Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death (Management of Patients with)

Joint ESC/ACC/AHA Guidelines

Developed in Collaboration with the European Heart Rhythm Association and the Heart Rhythm Society
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ACC/AHA/ESC 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death—executive summary

A report of the American College of Cardiology/American Heart Association Task Force and the European Society of Cardiology Committee for Practice Guidelines (Writing Committee to Develop Guidelines for Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death)

Developed in collaboration with the European Heart Rhythm Association and the Heart Rhythm Society

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## Levels of recommendation

<table>
<thead>
<tr>
<th>Strength of recommendation</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Class I</strong></td>
<td>Evidence and/or general agreement that a given treatment or procedure is beneficial, useful and effective</td>
</tr>
<tr>
<td><strong>Class II</strong></td>
<td>Conflicting evidence and/or divergence of opinion about the usefulness/efficacy of a treatment or procedure</td>
</tr>
<tr>
<td><strong>IIa</strong></td>
<td>Weight of evidence/opinion is in favour of usefulness/efficacy</td>
</tr>
<tr>
<td><strong>IIb</strong></td>
<td>Usefulness/efficacy is less well established by evidence/opinion</td>
</tr>
<tr>
<td><strong>Class III</strong></td>
<td>Evidence and/or general agreement that a treatment/procedure is not useful/effective and in some cases may be harmful</td>
</tr>
<tr>
<td>Level of evidence</td>
<td>Available evidence</td>
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<tr>
<td>-------------------</td>
<td>--------------------------------------------------------</td>
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<tr>
<td>A</td>
<td>Multiple randomized clinical trials or meta-analyses</td>
</tr>
<tr>
<td>B</td>
<td>Single randomized trial or large non-randomized studies</td>
</tr>
<tr>
<td>C</td>
<td>Consensus opinion of experts, case studies, or standard-of-care</td>
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### Classification of ventricular arrhythmias by electrocardiography (1)

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nonsustained VT</td>
<td>Three or more beats in duration, terminating spontaneously in &lt; 30 sec. VT is a cardiac arrhythmia of ≥ 3 consecutive complexes in duration emanating from the ventricles at a rate &gt; 100 bpm (cycle length &lt; 600 msec)</td>
</tr>
<tr>
<td>Monomorphic VT</td>
<td>Nonsustained VT with a single QRS morphology</td>
</tr>
<tr>
<td>Polymorphic VT</td>
<td>Nonsustained VT with a changing QRS morphology at cycle length between 600 and 180 msec</td>
</tr>
<tr>
<td>Sustained VT</td>
<td>VT &gt; 30 sec in duration and/or requiring termination due to haemodynamic compromise in &lt; 30 sec.</td>
</tr>
<tr>
<td>Monomorphic sustained VT</td>
<td>Sustained VT with a stable single QRS morphology</td>
</tr>
<tr>
<td>Polymorphic sustained VT</td>
<td>Sustained VT with a changing or multiform QRS morphology at cycle length between 600 and 180 msec</td>
</tr>
<tr>
<td>Bundle branch re-entrant tachycardia</td>
<td>VT due to reentry involving the His-Purkinje system, usually with LBBB morphology; this usually occurs in the setting of cardiomyopathy</td>
</tr>
</tbody>
</table>

LBBB = left bundle branch block; VT = ventricular tachycardia.
### Classification of ventricular arrhythmias by electrocardiography (2)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bidirectional VT</td>
<td>VT with a beat-to-beat alternans in the QRS frontal plane axis, often associated with digitalis toxicity</td>
</tr>
</tbody>
</table>
| 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:  
  - ‘Typical’ initiated following “short long short” coupling intervals  
  - Short coupled variant initiated by normal short coupling |
| Ventricular flutter              | A regular (cycle length variability 30 msec or less) ventricular arrhythmia ~300 bpm (cycle length 200 msec) with a monomorphic appearance; no isoelectric interval between successive QRS complexes |
| Ventricular fibrillation         | Rapid, usually > 300 bpm/200 msec (cycle length 180 msec or less), grossly irregular ventricular rhythm with marked variability in QRS cycle length, morphology and amplitude |

VT = ventricular tachycardia.
Incidence of sudden cardiac death (SCD)

- Varies geographically as a function of coronary heart disease (CHD) prevalence in different regions:
  - US: < 200,000 to > 450,000 SCDs annually
  - Europe: overall similar level of event rates, but wide geographic variations reported

