

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

Developed in Collaboration With the Heart Failure Society of America
Endorsed by Heart Failure Society of America

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Citation

This slide set is adapted from the 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. Published on [Date], available at: *Journal of the American College of Cardiology* [(insert full link)] and *Circulation* [(insert full link)]

The full-text guidelines are also available on the following Web sites: ACC (www.acc.org) and AHA (professional.heart.org)

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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: <ul style="list-style-type: none"> Is recommended Is indicated/useful/effective/beneficial Should be performed/administered/other Comparative-Effectiveness Phrases†: <ul style="list-style-type: none"> 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: <ul style="list-style-type: none"> Is reasonable Can be useful/effective/beneficial Comparative-Effectiveness Phrases†: <ul style="list-style-type: none"> 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: <ul style="list-style-type: none"> May/might be reasonable May/might be considered Usefulness/effectiveness is unknown/unclear/uncertain or not well established 	
CLASS III: No Benefit (MODERATE) <i>(Generally, LOE A or B use only)</i>	Benefit = Risk
Suggested phrases for writing recommendations: <ul style="list-style-type: none"> 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: <ul style="list-style-type: none"> Potentially harmful Causes harm Associated with excess morbidity/mortality Should not be performed/administered/other 	

LEVEL (QUALITY) OF EVIDENCE‡	
LEVEL A	<ul style="list-style-type: none"> 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) <ul style="list-style-type: none"> Moderate-quality evidence‡ from 1 or more RCTs Meta-analyses of moderate-quality RCTs
LEVEL B-NR	(Nonrandomized) <ul style="list-style-type: none"> 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) <ul style="list-style-type: none"> 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) <p>Consensus of expert opinion based on clinical experience</p>

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.

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General Evaluation of Patients with Documented or Suspected VA

History and Physical Examination

COR	LOE	Recommendation for Syncope*
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.

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

12-lead ECG and Exercise Testing

COR	LOE	Recommendations for 12-lead ECG and Exercise Testing
I	B-NR	1. In patients with sustained, hemodynamically stable, wide complex tachycardia, a 12-lead ECG during tachycardia should be obtained.
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.
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.

Ambulatory Electrocardiography

COR	LOE	Recommendation for Ambulatory Electrocardiography
I	B-NR	1. Ambulatory ECG monitoring is useful to evaluate whether symptoms, including palpitations, presyncope, or syncope, are caused by VA.

Implanted Cardiac Monitors

COR	LOE	Recommendation for Implanted Cardiac Monitors
Ila	B-NR	1. In patients with sporadic symptoms (including syncope) suspected to be related to VA implanted cardiac monitors can be useful.

Noninvasive Evaluation

Noninvasive Cardiac Imaging

COR	LOE	Recommendations for Noninvasive Cardiac Imaging
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.
Ila	C-EO	2. In patients presenting with VA who are suspected of having structural heart disease, cardiac MRI or CT can be useful to detect and characterize underlying structural heart disease.

Noninvasive Evaluation

Biomarkers

COR	LOE	Recommendation for Biomarkers
IIa	B-R	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.

Genetic Considerations in Arrhythmia Syndromes

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

Invasive Evaluation

Invasive Cardiac Imaging: Cardiac Catheterization or CT Angiography

COR	LOE	Recommendation for Invasive Imaging: Cardiac Catheterization
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.

Invasive Evaluation

Electrophysiological Study for VA

COR	LOE	Recommendations for Electrophysiological Study
Ila	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.
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.
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.

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Therapies for Treatment or Prevention of VA

Preventing SCD with HF Medications

COR	LOE	Recommendation for Pharmacological Prevention of SCD
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.

Surgery and Revascularization Procedures in Patients With Ischemic Heart Disease

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

Surgery and Revascularization Procedures in Patients With IHD

Surgery for Arrhythmia Management

COR	LOE	Recommendation for Surgery for Arrhythmia Management
IIb	C-LD	1. In patients with monomorphic VT refractory to antiarrhythmic medications and attempts at catheter ablation, surgical ablation may be reasonable.

Autonomic Modulation

COR	LOE	Recommendations for Autonomic Modulation
IIa	C-LD	1. In patients with symptomatic, non-life-threatening VA, treatment with a beta blocker is reasonable.
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.

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Acute Management of Specific VA

Acute Management of Specific VA

COR	LOE	Recommendations for Management of Cardiac Arrest
I	A	1. In patients in cardiac arrest, CPR should be performed according to published Basic and Advanced Cardiac Life Support algorithms.
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.
I	A	3. Patients presenting with VA with hemodynamic instability should undergo direct current cardioversion.
I	B-NR	4. In patients with polymorphic VT or VF with ST elevation MI, angiography with emergent revascularization is recommended.
I	C-EO	5. Patients with a wide-QRS tachycardia should be presumed to have VT if the diagnosis is unclear.

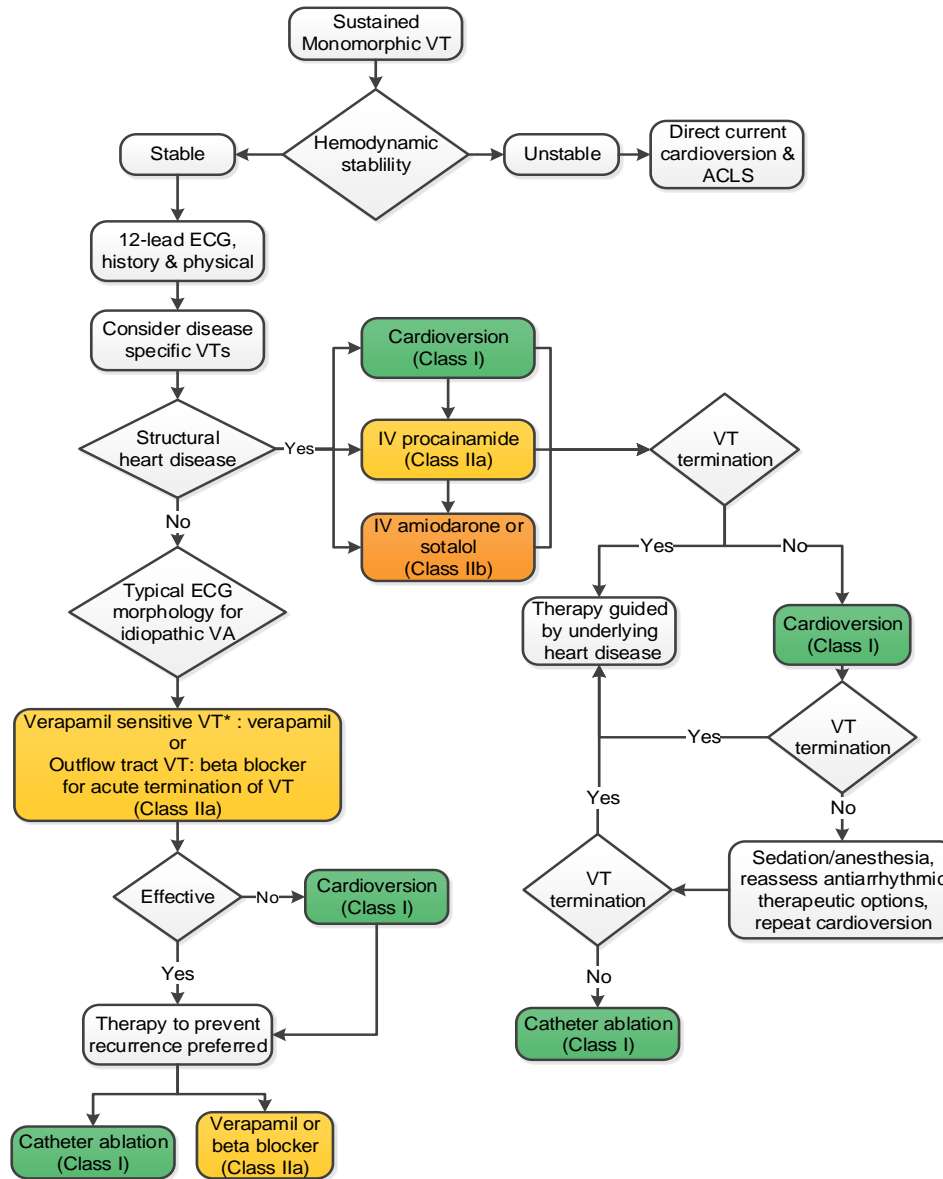
Acute Management of Specific VA (contd.)

COR	LOE	Recommendations for Management of Cardiac Arrest
IIa	A	6. In patients with hemodynamically stable VT, administration of intravenous procainamide can be useful to attempt to terminate VT .
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.
IIa	B-R	8. In patients with polymorphic VT due to myocardial ischemia, intravenous beta blockers can be useful.
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.
IIb	A	10. In patients in cardiac arrest, administration of epinephrine (1 mg every 3 to 5 minutes) during CPR may be reasonable.

Acute Management of Specific VA (contd.)

COR	LOE	Recommendations for Management of Cardiac Arrest
IIb	B-R	11. In patients with hemodynamically stable VT administration of intravenous amiodarone, or sotalol may be considered to attempt to terminate VT.
III: No Benefit	A	12. In patients with cardiac arrest, administration of high-dose epinephrine (>1 mg boluses) compared to standard doses is not beneficial.
III: No Benefit	A	13. In patients with refractory VF not related to torsades de pointes, administration of intravenous magnesium is not beneficial.
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.
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.

Management of Sustained Monomorphic VT



Colors correspond to Class of Recommendation in Table 1.

*Known history of Verapamil sensitive or classical ECG presentation.

