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

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

Developed in Collaboration With the Heart Failure Society of America

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This document was approved by the American College of Cardiology Clinical Policy Approval Committee, the American Heart Association Science Advisory and Coordinating Committee, and the Heart Rhythm Society in September 2017, and the American Heart Association Executive Committee in October 2017.

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Al-Khatib SM, et al.
2017 VA/SCD Guideline: Executive Summary

Kay GN, Matlock DD, Myerburg RJ, Page RL. 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 Foundation/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2017;●●:●●●●–●●●●.

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Table of Contents

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 (1, 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 (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 (4) and other methodology articles (5-8).

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 http://www.acc.org/guidelines/about-guidelines-and-clinical-documents/relationships-with-industry-policy. 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 http://jaccjacc.acc.org/Clinical_Document/2017_VASCD_Guideline_Comprehensive_Relationships.pdf, as is the comprehensive disclosure information for the Task Force http://www.acc.org/guidelines/about-guidelines-and-clinical-documents/guidelines-and-documents-task-forces.

Evidence Review and Evidence Review Committees

When developing recommendations, the writing committee uses evidence-based methodologies that are based on all available data (4-7). 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 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) (4, 6, 8).

The reader is encouraged to consult the full-text guideline (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

References

1. Committee on Standards for Developing Trustworthy Clinical Practice Guidelines, Institute of Medicine (U.S.). Clinical Practice Guidelines We Can Trust. Washington, DC: National Academies Press; 2011.
2. Committee on Standards for Systematic Reviews of Comparative Effectiveness Research, Institute of Medicine (U.S.). Finding What Works in Health Care: Standards for Systematic Reviews. Washington, DC: National Academies Press; 2011.
3. Anderson JL, Heidenreich PA, Barnett PG, et al. ACC/AHA statement on cost/value methodology in clinical practice guidelines and performance measures: a report of the American College of Cardiology/American Heart Association Task Force on Performance Measures and Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:2304-22.
4. ACCF/AHA Task Force on Practice Guidelines. Methodology Manual and Policies From the ACCF/AHA Task Force on Practice Guidelines. American College of Cardiology and American Heart Association, 2010. Available at: http://assets.cardiosource.com/Methodology_Manual_for_ACC_AHA_Writing_Committees.pdf and http://professional.heart.org/idc/groups/ahamah-public/@wcm/@sop/documents/downloadable/ucm_319826.pdf. Accessed October 1, 2017.
5. Arnett DK, Goodman RA, Halperin JL, et al. AHA/ACC/HHS strategies to enhance application of clinical practice guidelines in patients with cardiovascular disease and comorbid conditions: from the American Heart Association, American College of Cardiology, and U.S. Department of Health and Human Services. J Am Coll Cardiol. 2014;64:1851-6.
6. Halperin JL, Levine GN, Al-Khatib SM, et al. Further evolution of the ACC/AHA clinical practice guideline recommendation classification system: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2016;67:1572-4.
7. Jacobs AK, Anderson JL Halperin JL. The evolution and future of ACC/AHA clinical practice guidelines: a 30-year journey: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;64:1373-84.
8. Jacobs AK, Kushner FG, Ettinger SM, et al. ACCF/AHA clinical practice guideline methodology summit report: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2013;61:213-65.
9. Al-Khatib SM, Stevenson WG, Ackerman MJ, et al. 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2017. In Press.

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

CLASS (STRENGTH) OF RECOMMENDATION	
CLASS I (STRONG)	**Benefit >>> Risk**
Suggested phrases for writing recommendations: ▪ Is recommended ▪ Is indicated/useful/effective/beneficial ▪ Should be performed/administered/other ▪ Comparative-Effectiveness Phrases†: ○ Treatment/strategy A is recommended/indicated in preference to treatment B ○ Treatment A should be chosen over treatment B	
CLASS IIa (MODERATE)	**Benefit >> Risk**
Suggested phrases for writing recommendations: ▪ Is reasonable ▪ Can be useful/effective/beneficial ▪ Comparative-Effectiveness Phrases†: ○ Treatment/strategy A is probably recommended/indicated in preference to treatment B ○ It is reasonable to choose treatment A over treatment B	
CLASS IIb (WEAK)	**Benefit ≥ Risk**
Suggested phrases for writing recommendations: ▪ May/might be reasonable ▪ May/might be considered ▪ Usefulness/effectiveness is unknown/unclear/uncertain or not well established	
CLASS III: No Benefit (MODERATE) *(Generally, LOE A or B use only)*	**Benefit = Risk**
Suggested phrases for writing recommendations: ▪ Is not recommended ▪ Is not indicated/useful/effective/beneficial ▪ Should not be performed/administered/other	
CLASS III: Harm (STRONG)	**Risk > Benefit**
Suggested phrases for writing recommendations: ▪ Potentially harmful ▪ Causes harm ▪ Associated with excess morbidity/mortality ▪ Should not be performed/administered/other	

LEVEL (QUALITY) OF EVIDENCE‡	
LEVEL A	
▪ High-quality evidence‡ from more than 1 RCT ▪ Meta-analyses of high-quality RCTs ▪ One or more RCTs corroborated by high-quality registry studies	
LEVEL B-R	**(Randomized)**
▪ Moderate-quality evidence‡ from 1 or more RCTs ▪ Meta-analyses of moderate-quality RCTs	
LEVEL B-NR	**(Nonrandomized)**
▪ Moderate-quality evidence‡ from 1 or more well-designed, well-executed nonrandomized studies, observational studies, or registry studies ▪ Meta-analyses of such studies	
LEVEL C-LD	**(Limited Data)**
▪ Randomized or nonrandomized observational or registry studies with limitations of design or execution ▪ Meta-analyses of such studies ▪ Physiological or mechanistic studies in human subjects	
LEVEL C-EO	**(Expert Opinion)**
Consensus of expert opinion based on clinical experience	

COR and LOE are determined independently (any COR may be paired with any LOE).

A recommendation with LOE C does not imply that the recommendation is weak. Many important clinical questions addressed in guidelines do not lend themselves to clinical trials. Although RCTs are unavailable, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

* The outcome or result of the intervention should be specified (an improved clinical outcome or increased diagnostic accuracy or incremental prognostic information).

† For comparative-effectiveness recommendations (COR I and IIa; LOE A and B only), studies that support the use of comparator verbs should involve direct comparisons of the treatments or strategies being evaluated.

‡ The method of assessing quality is evolving, including the application of standardized, widely used, and preferably validated evidence grading tools; and for systematic reviews, the incorporation of an Evidence Review Committee.

COR indicates Class of Recommendation; EO, expert opinion; LD, limited data; LOE, Level of Evidence; NR, nonrandomized; R, randomized; and RCT, randomized controlled trial.

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 http://jaccjacc.acc.org/Clinical_Document/2017_VASCD_Data_Supplement.pdf 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 (1).

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?	7.9.1.3
2	What is the impact of ICD implantation for primary prevention in older patients and patients with significant comorbidities?	9.3

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

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 (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 (2). Available quality of life (QoL) data were deemed to be insufficient to support specific recommendations in this guideline.

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 U.S. GDP data from 2012 and were obtained from WHO-CHOICE Cost-Effectiveness Thresholds (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. (2).

1.2. Organization of the Writing Committee

The writing committee consisted of cardiac electrophysiologists (including those specialized in pediatrics), general 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 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" (4). It also supersedes some sections of the "ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities" (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" (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" (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.

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 (8). An AHA science advisory discusses the use of wearable cardioverter-defibrillators (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.

Table 4. Associated Guidelines and Statements

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

References

1. Kusumoto FM, Bailey KR, Chaouki AS, et al. Systematic review for the 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American

College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2017. In Press.

2. Anderson JL, Heidenreich PA, Barnett PG, et al. ACC/AHA statement on cost/value methodology in clinical practice guidelines and performance measures: a report of the American College of Cardiology/American Heart Association Task Force on Performance Measures and Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:2304-22.
3. World Health Organization. CHOosing Interventions that are Cost Effective (WHO-CHOICE): cost-effectiveness thresholds. Available at: http://www.who.int/choice/en/. Accessed March 26, 2013.
4. Zipes DP, Camm AJ, Borggrefe M, et al. ACC/AHA/ESC 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology/American Heart Association Task Force and the European Society of Cardiology Committee for Practice Guidelines (Writing Committee to Develop Guidelines for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death). Developed in collaboration with the European Heart Rhythm Association and the Heart Rhythm Society. J Am Coll Cardiol. 2006;48:e247-346.
5. Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the ACC/AHA/NASPE 2002 Guideline Update for Implantation of Cardiac Pacemakers and Antiarrhythmia Devices). Developed in collaboration with the American Association for Thoracic Surgery and Society of Thoracic Surgeons. J Am Coll Cardiol. 2008;51:e1-62.
6. Gersh BJ, Maron BJ, Bonow RO, et al. 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. Developed in collaboration with the American Association for Thoracic Surgery, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2011;58:e212-60.
7. Buxton AE, Calkins H, Callans DJ, et al. ACC/AHA/HRS 2006 key data elements and definitions for electrophysiological studies and procedures: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Data Standards (ACC/AHA/HRS Writing Committee to Develop Data Standards on Electrophysiology). J Am Coll Cardiol. 2006;48:2360-96.
8. Wilkoff BL, Fauchier L, Stiles MK, et al. 2015 HRS/EHRA/APHRS/SOLAECE expert consensus statement on optimal implantable cardioverter-defibrillator programming and testing. J Arrhythm. 2016;32:1-28.
9. Piccini JP Sr, Allen LA, Kudenchuk PJ, et al. Wearable cardioverter-defibrillator therapy for the prevention of sudden cardiac death: a science advisory from the American Heart Association. Circulation. 2016;133:1715-27.
10. Shen WK, Sheldon RS, Benditt DG, et al. 2017 ACC/AHA/HRS guideline for the evaluation and management of patients with syncope: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2017;70:e39-110.
11. Yancy CW, Jessup M, Bozkurt B, et al. 2017 ACC/AHA/HFSA focused update of the 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. J Am Coll Cardiol. 2017;70:776-803.
12. Yancy CW, Jessup M, Bozkurt B, et al. 2016 ACC/AHA/HFSA focused update on new pharmacological therapy for heart failure: an update of the 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. J Am Coll Cardiol. 2016;68:1476-88.
13. Yancy CW, Jessup M, Bozkurt B, et al. 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2013;62:e147-239.
14. Nishimura RA, Otto CM, Bonow RO, et al. 2017 AHA/ACC focused update of the 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2017;70:252-89.
15. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:e57-185.
16. Page RL, Joglar JA, Caldwell MA, et al. 2015 ACC/AHA/HRS Guideline for the management of adult patients with supraventricular tachycardia: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2016;67:e27-115.

17. Priori SG, Blomstrom-Lundqvist C, Mazzanti A, et al. 2015 ESC guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology. G Ital Cardiol (Rome). 2016;17:108-70.
18. Perkins GD, Jacobs IG, Nadkarni VM, et al. Cardiac arrest and cardiopulmonary resuscitation outcome reports: update of the Utstein Resuscitation Registry Templates for Out-of-Hospital Cardiac Arrest: a statement for healthcare professionals from a task force of the International Liaison Committee on Resuscitation (American Heart Association, European Resuscitation Council, Australian and New Zealand Council on Resuscitation, Heart and Stroke Foundation of Canada, InterAmerican Heart Foundation, Resuscitation Council of Southern Africa, Resuscitation Council of Asia);and the American Heart Association Emergency Cardiovascular Care Committee and the Council on Cardiopulmonary, Critical Care, Perioperative and Resuscitation. Circulation. 2015;132:1286-300.
19. January CT, Wann LS, Alpert JS, et al. 2014 AHA/ACC/HRS guideline for the management of patients with atrial fibrillation: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2014;64:e1-76.
20. Amsterdam EA, Wenger NK, Brindis RG, et al. 2014 AHA/ACC guideline for the management of patients with non-ST-elevation acute coronary syndromes: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;64:e139-228.
21. Andrus B, Lacaille D. 2013 ACC/AHA guideline on the assessment of cardiovascular risk. J Am Coll Cardiol. 2014;63:2886.
22. O'Gara PT, Kushner FG, Ascheim DD, et al. 2013 ACCF/AHA guideline for the management of ST-elevation myocardial infarction: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2013;61:e78-140.
23. Steg PG, James SK, Atar D, et al. ESC guidelines for the management of acute myocardial infarction in patients presenting with ST-segment elevation. Eur Heart J. 2012;33:2569-619.
24. Tracy CM, Epstein AE, Darbar D, et al. 2012 ACCF/AHA/HRS focused update of the 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2012;60:1297-313.
25. Hillis LD, Smith PK, Anderson JL, et al. 2011 ACCF/AHA guideline for coronary artery bypass graft surgery: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. Developed in collaboration with the American Association for Thoracic Surgery, Society of Cardiovascular Anesthesiologists, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2011;58:e123-210.
26. Levine GN, Bates ER, Blankenship JC, et al. 2011 ACCF/AHA/SCAI guideline for percutaneous coronary intervention: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Society for Cardiovascular Angiography and Interventions. Catheter Cardiovasc Interv. 2013;82:E266-355.
27. Smith SC Jr, Benjamin EJ, Bonow RO, et al. AHA/ACCF secondary prevention and risk reduction therapy for patients with coronary and other atherosclerotic vascular disease: 2011 update: a guideline from the American Heart Association and American College of Cardiology Foundation. J Am Coll Cardiol. 2011;58:2432-46.
28. Institute of Medicine. Committee on the Treatment of Cardiac Arrest: Current status and future directions: strategies to improve cardiac arrest survival: a time to act. Washington, DC: National Academic Press; 2015.
29. Link MS, Myerburg RJ, Estes NA 3rd. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 12: emergency action plans, resuscitation, cardiopulmonary resuscitation, and automated external defibrillators: a scientific statement from the American Heart Association and American College of Cardiology. Circulation. 2015;132:e334-8.
30. Pedersen CT, Kay GN, Kalman J, et al. EHRA/HRS/APHRS expert consensus on ventricular arrhythmias. Heart Rhythm. 2014;11:e166-96.
31. Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Heart Rhythm. 2014;11:e102-65.
32. Kusumoto FM, Calkins H, Boehmer J, et al. HRS/ACC/AHA expert consensus statement on the use of implantable cardioverter-defibrillator therapy in patients who are not included or not well represented in clinical trials. J Am Coll Cardiol. 2014;64:1143-77.
33. Birnie DH, Sauer WH, Bogun F, et al. HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. Heart Rhythm. 2014;11:1305-23.

34. Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. Heart Rhythm. 2013;10:1932-63.

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

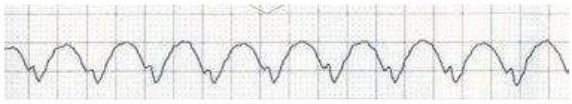
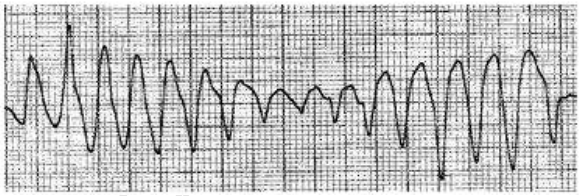
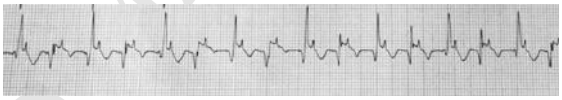
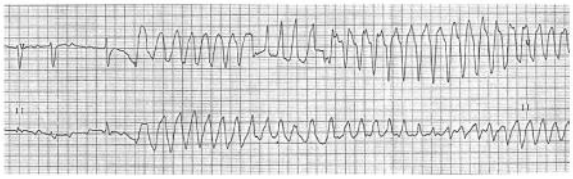
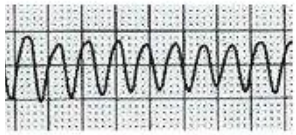
2. Epidemiology

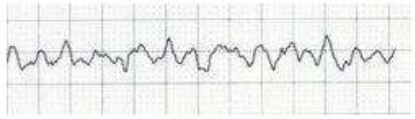
2.1. General Concepts

Table 5

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 (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.

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

Term	Definition or Description
Ventricular tachycardia (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 (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 (2)	A regular VA ≈300 bpm (cycle length: 200 ms) with a sinusoidal, monomorphic appearance; no isoelectric interval between successive QRS complexes. 

Ventricular fibrillation (2)	Rapid, grossly irregular electrical activity with marked variability in electrocardiographic waveform, ventricular rate usually >300 bpm (cycle length: <200 ms). 
Sudden cardiac arrest (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 (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 (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 (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 (2)	ICD placement in a patient with prior SCA, sustained VT, or syncope caused by VA.
Structural heart disease*	TThis term encompasses IHD, all types of cardiomyopathy, valvular heart disease, and adult congenital heart disease.
Cardiac channelopathy (4)	Arrhythmogenic disease due to a genetic abnormality that results in dysfunction of a cardiac ion channel (e.g., 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.

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 (5) on 12-lead ECGs, longer term monitoring shows PVCs in about 50% of all people with or without heart disease (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 (7, 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 (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 (10). In the same population, nonsustained ventricular tachycardia (NSVT) was independently associated with increased risk of death and other cardiovascular adverse outcomes, including stroke (11). An association of PVCs with increased risk of stroke was also seen in the ARIC population (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 (e.g., 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 (12, 13). In CAST (Cardiac Arrhythmia Suppression Trials), treatment of patients with post-myocardial infarction (MI) who took antiarrhythmic medications (e.g., flecainide, encainide, moricizine) increased the risk of death despite suppression of VA (14, 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 (e.g., quinidine, flecainide) increases the risk of death (15, 16). Likewise, in patients with a reduced LVEF class I, sodium channel–blocking medications and d-sotalol increase the risk of death (16, 17). Beta blockers, nondihydropyridines calcium channel blockers, and some antiarrhythmic medications may relieve symptoms of palpitations (18).

PVCs that occur during an exercise test are associated with a higher risk of death (19). In 1 study, PVCs that occur during recovery are a stronger predictor of death than PVCs occurring only during exercise (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 (21, 22). Complex PVCs may not represent a benign finding in endurance athletes. An electrophysiological study may be needed to assess patients' arrhythmogenic risk (22). Very frequent PVCs, >10,000 to 20,000 a day, can be associated with depressed LV function in some patients that is reversible with control of the PVCs, and has been referred to as PVC-induced cardiomyopathy (23, 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 (25, 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) (27). Of all out-of-hospital cardiac arrests, >50% will have significant coronary artery lesions on acute coronary angiography (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 (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 (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 (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 (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 (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) (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 (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 (35, 36), medication-induced long QT syndrome (36), or they may be idiopathic (37, 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 (1, 39), with at least 25% being first symptomatic cardiac events (1, 40, 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 (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 (41, 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 (44). An additional 209,000 in-hospital cardiac arrests occur annually (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 (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, 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 (46, 47). Survival to hospital discharge after in-hospital cardiac arrests is estimated to be 24% (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 (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 (40).

2.2.2. Population Subgroups and Risk Prediction

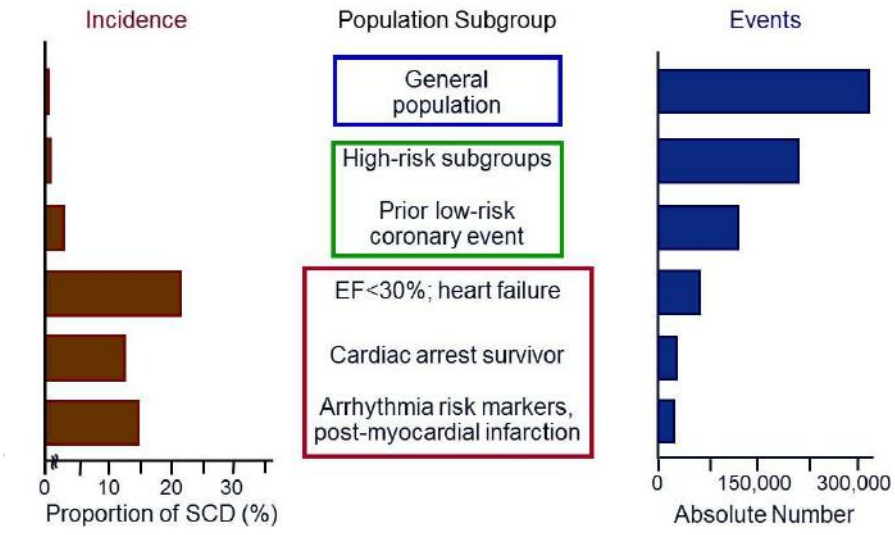
SRisk prediction for SCA and SCD is complex. Risk analysis is divided into 2 general categories: population risk prediction and individual risk prediction (41, 50). Conventional epidemiological markers provide insight into probabilities for the development of ischemic heart disease within a general class 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 (41). However, an analysis of 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 (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 (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 (52), with various forms of cardiomyopathy associated with myocardial fibrosis and LV hypertrophy increasing (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 (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 (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) (43).

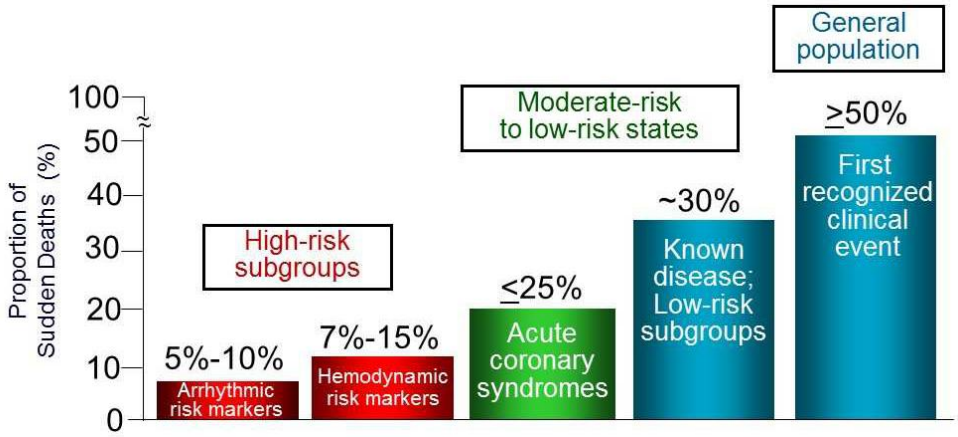
Despite the small progress that has been made in risk prediction of SCA and SCD, the greatest challenge is to identify the relatively small, high-risk subgroups concealed within the large general population who have no identified disease but are at risk of SCA as their first cardiac event (Figure 1)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

Figure 1A. SCD Incidence and Total Events (1)



EF indicates ejection fraction; and SCD, sudden cardiac death.

Figure 1B. SCD and Clinical Subsets (1)



SCD indicates sudden cardiac death.

References

1. Myerburg RJ, Junttila MJ. Sudden cardiac death caused by coronary heart disease. Circulation. 2012;125:1043-52.
2. Buxton AE, Calkins H, Callans DJ, et al. ACC/AHA/HRS 2006 key data elements and definitions for electrophysiological studies and procedures: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Data Standards. J Am Coll Cardiol. 2006;48:2360-96.
3. Sapp JL, Wells GA, Parkash R, et al. Ventricular tachycardia ablation versus escalation of antiarrhythmic drugs. N Engl J Med. 2016;375:111-21.
4. Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. Heart Rhythm. 2013;10:1932-63.
5. Hiss RG, Lamb LE. Electrocardiographic findings in 122,043 individuals. Circulation. 1962;25:947-61.
6. Brodsky M, Wu D, Denes P, et al. Arrhythmias documented by 24 hour continuous electrocardiographic monitoring in 50 male medical students without apparent heart disease. Am J Cardiol. 1977;39:390-5.
7. Massing MW, Simpson RJ Jr, Rautaharju PM, et al. Usefulness of ventricular premature complexes to predict coronary heart disease events and mortality (from the Atherosclerosis Risk In Communities cohort). Am J Cardiol. 2006;98:1609-12.
8. Ofoma U, He F, Shaffer ML, et al. Premature cardiac contractions and risk of incident ischemic stroke. J Am Heart Assoc. 2012;1:e002519.
9. Ataklte F, Erqou S, Laukkanen J, et al. Meta-analysis of ventricular premature complexes and their relation to cardiac mortality in general populations. Am J Cardiol. 2013;112:1263-70.
10. Lin CY, Chang SL, Lin YJ, et al. Long-term outcome of multiform premature ventricular complexes in structurally normal heart. Int J Cardiol. 2015;180:80-5.
11. Lin CY, Chang SL, Chung FP, et al. Long-term outcome of non-sustained ventricular tachycardia in structurally normal hearts. PloS One. 2016;11:e0160181.
12. Ruberman W, Weinblatt E, Goldberg JD, et al. Ventricular premature complexes and sudden death after myocardial infarction. Circulation. 1981;64:297-305.
13. Ruberman W, Weinblatt E, Goldberg JD, et al. Ventricular premature beats and mortality after myocardial infarction. N Engl J Med. 1977;297:750-7.
14. The Cardiac Arrhythmia Suppression Trial II Investigators. Effect of the antiarrhythmic agent moricizine on survival after myocardial infarction. The Cardiac Arrhythmia Suppression Trial II Investigators. N Engl J Med. 1992;327:227-33.
15. Echt DS, Liebson PR, Mitchell LB, et al. Mortality and morbidity in patients receiving encainide, flecainide, or placebo. The Cardiac Arrhythmia Suppression Trial. N Engl J Med. 1991;324:781-8.
16. Morganroth J Goin JE. Quinidine-related mortality in the short-to-medium-term treatment of ventricular arrhythmias. A meta-analysis. Circulation. 1991;84:1977-83.

