[S6.1.1.1-3–S6.1.1.1-6]

6.1.2. Primary Prevention of SCD in Patients With Ischemic Heart Disease

Recommendations for Primary Prevention of SCD in Patients With Ischemic Heart Disease References that support the recommendations are summarized in Online Data Supplement 21.		
COR	**LOE**	**Recommendations**
I	A	1. In patients with LVEF of 35% or less that is due to ischemic heart disease who are at least 40 days' post-MI and at least 90 days postrevascularization, and with NYHA class II or III HF despite GDMT, an ICD is recommended if meaningful survival of greater than 1 year is expected.[S6.1.2-1,S6.1.2-2]
I	A	2. In patients with LVEF of 30% or less that is due to ischemic heart disease who are at least 40 days' post-MI and at least 90 days postrevascularization, and with NYHA class I HF despite GDMT, an ICD is recommended if meaningful survival of greater than 1 year is expected.[S6.1.2-2,S6.1.2-3]
Value Statement: High Value (LOE: B-R)		3. A transvenous ICD provides high value in the primary prevention of SCD particularly when the patient's risk of death due to a VA is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient's burden of comorbidities and functional status.[S6.1.2-4]

Recommendations for Primary Prevention of SCD in Patients With Ischemic Heart Disease (Continued)		
COR	**LOE**	**Recommendations**
I	B-R	4. In patients with NSVT due to prior MI, LVEF of 40% or less and inducible sustained VT or VF at electrophysiological study, an ICD is recommended if meaningful survival of greater than 1 year is expected.[S6.1.2-5]
IIa	B-NR	5. In nonhospitalized patients with NYHA class IV symptoms who are candidates for cardiac transplantation or an LVAD, an ICD is reasonable if meaningful survival of greater than 1 year is expected.[S6.1.2-6–S6.1.2-9]
III: No Benefit	C-EO	6. An ICD is not indicated for NYHA class IV patients with medication-refractory HF who are not also candidates for cardiac transplantation, an LVAD, or a CRT defibrillator that incorporates both pacing and defibrillation capabilities.

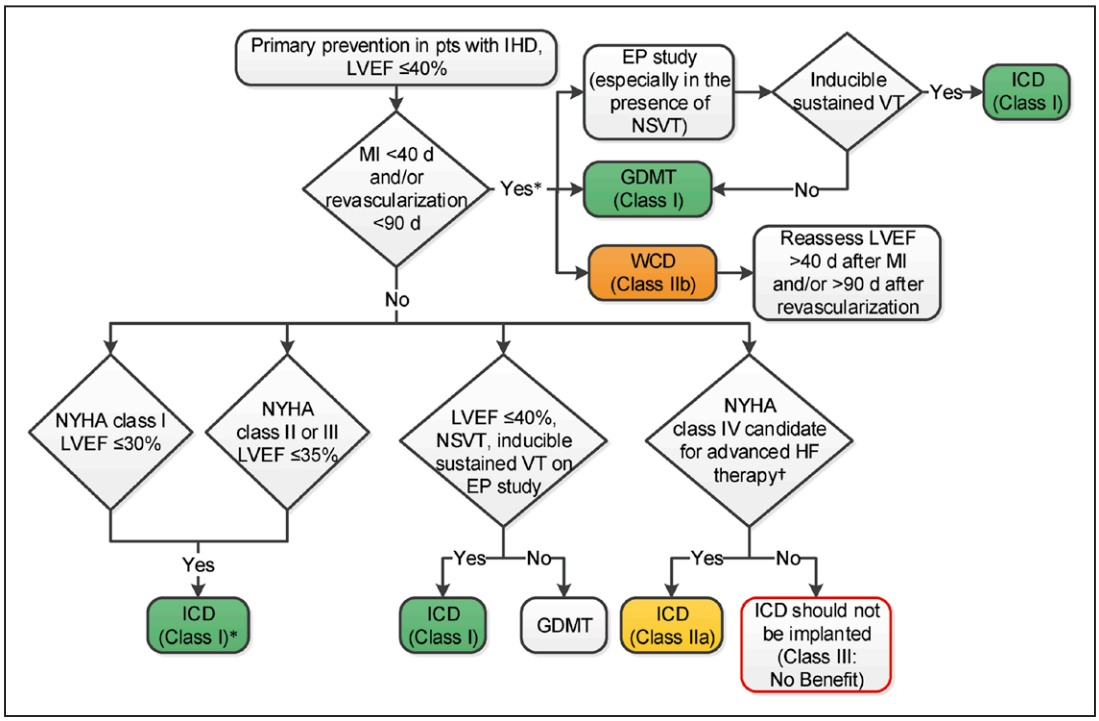


Figure 4. Primary prevention of SCD in patients with ischemic heart disease.
Colors correspond to Class of Recommendation in Table 1. See Section 7.1.2 in the full-text guideline for discussion. *Scenarios exist for early ICD placement in select circumstances such as patients with a pacing indication or syncope. †Advanced HF therapy includes CRT, cardiac transplant, and LVAD. thought due to VT. These are detailed elsewhere in an HRS/ACC/AHA expert consensus statement (24). CRT indicates cardiac resynchronization therapy; EP, electrophysiological; GDMT, guideline-directed management and therapy; HF, heart failure; ICD, implantable cardioverter-defibrillator; IHD, ischemic heart disease; LVEF, left ventricular ejection fraction; MI, myocardial infarction; NSVT, nonsustained ventricular tachycardia; NYHA, New York Heart Association; pts, patients; SCD, sudden cardiac death; VT, ventricular tachycardia; and WCD, wearable cardioverter-defibrillator.

6.1.3. Treatment and Prevention of Recurrent VA in Patients With Ischemic Heart Disease

Recommendations for Treatment of Recurrent VA in Patients With Ischemic Heart Disease

References that support the recommendations are summarized in Online Data Supplement 22 and 23.

COR	LOE	Recommendations
I	B-R	1. In patients with ischemic heart disease and recurrent VA, with significant symptoms or ICD shocks despite optimal device programming and ongoing treatment with a beta blocker, amiodarone or sotalol is useful to suppress recurrent VA.[S6.1.3-1–S6.1.3-3]
I	B-R	2. In patients with prior MI and recurrent episodes of symptomatic sustained VT, or who present with VT storm and have failed or are intolerant of amiodarone (LOE: B-R)[S6.1.3-4] or other antiarrhythmic medications (LOE: B-NR),[S6.1.3-5–S6.1.3-9] catheter ablation is recommended.[S6.1.3-10–S6.1.3-12]
	B-NR	

Recommendations for Treatment of Recurrent VA in Patients With Ischemic Heart Disease (Continued)

COR	LOE	Recommendations
IIb	C-LD	3. In patients with ischemic heart disease and ICD shocks for sustained monomorphic VT or symptomatic sustained monomorphic VT that is recurrent, or hemodynamically tolerated, catheter ablation as first-line therapy may be considered to reduce recurrent VA.[S6.1.3-10,S6.1.3-11]
III: Harm	B-R	4. In patients with prior MI, class IC antiarrhythmic medications (eg, flecainide and propafenone) should not be used.[S6.1.3-13]
III: Harm	C-LD	5. In patients with incessant VT or VF, an ICD should not be implanted until sufficient control of the VA is achieved to prevent repeated ICD shocks.[S6.1.3-14]
III: No Benefit	C-LD	6. In patients with ischemic heart disease and sustained monomorphic VT, coronary revascularization alone is an ineffective therapy to prevent recurrent VT.[S6.1.3-15,S6.1.3-16]

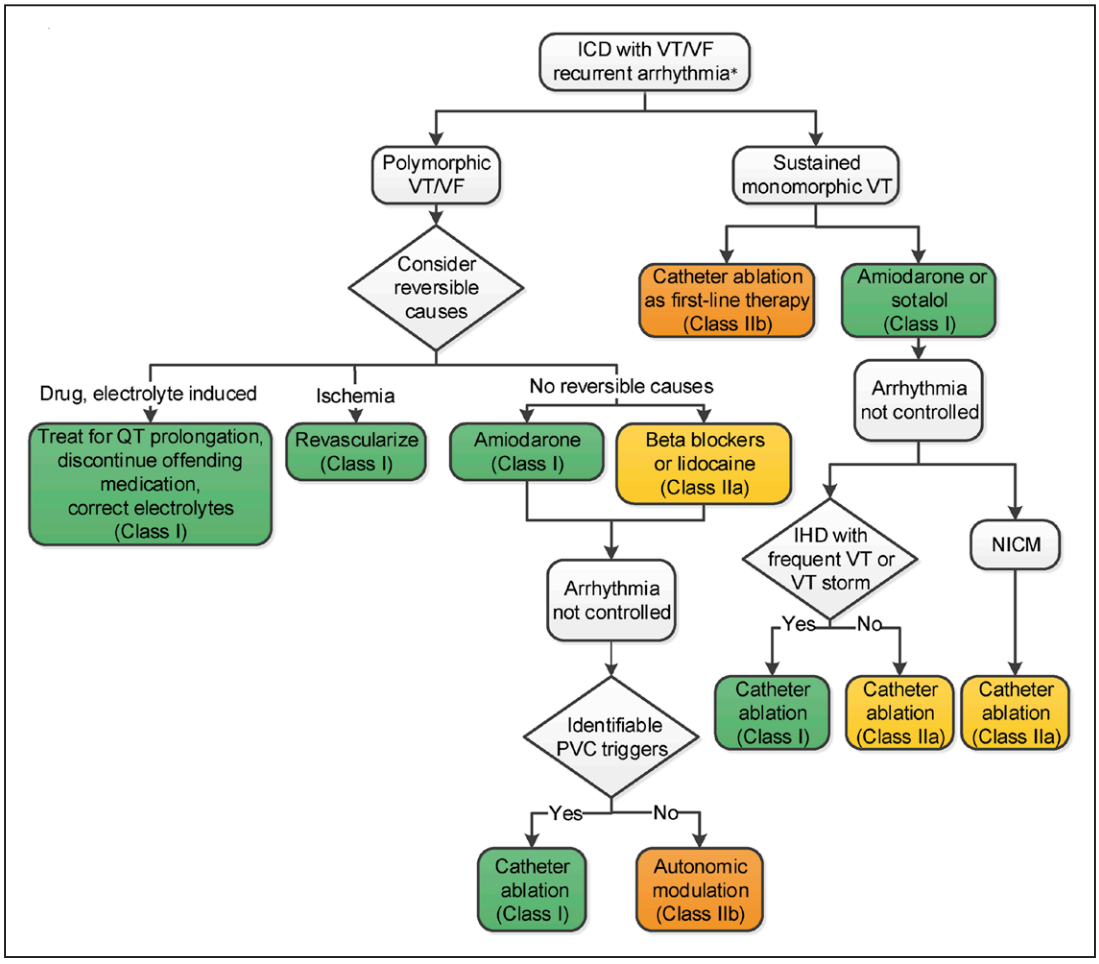


Figure 5. Treatment of recurrent VA in patients with ischemic heart disease or NICM.
Colors correspond to Class of Recommendation in Table 1. See Sections 5.6, 6, 7.1.3, and 7.2 in the full-text guideline for discussion. *Management should start with ensuring that the ICD is programmed appropriately and that potential precipitating causes, including heart failure exacerbation, are addressed. For information regarding optimal ICD programming, refer to the 2015 HRS/EHRA/APHRS/SOLAECE expert consensus statement.[S1.4-8] EHRA indicated European Heart Rhythm Association; HRS, Heart Rhythm Society; IHD, ischemic heart disease; ICD, implantable cardioverter-defibrillator; PVC, premature ventricular complex; NICM, nonischemic cardiomyopathy; SOLAECE, Sociedad Latinoamericana de Estimulación Cardíaca y Electrofisiología; VF, ventricular fibrillation; and VT, ventricular tachycardia.

6.2. Nonischemic Cardiomyopathy

Recommendations for Patients With NICM References that support the recommendations are summarized in Online Data Supplement 24.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with suspected NICM from myocardial infiltrative processess, cardiac MRI with late gadolinium enhancement is useful for diagnosis.[S6.2-1–S6.2-3]
IIa	B-NR	2. In patients with suspected NICM, cardiac MRI with late gadolinium enhancement can be useful for assessing risk of SCA/SCD.[S6.2-1–S6.2-3]
IIa	C-EO	3. In patients with NICM who develop conduction disease or LV dysfunction at less than 40 years of age, or who have a family history of NICM or SCD in a first-degree relative (<50 years of age), genetic counseling and genetic testing are reasonable to detect a heritable disease that may clarify prognosis and facilitate cascade screening of relatives.[S6.2-4,S6.2-5]

6.2.1. Secondary Prevention of SCD in Patients With NICM

Recommendations for Secondary Prevention of SCD in Patients With NICM References that support the recommendations are summarized in Online Data Supplement 25 and 26.		
COR	**LOE**	**Recommendations**
I	B-R B-NR	1. In patients with NICM who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R)[S6.2.1-1–S6.2.1-4] or stable sustained VT (LOE: B-NR)[S6.2.1-5] not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.
IIa	B-NR	2. In patients with NICM who experience syncope presumed to be due to VA and who do not meet indications for a primary prevention ICD, an ICD or an electrophysiological study for risk stratification for SCD can be beneficial if meaningful survival greater than 1 year is expected.[S6.2.1-6–S6.2.1-11]
IIb	B-R	3. In patients with NICM who survive a cardiac arrest, have sustained VT, or have symptomatic VA who are ineligible for an ICD (due to a limited life-expectancy and/or functional status or lack of access to an ICD), amiodarone may be considered for prevention of SCD.[S6.2.1-12,S6.2.1-13]

6.2.2. Primary Prevention of SCD in Patients With NICM

Recommendations for Primary Prevention of SCD in Patients With NICM References that support the recommendations are summarized in Online Data Supplement 27 and 28.		
COR	**LOE**	**Recommendations**
I	A	1. In patients with NICM, HF with NYHA class II–III symptoms and an LVEF of 35% or less, despite GDMT, an ICD is recommended if meaningful survival of greater than 1 year is expected.[S6.2.2-1–S6.2.2-6]
IIa	B-NR	2. In patients with NICM due to a *Lamin A/C* mutation who have 2 or more risk factors (NSVT, LVEF <45%, nonmissense mutation, and male sex), an ICD can be beneficial if meaningful survival of greater than 1 year is expected.[S6.2.2-7–S6.2.2-10]
IIb	B-R	3. In patients with NICM, HF with NYHA class I symptoms and an LVEF of 35% or less, despite GDMT, an ICD may be considered if meaningful survival of greater than 1 year is expected.[S6.2.2-5]
III: No Benefit	C-EO	4. In patients with medication-refractory NYHA class IV HF who are not also candidates for cardiac transplantation, an LVAD, or a CRT defibrillator that incorporates both pacing and defibrillation capabilities, an ICD should not be implanted.