ACLS indicates advanced cardiovascular life support; ECG, electrocardiogram;

VA, ventricular arrhythmia; and VT, ventricular tachycardia.

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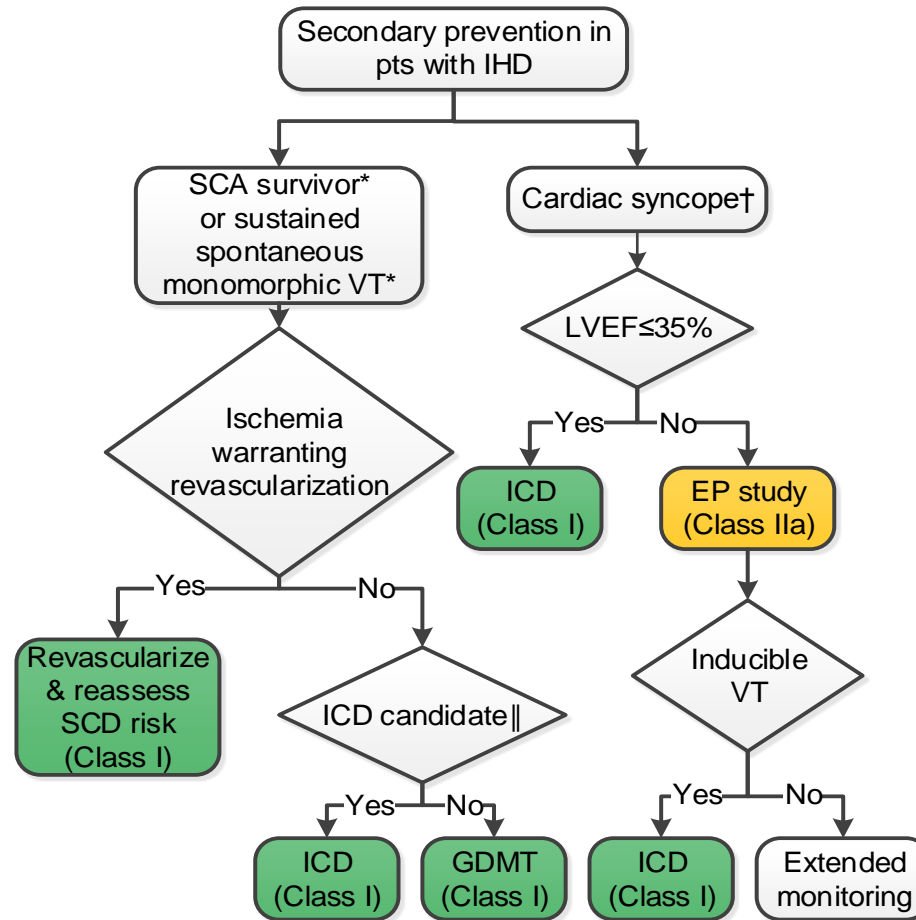
Ongoing Management of VA and SCD Risk Related to Specific Disease States

Ischemic Heart Disease

Secondary Prevention of SCD in Patients With Ischemic Heart Disease

COR	LOE	Recommendations for Secondary Prevention of SCD in Patients With Ischemic Heart Disease
I	B-R	1. In patients with ischemic heart disease who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R) or stable VT (LOE: B-NR) not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.
	B-NR	
<p>Value Statement: Intermediate Value (LOE: B-R)</p>		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.
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.

Secondary Prevention Patients With IHD



Colors correspond to Class of Recommendation in Table 1.

*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; SCA, sudden cardiac arrest; SCD, sudden cardiac death; and VT, ventricular tachycardia

Ischemic Heart Disease

Secondary Prevention of SCD in Patients With Ischemic Heart Disease Coronary Artery Spasm

COR	LOE	Recommendations for Patients With Coronary Artery Spasm
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.
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.
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.

Ischemic Heart Disease

Primary Prevention of SCD in Patients With Ischemic Heart Disease

COR	LOE	Recommendations for Primary Prevention of SCD in Patients With Ischemic Heart Disease
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.
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.

Ischemic Heart Disease

Primary Prevention of SCD in Patients With Ischemic Heart Disease (contd.)

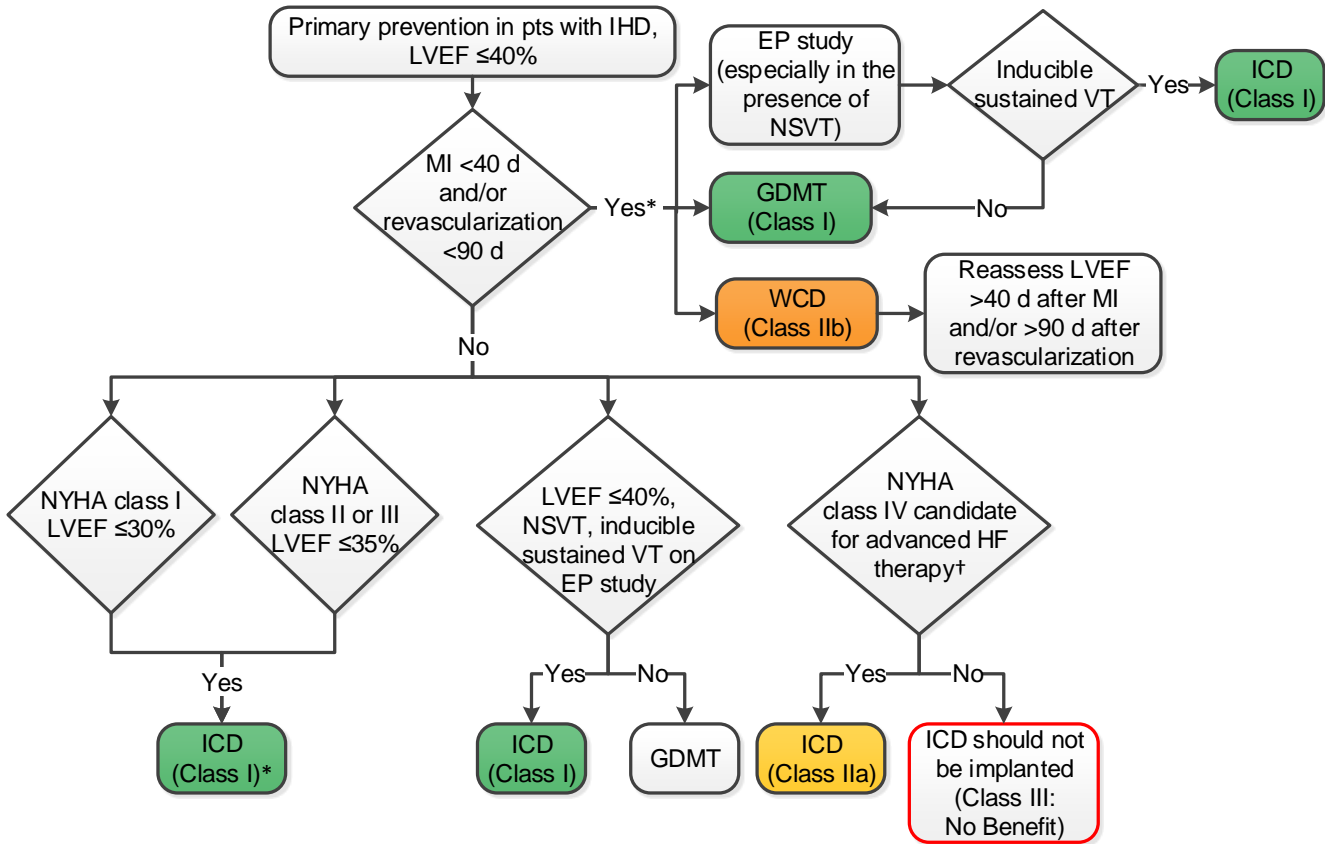
COR	LOE	Recommendations for Primary Prevention of SCD in Patients With Ischemic Heart Disease
		3. A transvenous ICD provides high value in the primary prevention of SCD particularly when the patient's risk of death due to a VA is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient's burden of comorbidities and functional status.
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.

Ischemic Heart Disease

Primary Prevention of SCD in Patients With Ischemic Heart Disease (contd.)

COR	LOE	Recommendations for Primary Prevention of SCD in Patients With Ischemic Heart Disease
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.
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.

Primary Prevention of SCD in Patients With Ischemic Heart Disease



Colors correspond to Class of Recommendation in Table 1.

*Advanced HF therapy includes CRT, cardiac transplant, and LVAD.

†Scenarios exist for early ICD placement in select circumstances such as patients with a pacing indication or syncope thought due to VT. These are detailed elsewhere in a HRS/ACC/AHA expert consensus statement.

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; SCD, sudden cardiac death; VT, ventricular tachycardia; and WCD, wearable cardioverter-defibrillator.



Ischemic Heart Disease

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

COR	LOE	Recommendations for Treatment of Recurrent VA in Patients With Ischemic Heart Disease
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.
I	B-R	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) or other antiarrhythmic medications (LOE: B-NR), catheter ablation is recommended.
	B-NR	

Ischemic Heart Disease

Treatment and Prevention of Recurrent VA in Patients With Ischemic Heart Disease (contd.)