17. Waldo AL, Camm AJ, deRuyter H, et al. Effect of d-sotalol on mortality in patients with left ventricular dysfunction after recent and remote myocardial infarction. The SWORD Investigators. Survival With Oral d-Sotalol. Lancet. 1996;348:7-12.
18. Ling Z, Liu Z, Su L, et al. Radiofrequency ablation versus antiarrhythmic medication for treatment of ventricular premature beats from the right ventricular outflow tract: prospective randomized study. Circ Arrhythm Electrophysiol. 2014;7:237-43.
19. Jouven X, Zureik M, Desnos M, et al. Long-term outcome in asymptomatic men with exercise-induced premature ventricular depolarizations. N Engl J Med. 2000;343:826-33.
20. Frolkis JP, Pothier CE, Blackstone EH, et al. Frequent ventricular ectopy after exercise as a predictor of death. N Engl J Med. 2003;348:781-90.
21. Biffi A, Pelliccia A, Verdile L, et al. Long-term clinical significance of frequent and complex ventricular tachyarrhythmias in trained athletes. J Am Coll Cardiol. 2002;40:446-52.
22. Heidbüchel H, Hoogsteen J, Fagard R, et al. High prevalence of right ventricular involvementin endurance athletes with ventricular arrhythmiasRole of an electrophysiologic study in risk stratification. Eur Heart J. 2003;24:1473-80.
23. Kanei Y, Friedman M, Ogawa N, et al. Frequent premature ventricular complexes originating from the right ventricular outflow tract are associated with left ventricular dysfunction. Ann Noninvasive Electrocardiol. 2008;13:81-5.
24. Lee GK, Klarich KW, Grogan M, et al. Premature ventricular contraction-induced cardiomyopathy: a treatable condition. Circ Arrhythm Electrophysiol. 2012;5:229-36.
25. Viskin S, Rosso R, Rogowski O, et al. The "short-coupled" variant of right ventricular outflow ventricular tachycardia: a not-so-benign form of benign ventricular tachycardia? J Cardiovasc Electrophysiol. 2005;16:912-6.
26. Noda T, Shimizu W, Taguchi A, et al. Malignant entity of idiopathic ventricular fibrillation and polymorphic ventricular tachycardia initiated by premature extrasystoles originating from the right ventricular outflow tract. J Am Coll Cardiol. 2005;46:1288-94.
27. Dumas F, Cariou A, Manzo-Silberman S, et al. Immediate percutaneous coronary intervention is associated with better survival after out-of-hospital cardiac arrest: insights from the PROCAT (Parisian Region Out of hospital Cardiac ArresT) registry. Circ Cardiovasc Interv. 2010;3:200-7.
28. Gupta S, Pressman GS, Figueredo VM. Incidence of, predictors for, and mortality associated with malignant ventricular arrhythmias in non-ST elevation myocardial infarction patients. Coron. Artery Dis. 2010;21:460-5.
29. Terkelsen CJ, Sorensen JT, Kaltoft AK, et al. Prevalence and significance of accelerated idioventricular rhythm in patients with ST-elevation myocardial infarction treated with primary percutaneous coronary intervention. Am J Cardiol. 2009;104:1641-6.
30. Al-Khatib SM, Granger CB, Huang Y, et al. Sustained ventricular arrhythmias among patients with acute coronary syndromes with no ST-segment elevation: incidence, predictors, and outcomes. Circulation. 2002;106:309-12.
31. Jabbari R, Engstrom T, Glinge C, et al. Incidence and risk factors of ventricular fibrillation before primary angioplasty in patients with first ST-elevation myocardial infarction: a nationwide study in Denmark. J Am Heart Assoc. 2015;4:e001399.
32. Mehta RH, Starr AZ, Lopes RD, et al. Incidence of and outcomes associated with ventricular tachycardia or fibrillation in patients undergoing primary percutaneous coronary intervention. JAMA. 2009;301:1779-89.
33. Volpi A, Cavalli A, Franzosi MG, et al. One-year prognosis of primary ventricular fibrillation complicating acute myocardial infarction. The GISSI (Gruppo Italiano per lo Studio della Streptochinasi nell'Infarto miocardico) investigators. Am J Cardiol. 1989;63:1174-8.
34. El-Sherif N, Smith RA, Evans K. Canine ventricular arrhythmias in the late myocardial infarction period. 8. Epicardial mapping of reentrant circuits. Circ Res. 1981;49:255-65.
35. Ackerman MJ, Priori SG, Willems S, et al. HRS/EHRA expert consensus statement on the state of genetic testing for the channelopathies and cardiomyopathies: this document was developed as a partnership between the Heart Rhythm Society (HRS) and the European Heart Rhythm Association (EHRA). Europace. 2011;13:1077-109.
36. Nannenberg EA, Sijbrands EJ, Dijksman LM, et al. Mortality of inherited arrhythmia syndromes: insight into their natural history. Circ Cardiovasc Genet. 2012;5:183-9.
37. Haïssaguerre M, Shah DC, Jais P, et al. Role of Purkinje conducting system in triggering of idiopathic ventricular fibrillation. Lancet. 2002;359:677-8.
38. Haïssaguerre M, Shoda M, Jais P, et al. Mapping and ablation of idiopathic ventricular fibrillation. Circulation. 2002;106:962-7.

39. Goldberger JJ, Buxton AE, Cain M, et al. Risk stratification for arrhythmic sudden cardiac death: identifying the roadblocks. Circulation. 2011;123:2423-30.
40. Fishman GI, Chugh SS, DiMarco JP, et al. Sudden cardiac death prediction and prevention: report from a National Heart, Lung, and Blood Institute and Heart Rhythm Society Workshop. Circulation. 2010;122:2335-48.
41. Myerburg RJ. Sudden cardiac death: exploring the limits of our knowledge. J Cardiovasc Electrophysiol. 2001;12:369-81.
42. Kong MH, Fonarow GC, Peterson ED, et al. Systematic review of the incidence of sudden cardiac death in the United States. J Am Coll Cardiol. 2011;57:794-801.
43. Mozaffarian D, Benjamin EJ, Go AS, et al. Heart disease and stroke statistics—2016 update: a report from the American Heart Association. Circulation. 2016;133:e38-360.
44. Benjamin EJ, Blaha MJ, Chiuve SE, et al. Heart disease and stroke statistics—2017 update: a report from the American Heart Association. Circulation. 2017;135:e146-603.
45. Merchant RM, Yang L, Becker LB, et al. Incidence of treated cardiac arrest in hospitalized patients in the United States. Crit Care Med. 2011;39:2401-6.
46. Institute of Medicine. Committee on the Treatment of Cardiac Arrest: current status and future directions: strategies to improve cardiac arrest survival: a time to act. Washington, DC: National Academic Press; 2015.
47. Jollis JG, Granger CB. Improving care of out-of-hospital cardiac arrest: next steps. Circulation. 2016;134:2040-2.
48. Daya MR, Schmicker R, May SH, et al. Current burden of cardiac arrest in the United States: report from the Resuscitation Outcomes Consortium. Paper commissioned by the Committee on the Treatment of Cardiac Arrest: Current Status and Future Directions. 2015.
49. Myerburg RJ, Halperin H, Egan DA, et al. Pulseless electric activity: definition, causes, mechanisms, management, and research priorities for the next decade: report from a National Heart, Lung, and Blood Institute workshop. Circulation. 2013;128:2532-41.
50. Myerburg RJ, Goldberger JJ. Sudden cardiac arrest risk assessment: Population science and the individual risk mandate. JAMA Cardiol. 2017;2:689-94.
51. Bogle BM, Ning H, Mehrotra S, et al. Lifetime risk for sudden cardiac death in the community. J Am Heart Assoc. 2016;5:e002398.
52. Junttila MJ, Hookana E, Kaikkonen KS, et al. Temporal trends in the clinical and pathological characteristics of victims of sudden cardiac death in the absence of previously identified heart disease. Circ Arrhythm Electrophysiol. 2016;9:e003723.
53. Hookana E, Junttila MJ, Puurunen VP, et al. Causes of nonischemic sudden cardiac death in the current era. Heart Rhythm. 2011;8:1570-5.
54. Wong MK, Morrison LJ, Qiu F, et al. Trends in short- and long-term survival among out-of-hospital cardiac arrest patients alive at hospital arrival. Circulation. 2014;130:1883-90.

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 (1-4).**

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

Table 6

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 (e.g., 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
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.

References
1. Middlekauff HR, Stevenson WG, Stevenson LW, et al. Syncope in advanced heart failure: high risk of sudden death regardless of origin of syncope. J Am Coll Cardiol. 1993;21:110-6.
2. Myerburg RJ, Castellanos A. Cardiac arrest and sudden cardiac death - Chapter 39. In: Mann DL, Zipes DP, Libby P, et al, eds. Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine. 10th ed. Oxford, UK: Elsevier; 2015:821-60.
3. Shen WK, Sheldon RS, Benditt DG, et al. 2017 ACC/AHA/HRS guideline for the evaluation and management of patients with syncope: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2017;70:e39-110.
4. Soteriades ES, Evans JC, Larson MG, et al. Incidence and prognosis of syncope. N Engl J Med. 2002;347:878-85.

3.2. Noninvasive Evaluation

3.2.1. 12-lead ECG and Exercise Testing

Recommendations for 12-lead ECG and Exercise Testing References 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 (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 (4, 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 (6).**

References
1. Brugada P, Brugada J, Mont L, et al. A new approach to the differential diagnosis of a regular tachycardia with a wide QRS complex. Circulation. 1991;83:1649-59.
2. Wellens HJ, Bar FW, Lie KI. The value of the electrocardiogram in the differential diagnosis of a tachycardia with a widened QRS complex. Am J Med. 1978;64:27-33.
3. Steinman RT, Herrera C, Schuger CD, et al. Wide QRS tachycardia in the conscious adult. Ventricular tachycardia is the most frequent cause. JAMA. 1989;261:1013-6.
4. Elhendy A, Chandrasekaran K, Gersh BJ, et al. Functional and prognostic significance of exercise-induced ventricular arrhythmias in patients with suspected coronary artery disease. Am J Cardiol. 2002;90:95-100.
5. Grady TA, Chiu AC, Snader CE, et al. Prognostic significance of exercise-induced left bundle-branch block. JAMA. 1998;279:153-6.
6. Perez-Rodon J, Martinez-Alday J, Baron-Esquivias G, et al. Prognostic value of the electrocardiogram in patients with syncope: data from the group for syncope study in the emergency room (GESINUR). Heart Rhythm. 2014;11:2035-44.

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 (1-4).**

References

1. Barrett PM, Komatireddy R, Haaser S, et al. Comparison of 24-hour Holter monitoring with 14-day novel adhesive patch electrocardiographic monitoring. Am J Med. 2014;127:95-7.
2. de Asmundis C, Conte G, Sieira J, et al. Comparison of the patient-activated event recording system vs. traditional 24 h Holter electrocardiography in individuals with paroxysmal palpitations or dizziness. Europace. 2014;16:1231-5.
3. Linzer M, Pritchett EL, Pontinen M, et al. Incremental diagnostic yield of loop electrocardiographic recorders in unexplained syncope. Am J Cardiol. 1990;66:214-9.
4. Turakhia MP, Hoang DD, Zimetbaum P, et al. Diagnostic utility of a novel leadless arrhythmia monitoring device. Am J Cardiol. 2013;112:520-4.

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 (1-4).**

References

1. Bloch Thomsen PE, Jons C, Raatikainen MJ, et al. Long-term recording of cardiac arrhythmias with an implantable cardiac monitor in patients with reduced ejection fraction after acute myocardial infarction: the Cardiac Arrhythmias and Risk Stratification After Acute Myocardial Infarction (CARISMA) study. Circulation. 2010;122:1258-64.
2. Krahn AD, Klein GJ, Yee R, et al. Use of an extended monitoring strategy in patients with problematic syncope. Reveal Investigators. Circulation. 1999;99:406-10.
3. Solbiati M, Costantino G, Casazza G, et al. Implantable loop recorder versus conventional diagnostic workup for unexplained recurrent syncope. Cochrane Database Syst Rev. 2016;4:CD011637.
4. Volosin K, Stadler RW, Wyszynski R, et al. Tachycardia detection performance of implantable loop recorders: results from a large 'real-life' patient cohort and patients with induced ventricular arrhythmias. Europace. 2013;15:1215-22.

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 (1, 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.**

References

1. Solomon SD, Zelenkofske S, McMurray JJ, et al. Sudden death in patients with myocardial infarction and left ventricular dysfunction, heart failure, or both. N Engl J Med. 2005;352:2581-8.
2. Gula LJ, Klein GJ, Hellkamp AS, et al. Ejection fraction assessment and survival: an analysis of the Sudden Cardiac Death in Heart Failure Trial (SCD-HeFT). Am Heart J. 2008;156:1196-200.

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 (1-4).**

References

1. Ahmad T, Fiuzat M, Neely B, et al. Biomarkers of myocardial stress and fibrosis as predictors of mode of death in patients with chronic heart failure. JACC Heart Fail. 2014;2:260-8.
2. Scott PA, Barry J, Roberts PR, et al. Brain natriuretic peptide for the prediction of sudden cardiac death and ventricular arrhythmias: a meta-analysis. Eur J Heart Fail. 2009;11:958-66.
3. Levine YC, Rosenberg MA, Mittleman M, et al. B-type natriuretic peptide is a major predictor of ventricular tachyarrhythmias. Heart Rhythm. 2014;11:1109-16.
4. Berger R, Huelsman M, Strecker K, et al. B-type natriuretic peptide predicts sudden death in patients with chronic heart failure. Circulation. 2002;105:2392-7.

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 (1-7).**
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 (8-11).**
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 (12-16).**

References

1. Buxton AE, Lee KL, DiCarlo L, et al. Electrophysiologic testing to identify patients with coronary artery disease who are at risk for sudden death. Multicenter Unsustained Tachycardia Trial Investigators. N Engl J Med 2000;342:1937-45.
2. Buxton AE, Lee KL, Hafley GE, et al. Relation of ejection fraction and inducible ventricular tachycardia to mode of death in patients with coronary artery disease: an analysis of patients enrolled in the multicenter unsustained tachycardia trial. Circulation. 2002;106:2466-72.
3. Costantini O, Hohnloser SH, Kirk MM, et al. The ABCD (Alternans Before Cardioverter Defibrillator) Trial: strategies using T-wave alternans to improve efficiency of sudden cardiac death prevention. J Am Coll Cardiol. 2009;53:471-9.
4. Bourke JP, Richards DA, Ross DL, et al. Routine programmed electrical stimulation in survivors of acute myocardial infarction for prediction of spontaneous ventricular tachyarrhythmias during follow-up: results, optimal stimulation protocol and cost-effective screening. J Am Coll Cardiol. 1991;18:780-8.
5. Bailey JJ, Berson AS, Handelsman H, et al. Utility of current risk stratification tests for predicting major arrhythmic events after myocardial infarction. J Am Coll Cardiol. 2001;38:1902-11.
6. Schmitt C, Barthel P, Ndrepepa G, et al. Value of programmed ventricular stimulation for prophylactic internal cardioverter-defibrillator implantation in postinfarction patients preselected by noninvasive risk stratifiers. J Am Coll Cardiol. 2001;37:1901-7.
7. Hilfiker G, Schoenenberger AW, Erne P, et al. Utility of electrophysiological studies to predict arrhythmic events. World J Cardiol. 2015;7:344-50.
8. Bardy GH, Lee KL, Mark DB, et al. Amiodarone or an implantable cardioverter-defibrillator for congestive heart failure. N Engl J Med. 2005;352:225-37.

9. Buxton AE, Lee KL, Fisher JD, et al. A randomized study of the prevention of sudden death in patients with coronary artery disease. Multicenter Unsustained Tachycardia Trial Investigators. N Engl J Med. 1999;341:1882-90.
10. Moss AJ, Hall WJ, Cannom DS, et al. Improved survival with an implanted defibrillator in patients with coronary disease at high risk for ventricular arrhythmia. Multicenter Automatic Defibrillator Implantation Trial Investigators. N Engl J Med. 1996;335:1933-40.
11. Moss AJ, Zareba W, Hall WJ, et al. Prophylactic implantation of a defibrillator in patients with myocardial infarction and reduced ejection fraction. N Engl J Med. 2002;346:877-83.
12. Bhandari AK, Shapiro WA, Morady F, et al. Electrophysiologic testing in patients with the long QT syndrome. Circulation. 1985;71:63-71.
13. Giustetto C, Di MF, Wolpert C, et al. Short QT syndrome: clinical findings and diagnostic-therapeutic implications. Eur. Heart J. 2006;27:2440-7.
14. Giustetto C, Schimpf R, Mazzanti A, et al. Long-term follow-up of patients with short QT syndrome. J Am Coll Cardiol. 2011;58:587-95.
15. Mahida S, Derval N, Sacher F, et al. Role of electrophysiological studies in predicting risk of ventricular arrhythmia in early repolarization syndrome. J Am Coll Cardiol. 2015;65:151-9.
16. Priori SG, Napolitano C, Memmi M, et al. Clinical and molecular characterization of patients with catecholaminergic polymorphic ventricular tachycardia. Circulation. 2002;106:69-74.

4. Therapies for Treatment or Prevention of VA

4.1. Medication Therapy

Table 7

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 upto 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-	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, slowsVT below programmed ICD detection rate, increases defibrillation threshold Other: Corneal microdeposits, thyroid abnormalities, ataxia, nausea, emesis, constipation, photosensitivity, skin discoloration, ataxia,

400 mg daily; reduce dose to 200 mg daily if possible					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
Diltiazem (IV) IV: 5-10 mg Qq 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	VT, VF	I_{Na}	No marked effect on most intervals; QTc can slightly	Initial $t_{1/2}$ 7–30 min; terminal 90–120 min.	Cardiac : Bradycardia, hemodynamic collapse, AVB, sinus arrest

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			shorten	Prolonged in HF, liver disease, shock, severe renal disease Metab: H Excr: U	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)	VT, PVC (in the absence of	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	Cardiac: HF, AVB, drug-induced Brugada syndrome

PO: Immediate release 150–300 mg q 8 h Extended release 225–425 mg q 12 h	structural heart disease)			metabolizers 2–10 h; poor metabolizers 10–32 h. Metab: H Excr: U	Other: Dizziness, fatigue, nausea, diarrhea, xerostomia, tremor, blurred vision
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 gGluconate 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	$_{1/2}$: 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	VT (specifically RVOT, verapamil-sensitive	$I_{Ca\text{-}L}$	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,

Sustained release PO: 240–480 mg/d	idiopathic LVT)				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.

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.

Modified from Shleifer JW, et al (1).

Reference

1. Schleifer JW, Sorajja D, Shen WK. Advances in the pharmacologic treatment of ventricular arrhythmias. Expert Opin Pharmacother. 2015;16:2637-51.

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 (1-8).**

References

1. Yancy CW, Jessup M, Bozkurt B, et al. 2016 ACC/AHA/HFSA Focused update on new pharmacological therapy for heart failure: an update of the 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. J Am Coll Cardiol. 2016;68:1476-88.
2. Committees for Cardiac insufficiency Bisoprolol Study II. The cardiac insufficiency Bisoprolol Study II (CIBIS-II): a randomised trial. Lancet. 1999;353:9-13.
3. Cohn JN, Johnson G, Ziesche S, et al. A comparison of enalapril with hydralazine-isosorbide dinitrate in the treatment of chronic congestive heart failure. N Engl J Med. 1991;325:303-10.
4. Packer M, Bristow MR, Cohn JN, et al. The effect of carvedilol on morbidity and mortality in patients with chronic heart failure. U.S. Carvedilol Heart Failure Study Group. N Engl J Med. 1996;334:1349-55.
5. Dargie HJ. Effect of carvedilol on outcome after myocardial infarction in patients with left-ventricular dysfunction: the CAPRICORN randomised trial. Lancet. 2001;357:1385-90.
6. Pitt B, Remme W, Zannad F, et al. Eplerenone, a selective aldosterone blocker, in patients with left ventricular dysfunction after myocardial infarction. N Engl J Med. 2003;348:1309-21.
7. Al Chekakie MO. Traditional heart failure medications and sudden cardiac death prevention: a review. J Cardiovasc Pharmacol Ther. 2013;18:412-26.
8. Pfeffer MA, McMurray JJ, Velazquez EJ, et al. Valsartan, captopril, or both in myocardial infarction complicated by heart failure, left ventricular dysfunction, or both. N Engl J Med. 2003;349:1893-906.

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 (1-4).**
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.**

References

1. Cook JR, Rizo-Patron C, Curtis AB, et al. Effect of surgical revascularization in patients with coronary artery disease and ventricular tachycardia or fibrillation in the Antiarrhythmics Versus Implantable Defibrillators (AVID) Registry. Am Heart J. 2002;143:821-6.
2. Velazquez EJ, Lee KL, Jones RH, et al. Coronary-artery bypass surgery in patients with ischemic cardiomyopathy. N Engl J Med. 2016;374:1511-20.
3. Every NR, Fahrenbruch CE, Hallstrom AP, et al. Influence of coronary bypass surgery on subsequent outcome of patients resuscitated from out of hospital cardiac arrest. J Am Coll Cardiol. 1992;19:1435-9.
4. Dumas F, Bougouin W, Geri G, et al. Emergency PCI in post-cardiac arrest patients without ST-segment elevation pattern: insights from the PROCAT II registry. JACC Cardiovasc Interv. 2016;9:1011-8.

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 (1-7).**

References

1. Anter E, Hutchinson MD, Deo R, et al. Surgical ablation of refractory ventricular tachycardia in patients with nonischemic cardiomyopathy. Circ Arrhythm Electrophysiol. 2011;4:494-500.
2. Bhavani SS, Tchou P, Saliba W, et al. Surgical options for refractory ventricular tachycardia. J Card Surg. 2007;22:533-4.
3. Choi EK, Nagashima K, Lin KY, et al. Surgical cryoablation for ventricular tachyarrhythmia arising from the left ventricular outflow tract region. Heart Rhythm. 2015;12:1128-36.
4. Kumar S, Barbhaiya CR, Sobieszczyk P, et al. Role of alternative interventional procedures when endo- and epicardial catheter ablation attempts for ventricular arrhythmias fail. Circ Arrhythm Electrophysiol. 2015;8:606-15.
5. Mulloy DP, Bhamidipati CM, Stone ML, et al. Cryoablation during left ventricular assist device implantation reduces postoperative ventricular tachyarrhythmias. J Thorac Cardiovasc Surg. 2013;145:1207-13.
6. Patel M, Rojas F, Shabari FR, et al. Safety and feasibility of open chest epicardial mapping and ablation of ventricular tachycardia during the period of left ventricular assist device implantation. J Cardiovasc Electrophysiol. 2016;27:95-101.
7. Sartipy U, Albage A, Straat E, et al. Surgery for ventricular tachycardia in patients undergoing left ventricular reconstruction by the Dor procedure. Ann Thorac Surg. 2006;81:65-71.

