



AMERICAN
COLLEGE *of*
CARDIOLOGY
FOUNDATION

American Heart
Association®



Learn and Live™

ACC/AHA/ESC Pocket Guideline

**Based on the ACC/AHA/ESC
2006 Guidelines**

**Management
of Patients With
Ventricular
Arrhythmias and
the Prevention of
Sudden Cardiac
Death**

September 2006

Special thanks to



Medtronic

**Distributed through support
from Medtronic Inc.**

**Medtronic Inc.
was not involved in the
development of this
publication and
in no way influenced
its contents.**

Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death

September 2006

ACC/AHA/ESC Writing Committee

Douglas P. Zipes, MD, MACC, FAHA, FESC, Co-Chair

A. John Camm, MD, FACC, FAHA, FESC, Co-Chair

Martin Borggrefe, MD, FESC

Alfred E. Buxton, MD, FACC, FAHA

Bernard Chaitman, MD, FACC, FAHA

Martin Fromer, MD

Gabriel Gregoratos, MD, FACC, FAHA

George Klein, MD, FACC

Arthur J. Moss, MD, FACC, FAHA

Robert J. Myerburg, MD, FACC, FAHA

Silvia G. Priori, MD, PhD, FESC

Miguel A. Quinones, MD, FACC

Dan M. Roden, MD, CM, FACC, FAHA

Michael J. Silka, MD, FACC, FAHA

Cynthia Tracy, MD, FACC, FAHA



© 2006 American College of Cardiology Foundation,
American Heart Association, Inc., and the European
Society of Cardiology.

The following material was adapted from the ACC/
AHA/ESC 2006 Guidelines for the Management of Patients
With Ventricular Arrhythmias and the Prevention of
Sudden Cardiac Death: Executive Summary (*Journal
of the American College of Cardiology* 2006; 48:1064–
1108, *Circulation* 2006; 114:1088-1132, and *European
Heart Journal* 2006; 27:2099-2140).

For a copy of the full report or published executive
summary, visit our Web sites at <http://www.acc.org>,
<http://www.americanheart.org>, or <http://www.escardio.org>
or call the ACC Resource Center at 1-800-253-4636,
ext. 694.

Contents

I. Introduction	4	Introduction
A. Prophylactic Implantable Cardioverter Device Recommendations Across Published Guidelines	9	
B. Classification of Ventricular Arrhythmias and Sudden Cardiac Death	10	
II. Incidence of Sudden Cardiac Death	11	SCD
III. Clinical Presentations of Patients With Ventricular Arrhythmias and Sudden Cardiac Death	11	Clin. Presentation
IV. General Evaluation of Patients With Documented or Suspected Ventricular Arrhythmias	12	Patient Evaluation
A. Resting Electrocardiogram	12	
B. Exercise Testing	13	
C. Ambulatory Electrocardiography	14	
D. Electrocardiogram Techniques and Measurements	14	
E. Left Ventricular Function and Imaging	15	
F. Electrophysiological Testing	16	
V. Therapies for Ventricular Arrhythmias	18	Therapies
A. Introduction	18	
B. Ablation	18	
VI. Acute Management of Specific Arrhythmias	20	Acute Management
A. Management of Cardiac Arrest	20	
B. Ventricular Tachycardia Associated With Low Troponin MI	21	
C. Sustained Monomorphic Ventricular Tachycardia	22	
D. Repetitive Monomorphic Ventricular Tachycardia	23	
E. Polymorphic Ventricular Tachycardia	23	
F. Torsades de Pointes	24	
G. Incessant Ventricular Tachycardia	26	

VII. Ventricular Arrhythmias and Sudden

Cardiac Death Related to Specific Pathology	27
A. Left Ventricular Dysfunction Due to Prior MI	27
B. Valvular Heart Disease	30
C. Congenital Heart Disease	30
D. Pericardial Diseases	32
E. Pulmonary Arterial Hypertension	32
F. Transient Arrhythmias of Reversible Cause	33

VIII. Ventricular Arrhythmias Associated With Cardiomyopathies 34

A. Dilated Cardiomyopathy (Nonischemic)	34
B. Hypertrophic Cardiomyopathy	36
C. Arrhythmogenic Right Ventricular Cardiomyopathy	38

IX. Heart Failure 39

X. Genetic Arrhythmia Syndromes 42

A. Long QT Syndrome	42
B. Brugada Syndrome	43
C. Catecholaminergic Polymorphic Ventricular Tachycardia	45

XI. Ventricular Arrhythmias and Sudden

Cardiac Death Related to Specific Populations 46

A. Athletes	46
B. Gender and Pregnancy	47
C. Elderly Patients	47
D. Patients With Implantable Cardioverter Devices	48
E. Drug-Induced Arrhythmias	49

I. Introduction

The reader should note that the recommendations, text, figures, and tables included in these pocket guidelines represent a succinct summary of the more extensive evidence base, critical evaluation, supporting text, tables, figures, and references that are included in the full-text guidelines. Readers are strongly encouraged to refer to the full-text guidelines.

Classification of Recommendations and Level of Evidence are expressed in the American College of Cardiology (ACC)/American Heart Association (AHA)/European Society of Cardiology (ESC) format as follows:

Classification of Recommendations

Class I Conditions for which there is evidence and/or general agreement that a given procedure or treatment is beneficial, useful, and effective.

Class II Conditions for which there is conflicting evidence and/or divergence of opinion about the usefulness/efficacy of a procedure or treatment.

Class IIa Weight of evidence/opinion is in favor of usefulness/efficacy.

Class IIb Usefulness/efficacy is less well established by evidence/opinion.

Class III Conditions for which there is evidence and/or general agreement that a procedure/treatment is not useful/effective and in some cases may be harmful.

Level of Evidence **Level of Evidence A** Data derived from multiple randomized clinical trials or meta-analyses.

Level of Evidence B Data derived from a single randomized trial or nonrandomized studies.

Level of Evidence C Only consensus opinion of experts, case studies, or standard-of-care.

The schema for classification of recommendations and level of evidence is summarized in *Table 1*.

Table 1. Applying Classification of Recommendations and Level of Evidence[†]

		SIZE OF TREATMENT EFFECT	
		CLASS I <i>Benefit >>> Risk</i> Procedure/Treatment SHOULD be performed/ administered	CLASS IIA <i>Benefit >> Risk</i> <i>Additional studies with focused objectives needed</i> IT IS REASONABLE to per- form procedure/administer treatment
ESTIMATE OF CERTAINTY (PRECISION) OF TREATMENT EFFECT	LEVEL A Multiple (3-5) population risk strata evaluated* General consistency of direction and magnitude of effect	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is useful/effective ■ Sufficient evidence from multiple randomized trials or meta-analyses 	<ul style="list-style-type: none"> ■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from multiple randomized trials or meta-analyses
	LEVEL B Limited (2-3) population risk strata evaluated*	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is useful/effective ■ Limited evidence from single randomized trial or nonrandomized studies 	<ul style="list-style-type: none"> ■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from single randomized trial or nonrandomized studies
	LEVEL C Very limited (1-2) population risk strata evaluated*	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is useful/effective ■ Only expert opinion, case studies, or standard-of-care 	<ul style="list-style-type: none"> ■ Recommendation in favor of treatment or procedure being useful/effective ■ Only diverging expert opinion, case studies, or standard-of-care



→	
<p>Class IIb <i>Benefit ≥ Risk</i> <i>Additional studies with broad objectives needed; additional registry data would be helpful</i> Procedure/Treatment MAY BE CONSIDERED</p>	<p>Class III <i>Risk ≥ Benefit</i> <i>No additional studies needed</i> Procedure/Treatment should NOT be performed/administered SINCE IT IS NOT HELPFUL AND MAY BE HARMFUL</p>
<ul style="list-style-type: none"> ■ Recommendation's usefulness/efficacy less well established ■ Greater conflicting evidence from multiple randomized trials or meta-analyses 	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Sufficient evidence from multiple randomized trials or meta-analyses
<ul style="list-style-type: none"> ■ Recommendation's usefulness/efficacy less well established ■ Greater conflicting evidence from single randomized trial or nonrandomized studies 	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Limited evidence from single randomized trial or nonrandomized studies
<ul style="list-style-type: none"> ■ Recommendation's usefulness/efficacy less well established ■ Only diverging expert opinion, case studies, or standard-of-care 	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Only expert opinion, case studies, or standard-of-care

* Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use.