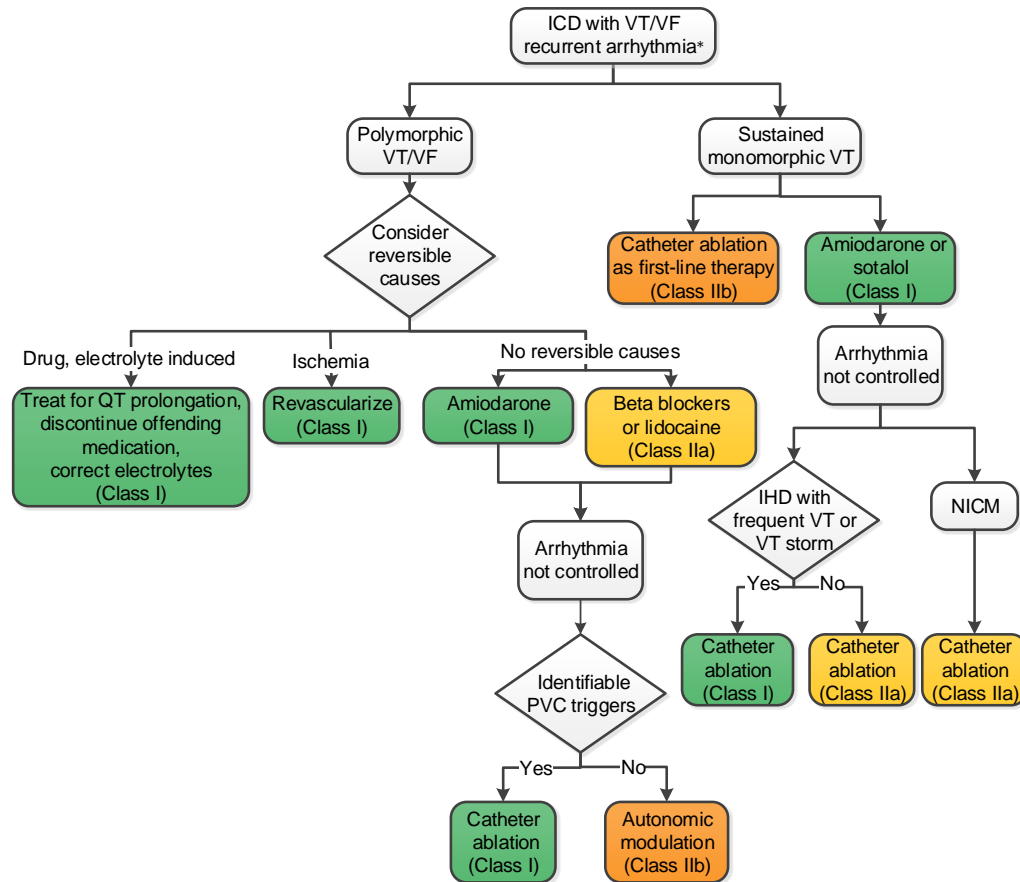
COR	LOE	Recommendations for Treatment of Recurrent VA in Patients With Ischemic Heart Disease
IIb	C-LD	3. In patients with ischemic heart disease and ICD shocks for sustained monomorphic VT or symptomatic sustained monomorphic VT that is recurrent, or hemodynamically tolerated, catheter ablation as first-line therapy may be considered to reduce recurrent VA.
III: Harm	B-R	4. In patients with prior MI, class IC antiarrhythmic medications (e.g., flecainide and propafenone) should not be used.
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.

Ischemic Heart Disease

Treatment and Prevention of Recurrent VA in Patients With Ischemic Heart Disease (contd.)

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

Treatment of Recurrent VA in Patients With Ischemic Heart Disease or NICM



Colors correspond to Class of Recommendation in Table 1.

*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.

APHRS indicates Asia Pacific Heart Rhythm Society; EHRA, 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.

Nonischemic Cardiomyopathy

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

Nonischemic Cardiomyopathy

Secondary Prevention of SCD in Patients With NICM

COR	LOE	Recommendations for Secondary Prevention of SCD in Patients With NICM
I	B-R	1. In patients with NICM who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R) or stable VT (LOE: B-NR) not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.
	B-NR	
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.
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.

Nonischemic Cardiomyopathy

Primary Prevention of SCD in Patients With NICM

COR	LOE	Recommendations for Primary Prevention of SCD in Patients With NICM
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.
IIa	B-NR	2. In patients with NICM due to a <i>Lamin A/C</i> 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.

Nonischemic Cardiomyopathy

Primary Prevention of SCD in Patients With NICM (contd.)

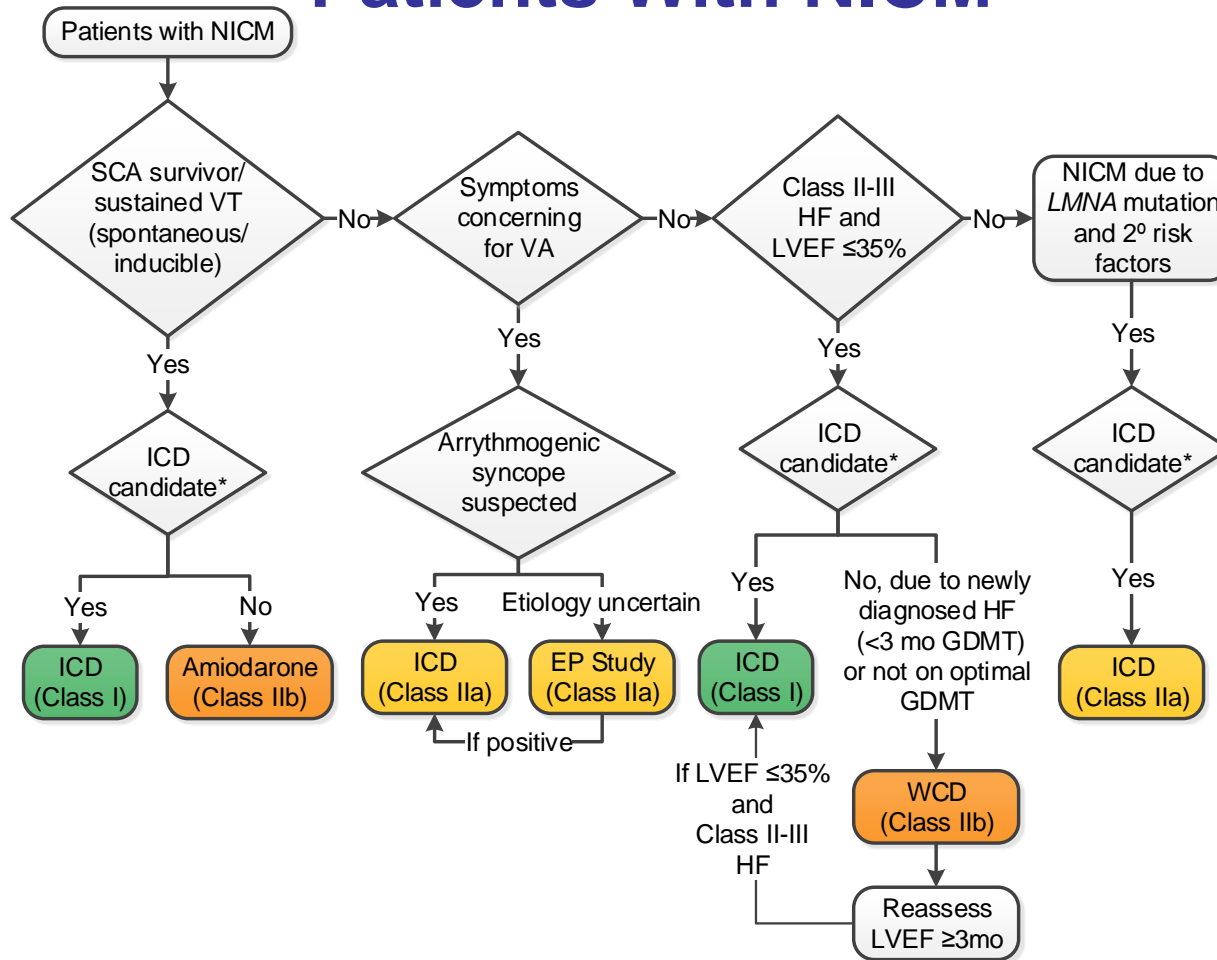
COR	LOE	Recommendations for Primary Prevention of SCD in Patients With NICM
IIb	B-R	3. In patients with NICM, HF with NYHA class I symptoms and an LVEF of 35% or less, despite GDMT, an ICD may be considered if meaningful survival of greater than 1 year is expected.
III: No Benefit	C-EO	4. In patients 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.

Nonischemic Cardiomyopathy

Treatment of Recurrent VA in Patients With NICM

COR	LOE	Recommendations for Treatment of Recurrent VA in Patients With NICM
Ila	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.
Ila	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.

Secondary and Primary Prevention of SCD in Patients With NICM



Colors correspond to Class of Recommendation in Table 1.

*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.

Arrhythmogenic Right Ventricular Cardiomyopathy

COR	LOE	Recommendations for Arrhythmogenic Right Ventricular Cardiomyopathy
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.
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.
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 $\leq 35\%$), an ICD is recommended if meaningful survival greater than 1 year is expected.

Arrhythmogenic Right Ventricular Cardiomyopathy (contd.)

COR	LOE	Recommendations for Arrhythmogenic Right Ventricular Cardiomyopathy
I	B-NR	4. In patients with arrhythmogenic right ventricular cardiomyopathy and VA, a beta blocker is recommended.
I	B-NR	5. In patients with a clinical diagnosis of arrhythmogenic right ventricular cardiomyopathy, avoiding intensive exercise is recommended.
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.
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.

Arrhythmogenic Right Ventricular Cardiomyopathy (contd.)

