

AMERICAN COLLEGE of CARDIOLOGY FOUNDATION



ACC/AHA Pocket Guideline

Based on the ACC/AHA/HRS 2008 Guidelines

Device-Based Therapy of Cardiac Rhythm Abnormalities

May 2008

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Device-Based Therapy of Cardiac Rhythm Abnormalities

May 2008

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I. Introduction

The focus of these guidelines is the appropriate use of heart rhythm devices (pacemakers for bradyarrhythmias and heart failure management, e.g., cardiac resynchronization, and implantable cardioverter-defibrillators [ICDs]), not the treatment of cardiac arrhythmias. The fact that the use of a device for treatment of a particular condition is listed as a Class I indication (beneficial, useful, and effective) does not preclude the use of other therapeutic modalities that may be equally effective. As with all clinical practice guidelines, the recommendations in this document focus on treatment of an average patient with a specific



disorder and may be modified by patient comorbidities, limitation of life expectancy because of coexisting diseases, and other situations that only the primary treating physician may evaluate appropriately.

The recommendations listed in this document are, whenever possible, evidence based. The schema for classification of recommendations and level of evidence is summarized in *Table 1*, which also illustrates how the grading system provides an estimate of the size of the treatment effect and an estimate of the certainty of the treatment effect.

The following represents a subset of recommendations for the implantation of antiarrhythmia devices. We chose to provide only recommendations in the pocket guide and recommend readers access the full-text and executive summary documents for more explanation. *Table 2* and *Figures 1 and 2* are provided to help practitioners choose which pacing device is appropriate for an individual patient.

Table 1. Applying Classification ofRecommendations and Level of Evidence

SIZE OF TREATMENT EFFECT

| | CLASS I Benefit >>> Risk Procedure/Treatment SHOULD be performed/ administered | CLASS IIA Benefit >> Risk Additional studies with focused objectives needed IT IS REASONABLE to per- form procedure/administer treatment |
|---|--|---|
| LEVEL A Multiple populations evaluated* Data derived from multi- ple randomized clinical trials or meta-analyses | Recommendation that procedure or treatment is useful/effective Sufficient evidence from multiple randomized trials or meta-analyses | Recommendation in favor of treatment or procedure being useful/effective Some conflicting evidence from multiple randomized trials or meta-analyses |
| LEVEL B Limited populations evaluated* Data derived from a single randomized trial or nonrandomized studies | Recommendation that procedure or treatment is useful/effective Limited evidence from single randomized trial or nonrandomized studies | Recommendation in favor of treatment or procedure being useful/effective Some conflicting evidence from single randomized trial or nonrandomized studies |
| LEVEL C Very limited populations evaluated* Only consensus opinion of experts, case studies, or standard of care | Recommendation that procedure or treatment is useful/effective Only expert opinion, case studies, or standard of care | Recommendation in favor of treatment or procedure being useful/effective Only diverging expert opinion, case studies, or standard of care |
| Suggested phrases for writing recommendations [†] | should is recommended is indicated is useful/effective/beneficial | is reasonable can be useful/effective/beneficial is probably recommended or indicated |

Class IIb

Benefit \geq Risk Additional studies with broad objectives needed; additional registry data would be helpful

Procedure/Treatment MAY BE CONSIDERED

 Recommendation's usefulness/efficacy less well established

 Greater conflicting evidence from multiple randomized trials or meta-analyses

Recommendation's usefulness/efficacy less well established

 Greater conflicting evidence from single randomized trial or nonrandomized studies

 Recommendation's usefulness/efficacy less well established

Only diverging expert opinion, case studies, or standard of care

may/might be considered may/might be reasonable usefulness/effectiveness is unknown /unclear/uncertain or not well established

Class III Risk≥Benefit

Procedure/Treatment should NOT be performed/administered SINCE IT IS NOT HELP-FUL AND MAY BE HARMFUL

Recommendation that procedure or treatment is not useful/effective and may be harmful

