Implantable Cardioverter Defibrillator

Description
The automatic implantable cardioverter defibrillator (ICD) is a device designed to monitor a patient's heart rate, recognize ventricular fibrillation or ventricular tachycardia, and deliver an electric shock to terminate these arrhythmias to reduce the risk of sudden death. A subcutaneous ICD (S-ICD) has been developed that does not employ transvenous leads, with the goal of reducing lead-related complications.

Background
Automatic implantable cardioverter defibrillators (ICD) monitor a patient’s heart rate, recognize ventricular fibrillation (VF) or ventricular tachycardia (VT), and deliver an electric shock to terminate these arrhythmias to reduce the risk of sudden death. Indications for ICD implantation can be broadly subdivided into 1) secondary prevention, i.e., their use in patients who have experienced a potentially life-threatening episode of ventricular tachyarrhythmia (near sudden cardiac death); and 2) primary prevention; i.e., their use in patients who are considered at high risk for sudden cardiac death but who have not yet experienced life-threatening VT or VF.

The standard implantable cardioverter defibrillator (ICD) involves placement of a generator in the subcutaneous tissue of the chest wall. Transvenous leads are attached to the generator and threaded intravenously into the endocardium. The leads sense and transmit information on cardiac rhythm to the generator, which analyzes the rhythm information and produces an electrical shock when a malignant arrhythmia is recognized.

A subcutaneous ICD (S-ICD®) has been developed. This device does not employ transvenous leads and thus avoids the need for venous access and complications associated with the venous leads. Rather, the S-ICD® uses a subcutaneous electrode implanted adjacent to the left sternum. The electrodes sense the cardiac rhythm and deliver countershocks through the subcutaneous tissue of the chest wall.

Several automatic ICDs are approved by the U.S. Food and Drug Administration (FDA) through the premarket approval process. FDA-labeled indications generally include patients who have experienced life-threatening VT associated with cardiac arrest or VT associated with hemodynamic compromise and resistance to pharmacologic treatment. In addition, devices
typically have approval in the secondary prevention setting in patients with a previous myocardial infarction and reduced injection fraction.

Regulatory Status

Transvenous ICDs

The Food and Drug Administration (FDA) has approved a large number of ICDs through the PMA process (FDA product code: LWS). A 2014 review of FDA approvals of cardiac implantable devices reported that between 1979 and 2012, FDA approved 19 ICDs (7 pulse generators, 3 leads, 9 combined systems) through new PMA applications. (1) Many originally-approved ICDs have undergone multiple supplemental applications. A summary of some currently-available ICDs is provided in Table 1 (not an exhaustive list).

Table 1: Implantable Cardioverter Defibrillator With FDA Approval

<table>
<thead>
<tr>
<th>Device</th>
<th>Manufacturer</th>
<th>Original PMA Approval Date</th>
<th>Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ellipse/Fortify Assura Family (originally: Cadence Tiered Therapy Defibrillation System)</td>
<td>St. Jude Medical Inc. (St. Paul, MN)</td>
<td>Jul 1993</td>
<td>Transvenous</td>
</tr>
<tr>
<td>Dynagen, Inogen, Origen, and Teligen Family (originally: Ventak, Vitality, Cofient family)</td>
<td>Boston Scientific Inc. (Marlborough, MA)</td>
<td>Jan 1998</td>
<td>Transvenous</td>
</tr>
<tr>
<td>Evera Family (originally: Virtuosos/Entrust/Maximo/Intrinsic/ Marquis family)</td>
<td>Medtronic Inc. (Minneapolis, MN)</td>
<td>Dec 1998</td>
<td>Transvenous</td>
</tr>
<tr>
<td>Subcutaneous Implantable Defibrillator System</td>
<td>Cameron Health Inc. (San Clemente, CA); acquired by Boston Scientific Inc.</td>
<td>Sep 2012</td>
<td>Subcutaneous</td>
</tr>
</tbody>
</table>

Subcutaneous ICDs

In September 2012, the FDA approved the Subcutaneous Implantable Defibrillator (S-ICD®) System (Cameron Health, Inc, San Clemente, CA; acquired by Boston Scientific, Inc., Marlborough, MA), through the PMA process for the treatment of life-threatening ventricular tachyarrhythmias in patients who do not have symptomatic bradycardia, incessant ventricular
tachycardia, or spontaneous, frequently recurring ventricular tachycardia that is reliably terminated with anti-tachycardia pacing.

In March 2015, the Emblem S-ICD™ (Boston Scientific, Inc., Marlborough, MA), which is smaller and longer-lasting than the original S-ICD, was cleared for marketing through a PMA supplement.

NOTE: ICDs may be combined with other pacing devices, such as pacemakers for atrial fibrillation, or biventricular pacemakers designed to treat heart failure. This evidence review addresses ICDs alone, when used solely to treat patients at risk for ventricular arrhythmias.