COR	LOE	Recommendations for Arrhythmogenic Right Ventricular Cardiomyopathy
IIa	B-NR	8. In patients with clinical evidence of arrhythmogenic right ventricular cardiomyopathy but not VA, a beta blocker can be useful.
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.
IIa	B-NR	10. In patients with suspected arrhythmogenic right ventricular cardiomyopathy, a signal averaged ECG can be useful for diagnosis and risk stratification.
IIb	B-NR	11. In asymptomatic patients with clinical evidence of arrhythmogenic right ventricular cardiomyopathy, an electrophysiological study may be considered for risk stratification.

Hypertrophic Cardiomyopathy

COR	LOE	Recommendations for HCM
I	B-NR	1. In patients with HCM, SCD risk stratification should be performed at the time of initial evaluation and periodically thereafter.
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.
I	B-NR	3. In first-degree relatives of patients with HCM, an ECG and echocardiogram should be performed.
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.

Hypertrophic Cardiomyopathy (contd.)

COR	LOE	Recommendations for HCM
Ila	B-NR	5. In patients with clinically suspected or diagnosed HCM, genetic counseling and genetic testing are reasonable.
Ila	B-NR	6. In patients with HCM and 1 or more of the following risk factors, an ICD is reasonable if meaningful survival of greater than 1 year is expected:
	C-LD	a. Maximum LV wall thickness ≥ 30 mm (LOE: B-NR).
	C-LD	b. SCD in 1 or more first-degree relatives presumably caused by HCM (LOE: C-LD).
	C-LD	c. 1 or more episodes of unexplained syncope within the preceding 6 months (LOE: C-LD).

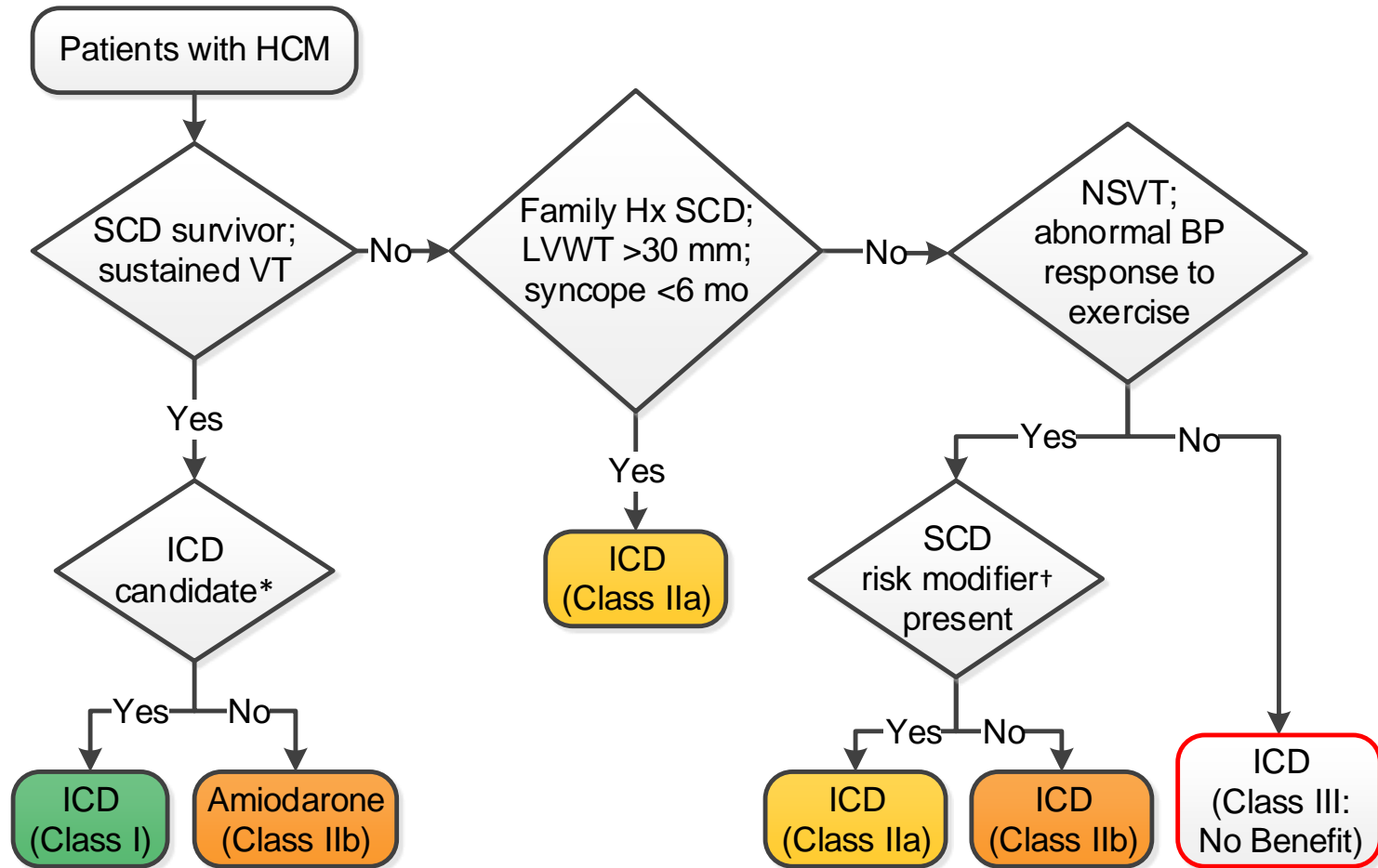
Hypertrophic Cardiomyopathy (contd.)

COR	LOE	Recommendations for HCM
IIa	B-NR	7. In patients with HCM who have spontaneous NSVT (LOE: C-LD) or an abnormal blood pressure response with exercise (LOE: B-NR), who also have additional SCD risk modifiers or high risk features, an ICD is reasonable if meaningful survival greater than 1 year is expected.
	C-LD	
IIb	B-NR	8. In patients with HCM who have NSVT (LOE: B-NR) or an abnormal blood pressure response with exercise (LOE: B-NR) but do not have any other SCD risk modifiers, an ICD may be considered, but its benefit is uncertain.
	B-NR	
IIb	C-LD	9. In patients with HCM and a history of sustained VT or VF, amiodarone may be considered when an ICD is not feasible or not preferred by the patient .

Hypertrophic Cardiomyopathy (contd.)

COR	LOE	Recommendations for HCM
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.
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.

Prevention of SCD in Patients With HCM



Colors correspond to Class of Recommendation in Table 1.

*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.

Myocarditis

COR	LOE	Recommendations for Myocarditis
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.
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.

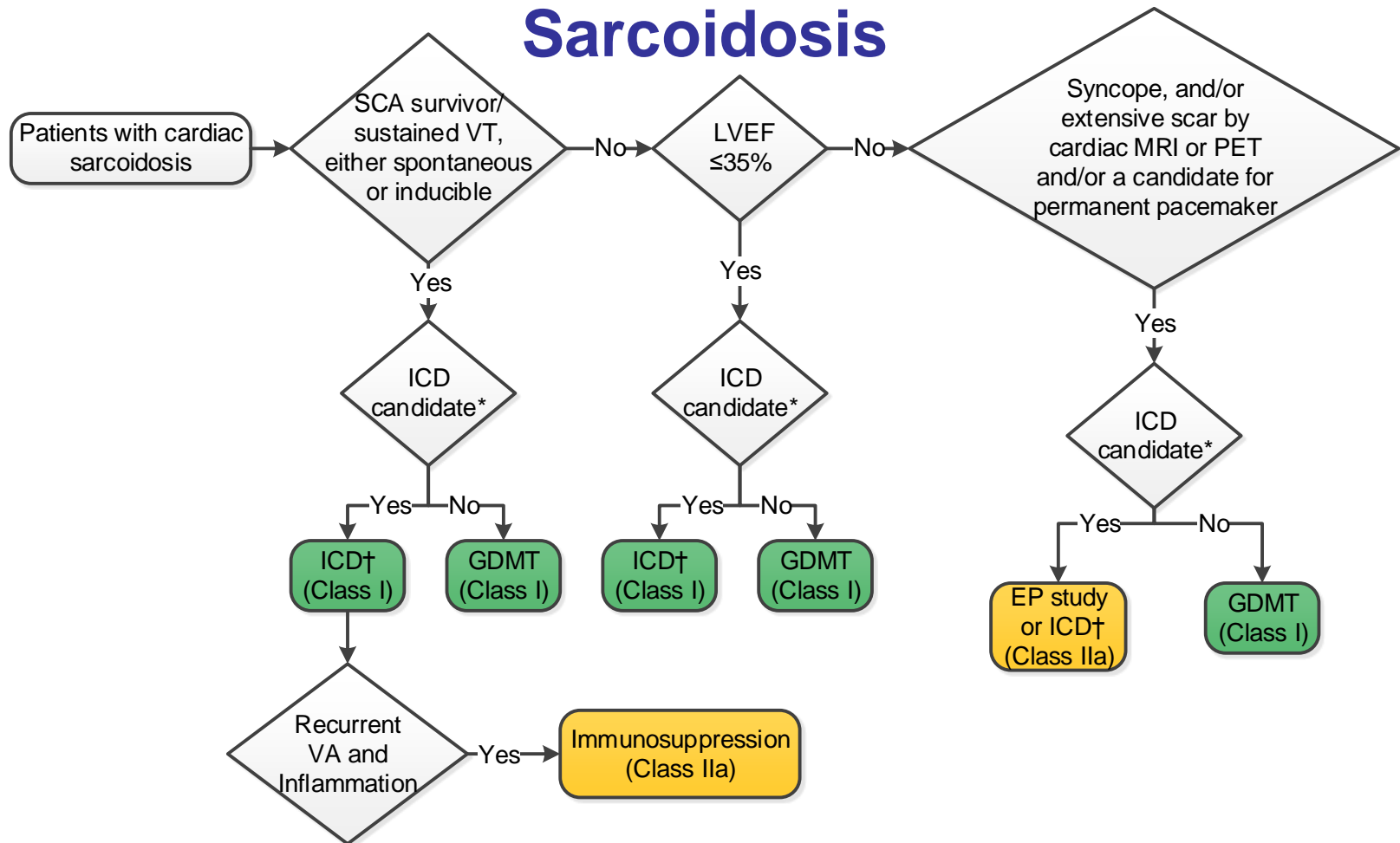
Cardiac Sarcoidosis

COR	LOE	Recommendations for Cardiac Sarcoidosis
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.
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.