† A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

Table 2. Inconsistencies Between ACC/AHA/ESC Guidelines for the Management of Patients With Ventricular Arrhythmias and the Prevention of SCD and Other Published ACC/AHA and ESC Guidelines With Respect to ICD Therapy for Primary Prevention to Reduce Total Mortality by a Reduction in SCD

Group addressed in recommendation	Guideline and Class of Recommendation with Level of Evidence* for Each Group		
	2005 ACC/AHA HF	2005 ESC HF	2004 ACC/AHA STEMI
LVD d/t MI, LVEF \leq 30%, NYHA II, III	<i>Class I;</i> <i>LOE: B</i>	<i>Class I;</i> <i>LOE: A</i>	<i>Class IIa;</i> <i>LOE: B</i>
LVD d/t MI, LVEF 30% to 35%, NYHA II, III	<i>Class IIa;</i> <i>LOE: B</i>	<i>Class I;</i> <i>LOE: A</i>	N/A
LVD d/t MI, LVEF 30% to 40%, NSVT, positive EP study	N/A	N/A	<i>Class I;</i> <i>LOE: B</i>
LVD d/t MI, LVEF \leq 30%, NYHA I	<i>Class IIa;</i> <i>LOE: B</i>	N/A	N/A
LVD d/t MI, LVEF \leq 31% to 35%, NYHA I	N/A	N/A	N/A
NICM, LVEF \leq 30%, NYHA II, III	<i>Class I;</i> <i>LOE: B</i>	<i>Class I;</i> <i>LOE: A</i>	N/A
NICM, LVEF 30% to 35%, NYHA II, III	<i>Class IIa;</i> <i>LOE: B</i>	<i>Class I;</i> <i>LOE: A</i>	N/A
NICM, LVEF \leq 30%, NYHA I	<i>Class IIb;</i> <i>LOE: C</i>	N/A	N/A
NICM, LVEF \leq 31% to 35%, NYHA I	N/A	N/A	N/A

*For an explanation of class of recommendation and level of evidence, see Table 1.

ACC/AHA HF = ACC/AHA 2005 Guideline Update for the Diagnosis and Management of Chronic Heart Failure in the Adult; **ACC/AHA/NASPE PM and ICD** = ACC/AHA/NASPE 2002 Guidelines Update for Implantation of Cardiac Pacemakers and Antiarrhythmia Devices; **ACC/AHA STEMI** = ACC/AHA 2004 Guidelines for the Management of

A. Prophylactic Implantable Cardioverter Device Recommendations Across Published Guidelines

Please see *Table 2* for prophylactic implantable cardioverter defibrillator (ICD) therapy recommendations across published guidelines. A detailed explanation of the rationale used in formulating these recommendations can be found in the full-text guidelines.

2002 ACC/AHA/NASPE PM and ICD	Comment from the ACC/AHA/ESC VA & SCD Guidelines
<i>Class IIa; LOE: B</i>	VA & SCD has combined all trials that enrolled patients with LVD d/t MI into one recommendation, <i>Class I; LOE: A</i>
N/A	
<i>Class IIb; LOE: B</i>	
N/A	VA & SCD has expanded the range of LVEF to $\leq 30\%$ to 35% for patients with LVD d/t MI and NYHA functional class I into one recommendation, <i>Class IIa; LOE: B</i> .
N/A	
N/A	VA & SCD has combined all trials of NICM, NYHA II, III into one recommendation, <i>Class I; LOE: B</i>
N/A	
N/A	VA & SCD has expanded the range of LVEF to $\leq 30\%$ to 35% for patients with NICM and NYHA functional class I into one recommendation, <i>Class IIb; LOE: C</i> .
N/A	

Patients with ST-Elevation Myocardial Infarction; **EP** = electrophysiological; **ESC HF** = ESC 2005 Guidelines for the Diagnosis and Treatment of Chronic Heart Failure; **LOE** = level of evidence; **LVD d/t MI** = left ventricular dysfunction due to prior myocardial infarction; **LVEF** = left ventricular ejection fraction; **N/A** = populations not addressed; **NICM** = non-ischemic cardiomyopathy; **NSVT** = nonsustained ventricular tachycardia; **NYHA** = New York Heart Association functional class; **SCD** = sudden cardiac death; **VA** = ventricular arrhythmias.

B. Classification of Ventricular Arrhythmias and Sudden Cardiac Death

This classification table is provided for direction and introduction to these pocket guidelines (*Table 3*).

Table 3. Classification of Ventricular Arrhythmias

Classification by Electrocardiography

Nonsustained VT	Three or more beats in duration, terminating spontaneously in less than 30 seconds. VT is a cardiac arrhythmia of 3 or more consecutive complexes in duration emanating from the ventricles at a rate of greater than 100 bpm (cycle length less than 600 ms).
Monomorphic	Nonsustained VT with a single QRS morphology.
Polymorphic	Nonsustained VT with a changing QRS morphology at cycle length between 600 and 180 ms.
Sustained VT	VT greater than 30 seconds in duration and/or requiring termination due to hemodynamic compromise in less than 30 seconds.
Monomorphic	Sustained VT with a stable single QRS morphology.
Polymorphic	Sustained VT with a changing or multiform QRS morphology at cycle length between 600 and 180 ms.
Bundle branch reentrant tachycardia	VT due to reentry involving the His-Purkinje system, usually with LBBB morphology; this usually occurs in the setting of cardiomyopathy.
Bidirectional VT	VT with a beat-to-beat alternans in the QRS frontal plane axis, often associated with digitalis toxicity.
Torsades de pointes	Characterized by VT associated with a long QT or QTc, and electrocardiographically characterized by twisting of the peaks of the QRS complexes around the isoelectric line during the arrhythmia: <ul style="list-style-type: none"> ■ "Typical" initiated following "short-long-short" coupling intervals ■ Short coupled variant initiated by normal-short coupling.
Ventricular flutter	A regular (cycle length variability 30 ms or less) ventricular arrhythmia approximately 300 bpm (cycle length 200 ms) with a monomorphic appearance; no isoelectric interval between successive QRS complexes.
Ventricular fibrillation	Rapid, usually more than 300 bpm / 200 ms (cycle length 180 ms or less), grossly irregular ventricular rhythm with marked variability in QRS cycle length, morphology, and amplitude.

This table has been extracted from Table 4 of the full-text guidelines.

LBBB = left bundle-branch block; **VT** = ventricular tachycardia.



II. Incidence of Sudden Cardiac Death

The geographic incidence of sudden cardiac death (SCD) varies as a function of coronary heart disease (CHD) prevalence in different regions. Estimates for the United States range from less than 200,000 to more than 450,000 SCDs annually, with the most widely used estimates in the range of 300,000 to 350,000 SCDs annually. The variation is based, in part, on the inclusion criteria used in individual studies. Overall, event rates in Europe are similar to those in the United States, with significant geographic variations reported.