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 (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 (2-4).**

References

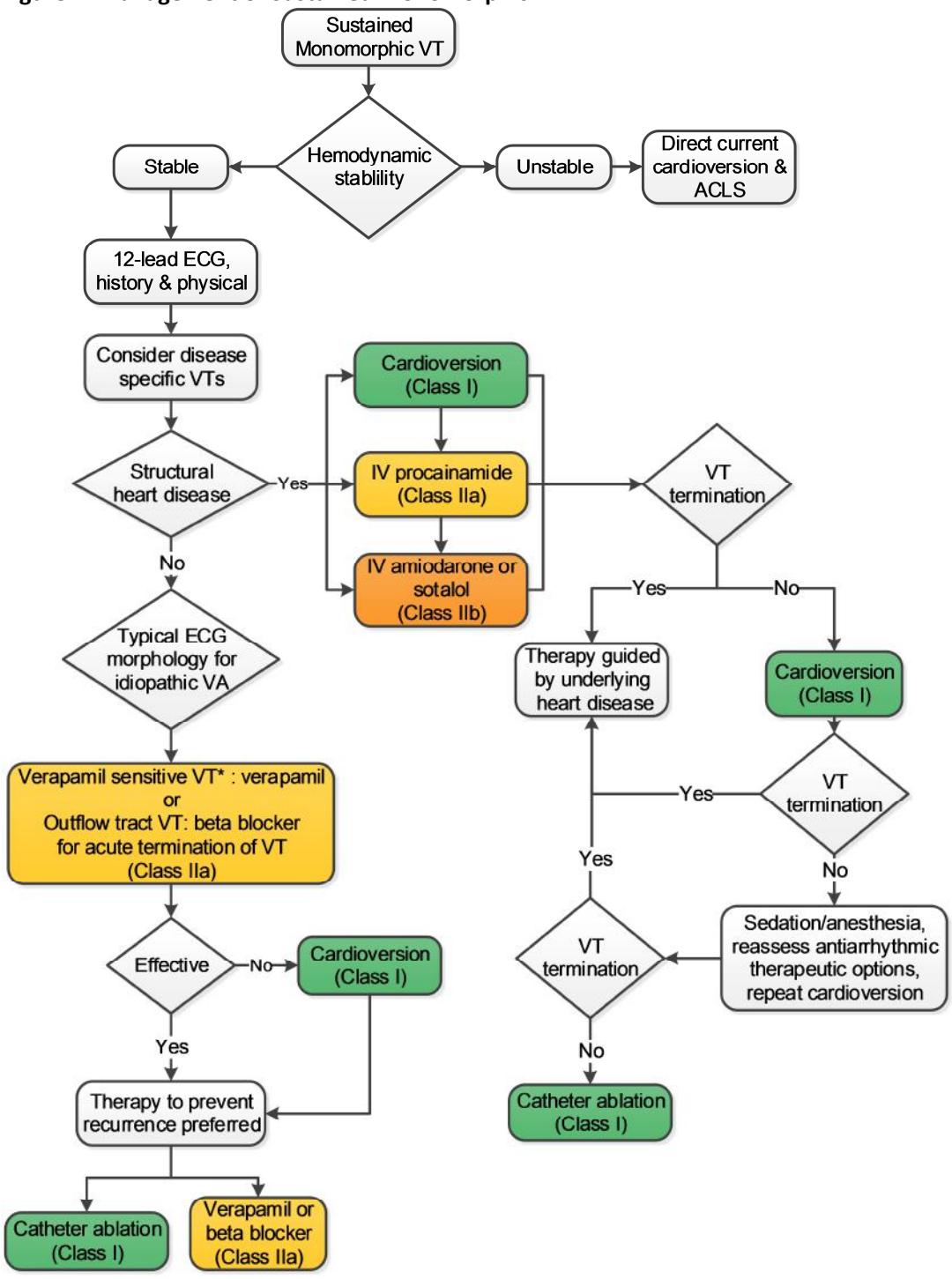
1. Krittayaphong R, Bhuripanyo K, Punlee K, et al. Effect of atenolol on symptomatic ventricular arrhythmia without structural heart disease: a randomized placebo-controlled study. Am Heart J. 2002;144:e10.
2. Vaseghi M, Barwad P, Malavassi Corrales FJ, et al. Cardiac sympathetic denervation for refractory ventricular arrhythmias. J Am Coll Cardiol. 2017;69:3070-80.
3. Vaseghi M, Gima J, Kanaan C, et al. Cardiac sympathetic denervation in patients with refractory ventricular arrhythmias or electrical storm: intermediate and long-term follow-up. Heart Rhythm. 2014;11:360-6.
4. Schwartz PJ, Motolese M, Pollavini G. Prevention of sudden cardiac death after a first myocardial infarction by pharmacologic or surgical antiadrenergic interventions. J Cardiovasc Electrophysiol. 1992;3:2-16.

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 (1-3).
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 (1, 4-6).
I	A	3. Patients presenting with VA with hemodynamic instability should undergo direct current cardioversion (1-3).
I	B-NR	4. In patients with polymorphic VT or VF with ST-elevation MI, angiography with emergency revascularization is recommended (7-10).
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 (11-13).
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 (1, 4, 5, 14, 15).
IIa	B-R	8. In patients with polymorphic VT due to myocardial ischemia, intravenous beta blockers can be useful (16, 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 (17, 18).
IIb	A	10. In patients in cardiac arrest, administration of epinephrine (1 mg every 3 to 5 minutes) during CPR may be reasonable (1, 19-24).
IIb	B-R	11. In patients with hemodynamically stable VT, administration of intravenous amiodarone or sotalol may be considered to attempt to terminate VT (5, 13, 25, 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 (19, 21).
III: No Benefit	A	13. In patients with refractory VF not related to torsades de pointes, administration of intravenous magnesium is not beneficial (27, 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 (16, 29).
III: Harm	C-LD	15. In patients with a wide QRS complex tachycardia of unknown origin, calcium channel blockers (e.g., verapamil and diltiazem) are potentially harmful (30, 31).

Figure 2

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.

References

1. Link MS, Berkow LC, Kudenchuk PJ, et al. Part 7: adult advanced cardiovascular life support: 2015 American Heart Association guidelines update for cardiopulmonary resuscitation and emergency cardiovascular care. Circulation. 2015;132(suppl 2):S444-64.
2. Stiell IG, Wells GA, Field B, et al. Advanced cardiac life support in out-of-hospital cardiac arrest. N Engl J Med. 2004;351:647-56.
3. Sasson C, Rogers MA, Dahl J, et al. Predictors of survival from out-of-hospital cardiac arrest: a systematic review and meta-analysis. Circ Cardiovasc Qual Outcomes. 2010;3:63-81.
4. Dorian P, Cass D, Schwartz B, et al. Amiodarone as compared with lidocaine for shock-resistant ventricular fibrillation. N Engl J Med. 2002;346:884-90.
5. Kudenchuk PJ, Brown SP, Daya M, et al. Amiodarone, lidocaine, or placebo in out-of-hospital cardiac arrest. N Engl J Med. 2016;374:1711-22.
6. Kudenchuk PJ, Cobb LA, Copass MK, et al. Amiodarone for resuscitation after out-of-hospital cardiac arrest due to ventricular fibrillation. N Engl J Med. 1999;341:871-8.
7. Spaulding CM, Joly LM, Rosenberg A, et al. Immediate coronary angiography in survivors of out-of-hospital cardiac arrest. N Engl J Med. 1997;336:1629-33.
8. Cronier P, Vignon P, Bouferrache K, et al. Impact of routine percutaneous coronary intervention after out-of-hospital cardiac arrest due to ventricular fibrillation. Crit Care. 2011;15:R122.
9. Zanuttini D, Armellini I, Nucifora G, et al. Impact of emergency coronary angiography on in-hospital outcome of unconscious survivors after out-of-hospital cardiac arrest. Am J Cardiol. 2012;110:1723-8.
10. Dumas F, Bougouin W, Geri G, et al. Emergency PCI in post-cardiac arrest patients without ST-segment elevation pattern: insights from the PROCAT II registry. JACC Cardiovasc Interv. 2016;9:1011-8.
11. Gorgels AP, van den Dool A, Hofs A, et al. Comparison of procainamide and lidocaine in terminating sustained monomorphic ventricular tachycardia. Am J Cardiol. 1996;78:43-6.
12. Markel DT, Gold LS, Allen J, et al. Procainamide and survival in ventricular fibrillation out-of-hospital cardiac arrest. Acad Emerg Med. 2010;17:617-23.
13. Ortiz M, Martin A, Arribas F, et al. Randomized comparison of intravenous procainamide vs. intravenous amiodarone for the acute treatment of tolerated wide QRS tachycardia: the PROCAMIO study. Eur Heart J. 2017;38:1329-35.
14. Herlitz J, Ekstrom L, Wennerblom B, et al. Lidocaine in out-of-hospital ventricular fibrillation. Does it improve survival? Resuscitation. 1997;33:199-205.
15. Kudenchuk PJ, Newell C, White L, et al. Prophylactic lidocaine for post resuscitation care of patients with out-of-hospital ventricular fibrillation cardiac arrest. Resuscitation. 2013;84:1512-8.
16. Teo KK, Yusuf S, Furberg CD. Effects of prophylactic antiarrhythmic drug therapy in acute myocardial infarction. An overview of results from randomized controlled trials. JAMA. 1993;270:1589-95.
17. Nademanee K, Taylor R, Bailey WE, et al. Treating electrical storm : sympathetic blockade versus advanced cardiac life support-guided therapy. Circulation. 2000;102:742-7.
18. Piccini JP, Hranitzky PM, Kilaru R, et al. Relation of mortality to failure to prescribe beta blockers acutely in patients with sustained ventricular tachycardia and ventricular fibrillation following acute myocardial infarction (from the VALsartan In Acute myocardial iNfarcTion trial [VALIANT] Registry). Am J Cardiol. 2008;102:1427-32.
19. Callaham M, Madsen CD, Barton CW, et al. A randomized clinical trial of high-dose epinephrine and norepinephrine vs standard-dose epinephrine in prehospital cardiac arrest. JAMA. 1992;268:2667-72.
20. Donnino MW, Salciccioli JD, Howell MD, et al. Time to administration of epinephrine and outcome after in-hospital cardiac arrest with non-shockable rhythms: retrospective analysis of large in-hospital data registry. BMJ. 2014;348:g3028.
21. Gueugniaud PY, Mols P, Goldstein P, et al. A comparison of repeated high doses and repeated standard doses of epinephrine for cardiac arrest outside the hospital. European Epinephrine Study Group. N Engl J Med. 1998;339:1595-601.
22. Hagihara A, Hasegawa M, Abe T, et al. Prehospital epinephrine use and survival among patients with out-of-hospital cardiac arrest. JAMA. 2012;307:1161-8.
23. Jacobs IG, Finn JC, Jelinek GA, et al. Effect of adrenaline on survival in out-of-hospital cardiac arrest: a randomised double-blind placebo-controlled trial. Resuscitation. 2011;82:1138-43.

24. Koscik C, Pinawin A, McGovern H, et al. Rapid epinephrine administration improves early outcomes in out-of-hospital cardiac arrest. Resuscitation. 2013;84:915-20.
25. Ho DS, Zecchin RP, Richards DA, et al. Double-blind trial of lignocaine versus sotalol for acute termination of spontaneous sustained ventricular tachycardia. Lancet. 1994;344:18-23.
26. Somberg JC, Bailin SJ, Haffajee CI, et al. Intravenous lidocaine versus intravenous amiodarone (in a new aqueous formulation) for incessant ventricular tachycardia. Am J Cardiol 2002;90:853-9.
27. Hassan TB, Jagger C, Barnett DB. A randomised trial to investigate the efficacy of magnesium sulphate for refractory ventricular fibrillation. Emerg Med J. 2002;19:57-62.
28. Thel MC, Armstrong AL, McNulty SE, et al. Randomised trial of magnesium in in-hospital cardiac arrest. Duke Internal Medicine Housestaff. Lancet. 1997;350:1272-6.
29. Elizari MV, Martinez JM, Belziti C, et al. Morbidity and mortality following early administration of amiodarone in acute myocardial infarction. GEMICA study investigators, GEMA Group, Buenos Aires, Argentina. Grupo de Estudios Multicentricos en Argentina. Eur Heart J. 2000;21:198-205.
30. Belhassen B, Horowitz LN. Use of intravenous verapamil for ventricular tachycardia. Am J Cardiol. 1984;54:1131-3.
31. Buxton AE, Marchlinski FE, Doherty JU, et al. Hazards of intravenous verapamil for sustained ventricular tachycardia. Am J Cardiol. 1987;59:1107-10.

6. Ongoing Management of VA and SCD Risk Related to Specific Disease States

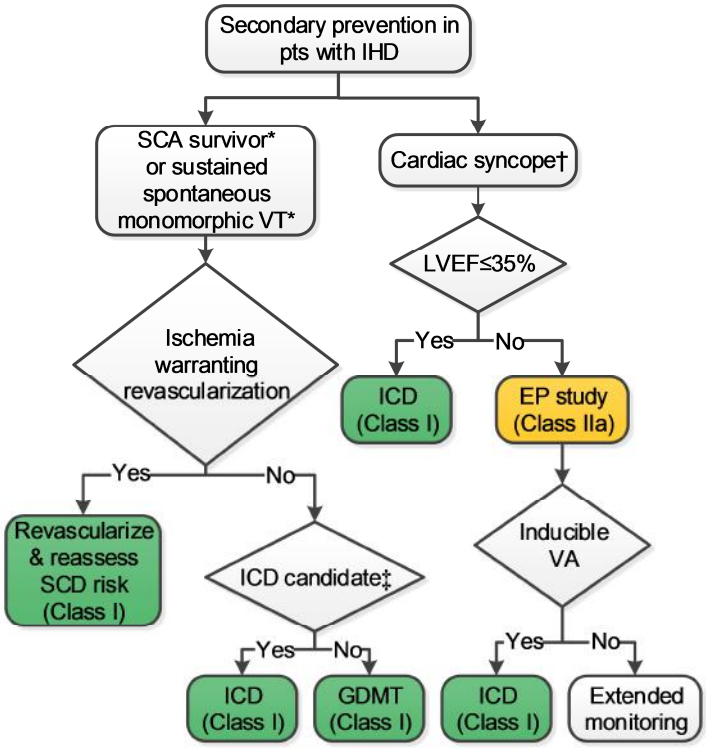
6.1. Ischemic Heart Disease

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** **B-NR**	**1. In patients with ischemic heart disease, who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R) (1-4) or stable VT (LOE: B-NR) (5) not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.**
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 (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 (7).**

Figure 3

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.

References

1. The AVID Investigators. A comparison of antiarrhythmic-drug therapy with implantable defibrillators in patients resuscitated from near-fatal ventricular arrhythmias. N Engl J Med. 1997;337:1576-83.
2. Connolly SJ, Gent M, Roberts RS, et al. Canadian implantable defibrillator study (CIDS) : a randomized trial of the implantable cardioverter defibrillator against amiodarone. Circulation. 2000;101:1297-302.
3. Connolly SJ, Hallstrom AP, Cappato R, et al. Meta-analysis of the implantable cardioverter defibrillator secondary prevention trials. AVID, CASH and CIDS studies. Antiarrhythmics vs Implantable Defibrillator study. Cardiac Arrest Study Hamburg . Canadian Implantable Defibrillator Study. Eur Heart J. 2000;21:2071-8.
4. Kuck KH, Cappato R, Siebels J, et al. Randomized comparison of antiarrhythmic drug therapy with implantable defibrillators in patients resuscitated from cardiac arrest: the Cardiac Arrest Study Hamburg (CASH). Circulation. 2000;102:748-54.
5. Raitt MH, Renfroe EG, Epstein AE, et al. "Stable" ventricular tachycardia is not a benign rhythm : insights from the antiarrhythmics versus implantable defibrillators (AVID) registry. Circulation. 2001;103:244-52.
6. Owens DK, Sanders GD, Heidenreich PA, et al. Effect of risk stratification on cost-effectiveness of the implantable cardioverter defibrillator. Am Heart J. 2002;144:440-8.
7. Bass EB, Elson JJ, Fogoros RN, et al. Long-term prognosis of patients undergoing electrophysiologic studies for syncope of unknown origin. Am J Cardiol. 1988;62:1186-91.

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 (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 (3-6).**
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 (3-6).**

References

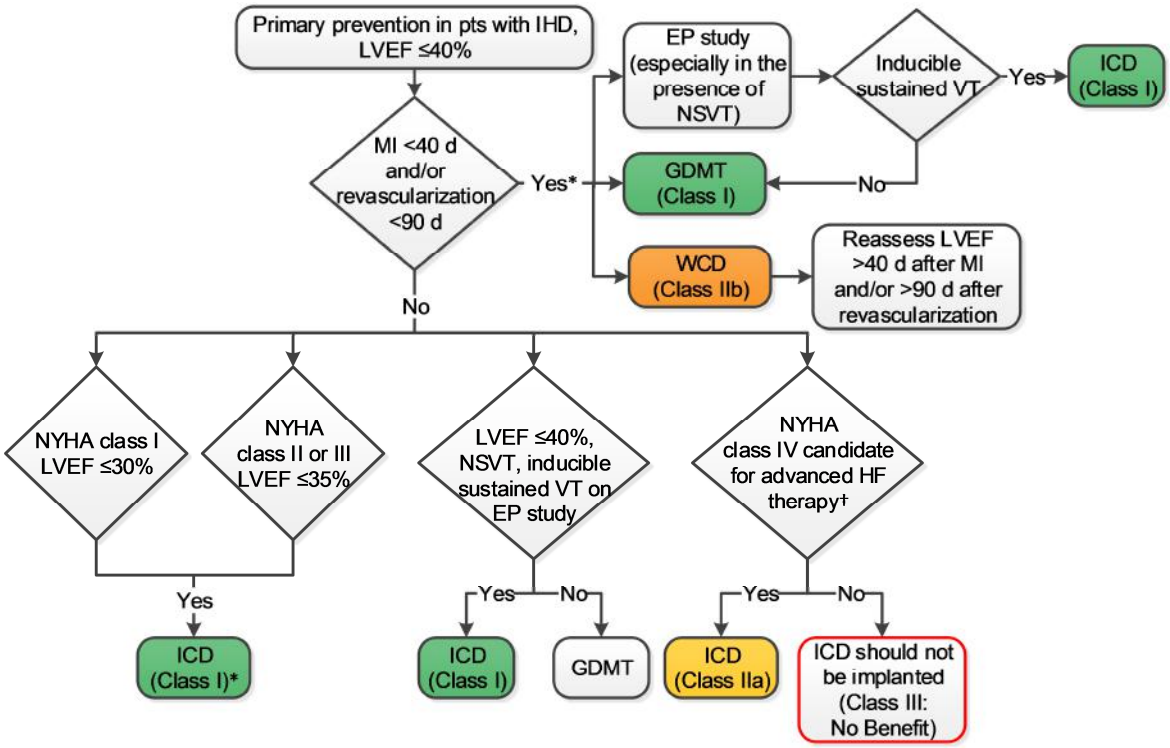
1. Chevalier P, Dacosta A, Defaye P, et al. Arrhythmic cardiac arrest due to isolated coronary artery spasm: long-term outcome of seven resuscitated patients. J Am Coll Cardiol. 1998;31:57-61.
2. Myerburg RJ, Kessler KM, Mallon SM, et al. Life-threatening ventricular arrhythmias in patients with silent myocardial ischemia due to coronary-artery spasm. N Engl J Med. 1992;326:1451-5.
3. Ahn JM, Lee KH, Yoo SY, et al. Prognosis of variant angina manifesting as aborted sudden cardiac death. J Am Coll Cardiol. 2016;68:137-45.
4. Matsue Y, Suzuki M, Nishizaki M, et al. Clinical implications of an implantable cardioverter-defibrillator in patients with vasospastic angina and lethal ventricular arrhythmia. J Am Coll Cardiol. 2012;60:908-13.
5. Takagi Y, Yasuda S, Tsunoda R, et al. Clinical characteristics and long-term prognosis of vasospastic angina patients who survived out-of-hospital cardiac arrest: multicenter registry study of the Japanese Coronary Spasm Association. Circ Arrhythm Electrophysiol. 2011;4:295-302.
6. Meisel SR, Mazur A, Chetboun I, et al. Usefulness of implantable cardioverter-defibrillators in refractory variant angina pectoris complicated by ventricular fibrillation in patients with angiographically normal coronary arteries. Am J Cardiol. 2002;89:1114-6.

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 (1, 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 (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 (4).**
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 (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 (6-9).**
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

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.

References

1. Bardy GH, Lee KL, Mark DB, et al. Amiodarone or an implantable cardioverter-defibrillator for congestive heart failure. N Engl J Med. 2005;352:225-37.
2. Moss AJ, Zareba W, Hall WJ, et al. Prophylactic implantation of a defibrillator in patients with myocardial infarction and reduced ejection fraction. N Engl J Med. 2002;346:877-83.
3. Moss AJ, Hall WJ, Cannom DS, et al. Improved survival with an implanted defibrillator in patients with coronary disease at high risk for ventricular arrhythmia. Multicenter Automatic Defibrillator Implantation Trial Investigators. N Engl J Med. 1996;335:1933-40.
4. Owens DK, Sanders GD, Heidenreich PA, et al. Effect of risk stratification on cost-effectiveness of the implantable cardioverter defibrillator. Am Heart J. 2002;144:440-8.
5. Buxton AE, Lee KL, Fisher JD, et al. A randomized study of the prevention of sudden death in patients with coronary artery disease. Multicenter Unsustained Tachycardia Trial Investigators. N Engl J Med. 1999;341:1882-90.
6. Cantero-Pérez EM, Sobrino-Márquez JM, Grande-Trillo A, et al. Implantable cardioverter defibrillator for primary prevention in patients with severe ventricular dysfunction awaiting heart transplantation. Transplant Proc. 2013;45:3659-61.
7. Frohlich GM, Holzmeister J, Hubler M, et al. Prophylactic implantable cardioverter defibrillator treatment in patients with end-stage heart failure awaiting heart transplantation. Heart. 2013;99:1158-65.

8. Gandjbakhch E, Rovani M, Varnous S, et al. Implantable cardioverter-defibrillators in end-stage heart failure patients listed for heart transplantation: results from a large retrospective registry. Arch Cardiovasc Diseases. 2016;109:476-85.
9. Vakil K, Duval S, Cogswell R, et al. Impact of implantable cardioverter-defibrillators on waitlist mortality among patients awaiting heart transplantation: an UNOS/OPTN analysis. JACC Clin Electrophysiol. 2017;3:33-40.

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

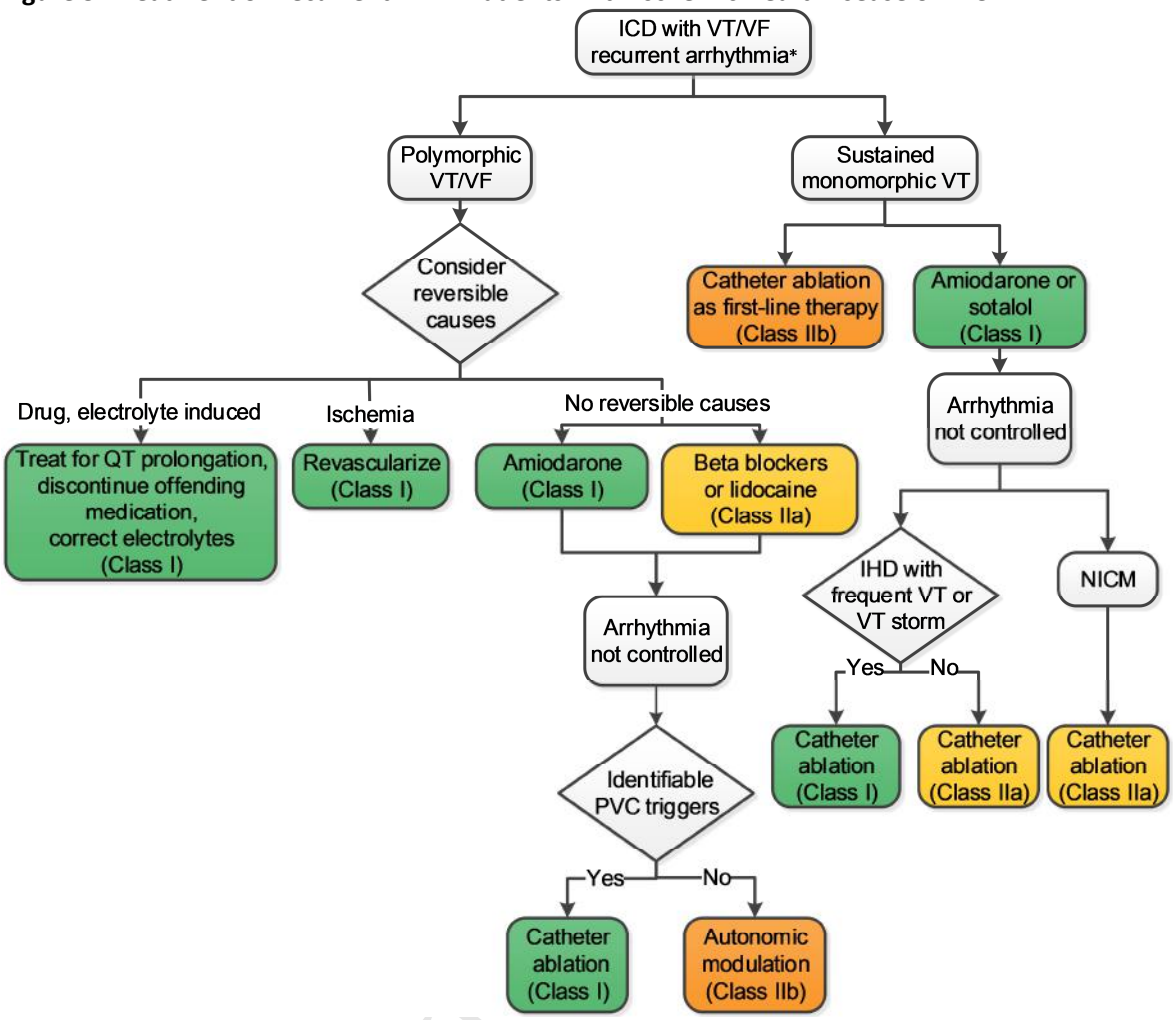
Recommendations for Treatment of Recurrent VA in Patients With Ischemic Heart Disease

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 (1-3).**
I	B-R B-NR	**2. In patients with prior MI and recurrent episodes of symptomatic sustained VT, or who present with VT or VF storm and have failed or are intolerant of amiodarone (LOE: B-R) (4) or other antiarrhythmic medications (LOE: B-NR) (5-9), catheter ablation is recommended (10-12).**
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 (10, 11).**
III: Harm	B-R	**4. In patients with prior MI, class IC antiarrhythmic medications (e.g., flecainide and propafenone) should not be used (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 (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 (15, 16).**

Figure 5

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 (26).
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.