Cardiac Sarcoidosis (contd.)

COR	LOE	Recommendations for Cardiac Sarcoidosis
Ila	C-LD	3. In patients with cardiac sarcoidosis and LVEF greater than 35%, it is reasonable to perform an electrophysiological study and to implant an ICD, if sustained VA is inducible, provided that meaningful survival of greater than 1 year is expected.
Ila	C-LD	4. In patients with cardiac sarcoidosis who have an indication for permanent pacing, implantation of an ICD can be beneficial.
Ila	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.

Prevention of SCD in Patients With Cardiac Sarcoidosis



Colors correspond to Class of Recommendation in Table 1.

*ICD candidacy as determined by functional status, life expectancy, or patient preference.

†For recurrent sustained monomorphic VT, refer to Figure Management of Sustained Monomorphic VT.

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.

Heart Failure

HF With Reduced Ejection Fraction

COR	LOE	Recommendation for HFrEF
IIa	B-NR	1. In patients with HFrEF 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.

Left Ventricular Assist Device

COR	LOE	Recommendation for Patients With an LVAD
IIa	C-LD	1. In patients with an LVAD and sustained VA, an ICD can be beneficial.

ICD Use After Heart Transplantation

COR	LOE	Recommendation for ICD Use After Heart Transplantation
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.

Neuromuscular Disorders

COR	LOE	Recommendations for Neuromuscular Disorders
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.
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.
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.
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.

Cardiac Channelopathies

COR	LOE	Recommendations for Cardiac Channelopathies
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.
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.

Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes

Congenital Long QT Syndrome

COR	LOE	Recommendations for Long QT Syndrome
I	B-NR	1. In patients with long QT syndrome with a resting QTc greater than 470 ms, a beta blocker is recommended.
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.

Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes Congenital Long QT Syndrome (contd.)

COR	LOE	Recommendations for Long QT Syndrome
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.
I	B-NR	4. In patients with clinically diagnosed long QT syndrome, genetic counseling and genetic testing are recommended.

Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes Congenital Long QT Syndrome (contd.)

COR	LOE	Recommendations for Long QT Syndrome
Ila	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.
Ila	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.

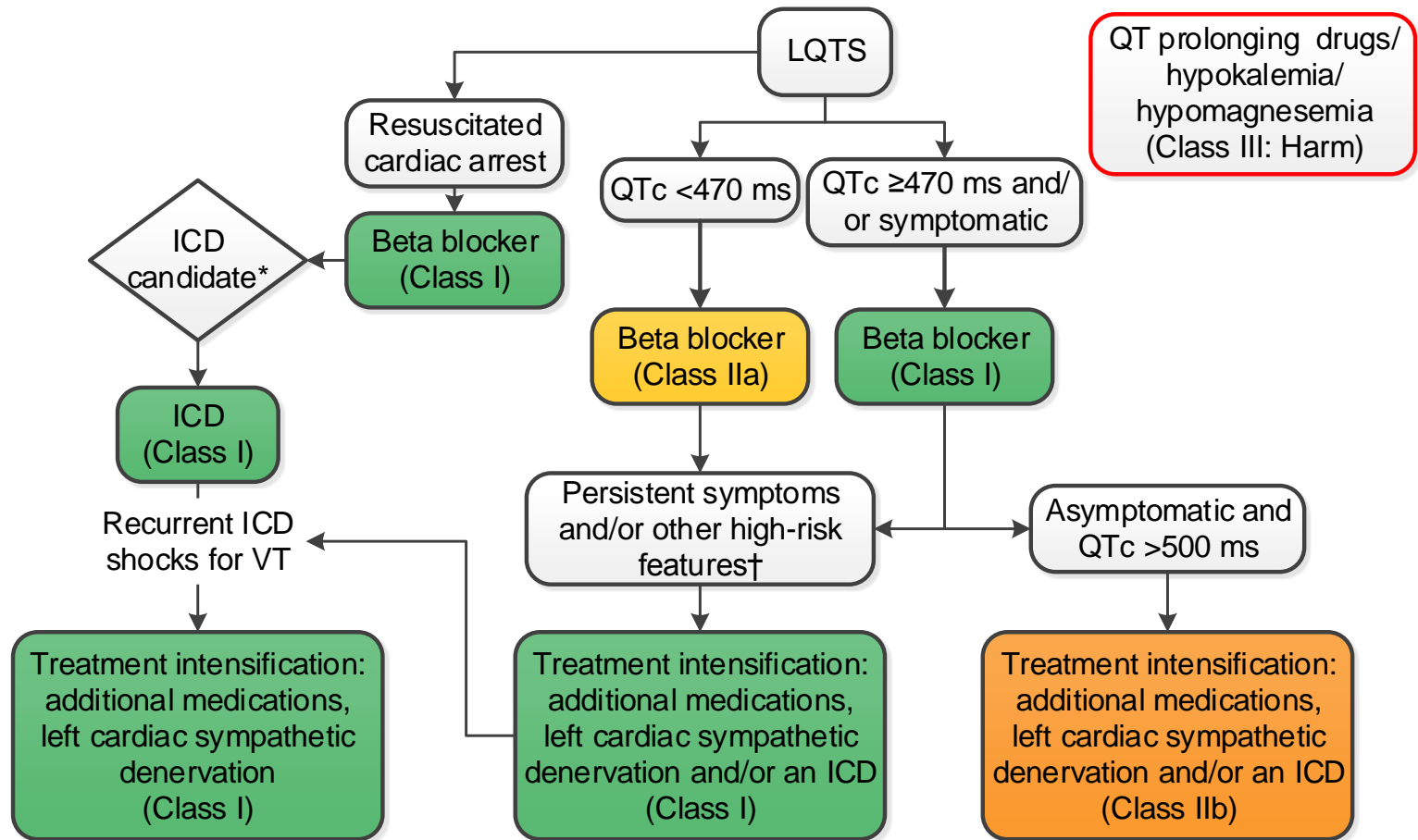
Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes

Congenital Long QT Syndrome (contd.)

COR	LOE	Recommendations for Long QT Syndrome
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.
III: Harm	B-NR	8. In patients with long QT syndrome, QT-prolonging medications are potentially harmful.

Prevention of SCD in Patients With Long QT Syndrome



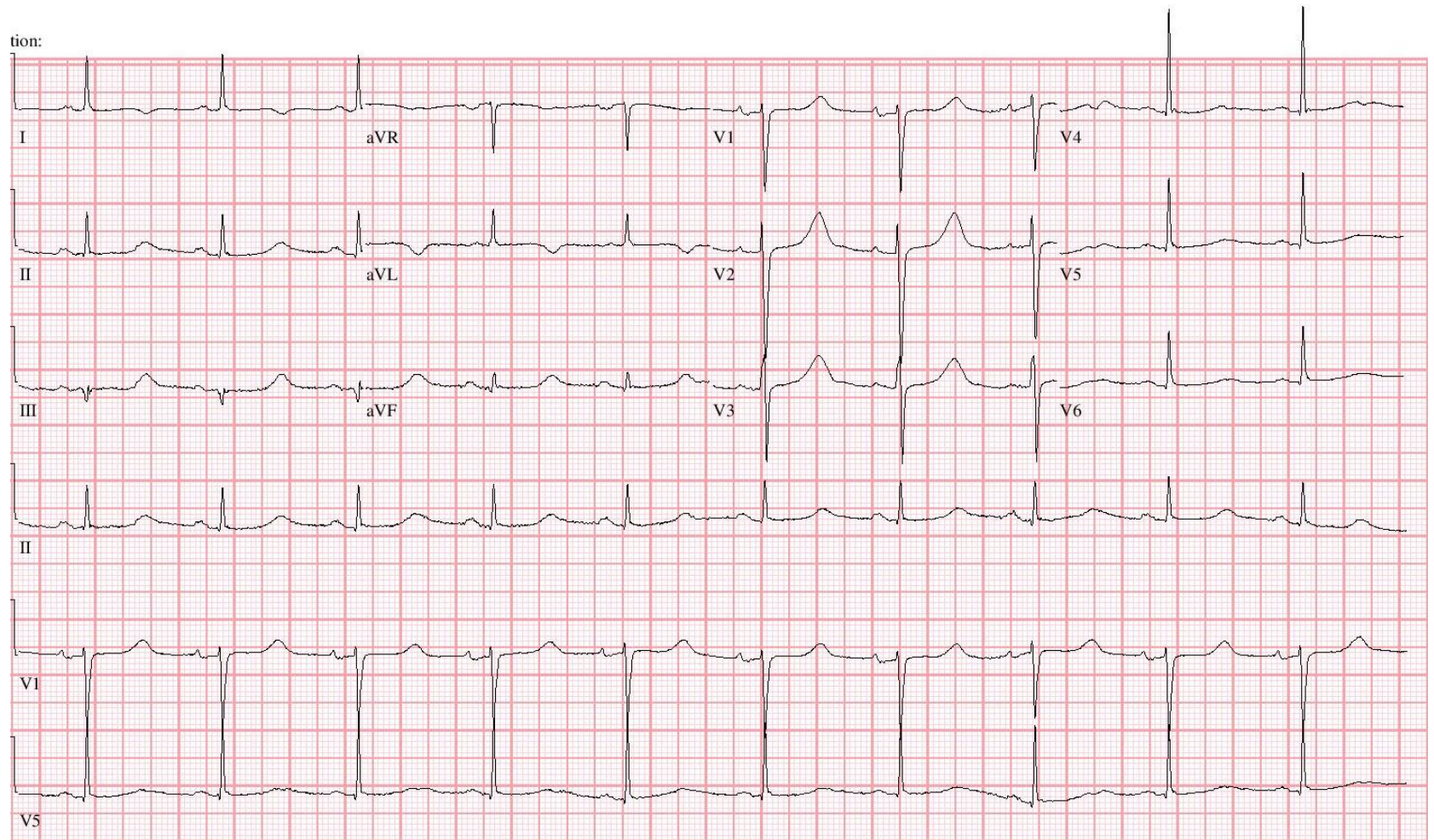
Colors correspond to Class of Recommendation in Table 1.