Related Policies

2.02.10 Biventricular Pacemakers (Cardiac Resynchronization Therapy) for the Treatment of Heart Failure
2.02.15 Wearable Cardioverter Defibrillator

Policy

*This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Adults

The use of the automatic implantable cardioverter defibrillator (ICD) may be considered medically necessary in adults who meet the following criteria:

Primary Prevention

- ischemic cardiomyopathy with New York Heart Association (NYHA) functional Class II or Class III symptoms, a history of myocardial infarction at least 40 days before ICD treatment, and left ventricular ejection fraction of 35% or less; or
- ischemic cardiomyopathy with NYHA functional Class I symptoms, a history of myocardial infarction at least 40 days before ICD treatment, and left ventricular ejection fraction of 30% or less; or
- nonischemic dilated cardiomyopathy and left ventricular ejection fraction of 35% or less, after reversible causes have been excluded, and the response to optimal medical therapy has been adequately determined; or
- hypertrophic cardiomyopathy (HCM) with 1 or more major risk factors for sudden cardiac death (history of premature HCM-related sudden death in 1 or more first-degree relatives younger than 50 years; left ventricular hypertrophy greater than 30 mm; 1 or more runs of nonsustained ventricular tachycardia at heart rates of 120 beats per minute or greater on 24-hour Holter monitoring; prior unexplained syncope inconsistent with neurocardiogenic origin) and judged to be at high risk for sudden cardiac death by a physician experienced in the care of patients with HCM
- diagnosis of any one of the following cardiac ion channelopathies and considered to
be at high risk for sudden cardiac death (see “Policy Guidelines”):
  o congenital long QT syndrome; OR
  o Brugada syndrome; OR
  o short QT syndrome; OR
  o catecholaminergic polymorphic ventricular tachycardia.

Secondary Prevention

- Patients with a history of a life-threatening clinical event associated with ventricular arrhythmic events such as sustained ventricular tachyarrhythmia after reversible causes (e.g., acute ischemia) have been excluded. See Policy Guidelines.

The use of the ICD is considered investigational in primary prevention patients who:
- have had an acute myocardial infarction (i.e., less than 40 days before ICD treatment);
- have New York Heart Association (NYHA) Class IV congestive heart failure (unless patient is eligible to receive a combination cardiac resynchronization therapy ICD device);
- have had a cardiac revascularization procedure in past 3 months (coronary artery bypass graft [CABG] or percutaneous coronary intervention [PCI] or are candidates for a cardiac revascularization procedure;
- In the absence of documented arrhythmias, patients with newly diagnosed non-ischemic cardiomyopathy (less then 3 months duration); or
- have noncardiac disease that would be associated with life expectancy less than 1 year.

The use of the ICD for secondary prevention is considered investigational for patients who do not meet the criteria for secondary prevention.

Pediatrics

The use of the ICD may be considered medically necessary in children who meet any of the following criteria:

- survivors of cardiac arrest, after reversible causes have been excluded;
- symptomatic, sustained ventricular tachycardia in association with congenital heart disease in patients who have undergone hemodynamic and electrophysiologic evaluation; or
- congenital heart disease with recurrent syncope of undetermined origin in the presence of either ventricular dysfunction or inducible ventricular arrhythmias.
- hypertrophic cardiomyopathy (HCM) with 1 or more major risk factors for sudden cardiac death (history of premature HCM-related sudden death in 1 or more first-degree relatives younger than 50 years; massive left ventricular hypertrophy based on age-specific norms; prior unexplained syncope inconsistent with neurocardiogenic origin) and judged to be at high risk for sudden cardiac death by a physician experienced in the care of patients with HCM.
• diagnosis of any one of the following cardiac ion channelopathies and considered to be at high risk for sudden cardiac death (see "Policy Guidelines"):
  o congenital long QT syndrome; OR
  o Brugada syndrome; OR
  o short QT syndrome; OR
  o catecholaminergic polymorphic ventricular tachycardia.

The use of the ICD is considered **investigational** for all other indications in pediatric patients.

**Subcutaneous ICD**

The use of a subcutaneous ICD may be considered **medically necessary** for adults or children who have an indication for ICD implantation for primary or secondary prevention for any of the above reasons and meet all of the following criteria:

- Have a contraindication to a transvenous ICD due to one or more of the following: (1) lack of adequate vascular access; (2) compelling reason to preserve existing vascular access (ie, need for chronic dialysis; younger patient with anticipated long-term need for ICD therapy); or (3) history of need for explantation of a transvenous ICD due to a complication, with ongoing need for ICD therapy.
- Have no indication for antibradycardia pacing; AND
- Do not have ventricular arrhythmias that are known or anticipated to respond to antitachycardia pacing.

The use of a subcutaneous ICD is considered **investigational** in adult and pediatric patients for individuals who do not meet the criteria outlined above.

**Policy Guidelines**

This policy addressed the use of ICD devices as stand-alone interventions, not as combination devices to treat heart failure (i.e., cardiac resynchronization devices) or in combination with pacemakers. Unless specified, the policy statements and policy rationale are referring to transvenous ICDs.