6.2.3. Treatment of Recurrent VA in Patients With NICM

Recommendations for Treatment of Recurrent VA in Patients With NICM References that support the recommendations are summarized in Online Data Supplement 29.		
COR	**LOE**	**Recommendations**
IIa	B-R	1. In patients with NICM and an ICD who experience spontaneous VA or recurrent appropriate shocks despite optimal device programming and treatment with a beta blocker, amiodarone or sotalol can be beneficial[S6.2.3-1].
IIa	B-NR	2. In patients with NICM and recurrent sustained monomorphic VT who fail or are intolerant of antiarrhythmic medications, catheter ablation can be useful for reducing recurrent VT and ICD shocks.[S6.2.3-2,S6.2.3-3]

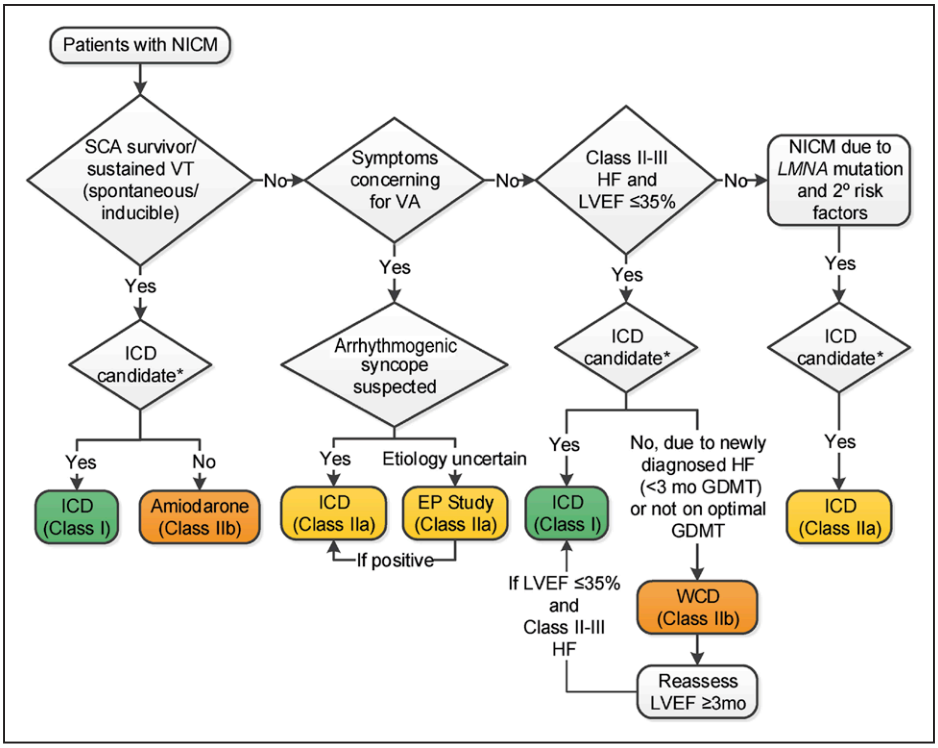


Figure 6. Secondary and primary prevention of SCD in patients with NICM.
Colors correspond to Class of Recommendation in Table 1. See Section 7.2 in the full-text guideline for discussion. *ICD candidacy as determined by functional status, life expectancy or patient preference. 2° indicates secondary; EP, electrophysiological; GDMT, guideline-directed management and therapy; HF, heart failure; ICD, implantable cardioverter-defibrillator; LVEF, left ventricular ejection fraction; NICM, nonischemic cardiomyopathy; SCA, sudden cardiac arrest; SCD, sudden cardiac death; VA, ventricular arrhythmia; and WCD, wearable cardioverter-defibrillator.

6.3. Arrhythmogenic Right Ventricular Cardiomyopathy

Recommendations for Arrhythmogenic Right Ventricular Cardiomyopathy

References that support the recommendations are summarized in Online Data Supplement 30.

COR	LOE	Recommendations
I	B-NR	1. In selected first-degree relatives of patients with arrhythmogenic right ventricular cardiomyopathy, clinical screening for the disease is recommended along with genetic counseling and genetic testing, if the proband has a disease causing mutation.[S6.3-1–S6.3-4]
I	B-NR	2. In patients with suspected arrhythmogenic right ventricular cardiomyopathy and VA or electrocardiographic abnormalities, cardiac MRI is useful for establishing a diagnosis and for risk stratification.[S6.3-5–S6.3-8]
I	B-NR	3. In patients with arrhythmogenic right ventricular cardiomyopathy and an additional marker of increased risk of SCD (resuscitated SCA, sustained VT, significant ventricular dysfunction with RVEF or LVEF ≤35%), an ICD is recommended if meaningful survival greater than 1 year is expected.[S6.3-9–S6.3-13]

Recommendations for Arrhythmogenic Right Ventricular Cardiomyopathy (Continued)

COR	LOE	Recommendations
I	B-NR	4. In patients with arrhythmogenic right ventricular cardiomyopathy and VA, a beta blocker is recommended.[S6.3-11,S6.3-14,S6.3-15]
I	B-NR	5. In patients with a clinical diagnosis of arrhythmogenic right ventricular cardiomyopathy, avoiding intensive exercise is recommended.[S6.3-11,S6.3-12,S6.3-16–S6.3-21]
IIa	B-NR	6. In patients with clinically diagnosed or suspected arrhythmogenic right ventricular cardiomyopathy, genetic counseling and genetic testing can be useful for diagnosis and for gene-specific targeted family screening.[S6.3-1,S6.3-4,S6.3-22–S6.3-26]
IIa	B-NR	7. In patients with arrhythmogenic right ventricular cardiomyopathy and syncope presumed due to VA, an ICD can be useful if meaningful survival greater than 1 year is expected.[S6.3-10,S6.3-11,S6.3-13]
IIa	B-NR	8. In patients with clinical evidence of arrhythmogenic right ventricular cardiomyopathy but not VA, a beta blocker can be useful.[S6.3-14,S6.3-15]

Recommendations for Arrhythmogenic Right Ventricular Cardiomyopathy (Continued)		
COR	**LOE**	**Recommendations**
IIa	B-NR	9. In patients with arrhythmogenic right ventricular cardiomyopathy and recurrent symptomatic sustained VT in whom a beta blocker is ineffective or not tolerated, catheter ablation with availability of a combined endocardial/epicardial approach can be beneficial.[S6.3-27–S6.3-33]
IIa	B-NR	10. In patients with suspected arrhythmogenic right ventricular cardiomyopathy, a signal averaged ECG can be useful for diagnosis and risk stratification.[S6.3-14,S6.3-34,S6.3-35]
IIb	B-NR	11. In asymptomatic patients with clinical evidence of arrhythmogenic right ventricular cardiomyopathy, an electrophysiological study may be considered for risk stratification.[S6.3-9,S6.3-36]

6.4. Hypertrophic Cardiomyopathy

Recommendations for HCM References that support the recommendations are summarized in Online Data Supplement 31.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with HCM, SCD risk stratification should be performed at the time of initial evaluation and periodically thereafter.[S6.4-1–S6.4-8]
I	B-NR	2. In patients with HCM who have survived an SCA due to VT or VF, or have spontaneous sustained VT causing syncope or hemodynamic compromise, an ICD is recommended if meaningful survival greater than 1 year is expected.[S6.4-1,S6.4-6,S6.4-9,S6.4-10]
I	B-NR	3. In first-degree relatives of patients with HCM, an ECG and echocardiogram should be performed.[S6.4-11–S6.4-17]
I	B-NR	4. In first-degree relatives of patients with HCM due to a known causative mutation, genetic counseling and mutation-specific genetic testing are recommended.[S6.4-13–S6.4-15,S6.4-18,S6.4-19]
IIa	B-NR	5. In patients with clinically suspected or diagnosed HCM, genetic counseling and genetic testing are reasonable.[S6.4-13–S6.4-15,S6.4-18–S6.4-22]
IIa	B-NR	6. In patients with HCM and 1 or more of the following risk factors, an ICD is reasonable if meaningful survival of greater than 1 year is expected: a. Maximum LV wall thickness ≥30 mm (LOE: B-NR).[S6.4-2,S6.4-3,S6.4-23,S6.4-24]
IIa	C-LD	b. SCD in 1 or more first-degree relatives presumably caused by HCM (LOE: C-LD).[S6.4-25,S6.4-26]
IIa	C-LD	c. 1 or more episodes of unexplained syncope within the preceding 6 months (LOE: C-LD).[S6.4-8,S6.4-26]
IIa	B-NR	7. In patients with HCM who have spontaneous NSVT (LOE: C-LD)[S6.4-2,S6.4-26,S6.4-27] or an abnormal blood pressure response with exercise (LOE: B-NR),[S6.4-5,S6.4-28,S6.4-29] who also have additional SCD risk modifiers or high-risk features, an ICD is reasonable if meaningful survival greater than 1 year is expected.
IIa	C-LD	
IIb	B-NR	8. In patients with HCM who have NSVT (LOE: B-NR)[S6.4-2,S6.4-26,S6.4-27] or an abnormal blood pressure response with exercise (LOE: B-NR)[S6.4-5,S6.4-28,S6.4-29] but do not have any other SCD risk modifiers, an ICD may be considered, but its benefit is uncertain.
IIb	B-NR	
IIb	C-LD	9. In patients with HCM and a history of sustained VT or VF, amiodarone may be considered when an ICD is not feasible or not preferred by the patient.[S6.4-30,S6.4-31]
III: No Benefit	B-NR	10. In patients with HCM, an invasive electrophysiological study with programmed ventricular stimulation should not be performed for risk stratification.[S6.4-32,S6.4-33]
III: No Benefit	B-NR	11. In patients with an identified HCM genotype in the absence of SCD risk factors, an ICD should not be implanted.[S6.4-7,S6.4-34,S6.4-35]

Refer to the ACCF/AHA HCM guideline for the definition of HCM.[S6.4-36]

Table 8. Major Clinical Features Associated With Increased Risk of SCD in Patients With HCM

Established risk factors*
Survival from a cardiac arrest due to VT or VF[S6.4-1,S6.4-5,S6.4-6]
Spontaneous sustained VT causing syncope or hemodynamic compromise[S6.4-1,S6.4-5,S6.4-6]
Family history of SCD associated with HCM[S6.4-25,S6.4-26]
LV wall thickness ≥30 mm[S6.4-2,S6.4-3,S6.4-23,S6.4-24]
Unexplained syncope within 6 mo[S6.4-8,S6.4-26]
NSVT ≥3 beats[S6.4-2,S6.4-26,S6.4-27]
Abnormal blood pressure response during exercise†[S6.4-5,S6.4-28,S6.4-29]
Potential risk modifiers‡
<30 y[S6.4-5,S6.4-26]
Delayed hyperenhancement on cardiac MRI[S6.4-37–S6.4-40]
LVOT obstruction[S6.4-2,S6.4-4]
Syncope >5 y ago[S6.4-8,S6.4-26]
High-risk subsets§
LV aneurysm[S6.4-41–S6.4-43]
LVEF <50%[S6.4-44]

*There is general agreement in the literature that these factors independently convey an increased risk for SCD in patients with HCM.

†Decrease in blood pressure of 20 mm Hg or failure to increase systolic blood pressure >20 mm Hg during exertion.

‡There is a lack of agreement in the literature that these modifiers independently convey an increased risk of SCD in patients with HCM; however, a risk modifier when combined with a risk factor often identifies a patient with HCM at increased risk for SCD beyond the risk conveyed by the risk factor alone.

§A small subset of patients with an LVEF <50% (end-stage disease) or an LV aneurysm warrant consideration for ICD implantation.[S6.4-44]

HCM indicates hypertrophic cardiomyopathy; ICD, implantable cardioverter-defibrillator; LV, left ventricular; LVEF, left ventricular ejection fraction; LVOT, left ventricular outflow tract; NSVT, nonsustained ventricular tachycardia; SCD, sudden cardiac death; VT, ventricular tachycardia; and VF, ventricular fibrillation.