Approximately 50% of all CHD deaths are sudden and unexpected, occurring shortly (instantaneous to 1 hr) after the onset of a change in clinical status, with some geographical variation in the fraction of coronary deaths that are sudden.

III. Clinical Presentations of Patients With Ventricular Arrhythmias and Sudden Cardiac Death

Ventricular arrhythmias (VA) can occur in individuals with or without cardiac disorders. There is a great deal of overlap between clinical presentations (*Table 4*) and severity and type of heart disease. The prognosis and management are individualized according to symptom burden and severity of underlying heart disease in addition to the clinical presentation.



IV. General Evaluation of Patients With Documented or Suspected Ventricular Arrhythmias

A. Resting Electrocardiogram

Recommendations

Class I Resting 12-lead electrocardiogram (ECG) is indicated in all patients who are evaluated for VA. (*Level of Evidence: A*)

Table 4. Clinical Presentations of Patients With Ventricular Arrhythmias and Sudden Cardiac Death

- Asymptomatic individuals with or without electrocardiographic abnormalities
 - Persons with symptoms potentially attributable to ventricular arrhythmias
 - Palpitations
 - Dyspnea
 - Chest pain
 - Syncope and presyncope
 - Ventricular tachycardia that is hemodynamically stable
 - Ventricular tachycardia that is not hemodynamically stable
 - Cardiac arrest
 - Asystolic (sinus arrest, atrioventricular block)
 - Ventricular tachycardia
 - Ventricular fibrillation
 - Pulseless electrical activity
-

B. Exercise Testing

Recommendations

Class I 1. Exercise testing (ET) is recommended in adult patients with VA who have an intermediate or greater probability of having CHD by age, gender, and symptoms* to provoke ischemic changes or VA. *(Level of Evidence: B)*

2. ET, regardless of age, is useful in patients with known or suspected exercise-induced VA, including catecholaminergic ventricular tachycardia (VT) to provoke the arrhythmia, achieve a diagnosis, and determine the patient's response to tachycardia. *(Level of Evidence: B)*

Class IIa ET can be useful in evaluating response to medical or ablation therapy in patients with known exercise-induced VA. *(Level of Evidence: B)*

Class IIb 1. ET may be useful in patients with VA and a low probability of CHD by age, gender, and symptoms.* *(Level of Evidence: C)*

2. ET may be useful in the investigation of isolated premature ventricular complexes (PVCs) in middle-aged or older patients without other evidence of CHD. *(Level of Evidence: C)*

Class III See Table 1 in the ACC/AHA 2002 Guideline Update for Exercise Testing for contraindications. *(Level of Evidence: B)*

*See Table 4 in the ACC/AHA 2002 Guideline Update for Exercise Testing for further explanation of CHD probability.

C. Ambulatory Electrocardiography

Recommendations

- Class I**
1. Ambulatory ECG is indicated when there is a need to clarify the diagnosis by detecting arrhythmias, QT interval changes, T-wave alternans or ST-changes, evaluate risk, or judge therapy. *(Level of Evidence: A)*
 2. Event monitors are indicated when symptoms are sporadic to establish whether they are caused by transient arrhythmias. *(Level of Evidence: B)*
 3. Implantable recorders are useful in patients with sporadic symptoms suspected to be related to arrhythmias such as syncope, when a symptom-rhythm correlation cannot be established by conventional diagnostic techniques. *(Level of Evidence: B)*
-

D. Electrocardiogram Techniques and Measurements

Recommendations

- Class IIa**
- It is reasonable to use T-wave alternans for improving the diagnosis and risk stratification of patients with VA or who are at risk for developing life-threatening VA. *(Level of Evidence: A)*
-

- Class IIb**
- ECG techniques such as signal-averaged ECG, heart rate variability, baroflex sensitivity, and heart rate turbulence may be useful for improving the diagnosis and risk stratification of patients with

ventricular arrhythmias or who are at risk of developing life-threatening ventricular arrhythmias.

(Level of Evidence: B)

E. Left Ventricular Function and Imaging

Recommendations

Class I

1. Echocardiography is recommended in patients with VA who are suspected of having structural heart disease. *(Level of Evidence: B)*

2. Echocardiography is recommended for the subset of patients at high risk for development of serious VA or SCD, such as those with dilated, hypertrophic, or right ventricular (RV) cardiomyopathies, acute myocardial infarction (MI) survivors, or relatives of patients with inherited disorders associated with SCD. *(Level of Evidence: B)*

3. ET with an imaging modality (echocardiography or nuclear perfusion [single-photon emission computed tomography (SPECT)]) is recommended to detect silent ischemia in patients with VA who have an intermediate probability of having CHD by age, symptoms, and gender, and in whom ECG assessment is less reliable because of digoxin use, left ventricular (LV) hypertrophy, greater than 1 mm ST-segment depression at rest, Wolff-Parkinson-White Syndrome or left bundle-branch block. *(Level of Evidence: B)*

4. Pharmacological stress testing with an imaging modality (echocardiography or myocardial perfusion

SPECT) is recommended to detect silent ischemia in patients with VA who have an intermediate probability of having CHD by age, symptoms, and gender and are physically unable to perform a symptom-limited exercise test. *(Level of Evidence: B)*

- Class IIa**
1. Magnetic resonance imaging, cardiac computed tomography, or radionuclide angiography can be useful in patients with VA when echocardiography does not provide accurate assessment of LV and RV function, and/or evaluation of structural changes. *(Level of Evidence: B)*
 2. Coronary angiography can be useful in establishing or excluding the presence of significant obstructive CHD in patients with life-threatening VA or in survivors of SCD, who have an intermediate or greater probability of having CHD by age, symptoms, and gender. *(Level of Evidence: C)*
 3. LV imaging can be useful in patients undergoing biventricular pacing. *(Level of Evidence: C)*
-

F. Electrophysiological Testing

Electrophysiological (EP) testing with intracardiac recording and electrical stimulation at baseline and with drugs, has been used for arrhythmia assessment and risk stratification for SCD. EP testing is used to document inducibility of VT, guide ablation, evaluate drug effects, assess the risks of recurrent VT or SCD, evaluate loss of consciousness in selected patients with arrhythmias suspected as a cause and assess the indications for ICD therapy.

I. EP Testing in Patients With CHD

Recommendations

- Class I**
1. EP testing is recommended for diagnostic evaluation of patients with remote MI with symptoms suggestive of ventricular tachyarrhythmias including palpitations, presyncope, and syncope. *(Level of Evidence: B)*
 2. EP testing is recommended in patients with CHD to guide and assess efficacy of VT ablation. *(Level of Evidence: B)*
 3. EP testing is useful in patients with CHD for the diagnostic evaluation of wide-QRS-complex tachycardias of unclear mechanism. *(Level of Evidence: C)*
-

- Class IIa**
- EP testing is reasonable for risk stratification in patients with remote MI, nonsustained VT, and LV ejection fraction (LVEF) $\leq 40\%$. *(Level of Evidence: B)*
-

2. EP Testing in Patients With Syncope

Recommendations

- Class I**
- EP testing is recommended in patients with syncope of unknown cause with impaired LV function or structural heart disease. *(Level of Evidence: B)*
-
- Class IIa**
- EP testing can be useful in patients with syncope when brady- or tachyarrhythmias are suspected, and in whom noninvasive diagnostic studies are not conclusive. *(Level of Evidence: B)*
-

V. Therapies for Ventricular Arrhythmias

A. Introduction

Therapies for VA include antiarrhythmic drugs (e.g., beta blockers, amiodarone, sotalol), devices (e.g., ICDs), ablation, surgery, and revascularization. With the exception of ablation, recommendations for each of these modalities can be found within specific disease-based sections (e.g., Heart Failure) of these pocket guidelines. The recommendations for ablation therapy are described below.