References

1. Connolly SJ, Dorian P, Roberts RS, et al. Comparison of beta-blockers, amiodarone plus beta-blockers, or sotalol for prevention of shocks from implantable cardioverter defibrillators: the OPTIC Study: a randomized trial. JAMA. 2006;295:165-71.
2. Pacifico A, Hohnloser SH, Williams JH, et al. Prevention of implantable-defibrillator shocks by treatment with sotalol. d,l-Sotalol Implantable Cardioverter-Defibrillator Study Group. N Engl J Med. 1999;340:1855-62.
3. Kettering K, Mewis C, Dornberger V, et al. Efficacy of metoprolol and sotalol in the prevention of recurrences of sustained ventricular tachyarrhythmias in patients with an implantable cardioverter defibrillator. Pacing Clin Electrophysiol. 2002;25:1571-6.
4. Sapp JL, Wells GA, Parkash R, et al. Ventricular tachycardia ablation versus escalation of antiarrhythmic drugs. N Engl J Med. 2016;375:111-21.

5. Mallidi J, Nadkarni GN, Berger RD, et al. Meta-analysis of catheter ablation as an adjunct to medical therapy for treatment of ventricular tachycardia in patients with structural heart disease. Heart Rhythm. 2011;8:503-10.
6. Marchlinski FE, Haffajee CI, Beshai JF, et al. Long-term success of irrigated radiofrequency catheter ablation of sustained ventricular tachycardia: post-approval THERMOCOOL VT trial. J Am Coll Cardiol. 2016;67:674-83.
7. Stevenson WG, Wilber DJ, Natale A, et al. Irrigated radiofrequency catheter ablation guided by electroanatomic mapping for recurrent ventricular tachycardia after myocardial infarction: the multicenter thermocool ventricular tachycardia ablation trial. Circulation. 2008;118:2773-82.
8. Tanner H, Hindricks G, Volkmer M, et al. Catheter ablation of recurrent scar-related ventricular tachycardia using electroanatomical mapping and irrigated ablation technology: results of the prospective multicenter Euro-VT-study. J Cardiovasc Electrophysiol. 2010;21:47-53.
9. Tung R, Vaseghi M, Frankel DS, et al. Freedom from recurrent ventricular tachycardia after catheter ablation is associated with improved survival in patients with structural heart disease: an International VT Ablation Center Collaborative Group study. Heart Rhythm. 2015;12:1997-2007.
10. Al-Khatib SM, Daubert JP, Anstrom KJ, et al. Catheter ablation for ventricular tachycardia in patients with an implantable cardioverter defibrillator (CALYPSO) pilot trial. J Cardiovasc Electrophysiol. 2015;26:151-7.
11. Kuck KH, Schaumann A, Eckardt L, et al. Catheter ablation of stable ventricular tachycardia before defibrillator implantation in patients with coronary heart disease (VTACH): a multicentre randomised controlled trial. Lancet. 2010;375:31-40.
12. Sesselberg HW, Moss AJ, McNitt S, et al. Ventricular arrhythmia storms in postinfarction patients with implantable defibrillators for primary prevention indications: a MADIT-II substudy. Heart Rhythm. 2007;4:1395-402.
13. Echt DS, Liebson PR, Mitchell LB, et al. Mortality and morbidity in patients receiving encainide, flecainide, or placebo. The Cardiac Arrhythmia Suppression Trial. N Engl J Med. 1991;324:781-8.
14. Sears SF Jr, Todaro JF, Lewis TS, et al. Examining the psychosocial impact of implantable cardioverter defibrillators: a literature review. Clinical Cardiol. 1999;22:481-9.
15. Steinberg JS, Gaur A, Sciacca R, et al. New-onset sustained ventricular tachycardia after cardiac surgery. Circulation. 1999;99:903-8.
16. Brugada J, Aguinaga L, Mont L, et al. Coronary artery revascularization in patients with sustained ventricular arrhythmias in the chronic phase of a myocardial infarction: effects on the electrophysiologic substrate and outcome. J Am Coll Cardiol. 2001;37:529-33.

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 (1-3).**
IIa	**B-NR**	**2. In patients with suspected NICM, cardiac MRI with late gadolinium enhancement can be useful for assessing risk of SCA/SCD (1-3).**
IIa	**C-EO**	**3. In patients with NICMwho develop conduction disease or LV dysfunction at less than 40 years of age, or who have a family history of NICMor 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 (4, 5).**

References

1. Greulich S, Deluigi CC, Gloekler S, et al. CMR imaging predicts death and other adverse events in suspected cardiac sarcoidosis. JACC Cardiovasc Imaging. 2013;6:501-11.
2. Kuruvilla S, Adenaw N, Katwal AB, et al. Late gadolinium enhancement on cardiac magnetic resonance predicts adverse cardiovascular outcomes in nonischemic cardiomyopathy: a systematic review and meta-analysis. Circ Cardiovasc Imaging. 2014;7:250-8.

3. Piers SR, Tao Q, van Huls van Taxis CFB, et al. Contrast-enhanced MRI-derived scar patterns and associated ventricular tachycardias in nonischemic cardiomyopathy: implications for the ablation strategy. Circ Arrhythm Electrophysiol. 2013;6:875-83.
4. Ackerman MJ, Priori SG, Willems S, et al. HRS/EHRA expert consensus statement on the state of genetic testing for the channelopathies and cardiomyopathies: this document was developed as a partnership between the Heart Rhythm Society (HRS) and the European Heart Rhythm Association (EHRA). Europace. 2011;13:1077-109.
5. Hershberger RE, Morales A, Siegfried JD. Clinical and genetic issues in dilated cardiomyopathy: a review for genetics professionals. Genet Med. 2010;12:655-67.

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) (1-4) or stable VT (LOE: B-NR) (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 (6-11).**
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 (12, 13).**

Figure 6

References

1. The Antiarrhythmics versus Implantable Defibrillators (AVID) Investigators. A comparison of antiarrhythmic-drug therapy with implantable defibrillators in patients resuscitated from near-fatal ventricular arrhythmias. N Engl J Med. 1997;337:1576-83.
2. Connolly SJ, Gent M, Roberts RS, et al. Canadian implantable defibrillator study (CIDS): a randomized trial of the implantable cardioverter defibrillator against amiodarone. Circulation. 2000;101:1297-302.
3. Desai AS, Fang JC, Maisel WH, et al. Implantable defibrillators for the prevention of mortality in patients with nonischemic cardiomyopathy: a meta-analysis of randomized controlled trials. JAMA. 2004;292:2874-9.
4. Kuck KH, Cappato R, Siebels J, et al. Randomized comparison of antiarrhythmic drug therapy with implantable defibrillators in patients resuscitated from cardiac arrest : the Cardiac Arrest Study Hamburg (CASH). Circulation. 2000;102:748-54.
5. Raitt MH, Renfroe EG, Epstein AE, et al. "Stable" ventricular tachycardia is not a benign rhythm : insights from the Antiarrhythmics versus Implantable Defibrillators (AVID) registry. Circulation. 2001;103:244-52.
6. Brilakis ES, Shen WK, Hammill SC, et al. Role of programmed ventricular stimulation and implantable cardioverter defibrillators in patients with idiopathic dilated cardiomyopathy and syncope. Pacing Clin Electrophysiol. 2001;24:1623-30.
7. Fonarow GC, Feliciano Z, Boyle NG, et al. Improved survival in patients with nonischemic advanced heart failure and syncope treated with an implantable cardioverter-defibrillator. Am J Cardiol. 2000;85:981-5.
8. Knight BP, Goyal R, Pelosi F, et al. Outcome of patients with nonischemic dilated cardiomyopathy and unexplained syncope treated with an implantable defibrillator. J Am Coll Cardiol. 1999;33:1964-70.
9. Middlekauff HR, Stevenson WG, Stevenson LW, et al. Syncope in advanced heart failure: high risk of sudden death regardless of origin of syncope. J Am Coll Cardiol. 1993;21:110-6.

10. Olshansky B, Poole JE, Johnson G, et al. Syncope predicts the outcome of cardiomyopathy patients: analysis of the SCD-HeFT study. J Am Coll Cardiol. 2008;51:1277-82.
11. Ruwald MH, Okumura K, Kimura T, et al. Syncope in high-risk cardiomyopathy patients with implantable defibrillators: frequency, risk factors, mechanisms, and association with mortality: results from the multicenter automatic defibrillator implantation trial-reduce inappropriate therapy (MADIT-RIT) study. Circulation. 2014;129:545-52.
12. Piccini JP, Berger JS, O'Connor CM. Amiodarone for the prevention of sudden cardiac death: a meta-analysis of randomized controlled trials. Eur Heart J. 2009;30:1245-53.
13. Claro JC, Candia R, Rada G, et al. Amiodarone versus other pharmacological interventions for prevention of sudden cardiac death. Cochrane Database Syst Rev. 2015;12:CD008093.

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 (1-6).**
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 (7-10).**
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 (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.**

Figure 6

References

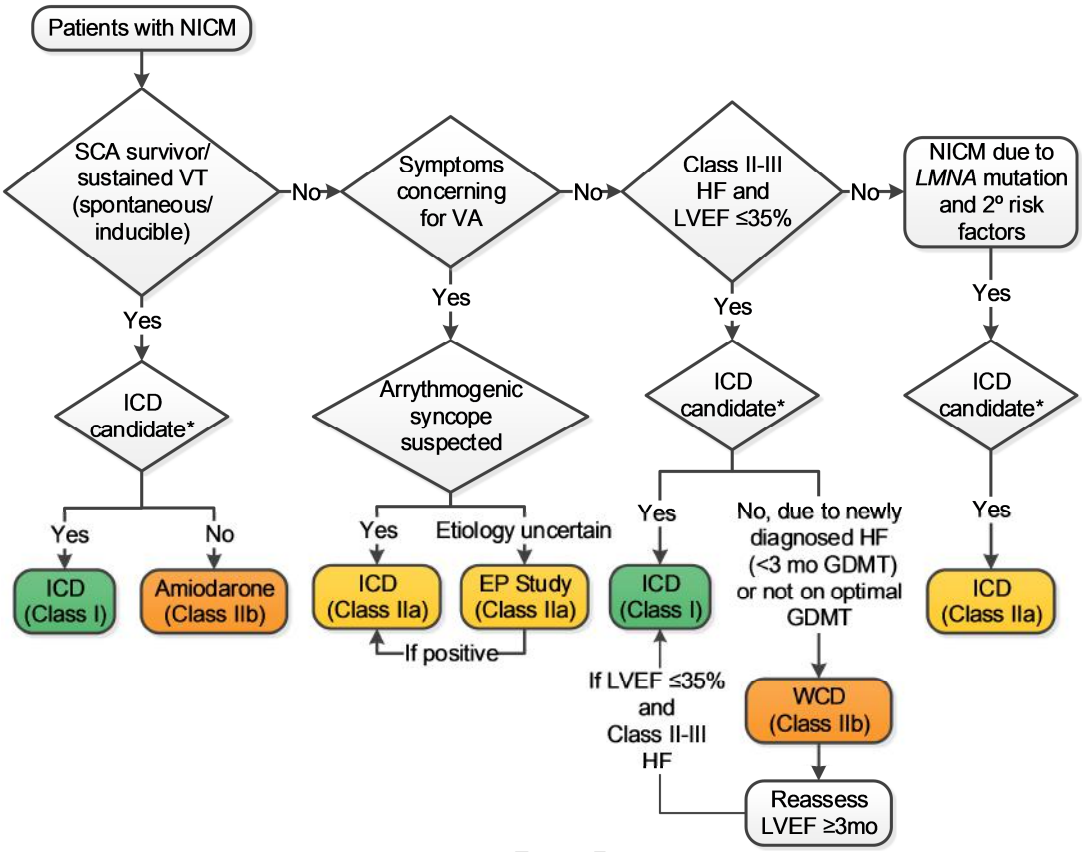
1. Bardy GH, Lee KL, Mark DB, et al. Amiodarone or an implantable cardioverter-defibrillator for congestive heart failure. N Engl J Med. 2005;352:225-37.
2. Bänsch D, Antz M, Boczor S, et al. Primary prevention of sudden cardiac death in idiopathic dilated cardiomyopathy: the Cardiomyopathy Trial (CAT). Circulation. 2002;105:1453-8.
3. Bristow MR, Saxon LA, Boehmer J, et al. Cardiac-resynchronization therapy with or without an implantable defibrillator in advanced chronic heart failure. N Engl J Med. 2004;350:2140-50.
4. Desai AS, Fang JC, Maisel WH, et al. Implantable defibrillators for the prevention of mortality in patients with nonischemic cardiomyopathy: a meta-analysis of randomized controlled trials. JAMA. 2004;292:2874-9.
5. Kadish A, Dyer A, Daubert JP, et al. Prophylactic defibrillator implantation in patients with nonischemic dilated cardiomyopathy. N Engl J Med. 2004;350:2151-8.
6. Strickberger SA, Hummel JD, Bartlett TG, et al. Amiodarone versus implantable cardioverter-defibrillator: randomized trial in patients with nonischemic dilated cardiomyopathy and asymptomatic nonsustained ventricular tachycardia--AMIOVIRT. J Am Coll Cardiol. 2003;41:1707-12.
7. Anselme F, Moubarak G, Savoure A, et al. Implantable cardioverter-defibrillators in lamin A/C mutation carriers with cardiac conduction disorders. Heart Rhythm. 2013;10:1492-8.
8. Pasotti M, Klersy C, Pilotto A, et al. Long-term outcome and risk stratification in dilated cardiolaminopathies. J Am Coll Cardiol. 2008;52:1250-60.

9. Van Berlo JH, de Voogt WG, van der Kooi AJ, et al. Meta-analysis of clinical characteristics of 299 carriers of LMNA gene mutations: do lamin A/C mutations portend a high risk of sudden death? J Mol Med. 2005;83:79-83.
10. van Rijsingen IA, Arbustini E, Elliott PM, et al. Risk factors for malignant ventricular arrhythmias in lamin A/C mutation carriers a European cohort study. J Am Coll Cardiol. 2012;59:493-500.

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 (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 (2, 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 cardiac-defibrillator.

References

1. Connolly SJ, Dorian P, Roberts RS, et al. Comparison of beta-blockers, amiodarone plus beta-blockers, or sotalol for prevention of shocks from implantable cardioverter defibrillators: the OPTIC Study: a randomized trial. JAMA. 2006;295:165-71.
2. Dinov B, Fiedler L, Schonbauer R, et al. Outcomes in catheter ablation of ventricular tachycardia in dilated nonischemic cardiomyopathy compared with ischemic cardiomyopathy: results from the Prospective Heart Centre of Leipzig VT (HELP-VT) Study. Circulation. 2014;129:728-36.
3. Tung R, Vaseghi M, Frankel DS, et al. Freedom from recurrent ventricular tachycardia after catheter ablation is associated with improved survival in patients with structural heart disease: an International VT Ablation Center Collaborative Group study. Heart Rhythm. 2015;12:1997-2007.

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 (1-4).**
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 (5-8).**
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 (9-13).**
I	**B-NR**	**4. In patients with arrhythmogenic right ventricular cardiomyopathy and VA, a beta blocker is recommended (11, 14, 15).**
I	**B-NR**	**5. In patients with a clinical diagnosis of arrhythmogenic right ventricular cardiomyopathy , avoiding intensive exercise is recommended (11, 12, 16-21).**
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 (1, 4, 22-26).**
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 (10, 11, 13).**
IIa	**B-NR**	**8. In patients with clinical evidence of arrhythmogenic right ventricular cardiomyopathy but not VA, a beta blocker can be useful (14, 15).**
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 (27-33).**
IIa	**B-NR**	**10. In patients with suspected arrhythmogenic right ventricular cardiomyopathy , a signal averaged ECG can be useful for diagnosis and risk stratification (14, 34, 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 (9, 36).**

References

1. Groeneweg JA, Bhonsale A, James CA, et al. Clinical presentation, long-term follow-up, and outcomes of 1001 arrhythmogenic right ventricular dysplasia/cardiomyopathy patients and family members. Circ Cardiovasc Genet. 2015;8:437-46.
2. Marcus FI, Edson S, Towbin JA. Genetics of arrhythmogenic right ventricular cardiomyopathy: a practical guide for physicians. J Am Coll Cardiol. 2013;61:1945-8.

3. Quarta G, Muir A, Pantazis A, et al. Familial evaluation in arrhythmogenic right ventricular cardiomyopathy: impact of genetics and revised task force criteria. Circulation. 2011;123:2701-9.
4. te Riele AS, James CA, Groeneweg JA, et al. Approach to family screening in arrhythmogenic right ventricular dysplasia/cardiomyopathy. Eur Heart J. 2016;37:755-63.
5. Liu T, Pursnani A, Sharma UC, et al. Effect of the 2010 task force criteria on reclassification of cardiovascular magnetic resonance criteria for arrhythmogenic right ventricular cardiomyopathy. J Cardiovasc Magn Reson. 2014;16:47.
6. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the task force criteria. Circulation. 2010;121:1533-41.
7. te Riele AS, Bhonsale A, James CA, et al. Incremental value of cardiac magnetic resonance imaging in arrhythmic risk stratification of arrhythmogenic right ventricular dysplasia/cardiomyopathy-associated desmosomal mutation carriers. J Am Coll Cardiol. 2013;62:1761-9.
8. Vermes E, Strohm O, Otmani A, et al. Impact of the revision of arrhythmogenic right ventricular cardiomyopathy/dysplasia task force criteria on its prevalence by CMR criteria. JACC Cardiovasc Imag. 2011;4:282-7.
9. Bhonsale A, James CA, Tichnell C, et al. Incidence and predictors of implantable cardioverter-defibrillator therapy in patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy undergoing implantable cardioverter-defibrillator implantation for primary prevention. J Am Coll Cardiol. 2011;58:1485-96.
10. Corrado D, Calkins H, Link MS, et al. Prophylactic implantable defibrillator in patients with arrhythmogenic right ventricular cardiomyopathy/dysplasia and no prior ventricular fibrillation or sustained ventricular tachycardia. Circulation. 2010;122:1144-52.
11. Corrado D, Wichter T, Link MS, et al. Treatment of arrhythmogenic right ventricular cardiomyopathy/dysplasia: an International Task Force Consensus Statement. Circulation. 2015;132:441-53.
12. Link MS, Laidlaw D, Polonsky B, et al. Ventricular arrhythmias in the North American multidisciplinary study of ARVC: predictors, characteristics, and treatment. J Am Coll Cardiol. 2014;64:119-25.
13. Piccini JP, Dalal D, Roguin A, et al. Predictors of appropriate implantable defibrillator therapies in patients with arrhythmogenic right ventricular dysplasia. Heart Rhythm. 2005;2:1188-94.
14. Marcus FI, Zareba W, Calkins H, et al. Arrhythmogenic right ventricular cardiomyopathy/dysplasia clinical presentation and diagnostic evaluation: results from the North American Multidisciplinary Study. Heart Rhythm. 2009;6:984-92.
15. Marcus GM, Glidden DV, Polonsky B, et al. Efficacy of antiarrhythmic drugs in arrhythmogenic right ventricular cardiomyopathy: a report from the North American ARVC Registry. J Am Coll Cardiol. 2009;54:609-15.
16. Corrado D, Basso C, Thiene G, et al. Spectrum of clinicopathologic manifestations of arrhythmogenic right ventricular cardiomyopathy/dysplasia: a multicenter study. J Am Coll Cardiol. 1997;30:1512-20.
17. James CA, Bhonsale A, Tichnell C, et al. Exercise increases age-related penetrance and arrhythmic risk in arrhythmogenic right ventricular dysplasia/cardiomyopathy-associated desmosomal mutation carriers. J Am Coll Cardiol. 2013;62:1290-7.
18. Ruwald AC, Marcus F, Estes NA 3rd, et al. Association of competitive and recreational sport participation with cardiac events in patients with arrhythmogenic right ventricular cardiomyopathy: results from the North American multidisciplinary study of arrhythmogenic right ventricular cardiomyopathy. Eur Heart J. 2015;36:1735-43.
19. Saberniak J, Hasselberg NE, Borgquist R, et al. Vigorous physical activity impairs myocardial function in patients with arrhythmogenic right ventricular cardiomyopathy and in mutation positive family members. Eur J Heart Fail. 2014;16:1337-44.
20. Sawant AC, Bhonsale A, te Riele AS, et al. Exercise has a disproportionate role in the pathogenesis of arrhythmogenic right ventricular dysplasia/cardiomyopathy in patients without desmosomal mutations. J Am Heart Assoc. 2014;3:e001471.
21. Sawant AC, te Riele AS, Tichnell C, et al. Safety of American Heart Association-recommended minimum exercise for desmosomal mutation carriers. Heart Rhythm. 2016;13:199-207.
22. Bhonsale A, Groeneweg JA, James CA, et al. Impact of genotype on clinical course in arrhythmogenic right ventricular dysplasia/cardiomyopathy-associated mutation carriers. Eur Heart J. 2015;36:847-55.

23. Bhonsale A, James CA, Tichnell C, et al. Risk stratification in arrhythmogenic right ventricular dysplasia/cardiomyopathy-associated desmosomal mutation carriers. Circ Arrhythm Electrophysiol. 2013;6:569-78.
24. Hershberger RE, Lindenfeld J, Mestroni L, et al. Genetic evaluation of cardiomyopathy--a Heart Failure Society of America practice guideline. J Card Fail. 2009;15:83-97.
25. Kapplinger JD, Landstrom AP, Salisbury BA, et al. Distinguishing arrhythmogenic right ventricular cardiomyopathy/dysplasia-associated mutations from background genetic noise. J Am Coll Cardiol. 2011;57:2317-27.
26. Rigato I, Bauce B, Rampazzo A, et al. Compound and digenic heterozygosity predicts lifetime arrhythmic outcome and sudden cardiac death in desmosomal gene-related arrhythmogenic right ventricular cardiomyopathy. Circ Cardiovasc Genet. 2013;6:533-42.
27. Philips B, te Riele AS, Sawant A, et al. Outcomes and ventricular tachycardia recurrence characteristics after epicardial ablation of ventricular tachycardia in arrhythmogenic right ventricular dysplasia/cardiomyopathy. Heart Rhythm. 2015;12:716-25.
28. Philips B, Madhavan S, James C, et al. Outcomes of catheter ablation of ventricular tachycardia in arrhythmogenic right ventricular dysplasia/cardiomyopathy. Circ Arrhythm Electrophysiol. 2012;5:499-505.
29. Dalal D, Jain R, Tandri H, et al. Long-term efficacy of catheter ablation of ventricular tachycardia in patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy. J Am Coll Cardiol. 2007;50:432-40.
30. Garcia FC, Bazan V, Zado ES, et al. Epicardial substrate and outcome with epicardial ablation of ventricular tachycardia in arrhythmogenic right ventricular cardiomyopathy/dysplasia. Circulation. 2009;120:366-75.
31. Bai R, Di BL, Shivkumar K, et al. Ablation of ventricular arrhythmias in arrhythmogenic right ventricular dysplasia/cardiomyopathy: arrhythmia-free survival after endo-epicardial substrate based mapping and ablation. Circ Arrhythm Electrophysiol. 2011;4:478-85.
32. Berruezo A, Fernandez-Armenta J, Mont L, et al. Combined endocardial and epicardial catheter ablation in arrhythmogenic right ventricular dysplasia incorporating scar dechanneling technique. Circ Arrhythm Electrophysiol. 2012;5:111-21.
33. Santangeli P, Zado ES, Supple GE, et al. Long-term outcome with catheter ablation of ventricular tachycardia in patients with arrhythmogenic right ventricular cardiomyopathy. Circ Arrhythm Electrophysiol. 2015;8:1413-21.
34. Choudhary N, Tompkins C, Polonsky B, et al. Clinical presentation and outcomes by sex in arrhythmogenic right ventricular cardiomyopathy: findings from the North American ARVC registry. J Cardiovasc Electrophysiol. 2016;27:555-62.
35. Kamath GS, Zareba W, Delaney J, et al. Value of the signal-averaged electrocardiogram in arrhythmogenic right ventricular cardiomyopathy/dysplasia. Heart Rhythm. 2011;8:256-62.
36. Saguner AM, Medeiros-Domingo A, Schwyzer MA, et al. Usefulness of inducible ventricular tachycardia to predict long-term adverse outcomes in arrhythmogenic right ventricular cardiomyopathy. Am J Cardiol. 2013;111:250-7.