*ICD candidacy as determined by functional status, life expectancy, or patient preference.

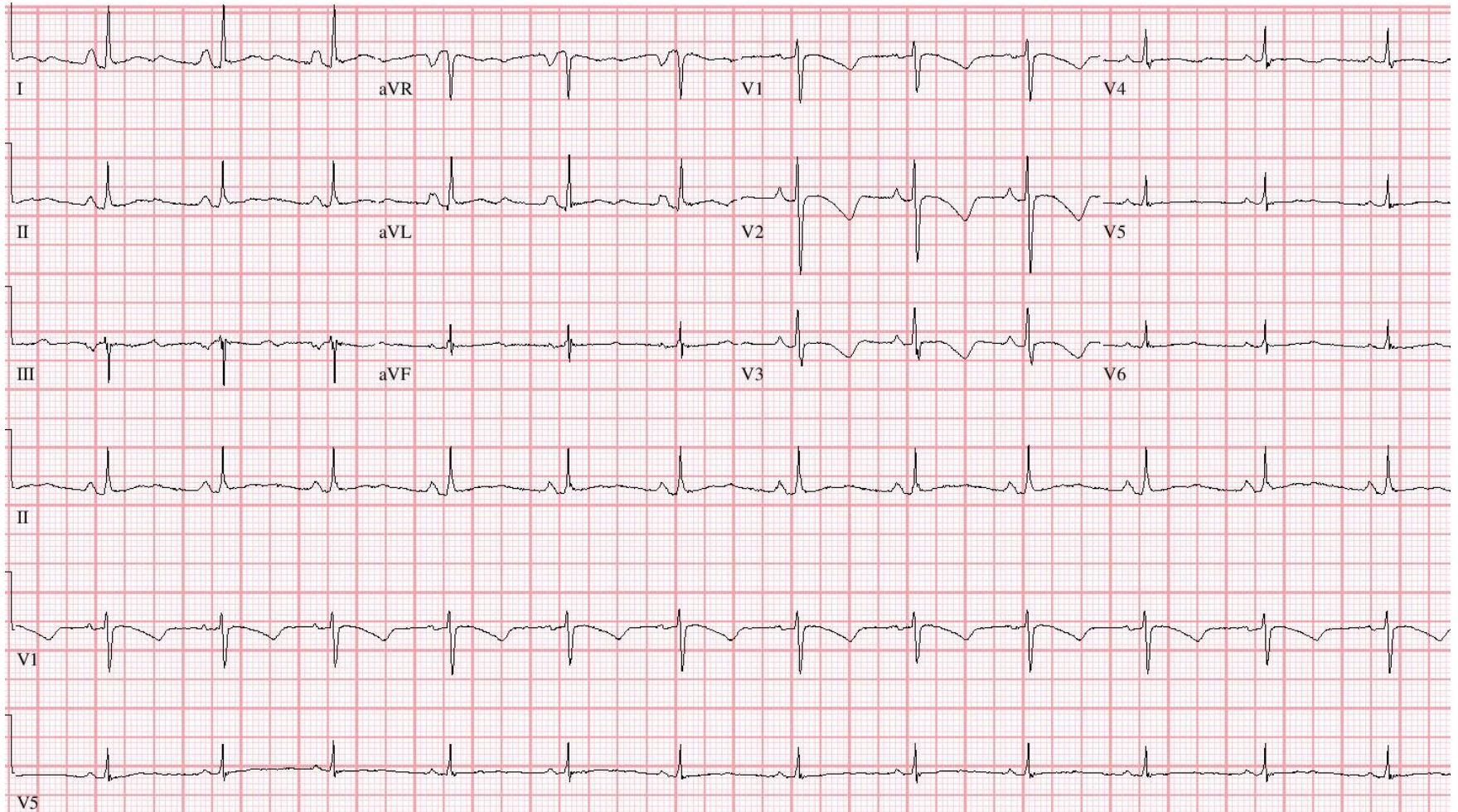
†High-risk patients with LQTS include those with QTc >500 ms, genotypes LQT2 and LQT3, females with genotype LQT2, <40 years of age, onset of symptoms at <10 years of age, and patients with recurrent syncope

ICD indicates implantable cardioverter-defibrillator; LQTS, long-QT syndrome; VT, ventricular tachycardia.