B. Ablation

Recommendations

- Class I**
1. Ablation is indicated in patients who are otherwise at low risk for SCD and have sustained predominantly monomorphic VT that is drug resistant, or who are drug intolerant, or who do not wish long-term drug therapy. (*Level of Evidence: C*)
 2. Ablation is indicated in patients with bundle-branch reentrant VT. (*Level of Evidence: C*)
 3. Ablation is indicated as adjunctive therapy in patients with an ICD who are receiving multiple shocks as a result of sustained VT that is not manageable by reprogramming or changing drug therapy, or who do not wish long-term drug-therapy. (*Level of Evidence: C*)
 4. Ablation is indicated in patients with Wolff-Parkinson-White syndrome resuscitated from sudden cardiac arrest due to atrial fibrillation and rapid conduction over the accessory pathway causing ventricular fibrillation (VF). (*Level of Evidence: B*)

-
- Class IIa**
1. Ablation can be useful therapy in patients who are otherwise at low risk for SCD and have symptomatic nonsustained monomorphic VT that is drug resistant, who are drug intolerant, or who do not wish long-term drug therapy. *(Level of Evidence: C)*
 2. Ablation can be useful therapy in patients who are otherwise at low risk for SCD and have frequent symptomatic predominantly monomorphic PVCs that are drug resistant, who are drug intolerant, or who do not wish long-term drug therapy.
(Level of Evidence: C)
 3. Ablation can be useful in symptomatic patients with Wolff-Parkinson-White syndrome who have accessory pathways with refractory periods less than 240 ms in duration. *(Level of Evidence: B)*
-

- Class IIb**
1. Ablation of Purkinje fiber potentials may be considered in patients with VA storm consistently provoked by PVCs of similar morphology.
(Level of Evidence: C)
 2. Ablation of asymptomatic PVCs may be considered when the PVCs are very frequent to avoid or treat tachycardia-induced cardiomyopathy.
(Level of Evidence: C)
-

- Class III**
- Ablation of asymptomatic relatively infrequent PVCs is not indicated. *(Level of Evidence: C)*
-



VI. Acute Management of Specific Arrhythmias

A. Management of Cardiac Arrest

Recommendations

- Class I**
1. After establishing the presence of definite, suspected, or impending cardiac arrest, the first priority should be activation of a response team capable of identifying the specific mechanism and carrying out prompt intervention. *(Level of Evidence: B)*
 2. Cardiopulmonary resuscitation (CPR) should be implemented immediately after contacting a response team. *(Level of Evidence: A)*
 3. In an out-of-hospital setting, if an automated external defibrillator is available, it should be applied immediately and shock therapy administered according to the algorithms contained in the documents on CPR developed by the AHA in association with the International Liaison Committee on Resuscitation (ILCOR) and/or the European Resuscitation Council (ERC).
(Level of Evidence: C)
 4. For victims with ventricular tachyarrhythmic mechanisms of cardiac arrest, when recurrences occur after a maximally defibrillating shock (generally 360 J for monophasic defibrillators), intravenous amiodarone should be the preferred antiarrhythmic drug for attempting a stable rhythm after further defibrillations. *(Level of Evidence: B)*

5. For recurrent ventricular tachyarrhythmias or non-tachyarrhythmic mechanisms of cardiac arrest, it is recommended to follow the algorithms contained in the documents on CPR developed by the AHA in association with the ILCOR and/or the ERC.

(Level of Evidence: C)

6. Reversible causes and factors contributing to cardiac arrest should be managed during advanced life support, including management of hypoxia, electrolyte disturbances, mechanical factors, and volume depletion. *(Level of Evidence: C)*

Class IIa For response times ≥ 5 min, a brief (< 90 to 180 s) period of CPR is reasonable prior to attempting defibrillation. *(Level of Evidence: B)*

Class IIb A single precordial thump may be considered by healthcare professional providers when responding to a witnessed cardiac arrest. *(Level of Evidence: C)*

B. Ventricular Tachycardia Associated With Low Troponin MI Recommendations

Class I Patients presenting with sustained VT in whom low level elevations in cardiac biomarkers of myocyte injury/necrosis are documented, should be treated similarly to patients that have sustained VT and in whom no biomarker rise is documented. *(Level of Evidence: C)*

C. Sustained Monomorphic Ventricular Tachycardia

Recommendations

- Class I**
1. Wide-QRS tachycardia should be presumed to be VT if the diagnosis is unclear. *(Level of Evidence: C)*
 2. Direct current cardioversion with appropriate sedation is recommended at any point in the treatment cascade in patients with suspected sustained monomorphic VT with hemodynamic compromise. *(Level of Evidence: C)*
-

- Class IIa**
1. Intravenous (IV) procainamide (or ajmaline in some European countries) is reasonable for initial treatment of patients with stable sustained monomorphic VT. *(Level of Evidence: B)*
 2. IV amiodarone is reasonable in patients with sustained monomorphic VT that is hemodynamically unstable, that is refractory to conversion with countershock, or recurrent despite procainamide or other agents. *(Level of Evidence: C)*
 3. Transvenous catheter pace termination can be useful to treat patients with sustained monomorphic VT that is refractory to cardioversion or is frequently recurrent despite antiarrhythmic medication. *(Level of Evidence: C)*

Class IIb IV lidocaine might be reasonable for the initial treatment of patients with stable sustained monomorphic VT specifically associated with acute myocardial ischemia or infarction. *(Level of Evidence: C)*

Class III Calcium channel blockers such as verapamil and diltiazem should not be used in patients to terminate wide-QRS-complex tachycardia of unknown origin, especially in patients with a history of myocardial dysfunction. *(Level of Evidence: C)*

D. Repetitive Monomorphic Ventricular Tachycardia

Recommendations

Class IIa IV amiodarone, beta blockers, and IV procainamide (or sotalol or ajmaline in Europe) can be useful for treating repetitive monomorphic VT in the context of CHD and idiopathic VT. *(Level of Evidence: C)*

E. Polymorphic Ventricular Tachycardia

Recommendations

Class I 1. Direct current cardioversion with appropriate sedation as necessary is recommended for patients with sustained polymorphic VT with hemodynamic compromise and is reasonable at any point in the treatment cascade. *(Level of Evidence: B)*

2. IV beta blockers are useful for patients with recurrent polymorphic VT, especially if ischemia is suspected or cannot be excluded. *(Level of Evidence: B)*
 3. IV loading with amiodarone is useful for patients with recurrent polymorphic VT in the absence of abnormal repolarization related to congenital or acquired QT syndrome. *(Level of Evidence: C)*
 4. Urgent angiography with a view to revascularization should be considered for patients with polymorphic VT when myocardial ischemia cannot be excluded. *(Level of Evidence: C)*
-

Class IIb IV lidocaine may be reasonable for treatment of polymorphic VT specifically associated with acute myocardial ischemia or infarction. *(Level of Evidence: C)*

F. Torsades de Pointes

Recommendations

Class I 1. Withdrawal of any offending drugs and correction of electrolyte abnormalities are recommended in patients presenting with torsades de pointes. *(Level of Evidence: A)*

2. Acute and long-term pacing is recommended for patients presenting with torsades de pointes due to heart block and symptomatic bradycardia.