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 (1-8).
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 (1, 6, 9, 10).
I	B-NR	3. In first-degree relatives of patients with HCM, an ECG and echocardiogram should be performed (11-17).
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 (13-15, 18, 19).
IIa	B-NR	5. In patients with clinically suspected or diagnosed HCM, genetic counseling and genetic testing are reasonable (13-15, 18-22).
IIa	B-NR C-LD C-LD	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) (2, 3, 23, 24). b. SCD in 1 or more first-degree relatives presumably caused by HCM (LOE: C-LD) (25, 26). c. 1 or more episodes of unexplained syncope within the preceding 6 months (LOE: C-LD) (8, 26).
IIa	B-NR C-LD	7. In patients with HCM who have spontaneous NSVT (LOE: C-LD) (2, 26, 27) or an abnormal blood pressure response with exercise (LOE: B-NR) (5, 28, 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.
IIb	B-NR B-NR	8. In patients with HCM who have NSVT (LOE: B-NR) (2, 26, 27) or an abnormal blood pressure response with exercise (LOE: B-NR) (5, 28, 29) but do not have any other SCD risk modifiers, an ICD may be considered, but its benefit is uncertain.
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 (30, 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 (32, 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 (7, 34, 35).

Table 8 and Figure7

Refer to the ACCF/AHA HCM guideline for the definition of HCM (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 (1, 5, 6)
- Spontaneous sustained VT causing syncope or hemodynamic compromise (1, 5, 6)
- Family history of SCD associated with HCM (25, 26)
- LV wall thickness ≥30 mm (2, 3, 23, 24)
- Unexplained syncope within 6 mo (8, 26)
- NSVT ≥3 beats (2, 26, 27)
- Abnormal blood pressure response during exercise† (5, 28, 29)

Potential risk modifiers‡

- <30 y (5, 26)
- Delayed hyperenhancement on cardiac MRI (37-39, 54)
- LVOT obstruction (2, 4)
- Syncope >5 y ago (8, 26)

High-risk subsets§

- LV aneurysm (40, 55, 56)
- LVEF <50% (52)

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

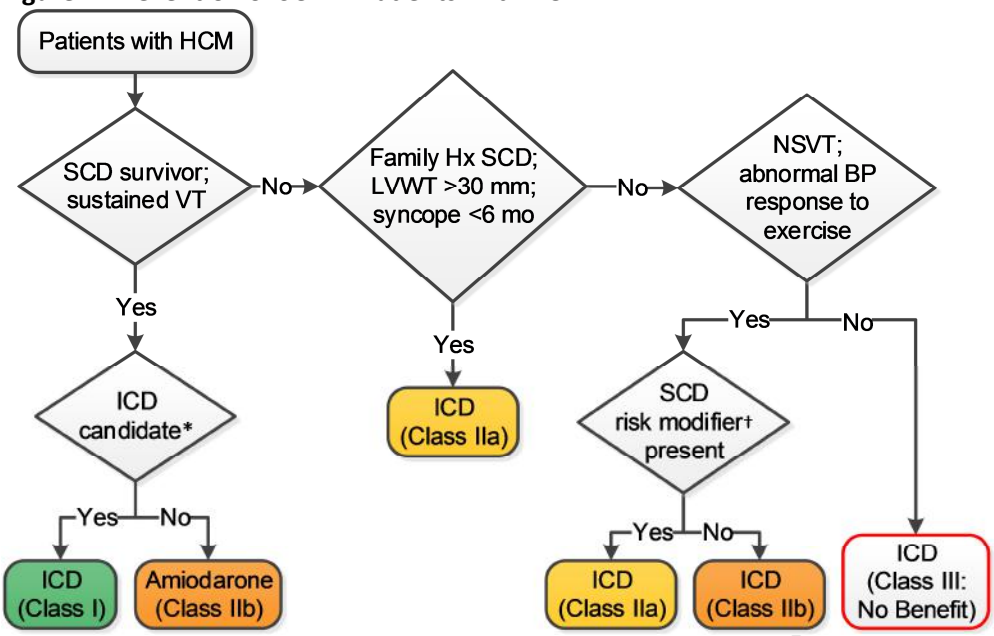
†Decrease in blood pressure of 20 mm Hg or failure to increase systolic blood pressure >20 mm Hg during exertion.

‡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 (52).

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.

References

1. Elliott PM, Sharma S, Varnava A, et al. Survival after cardiac arrest or sustained ventricular tachycardia in patients with hypertrophic cardiomyopathy. J Am Coll Cardiol. 1999;33:1596-601.
2. Elliott PM, Poloniecki J, Dickie S, et al. Sudden death in hypertrophic cardiomyopathy: identification of high risk patients. J Am Coll Cardiol. 2000;36:2212-8.
3. Elliott PM, Gimeno Blanes JR, Mahon NG, et al. Relation between severity of left-ventricular hypertrophy and prognosis in patients with hypertrophic cardiomyopathy. Lancet. 2001;357:420-4.
4. Elliott PM, Gimeno JR, Tome MT, et al. Left ventricular outflow tract obstruction and sudden death risk in patients with hypertrophic cardiomyopathy. Eur Heart J. 2006;27:1933-41.
5. Maki S, Ikeda H, Muro A, et al. Predictors of sudden cardiac death in hypertrophic cardiomyopathy. Am J Cardiol. 1998;82:774-8.
6. Maron BJ, Spirito P, Shen WK, et al. Implantable cardioverter-defibrillators and prevention of sudden cardiac death in hypertrophic cardiomyopathy. JAMA. 2007;298:405-12.
7. O'Mahony C, Jichi F, Pavlou M, et al. A novel clinical risk prediction model for sudden cardiac death in hypertrophic cardiomyopathy (HCM risk-SCD). Eur Heart J. 2014;35:2010-20.
8. Spirito P, Autore C, Rapezzi C, et al. Syncope and risk of sudden death in hypertrophic cardiomyopathy. Circulation. 2009;119:1703-10.
9. O'Mahony C, Lambiase PD, Quarta G, et al. The long-term survival and the risks and benefits of implantable cardioverter defibrillators in patients with hypertrophic cardiomyopathy. Heart. 2012;98:116-25.
10. Syska P, Przybylski A, Chojnowska L, et al. Implantable cardioverter-defibrillator in patients with hypertrophic cardiomyopathy: efficacy and complications of the therapy in long-term follow-up. J Cardiovasc Electrophysiol. 2010;21:883-9.
11. Adabag AS, Kuskowski MA, Maron BJ. Determinants for clinical diagnosis of hypertrophic cardiomyopathy. Am J Cardiol. 2006;98:1507-11.

12. Afonso LC, Bernal J, Bax JJ, et al. Echocardiography in hypertrophic cardiomyopathy: the role of conventional and emerging technologies. JACC Cardiovasc Imag. 2008;1:787-800.
13. Girolami F, Ho CY, Semsarian C, et al. Clinical features and outcome of hypertrophic cardiomyopathy associated with triple sarcomere protein gene mutations. J Am Coll Cardiol. 2010;55:1444-53.
14. Ingles J, Sarina T, Yeates L, et al. Clinical predictors of genetic testing outcomes in hypertrophic cardiomyopathy. Genet Med. 2013;15:972-7.
15. Jensen MK, Havndrup O, Christiansen M, et al. Penetrance of hypertrophic cardiomyopathy in children and adolescents: a 12-year follow-up study of clinical screening and predictive genetic testing. Circulation. 2013;127:48-54.
16. Klues HG, Schiffers A, Maron BJ. Phenotypic spectrum and patterns of left ventricular hypertrophy in hypertrophic cardiomyopathy: morphologic observations and significance as assessed by two-dimensional echocardiography in 600 patients. J Am Coll Cardiol. 1995;26:1699-708.
17. Wigle ED, Rakowski H, Kimball BP, et al. Hypertrophic cardiomyopathy. Clinical spectrum and treatment. Circulation. 1995;92:1680-92.
18. Christiaans I, Birnie E, van Langen IM, et al. The yield of risk stratification for sudden cardiac death in hypertrophic cardiomyopathy myosin-binding protein C gene mutation carriers: focus on predictive screening. Eur Heart J. 2010;31:842-8.
19. Olivotto I, Girolami F, Ackerman MJ, et al. Myofilament protein gene mutation screening and outcome of patients with hypertrophic cardiomyopathy. Mayo Clin Proc. 2008;83:630-8.
20. Christiaans I, van Langen IM, Birnie E, et al. Genetic counseling and cardiac care in predictively tested hypertrophic cardiomyopathy mutation carriers: the patients' perspective. Am J Med Genet. 2009;149a:1444-51.
21. Hamang A, Eide GE, Rokne B, et al. Predictors of heart-focused anxiety in patients undergoing genetic investigation and counseling of long QT syndrome or hypertrophic cardiomyopathy: a one year follow-up. J Genet Counsel. 2012;21:72-84.
22. Bos JM, Will ML, Gersh BJ, et al. Characterization of a phenotype-based genetic test prediction score for unrelated patients with hypertrophic cardiomyopathy. Mayo Clin Proc. 2014;89:727-37.
23. Sorajja P, Nishimura RA, Ommen SR, et al. Use of echocardiography in patients with hypertrophic cardiomyopathy: clinical implications of massive hypertrophy. J Am Soc Echocardiogr. 2006;19:788-95.
24. Spirito P, Bellone P, Harris KM, et al. Magnitude of left ventricular hypertrophy and risk of sudden death in hypertrophic cardiomyopathy. N Engl J Med. 2000;342:1778-85.
25. Bos JM, Maron BJ, Ackerman MJ, et al. Role of family history of sudden death in risk stratification and prevention of sudden death with implantable defibrillators in hypertrophic cardiomyopathy. Am J Cardiol. 2010;106:1481-6.
26. Maron BJ, Shen WK, Link MS, et al. Efficacy of implantable cardioverter-defibrillators for the prevention of sudden death in patients with hypertrophic cardiomyopathy. N Engl J Med. 2000;342:365-73.
27. Monserrat L, Elliott PM, Gimeno JR, et al. Non-sustained ventricular tachycardia in hypertrophic cardiomyopathy: an independent marker of sudden death risk in young patients. J Am Coll Cardiol. 2003;42:873-9.
28. Olivotto I, Maron BJ, Montereggi A, et al. Prognostic value of systemic blood pressure response during exercise in a community-based patient population with hypertrophic cardiomyopathy. J Am Coll Cardiol. 1999;33:2044-51.
29. Sadoul N, Prasad K, Elliott PM, et al. Prospective prognostic assessment of blood pressure response during exercise in patients with hypertrophic cardiomyopathy. Circulation. 1997;96:2987-91.
30. McKenna WJ, Oakley CM, Krikler DM, et al. Improved survival with amiodarone in patients with hypertrophic cardiomyopathy and ventricular tachycardia. Br Heart J. 1985;53:412-6.
31. Melacini P, Maron BJ, Bobbo F, et al. Evidence that pharmacological strategies lack efficacy for the prevention of sudden death in hypertrophic cardiomyopathy. Heart. 2007;93:708-10.
32. Kuck KH, Kunze KP, Schluter M, et al. Programmed electrical stimulation in hypertrophic cardiomyopathy. Results in patients with and without cardiac arrest or syncope. Eur Heart J. 1988;9:177-85.
33. Zhu DW, Sun H, Hill R, et al. The value of electrophysiology study and prophylactic implantation of cardioverter defibrillator in patients with hypertrophic cardiomyopathy. Pacing Clin Electrophysiol. 1998;21:299-302.
34. Ackerman MJ, VanDriest SL, Ommen SR, et al. Prevalence and age-dependence of malignant mutations in the beta-myosin heavy chain and troponin T genes in hypertrophic cardiomyopathy: a comprehensive outpatient perspective. J Am Coll Cardiol. 2002;39:2042-8.
35. Lopes LR, Rahman MS, Elliott PM. A systematic review and meta-analysis of genotype-phenotype associations in patients with hypertrophic cardiomyopathy caused by sarcomeric protein mutations. Heart. 2013;99:1800-11.

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 (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 (2-4).**

References

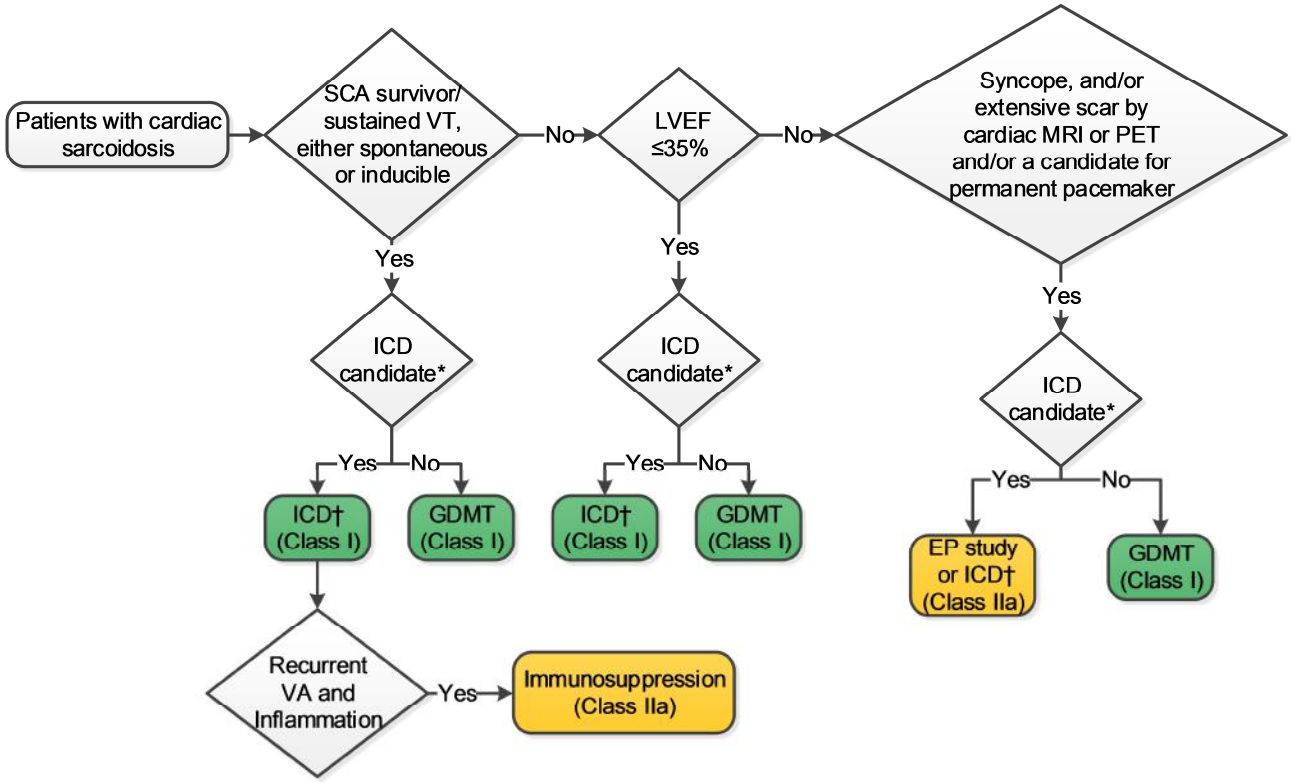
1. Aoyama N, Izumi T, Hiramori K, et al. National survey of fulminant myocarditis in Japan: therapeutic guidelines and long-term prognosis of using percutaneous cardiopulmonary support for fulminant myocarditis (special report from a scientific committee). Circ J. 2002;66:133-44.
2. Maleszewski JJ, Orellana VM, Hodge DO, et al. Long-term risk of recurrence, morbidity and mortality in giant cell myocarditis. Am J Cardiol. 2015;115:1733-8.
3. Cooper LT Jr, Berry GJ, Shabetai R. Idiopathic giant-cell myocarditis--natural history and treatment. Multicenter Giant Cell Myocarditis Study Group Investigators. N Engl J Med. 1997;336:1860-6.
4. Kandolin R, Lehtonen J, Salmenkivi K, et al. Diagnosis, treatment, and outcome of giant-cell myocarditis in the era of combined immunosuppression. Circ Heart Fail. 2013;6:15-22.

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 (1-5).**
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 (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 impant an ICD, if sustained VA is inducible, provided that meaningful survival of greater than 1 year is expected (11, 12).**
IIa	**C-LD**	**4. In patients with cardiac sarcoidosis who have an indication for permanent pacing, implantation of an ICD can be beneficial (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 (14-16).**

Figure 8

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 cardiac-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.

References

1. Kandolin R, Lehtonen J, Airaksinen J, et al. Cardiac sarcoidosis: epidemiology, characteristics, and outcome over 25 years in a nationwide study. Circulation. 2015;131:624-32.
2. Yazaki Y, Isobe M, Hiroe M, et al. Prognostic determinants of long-term survival in Japanese patients with cardiac sarcoidosis treated with prednisone. Am J Cardiol. 2001;88:1006-10.
3. Kron J, Sauer W, Schuller J, et al. Efficacy and safety of implantable cardiac defibrillators for treatment of ventricular arrhythmias in patients with cardiac sarcoidosis. Europace. 2013;15:347-54.
4. Mohsen A, Jimenez A, Hood RE, et al. Cardiac sarcoidosis: electrophysiological outcomes on long-term follow-up and the role of the implantable cardioverter-defibrillator. J Cardiovasc Electrophysiol. 2014;25:171-6.
5. Schuller JL, Zipse M, Crawford T, et al. Implantable cardioverter defibrillator therapy in patients with cardiac sarcoidosis. J Cardiovasc Electrophysiol. 2012;23:925-9.
6. Coleman GC, Shaw PW, Balfour PC Jr, et al. Prognostic value of myocardial scarring on CMR in patients with cardiac sarcoidosis: a systematic review and meta-analysis. JACC Cardiovasc Imaging. 2016;10:411-20.
7. Murtagh G, Laffin LJ, Beshai JF, et al. Prognosis of myocardial damage in sarcoidosis patients with preserved left ventricular ejection fraction: risk stratification using cardiovascular magnetic resonance. Circ Cardiovasc Imaging. 2016;9:e003738.

8. Crawford T, Mueller G, Sarsam S, et al. Magnetic resonance imaging for identifying patients with cardiac sarcoidosis and preserved or mildly reduced left ventricular function at risk of ventricular arrhythmias. Circ Arrhythm Electrophysiol. 2014;7:1109-15.
9. Greulich S, Deluigi CC, Gloekler S, et al. CMR imaging predicts death and other adverse events in suspected cardiac sarcoidosis. JACC Cardiovasc Imaging. 2013;6:501-11.
10. Blankstein R, Osborne M, Naya M, et al. Cardiac positron emission tomography enhances prognostic assessments of patients with suspected cardiac sarcoidosis. J Am Coll Cardiol. 2014;63:329-36.
11. Aizer A, Stern EH, Gomes JA, et al. Usefulness of programmed ventricular stimulation in predicting future arrhythmic events in patients with cardiac sarcoidosis. Am J Cardiol. 2005;96:276-82.
12. Mehta RH, Starr AZ, Lopes RD, et al. Incidence of and outcomes associated with ventricular tachycardia or fibrillation in patients undergoing primary percutaneous coronary intervention. JAMA. 2009;301:1779-89.
13. Takaya Y, Kusano KF, Nakamura K, et al. Outcomes in patients with high-degree atrioventricular block as the initial manifestation of cardiac sarcoidosis. Am J Cardiol 2015;115:505-9.
14. Naruse Y, Sekiguchi Y, Nogami A, et al. Systematic treatment approach to ventricular tachycardia in cardiac sarcoidosis. Circ Arrhythm Electrophysiol. 2014;7:407-13.
15. Segawa M, Fukuda K, Nakano M, et al. Time course and factors correlating with ventricular tachyarrhythmias after introduction of steroid therapy in cardiac sarcoidosis. Circ Arrhythm Electrophysiol. 2016;9:e003353.
16. Yodogawa K, Seino Y, Ohara T, et al. Effect of corticosteroid therapy on ventricular arrhythmias in patients with cardiac sarcoidosis. Ann Noninvasive Electrocardiol. 2011;16:140-7.

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 (e.g., NYHA class IV and/or use of inotropes) with a plan to discharge home, an ICD is reasonable (1-5).**

References

1. Frohlich GM, Holzmeister J, Hubler M, et al. Prophylactic implantable cardioverter defibrillator treatment in patients with end-stage heart failure awaiting heart transplantation. Heart. 2013;99:1158-65.
2. Kao AC, Krause SW, Handa R, et al. Wearable defibrillator use in heart failure (WIF): results of a prospective registry. BMC Cardiovasc Disord. 2012;12:123.
3. Opreanu M, Wan C, Singh V, et al. Wearable cardioverter-defibrillator as a bridge to cardiac transplantation: a national database analysis. J Heart Lung Transplant. 2015;34:1305-9.
4. Sandner SE, Wieselthaler G, Zuckermann A, et al. Survival benefit of the implantable cardioverter-defibrillator in patients on the waiting list for cardiac transplantation. Circulation. 2001;104:I171-6.
5. Vakil K, Duval S, Cogswell R, et al. Impact of implantable cardioverter-defibrillators on waitlist mortality among patients awaiting heart transplantation. An UNOS/OPTN Analysis. JACC Clin Electrophysiol. 2017;3:33-40.

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 (1).**

References
1. Vakil K, Kazmirczak F, Sathnur N, et al. Implantable cardioverter-defibrillator use in patients with left ventricular assist devices: a systematic review and meta-analysis. JACC Heart Fail. 2016;4:772-9.

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 (1-3).**

References
1. Tsai VW, Cooper J, Garan H, et al. The efficacy of implantable cardioverter-defibrillators in heart transplant recipients: results from a multicenter registry. Circ Heart Fail. 2009;2:197-201.
2. McDowell DL, Hauptman PJ. Implantable defibrillators and cardiac resynchronization therapy in heart transplant recipients: results of a national survey. J Heart Lung Transplant. 2009;28:847-50.
3. Neylon A, Canniffe C, Parlon B, et al. Implantable cardioverter-defibrillators in a heart transplant population: a single-center experience. J Heart Lung Transplant. 2016;35:682-4.

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 (1, 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 a meaningful survival of greater than 1 year is expected (3-8).**
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 (9-12).**
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 (9, 13, 14).**

Table 9

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

HF indicates heart failure; and NICM, nonischemic cardiomyopathy.
Adapted with permission from Groh, et al. (15).

References

1. Bhakta D, Groh MR, Shen C, et al. Increased mortality with left ventricular systolic dysfunction and heart failure in adults with myotonic dystrophy type 1. Am Heart J. 2010;160:1137-41, 41.
2. Merino JL, Carmona JR, Fernandez-Lozano I, et al. Mechanisms of sustained ventricular tachycardia in myotonic dystrophy: implications for catheter ablation. Circulation. 1998;98:541-6.
3. Anselme F, Moubarak G, Savoure A, et al. Implantable cardioverter-defibrillators in lamin A/C mutation carriers with cardiac conduction disorders. Heart Rhythm. 2013;10:1492-8.
4. van Rijsingen IA, Arbustini E, Elliott PM, et al. Risk factors for malignant ventricular arrhythmias in lamin A/C mutation carriers a European cohort study. J Am Coll Cardiol. 2012;59:493-500.
5. Meune C, Van Berlo JH, Anselme F, et al. Primary prevention of sudden death in patients with lamin A/C gene mutations. N Engl J Med. 2006;354:209-10.
6. Pasotti M, Klersy C, Pilotto A, et al. Long-term outcome and risk stratification in dilated cardiolaminopathies. J Am Coll Cardiol. 2008;52:1250-60.
7. Van Berlo JH, de Voogt WG, van der Kooi AJ, et al. Meta-analysis of clinical characteristics of 299 carriers of LMNA gene mutations: do lamin A/C mutations portend a high risk of sudden death? J Mol Med. 2005;83:79-83.
8. Russo V, Nigro G. ICD role in preventing sudden cardiac death in Emery-Dreifuss muscular dystrophy with preserved myocardial function: 2013 ESC Guidelines on Cardiac Pacing and Cardiac Resynchronization Therapy. Europace. 2015;17:337.