 Sufficient evidence from multiple randomized trials or meta-analyses

Recommendation that procedure or treatment is not useful/effective and may be harmful

■ Limited evidence from single randomized trial or nonrandomized studies

Recommendation that procedure or treatment is not useful/effective and may be harmful

Only expert opinion, case studies, or standard of care

is not recommended is not indicated should not is not useful/effective/beneficial may be harmful * Data available from clinical trials or registries about the usefulness/ efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective

t In 2003 the ACC/AHA Task Force on Practice Guidelines provided a list of suggested phrases to use when writing recommendations. All recommendations in this quideline have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers' comprehension of the guidelines and will allow gueries at the individual recommendation level

Table 2. Choice of Pacemaker Generatorin Selected Indications for Pacing

| Pacemaker Generator | Sinus Node Dysfunction |
|--|--|
| Single-chamber atrial pacemaker | No suspected abnormality of atrioventricular conduction and not at increased risk for future atrioventricular block Maintenance of atrioventricular synchrony during pacing desired |
| Single-chamber ventricular pacemaker | Maintenance of atrioventricular synchrony during pacing not necessary Rate response available if desired |
| Dual-chamber pacemaker | Atrioventricular synchrony during pacing desired Suspected abnormality of atrioventricular conduction or increased risk for future atrioventricular block Rate response available if desired |
| Single-lead, atrial-sensing ventricular pacemaker | Not appropriate |

Atrioventricular Block

Neurally Mediated Syncope or Carotid Sinus Hypersensitivity

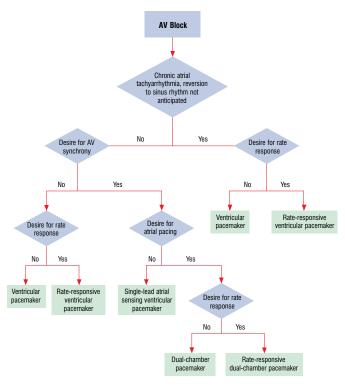
| Not | | |
|-----|--|--|
| | | |
| | | |

Not appropriate

| Chronic atrial fibrillation or other atrial tachyarrhythmia or maintenance of atrioventricular synchrony during pacing not necessary Rate response available if desired | Chronic atrial fibrillation or other atrial tachyarrhythmia Rate response available if desired |
|---|--|
| Rate response available if desired Atrioventricular synchrony during pacing desired Atrial pacing desired Rate response available if desired | Sinus mechanism present Rate response available if desired |
| Desire to limit the number of pacemaker leads | Not appropriate |

Figure 1. Selection of Pacemaker Systems for Patients With Atrioventricular Block

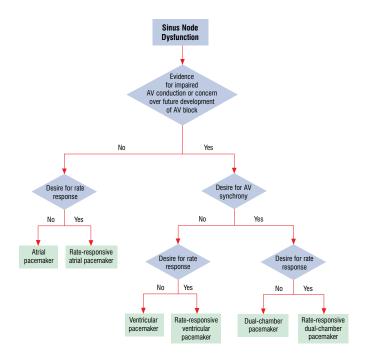
Decisions are illustrated by diamonds. Green shaded boxes indicate type of pacemaker.



AV indicates atrioventricular.

Figure 2. Selection of Pacemaker Systems for Patients With Sinus Node Dysfunction

Decisions are illustrated by diamonds. Green shaded boxes indicate type of pacemaker.



AV indicates atrioventricular.