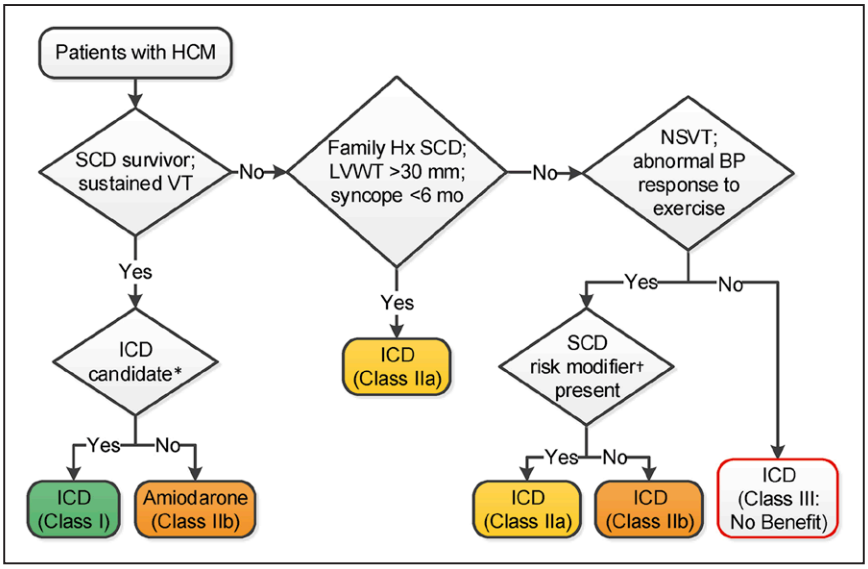


Figure 7. Prevention of SCD in patients with HCM.
Colors correspond to Class of Recommendation in Table 1. See Section 7.4 in the full-text guideline for discussion. *ICD candidacy as determined by functional status, life expectancy, or patient preference. †Risk modifiers: Age <30 y, late gadolinium enhancement on cardiac MRI, LVOT obstruction, LV aneurysm, syncope >5 y. BP indicates blood pressure; HCM, hypertrophic cardiomyopathy; Hx, history; ICD, implantable cardioverter-defibrillator; LVOT, left ventricular outflow tract; LVWT, left ventricular wall thickness; MRI, magnetic resonance imaging; NSVT, nonsustained ventricular tachycardia; SCD, sudden cardiac death; and VT, ventricular tachycardia.

6.5. Myocarditis

Recommendations for Myocarditis References that support the recommendations are summarized in Online Data Supplement 32.		
COR	**LOE**	**Recommendations**
I	C-LD	1. In patients with life-threatening VT or VF associated with confirmed or clinically suspected myocarditis, referral to centers with mechanical hemodynamic support and advanced arrhythmia management is recommended.[S6.5-1]
IIb	C-LD	2. In patients with giant cell myocarditis with VF or hemodynamically unstable VT treated according to GDMT, an ICD and/or an antiarrhythmic medication may be considered if meaningful survival of greater than 1 year is expected.[S6.5-2–S6.5-4]

6.6. Cardiac Sarcoidosis

Recommendations for Cardiac Sarcoidosis References that support the recommendations are summarized in Online Data Supplement 33.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with cardiac sarcoidosis who have sustained VT or are survivors of SCA or have an LVEF of 35% or less, an ICD is recommended, if meaningful survival of greater than 1 year is expected.[S6.6-1–S6.6-5]

Recommendations for Cardiac Sarcoidosis (Continued)		
COR	**LOE**	**Recommendations**
IIa	B-NR	2. In patients with cardiac sarcoidosis and LVEF greater than 35% who have syncope and/or evidence of myocardial scar by cardiac MRI or positron emission tomographic (PET) scan, and/or have an indication for permanent pacing, implantation of an ICD is reasonable, provided that meaningful survival of greater than 1 year is expected.[S6.6-6–S6.6-10]
IIa	C-LD	3. In patients with cardiac sarcoidosis and LVEF greater than 35%, it is reasonable to perform an electrophysiological study and to implant an ICD, if sustained VA is inducible, provided that meaningful survival of greater than 1 year is expected.[S6.6-11,S6.6-12]
IIa	C-LD	4. In patients with cardiac sarcoidosis who have an indication for permanent pacing, implantation of an ICD can be beneficial.[S6.6-13]
IIa	C-LD	5. In patients with cardiac sarcoidosis with frequent symptomatic VA and evidence of myocardial inflammation, immunosuppression in combination with antiarrhythmic medication therapy can be useful to reduce VA burden.[S6.6-14–S6.6-16]

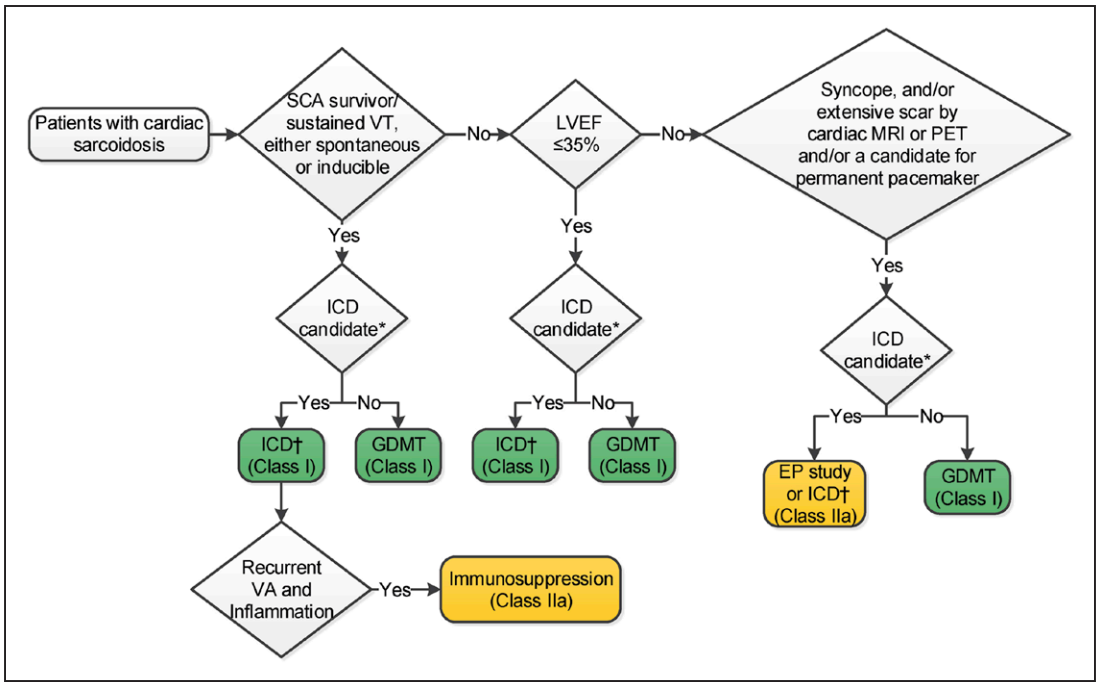


Figure 8. Prevention of SCD in patients with cardiac sarcoidosis.
Colors correspond to Class of Recommendation in Table 1. See Section 7.6 in the full-text guideline for discussion. *ICD candidacy as determined by functional status, life expectancy, or patient preference. †For recurrent sustained monomorphic VT, refer to Figure 2. CEP indicates electrophysiological; GDMT, guideline-directed management and therapy; ICD, implantable cardioverter-defibrillator; LVEF, left ventricular ejection fraction; MRI, magnetic resonance imaging; PET, positron emission tomography; SCA, sudden cardiac arrest; SCD, sudden cardiac death; VA, ventricular arrhythmia; and VT, ventricular tachycardia.

6.7. Heart Failure

6.7.1. HF With Reduced Ejection Fraction

Recommendation for HF*r*EF References that support the recommendation are summarized in Online Data Supplement 35.		
COR	**LOE**	**Recommendation**
IIa	B-NR	1. In patients with HF*r*EF who are awaiting heart transplant and who otherwise would not qualify for an ICD (eg, NYHA class IV and/or use of inotropes) with a plan to discharge home, an ICD is reasonable.[S6.7.1-1–S6.7.1-5]

6.7.2. Left Ventricular Assist Device

Recommendation for Patients With an LVAD References that support the recommendation are summarized in Online Data Supplement 36.		
COR	**LOE**	**Recommendation**
IIa	C-LD	1. In patients with an LVAD and sustained VA, an ICD can be beneficial.[S6.7.2-1]

6.7.3. ICD Use After Heart Transplantation

Recommendation for ICD Use After Heart Transplantation References that support the recommendation are summarized in Online Data Supplement 37.		
COR	**LOE**	**Recommendation**
IIb	B-NR	1. In patients with a heart transplant and severe allograft vasculopathy with LV dysfunction, an ICD may be reasonable if meaningful survival of greater than 1 year is expected.[S6.7.3-1–S6.7.3-3]

6.8. Neuromuscular Disorders

Recommendations for Neuromuscular Disorders References that support the recommendations are summarized in Online Data Supplement 38.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with neuromuscular disorders, primary and secondary prevention ICDs are recommended for the same indications as for patients with NICM if meaningful survival of greater than 1 year is expected.[S6.8-1,S6.8-2]
IIa	B-NR	2. In patients with Emery-Dreifuss and limb-girdle type IB muscular dystrophies with progressive cardiac involvement, an ICD is reasonable if meaningful survival of greater than 1 year is expected.[S6.8-3–S6.8-8]
IIa	B-NR	3. In patients with muscular dystrophy, follow-up for development of cardiac involvement is reasonable, even if the patient is asymptomatic at presentation.[S6.8-9–S6.8-12]
IIb	B-NR	4. In patients with myotonic dystrophy type 1 with an indication for a permanent pacemaker, an ICD may be considered to minimize the risk of SCA from VT if meaningful survival of greater than 1 year is expected.[S6.8-9,S6.8-13,S6.8-14]

Table 9. Neuromuscular Disorders Associated With Heart Disease

Muscular Dystrophy	Inheritance	Gene/Protein Affected	Primary Cardiac Pathology	Frequency of Cardiac Involvement	Causes of Death	Associated With Sudden Death?
Duchenne	X-linked recessive	Dystrophin	NICM	>90%	Respiratory, HF	Yes, uncertain etiology
Becker	X-linked recessive	Dystrophin	NICM	60%–75%	HF, respiratory	Yes, uncertain etiology
Limb-girdle type 1B	Autosomal dominant	*Lamin A/C*	Conduction system disease and NICM	>90%	Sudden, HF	Yes
Limb-girdle type 2C-2F	Autosomal recessive	Sarcoglycan	NICM	<25%	Respiratory, HF	Uncertain
Limb-girdle type 2I	Autosomal recessive	Fukutin-related protein	NICM	20%–80%	Respiratory, HF	Uncertain
Myotonic type 1	Autosomal dominant	CTG repeat expansion	Conduction system disease and NICM	60%–80%	Respiratory, sudden, HF	30% of deaths, uncertain bradycardia versus tachycardia
Myotonic type 2	Autosomal dominant	CCTG repeat expansion	Conduction system disease	10%–25%	Normal causes	Reported
Emery-Dreifuss	X-linked and autosomal dominant or recessive	Emerin, *Lamin A/C*	Conduction system disease and NICM	>90%	Sudden, HF	Yes
Facioscapulohumeral	Autosomal dominant	D4Z4 repeat contaction	Possibly conduction disease	5%–15%	Normal causes, respiratory rarely	Not reported

Adapted with permission from Groh, et al.[S6.8-5]
HF indicates heart failure; and NICM, nonischemic cardiomyopathy.

6.9. Cardiac Channelopathies

Recommendations for Cardiac Channelopathies References that support the recommendations are summarized in Online Data Supplement 39.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In first-degree relatives of patients who have a causative mutation for long QT syndrome, catecholaminergic polymorphic ventricular tachycardia, short QT syndrome, or Brugada syndrome, genetic counseling and mutation-specific genetic testing are recommended.[S6.9-1–S6.9-6]
I	B-NR	2. In patients with a cardiac channelopathy and SCA, an ICD is recommended if meaningful survival of greater than 1 year is expected.[S6.9-7–S6.9-13]

6.9.1. Specific Cardiac Channelopathy Syndromes

6.9.1.1. Congenital Long QT Syndrome

Recommendations for Long QT Syndrome References that support the recommendations are summarized in Online Data Supplement 40.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with long QT syndrome with a resting QTc greater than 470 ms, a beta blocker is recommended.[S6.9.1.1-1–S6.9.1.1-5]
I	B-NR	2. In high-risk patients with symptomatic long QT syndrome in whom a beta blocker is ineffective or not tolerated, intensification of therapy with additional medications (guided by consideration of the particular long QT syndrome type), left cardiac sympathetic denervation, and/or an ICD is recommended.[S6.9.1.1-2,S6.9.1.1-6–S6.9.1.1-12]
I	B-NR	3. In patients with long QT syndrome and recurrent appropriate ICD shocks despite maximum tolerated doses of a beta blocker, intensification of medical therapy with additional medications (guided by consideration of the particular long QT syndrome type) or left cardiac sympathetic denervation, is recommended.[S6.9.1.1-6,S6.9.1.1-7,S6.9.1.1-10,S6.9.1.1-13–S6.9.1.1-16]
I	B-NR	4. In patients with clinically diagnosed long QT syndrome, genetic counseling and genetic testing are recommended.[S6.9.1.1-17–S6.9.1.1-21]
IIa	B-NR	5. In patients with suspected long QT syndrome, ambulatory electrocardiographic monitoring, recording the ECG lying and immediately on standing, and/or exercise treadmill testing can be useful for establishing a diagnosis and monitoring the response to therapy.[S6.9.1.1-22–S6.9.1.1-29]
IIa	B-NR	6. In asymptomatic patients with long QT syndrome and a resting QTc less than 470 ms, chronic therapy with a beta blocker is reasonable.[S6.9.1.1-3,S6.9.1.1-30,S6.9.1.1-31]
IIb	B-NR	7. In asymptomatic patients with long QT syndrome and a resting QTc greater than 500 ms while receiving a beta blocker, intensification of therapy with medications (guided by consideration of the particular long QT syndrome type), left cardiac sympathetic denervation or an ICD may be considered.[S6.9.1.1-2,S6.9.1.1-8,S6.9.1.1-11,S6.9.1.1-30]
III: Harm	B-NR	8. In patients with long QT syndrome, QT-prolonging medications are potentially harmful.[S6.9.1.1-5,S6.9.1.1-12,S6.9.1.1-32–S6.9.1.1-34]

Table 10. Commonly Used QT-Prolonging Medications[S6.9.1.1-35,S6.9.1.1-36]

Examples of QT Prolonging Medications*			
Antiarrhythmic Medications	**Psychotropic Medications**	**Antibiotics**	**Others**
Disopyramide	Haloperidol	Erythromycin	Methadone
Procainamide (N-acetylprocainamide)	Phenothiazines	Pentamidine	Probucol
Quinidine	Citalopram	Azithromycin	Droperidol
Dofetilide	Tricyclic antidepressants	Chloroquine	Ondansetron
Dronedarone		Ciprofloxacin	
Ibutilide		Fluconazole	
Sotalol		Levofloxacin	
Amiodarone†		Moxifloxacin	
		Clarithromycin	
		Itraconazole	
		Ketoconazole	

*A more complete list is maintained at: www.crediblemeds.org.[S6.9.1.1-35]
†Amiodarone rarely causes torsades de pointes.