9. Groh WJ, Groh MR, Saha C, et al. Electrocardiographic abnormalities and sudden death in myotonic dystrophy type 1. N Engl J Med. 2008;358:2688-97.
10. Lallemand B, Clementy N, Bernard-Brunet A, et al. The evolution of infrahissian conduction time in myotonic dystrophy patients: clinical implications. Heart. 2012;98:291-6.
11. Nazarian S, Wagner KR, Caffo BS, et al. Clinical predictors of conduction disease progression in type I myotonic muscular dystrophy. Pacing Clin Electrophysiol. 2011;34:171-6.
12. Tanawuttiwat T, Wagner KR, Tomaselli G, et al. Left ventricular dysfunction and conduction disturbances in patients with myotonic muscular dystrophy type I and II. JAMA Cardiology. 2017;2:225-8.
13. Bhakta D, Shen C, Kron J, et al. Pacemaker and implantable cardioverter-defibrillator use in a US myotonic dystrophy type 1 population. J Cardiovasc Electrophysiol. 2011;22:1369-75.
14. Laurent V, Pellieux S, Corcia P, et al. Mortality in myotonic dystrophy patients in the area of prophylactic pacing devices. Int J Cardiol. 2011;150:54-8.
15. Groh WJ. Arrhythmias in the muscular dystrophies. Heart Rhythm. 2012;9:1890-95.

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 (1-6).**
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 (7-13).**

References

1. Bai R, Napolitano C, Bloise R, et al. Yield of genetic screening in inherited cardiac channelopathies: how to prioritize access to genetic testing. Circ Arrhythm Electrophysiol. 2009;2:6-15.
2. Goldenberg I, Horr S, Moss AJ, et al. Risk for life-threatening cardiac events in patients with genotype-confirmed long-QT syndrome and normal-range corrected QT intervals. J Am Coll Cardiol. 2011;57:51-9.
3. Nannenberg EA, Sijbrands EJ, Dijksman LM, et al. Mortality of inherited arrhythmia syndromes: insight into their natural history. Circ Cardiovasc Genet. 2012;5:183-9.
4. Priori SG, Gasparini M, Napolitano C, et al. Risk stratification in Brugada syndrome: results of the PRELUDE (PRogrammed ELectrical stimUlation preDictive valuE) registry. J Am Coll Cardiol. 2012;59:37-45.
5. Priori SG, Napolitano C, Memmi M, et al. Clinical and molecular characterization of patients with catecholaminergic polymorphic ventricular tachycardia. Circulation. 2002;106:69-74.
6. Wilde AA, Moss AJ, Kaufman ES, et al. Clinical aspects of type 3 long-QT syndrome: an international multicenter study. Circulation. 2016;134:872-82.
7. Wedekind H, Burde D, Zumhagen S, et al. QT interval prolongation and risk for cardiac events in genotyped LQTS-index children. Eur J Pediatr. 2009;168:1107-15.
8. Moss AJ, Zareba W, Hall WJ, et al. Effectiveness and limitations of beta-blocker therapy in congenital long-QT syndrome. Circulation. 2000;101:616-23.
9. Zareba W, Moss AJ, Daubert JP, et al. Implantable cardioverter defibrillator in high-risk long QT syndrome patients. J Cardiovasc Electrophysiol. 2003;14:337-41.
10. Monnig G, Kobe J, Loher A, et al. Implantable cardioverter-defibrillator therapy in patients with congenital long-QT syndrome: a long-term follow-up. Heart Rhythm. 2005;2:497-504.
11. Hayashi M, Denjoy I, Extramiana F, et al. Incidence and risk factors of arrhythmic events in catecholaminergic polymorphic ventricular tachycardia. Circulation. 2009;119:2426-34.
12. Gehi AK, Duong TD, Metz LD, et al. Risk stratification of individuals with the Brugada electrocardiogram: a meta-analysis. J Cardiovasc Electrophysiol. 2006;17:577-83.
13. Probst V, Veltmann C, Eckardt L, et al. Long-term prognosis of patients diagnosed with Brugada syndrome: results from the FINGER Brugada syndrome registry. Circulation. 2010;121:635-43.

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 (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 (2, 6-12).**
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 according to the particular long QT syndrome type) or left cardiac sympathetic denervation, is recommended (6, 7, 10, 13-16).**
I	**B-NR**	**4. In patients with clinically diagnosed long QT syndrome, genetic counseling and genetic testing are recommended (17-21).**
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 (22-29).**
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 (3, 30, 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 (2, 8, 11, 30).**
III: Harm	**B-NR**	**8. In patients with long QT syndrome, QT-prolonging medications are potentially harmful (5, 12, 32-34).**

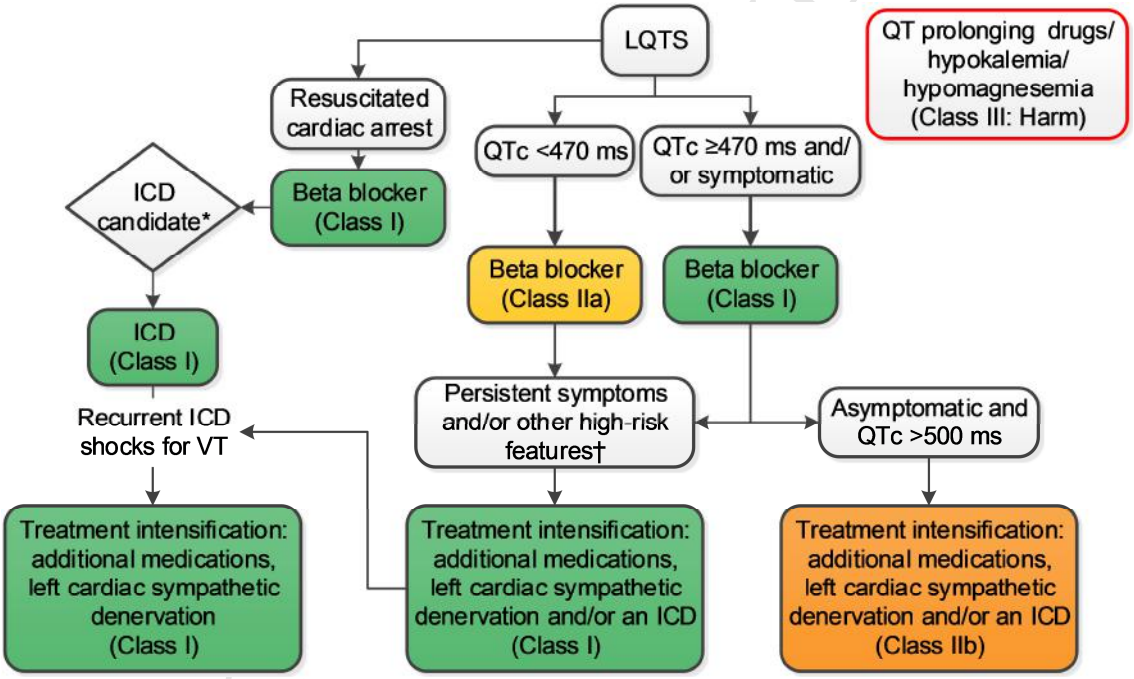
Table 10 and Figures 9, 10 (LQT1), 11 (LQT2), and 12 (LQT3)

Table 10. Commonly Used QT-Prolonging Medications (35,36)

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

*A more complete list is maintained at: www.crediblemeds.org (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

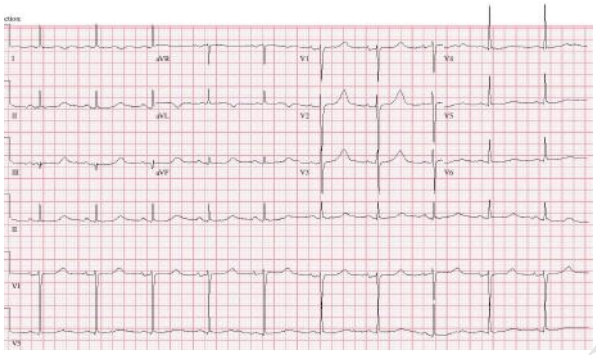


Figure 11. Long-QT Syndrome Type 2

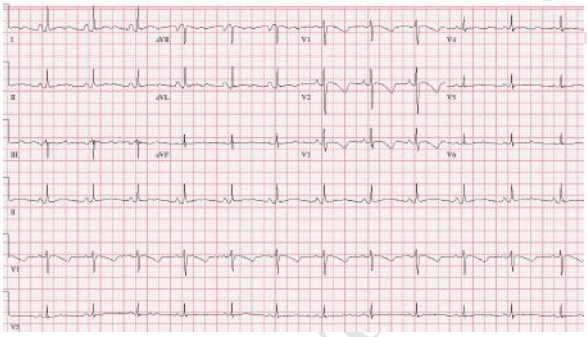
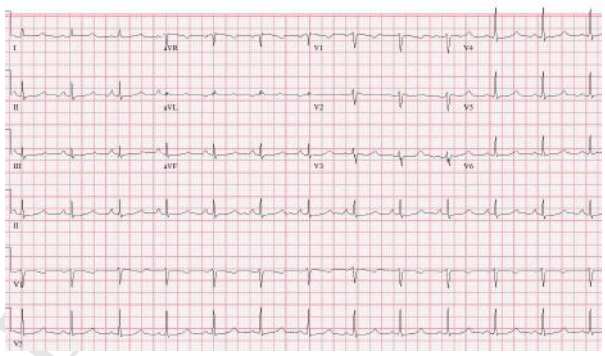


Figure 12. Long-QT Syndrome Type 3



References

1. Abu-Zeitone A, Peterson DR, Polonsky B, et al. Efficacy of different beta-blockers in the treatment of long QT syndrome. J Am Coll Cardiol. 2014;64:1352-8.
2. Goldenberg I, Bradley J, Moss A, et al. Beta-blocker efficacy in high-risk patients with the congenital long-QT syndrome types 1 and 2: implications for patient management. J Cardiovasc Electrophysiol. 2010;21:893-901.
3. Moss AJ, Zareba W, Hall WJ, et al. Effectiveness and limitations of beta-blocker therapy in congenital long-QT syndrome. Circulation. 2000;101:616-23.
4. Sauer AJ, Moss AJ, McNitt S, et al. Long QT syndrome in adults. J Am Coll Cardiol. 2007;49:329-37.
5. Vincent GM, Schwartz PJ, Denjoy I, et al. High efficacy of beta-blockers in long-QT syndrome type 1: contribution of noncompliance and QT-prolonging drugs to the occurrence of beta-blocker treatment "failures". Circulation. 2009;119:215-21.
6. Chorin E, Hu D, Antzelevitch C, et al. Ranolazine for congenital long-qt syndrome type iii: experimental and long-term clinical data. Circ Arrhythm Electrophysiol. 2016;9:e004370.

7. Chorin E, Taub R, Medina A, et al. Long-term flecainide therapy in type 3 long QT syndrome. Europace. 2017;euw439 [Epub ahead of print].
8. Hobbs JB, Peterson DR, Moss AJ, et al. Risk of aborted cardiac arrest or sudden cardiac death during adolescence in the long-QT syndrome. JAMA. 2006;296:1249-54.
9. Jons C, Moss AJ, Goldenberg I, et al. Risk of fatal arrhythmic events in long QT syndrome patients after syncope. J Am Coll Cardiol. 2010;55:783-8.
10. Mazzanti A, Maragna R, Faragli A, et al. Gene-specific therapy with mexiletine reduces arrhythmic events in patients with long QT syndrome type 3. J Am Coll Cardiol. 2016;67:1053-8.
11. Nannenberg EA, Sijbrands EJ, Dijksman LM, et al. Mortality of inherited arrhythmia syndromes: insight into their natural history. Circ Cardiovasc Genet. 2012;5:183-9.
12. Wedekind H, Burde D, Zumhagen S, et al. QT interval prolongation and risk for cardiac events in genotyped LQTS-index children. Eur J Pediatr. 2009;168:1107-15.
13. Collura CA, Johnson JN, Moir C, et al. Left cardiac sympathetic denervation for the treatment of long QT syndrome and catecholaminergic polymorphic ventricular tachycardia using video-assisted thoracic surgery. Heart Rhythm. 2009;6:752-9.
14. Hofferberth SC, Cecchin F, Loberman D, et al. Left thoracoscopic sympathectomy for cardiac denervation in patients with life-threatening ventricular arrhythmias. J Thorac Cardiovasc Surg. 2014;147:404-9.
15. Schneider HE, Steinmetz M, Krause U, et al. Left cardiac sympathetic denervation for the management of life-threatening ventricular tachyarrhythmias in young patients with catecholaminergic polymorphic ventricular tachycardia and long QT syndrome. Clin Res Cardiol. 2013;102:33-42.
16. Schwartz PJ, Priori SG, Cerrone M, et al. Left cardiac sympathetic denervation in the management of high-risk patients affected by the long-QT syndrome. Circulation. 2004;109:1826-33.
17. Bai R, Napolitano C, Bloise R, et al. Yield of genetic screening in inherited cardiac channelopathies: how to prioritize access to genetic testing. Circ Arrhythm Electrophysiol. 2009;2:6-15.
18. Costa J, Lopes CM, Barsheshet A, et al. Combined assessment of sex- and mutation-specific information for risk stratification in type 1 long QT syndrome. Heart Rhythm. 2012;9:892-8.
19. Kim JA, Lopes CM, Moss AJ, et al. Trigger-specific risk factors and response to therapy in long QT syndrome type 2. Heart Rhythm. 2010;7:1797-805.
20. Migdalovich D, Moss AJ, Lopes CM, et al. Mutation and gender-specific risk in type 2 long QT syndrome: implications for risk stratification for life-threatening cardiac events in patients with long QT syndrome. Heart Rhythm. 2011;8:1537-43.
21. Tester DJ, Will ML, Haglund CM, et al. Effect of clinical phenotype on yield of long QT syndrome genetic testing. J Am Coll Cardiol. 2006;47:764-8.
22. Adler A, van der Werf C, Postema PG, et al. The phenomenon of "QT stunning": the abnormal QT prolongation provoked by standing persists even as the heart rate returns to normal in patients with long QT syndrome. Heart Rhythm. 2012;9:901-8.
23. Aziz PF, Wieand TS, Ganley J, et al. Genotype- and mutation site-specific QT adaptation during exercise, recovery, and postural changes in children with long-QT syndrome. Circ Arrhythm Electrophysiol. 2011;4:867-73.
24. Chattha IS, Sy RW, Yee R, et al. Utility of the recovery electrocardiogram after exercise: a novel indicator for the diagnosis and genotyping of long QT syndrome? Heart Rhythm. 2010;7:906-11.
25. Laksman ZW, Hamilton RM, Chockalingam P, et al. Mutation location effect on severity of phenotype during exercise testing in type 1 long-QT syndrome: impact of transmembrane and C-loop location. J Cardiovasc Electrophysiol. 2013;24:1015-20.
26. Moltedo JM, Kim JJ, Friedman RA, et al. Use of a cardioselective beta-blocker for pediatric patients with prolonged QT syndrome. Pediatr Cardiol. 2011;32:63-6.
27. Sy RW, van der Werf C, Chattha IS, et al. Derivation and validation of a simple exercise-based algorithm for prediction of genetic testing in relatives of LQTS probands. Circulation. 2011;124:2187-94.
28. Villain E, Denjoy I, Lupoglazoff JM, et al. Low incidence of cardiac events with beta-blocking therapy in children with long QT syndrome. Eur Heart J. 2004;25:1405-11.
29. Viskin S, Postema PG, Bhuiyan ZA, et al. The response of the QT interval to the brief tachycardia provoked by standing: a bedside test for diagnosing long QT syndrome. J Am Coll Cardiol. 2010;55:1955-61.
30. Priori SG, Napolitano C, Schwartz PJ, et al. Association of long QT syndrome loci and cardiac events among patients treated with beta-blockers. JAMA. 2004;292:1341-4.

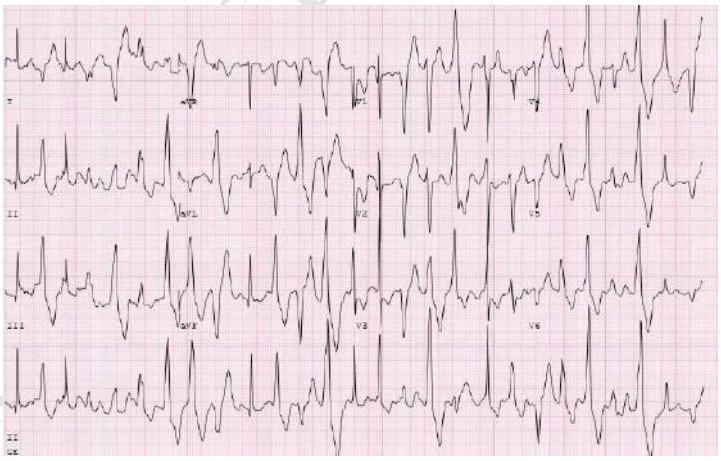
31. Goldenberg I, Horr S, Moss AJ, et al. Risk for life-threatening cardiac events in patients with genotype-confirmed long-QT syndrome and normal-range corrected QT intervals. J Am Coll Cardiol. 2011;57:51-9.
32. Choy AM, Lang CC, Chomsky DM, et al. Normalization of acquired QT prolongation in humans by intravenous potassium. Circulation. 1997;96:2149-54.
33. Kannankeril P, Roden DM, Darbar D. Drug-induced long QT syndrome. Pharmacol Rev. 2010;62:760-81.
34. Zhang C, Kutyifa V, Moss AJ, et al. Long-QT syndrome and therapy for attention deficit/hyperactivity disorder. J Cardiovasc Electrophysiol. 2015;26:1039-44.
35. Credible meds. Available at: http://www.crediblemeds.org. Accessed December 26, 2016.
36. Roden DM. Drug-induced prolongation of the QT interval. N Engl J Med. 2004;350:1013-22.

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 (1, 2).**
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 (e.g., beta blocker, flecainide), left cardiac sympathetic denervation, and/or an ICD is recommended (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 (7).**

Figure 13

Figure 13. Exercise-Induced Polymorphic VT in Catecholaminergic Polymorphic Ventricular Tachycardia



References

1. Hayashi M, Denjoy I, Extramiana F, et al. Incidence and risk factors of arrhythmic events in catecholaminergic polymorphic ventricular tachycardia. Circulation. 2009;119:2426-34.
2. Roston TM, Vinocur JM, Maginot KR, et al. Catecholaminergic polymorphic ventricular tachycardia in children: analysis of therapeutic strategies and outcomes from an international multicenter registry. Circ Arrhythm Electrophysiol. 2015;8:633-42.

3. Collura CA, Johnson JN, Moir C, et al. Left cardiac sympathetic denervation for the treatment of long QT syndrome and catecholaminergic polymorphic ventricular tachycardia using video-assisted thoracic surgery. Heart Rhythm. 2009;6:752-9.
4. Hofferberth SC, Cecchin F, Loberman D, et al. Left thoracoscopic sympathectomy for cardiac denervation in patients with life-threatening ventricular arrhythmias. J Thorac Cardiovasc Surg. 2014;147:404-9.
5. Schneider HE, Steinmetz M, Krause U, et al. Left cardiac sympathetic denervation for the management of life-threatening ventricular tachyarrhythmias in young patients with catecholaminergic polymorphic ventricular tachycardia and long QT syndrome. Clin Res Cardiol. 2013;102:33-42.
6. van der Werf C, Kannankeril PJ, Sacher F, et al. Flecainide therapy reduces exercise-induced ventricular arrhythmias in patients with catecholaminergic polymorphic ventricular tachycardia. J Am Coll Cardiol. 2011;57:2244-54.
7. Priori SG, Napolitano C, Memmi M, et al. Clinical and molecular characterization of patients with catecholaminergic polymorphic ventricular tachycardia. Circulation. 2002;106:69-74.

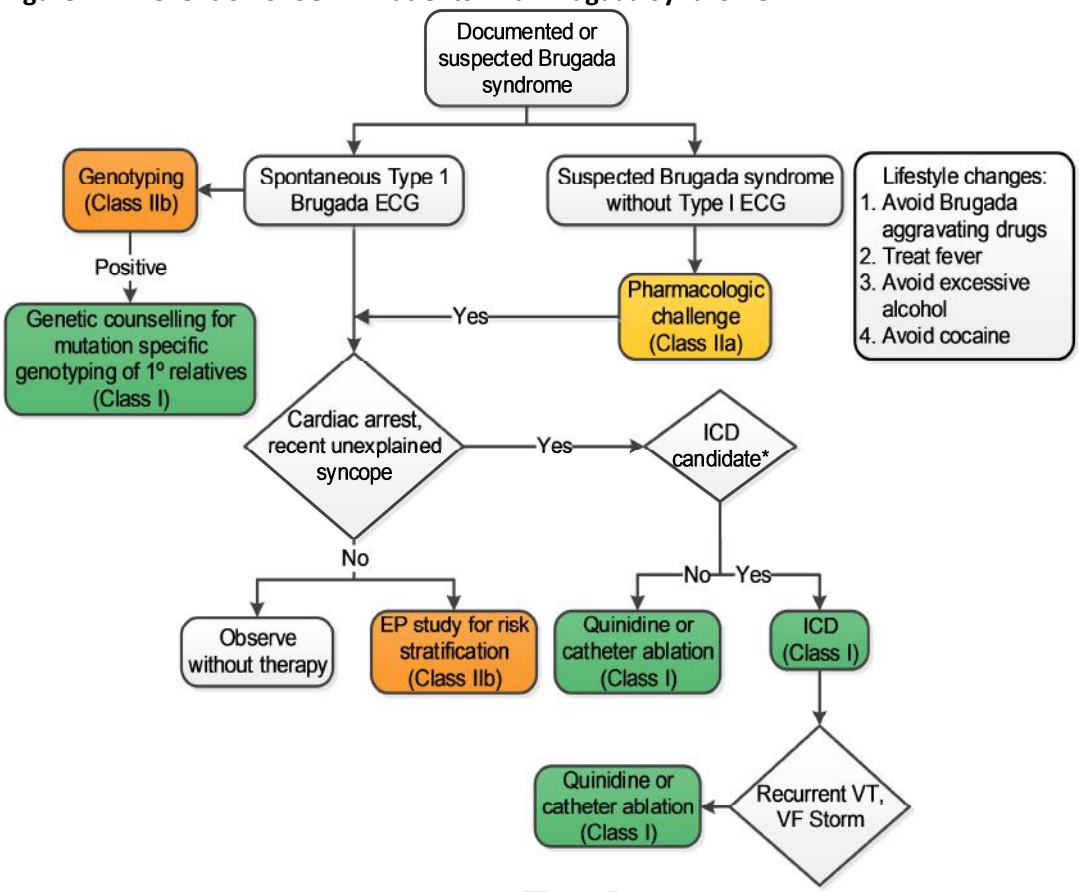
6.9.1.3. Brugada Syndrome

Recommendations for Brugada Syndrome References that support the recommendations are summarized in Online Data Supplement 42 and 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 (1-5).**
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 a meaningful survival of greater than 1 year is expected (4, 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 (7-11).**
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 (7, 9-11).**
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 (12-14).**
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 (1, 6, 13, 15-17).**
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 (18-20).**

SR indicated systematic review.

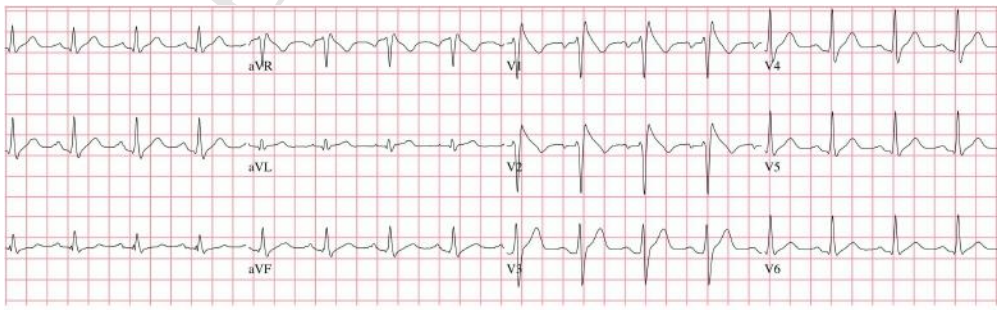
Figures 14 and 15

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



References

1. Casado-Arroyo R, Berne P, Rao JY, et al. Long-term trends in newly diagnosed Brugada syndrome: implications for risk stratification. J Am Coll Cardiol. 2016;68:614-23.
2. Gehi AK, Duong TD, Metz LD, et al. Risk stratification of individuals with the Brugada electrocardiogram: a meta-analysis. J Cardiovasc Electrophysiol. 2006;17:577-83.