The following abbreviations are used throughout this pocket guide:

| AV | Atrioventricular |
|------|---|
| CRT | Cardiac resynchronization therapy |
| DDD | Dual-chamber pacemaker that senses/paces in the atrium/ventricle and is inhibited/triggered by intrinsic rhythm |
| ICD | Implantable cardioverter-defibrillator |
| LOE | Level of evidence |
| LV | Left ventricular/left ventricle |
| LVEF | Left ventricular ejection fraction |
| NYHA | New York Heart Association |
| QRS | Letters used to describe the ECG waves associated with ventricular conduction |
| SCD | Sudden cardiac death |
| SND | Sinus node dysfunction |
| νт | Ventricular tachycardia |

II. Recommendations for Permanent Pacing in Sinus Node Dysfunction

Class I Permanent pacemaker implantation is indicated for:
 Sinus node dysfunction (SND) with documented symptomatic bradycardia, including frequent sinus pauses that produce symptoms. (Level of Evidence [LOE]: C)

Symptomatic chronotropic incompetence. (LOE: C)

Symptomatic sinus bradycardia that results from required drug therapy for medical conditions. (LOE: C)

Class IIa Permanent pacemaker implantation is reasonable for:
 SND with heart rate less than 40 bpm when a clear association between significant symptoms consistent with bradycardia and the actual presence of bradycardia has not been documented. (LOE: C)
 Syncope of unexplained origin when clinically significant abnormalities of sinus node function are discovered or provoked in electrophysiological

studies. (LOE: C)

| Class IIb | Permanent pacemaker implantation may be considered in minimally symptomatic patients with chronic heart rate less than 40 bpm while awake. <i>(LOE: C)</i> |
|-----------|---|
| | |

- Class III Permanent pacemaker implantation is not indicated for:
 - SND in asymptomatic patients. (LOE: C)
 - SND in patients for whom the symptoms suggestive of bradycardia have been clearly documented to occur in the absence of bradycardia. (*LOE: C*)
 - SND with symptomatic bradycardia due to nonessential drug therapy. (LOE: C)



III. Recommendations for Acquired Atrioventricular Block in Adults

Class I Permanent pacemaker implantation is indicated for:
 Third-degree and advanced second-degree atrioventricular (AV) block at any anatomic level associated with bradycardia with symptoms (including heart failure) or ventricular arrhythmias presumed to be due to AV block. (LOE: C)

• Third-degree and advanced second-degree AV block at any anatomic level associated with arrhythmias and other medical conditions that require drug therapy that results in symptomatic bradycardia. (LOE: C)

• Third-degree and advanced second-degree AV block at any anatomic level in awake, symptom-free patients in sinus rhythm, with documented periods of asystole greater than or equal to 3.0 seconds or any escape rate less than 40 bpm, or with an escape rhythm that is below the AV node. (*LOE: C*)

Third-degree and advanced second-degree AV block at any anatomic level in awake, symptom-free patients with atrial fibrillation and bradycardia with 1 or more pauses of at least 5 seconds or longer. (LOE: C) Third-degree and advanced second-degree AV block at any anatomic level after catheter ablation of the AV junction. (*LOE: C*)

• Third-degree and advanced second-degree AV block at any anatomic level associated with postoperative AV block that is not expected to resolve after cardiac surgery. (*LOE: C*)

• Third-degree and advanced second-degree AV block at any anatomic level associated with neuromuscular diseases with AV block, such as myotonic muscular dystrophy, Kearns-Sayre syndrome, Erb dystrophy (limb-girdle muscular dystrophy), and peroneal muscular atrophy, with or without symptoms. (*LOE: B*)

Second-degree AV block with associated symptomatic bradycardia regardless of type or site of block. (LOE: B)

Asymptomatic persistent third-degree AV block at any anatomic site with average awake ventricular rates of 40 bpm or faster if cardiomegaly or left ventricular (LV) dysfunction is present or if the site of block is below the AV node. (*LOE: B*)

Second- or third-degree AV block during exercise in the absence of myocardial ischemia. (LOE: C)

Permanent pacemaker implantation is reasonable for: Class IIa Persistent third-degree AV block with an escape rate greater than 40 bpm in asymptomatic adult patients without cardiomegaly. (LOE: C) Asymptomatic second-degree AV block at intraor infra-His levels found at electrophysiological study. (LOE: B) First- or second-degree AV block with symptoms similar to those of pacemaker syndrome or hemodynamic compromise. (LOE: B) Asymptomatic type II second-degree AV block with a narrow QRS. When type II second-degree AV block occurs with a wide QRS, including isolated right bundle-branch block, pacing becomes a Class I recommendation (see Section 2.1.3, "Chronic Bifascicular Block," in the full-text guidelines). (LOE: B)