- ~50% of all CHD deaths are sudden and unexpected:
  - Occur shortly (instantaneous to 1 h) after the onset of a change in clinical status
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
  - Dyspnoea
  - Chest pain
  - Syncope and presyncope
- Ventricular tachycardia that is haemodynamically stable
- Ventricular tachycardia that is not haemodynamically stable
- Cardiac arrest
  - Asystolic (sinus arrest, atrioventricular block)
  - Ventricular tachycardia
  - Ventricular fibrillation
  - Pulseless electrical activity
General evaluation of patients with documented or suspected ventricular arrhythmias

Resting ECG

Class I  Resting 12-lead ECG is indicated in all patients who are evaluated for ventricular arrhythmias (VA) (LEVEL A)

Exercise testing

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 ischaemic changes or VA (LEVEL B)

2. ET is useful in patients regardless of age 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 B)

Class IIa  1. ET can be useful in evaluating response to medical or ablation therapy in patients with known exercise-induced VA (LEVEL B)
General evaluation of patients with documented or suspected ventricular arrhythmias

Exercise testing

Class IIb  1. ET might be useful in patients with VA and a low probability of CHD by age, gender, and symptoms (LEVEL C)
2. ET might be useful in the investigation of isolated premature ventricular complexes (PVCs) in middle-aged or older patients without other evidence of CHD (LEVEL C)

Ambulatory electrocardiography

Class I
1. 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 A)
2. Event monitors are indicated when symptoms are sporadic, to establish whether they are caused by transient arrhythmias (LEVEL 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 B)
General evaluation of patients with documented or suspected ventricular arrhythmias

ECG techniques and measurements

Class IIa  It is reasonable to use T-wave alternans for improving the diagnosis and risk stratification of patients with VA or at risk for developing life-threatening VA (LEVEL 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 VA or who are at risk of developing life-threatening VA (LEVEL B)

Left ventricular function and imaging

Class I  1. Echocardiography is recommended in patients with VA who are suspected of having structural heart disease (LEVEL 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 cardiomyopathies, acute myocardial infarction (AMI) survivors, or relatives of patients with inherited disorders associated with SCD (LEVEL B)
General evaluation of patients with documented or suspected ventricular arrhythmias

Left ventricular function and imaging

Class I
3. ET with an imaging modality (echocardiography or nuclear perfusion [single-photon emission computed tomography (SPECT)]) is recommended to detect silent ischaemia 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, > 1 mm ST-segment depression at rest, Wolff-Parkinson-White Syndrome or LBBB (LEVEL B)

4. Pharmacological stress testing with an imaging modality (echocardiography or myocardial perfusion SPECT) is recommended to detect silent ischaemia 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 B)

Class IIa
1. Magnetic resonance imaging (MRI), cardiac computed tomography (CT), or radionuclide angiography can be useful in patients with VA when echocardiography does not provide accurate assessment of LV and right ventricular (RV) function, and/or evaluation of structural changes (LEVEL 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 C)

3. LV imaging can be useful in patients undergoing biventricular pacing (LEVEL C)
General evaluation of patients with documented or suspected ventricular arrhythmias

Electrophysiological (EP) testing in patients with CHD

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

Class IIa
4. EP testing is reasonable for risk stratification in patients with remote MI, non sustained (NSVT) and LV ejection fraction (LVEF) ≤ 40% (LEVEL B)

Electrophysiological (EP) testing in patients with syncope

Class I
EP testing is recommended in patients with syncope of unknown cause with impaired LV function or structural heart disease (LEVEL B)
EP testing can be useful in patients with syncope when brady- or tachyarrhythmias are suspected, and in whom non-invasive diagnostic studies are not conclusive (LEVEL B)
Ablation for ventricular arrhythmias (1)

**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 C)
2. Ablation is indicated in patients with bundle-branch reentrant VT (LEVEL 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 the patient does not wish long-term drug-therapy (LEVEL C)
4. Ablation is indicated in patients with Wolff-Parkinson-White syndrome resuscitated from sudden cardiac arrest due to atrial fibrillation (AF) and rapid conduction over the accessory pathway causing ventricular fibrillation (VF) (LEVEL 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, or who are drug intolerant, or who do not wish long-term drug therapy (LEVEL C)
Ablation for ventricular arrhythmias (2)

Class IIa
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, or who are drug intolerant, or who do not wish long-term drug therapy (LEVEL C)
3. Ablation can be useful in symptomatic patients with Wolff-Parkinson-White syndrome who have accessory pathways with refractory periods < 240 ms in duration (LEVEL B)