3. Hiraoka M, Takagi M, Yokoyama Y, et al. Prognosis and risk stratification of young adults with Brugada syndrome. J Electrocardiol. 2013;46:279-83.
4. Priori SG, Gasparini M, Napolitano C, et al. Risk stratification in Brugada syndrome: results of the PRELUDE (PRogrammed ELectrical stimUlation preDictive valuE) registry. J Am Coll Cardiol. 2012;59:37-45.
5. Probst V, Veltmann C, Eckardt L, et al. Long-term prognosis of patients diagnosed with Brugada syndrome: results from the FINGER Brugada syndrome registry. Circulation. 2010;121:635-43.
6. Sroubek J, Probst V, Mazzanti A, et al. Programmed ventricular stimulation for risk stratification in the Brugada syndrome: a pooled analysis. Circulation. 2016;133:622-30.
7. Belhassen B, Rahkovich M, Michowitz Y, et al. Management of Brugada syndrome: thirty-three-year experience using electrophysiologically guided therapy with class 1a antiarrhythmic drugs. Circ Arrhythm Electrophysiol. 2015;8:1393-402.
8. Brugada J, Pappone C, Berruezo A, et al. Brugada syndrome phenotype elimination by epicardial substrate ablation. Circ Arrhythm Electrophysiol. 2015;8:1373-81.
9. Nademanee K, Veerakul G, Chandanamattha P, et al. Prevention of ventricular fibrillation episodes in Brugada syndrome by catheter ablation over the anterior right ventricular outflow tract epicardium. Circulation. 2011;123:1270-9.
10. Sunsaneewitayakul B, Yao Y, Thamaree S, et al. Endocardial mapping and catheter ablation for ventricular fibrillation prevention in Brugada syndrome. J Cardiovasc Electrophysiol. 2012;23(suppl 1):S10-6.
11. Zhang P, Tung R, Zhang Z, et al. Characterization of the epicardial substrate for catheter ablation of Brugada syndrome. Heart Rhythm. 2016;13:2151-8.
12. Antzelevitch C, Yan GX, Ackerman MJ, et al. J-Wave syndromes expert consensus conference report: emerging concepts and gaps in knowledge. Europace. 2017;19:665-94.
13. Delise P, Allocca G, Marras E, et al. Risk stratification in individuals with the Brugada type 1 ECG pattern without previous cardiac arrest: usefulness of a combined clinical and electrophysiologic approach. Eur Heart J. 2011;32:169-76.
14. Somani R, Krahn AD, Healey JS, et al. Procainamide infusion in the evaluation of unexplained cardiac arrest: from the Cardiac Arrest Survivors with Preserved Ejection Fraction Registry (CASPER). Heart Rhythm. 2014;11:1047-54.
15. Kusumoto FM, Bailey KR, Chaouki AS, et al. Systematic review for the 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2017. In Press.
16. Sieira J, Ciconte G, Conte G, et al. Asymptomatic Brugada syndrome: clinical characterization and long-term prognosis. Circ Arrhythm Electrophysiol. 2015;8:1144-50.
17. Sieira J, Conte G, Ciconte G, et al. Prognostic value of programmed electrical stimulation in Brugada syndrome: 20 years experience. Circ Arrhythm Electrophysiol. 2015;8:777-84.
18. Crotti L, Marcou CA, Tester DJ, et al. Spectrum and prevalence of mutations involving BrS1- through BrS12-susceptibility genes in a cohort of unrelated patients referred for Brugada syndrome genetic testing: implications for genetic testing. J Am Coll Cardiol. 2012;60:1410-8.
19. Probst V, Wilde AA, Barc J, et al. SCN5A mutations and the role of genetic background in the pathophysiology of Brugada syndrome. Circ Cardiovasc Genet. 2009;2:552-7.
20. Risgaard B, Jabbari R, Refsgaard L, et al. High prevalence of genetic variants previously associated with Brugada syndrome in new exome data. Clin Genet. 2013;84:489-95.

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 (1, 2).**
I	**B-NR**	**2. In patients with early repolarization pattern on ECG and cardiac arrest or sustained VA, an ICD is recommended (3, 4).**
III: No Benefit	**B-NR**	**3. In patients with early repolarization pattern on ECG, genetic testing is not recommended (5).**

References

1. Rosso R, Kogan E, Belhassen B, et al. J-point elevation in survivors of primary ventricular fibrillation and matched control subjects: incidence and clinical significance. J Am Coll Cardiol. 2008;52:1231-8.
2. Adhikarla C, Boga M, Wood AD, et al. Natural history of the electrocardiographic pattern of early repolarization in ambulatory patients. Am J Cardiol. 2011;108:1831-5.
3. Haissaguerre M, Sacher F, Nogami A, et al. Characteristics of recurrent ventricular fibrillation associated with inferolateral early repolarization role of drug therapy. J Am Coll Cardiol. 2009;53:612-9.
4. Siebermair J, Sinner MF, Beckmann BM, et al. Early repolarization pattern is the strongest predictor of arrhythmia recurrence in patients with idiopathic ventricular fibrillation: results from a single centre long-term follow-up over 20 years. Europace. 2016;18:718-25.
5. Sinner MF, Porthan K, Noseworthy PA, et al. A meta-analysis of genome-wide association studies of the electrocardiographic early repolarization pattern. Heart Rhythm. 2012;9:1627-34.

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 (1, 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 (3-5).**
IIa	**C-LD**	**3. In patients with short QT syndrome and recurrent sustained VA, treatment with quinidine can be useful (3, 5, 6).**
IIa	**C-LD**	**4. In patients with short QT syndrome and VT/VF storm, isoproterenol infusion can be effective (7).**
IIb	**C-EO**	**5. In patients with short QT syndrome, genetic testing may be considered to facilitate screening of first-degree relatives (4).**

References

1. Dhutia H, Malhotra A, Parpia S, et al. The prevalence and significance of a short QT interval in 18,825 low-risk individuals including athletes. Br J Sports Med. 2016;50:124-9.
2. Gollob MH, Redpath CJ, Roberts JD. The short QT syndrome: proposed diagnostic criteria. J Am Coll Cardiol. 2011;57:802-12.
3. Giustetto C, Schimpf R, Mazzanti A, et al. Long-term follow-up of patients with short QT syndrome. J Am Coll Cardiol. 2011;58:587-95.

4. Mazzanti A, Kanthan A, Monteforte N, et al. Novel insight into the natural history of short QT syndrome. J Am Coll Cardiol. 2014;63:1300-8.
5. Villafane J, Atallah J, Gollob MH, et al. Long-term follow-up of a pediatric cohort with short QT syndrome. J Am Coll Cardiol. 2013;61:1183-91.
6. Giustetto C, Di MF, Wolpert C, et al. Short QT syndrome: clinical findings and diagnostic-therapeutic implications. Eur Heart J. 2006;27:2440-7.
7. Bun SS, Maury P, Giustetto C, et al. Electrical storm in short-QT syndrome successfully treated with Isoproterenol. J Cardiovasc Electrophysiol. 2012;23:1028-30.

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**	**Recommendation**
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 (1, 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 (3, 4).**

References

1. Gill JS, Blaszyk K, Ward DE, et al. Verapamil for the suppression of idiopathic ventricular tachycardia of left bundle branch block-like morphology. Am Heart J. 1993;126:1126-33.
2. Gill JS, Ward DE, Camm AJ. Comparison of verapamil and diltiazem in the suppression of idiopathic ventricular tachycardia. Pacing Clin Electrophysiol. 1992;15:2122-6.
3. Kontos MC, Diercks DB, Ho PM, et al. Treatment and outcomes in patients with myocardial infarction treated with acute beta-blocker therapy: results from the American College of Cardiology's NCDR(R). Am Heart J. 2011;161:864-70.
4. Levine JH, Massumi A, Scheinman MM, et al. Intravenous amiodarone for recurrent sustained hypotensive ventricular tachyarrhythmias. Intravenous Amiodarone Multicenter Trial Group. J Am Coll Cardiol. 1996;27:67-75.

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 (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 (1-3).**

References

1. Tada H, Ito S, Naito S, et al. Idiopathic ventricular arrhythmia arising from the mitral annulus: a distinct subgroup of idiopathic ventricular arrhythmias. J Am Coll Cardiol. 2005;45:877-86.

2. Yamada T, Litovsky SH, Kay GN. The left ventricular ostium: an anatomic concept relevant to idiopathic ventricular arrhythmias. Circ Arrhythm Electrophysiol. 2008;1:396-404.
3. Yamada T, Maddox WR, McElderry HT, et al. Radiofrequency catheter ablation of idiopathic ventricular arrhythmias originating from intramural foci in the left ventricular outflow tract: efficacy of sequential versus simultaneous unipolar catheter ablation. Circ Arrhythm Electrophysiol. 2015;8:344-52.

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 (1-5).**

References

1. Ban JE, Lee HS, Lee DI, et al. Electrophysiological characteristics related to outcome after catheter ablation of idiopathic ventricular arrhythmia originating from the papillary muscle in the left ventricle. Korean Circ J. 2013;43:811-8.
2. Crawford T, Mueller G, Good E, et al. Ventricular arrhythmias originating from papillary muscles in the right ventricle. Heart Rhythm. 2010;7:725-30.
3. Doppalapudi H, Yamada T, McElderry HT, et al. Ventricular tachycardia originating from the posterior papillary muscle in the left ventricle: a distinct clinical syndrome. Circ Arrhythm Electrophysiol. 2008;1:23-9.
4. Yamada T, Doppalapudi H, McElderry HT, et al. Electrocardiographic and electrophysiological characteristics in idiopathic ventricular arrhythmias originating from the papillary muscles in the left ventricle: relevance for catheter ablation. Circ Arrhythm Electrophysiol. 2010;3:324-31.
5. Yokokawa M, Good E, Desjardins B, et al. Predictors of successful catheter ablation of ventricular arrhythmias arising from the papillary muscles. Heart Rhythm. 2010;7:1654-9.

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 (1-3).**
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 (3-6).**
IIa	**C-LD**	**3. In patients with recurrent verapamil-sensitive idiopathic LVT, chronic therapy with oral verapamil can be useful (7-10).**

References

1. Lin D, Hsia HH, Gerstenfeld EP, et al. Idiopathic fascicular left ventricular tachycardia: linear ablation lesion strategy for noninducible or nonsustained tachycardia. Heart Rhythm. 2005;2:934-9.
2. Liu Y, Fang Z, Yang B, et al. Catheter ablation of fascicular ventricular tachycardia: long-term clinical outcomes and mechanisms of recurrence. Circ Arrhythm Electrophysiol. 2015;8:1443-51.
3. Nogami A, Naito S, Tada H, et al. Demonstration of diastolic and presystolic Purkinje potentials as critical potentials in a macroreentry circuit of verapamil-sensitive idiopathic left ventricular tachycardia. J Am Coll Cardiol. 2000;36:811-23.
4. Belhassen B, Rotmensch HH, Laniado S. Response of recurrent sustained ventricular tachycardia to verapamil. Br Heart J. 1981;46:679-82.
5. German LD, Packer DL, Bardy GH, et al. Ventricular tachycardia induced by atrial stimulation in patients without symptomatic cardiac disease. Am J Cardiol. 1983;52:1202-7.

6. Tsuchiya T, Okumura K, Honda T, et al. Effects of verapamil and lidocaine on two components of the re-entry circuit of verapamil-senstitive idiopathic left ventricular tachycardia. J Am Coll Cardiol. 2001;37:1415-21.
7. Anderson JH, Tester DJ, Will ML, et al. Whole-exome molecular autopsy after exertion-related sudden unexplained death in the young. Circ Cardiovasc Genet. 2016;9:259-65.
8. Ohe T, Shimomura K, Aihara N, et al. Idiopathic sustained left ventricular tachycardia: clinical and electrophysiologic characteristics. Circulation. 1988;77:560-8.
9. Snyder C, Bishara J, Darling R, et al. Verapamil-sensitive ventricular tachycardia in an infant. Congenit Heart Dis. 2006;1:124-6.
10. Wang JD, Fu YC, Jan SL, et al. Verapamil sensitive idiopathic ventricular tachycardia in an infant. Jpn Heart J. 2003;44:667-71.

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 (1-8).**
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 (9-13).**
I	**B-NR**	**3. For patients with recurrent episodes of idiopathic VF initiated by PVCs with a consistent QRS morphology, catheter ablation is useful (11, 14).**

References

1. Anderson JH, Tester DJ, Will ML, et al. Whole-exome molecular autopsy after exertion-related sudden unexplained death in the young. Circ Cardiovasc Genet. 2016;9:259-65.
2. Dalal A, Czosek RJ, Kovach J, et al. Clinical presentation of pediatric patients at risk for sudden cardiac arrest. J Pediatr. 2016;177:191-6.
3. Kumar S, Peters S, Thompson T, et al. Familial cardiological and targeted genetic evaluation: low yield in sudden unexplained death and high yield in unexplained cardiac arrest syndromes. Heart Rhythm. 2013;10:1653-60.
4. Linzer M, Pritchett EL, Pontinen M, et al. Incremental diagnostic yield of loop electrocardiographic recorders in unexplained syncope. Am J Cardiol. 1990;66:214-9.
5. Solomon SD, Zelenkofske S, McMurray JJ, et al. Sudden death in patients with myocardial infarction and left ventricular dysfunction, heart failure, or both. N Engl J Med. 2005;352:2581-8.
6. Tester DJ, Medeiros-Domingo A, Will ML, et al. Unexplained drownings and the cardiac channelopathies: a molecular autopsy series. Mayo Clin Proc. 2011;86:941-7.
7. Tzimas I, Zingraf JC, Bajanowski T, et al. The role of known variants of KCNQ1, KCNH2, KCNE1, SCN5A, and NOS1AP in water-related deaths. Int J Legal Med. 2016;130:1575-9.
8. Wang D, Shah KR, Um SY, et al. Cardiac channelopathy testing in 274 ethnically diverse sudden unexplained deaths. Forensic Sci Int. 2014;237:90-9.
9. Conte G, Caputo ML, Regoli F, et al. True idiopathic ventricular fibrillation in out-of-hospital cardiac arrest survivors in the Swiss Canton Ticino: prevalence, clinical features, and long-term follow-up. Europace. 2017;19:259-66.
10. Frommeyer G, Dechering DG, Kochhauser S, et al. Long-time "real-life" performance of the subcutaneous ICD in patients with electrical heart disease or idiopathic ventricular fibrillation. J Interv Card Electrophysiol. 2016;47:185-8.
11. Haïssaguerre M, Shoda M, Jais P, et al. Mapping and ablation of idiopathic ventricular fibrillation. Circulation. 2002;106:962-7.
12. Knecht S, Sacher F, Wright M, et al. Long-term follow-up of idiopathic ventricular fibrillation ablation: a multicenter study. J Am Coll Cardiol. 2009;54:522-8.

13. Leenhardt A, Glaser E, Burguera M, et al. Short-coupled variant of torsade de pointes. A new electrocardiographic entity in the spectrum of idiopathic ventricular tachyarrhythmias. Circulation. 1994;89:206-15.
14. Haïssaguerre M, Shah DC, Jais P, et al. Role of Purkinje conducting system in triggering of idiopathic ventricular fibrillation. Lancet. 2002;359:677-8.

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 (1, 2).**
IIa	**B-NR**	**2. In patients with PVC-induced cardiomyopathy, pharmacologic treatment (e.g., beta blocker, amiodarone) is reasonable to reduce recurrent arrhythmias, and improve symptoms and LV function (3, 4).**

References

1. Haïssaguerre M, Shah DC, Jais P, et al. Role of Purkinje conducting system in triggering of idiopathic ventricular fibrillation. Lancet. 2002; 359:677-8.
2. Haïssaguerre M, Shoda M, Jais P, et al. Mapping and ablation of idiopathic ventricular fibrillation. Circulation. 2002;106:962-7.
3. Lee GK, Klarich KW, Grogan M, et al. Premature ventricular contraction-induced cardiomyopathy: a treatable condition. Circ Arrhythm Electrophysiol. 2012;5:229-36.
4. Singh SN, Fletcher RD, Fisher SG, et al. Amiodarone in patients with congestive heart failure and asymptomatic ventricular arrhythmia. Survival Trial of Antiarrhythmic Therapy in Congestive Heart Failure. N Engl J Med. 1995;333:77-82.

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 (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 (2, 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 (4, 5).**

References

1. Seth R, Moss AJ, McNitt S, et al. Long QT syndrome and pregnancy. J Am Coll Cardiol. 2007;49:1092-8.
2. Jeejeebhoy FM, Zelop CM, Lipman S, et al. Cardiac arrest in pregnancy: a scientific statement from the American Heart Association. Circulation. 2015;132:1747-73.
3. Vanden Hoek TL, Morrison LJ, Shuster M, et al. Part 12: cardiac arrest in special situations: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care. Circulation. 2010;122(suppl 3):S829-61.
4. Natale A, Davidson T, Geiger MJ, et al. Implantable cardioverter-defibrillators and pregnancy: a safe combination? Circulation. 1997;96:2808-12.
5. Colletti PM, Lee KH, Elkayam U. Cardiovascular imaging of the pregnant patient. Am J Roentgenol. 2013;200:515-21.

9.2. Older Patients With Comorbidities

Recommendation for Older Patients With Comorbidities See Systematic Review Report (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 (1).**

SR indicates systematic review.

Reference

1. Kusumoto FM, Bailey KR, Chaouki AS, et al. Systematic review for the 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2017. In Press.

9.3. Medication-Induced Arrhythmias

Recommendations for Medication-Induced Arrhythmias References that support the recommendations are summarized in Online Data Supplement 52 and 53.		
Digoxin		
COR	**LOE**	**Recommendation**
I	**B-NR**	**1. Administration of digoxin antibodies is recommended for patients who present with sustained VA potentially due to digoxin toxicity (1, 2).**
Medication-Induced QT Prolongation and Torsades de Pointes		
COR	**LOE**	**Recommendations**
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 (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 (4, 5).**
I	**C-LD**	**4. For patients with torsades de pointes associated with acquired QT prolongation, potassium repletion to 4.0 mmol per L or more and magnesium repletion to normal values (e.g., ≥2.0 mmol/L) are beneficial (6, 7).**
Sodium Channel Blocker–Related Toxicity		
COR	**LOE**	**Recommendations**
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 (8, 9).**
III: Harm	**B-NR**	**6. In patients with congenital or acquired long QT syndrome, QT-prolonging medications are potentially harmful (10).**

Table 10

References

1. Antman EM, Wenger TL, Butler VP Jr, et al. Treatment of 150 cases of life-threatening digitalis intoxication with digoxin-specific Fab antibody fragments. Final report of a multicenter study. Circulation. 1990;81:1744-52.
2. Chan BS, Buckley NA. Digoxin-specific antibody fragments in the treatment of digoxin toxicity. Clin Toxicol. 2014;52:824-36.
3. Keren A, Tzivoni D, Gavish D, et al. Etiology, warning signs and therapy of torsade de pointes. A study of 10 patients. Circulation. 1981;64:1167-74.
4. Tzivoni D, Banai S, Schuger C, et al. Treatment of torsade de pointes with magnesium sulfate. Circulation. 1988;77:392-7.
5. Kannankeril P, Roden DMDarbar D. Drug-induced long QT syndrome. Pharmacol Rev. 2010;62:760-81.
6. Choy AM, Lang CC, Chomsky DM, et al. Normalization of acquired QT prolongation in humans by intravenous potassium. Circulation. 1997;96:2149-54.
7. Yang T, Roden DM. Extracellular potassium modulation of drug block of IKr. Implications for torsade de pointes and reverse use-dependence. Circulation. 1996;93:407-11.
8. Hellestrand KJ, Burnett PJ, Milne JR, et al. Effect of the antiarrhythmic agent flecainide acetate on acute and chronic pacing thresholds. Pacing Clin Electrophysiol. 1983;6:892-9.

9. Echt DS, Black JN, Barbey JT, et al. Evaluation of antiarrhythmic drugs on defibrillation energy requirements in dogs. Sodium channel block and action potential prolongation. Circulation. 1989;79:1106-17.
10. Schwartz PJ, Woosley RL. Predicting the unpredictable: drug-induced QT prolongation and torsades de pointes. J Am Coll Cardiol. 2016;67:1639-50.

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 (1-6).**
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 (3, 7-12).**
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 (13-17).**
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 (13-17).**
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 (18, 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 (1, 19, 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 (21-25).**
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 (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 (5, 16, 27-29).**
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 (14-16, 20).**
III: Harm	B-NR	**11. In patients with adult congenital heart disease who have asymptomatic VA, prophylactic antiarrhythmic therapy with class Ic medications (i.e., flecainide, propafenone) or amiodarone is potentially harmful (30-32).**

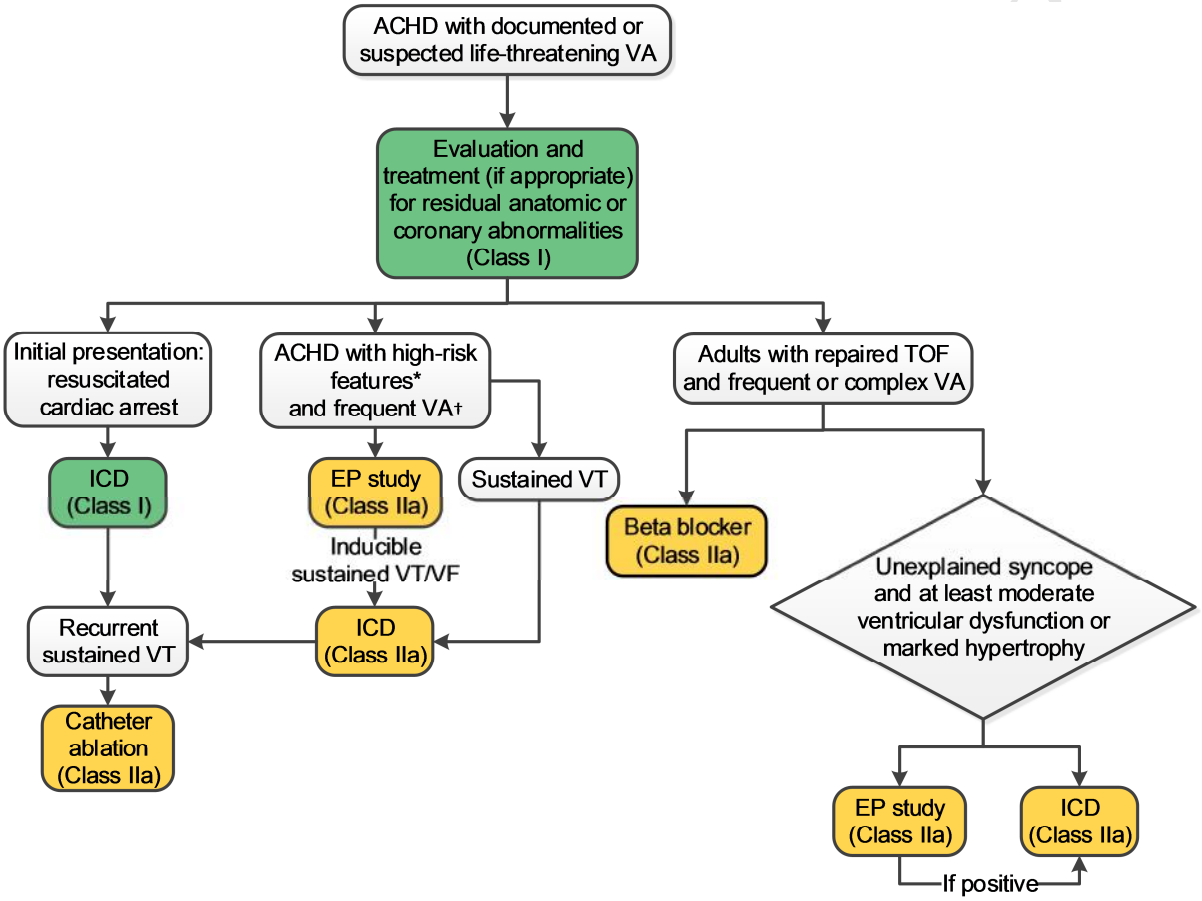
Table 11 and Figure 16

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 (44, 47, 57-62)	2%–6%	<1.5%	Ventricular pacing RV dilatation Pulmonary hypertension *NKX2.5* gene
VSD (27, 44, 47, 57-63)	3%–18%	<3%	
Moderate complexity			
Tetralogy of Fallot (1, 2, 5, 6, 28, 34, 36, 44, 46, 47, 54-56, 62-65)	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 (27, 44, 56)	10%–34%	3%–20%	Unexplained syncope Severe LV hypertrophy Aortic stenosis mean pressure gradient >40 mm Hg Ventricular dysfunction
Coarctation of aorta (28, 29, 44, 46, 56, 62)	2%	2%	Aneurysm at repair site Aortic stenosis Systemic hypertension Premature coronary artery disease
Ebstein's anomaly (45, 47, 55)	2%	3%–6%	Cardiomegaly Atrial fibrillation Wide complex tachycardia Mitral regurgitation Dilated RVOT
Severe complexity			
Transposition of the great arteries (27, 44-48, 51, 55, 56, 62)			Atrial switch Mustard repair Prior VSD closure Unexplained syncope Atrial tachycardia Coronary orifice stenosis Systemic ventricular dysfunction Severe tricuspid regurgitation
Atrial switch	2%	3%–9.5%	
Arterial switch	2%	1%	
cc-TGA	10%	17%–25%	
Truncus arteriosus (66, 67)	10%	4%	Multiple surgical repairs Coronary anomalies Ventricular dysfunction and/or hypertrophy
Fontan repair for univentricular physiology* (27, 37, 44, 45, 47, 55, 68)	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.