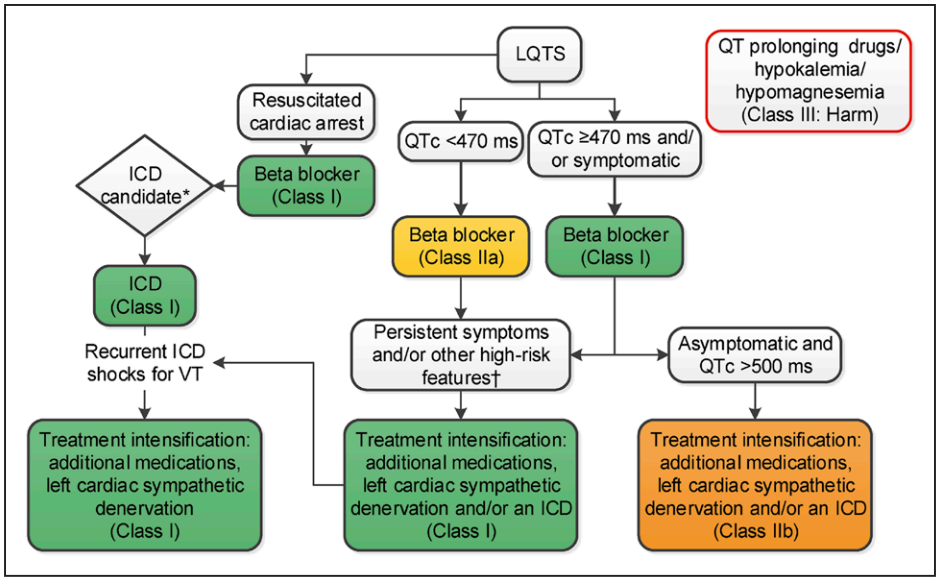


Figure 9. Prevention of SCD in patients with long QT syndrome.
Colors correspond to Class of Recommendation in Table 1. See Section 7.9.1.1 in the full-text guideline for discussion. *ICD candidacy as determined by functional status, life expectancy, or patient preference. †High-risk patients with LQTS include those with QTc >500 ms, genotypes LQT2 and LQT3, females with genotype LQT2, <40 years of age, onset of symptoms at <10 years of age, and patients with recurrent syncope. ICD indicates implantable cardioverter-defibrillator; LQTS, long QT syndrome; VT, ventricular tachycardia.

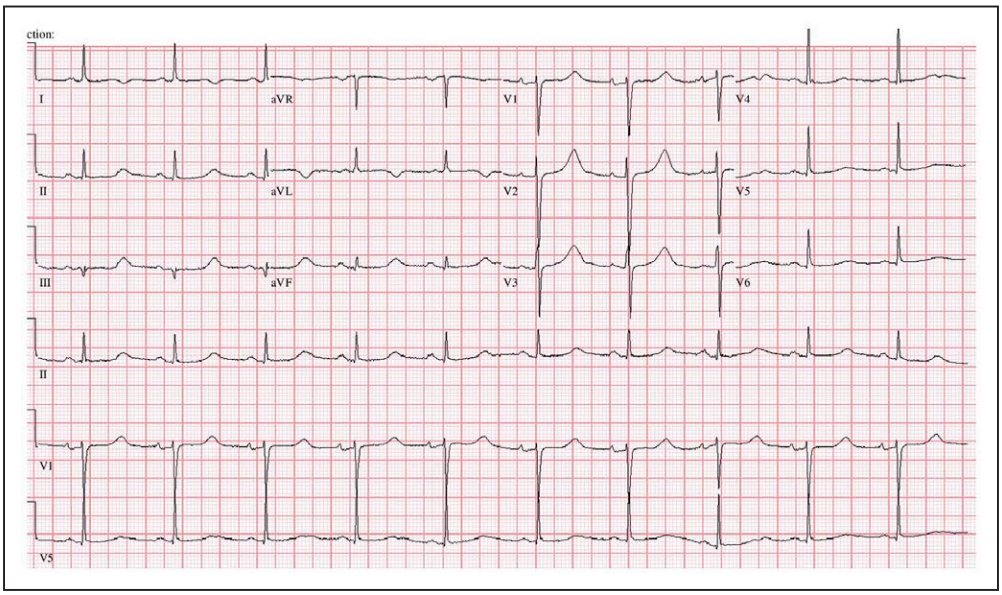


Figure 10. Long QT syndrome type 1.

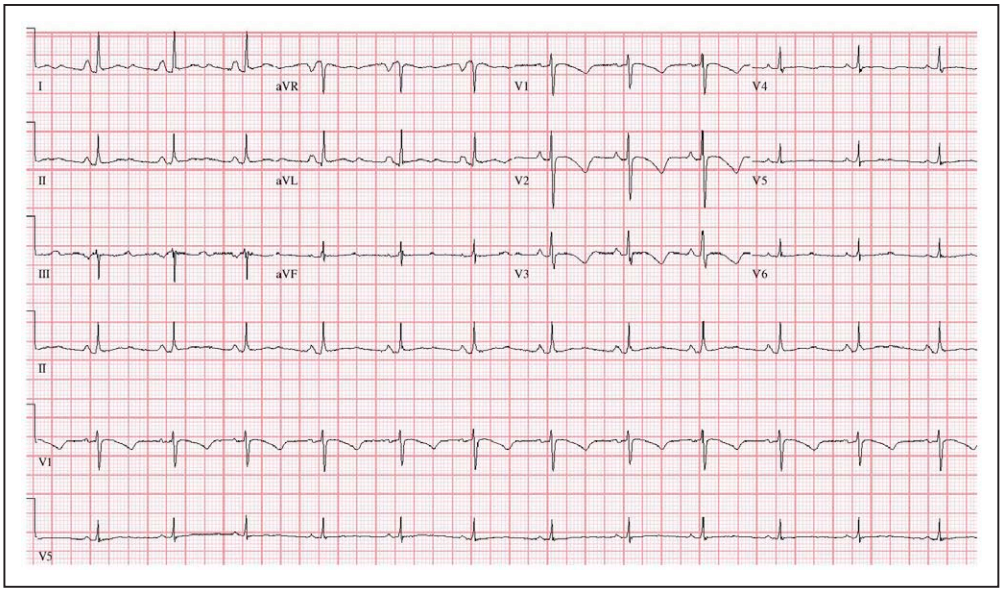


Figure 11. Long QT syndrome type 2.

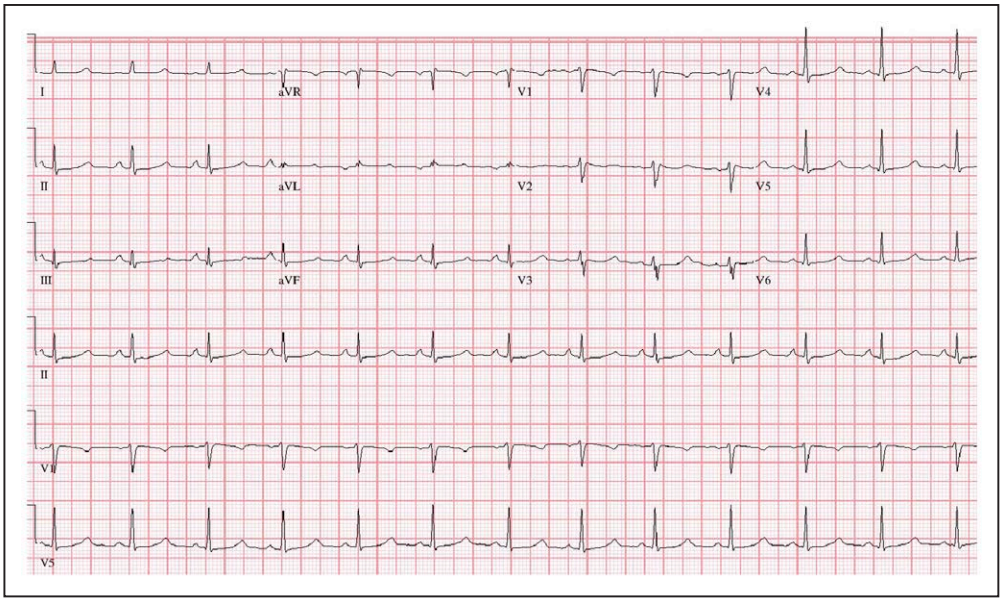


Figure 12. Long QT syndrome type 3.

6.9.1.2. Catecholaminergic Polymorphic Ventricular Tachycardia

Recommendations for Catecholaminergic Polymorphic Ventricular Tachycardia References that support the recommendations are summarized in Online Data Supplement 41.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with catecholaminergic polymorphic ventricular tachycardia, a beta blocker is recommended.[S6.9.1.2-1,S6.9.1.2-2]

Recommendations for Catecholaminergic Polymorphic Ventricular Tachycardia (Continued)		
COR	**LOE**	**Recommendations**
I	B-NR	2. In patients with catecholaminergic polymorphic ventricular tachycardia and recurrent sustained VT or syncope, while receiving adequate or maximally tolerated beta blocker, treatment intensification with either combination medication therapy (eg, beta blocker, flecainide), left cardiac sympathetic denervation, and/or an ICD is recommended.[S6.9.1.2-2–S6.9.1.2-6]
IIa	B-NR	3. In patients with catecholaminergic polymorphic ventricular tachycardia and with clinical VT or exertional syncope, genetic counseling and genetic testing are reasonable.[S6.9.1.2-7]

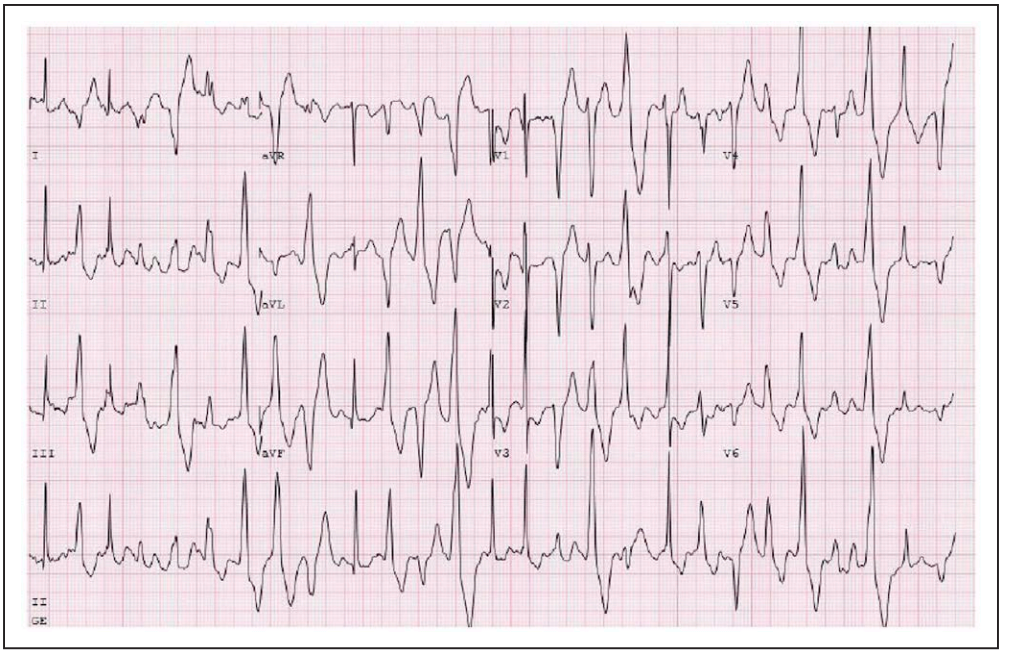


Figure 13. **Exercise-induced polymorphic VT in catecholaminergic polymorphic ventricular tachycardia.**

6.9.1.3. Brugada Syndrome

Recommendations for Brugada Syndrome References that support the recommendations are summarized in Online Data Supplement 42 and the Systematic Review Report.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In asymptomatic patients with only inducible type 1 Brugada electrocardiographic pattern, observation without therapy is recommended.[S6.9.1.3-1–S6.9.1.3-5]
I	B-NR	2. In patients with Brugada syndrome with spontaneous type 1 Brugada electrocardiographic pattern and cardiac arrest, sustained VA or a recent history of syncope presumed due to VA, an ICD is recommended if meaningful survival of greater than 1 year is expected.[S6.9.1.3-4,S6.9.1.3-6]
I	B-NR	3. In patients with Brugada syndrome experiencing recurrent ICD shocks for polymorphic VT, intensification of therapy with quinidine or catheter ablation is recommended.[S6.9.1.3-7–S6.9.1.3-11]
I	B-NR	4. In patients with spontaneous type 1 Brugada electrocardiographic pattern and symptomatic VA who either are not candidates for or decline an ICD, quinidine or catheter ablation is recommended.[S6.9.1.3-7,S6.9.1.3-9–S6.9.1.3-11]
IIa	B-NR	5. In patients with suspected Brugada syndrome in the absence of a spontaneous type 1 Brugada electrocardiographic pattern, a pharmacological challenge using a sodium channel blocker can be useful for diagnosis.[S6.9.1.3-12–S6.9.1.3-14]
IIb	B-NR^{SR}	6. In patients with asymptomatic Brugada syndrome and a spontaneous type 1 Brugada electrocardiographic pattern, an electrophysiological study with programmed ventricular stimulation using single and double extrastimuli may be considered for further risk stratification.[S6.9.1.3-1,S6.9.1.3-6,S6.9.1.3-13, S6.9.1.3-15–S6.9.1.3-17]
IIb	C-EO	7. In patients with suspected or established Brugada syndrome, genetic counseling and genetic testing may be useful to facilitate cascade screening of relatives.[S6.9.1.3-18–S6.9.1.3-20]

SR indicated systematic review.