Class IIb Permanent pacemaker implantation may be considered for:

Neuromuscular diseases such as myotonic muscular dystrophy, Erb dystrophy (limb-girdle muscular dystrophy), and peroneal muscular atrophy with any degree of AV block (including first-degree AV block), with or without symptoms, because there may be unpredictable progression of AV conduction disease. (LOE: B)

• AV block in the setting of drug use and/or drug toxicity when the block is expected to recur even after the drug is withdrawn. (*LOE: B*)

Class III Permanent pacemaker implantation is not indicated for:

Asymptomatic first-degree AV block. (LOE: B) (See Section 1.1.3, "Chronic Bifascicular Block," in the full-text guidelines.)

Asymptomatic type I second-degree AV block at the supra-His (AV node) level or which is not known to be intra- or infra-Hisian. *(LOE: C)*

• AV block that is expected to resolve and is unlikely to recur (e.g., drug toxicity, Lyme disease, or transient increases in vagal tone, or during hypoxia in sleep apnea syndrome in the absence of symptoms). (*LOE: B*)

IV. Recommendations for Permanent Pacing in Chronic Bifascicular Block

Class I Permanent pacemaker implantation is indicated for:
 Advanced second-degree AV block or intermittent third-degree AV block. (LOE: B)

■ Type II second-degree AV block. (LOE: B)

Alternating bundle-branch block. (LOE: C)

Class IIa Permanent pacemaker implantation is reasonable for:
 Syncope not demonstrated to be due to AV block when other likely causes have been excluded, specifically ventricular tachycardia (VT). (LOE: B)

• Incidental finding at electrophysiological study of a markedly prolonged HV interval (greater than or equal to 100 milliseconds) in asymptomatic patients. (LOE: B)

Incidental finding at electrophysiological study of pacing-induced infra-His block that is not physiological. (LOE: B) **Class IIb** Permanent pacemaker implantation may be considered in the setting of neuromuscular diseases such as myotonic muscular dystrophy, Erb dystrophy (limb-girdle muscular dystrophy), and peroneal muscular atrophy with bifascicular block or any fascicular block, with or without symptoms. *(LOE: C)*

Class III Permanent pacemaker implantation is not indicated for:

■ Fascicular block without AV block or symptoms. *(LOE: B)*

■ Fascicular block with first-degree AV block without symptoms. *(LOE: B)*



V. Recommendations for Permanent Pacing After the Acute Phase of Myocardial Infarction*

Class I Permanent ventricular pacing is indicated for:

 Persistent second-degree AV block in the His-Purkinje system with alternating bundle-branch block or third-degree AV block within or below the His-Purkinje system after ST-segment elevation myocardial infarction. (LOE: B)
 Transient advanced second- or third-degree

infranodal AV block and associated bundle-branch block. If the site of block is uncertain, an electrophysiological study may be necessary. (*LOE: B*)

Persistent and symptomatic second- or thirddegree AV block. (LOE: C)

Class IIb Permanent ventricular pacing may be considered for persistent second- or third-degree AV block at the AV node level, even in the absence of symptoms. (*LOE: B*)

> * These recommendations are consistent with the ACC/AHA Guidelines for the Management of Patients With ST-Elevation Myocardial Infarction.