Class IIb
1. Ablation of potentials from Purkinje fibres triggering VA storm may be considered in selected patients
2. Ablation of asymptomatic PVCs may be considered when the PVCs are very frequent to avoid or treat tachycardia-induced cardiomyopathy (LEVEL C)

Class III
Ablation of asymptomatic relatively infrequent PVCs is not indicated (LEVEL C)
Management of cardiac arrest (1)

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 B)

2. Cardiopulmonary resuscitation (CPR) should be implemented immediately after contacting a response team (LEVEL A)

3. In an out of hospital setting, if an automated external defibrillator (AED) is available, it should be applied immediately and shock therapy administered according to the algorithms contained in the documents on CPR developed by either the AHA in association with the International Liaison Committee on Resuscitation (ILCOR) and/or the European Resuscitation Council (ERC) (LEVEL C)

4. For victims with ventricular tachyarrhythmic mechanisms of cardiac arrest, when recurrences occur after a maximally defibrillating shock (generally 360 Joules for monophasic defibrillators), intravenous amiodarone should be the preferred antiarrhythmic drug for attempting to achieve a stable rhythm after further defibrillations (LEVEL B)
Management of cardiac arrest (2)

Class I
5. For recurrent ventricular tachyarrhythmias or nontachyarrhythmic mechanisms of cardiac arrest, it is recommended to follow the algorithms contained in the documents on CPR developed by either the AHA in association with the ILCOR and/or the ERC (LEVEL 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 C)

Class IIa
For response times ≥ 5 min, a brief (< 90 to 180 sec) period of CPR is reasonable prior to attempting defibrillation (LEVEL B)

Class IIb
A single precordial thump may be considered by healthcare professional providers when responding to a witnessed cardiac arrest (LEVEL C)
Ventricular tachycardia associated with low troponin MI

**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 C)

**Repetitive monomorphic VT**

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

Class I
1. Wide-QRS tachycardia should be presumed to be VT if the diagnosis is unclear (LEVEL C)
2. Direct-current (DC) cardioversion with appropriate sedation is recommended at any point in the treatment cascade in patients with suspected sustained monomorphic VT with haemodynamic compromise (LEVEL 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 B)
2. IV amiodarone is reasonable in patients with sustained monomorphic VT that is haemodynamically unstable, that is refractory to conversion with countershock, or recurrent despite procainamide or other agents (LEVEL 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 C)

Class IIb
IV lidocaine might be reasonable for the initial treatment of patients with stable sustained monomorphic VT specifically associated with acute myocardial ischaemia or infarction (LEVEL 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 C)
Polymorphic VT

Class I
1. DC cardioversion with appropriate sedation as necessary is recommended for patients with sustained polymorphic VT with haemodynamic compromise and is reasonable at any point in the treatment cascade (LEVEL B)
2. IV beta-blockers are useful for patients with recurrent polymorphic VT, especially if ischaemia is suspected or cannot be excluded (LEVEL 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 C)
4. Urgent angiography with a view to revascularization should be considered for patients with polymorphic VT when myocardial ischaemia cannot be excluded (LEVEL C)

Class IIb
IV lidocaine may be reasonable for treatment of polymorphic VT specifically associated with acute myocardial ischaemia or infarction (LEVEL C)
Torsades de Pointes

Class I
1. Withdrawal of any offending drugs and correction of electrolyte abnormalities are recommended in patients presenting with torsades de pointes (LEVEL A)
2. Acute and long-term pacing is recommended for patients presenting with torsades de pointes due to heart block and symptomatic bradycardia (LEVEL 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 B)
2. Acute and long-term pacing is reasonable for patients who present with recurrent pause-dependent torsades de pointes (LEVEL B)
3. Beta-blockade combined with pacing is reasonable acute therapy for patients who present with torsades de pointes and sinus bradycardia (LEVEL 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 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 B)
2. IV lidocaine or oral mexiletine may be considered in patients who present with LQT3 and torsades de pointes (LEVEL C)
Incessant VT

Class I  
Revascularization and beta-blockade, followed by antiarrhythmic drugs such as IV procainamide or IV amiodarone are recommended for patients with recurrent or incessant polymorphic VT due to acute myocardial ischaemia (LEVEL 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 B)

Class IIb  
1. IV amiodarone and IV beta-blockers separately or together may be reasonable in patients with VT storm (LEVEL C)
2. Overdrive pacing or general anesthesia may be considered for patients with frequently recurring or incessant VT (LEVEL C)
3. Spinal cord modulation may be considered for some patients with frequently recurring or incessant VT (LEVEL C)
Left ventricular dysfunction due to prior MI (1)