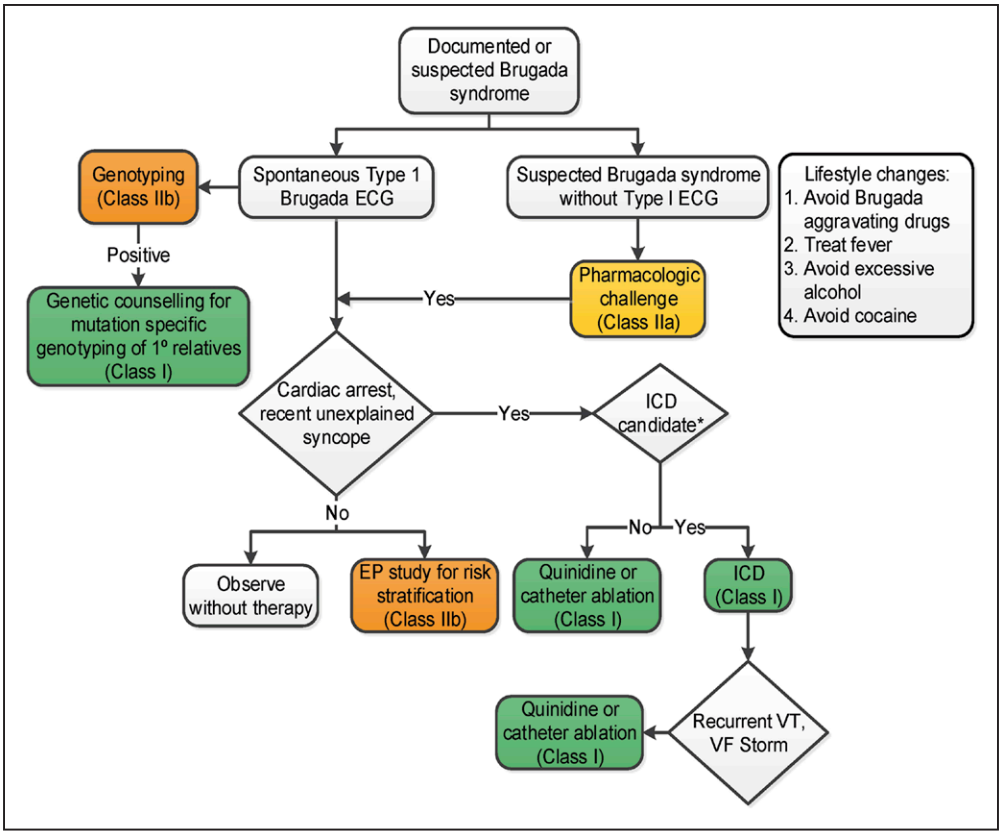


Figure 14. Prevention of SCD in patients with brugada syndrome.
Colors correspond to Class of Recommendation in Table 1. See Section 7.9.1.3 in the full-text guideline for discussion. *ICD candidacy as determined by functional status, life expectancy or patient preference. 1° indicates primary; ECG, electrocardiogram; EP, electrophysiological; ICD implantable cardioverter-defibrillator; SCD, sudden cardiac death; VT, ventricular tachycardia; and VF, ventricular fibrillation.

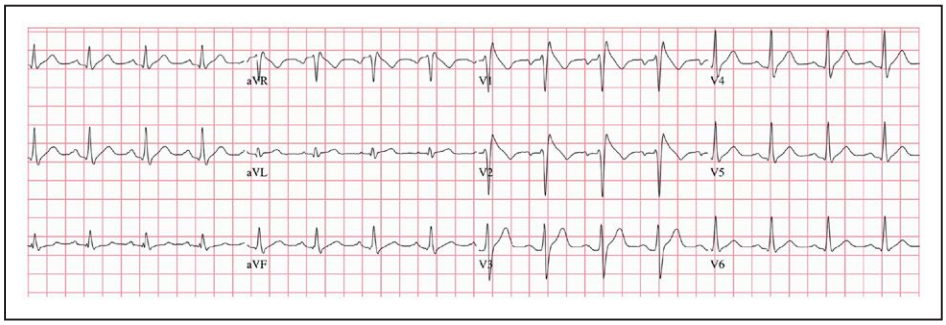


Figure 15. Brugada syndrome.

6.9.1.4. Early Repolarization "J-wave" Syndrome

Recommendations for Early Repolarization Syndrome References that support the recommendations are summarized in Online Data Supplement 43.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In asymptomatic patients with an early repolarization pattern on ECG, observation without treatment is recommended.[S6.9.1.4-1,S6.9.1.4-2]
I	B-NR	2. In patients with early repolarization pattern on ECG and cardiac arrest or sustained VA, an ICD is recommended if meaningful survival greater than 1 year is expected.[S6.9.1.4-3,S6.9.1.4-4]
III: No Benefit	B-NR	3. In patients with early repolarization pattern on ECG, genetic testing is not recommended.[S6.9.1.4-5]

6.9.1.5. Short QT Syndrome

Recommendations for Short QT Syndrome References that support the recommendations are summarized in Online Data Supplement 44.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In asymptomatic patients with a short QTc interval, observation without treatment is recommended.[S6.9.1.5-1,S6.9.1.5-2]
I	B-NR	2. In patients with short QT syndrome who have a cardiac arrest or sustained VA, an ICD is recommended if meaningful survival greater than 1 year is expected.[S6.9.1.5-3–S6.9.1.5-5]

Recommendations for Short QT Syndrome (Continued)		
COR	LOE	Recommendations
IIa	C-LD	3. In patients with short QT syndrome and recurrent sustained VA, treatment with quinidine can be useful.[S6.9.1.5-3,S6.9.1.5-5,S6.9.1.5-6]
IIa	C-LD	4. In patients with short QT syndrome and VT/VF storm, isoproterenol infusion can be effective.[S6.9.1.5-7]
IIb	C-EO	5. In patients with short QT syndrome, genetic testing may be considered to facilitate screening of first-degree relatives.[S6.9.1.5-4]

7. VA IN THE STRUCTURALLY NORMAL HEART

Recommendations for VA in the Structurally Normal Heart References that support the recommendations are summarized in Online Data Supplement 45.		
COR	LOE	Recommendations
I	B-R	1. In patients with symptomatic PVCs in an otherwise normal heart, treatment with a beta blocker or nondihydropyridine calcium channel blocker is useful to reduce recurrent arrhythmias and improve symptoms.[S7-1,S7-2]
IIa	B-R	2. In patients with symptomatic VA in an otherwise normal heart, treatment with an antiarrhythmic medication is reasonable to reduce recurrent symptomatic arrhythmias and improve symptoms if beta blockers and nondihydropyridine calcium channel blockers are ineffective or not tolerated.[S7-3,S7-4]

7.1. Outflow Tract and Atrioventricular Annular VA

Recommendations for Outflow Tract VA References that support the recommendations are summarized in Online Data Supplement 46.		
COR	LOE	Recommendations
I	B-NR	1. In patients with symptomatic outflow tract VA in an otherwise normal heart for whom antiarrhythmic medications are ineffective, not tolerated, or not the patient's preference, catheter ablation is useful.[S7.1-1–S7.1-3]
I	B-NR	2. In patients with symptomatic outflow tract VT in an otherwise normal heart, a beta blocker or a calcium channel blocker is useful.[S7.1-1–S7.1-3]

7.2. Papillary Muscle VA

Recommendation for Papillary Muscle VA (PVCs and VT) References that support the recommendation are summarized in Online Data Supplement 47.		
COR	LOE	Recommendation
I	B-NR	1. In patients with symptomatic VA arising from the papillary muscles for whom antiarrhythmic medications are ineffective, not tolerated, or not the patient's preference, catheter ablation is useful.[S7.2-1–S7.2-5]

7.3. Interfascicular Reentrant VT (Belhassen Tachycardia)

Recommendations for Interfascicular Reentrant VT (Belhassen Tachycardia) References that support the recommendations are summarized in Online Data Supplement 48.		
COR	LOE	Recommendations
I	B-NR	1. In patients with verapamil-sensitive, idiopathic LVT related to interfascicular reentry for whom antiarrhythmic medications are ineffective, not tolerated, or not the patient's preference, catheter ablation is useful.[S7.3-1–S7.3-3]
I	B-NR	2. In patients with sustained hemodynamically tolerated verapamil-sensitive, idiopathic LVT related to interfascicular reentry, intravenous verapamil is recommended for VT termination.[S7.3-3–S7.3-6]
IIa	C-LD	3. In patients with recurrent verapamil-sensitive idiopathic LVT, chronic therapy with oral verapamil can be useful.[S7.3-7–S7.3-10]

7.4. Idiopathic Polymorphic VT/VF

Recommendations for Idiopathic Polymorphic VT/VF References that support the recommendations are summarized in Online Data Supplement 49.		
COR	LOE	Recommendations
I	B-NR	1. In young patients (<40 years of age) with unexplained SCA, unexplained near drowning, or recurrent exertional syncope, who do not have ischemic or other structural heart disease, further evaluation for genetic arrhythmia syndromes is recommended.[S7.4-1–S7.4-8]
I	B-NR	2. In patients resuscitated from SCA due to idiopathic polymorphic VT or VF, an ICD is recommended if meaningful survival greater than 1 year is expected.[S7.4-9–S7.4-13]
I	B-NR	3. For patients with recurrent episodes of idiopathic VF initiated by PVCs with a consistent QRS morphology, catheter ablation is useful.[S7.4-11,S7.4-14]

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8. PVC-INDUCED CARDIOMYOPATHY

Recommendations for PVC-Induced Cardiomyopathy References that support the recommendations are summarized in Online Data Supplement 50.		
COR	**LOE**	**Recommendations**
I	B-NR	1. For patients who require arrhythmia suppression for symptoms or declining ventricular function suspected to be due to frequent PVCs (generally >15% of beats and predominately of 1 morphology) and for whom antiarrhythmic medications are ineffective, not tolerated, or not the patient's preference, catheter ablation is useful.[S8-1,S8-2]
IIa	B-NR	2. In patients with PVC-induced cardiomyopathy, pharmacologic treatment (eg, beta blocker, amiodarone) is reasonable to reduce recurrent arrhythmias, and improve symptoms and LV function.[S8-3,S8-4]

9. VA AND SCD RELATED TO SPECIFIC POPULATIONS

9.1. Pregnancy

Recommendations for Pregnancy References that support the recommendations are summarized in Online Data Supplement 51.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In mothers with long QT syndrome, a beta blocker should be continued during pregnancy and throughout the postpartum period including in women who are breastfeeding.[S9.1-1]
I	C-EO	2. In the pregnant patient with sustained VA, electrical cardioversion is safe and effective and should be used with standard electrode configuration.[S9.1-2,S9.1-3]
IIa	B-NR	3. In pregnant patients needing an ICD or VT ablation, it is reasonable to undergo these procedures during pregnancy, preferably after the first trimester.[S9.1-4,S9.1-5]

9.2. Older Patients With Comorbidities

Recommendation for Older Patients With Comorbidities See Systematic Review Report.[S9.2-1]		
COR	**LOE**	**Recommendation**
IIa	B-NR[SR]	1. For older patients and those with significant comorbidities, who meet indications for a primary prevention ICD, an ICD is reasonable if meaningful survival of greater than 1 year is expected.[S9.2-1]

SR indicates systematic review.