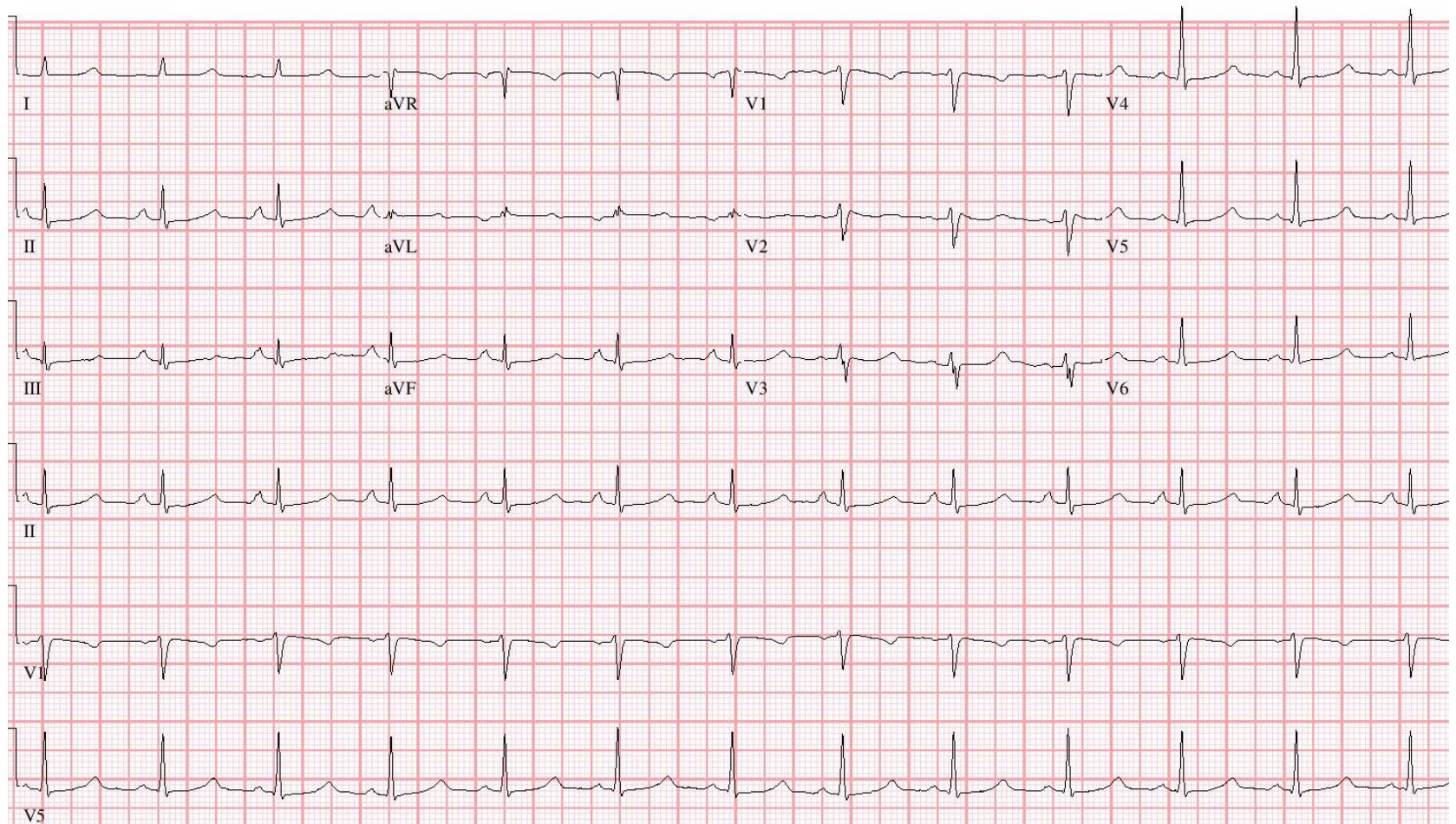
Long QT Syndrome Type 1



Long QT Syndrome Type 2



Long QT Syndrome Type 3



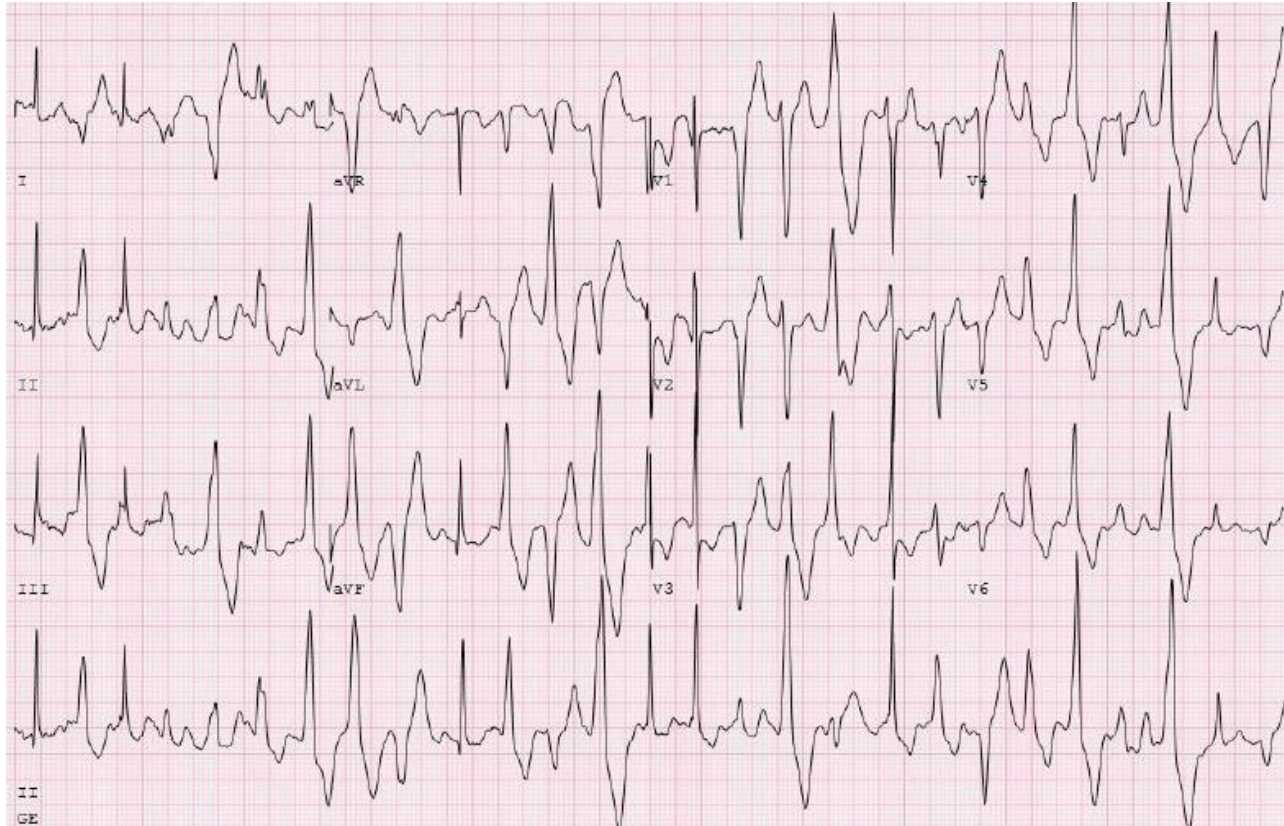
Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes

Catecholaminergic Polymorphic Ventricular Tachycardia

COR	LOE	Recommendations for Catecholaminergic Polymorphic Ventricular Tachycardia
I	B-NR	1. In patients with catecholaminergic polymorphic ventricular tachycardia, a beta blocker is recommended.
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.
Ia	B-NR	3. In patients with catecholaminergic polymorphic ventricular tachycardia and with clinical VT or exertional syncope, genetic counseling and genetic testing are reasonable.

Exercise-Induced Polymorphic VT in Catecholaminergic Polymorphic Ventricular Tachycardia



Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes

Brugada Syndrome

COR	LOE	Recommendations for Brugada Syndrome
I	B-NR	1. In asymptomatic patients with only inducible type 1 Brugada electrocardiographic pattern, observation without therapy is recommended.
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.
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.

Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes

Brugada Syndrome (contd.)

COR	LOE	Recommendations for Brugada Syndrome
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.
Ila	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.

Cardiac Channelopathies

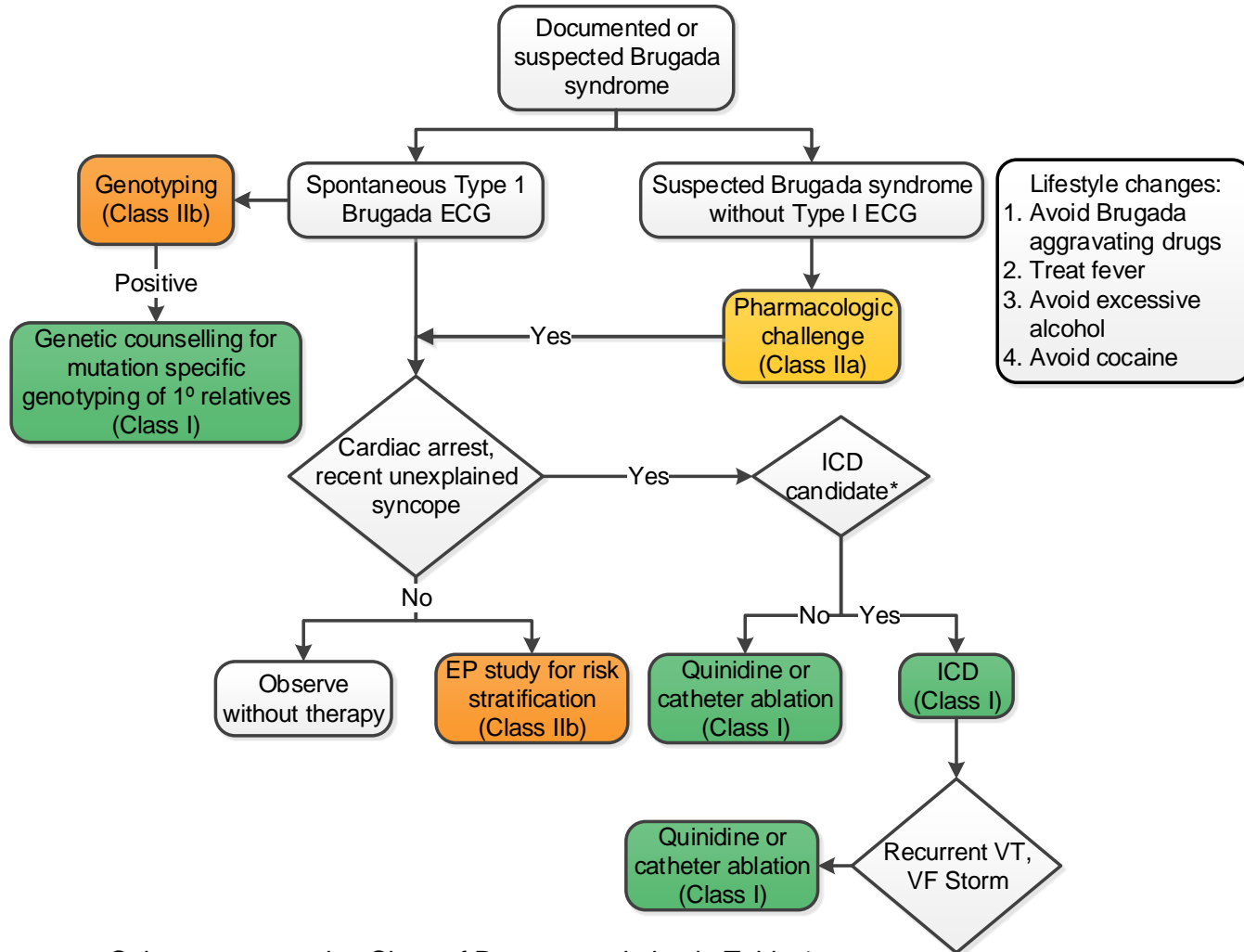
Specific Cardiac Channelopathy Syndromes

Brugada Syndrome (contd.)

COR	LOE	Recommendations for Brugada Syndrome
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.
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.

SR indicates systematic review.

Prevention of SCD in Patients With Brugada Syndrome

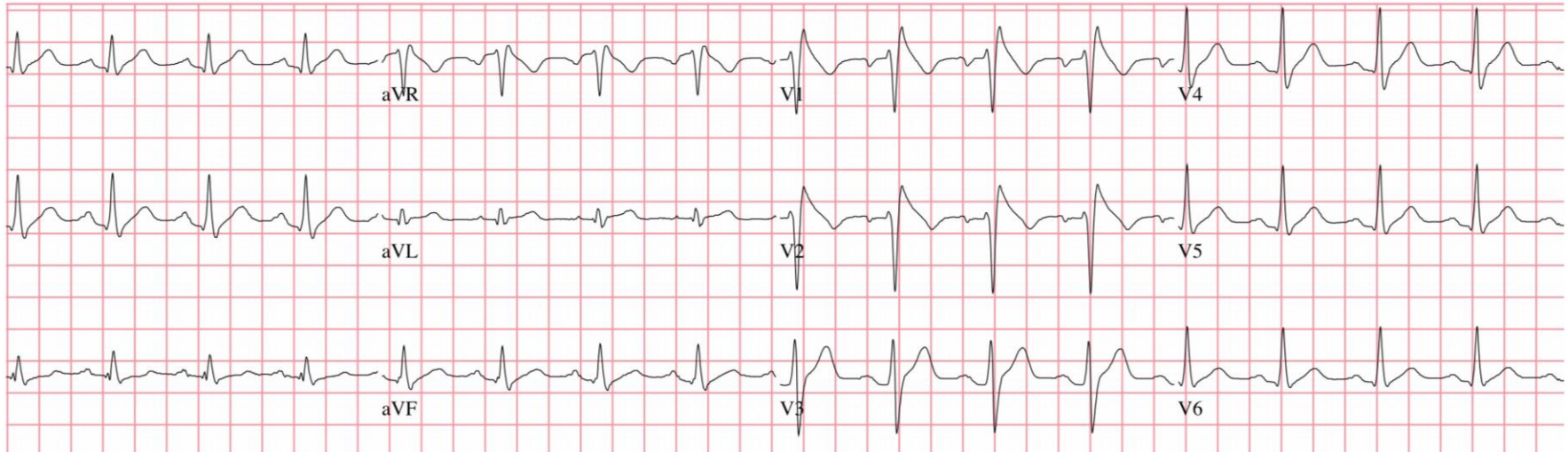


Colors correspond to Class of Recommendation in Table 1.