(Level of Evidence: A)

Class IIa

1. Management with IV magnesium sulfate is reasonable for patients who present with long QT syndrome (LQTS) and few episodes of torsades de pointes. Magnesium is not likely to be effective in patients with a normal QT interval. *(Level of Evidence: B)*

2. Acute and long-term pacing is reasonable for patients who present with recurrent pause-dependent torsades de pointes. *(Level of Evidence: B)*

3. Beta blockade combined with pacing is reasonable acute therapy for patients who present with torsades de pointes and sinus bradycardia. *(Level of Evidence: C)*

4. Isoproterenol is reasonable as temporary treatment in acute patients who present with recurrent pause-dependent torsades de pointes who do not have congenital LQTS. *(Level of Evidence: B)*

Class IIb

1. Potassium repletion to 4.5 to 5 mM/L may be considered for patients who present with torsades de pointes. *(Level of Evidence: B)*

2. IV lidocaine or oral mexiletine may be considered in patients who present with LQT3 and torsades de pointes. *(Level of Evidence: C)*

G. Incessant Ventricular Tachycardia

Recommendations

Class I Revascularization and beta blockade followed by IV antiarrhythmic drugs such as procainamide or amiodarone are recommended for patients with recurrent or incessant polymorphic VT due to acute myocardial ischemia. *(Level of Evidence: C)*

Class IIa IV amiodarone or procainamide followed by VT ablation can be effective in the management of patients with frequently recurring or incessant monomorphic VT. *(Level of Evidence: B)*

- Class IIb**
1. IV amiodarone and IV beta blockers separately or together may be reasonable in patients with VT storm. *(Level of Evidence: C)*
 2. Overdrive pacing or general anesthesia may be considered for patients with frequently recurring or incessant VT. *(Level of Evidence: C)*
 3. Spinal cord modulation may be considered for some patients with frequently recurring or incessant VT. *(Level of Evidence: C)*
-

VII. Ventricular Arrhythmias and Sudden Cardiac Death Related to Specific Pathology

A. Left Ventricular Dysfunction Due to Prior MI

Recommendations

- Class I**
1. Aggressive attempts should be made to treat heart failure (HF) that may be present in some patients with LV dysfunction (LVD) due to prior MI and ventricular tachyarrhythmias. (*Level of Evidence: C*)
 2. Aggressive attempts should be made to treat myocardial ischemia that may be present in some patients with ventricular tachyarrhythmias. (*Level of Evidence: C*)
 3. Coronary revascularization is indicated to reduce the risk of SCD in patients with VF when direct, clear evidence of acute myocardial ischemia is documented to immediately precede the onset of VF. (*Level of Evidence: B*)
 4. If coronary revascularization cannot be carried out, and there is evidence of prior MI and significant LVD, the primary therapy of patients resuscitated from VF should be the ICD in patients who are receiving chronic optimal medical therapy and those who have reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: A*)
 5. ICD therapy is recommended for primary prevention to reduce total mortality by a reduction

in SCD in patients with LVD due to prior MI who are at least 40 days post-MI, have an LVEF \leq 30% to 40%, are New York Heart Association (NYHA) functional class II or III, are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: A*)

6. The ICD is effective therapy to reduce mortality by a reduction in SCD in patients with LVD due to prior MI who present with hemodynamically unstable sustained VT, who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: A*)

Class IIa

1. Implantation of an ICD is reasonable in patients with LVD due to prior MI who are at least 40 days post-MI, have an LVEF of \leq 30% to 35%, are NYHA functional class I on chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: B*)

2. Amiodarone, often in combination with beta blockers, can be useful for patients with LVD due to prior MI and symptoms due to VT unresponsive to beta-adrenergic blocking agents. (*Level of Evidence: B*)

3. Sotalol is reasonable therapy to reduce symptoms resulting from VT for patients with LVD due to prior MI unresponsive to beta-blocking agents. (*Level of Evidence: C*)

4. Adjunctive therapies to the ICD, including catheter ablation or surgical resection, and pharmacological therapy with agents such as amiodarone or sotalol are reasonable to improve symptoms due to frequent episodes of sustained VT or VF in patients with LVD due to prior MI. *(Level of Evidence: C)*

5. Amiodarone is reasonable therapy to reduce symptoms due to recurrent hemodynamically stable VT for patients with LVD due to prior MI who cannot or refuse to have an ICD implanted. *(Level of Evidence: C)*

6. ICD implantation is reasonable for treatment of recurrent sustained VT in patients post-MI with normal or near normal ventricular function who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*

Class IIb

1. Curative catheter ablation or amiodarone may be considered in lieu of ICD therapy to improve symptoms in patients with LVD due to prior MI and recurrent hemodynamically stable VT whose LVEF is $> 40\%$. *(Level of Evidence: B)*

2. Amiodarone may be reasonable therapy for patients with LVD due to prior MI with an ICD indication, as defined above, in patients who cannot or refuse to have an ICD implanted. *(Level of Evidence: C)*

-
- Class III** 1. Prophylactic antiarrhythmic drug therapy is not indicated to reduce mortality in patients with asymptomatic nonsustained VA. *(Level of Evidence: B)*
2. Class IC antiarrhythmic drugs in patients with a past history of MI should not be used. *(Level of Evidence: A)*
-

B. Valvular Heart Disease

Recommendations

- Class I** Patients with valvular heart disease and VA should be evaluated and treated following current recommendations for each disorder. *(Level of Evidence: C)*
-

- Class IIb** The effectiveness of mitral valve repair or replacement to reduce the risk of SCD in patients with mitral valve prolapse, severe mitral regurgitation and serious VA is not well established. *(Level of Evidence: C)*
-

C. Congenital Heart Disease

Recommendations

- Class I** 1. ICD implantation is indicated in patients with congenital heart disease who are survivors of cardiac arrest after evaluation to define the cause of the event and exclude any reversible causes. ICD implantation is indicated in patients who are receiving

chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: B)*

2. Patients with congenital heart disease and spontaneous sustained VT should undergo invasive hemodynamic and EP evaluation. Recommended therapy includes catheter ablation or surgical resection to eliminate VT. If that is not successful, ICD implantation is recommended.

(Level of Evidence: C)

Class IIa Invasive hemodynamic and EP evaluation is reasonable in patients with congenital heart disease and unexplained syncope and impaired ventricular function. In the absence of a defined and reversible cause, ICD implantation is reasonable in patients who are receiving chronic optimal medical therapy and who have reasonable expectation of survival with a good functional status for more than 1 year.

(Level of Evidence: B)

Class IIb EP testing may be considered for patients with congenital heart disease and ventricular couplets or nonsustained VT to determine the risk of a sustained VA. *(Level of Evidence: C)*

Class III Prophylactic antiarrhythmic therapy is not indicated for asymptomatic patients with congenital heart disease and isolated PVCs. *(Level of Evidence: C)*

D. Pericardial Diseases

Recommendations

- Class I** VA that develop in patients with pericardial disease should be treated in the same manner that such arrhythmias are treated in patients with other diseases, including ICD pacemaker implantation as required. Patients receiving ICD implantation should be receiving chronic optimal medical therapy, and have reasonable expectation of survival with a good functional status for more than 1 year.