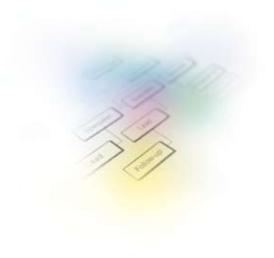
Class III Permanent ventricular pacing is not indicated for:

Transient AV block in the absence of intraventricular conduction defects. (*LOE: B*)

Transient AV block in the presence of isolated left anterior fascicular block. (*LOE: B*)

Acquired bundle-branch block or fascicular block in the absence of AV block. (*LOE: B*)

• Persistent first-degree AV block in the presence of bundle-branch block that is old or of indeterminate age. (*LOE: B*)



VI. Recommendations for Permanent Pacing in Hypersensitive Carotid Sinus Syndrome and Neurocardiogenic Syncope

| Class I | Permanent pacing is indicated for recurrent syncope caused by spontaneously occurring carotid sinus stimulation and carotid sinus pressure that induces ventricular asystole of more than 3 seconds. (<i>LOE: C</i>) |
|-----------|---|
| Class IIa | Permanent pacing is reasonable for syncope without clear, provocative events and with a hypersensitive cardioinhibitory response of 3 seconds or longer. <i>(LOE: C)</i> |
| Class IIb | Permanent pacing may be considered for signifi- cantly symptomatic neurocardiogenic syncope associated with bradycardia documented sponta- neously or at the time of tilt-table testing. (<i>LOE: B</i>) |
| Class III | Permanent pacing is not indicated for: A hypersensitive cardioinhibitory response to carotid sinus stimulation without symptoms or with vague symptoms. (LOE: C) Situational vasovagal syncope in which avoidance behavior is effective and preferred. (LOE: C) |

VII. Recommendations for Pacing After Cardiac Transplantation

| Class I | Permanent pacing is indicated for persistent |
|---------|---|
| | inappropriate or symptomatic bradycardia not |
| | expected to resolve and for other Class I indi- |
| | cations for permanent pacing. (LOE: C) |

Class IIb Permanent pacing may be considered:

• When relative bradycardia is prolonged or recurrent, limiting rehabilitation or discharge after postoperative recovery from cardiac transplantation. (*LOE: C*)

• For syncope after cardiac transplantation even when bradyarrhythmia has not been documented. (*LOE: C*) VIII. Recommendations for Permanent Pacemakers That Automatically Detect and Pace to Terminate Tachycardias

- **Class lla** Permanent pacing is reasonable for symptomatic recurrent supraventricular tachycardia that is reproducibly terminated by pacing when catheter ablation and/or drugs fail to control the arrhythmia or produce intolerable side effects. *(LOE: C)*
- **Class III** Permanent pacing is not indicated in the presence of an accessory pathway that has the capacity for rapid anterograde conduction. (*LOE: C*)



IX. Recommendations for Pacing to Prevent Tachycardia

| Class I | Permanent pacing is indicated for sustained pause-dependent VT, with or without QT pro-longation. <i>(LOE: C)</i> |
|-----------|--|
| Class IIa | Permanent pacing is reasonable for high-risk patients with congenital long-QT syndrome. (LOE: C) |
| Class IIb | Permanent pacing may be considered for prevention of symptomatic, drug-refractory, recurrent atrial fi- brillation in patients with coexisting SND. (<i>LOE: B</i>) |
| Class III | Permanent pacing is not indicated for: Frequent or complex ventricular ectopic activity without sustained VT in the absence of the long-QT syndrome. (LOE: C) Torsade de pointes VT due to reversible causes. (LOE: A) |

X. Recommendation for Pacing to Prevent Atrial Fibrillation

Class III Permanent pacing is not indicated for the prevention of atrial fibrillation in patients without any other indication for pacemaker implantation. (*LOE: B*)

XI. Recommendations for Cardiac Resynchronization Therapy in Patients With Severe Systolic Heart Failure

Class IFor patients who have LV ejection fraction (LVEF)
less than or equal to 35%, a QRS duration greater
than or equal to 0.12 seconds, and sinus rhythm,
cardiac resynchronization therapy (CRT) with or
without an ICD is indicated for the treatment of New
York Heart Association (NYHA) functional Class III
or ambulatory Class IV heart failure symptoms with
optimal recommended medical therapy. (LOE: A)