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 C)

2. Aggressive attempts should be made to treat myocardial ischaemia that may be present in some patients with ventricular tachyarrhythmias (LEVEL C)

3. Coronary revascularization is indicated to reduce the risk of SCD in patients with VF when direct, clear evidence of acute myocardial ischaemia is documented to immediately precede the onset of VF (LEVEL 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 who have reasonable expectation of survival with a good functional status for > 1 year (LEVEL 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 ≤ 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 > 1 year (LEVEL 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 haemodynamically unstable sustained VT, who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for > 1 year (LEVEL A)
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 ≤ 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 > 1 year (LEVEL 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 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 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 C)

5. Amiodarone is reasonable therapy to reduce symptoms due to recurrent haemodynamically stable VT for patients with LVD due to prior MI who cannot or refuse to have an ICD implanted (LEVEL 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 > 1 year (LEVEL C)
Left ventricular dysfunction due to prior MI (3)

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 haemodynamically stable VT whose LVEF is > 40% (LEVEL 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 C)

Class III
1. Prophylactic antiarrhythmic drug therapy is not indicated to reduce mortality in patients with asymptomatic nonsustained VA (LEVEL B)
2. Class IC antiarrhythmic drugs in patients with a past history of MI should not be used (LEVEL A)
Valvular heart disease

Class I  Patients with valvular heart disease and VA should be evaluated and treated following current recommendations for each disorder (LEVEL 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 C)
Congenital heart disease

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 > 1 year (LEVEL B)

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

Class IIa
Invasive haemodynamic 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 > 1 year (LEVEL B)

Class IIb
EP testing may be considered for patients with congenital heart disease and ventricular couplets or NSVT to determine the risk of a sustained VA (LEVEL C)

Class III
Prophylactic antiarrhythmic therapy is not indicated for asymptomatic patients with congenital heart disease and isolated PVCs (LEVEL C)
Pericardial diseases

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 > 1 year (LEVEL C)

Pulmonary arterial hypertension

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 C)
Transient arrhythmias of reversible cause

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 ischaemia or MI (LEVEL 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 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 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 (LEVEL B)
Dilated cardiomyopathy (DCM) (Nonischaemic) (1)

Class I

1. EP testing is useful to diagnose bundle branch-reentrant tachycardia, and to guide ablation in patients with nonischaemic DCM (LEVEL C)

2. EP testing is useful for diagnostic evaluation in patients with nonischaemic DCM with sustained palpitations, wide QRS-complex tachycardia, syncope or presyncope (LEVEL C)

3. An ICD should be implanted in patients with nonischaemic 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 > 1 year (LEVEL A)

4. ICD therapy is recommended for primary prevention to reduce total mortality by a reduction in SCD in patients with nonischaemic DCM who have an LVEF ≤ 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 > 1 year (LEVEL B)
Dilated cardiomyopathy (DCM) (Nonischaemic) (2)

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

Class IIb
1. Amiodarone may be considered for sustained VT or VF in patients with nonischaemic DCM (LEVEL C)
2. Placement of an ICD might be considered in patients who have nonischaemic DCM, LVEF ≤ 30% to 35%, are NYHA functional class I receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for > 1 year (LEVEL C)
Hypertrophic cardiomyopathy (HCM)

Class I
ICD therapy should be used for treatment in patients with 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 > 1 year (LEVEL B)

Class Ila
1. ICD implantation can be effective for primary prophylaxis against SCD in patients with HCM who have one or more major risk factor (See Slide 34) for SCD and who are receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for > 1 year (LEVEL 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 C)

Class Iib
1. EP testing may be considered for risk assessment for SCD in patients with HCM (LEVEL 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 Slide 34), if ICD implantation is not feasible (LEVEL C)
Risk factors for SCD in HCM

Major risk factors
- Cardiac arrest (VF)
- Spontaneous sustained VT
- Family history of premature sudden death
- Unexplained syncope
- LV thickness ≥ 30 mm
- Abnormal exercise BP
- Nonsustained spontaneous VT

Possible in individual patients
- AF
- Myocardial ischaemia
- LV outflow obstruction
- High-risk mutation
- Intense (competitive) physical exertion


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

**Class II**
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 > 1 year (LEVEL 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/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 > 1 year (LEVEL C)
2. Amiodarone or sotalol can be effective for treatment of sustained VT/VF in patients with ARVC when ICD implantation is not feasible (LEVEL C)
3. Ablation can be useful as adjunctive therapy in management of patients with ARVC with recurrent VT, despite optimal anti-arrhythmic drug therapy (LEVEL C)