(Level of Evidence: C)

E. Pulmonary Arterial Hypertension

Recommendations

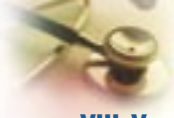
- Class III** Prophylactic antiarrhythmic therapy generally is not indicated for primary prevention of SCD in patients with pulmonary arterial hypertension or other pulmonary conditions. *(Level of Evidence: C)*
-



F. Transient Arrhythmias of Reversible Cause

Recommendations

- Class I**
1. Myocardial revascularization should be performed, when appropriate, to reduce the risk of SCD in patients experiencing cardiac arrest due to VF or polymorphic VT in the setting of acute ischemia or myocardial infarction. (*Level of Evidence: C*)
 2. Unless electrolyte abnormalities are proven to be the cause, survivors of cardiac arrest due to VF or polymorphic VT in whom electrolyte abnormalities are discovered in general should be evaluated and treated in a similar manner as survivors of cardiac arrest without electrolyte abnormalities. (*Level of Evidence: C*)
 3. Patients who experience sustained monomorphic VT in the presence of antiarrhythmic drugs or electrolyte abnormalities should be evaluated and treated in a manner similar to that of patients with VT without electrolyte abnormalities or antiarrhythmic drugs present. Antiarrhythmic drugs or electrolyte abnormalities should not be assumed to be the sole cause of sustained monomorphic VT. (*Level of Evidence: B*)
 4. Patients who experience polymorphic VT in association with prolonged QT interval due to antiarrhythmic medications or other drugs should be advised to avoid exposure to all agents associated with QT prolongation. A list of such drugs can be found on the Web sites www.qtdrugs.org and www.torsades.org. (*Level of Evidence: B*)
-



VIII. Ventricular Arrhythmias Associated With Cardiomyopathies

A. Dilated Cardiomyopathy (Nonischemic)

Recommendations

- Class I**
1. EP testing is useful to diagnose bundle branch-reentrant tachycardia, and to guide ablation in patients with nonischemic dilated cardiomyopathy (DCM). (*Level of Evidence: C*)
 2. EP testing is useful for diagnostic evaluation in patients with nonischemic DCM with sustained palpitations, wide-QRS-complex tachycardia, syncope or presyncope. (*Level of Evidence: C*)
 3. An ICD should be implanted in patients with nonischemic DCM and significant LVD who have sustained VT or VF, who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: A*)
 4. ICD therapy is recommended for primary prevention to reduce total mortality by a reduction in SCD in patients with nonischemic DCM who have an LVEF $\leq 30\%$ to 35% , are NYHA functional class II or III receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: B*)

-
- Class IIa**
1. ICD implantation can be beneficial for patients with unexplained syncope, significant LVD, and nonischemic DCM who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*
 2. ICD implantation can be effective for termination of sustained VT in patients with normal or near normal ventricular function and nonischemic DCM who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*

-
- Class IIb**
1. Amiodarone may be considered for sustained VT or VF in patients with nonischemic DCM. *(Level of Evidence: C)*
 2. Placement of an ICD might be considered in patients who have nonischemic DCM, LVEF \leq 30% to 35%, are NYHA functional class I receiving chronic optimal medical therapy, and in patients who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*
-

B. Hypertrophic Cardiomyopathy

Recommendations

Class I ICD therapy should be used for treatment in patients with hypertrophic cardiomyopathy (HCM) who have sustained VT and/or VF and who are receiving chronic optimal medical therapy and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: B)*

Class IIa

1. ICD implantation can be effective for primary prophylaxis against SCD in patients with HCM who have one or more major risk factor *(See Table 5)* for SCD and who are receiving chronic optimal medical therapy and in patients who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*
2. Amiodarone therapy can be effective for treatment in patients with HCM with a history of sustained VT and/or VF when ICD is not feasible. *(Level of Evidence: C)*

Class IIb

1. EP testing may be considered for risk assessment for SCD in patients with HCM. *(Level of Evidence: C)*
2. Amiodarone may be considered for primary prophylaxis against SCD in patients with HCM who have one or more major risk factor for SCD *(See Table 5)*, if ICD implantation is not feasible. *(Level of Evidence: C)*

Table 5. Risk Factors for SCD in Hypertrophic Cardiomyopathy

Major risk factors	Possible in individual patients
Cardiac arrest (VF)	AF
Spontaneous sustained VT	Myocardial ischemia
Family history of premature sudden death	LV outflow obstruction
Unexplained syncope	High-risk mutation
LV thickness greater than or equal to 30 mm	Intense (competitive) physical exertion
Abnormal exercise BP	
Nonsustained spontaneous VT	

Modified with permission from Maron BJ, McKenna WJ, Danielson GK, et al. American College of Cardiology/European Society of Cardiology clinical expert consensus document on hypertrophic cardiomyopathy. A report of the American College of Cardiology Foundation Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines. *J Am Coll Cardiol* 2003; 42:1687–713.

AF = atrial fibrillation; **BP** = blood pressure; **LV** = left ventricular; **SCD** = sudden cardiac death; **VF** = ventricular fibrillation; **VT** = ventricular tachycardia.



C. Arrhythmogenic Right Ventricular Cardiomyopathy

Recommendations

Class I ICD implantation is recommended for prevention of SCD in patients with arrhythmogenic RV cardiomyopathy (ARVC) with documented sustained VT or VF who are receiving chronic optimal medical therapy and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: B)*

Class IIa

1. ICD implantation can be effective for prevention of SCD in patients with ARVC with extensive disease, including those with LV involvement, one or more affected family member with SCD, or undiagnosed syncope when VT or VF has not been excluded as the cause of syncope, who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*
2. Amiodarone or sotalol can be effective for treatment of sustained VT or VF in patients with ARVC when ICD implantation is not feasible. *(Level of Evidence: C)*
3. Ablation can be useful as adjunctive therapy in management of patients with ARVC with recurrent VT, despite optimal antiarrhythmic drug therapy. *(Level of Evidence: C)*

Class IIb EP testing might be useful for risk assessment of SCD in patients with ARVC. *(Level of Evidence: C)*



IX. Heart Failure

Recommendations

- Class I**
1. ICD therapy is recommended for secondary prevention of SCD in patients who survived VF or hemodynamically unstable VT, or VT with syncope and have an LVEF $\leq 40\%$, who are receiving chronic optimal medical therapy and who have a reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: A*)
 2. ICD therapy is recommended for primary prevention to reduce total mortality by a reduction in SCD in patients with LVD due to prior MI who are at least 40 days post-MI, have an LVEF $\leq 30\%$ to 40% , are NYHA functional class II or III receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: A*)
 3. ICD therapy is recommended for primary prevention to reduce total mortality by a reduction in SCD in patients with nonischemic heart disease who have an LVEF $\leq 30\%$ to 35% , are NYHA functional class II or III receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. (*Level of Evidence: B*)
 4. Amiodarone, sotalol, and/or other beta blockers are recommended pharmacological adjuncts to ICD therapy to suppress symptomatic ventricular

tachyarrhythmias (both sustained and nonsustained) in otherwise optimally treated patients with HF.

(Level of Evidence: C)

5. Amiodarone is indicated for the suppression of acute hemodynamically compromising ventricular or supraventricular tachyarrhythmias when cardioversion and/or correction of reversible causes has failed to terminate the arrhythmia or prevent its early recurrence. *(Level of Evidence: B)*

Class IIa

1. ICD therapy combined with biventricular pacing can be effective for primary prevention to reduce total mortality by a reduction in SCD, in patients with NYHA functional class III or IV receiving optimal medical therapy, in sinus rhythm with a QRS complex of at least 120 ms and who have reasonable expectation of survival with a good functional status for more than 1 year.