Class IIa For patients who have:
 LVEF less than or equal to 35%, a QRS duration greater than or equal to 0.12 seconds, and atrial fibrillation, CRT with or without ICD is reasonable for the treatment of NYHA functional Class III or

ambulatory Class IV heart failure symptoms on optimal recommended medical therapy. (LOE: B)

• LVEF less than or equal to 35% with NYHA functional Class III or ambulatory Class IV symptoms who are receiving optimal recommended medical therapy and who have frequent dependence on ventricular pacing, CRT is reasonable. (LOE: C)

- **Class IIb** For patients with LVEF less than or equal to 35% with NYHA functional Class I or II symptoms who are receiving optimal recommended medical therapy and who are undergoing implantation of a permanent pacemaker and/or ICD with anticipated frequent ventricular pacing, CRT may be considered. (LOE: C)
- Class III CRT is not indicated for:

• Asymptomatic patients with reduced LVEF in the absence of other indications for pacing. (*LOE: B*)

Patients whose functional status and life expectancy are limited predominantly by chronic noncardiac conditions. (LOE: C)

XII. Recommendations for Pacing In Patients With Hypertrophic Cardiomyopathy

| Class I | Permanent pacing is indicated for SND or AV block in patients with hypertrophic cardiomyopathy as described previously (see Section 2.1.1, "Sinus Node Dysfunction" and 2.1.2, "Acquired Atrioventricular Block in Adults" in the full-text guidelines). <i>(LOE: C)</i> |
|-----------|---|
| Class IIb | Permanent pacing may be considered in medically refractory symptomatic patients with hypertrophic cardiomyopathy and significant resting or provoked LV outflow tract obstruction. <i>(LOE: A)</i> As for Class I indications, when risk factors for sudden cardiac death (SCD) are present, consider a DDD ICD (see Section 2, "Indications for Implantable Cardioverter- Defibrillator Therapy," in the full-text guidelines). |
| Class III | Permanent pacemaker implantation is not indicated for: Patients who are asymptomatic or whose symptoms are medically controlled. (<i>LOE: C</i>) Symptomatic patients without evidence of LV outflow tract obstruction. (<i>LOE: C</i>) |

XIII. Recommendations for Permanent Pacing in Children, Adolescents, and Patients With Congenital Heart Disease

Class I Permanent pacemaker implantation is indicated for:
 Advanced second- or third-degree AV block associated with symptomatic bradycardia, ventricular dysfunction, or low cardiac output. (LOE: C)

SND with correlation of symptoms during age-inappropriate bradycardia. The definition of bradycardia varies with the patient's age and expected heart rate. (*LOE: B*)

• Postoperative advanced second- or third-degree AV block that is not expected to resolve or that persists at least 7 days after cardiac surgery. (*LOE: B*)

• Congenital third-degree AV block with a wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction. (*LOE: B*)

• Congenital third-degree AV block in the infant with a ventricular rate less than 55 bpm or with congenital heart disease and a ventricular rate less than 70 bpm. (*LOE: C*)

Class IIa Permanent pacemaker implantation is reasonable for: Patients with congenital heart disease and sinus bradycardia for the prevention of recurrent episodes of intra-atrial reentrant tachycardia; SND may be intrinsic or secondary to antiarrhythmic treatment. (LOE: C)

• Patients with congenital third-degree AV block beyond the first year of life with an average heart rate less than 50 bpm, abrupt pauses in ventricular rate that are 2 or 3 times the basic cycle length, or associated with symptoms due to chronotropic incompetence. (LOE: B)

• Patients with sinus bradycardia with complex congenital heart disease with a resting heart rate less than 40 bpm or pauses in ventricular rate longer than 3 seconds. (*LOE: C*)

Patients with congenital heart disease and impaired hemodynamics due to sinus bradycardia or loss of AV synchrony. (LOE: C)