*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.

Brugada Syndrome



Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes

Early Repolarization “J-wave” Syndrome

COR	LOE	Recommendations for Early Repolarization Syndrome
I	B-NR	1. In asymptomatic patients with an early repolarization pattern on ECG, observation without treatment is recommended.
I	B-NR	2. In patients with early repolarization pattern on ECG and cardiac arrest or sustained VA, an ICD is recommended.
III: No Benefit	B-NR	3. In patients with early repolarization pattern on ECG, genetic testing is not recommended.

Cardiac Channelopathies

Specific Cardiac Channelopathy Syndromes

Short QT Syndrome

COR	LOE	Recommendations for Short QT Syndrome
I	B-NR	1. In asymptomatic patients with a short QTc interval, observation without treatment is recommended.
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.
IIa	C-LD	3. In patients with short QT syndrome and recurrent sustained VA, treatment with quinidine can be useful.
IIa	C-LD	4. In patients with short QT syndrome and VT/VF storm, isoproterenol infusion can be effective.
IIb	C-EO	5. In patients with short QT syndrome, genetic testing may be considered to facilitate screening of first-degree relatives.

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VA in the Structurally Normal Heart

VA in the Structurally Normal Heart

COR	LOE	Recommendations for VA in the Structurally Normal Heart
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.
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.

Outflow Tract and Atrioventricular Annular VA

COR	LOE	Recommendations for Outflow Tract VA
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
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.

Papillary Muscle VA

COR	LOE	Recommendation for Papillary Muscle VA (PVCs and VT)
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.

Interfascicular Reentrant VT (Belhassen Tachycardia)

COR	LOE	Recommendations for Interfascicular Reentrant VT (Belhassen Tachycardia)
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.
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.
IIa	C-LD	3. In patients with recurrent verapamil-sensitive idiopathic LVT, chronic therapy with oral verapamil can be useful.

Idiopathic Polymorphic VT/VF

COR	LOE	Recommendations for Idiopathic Polymorphic VT/VF
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.
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.
I	B-NR	3. For patients with recurrent episodes of idiopathic VF initiated by PVCs with a consistent QRS morphology, catheter ablation is useful.

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PVC-Induced Cardiomyopathy

PVC-Induced Cardiomyopathy

COR	LOE	Recommendations for PVC-Induced Cardiomyopathy
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.
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.

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VA and SCD Related to Specific Populations

Pregnancy

COR	LOE	Recommendations for Pregnancy
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 breast feeding.
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.
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.

Older Patients With Comorbidities

COR	LOE	Recommendation for Older Patients With Comorbidities
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.

SR indicates systematic review.

Medication-Induced Arrhythmias

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.
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.
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.

Medication-Induced Arrhythmias

Medication-Induced QT Prolongation and Torsades de Pointes

COR	LOE	Recommendations
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.
Sodium Channel Blocker-Related Toxicity		
COR	LOE	Recommendations
Ila	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.
III: Harm	B-NR	6. In patients with congenital or acquired long QT syndrome, QT-prolonging medications are potentially harmful.

Adult Congenital Heart Disease

COR	LOE	Recommendations for Adult Congenital Heart Disease
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.
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.
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 .

Adult Congenital Heart Disease (contd.)

COR	LOE	Recommendations for Adult Congenital Heart Disease
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.
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.
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.
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.

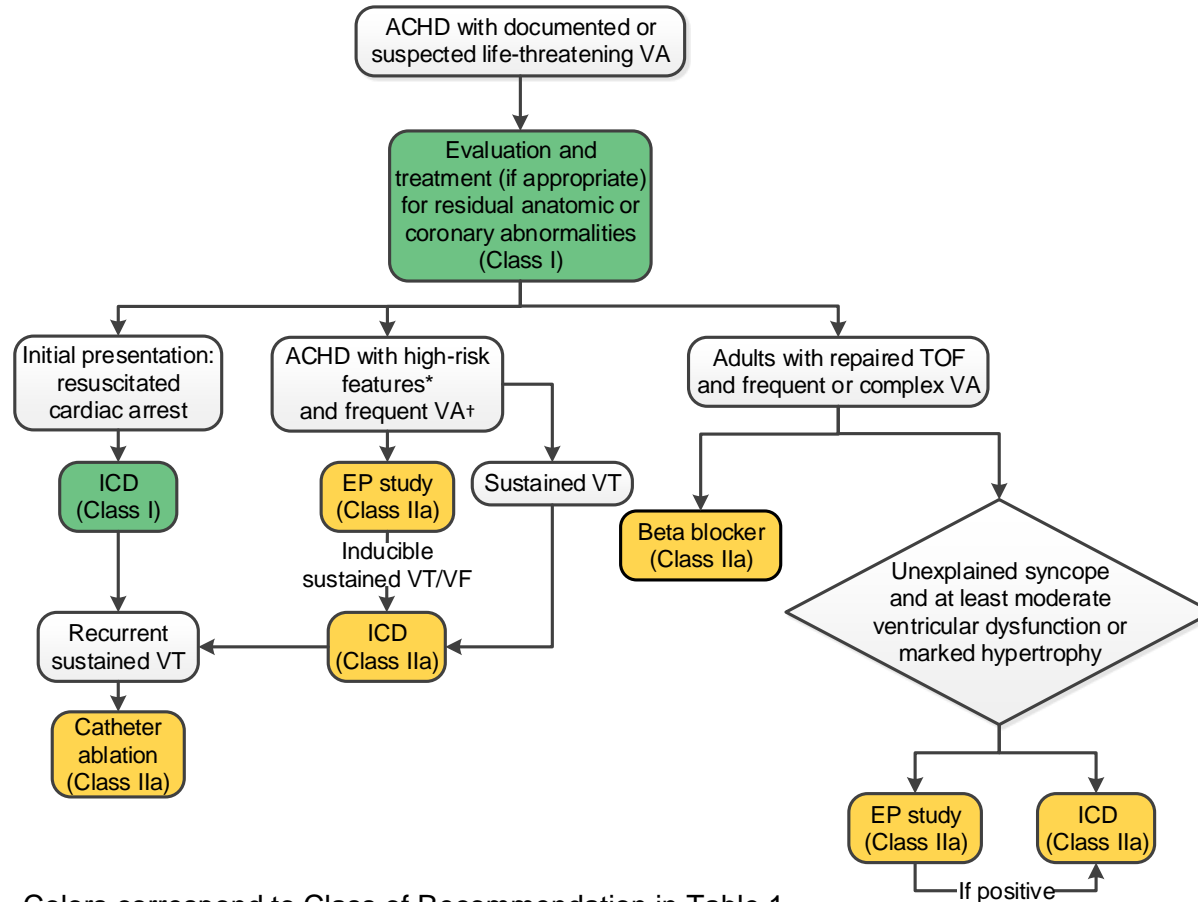
Adult Congenital Heart Disease (contd.)

COR	LOE	Recommendations for Adult Congenital Heart Disease
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.
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.
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.

Adult Congenital Heart Disease (contd.)

COR	LOE	Recommendations for Adult Congenital Heart Disease
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.

Prevention of SCD in Patients With Adult Congenital Heart Disease



Colors correspond to Class of Recommendation in Table 1.

*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.

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Defibrillators Other than Transvenous ICDs

Subcutaneous Implantable Cardioverter-Defibrillator

COR	LOE	Recommendations for Subcutaneous Implantable Cardioverter-Defibrillator
I	B-NR	1. In patients who meet criteria for an ICD who have inadequate vascular access or are at high risk for infection, and in whom pacing for bradycardia or VT termination or as part of CRT is neither needed nor anticipated, a subcutaneous implantable cardioverter-defibrillator is recommended.
Ia	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.
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 .

Wearable Cardioverter-Defibrillator

COR	LOE	Recommendations for Wearable Cardioverter-Defibrillator
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
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.

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Special Considerations for Catheter Ablation

Special Considerations for Catheter Ablation

COR	LOE	Recommendations for Catheter Ablation
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
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.

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Post-Mortem Evaluation of SCD

Post-Mortem Evaluation of SCD

COR	LOE	Recommendations for Postmortem Evaluation of SCD
I	B-NR	1. In victims of SCD without obvious causes, a standardized cardiac-specific autopsy is recommended.
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.
Ila	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.
Ila	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.

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Terminal Care

Terminal Care

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

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Shared Decision Making

Shared Decision Making

COR	LOE	Recommendations for Shared Decision-Making
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.
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.

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