(Level of Evidence: B)

2. ICD therapy is reasonable for primary prevention to reduce total mortality by a reduction in SCD in patients with LVD due to prior MI who are at least 40 days post-MI, have an LVEF of $\leq 30\%$ to 35% , are NYHA functional class I receiving chronic optimal medical therapy, and have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: B)*

3. ICD therapy is reasonable in patients with recurrent stable VT, a normal or near normal LVEF and optimally treated HF, and who have a reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*

4. Biventricular pacing in the absence of ICD therapy is reasonable for the prevention of SCD in patients with NYHA functional class III or IV HF, an LVEF $\leq 35\%$ and a QRS complex ≥ 160 ms (or at least 120 ms in the presence of other evidence of ventricular dyssynchrony) who are receiving chronic optimal medical therapy and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: B)*

Class IIb

1. Amiodarone, sotalol, and/or beta blockers may be considered as pharmacological alternatives to ICD therapy to suppress symptomatic ventricular tachyarrhythmias (both sustained and nonsustained) in optimally treated patients with HF for whom ICD therapy is not feasible. *(Level of Evidence: C)*

2. ICD therapy may be considered for primary prevention to reduce total mortality by a reduction in SCD in patients with nonischemic heart disease who have an LVEF of $\leq 30\%$ to 35% , are NYHA functional class I receiving chronic optimal medical therapy, and who have a reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: B)*



X. Genetic Arrhythmia Syndromes

A. Long QT Syndrome

Recommendations

- Class I**
1. Lifestyle modification (see full-text guidelines) is recommended for patients with an LQTS diagnosis (clinical and/or molecular). *(Level of Evidence: B)*
 2. Beta blockers are recommended for patients with an LQTS clinical diagnosis (i.e., in the presence of prolonged QT interval). *(Level of Evidence: B)*
 3. Implantation of an ICD along with use of beta blockers is recommended for LQTS patients with previous cardiac arrest and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: A)*
-

- Class IIa**
1. Beta blockers can be effective to reduce SCD in patients with a molecular LQTS analysis and normal QT interval. *(Level of Evidence: B)*
 2. Implantation of an ICD with continued use of beta blockers can be effective to reduce SCD in LQTS patients who are experiencing syncope and/or VT while receiving beta blockers and who have reasonable expectation of survival with a good functional status for more than 1 year.
(Level of Evidence: B)

-
- Class IIb**
1. Left cardiac sympathetic neural denervation may be considered for LQTS patients with syncope, torsades de pointes, or cardiac arrest while receiving beta blockers. *(Level of Evidence: B)*
 2. Implantation of an ICD with use of beta blockers may be considered for prophylaxis of SCD for patients who are in categories possibly associated with higher risk of cardiac arrest such as LQT2 and LQT3, and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: B)*
-

B. Brugada Syndrome

Recommendations

Class I An ICD is indicated for Brugada syndrome patients with previous cardiac arrest receiving chronic optimal medical therapy and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*

Class IIa

1. An ICD is reasonable for Brugada syndrome patients with spontaneous ST-segment elevation in V₁, V₂, or V₃ who have had syncope with or without mutations demonstrated in the SCN5A gene and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*

2. Clinical monitoring for the development of a spontaneous ST-segment elevation pattern is reasonable for the management of patients with ST-segment elevation induced only with provocative pharmacological challenge with or without symptoms. *(Level of Evidence: C)*

3. An ICD is reasonable for Brugada syndrome patients with documented VT that has not resulted in cardiac arrest and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*

4. Isoproterenol can be useful to treat an electrical storm in the Brugada syndrome. *(Level of Evidence: C)*

Class IIb

1. EP testing may be considered for risk stratification in asymptomatic Brugada syndrome patients with spontaneous ST-segment elevation with or without a mutation in the SCN5A gene. *(Level of Evidence: C)*

2. Quinidine might be reasonable for the treatment of electrical storm in patients with Brugada syndrome. *(Level of Evidence: C)*



C. Catecholaminergic Polymorphic Ventricular Tachycardia

Recommendations

- Class I**
1. Beta blockers are indicated for patients who are clinically diagnosed with catecholaminergic polymorphic VT (CPVT) on the basis of the presence of spontaneous or documented stress-induced VA. *(Level of Evidence: C)*
 2. Implantation of an ICD with use of beta blockers is indicated for patients with CPVT who are survivors of cardiac arrest and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*
-

- Class IIa**
1. Beta blockers can be effective in patients without clinical manifestations when the diagnosis of CPVT is established during childhood based on genetic analysis. *(Level of Evidence: C)*
 2. Implantation of an ICD with use of beta blockers can be effective for affected patients with CPVT with syncope and/or documented sustained VT who are receiving beta blockers and who have reasonable expectation of survival with a good functional status for more than 1 year. *(Level of Evidence: C)*
-

- Class IIb**
- Beta blockers may be considered for patients with CPVT who were genetically diagnosed in adulthood and never manifested clinical symptoms of tachyarrhythmias. *(Level of Evidence: C)*
-



XI. Ventricular Arrhythmias and Sudden Cardiac Death Related to Specific Populations

A. Athletes

Recommendations

Class I 1. Preparticipation history and physical examination, including family history of premature or sudden death and specific evidence of cardiovascular diseases such as cardiomyopathies and ion channel abnormalities is recommended in athletes.

(Level of Evidence: C)

2. Athletes presenting with rhythm disorders, structural heart disease, or other signs or symptoms suspicious for cardiovascular disorders, should be evaluated as any other patient but with recognition of the potential uniqueness of their activity. *(Level of Evidence: C)*

3. Athletes presenting with syncope should be carefully evaluated to uncover underlying cardiovascular disease or rhythm disorder. *(Level of Evidence: B)*

4. Athletes with serious symptoms should cease competition while cardiovascular abnormalities are being fully evaluated. *(Level of Evidence: C)*

Class IIb 12-lead ECG and possibly echocardiography may be considered as preparticipation screening for heart disorders in athletes. *(Level of Evidence: B)*

B. Gender & Pregnancy

Recommendations

- Class I**
1. Pregnant women developing hemodynamically unstable VT or VF should be electrically cardioverted or defibrillated. *(Level of Evidence: B)*
 2. In pregnant women with LQTS who have had symptoms, it is beneficial to continue beta-blocker medications throughout pregnancy and afterwards, unless there are definite contraindications. *(Level of Evidence: C)*
-

C. Elderly Patients

Recommendations

- Class I**
1. Elderly patients with VA should generally be treated in the same manner as younger individuals. *(Level of Evidence: A)*
 2. The dosing and titration schedule of antiarrhythmic drugs prescribed to elderly patients should be adjusted to the altered pharmacokinetics of such patients. *(Level of Evidence: C)*
-
- Class III**
- Elderly patients with projected life expectancy less than 1 year due to major comorbidities should not receive ICD therapy. *(Level of Evidence: C)*
-

Despite the demonstrated efficacy in reducing all-cause mortality and SCD, beta blockers are underused in the elderly. Several randomized prospective trials have demonstrated the efficacy of ICDs in primary and secondary prevention of SCD when compared with antiarrhythmic drug therapy across all age groups.