9.3. Medication-Induced Arrhythmias

Recommendations for Medication-Induced Arrhythmias References that support the recommendations are summarized in Online Data Supplement 52 and 53.		
COR	**LOE**	**Recommendation**
Digoxin		
I	B-NR	1. Administration of digoxin antibodies is recommended for patients who present with sustained VA potentially due to digoxin toxicity.[S9.3-1,S9.3-2]
Medication-induced QT prolongation and torsades de pointes		
I	B-NR	2. In patients with recurrent torsades de pointes associated with acquired QT prolongation and bradycardia that cannot be suppressed with intravenous magnesium administration, increasing the heart rate with atrial or ventricular pacing or isoproterenol are recommended to suppress the arrhythmia.[S9.3-3]
I	C-LD	3. For patients with QT prolongation due to a medication, hypokalemia, hypomagnesemia, or other acquired factor and recurrent torsades de pointes, administration of intravenous magnesium sulfate is recommended to suppress the arrhythmia.[S9.3-4,S9.3-5]
I	C-LD	4. For patients with torsades de pointes associated with acquired QT prolongation, potassium repletion to 4.0 mmol/L or more and magnesium repletion to normal values (eg, ≥2.0 mmol/L) are beneficial.[S9.3-6,S9.3-7]
Sodium channel blocker–related toxicity		
IIa	C-LD	5. In patients taking sodium channel blockers who present with elevated defibrillation or pacing thresholds, discontinuing the presumed responsible medication or reprogramming the device can be useful to restore effective device therapy.[S9.3-8,S9.3-9]
III: Harm	B-NR	6. In patients with congenital or acquired long QT syndrome, QT-prolonging medications are potentially harmful.[S9.3-10]

9.4. Adult Congenital Heart Disease

Recommendations for Adult Congenital Heart Disease References that support the recommendations are summarized in Online Data Supplement 54.		
COR	**LOE**	**Recommendations**
I	B-NR	1. Adult patients with repaired complex congenital heart disease presenting with frequent, complex, or sustained VA, or unexplained syncope should undergo evaluation for potential residual anatomic or coronary abnormalities.[S9.4-1–S9.4-6]
I	B-NR	2. In patients with adult congenital heart disease and complex or sustained VA in the presence of important residual hemodynamic lesions, treatment of hemodynamic abnormalities with catheter or surgical intervention as feasible is indicated prior to consideration of ablation or an ICD.[S9.4-3,S9.4-7–S9.4-12]

Recommendations for Adult Congenital Heart Disease (Continued)		
COR	LOE	Recommendations
I	B-NR	3. In patients with adult congenital heart disease and hemodynamically unstable VT, an ICD is recommended after evaluation and appropriate treatment for residual lesions/ventricular dysfunction if meaningful survival of greater than 1 year is expected.[S9.4-13–S9.4-17]
I	B-NR	4. In patients with adult congenital heart disease with SCA due to VT or VF in the absence of reversible causes, an ICD is recommended if meaningful survival of greater than 1 year is expected.[S9.4-13–S9.4-17]
IIa	B-NR	5. In adults with repaired tetralogy of Fallot physiology with high-risk characteristics and frequent VA, an electrophysiological study can be useful to evaluate the risk of sustained VT/VF.[S9.4-18,S9.4-19]
IIa	B-NR	6. In adults with repaired tetralogy of Fallot physiology and inducible VT/VF or spontaneous sustained VT, implantation of an ICD is reasonable if meaningful survival greater than 1 year is expected.[S9.4-1,S9.4-19,S9.4-20]
IIa	B-NR	7. In patients with adult congenital heart disease with recurrent sustained monomorphic VT or recurrent ICD shocks for VT, catheter ablation can be effective.[S9.4-21–S9.4-25]

Recommendations for Adult Congenital Heart Disease (Continued)		
COR	LOE	Recommendations
IIa	B-NR	8. In adults with repaired severe complexity adult congenital heart disease and frequent or complex VA, a beta blocker can be beneficial to reduce the risk of SCA[S9.4-26].
IIa	B-NR	9. In patients with repaired moderate or severe complexity adult congenital heart disease with unexplained syncope and at least moderate ventricular dysfunction or marked hypertrophy, either ICD implantation or an electrophysiological study with ICD implantation for inducible sustained VA is reasonable if meaningful survival of greater than 1 year is expected.[S9.4-5,S9.4-16,S9.4-27–S9.4-29]
IIb	B-NR	10. In patients with adult congenital heart disease and severe ventricular dysfunction (LVEF <35%) and symptoms of heart failure despite GDMT or additional risk factors, ICD implantation may be considered if meaningful survival of greater than 1 year is expected.[S9.4-14–S9.4-16,S9.4-20]
III: Harm	B-NR	11. In patients with adult congenital heart disease who have asymptomatic VA, prophylactic antiarrhythmic therapy with class Ic medications (ie, flecainide, propafenone) or amiodarone is potentially harmful.[S9.4-30–S9.4-32]

Table 11. Congenital Heart Disease: Risk Factors for VA/SCD

Congenital Heart Disease	Incidence of VA	Incidence of SCD	Higher Risk Characteristics
Simple complexity			
ASD[S9.4-33–S9.4-40]	2%–6%	<1.5%	Ventricular pacing
VSD[S9.4-27,S9.4-33–S9.4-41]	3%–18%	<3%	RV dilatation Pulmonary hypertension *NKX2.5* gene
Moderate complexity			
Tetralogy of Fallot[S9.4-1,S9.4-2,S9.4-5,S9.4-6,S9.4-28,S9.4-33,S9.4-34,S9.4-42–S9.4-51]	14%–31%	1.4%–8.3%	Unexplained syncope Frequent or complex VA Sustained VT QRS duration ≥180 ms Inducible sustained VT Atrial tachycardia Decreased LVEF Dilated right ventricle Severe PR Severe PS
Aortic stenosis[S9.4-27,S9.4-33,S9.4-47]	10%–34%	3%–20%	Unexplained syncope Severe LV hypertrophy Aortic stenosis mean pressure gradient >40 mm Hg Ventricular dysfunction
Coarctation of aorta[S9.4-28,S9.4-29,S9.4-33,S9.4-44,S9.4-47,S9.4-48]	2%	2%	Aneurysm at repair site Aortic stenosis Systemic hypertension Premature coronary artery disease
Ebstein's anomaly[S9.4-34,S9.4-46,S9.4-52]	2%	3%–6%	Cardiomegaly Atrial fibrillation Wide complex tachycardia Mitral regurgitation Dilated RVOT

(*Continued*)

Table 11. Continued

Congenital Heart Disease	Incidence of VA	Incidence of SCD	Higher Risk Characteristics
Severe complexity			
Transposition of the great arteries[S9.4-27,S9.4-33,S9.4-34,S9.4-44,S9.4-46–S9.4-48,S9.4-52–S9.4-54]			Atrial switch Mustard repair
Atrial switch	2%	3%–9.5%	Prior VSD closure
Arterial switch	2%	1%	Unexplained syncope
cc-TGA	10%	17%–25%	Atrial tachycardia Coronary orifice stenosis Systemic ventricular dysfunction Severe tricuspid regurgitation
Truncus arteriosus[S9.4-55,S9.4-56]	10%	4%	Multiple surgical repairs Coronary anomalies Ventricular dysfunction and/or hypertrophy
Fontan repair for univentricular physiology*[S9.4-27,S9.4-33,S9.4-34,S9.4-46,S9.4-52,S9.4-57,S9.4-58]	5%–17%	2.8%–5.4%	Atrial tachycardia Longer duration of follow-up Ascites Protein-losing enteropathy

*Univentricular physiology includes: Tricuspid atresia, Double inlet left ventricle, Mitral atresia, Hypoplastic left heart, Unbalanced AV septal defect.

ASD indicates atrial septal defect; cc-TGA, congenitally corrected transposition of the great arteries; LV, left ventricular; LVEF, left ventricular ejection fraction; PR, pulmonary regurgitation; PS, pulmonary stenosis; RV, right ventricular; RVOT, right ventricular outflow tract; SCD, sudden cardiac death; VA, ventricular arrhythmia; VSD, ventricular septal defect; and VT, ventricular tachycardia.

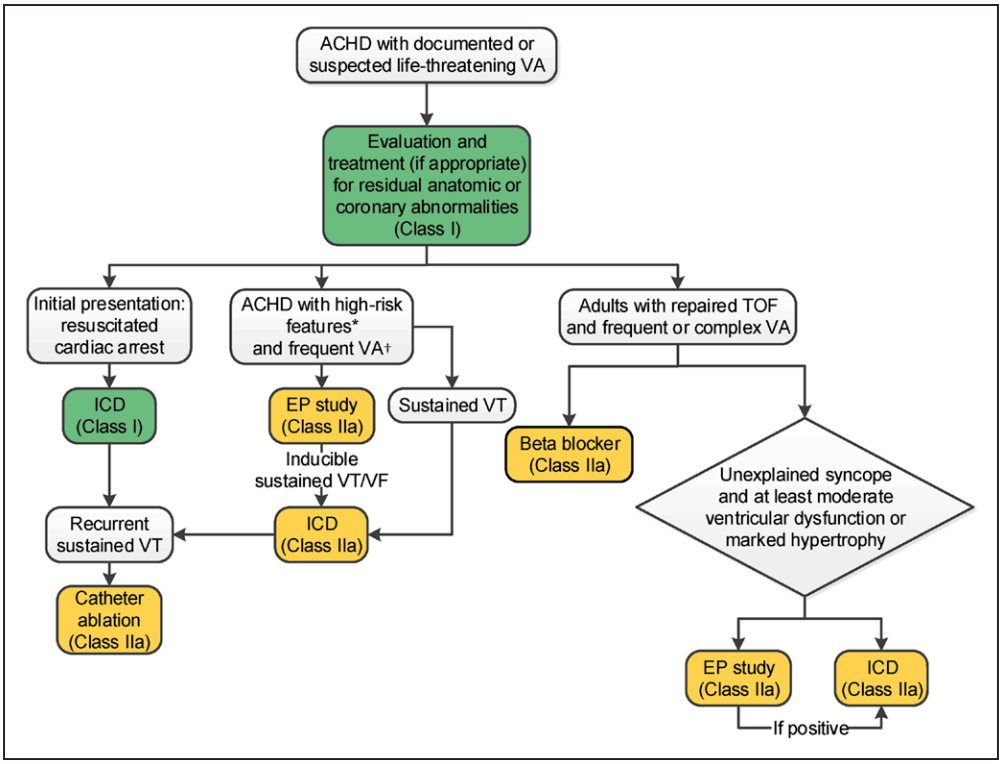

Figure 16. Prevention of SCD in patients with adult congenital heart disease.
Colors correspond to Class of Recommendation in Table 1. See Section 10.8 in the full-text guideline for discussion. *High-risk features: prior palliative systemic to pulmonary shunts, unexplained syncope, frequent PVC, atrial tachycardia, QRS duration ≥180 ms, decreased LVEF or diastolic dysfunction, dilated right ventricle, severe pulmonary regurgitation or stenosis, or elevated levels of BNP. †Frequent VA refers to frequent PVCs and/or nonsustained VT. ACHD indicates adult congenital heart disease; BNP, B-type natriuretic peptide; EP, electrophysiological; ICD, implantable cardioverter-defibrillator; LVEF, left ventricular ejection fraction; PVC, premature ventricular complexes; SCD, sudden cardiac death; TOF, tetralogy of Fallot; VA, ventricular arrhythmia; and VT, ventricular tachycardia.

10. DEFIBRILLATORS OTHER THAN TRANSVENOUS ICDs

10.1. Subcutaneous Implantable Cardioverter-Defibrillator

Recommendations for Subcutaneous Implantable Cardioverter-Defibrillator References that support the recommendations are summarized in Online Data Supplement 55.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients who meet criteria for an ICD who have inadequate vascular access or are at high risk for infection, and in whom pacing for bradycardia or VT termination or as part of CRT is neither needed nor anticipated, a subcutaneous implantable cardioverter-defibrillator is recommended.[S10.1-1–S10.1-5]
IIa	B-NR	2. In patients who meet indication for an ICD, implantation of a subcutaneous implantable cardioverter-defibrillator is reasonable if pacing for bradycardia or VT termination or as part of CRT is neither needed nor anticipated.[S10.1-1–S10.1-4]
III: Harm	B-NR	3. In patients with an indication for bradycardia pacing or CRT, or for whom antitachycardia pacing for VT termination is required, a subcutaneous implantable cardioverter-defibrillator should not be implanted.[S10.1-1–S10.1-4,S10.1-6–S10.1-8]

10.2. Wearable Cardioverter-Defibrillator

Recommendations for Wearable Cardioverter-Defibrillator References that support the recommendations are summarized in Online Data Supplement 56.		
COR	**LOE**	**Recommendations**
IIa	B-NR	1. In patients with an ICD and a history of SCA or sustained VA in whom removal of the ICD is required (as with infection), the wearable cardioverter-defibrillator is reasonable for the prevention of SCD.[S10.2-1–S10.2-4]
IIb	B-NR	2. In patients at an increased risk of SCD but who are not ineligible for an ICD, such as awaiting cardiac transplant, having an LVEF of 35% or less and are within 40 days from an MI, or have newly diagnosed NICM, revascularization within the past 90 days, myocarditis or secondary cardiomyopathy or a systemic infection, the wearable cardioverter-defibrillator may be reasonable.[S10.2-1–S10.2-5]

11. SPECIAL CONSIDERATIONS FOR CATHETER ABLATION

Recommendations for Catheter Ablation References that support the recommendations are summarized in Online Data Supplement 57.		
COR	**LOE**	**Recommendations**
I	C-LD	1. In patients with bundle-branch reentrant VT, catheter ablation is useful for reducing the risk of recurrent VT and ICD shocks.[S11-1–S11-3]
IIa	B-NR	2. In patients with structural heart disease who have failed endocardial catheter ablation, epicardial catheter ablation can be useful for reducing the risk of recurrent monomorphic VT.[S11-4–S11-6]

12. POSTMORTEM EVALUATION OF SCD

Recommendations for Postmortem Evaluation of SCD References that support the recommendations are summarized in Online Data Supplement 58.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In victims of SCD without obvious causes, a standardized cardiac-specific autopsy is recommended.[S12-1,S12-2]
I	B-NR	2. In first-degree relatives of SCD victims who were 40 years of age or younger, cardiac evaluation is recommended, with genetic counseling and genetic testing performed as indicated by clinical findings.[S12-3]
IIa	B-NR	3. In victims of SCD with an autopsy that implicates a potentially heritable cardiomyopathy or absence of structural disease, suggesting a potential cardiac channelopathy, postmortem genetic testing is reasonable.[S12-4–S12-7]
IIa	C-LD	4. In victims of SCD with a previously identified phenotype for a genetic arrhythmia-associated disorder, but without genotyping prior to death, postmortem genetic testing can be useful for the purpose of family risk profiling.[S12-8]

13. TERMINAL CARE

Recommendations for Terminal Care References that support the recommendations are summarized in Online Data Supplement 59.		
COR	**LOE**	**Recommendations**
I	C-EO	1. At the time of ICD implantation or replacement, and during advance care planning, patients should be informed that their ICD shock therapy can be deactivated at any time if it is consistent with their goals and preferences.
I	C-EO	2. In patients with refractory HF symptoms, refractory sustained VA, or nearing the end of life from other illness, clinicians should discuss ICD shock deactivation and consider the patients' goals and preferences.