**Class IIb**
EP testing might be useful for risk assessment of SCD in patients with ARVC (LEVEL C)
Heart failure (1)

Class I
1. ICD therapy is recommended for secondary prevention of SCD in patients who survived VF or haemodynamically unstable VT, or VT with syncope and have an LVEF ≤ 40%, who are receiving chronic optimal medical therapy and who have a reasonable expectation of survival with a good functional status for > 1 year (LEVEL 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 ≤ 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 > 1 year (LEVEL A)

3. ICD therapy is recommended for primary prevention to reduce total mortality by a reduction in SCD in patients with nonischaemic heart disease who have an LVEF ≤ 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 > 1 year (LEVEL 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 heart failure (HF) (LEVEL C)

5. Amiodarone is indicated for the suppression of acute haemodynamically 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 B)
Heart failure (2)

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 > 1 year (LEVEL 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 ≤ 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 > 1 year (LEVEL 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 > 1 year (LEVEL 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 ≤ 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 > 1 year (LEVEL B)
Heart failure (3)

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 C)

2. ICD therapy may be considered for primary prevention to reduce total mortality by a reduction in SCD in patients with nonischaemic heart disease who have an LVEF of ≤ 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 > 1 year (LEVEL B)
Long QT syndrome

Class I
1. Life style modification is recommended for patients with an LQTS diagnosis (clinical and/or molecular) (LEVEL B)
2. Beta-blockers are recommended for patients with an LQTS clinical diagnosis (i.e., in the presence of prolonged QT interval) (LEVEL 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 > 1 year (LEVEL A)

Class IIa
1. Beta-blockers can be effective to reduce SCD in patients with a molecular LQTS analysis and normal QT interval (LEVEL 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 > 1 year (LEVEL 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 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 > 1 year (LEVEL B)
**Brugada syndrome**

**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 > 1 year (LEVEL 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 > 1 year (LEVEL 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 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 > 1 year (LEVEL C)
4. Isoproterenol can be useful to treat an electrical storm in the Brugada syndrome (LEVEL 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 C)
2. Quinidine might be reasonable for the treatment of electrical storm in patients with Brugada syndrome (LEVEL C)
Catecholaminergic polymorphic ventricular tachycardia

Class I
1. Beta-blockers are indicated for patients who are clinically diagnosed with catecholaminergic polymorphic VT (CPVT) based on the presence of spontaneous or documented stress-induced VA (LEVEL 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 > 1 year (LEVEL C)

Class Ila
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 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 > 1 year (LEVEL 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 C)
Athletes

Class I
1. Pre-participation 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 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 recognizing the potential uniqueness of their activity (LEVEL C)
3. Athletes presenting with syncope should be carefully evaluated to uncover underlying cardiovascular disease or rhythm disorder (LEVEL B)
4. Athletes with serious symptoms should cease competition while cardiovascular abnormalities are being fully evaluated (LEVEL C)

Class IIb
12-lead ECG and possibly echocardiography may be considered as pre-participation screening for heart disorders in athletes (LEVEL B)
Gender & pregnancy

Class I 1. Pregnant women developing haemodynamically unstable VT or VF should be electrically cardioverted or defibrillated (LEVEL 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 C)

Elderly patients

Class I 1. Elderly patients with VA should generally be treated in the same manner as younger individuals (LEVEL 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 C)

Class III Elderly patients with projected life expectancy < 1 year due to major comorbidities should not receive ICD therapy (LEVEL C)
Patients with ICDs

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

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

Class I
An anti-digitalis antibody is recommended for patients who present with sustained ventricular arrhythmias, advanced AV block, and/or asystole that are considered due to digitalis toxicity (LEVEL 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 C)
2. Magnesium or pacing is reasonable for patients who take digitalis and present with severe toxicity* (LEVEL C)

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

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

* Sustained VA, advanced AV block, and/or asystole.
Drug-induced arrhythmias: 
drug-induced LQTS

**Class I**
In patients with drug-induced LQTS, removal of the offending agent is indicated (LEVEL 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 B)
2. Atrial or ventricular pacing or isoproterenol is reasonable for patients taking QT-prolonging drugs who present with recurrent torsades de pointes (LEVEL 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 C)
Drug-induced arrhythmias:
sodium-channel blocker-related toxicity

Class I
In patients with sodium-channel blocker-related toxicity, removal of the offending agent is indicated (LEVEL 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 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 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 C)