• Unexplained syncope in the patient with prior congenital heart surgery complicated by transient complete heart block with residual fascicular block after a careful evaluation to exclude other causes of syncope. (*LOE: B*)

| Class IIb | Permanent pacemaker implantation may be |
|-----------|---|
| | considered for: |

Transient postoperative third-degree AV block that reverts to sinus rhythm with residual bifascicular block. (*LOE: C*)

Congenital third-degree AV block in asymptomatic children or adolescents with an acceptable rate, a narrow QRS complex, and normal ventricular function. (*LOE: B*)

• Asymptomatic sinus bradycardia after biventricular repair of congenital heart disease with a resting heart rate less than 40 bpm or pauses in ventricular rate longer than 3 seconds. (*LOE: C*)

Class III Permanent pacemaker implantation is not indicated for:

Transient postoperative AV block with return of normal AV conduction in the otherwise asymptomatic patient. (*LOE: B*)

Asymptomatic bifascicular block with or without first-degree AV block after surgery for congenital heart disease in the absence of prior transient complete AV block. (*LOE: C*)

Asymptomatic type I second-degree AV block. (LOE: C)

• Asymptomatic sinus bradycardia with the longest relative risk interval less than 3 seconds and a minimum heart rate more than 40 bpm. *(LOE: C)*

XIV. Recommendations for Implantable Cardioverter-Defibrillators

Secondary prevention refers to the prevention of SCD in those patients who have survived a prior cardiac arrest or sustained VT. Primary prevention refers to the prevention of SCD in individuals without a history of cardiac arrest or sustained VT. Patients with cardiac conditions associated with a high risk of sudden death who have unexplained syncope that is likely to be due to ventricular arrhythmias are considered to have a secondary indication.

Recommendations for consideration of ICD therapy, particularly those for primary prevention, apply only to patients who are receiving optimal medical therapy and have a reasonable expectation of survival with good functional status for more than 1 year. It is difficult to estimate survival with heart failure in the general population, for whom comorbidities and age differ from those in trial populations from which the predictive models have been derived. Patients with repeated heart failure hospitalizations, particularly in the presence of reduced renal function, are at high risk for early death due to heart failure.

We acknowledge that the determination of LVEF lacks a "gold standard" and that there may be variation among the commonly used clinical techniques of LVEF determination. All clinical methods of LVEF determination lack

precision, and the accuracy of techniques varies among laboratories and institutions. On the basis of these considerations, the present writing committee recommends that clinicians use the LVEF determination that they believe is the most clinically accurate and appropriate in their institution.

Class I ICD therapy is indicated in patients:

• Who are survivors of cardiac arrest due to ventricular fibrillation or hemodynamically unstable sustained VT after evaluation to define the cause of the event and to exclude any completely reversible causes. (*LOE: A*)

• With structural heart disease and spontaneous sustained VT, whether hemodynamically stable or unstable. (*LOE: B*)

• With syncope of undetermined origin with clinically relevant, hemodynamically significant sustained VT or ventricular fibrillation induced at electrophysiological study. (*LOE: B*)

• With LVEF less than 35% due to prior myocardial infarction who are at least 40 days post-myocardial infarction and who are in NYHA functional Class II or III. (*LOE: A*)

• With nonischemic dilated cardiomyopathy who have an LVEF less than or equal to 35% and who are in NYHA functional Class II or III. (LOE: B)

• With LV dysfunction due to prior myocardial infarction who are at least 40 days post-myocardial infarction, have an LVEF less than or equal to 30%, and are in NYHA functional Class I. (*LOE: A*)

• With nonsustained VT due to prior myocardial infarction, LVEF less than 40%, and inducible ventricular fibrillation or sustained VT at electrophysiological study. *(LOE: B)*

Class IIa ICD implantation is reasonable:

• For patients with unexplained syncope, significant LV dysfunction, and nonischemic dilated cardiomyopathy. (LOE: C)

■ For patients with sustained VT and normal or nearnormal ventricular function. (*LOE: C*)