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14. SHARED DECISION-MAKING

Recommendations for Shared Decision-Making References that support the recommendations are summarized in Online Data Supplement 60.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with VA or at increased risk for SCD, clinicians should adopt a shared decision-making approach in which treatment decisions are based not only on the best available evidence, but also on the patients' health goals, preferences, and values.[S14-1–S14-5]
I	B-NR	2. Patients considering implantation of a new ICD or replacement of an existing ICD for a low battery should be informed of their individual risk of SCD and nonsudden death from HF or noncardiac conditions and the effectiveness, safety, and potential complications of the ICD in light of their health goals, preferences, and values.[S14-1–S14-5]

15. COST AND VALUE CONSIDERATIONS

The key principles of value assessment as part of clinical practice guidelines have been discussed in detail.[S15-1] Economic outcomes of clinical management strategies can be documented empirically using the same research designs as used in establishing clinical outcomes, including RCTs and observational comparisons. In addition, simulation models are often used to assess the value of management strategies, because the standard for cost-effectiveness studies is to compare life-time outcomes, and clinical studies usually have follow-up of a few years at most. Standards for economic modeling in health care have been published by an expert group.[S15-2]

Economic assessments of alternative management strategies for VA and prevention of SCD have primarily evaluated ICDs, including several RCTs[S15-3–S15-7] and observational studies,[S15-8,S15-9] and simulation models.[S15-10–S15-14] In all studies, patients who received ICDs had higher long-term costs. The high initial cost of the ICD device and the implantation procedure leads to higher long-term costs, because there are few, if any, subsequent cost-savings from implanting an ICD. ICDs without resynchronization capability do not reduce hospital readmissions, and may increase late costs due to device monitoring, complications, and replacement. However, the cost of the device and the procedure may change significantly over time.

The trial based assessments of the cost-effectiveness of the ICD are based on 3 to 6 years of follow-up, which is considerably shorter than the lifetime perspective that is standard in cost-effectiveness models. Because most of the incremental cost of the ICD is incurred immediately, while most of the potential effectiveness (life-years of survival added by the ICD) is accrued over many years, estimates of ICD cost effectiveness based on limited trial follow-up have a systematic bias toward showing lower value. Trial based economic studies that projected long-term ICD outcomes have consistently found more favorable cost-effectiveness ratios than estimates restricted to the duration of trial follow-up.[S15-4–S15-7] A lifetime simulation model applied to each major trial of primary prevention ICDs also reported consistently more favorable estimates of cost effectiveness than the estimates based on limited trial follow-up.[S15-11] Because the framework proposed for assessing value in ACC/AHA clinical practice guidelines uses benchmarks based on lifetime estimates,[S15-1] we have generally relied on the model-based estimates of ICD cost-effectiveness in applying value ratings to recommendations in this guideline.

The initial cost of an ICD device is similar regardless of the clinical indication, so variations in ICD cost effectiveness are driven primarily by potential differences in clinical effectiveness in extending survival in different patient populations. The effect of the years of life added by an ICD on its incremental cost-effectiveness ratio is illustrated in Figure 17: the cost-effectiveness ratio becomes rapidly unfavorable as the extension in survival time falls below 1 year, particularly below 0.5 year. This inverse relation strongly suggests that the value provided by an ICD will be highest when the risk of arrhythmic death due to VT/VF is relatively high and the risk of nonarrhythmic death (either cardiac or noncardiac) is relatively low, such that a meaningful increase in survival can be expected from the ICD. Thus, appropriate patient selection is fundamental to high value care in using the ICD to prevent SCD. It should also be recognized that, cost-effectiveness is also influenced by the costs for the ICD and implantation procedure, which are likely to change significantly over time.

The empirical evidence suggests that ICDs are not effective for primary prevention of SCD when implanted early after coronary artery bypass graft[S15-15] or an acute MI.[S15-16,S15-17] An analysis of individual patient level data from 3 secondary prevention trials[S15-18] showed a significant variation (P=0.011) in the clinical effectiveness of ICDs between patients with an LVEF ≤35% (hazard ratio: 0.66) and an LVEF >35% (hazard ratio: 1.2). Some studies and simulation models suggest that ICDs might prolong life expectancy to a greater extent when used in higher-risk patients than in lower-risk patients.[S15-19] In contrast, there is little evidence of variation in the effectiveness or cost-effectiveness of the ICD based on factors such as age or sex.[S15-20] Most studies of ICD effectiveness and value have been performed on patients with reduced LV function due to prior MI or NICM. There are few data on the effectiveness or value of an ICD for other potential clinical

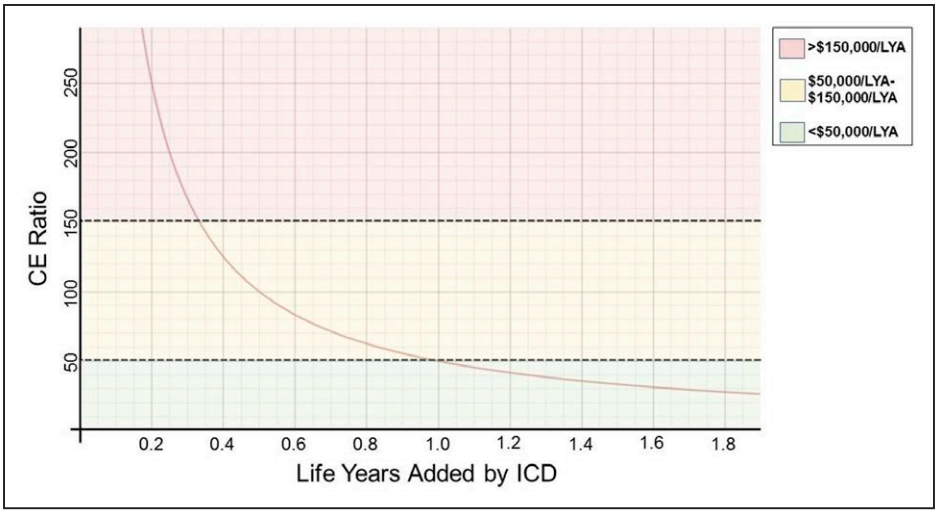

Figure 17. Incremental cost-effectiveness of ICD by years of life added* (example).
*Figure based on formula: Incremental cost-effectiveness ratio = $50 000/QALYs. CE indicated cost effectiveness; ICD, implantable cardioverter-defibrillator; LYA, life year added; and QALYs, quality-adjusted life-years.

indications, such as cardiac channelopathies or HCM, although studies have suggested that their potential cost effectiveness in such patients will depend on their underlying risk of SCD, with little evidence of value in low-risk patients.[S15-14]

16. QUALITY OF LIFE

ICD implantation has not had a significant effect on QoL in the overall population of patients enrolled in RCTs.[S16-1–S16-3] Several studies have, however, demonstrated that the subset of patients who receive inappropriate ICD shocks have worse QoL than patients who have an ICD but have not had inappropriate shocks.[S16-2] Because an ICD is designed to prevent SCD rather than to reduce symptoms, it would not be expected to improve QoL or functional status directly, but may have indirect, negative effects in some patients due to device complications, or indirect, positive effects in some patients due to reassurance of having a protective device in place.

17. EVIDENCE GAPS AND FUTURE RESEARCH NEEDS

Despite the numerous advances in risk stratification for SCD and prevention and treatment of SCD and VA, many gaps in knowledge remain. These gaps include:

- Identification of patients who are most likely to benefit from an ICD among all ICD-eligible patients. The role of novel markers (including genetic and imaging markers) and combinations of markers should be studied.
- Characterizing the role of the ICD in patient subgroups not well-represented in the pivotal ICD trials. Such subgroups include patients ≥80 years of age and those with kidney disease, especially patients with end-stage renal disease on dialysis, or multiple comorbidities.
- Methods to identify and treat patients at high individual risk for SCD who are not identified by current ICD eligibility criteria, including those who are within 40 days of an MI.
- Defining the role of the ICD in patients with HCM, ARVC, cardiac sarcoidosis, and inherited cardiac channelopathies in prospective studies (preferably RCT).
- Determining the best approach to patients due for elective ICD generator replacement due to battery depletion, but who may now be at low risk for SCA, such as if significant LVEF improvement has occurred.
- Obtaining more data on the efficacy and effectiveness of the S-ICD, compared with transvenous ICDs and on the extent of testing required, and its use with other novel technologies, including leadless pacemakers.
- Conducting RCTs on catheter ablation of VT in IHD, and cardiomyopathies that evaluates procedural end points, mortality, arrhythmia suppression, QoL, and costs.
- Improving identification of individuals without significant ventricular dysfunction who are at risk of SCD.
- Identifying mechanisms and risk factors for SCD in patients with HF*p*EF.
- Improving emergency response to out-of-hospital cardiac arrest.

- Developing better methods for identifying and ablating the arrhythmia substrate in structural heart disease.
- Developing better risk stratification of diseases and syndromes associated with sudden death, including IHD, NICM, ACHD, and Brugada syndrome.
- Identifying what causes different types of LQTS, CPVT, Brugada syndrome, HCM, and ARVC and advancing the genotype-phenotype relationships, genotype-dependent risk, and genotype-based tailoring of therapies for patients with inherited cardiomyopathies and inherited channelopathies.
- Defining the most appropriate and beneficial use of WCDs.
- Developing methods to identify and treat patients at high personal risk for SCD who are not identified by current ICD eligibility criteria.
- Defining the role of CMR in enhancing risk stratification for SCD.

Increasing research funding in this area, through existing and new mechanisms is critically important. Some have proposed research funding strategies that would offer business incentives to the insurance industries, while providing support for unresolved research goals. Such approaches should be tested.

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ARTICLE INFORMATION

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1. INTRODUCTION

1.4. Scope of the Guideline

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2. EPIDEMIOLOGY

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Appendix 1. Author Relationships With Industry and Other Entities (Relevant)—2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death (October 2017)

Committee Member	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness	Voting Recusals by Section*
Sana M. Al-Khatib, Chair	Duke Clinical Research Institute; Duke University—Professor of Medicine	None	None	None	None	None	None	None
William G. Stevenson, Vice Chair	Vanderbilt University Medical Center—Professor of Medicine—Brigham and Women's Hospital—Director of Clinical Cardiac EP	• St. Jude Medical	• Boston Scientific	• Biosense Webster‡	None	None	None	4.1, 4.2.2, 4.2.3, 5, 10.1, 5.4, 5.6, 6, 7, 8, 9 (except 9.7), 13, 15
Michael J. Ackerman	Mayo Clinic—Professor of Medicine, Pediatrics, and Pharmacology; Long QT Syndrome/Genetic Heart Rhythm Clinic and the Mayo Clinic Windland Smith Rice Sudden Death Genomics Laboratory—Director	• Audentes Therapeutics • Boston Scientific • Gilead Sciences • Invitae • Medtronic • MyoKardia • St. Jude Medical	None	None	None	• Transgenomic (Familion)† • Blue Ox Health Corporation‡ • AliveCor‡ • StemoniX‡	None	4.1, 4.2.2, 4.2.3, 4.2.6, 5 (except 5.1.5.2, 5.5), 6, 7, 8, 9, 10 (except 10.2) 11, 13, 15
William J. Bryant	Dominick Feld Hyde—Attorney at Law	None	None	None	None	None	None	None
David J. Callans	University of Pennsylvania Health System—Professor of Medicine; Associate Director of EP	• Biosense Webster† • Biotronik • Boston Scientific† • Medtronic • St. Jude Medical	None	None	• Biosense Webster (PI)‡ • Endosense (PI)‡	• Acutus	None	4.1, 4.2.2, 4.2.3, 5.3, 5.4, 5.5.1, 5.6, 6, 7, 8, 9 (expect 9.7), 10 (except 10.3), 13, 15
Anne B. Curtis	University at Buffalo—SUNY Distinguished Professor; Charles and Mary Bauer Professor and Chair	• Medtronic • St. Jude Medical	None	None	None	None	None	4.1, 4.2.2, 4.2.3, 5.1.1, 5.1.2, 5.1.3, 5.1.4, 5.2, 5.4, 5.6, 6, 7, 8, 9, 10, 12, 13, 15
Barbara J. Deal	Getz Professor of Cardiology Feinberg School of Medicine Northwestern University	None	None	None	None	None	None	None
Timm Dickfeld	University of Maryland—Professor of Medicine	• Biosense • St. Jude Medical • Siemens	None	None	• Biosense† • General Electric†	• Impulse Dynamics‡ • Siemens†	None	4.1, 4.2 (except 4.2.6), 4.3, 5.3, 5.4, 5.6, 6, 7, 8, 9 (except 9.7), 10.1, 11, 13, 15
Anne M. Gillis	University of Calgary—Professor of Medicine	None	None	None	• Medtronic	None	None	4.2, 5.2.2, 5.3.2, 6.4.1, 6.4.2, 6.4.4, 6.5, 6.7, 7, 8, 9, 10, 11 (except 11.7), 13, 15
Christopher B. Granger	Duke Clinical Research Institute; Duke University—Professor of Medicine; Director, Cardiac Care Unit	• AstraZeneca† • Gilead Sciences† • GlaxoSmithKline† • Janssen Pharmaceuticals† • Medtronic† • Pfizer† • Sanofi-aventis†	None	None	• AstraZeneca† • GlaxoSmithKline • Janssen Pharmaceuticals† • Medtronic† • Pfizer • Sanofi-aventis†	• GE Healthcare† • Medtronic† • ZOLL Medical† • Spacelabs† • Phillips†	None	4, 5.1 (except 5.1.5), 5.2, 5.3, 5.4, 5.6, 6, 7, 8, 9, 12, 13, 15
Stephen C. Hammill	Mayo Clinic—Professor Emeritus of Medicine	None	None	None	None	None	None	None
Mark A. Hlatky	Stanford University School of Medicine—Professor of Health and Research Policy, and of Cardiovascular Medicine	None	None	None	None	None	None	None
José A. Joglar	UT Southwestern Medical Center—Professor of Internal Medicine; Clinical Cardiac EP—Fellowship Program Director	None	None	None	None	None	None	None
G. Neal Kay	University of Alabama at Birmingham—Professor Emeritus	None	None	None	None	None	None	None
Michael E. Field	University of Wisconsin School of Medicine and Public Health—Director, Clinical EP and Cardiac Arrhythmia Service, Associate Professor of Medicine	None	None	None	None	None	None	None

(*Continued*)

Appendix 1. Continued

Committee Member	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness	Voting Recusals by Section*
Gregg C. Fonarow	Ahmanson-UCLA Cardiomyopathy Center—Director; UCLA Division of Cardiology—Co-Chief	• Amgen • Janssen Pharmaceuticals • Medtronic • ZS Pharma	None	None	• Medtronic–IMPROVE-HF (Steering Committee)‡ • Medtronic†	None	None	4.1, 4.2.2, 4.2.3, 5.1 (except 5.1.5.1), 5.2, 5.3, 5.4, 5.6, 6, 7, 8, 9, 10, 12, 13, 15
Daniel D. Matlock	University of Colorado School of Medicine—Associate Professor of Medicine	None	None	None	None	None	None	None
Robert J. Myerburg	University of Miami Miller School of Medicine—Professor of Medicine and Physiology	None	None	None	None	None	None	None
Richard L. Page	University of Wisconsin Hospital and Clinics—Chair, Department of Medicine	None	None	None	None	None	None	None

This table represents the relationships of committee members with industry and other entities that were determined to be relevant to this document. These relationships were reviewed and updated in conjunction with all meetings and/or conference calls of the writing committee during the document development process. The table does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of ≥5% of the voting stock or share of the business entity, or ownership of ≥$5000 of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. Relationships that exist with no financial benefit are also included for the purpose of transparency. Relationships in this table are modest unless otherwise noted.