D. Patients With Implantable Cardioverter Devices

Recommendations

- Class I**
1. Patients with implanted ICDs should receive regular follow-up and analysis of the device status. *(Level of Evidence: C)*
 2. Implanted ICDs should be programmed to obtain optimal sensitivity and specificity. *(Level of Evidence: C)*
 3. Measures should be undertaken to minimize the risk of inappropriate ICD therapies. *(Level of Evidence: C)*
 4. Patients with implanted ICDs who present with incessant VT should be hospitalized for management. *(Level of Evidence: C)*
-

- Class IIa**
1. Catheter ablation can be useful for patients with implanted ICDs who experience incessant or frequently recurring VT. *(Level of Evidence: B)*
 2. In patients experiencing inappropriate ICD therapy, electrophysiologic evaluation can be useful for diagnostic and therapeutic purposes. *(Level of Evidence: C)*
-

E. Drug-Induced Arrhythmias

1. Digitalis Toxicity

Recommendations

Class I An anti-digitalis antibody is recommended for patients who present with sustained ventricular arrhythmias, advanced atrioventricular (AV) block, and/or asystole that are considered due to digitalis toxicity. *(Level of Evidence: A)*

Class IIa 1. Patients taking digitalis who present with mild cardiac toxicity (e.g., isolated ectopic beats only), can be managed effectively with recognition, continuous monitoring of cardiac rhythm, withdrawal of digitalis, restoration of normal electrolyte levels (including serum potassium > 4 mM/L) and oxygenation. *(Level of Evidence: C)*

2. Magnesium or pacing is reasonable for patients who take digitalis and present with severe toxicity.* *(Level of Evidence: C)*

Class IIb Dialysis for the management of hyperkalemia may be considered for patients who take digitalis and present with severe toxicity.* *(Level of Evidence: C)*

* Sustained VA, advanced AV block, and/or asystole.

Class III Management by lidocaine or phenytoin is not recommended for patients taking digitalis and who present with severe toxicity.* (*Level of Evidence: C*)

2. Drug-Induced Long QT Syndrome

Recommendations

Class I In patients with drug-induced LQTS, removal of the offending agent is indicated. (*Level of Evidence: A*)

- Class IIa**
1. Management with IV magnesium sulfate is reasonable for patients who take QT-prolonging drugs and present with few episodes of torsades de pointes in which the QT remains long.
(*Level of Evidence: B*)
 2. Atrial or ventricular pacing or isoproterenol is reasonable for patients taking QT-prolonging drugs who present with recurrent torsades de pointes.
(*Level of Evidence: B*)
-

Class IIb Potassium ion repletion to 4.5 to 5 mM/L may be reasonable for patients who take QT-prolonging drugs and present with few episodes of torsades de pointes in whom the QT remains long.
(*Level of Evidence: C*)

* Sustained VA, advanced AV block, and/or asystole.

3. Sodium Channel Blocker–Related Toxicity

Recommendations

Class I In patients with sodium channel blocker–related toxicity, removal of the offending agent is indicated.
(*Level of Evidence: A*)

Class IIa 1. Stopping the drug, reprogramming the pacemaker or repositioning leads can be useful in patients taking sodium channel blockers who present with elevated defibrillation thresholds or pacing requirement. (*Level of Evidence: C*)

2. In patients taking sodium channel blockers who present with atrial flutter with 1:1 AV conduction, withdrawal of the offending agent is reasonable. If the drug needs to be continued, additional A-V nodal blockade with diltiazem, verapamil or beta blocker or atrial flutter ablation can be effective.
(*Level of Evidence: C*)

Class IIb Administration of a beta blocker and a sodium bolus may be considered for patients taking sodium channel blockers if the tachycardia becomes more frequent or more difficult to cardiovert.
(*Level of Evidence: C*)

Drug interactions causing arrhythmias are included in *Table 6*. Syndromes of drug-induced arrhythmias and their management are included in *Table 7*.

Table 6. Drug Interactions Causing Arrhythmias

Drugs	Interacting Drug	Effect
Increased Concentration of Arrhythmogenic Drug		
Digoxin	Some antibiotics	By eliminating gut flora that metabolize digoxin, some antibiotics may increase digoxin bioavailability. Note: some antibiotics also interfere with P-glycoprotein (expressed in the intestine and elsewhere), another effect that can elevate digoxin concentration
Digoxin	Amiodarone Quinidine Verapamil Cyclosporine Itraconazole Erythromycin	Increased digoxin bioavailability, reduced biliary and renal excretion due to P-glycoprotein inhibition Digoxin toxicity
Quinidine	Ketoconazole	Increased drug levels
Cisapride	Itraconazole	
Terfenadine, astemizole	Erythromycin* Clarithromycin Some calcium blockers* Some HIV protease inhibitors (especially ritanovir)	
Beta blockers, propafenone	Quinidine (even ultra-low dose) Fluoxetine	Increased beta blockade Increased beta blockade
Flecainide	Some tricyclic antidepressants	Increased adverse effects Decreased analgesia (due to failure of biotransformation to the active metabolite morphine)
Dofetilide	Verapamil Cimetidine Trimethoprim Ketoconazole Megestrol	Increased plasma dofetilide concentration due to inhibition of renal excretion

Drugs	Interacting Drug	Effect
Decreased Concentration of Arrhythmogenic Drug		
Digoxin	Antacids	Decreased digoxin effect due to decreased absorption
	Rifampin	Increased P-glycoprotein activity
Quinidine, mexiletine	Rifampin, barbiturates	Induced drug metabolism
Synergistic Pharmacological Activity Causing Arrhythmias		
QT-prolonging antiarrhythmics	Diuretics	Increased torsades de pointes risk due to diuretic-induced hypokalemia
Beta blockers	Amiodarone, clonidine, digoxin, diltiazem, verapamil	Bradycardia when used in combination
Digoxin	Amiodarone, beta blockers, clonidine, diltiazem, verapamil	
Verapamil	Amiodarone, beta blockers, clonidine, digoxin, diltiazem	
Diltiazem	Amiodarone, beta blockers, clonidine, digoxin, verapamil	
Clonidine	Amiodarone, beta blockers, digoxin, diltiazem, verapamil	
Amiodarone	Beta blockers, clonidine, digoxin, diltiazem, verapamil	
Sildenafil	Nitrates	Increased and persistent vasodilation; risk of myocardial ischemia

* These may also accumulate to toxic levels with co-administration of inhibitor drugs like ketoconazole.

Data are from Roden DM, Anderson ME. Proarrhythmia. In Kass RS, Clancey CE, editors. Handbook of Pharmacology: vol. 171. Basis and Treatment of Cardiac Arrhythmias. Boston: Springer Verlag, 2006:288-304.

Table 7. Syndromes of Drug-Induced Arrhythmias and Their Management

Drugs	Clinical setting	Management*
Digitalis	Mild cardiac toxicity (isolated arrhythmias only)	
	Severe toxicity: Sustained ventricular arrhythmias; advanced AV block; asystole	Anti-digitalis antibody Pacing Dialysis for hyperkalemia
QT-prolonging drugs	Torsades de pointes: few episodes, QT remains long	IV magnesium sulfate (MgSO ₄) Replete potassium (K ⁺) to 4.5 to 5 mEq/L
	Recurrent torsades de pointes	Ventricular pacing Isoproterenol
Sodium-channel blockers	Elevated defibrillation or pacing requirement	Stop drug; reposition leads
	Atrial flutter with 1:1 AV conduction	Diltiazem, verapamil, beta blocker (IV)
	Ventricular tachycardia (more frequent; difficult to cardiovert)	Beta blocker; sodium
	Brugada syndrome	Stop drug; treat arrhythmia

*Always includes recognition, continuous monitoring of cardiac rhythm, withdrawal of offending agents, restoration of normal electrolytes (including serum potassium to greater than 4 mEq/L) and oxygenation. The order shown is not meant to represent the preferred sequence when more than one treatment is listed.

AV = atrioventricular; **IV** = intravenous.