References

1. Diller GP, Kempny A, Liodakis E, et al. Left ventricular longitudinal function predicts life-threatening ventricular arrhythmia and death in adults with repaired tetralogy of Fallot. Circulation. 2012;125:2440-6.

2. Gatzoulis MA, Till JA, Somerville J, et al. Mechanoelectrical interaction in tetralogy of Fallot. QRS prolongation relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death. Circulation. 1995;92:231-7.
3. Harrison DA, Harris L, Siu SC, et al. Sustained ventricular tachycardia in adult patients late after repair of tetralogy of Fallot. J Am Coll Cardiol. 1997;30:1368-73.
4. Knauth AL, Gauvreau K, Powell AJ, et al. Ventricular size and function assessed by cardiac MRI predict major adverse clinical outcomes late after tetralogy of Fallot repair. Heart. 2008;94:211-6.
5. Koyak Z, de Groot JR, Bouma BJ, et al. Sudden cardiac death in adult congenital heart disease: can the unpredictable be foreseen? Europace. 2017;19:401-6.
6. Koyak Z, Harris L, de Groot JR, et al. Sudden cardiac death in adult congenital heart disease. Circulation. 2012;126:1944-54.
7. Adamson L, Vohra HA, Haw MP. Does pulmonary valve replacement post repair of tetralogy of Fallot improve right ventricular function? Interact Cardiovasc Thorac Surg. 2009;9:520-7.
8. Deal BJ, Scagliotti D, Miller SM, et al. Electrophysiologic drug testing in symptomatic ventricular arrhythmias after repair of tetralogy of Fallot. Am J Cardiol. 1987;59:1380-5.
9. Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS Expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Heart Rhythm. 2014;11:e102-65.
10. Sabate Rotes A, Connolly HM, Warnes CA, et al. Ventricular arrhythmia risk stratification in patients with tetralogy of Fallot at the time of pulmonary valve replacement. Circ Arrhythm Electrophysiol. 2015;8:110-6.
11. Therrien J, Provost Y, Merchant N, et al. Optimal timing for pulmonary valve replacement in adults after tetralogy of Fallot repair. Am J Cardiol. 2005;95:779-82.
12. Therrien J, Siu SC, Harris L, et al. Impact of pulmonary valve replacement on arrhythmia propensity late after repair of tetralogy of Fallot. Circulation. 2001;103:2489-94.
13. Kella DK, Merchant FM, Veledar E, et al. Lesion-specific differences for implantable cardioverter defibrillator therapies in adults with congenital heart disease. Pacing Clin Electrophysiol. 2014;37:1492-8.
14. Koyak Z, de Groot JR, Van Gelder IC, et al. Implantable cardioverter defibrillator therapy in adults with congenital heart disease: who is at risk of shocks? Circ Arrhythm Electrophysiol. 2012;5:101-10.
15. Santharam S, Hudsmith L, Thorne S, et al. Long-term follow-up of implantable cardioverter-defibrillators in adult congenital heart disease patients: indications and outcomes. Europace. 2017;19:407-13.
16. Vehmeijer JT, Brouwer TF, Limpens J, et al. Implantable cardioverter-defibrillators in adults with congenital heart disease: a systematic review and meta-analysis. Eur Heart J. 2016;37:1439-48.
17. Yap SC, Roos-Hesselink JW, Hoendermis ES, et al. Outcome of implantable cardioverter defibrillators in adults with congenital heart disease: a multi-centre study. Eur Heart J. 2007;28:1854-61.
18. Chandar JS, Wolff GS, Garson A Jr, et al. Ventricular arrhythmias in postoperative tetralogy of Fallot. Am J Cardiol. 1990;65:655-61.
19. Khairy P, Landzberg MJ, Gatzoulis MA, et al. Value of programmed ventricular stimulation after tetralogy of Fallot repair: a multicenter study. Circulation. 2004;109:1994-2000.
20. Khairy P, Harris L, Landzberg MJ, et al. Implantable cardioverter-defibrillators in tetralogy of Fallot. Circulation. 2008;117:363-70.
21. Kapel GF, Reichlin T, Wijnmaalen AP, et al. Re-entry using anatomically determined isthmuses: a curable ventricular tachycardia in repaired congenital heart disease. Circ Arrhythm Electrophysiol. 2015;8:102-9.
22. Kapel GF, Reichlin T, Wijnmaalen AP, et al. Left-sided ablation of ventricular tachycardia in adults with repaired tetralogy of Fallot: a case series. Circ Arrhythm Electrophysiol. 2014;7:889-97.
23. Kapel GF, Sacher F, Dekkers OM, et al. Arrhythmogenic anatomical isthmuses identified by electroanatomical mapping are the substrate for ventricular tachycardia in repaired tetralogy of Fallot. Eur Heart J. 2017;38:268-76.
24. van Zyl M, Kapa S, Padmanabhan D, et al. Mechanism and outcomes of catheter ablation for ventricular tachycardia in adults with repaired congenital heart disease. Heart Rhythm. 2016;13:1449-54.
25. Zeppenfeld K, Schalij MJ, Bartelings MM, et al. Catheter ablation of ventricular tachycardia after repair of congenital heart disease: electroanatomic identification of the critical right ventricular isthmus. Circulation. 2007;116:2241-52.
26. Khairy P, Harris L, Landzberg MJ, et al. Sudden death and defibrillators in transposition of the great arteries with intra-atrial baffles: a multicenter study. Circ Arrhythm Electrophysiol. 2008;1:250-7.
27. Engelings CC, Helm PC, Abdul-Khaliq H, et al. Cause of death in adults with congenital heart disease - an analysis of the German National Register for Congenital Heart Defects. Int J Cardiol. 2016;211:31-6.

28. Gallego P, Gonzalez AE, Sanchez-Recalde A, et al. Incidence and predictors of sudden cardiac arrest in adults with congenital heart defects repaired before adult life. Am J Cardiol. 2012;110:109-17.
29. Tutarel O, Kempny A, Alonso-Gonzalez R, et al. Congenital heart disease beyond the age of 60: emergence of a new population with high resource utilization, high morbidity, and high mortality. Eur Heart J. 2014;35:725-32.
30. Fish FA, Gillette PC, Benson DW Jr. Proarrhythmia, cardiac arrest and death in young patients receiving encainide and flecainide. The Pediatric Electrophysiology Group. J Am Coll Cardiol. 1991;18:356-65.
31. Stan MN, Sathananthan M, Warnes C, et al. Amiodarone-induced thyrotoxicosis in adults with congenital heart disease-clinical presentation and response to therapy. Endocr Pract. 2014;21:33-40.
32. Thorne SA, Barnes I, Cullinan P, et al. Amiodarone-associated thyroid dysfunction: risk factors in adults with congenital heart disease. Circulation. 1999;100:149-54.

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. n 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 (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 (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 (1-4, 6-8).**

References

1. Bardy GH, Smith WM, Hood MA, et al. An entirely subcutaneous implantable cardioverter-defibrillator. N Engl J Med. 2010;363:36-44.
2. Weiss R, Knight BP, Gold MR, et al. Safety and efficacy of a totally subcutaneous implantable-cardioverter defibrillator. Circulation. 2013;128:944-53.
3. Lambiase PD, Barr C, Theuns DA, et al. Worldwide experience with a totally subcutaneous implantable defibrillator: early results from the EFFORTLESS S-ICD Registry. Eur Heart J. 2014;35:1657-65.
4. Burke MC, Gold MR, Knight BP, et al. Safety and efficacy of the totally subcutaneous implantable defibrillator: 2-year results from a pooled analysis of the IDE study and EFFORTLESS registry. J Am Coll Cardiol. 2015;65:1605-15.
5. El-Chami MF, Levy M, Kelli HM, et al. Outcome of subcutaneous implantable cardioverter defibrillator implantation in patients with end-stage renal disease on dialysis. J Cardiovasc Electrophysiol. 2015;26:900-4.
6. de Bie MK, Thijssen J, van Rees JB, et al. Suitability for subcutaneous defibrillator implantation: results based on data from routine clinical practice. Heart. 2013;99:1018-23.
7. Olde Nordkamp LR, Dabiri AL, Boersma LV, et al. The entirely subcutaneous implantable cardioverter-defibrillator: initial clinical experience in a large Dutch cohort. J Am Coll Cardiol. 2012;60:1933-9.
8. Köbe J, Reinke F, Meyer C, et al. Implantation and follow-up of totally subcutaneous versus conventional implantable cardioverter-defibrillators: a multicenter case-control study. Heart Rhythm. 2013;10:29-36.

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 (1-4).**
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, wearable cardioverter-defibrillator may be reasonable (1-5).**

References

1. Chung MK. The role of the wearable cardioverter defibrillator in clinical practice. Cardiol Clin. 2014;32:253-70.
2. Chung MK, Szymkiewicz SJ, Shao M, et al. Aggregate national experience with the wearable cardioverter-defibrillator: event rates, compliance, and survival. J Am Coll Cardiol. 2010;56:194-203.
3. Klein HU, Meltendorf U, Reek S, et al. Bridging a temporary high risk of sudden arrhythmic death. Experience with the wearable cardioverter defibrillator (WCD). Pacing Clin Electrophysiol. 2010;33:353-67.
4. Piccini JP Sr, Allen LA, Kudenchuk PJ, et al. Wearable cardioverter-defibrillator therapy for the prevention of sudden cardiac death: a science advisory from the American Heart Association. Circulation. 2016;133:1715-27.
5. Ellenbogen KA, Koneru JN, Sharma PS, et al. Benefit of the wearable cardioverter-defibrillator in protecting patients after implantable-cardioverter defibrillator explant. Results from the National Registry. JACC Clin Electrophysiol. 2017;3:243-50.

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 (1-3).**
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 (4-6).**

References

1. Blanck Z, Dhala A, Deshpande S, et al. Bundle branch reentrant ventricular tachycardia: cumulative experience in 48 patients. J Cardiovasc Electrophysiol. 1993;4:253-62.
2. Lopera G, Stevenson WG, Soejima K, et al. Identification and ablation of three types of ventricular tachycardia involving the his-purkinje system in patients with heart disease. J Cardiovasc Electrophysiol. 2004;15:52-8.
3. Mehdirad AA, Keim S, Rist K, et al. Long-term clinical outcome of right bundle branch radiofrequency catheter ablation for treatment of bundle branch reentrant ventricular tachycardia. Pacing Clin Electrophysiol. 1995;18:2135-43.

4. Dinov B, Fiedler L, Schonbauer R, et al. Outcomes in catheter ablation of ventricular tachycardia in dilated nonischemic cardiomyopathy compared with ischemic cardiomyopathy: results from the Prospective Heart Centre of Leipzig VT (HELP-VT) Study. Circulation. 2014;129:728-36.
5. Tanner H, Hindricks G, Volkmer M, et al. Catheter ablation of recurrent scar-related ventricular tachycardia using electroanatomical mapping and irrigated ablation technology: results of the prospective multicenter Euro-VT-study. J Cardiovasc Electrophysiol. 2010;21:47-53.
6. Marchlinski FE, Haffajee CI, Beshai JF, et al. Long-term success of irrigated radiofrequency catheter ablation of sustained ventricular tachycardia: post-approval THERMOCOOL VT trial. J Am Coll Cardiol. 2016;67:674-83.

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 (1, 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 (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 (4-7).**
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 (8).**

References

1. Basso C, Burke M, Fornes P, et al. Guidelines for autopsy investigation of sudden cardiac death. Virchows Arch. 2008;452:11-8.
2. de Noronha SV, Behr ER, Papadakis M, et al. The importance of specialist cardiac histopathological examination in the investigation of young sudden cardiac deaths. Europace. 2014;16:899-907.
3. Wilde AA, Behr ER. Genetic testing for inherited cardiac disease. Nat Rev Cardiol. 2013;10:571-83.
4. Tester DJ, Medeiros-Domingo A, Will ML, et al. Cardiac channel molecular autopsy: insights from 173 consecutive cases of autopsy-negative sudden unexplained death referred for postmortem genetic testing. Mayo Clin Proc. 2012;87:524-39.
5. Tang Y, Stahl-Herz J, Sampson BA. Molecular diagnostics of cardiovascular diseases in sudden unexplained death. Cardiovasc Pathol. 2014;23:1-4.
6. Bagnall RD, Das KJ, Duflou J, et al. Exome analysis-based molecular autopsy in cases of sudden unexplained death in the young. Heart Rhythm. 2014;11:655-62.
7. Bagnall RD, Weintraub RG, Ingles J, et al. A prospective study of sudden cardiac death among children and young adults. N Engl J Med. 2016;374:2441-52.
8. Semsarian C, Ingles J. Molecular autopsy in victims of inherited arrhythmias. J Arrhythmia. 2016;32:359-65.

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.**

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 (1-5).**
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 (1-5).**

References

1. Lewis KB, Stacey D, Matlock DD. Making decisions about implantable cardioverter-defibrillators from implantation to end of life: an integrative review of patients' perspectives. Patient. 2014;7:243-60.
2. Stewart GC, Weintraub JR, Pratibhu PP, et al. Patient expectations from implantable defibrillators to prevent death in heart failure. J Cardiac Fail. 2010;16:106-13.
3. Hauptman PJ, Chibnall JT, Guild C, et al. Patient perceptions, physician communication, and the implantable cardioverter-defibrillator. JAMA Intern Med. 2013;173:571-7.
4. Ottenberg AL, Mueller PS, Topazian RJ, et al. "It's not broke, so let's not try to fix it": why patients decline a cardiovascular implantable electronic device. Pacing Clin Electrophysiol. 2014;37:1306-14.
5. Yuhas J, Mattocks K, Gravelin L, et al. Patients' attitudes and perceptions of implantable cardioverter-defibrillators: potential barriers to appropriate primary prophylaxis. Pacing Clin Electrophysiol. 2012;35:1179-87.

15. Cost and Value Considerations

The key principles of value assessment as part of clinical practice guidelines have been discussed in detail (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 (2).

Economic assessments of alternative management strategies for VA and prevention of SCD have primarily evaluated ICDs, including several RCTs (3-7) and observational studies (8, 9), and simulation models (10-14). 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 (4-7). 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 (11). Because the framework proposed for assessing value in ACC/AHA clinical practice guidelines uses benchmarks based on lifetime estimates (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 (15) or an acute MI (16, 17). An analysis of individual patient level data from 3 secondary prevention trials (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 (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 (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 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 (14).

Figure 17. Incremental Cost-Effectiveness of ICD by Years of Life Added* (Example)

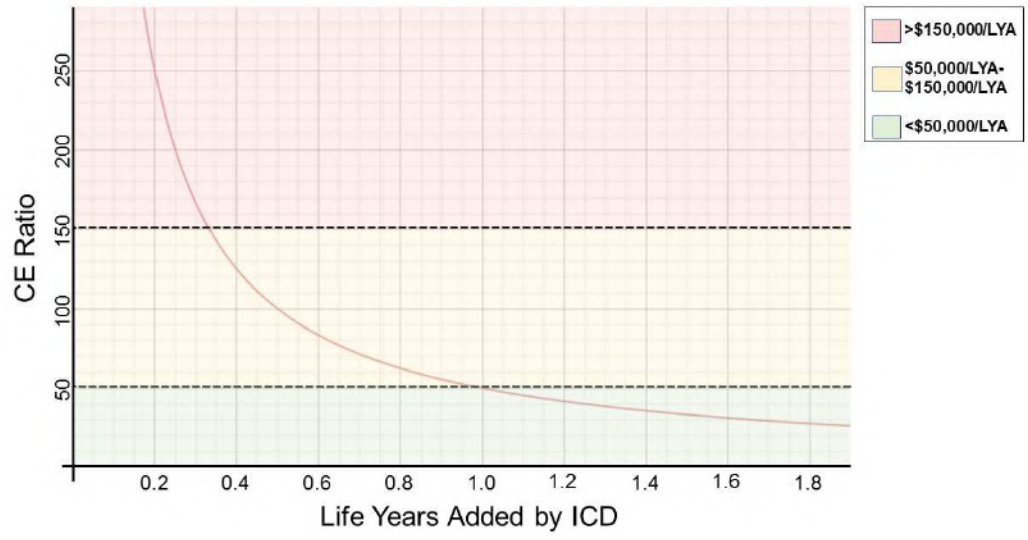


*Figure based on formula: Incremental cost-effectiveness ratio = $50,000/QALYs.
CE indicated cost effectiveness; ICD, implantable cardioverter-defibrillator; LYA, life year added; and QALYs, quality-adjusted life-years.

References

1. Anderson JL, Heidenreich PA, Barnett PG, et al. ACC/AHA statement on cost/value methodology in clinical practice guidelines and performance measures: a report of the American College of Cardiology/American Heart Association Task Force on Performance Measures and Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:2304-22.
2. Sanders GD, Neumann PJ, Basu A, et al. Recommendations for conduct, methodological practices, and reporting of cost-effectiveness analyses: second panel on cost-effectiveness in health and medicine. JAMA. 2016;316:1093-103.
3. Mushlin AI, Hall J, Zwanziger J, et al. The cost-effectiveness of automatic implantable cardiac defibrillators. Results from MADIT. Circulation. 1998;97:2129-35.
4. O'Brien BJ, Connolly SJ, Goeree R, et al. Cost-effectiveness of the implantable cardioverter-defibrillator. Results from the Canadian Implantable Defibrillator Study (CIDS). Circulation. 2001;103:1416-21.
5. Larson G, Hallstrom A, McAnulty J, et al. Cost-effectiveness of the implantable cardioverter-defibrillator versus antiarrhythmic drugs in survivors of serious ventricular tachyarrhythmias. Results of the Antiarrhythmics Versus Implantable Defibrillators (AVID) Economic Analysis Substudy. Circulation. 2002;105:2049-57.
6. Zwanziger J, Hall WJ, Dick AW, et al. The cost-effectiveness of implantable cardioverter-defibrillators. Results from the Multicenter Automatic Defibrillator Implantation Trial (MADIT)-II. J Am Coll Cardiol. 2006;47:2310-8.
7. Mark DB, Nelson CL, Anstrom KJ, et al. Cost-effectiveness of defibrillator therapy or amiodarone in chronic stable heart failure. Results from the Sudden Cardiac Death in Heart Failure Trial (SCD-HeFT). Circulation. 2006;114:135-42.
8. Weiss JP, Saynina O, McDonald KM, et al. Effectiveness and cost-effectiveness of implantable cardioverter defibrillators in the treatment of ventricular arrhythmias among Medicare beneficiaries. Am J Med. 2002;112:519-27.
9. Al-Khatib SM, Anstrom KJ, Eisenstein EL, et al. Clinical and economic implications of the Multicenter Automatic Defibrillator Implantation Trial-II. Ann Intern Med. 2005;142:593-600.
10. Buxton M, Caine N, Chase D, et al. A review of the evidence on the effects and costs of implantable cardioverter defibrillator therapy in different patient groups, and modelling of cost-effectiveness and cost-utility for these groups in a UK context. Health Technol Assess. 2006;10:iii-iv, ix-xi, 1-164.
11. Sanders GD, Hlatky MA, Owens DK. Cost effectiveness of implantable cardioverter-defibrillators. N Engl J Med. 2005;353:1471-80.
12. Smith T, Jordaens L, Theuns DAMJ, et al. The cost-effectiveness of primary prophylactic implantable defibrillator therapy in patients with ischaemic or non-ischaemic heart disease: a European analysis. Eur Heart J. 2013;34:211-9.
13. Cowie MR, Marshall D, Drummond M, et al. Lifetime cost-effectiveness of prophylactic implantation of cardioverter defibrillator in patients with reduced left ventricular systolic function: results of Markov modelling in a European population. Europace. 2008;11:716-26.
14. Goldenberg I, Moss AJ, Maron BJ, et al. Cost-effectiveness of implanted defibrillators in young people with inherited cardiac arrhythmias. Ann Noninvasive Electrocardiol. 2005;10(suppl):67-83.
15. Bigger JT. Prophylactic use of implanted cardiac defibrillators in patients at high risk for ventricular arrhythmias after coronary-artery bypass graft surgery. N Engl J Med. 1997;337:1569-75.
16. Hohnloser SH, Kuck KH, Dorian P, et al. Prophylactic use of an implantable cardioverter-defibrillator after acute myocardial infarction. N Engl J Med. 2004;351:2481-8.
17. Steinbeck G, Andresen D, Seidl K, et al. Defibrillator implantation early after myocardial infarction. N Engl J Med. 2009 361:1427-36.
18. Connolly SJ, Hallstrom AP, Cappato R, et al. Meta-analysis of the implantable cardioverter defibrillator secondary prevention trials. Eur Heart J. 2000;21:2071-8.
19. Owens DK, Sanders GD, Heidenreich PA, et al. Effect of risk stratification on cost-effectiveness of the implantable cardioverter defibrillator. Am Heart J. 2002;144:440-8.
20. Ezekowitz JA, Rowe BH, Dryden DM, et al. Systematic review: implantable cardioverter defibrillators for adults with left ventricular systolic dysfunction. Ann Intern Med. 2007;147:251-62.

16. Quality of Life

ICD implantation has not had a significant effect on QoL in the overall population of patients enrolled in RCTs (1-3). 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 (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.

References

1. Mark DB, Anstrom KJ, Sun JL, et al. Quality of life with defibrillator therapy or amiodarone in heart failure. N Engl J Med. 2008;359:999-1008.
2. Noyes K, Corona E, Veazie P, et al. Examination of the effect of implantable cardioverter-defibrillators on health-related quality of life: based on results from the Multicenter Automatic Defibrillator Trial-II. Am J Cardiovasc Drugs. 2009;9:393-400.
3. Passman R, Subacius H, Ruo B, et al. Implantable cardioverter defibrillators and quality of life: results from the defibrillators in nonischemic cardiomyopathy treatment evaluation study. Arch Intern Med. 2007;167:2226-32.

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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Key Words: ACC/AHA Clinical Practice Guidelines ■ acute coronary syndrome ■ ambulatory ECG monitoring ■ antiarrhythmic drug therapy ■ arrhythmogenic cardiomyopathy ■ athletes ■ cardiac electrophysiology ■ cardiac resynchronization therapy ■ cardiomyopathy ■ catheter ablation ■ congenital heart disease ■ CT imaging ■ ECG ■ echocardiography ■ electrophysiological testing ■ genetic arrhythmias ■ Guidelines ■ heart failure ■ imaging ■ implantable cardioverter-defibrillator ■ implantable and external cardioverter devices ■ medication-induced arrhythmias ■ MR imaging ■ myocardial infarction ■ premature ventricular beats ■ resuscitation ■ sarcoidosis ■ specific pathology (e.g., congenital heart disease, myocarditis, renal failure) ■ stable coronary artery disease ■ sudden cardiac arrest ■ sudden cardiac death ■ torsades de pointes ■ ventricular fibrillation ■ ventricular tachycardia.

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
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
Stephen C. Hammill	Mayo Clinic—Professor Emeritus of 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
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 ≥$5,000 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, principle 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

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
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.