According to the ACC/AHA, a person has a *relevant* relationship IF: a) the *relationship or interest* relates to the same or similar subject matter, intellectual property or asset, topic, or issue addressed in the *document*; or b) the *company/entity* (with whom the relationship exists) makes a drug, drug class, or device addressed in the *document*, or makes a competing drug or device addressed in the *document*; or c) the *person or a member of the person's household*, has a reasonable potential for financial, professional or other personal gain or loss as a result of the issues/content addressed in the *document*.

*Writing committee members are required to recuse themselves from voting on sections to which their specific relationships with industry and other entities may apply. Section numbers pertain to those in the full-text guideline.

†Significant relationship.

‡No financial benefit.

ACC indicates American College of Cardiology; ACTION, Acute Coronary Treatment and Intervention Outcomes Network; AHA, American Heart Association; DSMB, data safety monitoring board; EP, Electrophysiology; HRS, Heart Rhythm Society; IMPROVE-HF, Improve the Use of Evidence-Based Heart Failure Therapies in the Outpatient Setting; and PI, principal investigator.

Appendix 2. Reviewer Relationships With Industry and Other Entities (Comprehensive)—2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death (July 2017)

Reviewer	Representation	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Salary	Expert Witness
Alfred E. Buxton	Content Reviewer	Professor of Medicine—Harvard Medical School—Beth Israel Deaconess Medical Center	None	None	None	• NHLBI (DSMB)†	• Medtronic† • Biosense Webster†	None	None
Andrew E. Epstein	Content Reviewer	Professor of Medicine—Cardiovascular Division University of Pennsylvania—Chief of Cardiology Section—Philadelphia VA Medical Center	• Zoll*	None	None	• Biotronik* • Boston Scientific* • Boston Scientific (DSMB)* • Medtronic* • Medtronic (DSMB) • St Jude Medical/ Abbott* • St Jude Medical/ Abbott (DSMB)*	None	None	• Defendant, Amiodarone pulmonary toxicity, 2016 • Defendant, Appropriateness of pacemaker implantation, 2016*
Brian Olshansky	Content Reviewer	Adjunct Professor of Medicine—Des Moines University—Professor Emeritus—University of Iowa	• Boehringer Ingelheim • Lundbeck Inc* • On-X/Cryolife	• Lundbeck Inc* • On-X/Cryolife	None	• Amarin (DSMB)*	None	None	• Plaintiff, Long QT sudden death, 2017
Bulent Gorenek	Content Reviewer—ACC EP Council	None	None	None	None	None	None	None	
Charles I. Berul	Content Reviewer	Division Chief of Pediatric Cardiology—Children's National Medical Center	None	None	None	None	• Circulation*	None	None
Darren Sudman	Content Reviewer	Executive Director—Simon's Fund	None	None	None	None	None	None	None
George J. Klein	Content Reviewer	Chief of Cardiology—London Health Sciences Center	• Biotronik • Boston Scientific • Medtronic*	None	None	None	None	None	None
Glenn N. Levine	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Professor of Medicine—Baylor College of Medicine Director—Cardiac Care Unit—Michael E. DeBakey Medical Center	None	None	None	None	None	None	• Defendant, Catheterization Laboratory Procedure, 2016 • Defendant, Out of hospital death, 2016
Gurusher S. Panjrath	Content Reviewer—ACC Heart Failure and Transplant Council	Director Heart Failure and Mechanical Support Program—George Washington University	• Amgen Inc.*	None	None	None	• BEAT HF‡ • ENDEAVOUR‡	None	None
James P. Daubert	Official Reviewer—AHA	Duke University Medical Center	• Biosense Webster • Boston Scientific • CardioFocus • Gilead • Heart Metabolics • Medtronic* • St. Jude Medical • Zoll	None	None	• ARCA biopharma • Biosense Webster* • Boston Scientific* • Gilead* • Gilead (DSMB) • Medtronic* • NHLBI* • NHLBI (DSMB) • Northwestern University • St. Jude Medical (DSMB) • VytronUS (DSMB)	• Biosense* • Biotronik* • Boston Scientific* • Gilead Scienes, Inc.* • Medtronic* • St. Jude Medical*	• ACC	None
James Tisdale	Content Reviewer—ACC EP Council	Professor—College of Pharmacy Purdue University—Adjunct Professor—School of Medicine Indiana University	None	None	None	• AHA* • HRS* • Indiana Clinical Translational Sciences Institute/ Strategic Research Initiative*	• ACC† • AHA† • AZCert† • QT drugs list, credible meds. org†	None	• Plaintiff, Drug-induced torsades de pointes, 2017*
John L. Sapp	Official Reviewer—HRS	Interim Head—Division of Cardiology QEII Health Sciences Centre—Professor of Medicine—Dalhousie University	• Biosense Webster* • Medtronic • St. Jude	None	None	• Biosense Webster* • Canadian Institute of Health Research* • DSMB† • Phillips healthcare* • St. Jude Medical*	• ARTESiA‡ • Medtronic‡ • Optisure Registry‡ • St. Jude‡	None	None
Joseph Edward Marine	Official Reviewer—ACC	Associate Professor of Medicine—Johns Hopkins University School of Medicine	None	None	None	None	• UpToDate	None	None

(*Continued*)

Appendix 2. Continued

Reviewer	Representation	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Salary	Expert Witness
Kathleen T. Hickey	Official Reviewer—AHA	Professor of Nursing—Columbia University Medical Center	None	None	None	None	None	None	None
Kenneth A. Ellenbogen	Content Reviewer	Chief of Cardiology—Virginia Commonwealth University Medical Center	• AHA • AtriCure* • Biosense Webster* • Biotronik* • Boston Science* • Capricor • HRS • Janssen • Medtronic* • Pfizer* • Sentra heart • St. Jude Medical*	None	None	• AtriCure* • Biosense Webster* • Boston Science* • Daiichi Sankyo • Medtronic* • Medtronic (DSMB)* • NIH* • Pfizer*	• Biosense Webster* • Boston Science* • Circulation† • Heart Rhythm† • JACC† • Medtronic* • PACE† • Sanofi Aventis	None	None
Kim K. Birtcher	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	University of Houston—College of Pharmacology	• Jones and Bartlett Learning	None	None	None	• Accreditation Council for Clinical Lipidology	• University of Houston College of Pharmacology* • Walgreens*	None
Kristen B. Campbell	Content Reviewer	Duke University Hospital	None	None	None	None	None	None	None
Kristen K. Patton	Content Reviewer	Professor of Medicine—University of Washington	None	None	None	None	• ABIM • ACGME† • AHA† • FDA • HRS†	None	None
L. Brent Mitchell	Content Reviewer	Professor—Department of Cardiac Sciences—Libin Cardiovascular Institute of Alberta—University of Calgary—Alberta Health Services	• Boehringer Ingelheim* • Forest Pharmaceuticals • Guidnat Canada* • Medtronic Canada* • Medtronic Inc* • Merck • Pfizer* • Servier Canada*	None	None	• Boston Scientific*	• ARTESIA‡ • Health Protection Branch, Government of Canada	None	None
Martin Borggrefe	Content Reviewer	I Medizinische KlinikKlinikum Mannheim GmbHUniversitätsklinikum	• Bayer Health Care • Boehringer Ingelheim • Impulse Dynamics • Sanofi Aventis • St. Jude Medical	None	None	• German Centre for Cardiovascular Research*	None	None	None
Mathew D. Hutchinson	Official Reviewer—HRS	Professor of Medicine—University of Arizona College of Medicine—Tucson	• St. Jude Medical	None	None	None	None	None	None
Matthew W. Martinez	Content Reviewer—Sports and Exercise EP Council	Lehigh Valley Health Network	None	None	None	None	None	None	None
Melissa R. Robinson	Content Reviewer	Director—Complex Ablation Program—University of Washington	• Medtronic* • Abbott* • Boston Scientific*	None	None	None	None	None	None
Michael J. Silka	Content Reviewer	Children's Hospital Los Angeles	None	None	None	None	None	None	• Defendant, ICD implantation, 2017
Miguel A. Quinones	Content Reviewer	Methodist DeBakey Heart and Vascular Center	None	None	None	None	• Houston Methodist Hospital*	None	None
Mitchell T. Saltzberg	Organizational Reviewer—HFSA	Jefferson Medical College—Christiana Care Health System	None	None	• Nephroceuticals* • Stem Cell Theranostics*	None	None	None	None
N.A. Mark Estes III	Content Reviewer	Professor of Medicine—Tufts University School of Medicine	• Boston Scientific* • Medtronic* • St. Jude Medical*	None	None	• Boston Scientific* • International Board of Heart Rhythm Examiners† • Medtronic* • St. Jude Medical*	None	None	None
Norma M. Keller	Official Reviewer—ACC	New York University Medical Center	None	None	None	None	None	None	None

(*Continued*)

Appendix 2. Continued

Reviewer	Representation	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Salary	Expert Witness
Peter Leong-Sit	Content Reviewer—HRS	Associate Professor of Medicine—Western University—London Health Sciences Centre	• Medtronic Canada	• Bayer Healthcare Pharmaceuticals • Biosense Webster • Johnson and Johnson	None	None	None	• Bayer Healthcare Pharmaceuticals*	None
Rachel J. Lampert	Content Reviewer	Yale University School of Medicine—Section of Cardiology	• Medtronic*	None	None	• Boston Scientific* • GE Medical* • Medtronic, Inc.* • St. Jude Medical*	None	None	None
Sami Viskin	Content Reviewer	Tel Aviv Medical Center—Department of Cardiology	• Boston Scientific European Strategy Advisory Board	None	None	None	None	None	None
Samuel S. Gidding	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Dupont Hospital for Children—Nemours Cardiac Center	• Familial Hypercholesterolemia Foundation† • Regenxbio	None	None	• Familial Hypercholestrolemia Foundation† • NIH Grants*	• Cardiology Division Head†	None	None
Silvia G. Priori	Content Reviewer	Professore Ordinario di Cardiologia—Università di Pavia—Direttore Scientifico—Istituti Clinici Scientifici Maugeri—Pavia, Italia	• Ambry Genetics • Boston Scientific • Medtronic • Medtronic, Inc.	None	• Audentes Therapeutics Inc*	• Gilead Sciences*	• HRS • GS-US-372–1234‡	None	None
Susan Strong	Official Reviewer—AHA	Sabin Middle School	None	None	None	None	None	None	None
Win-Kuang Shen	Content Reviewer	Professor of Medicine—Consultant—Mayo Clinic Arizona, Phoenix Campus	None	None	None	None	None	None	None
Zachary D. Goldberger	Official Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines Lead Reviewer	Assistant Professor of Medicine—Division of Cardiology—Harborview Medical Center—University of Washington School of Medicine	• RubiconMD	None	None	None	None	None	None

This table represents the relationships of reviewers with industry and other entities that were disclosed at the time of peer review, including those not deemed to be relevant to this document, at the time this document was under review. The table does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of ≥5% of the voting stock or share of the business entity, or ownership of ≥$5000 of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. Relationships that exist with no financial benefit are also included for the purpose of transparency. Relationships in this table are modest unless otherwise noted. Names are listed in alphabetical order within each category of review. Please refer to http://www.acc.org/guidelines/about-guidelines-and-clinical-documents/relationships-with-industry-policy for definitions of disclosure categories or additional information about the ACC/AHA Disclosure Policy for Writing Committees.

*Significant relationship.

†No financial benefit.

‡This disclosure was entered under the Clinical Trial Enroller category in the ACC's disclosure system. To appear in this category, the author acknowledges that there is no direct or institutional relationship with the trial sponsor as defined in the ACC/AHA Disclosure Policy for Writing Committees.

ACC indicates American College of Cardiology; AHA, American Heart Association; CPVT, catecholaminergic polymorphic ventricular tachycardia; DSMB, data safety monitoring board; EP, electrophysiology; HFSA, Heart Failure Society of America; HRS, Heart Rhythm Society; ICD, implantable cardioverter-defibrillator; NHLBI, National Heart, Lung, and Blood Institute; and NIH, National Institutes of Health.