■ For patients with hypertrophic cardiomyopathy who have 1 or more major[†] risk factor for SCD. *(LOE: C)*

• For the prevention of SCD in patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy who have 1 or more risk factor for SCD. (*LOE: C*)

• To reduce SCD in patients with long-QT syndrome who are experiencing syncope and/or VT while receiving beta blockers. (*LOE: B*)

For nonhospitalized patients awaiting transplantation. (LOE: C)

[†] See Section 2.2.4, "Hypertrophic Cardiomyopathy," in the full-text guidelines for definition of major risk factors.

| For patients with Brugada syndrome who have |
|---|
| had syncope. (LOE: C) |

• For patients with Brugada syndrome who have documented VT that has not resulted in cardiac arrest. *(LOE: C)*

• For patients with catecholaminergic polymorphic VT who have syncope and/or documented sustained VT while receiving beta blockers. (*LOE: C*)

• For patients with cardiac sarcoidosis, giant cell myocarditis, or Chagas disease. (LOE: C)

Class IIb ICD therapy may be considered in patients with:
 Nonischemic heart disease who have an LVEF of less than or equal to 35% and who are in NYHA functional Class I. (LOE: C)

Long-QT syndrome and risk factors for SCD. (LOE: B)

• Syncope and advanced structural heart disease in whom thorough invasive and noninvasive investigations have failed to define a cause. (*LOE: C*)

A familial cardiomyopathy associated with sudden death. (LOE: C)

LV noncompaction. (LOE: C)

Class III ICD therapy is not indicated for patients:

• Who do not have a reasonable expectation of survival with an acceptable functional status for at least 1 year, even if they meet ICD implantation criteria specified in the Class I, IIa, and IIb recommendations above. *(LOE: C)*

With incessant VT or ventricular fibrillation. (LOE: C)

• With significant psychiatric illnesses that may be aggravated by device implantation or that may preclude systematic follow-up. (*LOE: C*)

• With NYHA Class IV symptoms and drug-refractory congestive heart failure who are not candidates for cardiac transplantation or implantation of a CRT device that incorporates both pacing and defibrillation capabilities. (*LOE: C*)

• With syncope of undetermined cause without inducible ventricular tachyarrhythmias and without structural heart disease. *(LOE: C)*

• When ventricular fibrillation or VT is amenable to surgical or catheter ablation (e.g., atrial arrhythmias associated with Wolff-Parkinson-White syndrome, right ventricular or LV outflow tract VT, idiopathic VT, or fascicular VT in the absence of structural heart disease). (*LOE: C*)

• With ventricular tachyarrhythmias due to a completely reversible disorder in the absence of structural heart disease (e.g., electrolyte imbalance, drugs, or trauma). (*LOE: B*)

XV. Recommendations for Implantable Cardioverter-Defibrillators in Pediatric Patients and Patients With Congenital Heart Disease

Class I ICD implantation is indicated:

• In the survivor of cardiac arrest after evaluation to define the cause of the event and to exclude any reversible causes. (*LOE: B*)

• For patients with symptomatic sustained VT in association with congenital heart disease who have undergone hemodynamic and electrophysiological evaluation. Catheter ablation or surgical repair may offer possible alternatives in carefully selected patients. (LOE: C)

Class IIa ICD implantation is reasonable for patients with congenital heart disease with recurrent syncope of undetermined origin in the presence of either ventricular dysfunction or inducible ventricular arrhythmias at electrophysiological study. (*LOE: B*)

- **Class IIb** ICD implantation may be considered for patients with recurrent syncope associated with complex congenital heart disease and advanced systemic ventricular dysfunction when thorough invasive and noninvasive investigations have failed to define a cause. (*LOE: C*)
- **Class III** All Class III recommendations found in Section 3 of the full-text guidelines, "Indications for Implantable Cardioverter-Defibrillator Therapy," apply to pediatric patients with congenital heart disease, and ICD implantation is not indicated in these patient populations. (*LOE: C*)