**Secondary Prevention Criteria for ICD**

The 2014 HRS/ACC/AHA Expert Consensus Statement on the Use of Implantable Cardioverter Defibrillator Therapy in Patients Who Are Not Included or Not Well Represented in Clinical Trials and the 2013 ACCF/HRS/AHA/ASE/HFSA/SCAI/SCCT/SCMR Appropriate Use Criteria for Implantable Cardioverter-Defibrillators and Cardiac Resynchronization Therapy provide guidance on the use of ICD therapy in the management of some common populations of patients who are not represented in clinical trials. Therefore, they are not specifically included in the various guidelines that provide indications for ICD therapy. Guidance from the Expert Consensus Statement and Appropriate Use Criteria indicate ICD implantation may be appropriate for,
Patients who, within 40 days of an MI (but > 48 hours), present with syncope that is thought to be due to ventricular tachyarrhythmia (by clinical history, documented non-sustained ventricular tachycardia (NSVT), or electrophysiologic study) and LVEF <35%.

Patients with cardiac conditions associated with a high risk of sudden death who have unexplained syncope that is likely to be due to self-terminating ventricular arrhythmias, they are considered to have a prevention indication of sudden cardiac death.

Patients who, within 40 days of an MI, develop sustained (or hemodynamically significant) ventricular tachyarrhythmias >48 hours after an MI and in the absence of ongoing ischemia, and LVEF <35%.

Patients within 90 days of revascularization who have previously qualified for the implantation of an ICD for secondary prevention of sudden cardiac death (resuscitated from cardiac arrest due to ventricular tachyarrhythmia) and LV EF <35%.

Patients within 90 days of revascularization with structural heart disease and sustained (or hemodynamically significant) ventricular tachyarrhythmia that was not clearly related to acute myocardial infarction or ischemia.

In patients less than 9 months from the initial diagnosis of NICM with sustained (or hemodynamically significant) ventricular tachyarrhythmia.

**Pediatric Criteria for ICD**

Indications for pediatric ICD use are based on American College of Cardiology/American Heart Association/Heart Rhythm Society (ACC/AHA/HRS) guidelines published in 2008, which acknowledged the lack of primary research in this field on pediatric patients (see Rationale section). These are derived from nonrandomized studies, extrapolation from adult clinical trials, and expert consensus.

**Criteria for ICD Implantation in Patients with Cardiac Ion Channelopathies**

Individuals with cardiac ion channelopathies may have a history of a life-threatening clinical event associated with ventricular arrhythmic events such as sustained ventricular tachyarrhythmia, after reversible causes, in which case they should be considered for ICD implantation for *secondary* prevention, even if they do not meet criteria for primary prevention.

Criteria for ICD implantation in patients with cardiac ion channelopathies are derived from results of clinical input, a 2013 consensus statement from the HRS, European Heart Rhythm Association (EHRA), and the Asia-Pacific Heart Rhythm Society on the diagnosis and management of patients with inherited primary arrhythmia syndromes (Priori et al, 2013), 2013 guidelines from the ACC, AHA, HRS, the American Association of Thoracic Surgeons, and the Society of Thoracic Surgeons on device-based therapy of cardiac rhythm abnormalities (Tracy et al, 2013), and a report from the HRS/EHRA’s Second Consensus Conference on Brugada syndrome (Antzelevitch et al, 2005).
Indications for consideration for ICD implantation for each cardiac ion channelopathy are as follows:

- **Long QT syndrome:**
  - Patients with a diagnosis of LQTS who are survivors of cardiac arrest.
  - Patients with a diagnosis of LQTS who experience recurrent syncopal events while on beta-blocker therapy.

- **Brugada syndrome:**
  - Patients with a diagnosis of BrS who are survivors of cardiac arrest.
  - Patients with a diagnosis of BrS who have documented spontaneous sustained ventricular tachycardia (VT) with or without syncope.
  - Patients with a spontaneous diagnostic type 1 ECG who have a history of syncope, seizure, or nocturnal agonal respiration judged to be likely caused by ventricular arrhythmias (after noncardiac causes have been ruled out).
  - Patients with a diagnosis of BrS who develop ventricular fibrillation (VF) during programmed electrical stimulation.

- **Catecholaminergic polymorphic ventricular tachycardia:**
  - Patients with a diagnosis of CPVT who are survivors of cardiac arrest.
  - Patients with a diagnosis of CPVT who experience recurrent syncope or polymorphic/bidirectional VT despite optimal medical management, and/or left cardiac sympathetic denervation.

- **Short QT syndrome:**
  - Patients with a diagnosis of SQTS who are survivors of cardiac arrest.
  - Patients with a diagnosis of SQTS who are symptomatic and have documented spontaneous VT with or without syncope.
  - Patients with a diagnosis of SQTS or are asymptomatic or symptomatic and have a family history of sudden cardiac death.

**NOTE:** For congenital LQTS, patients may have one or more clinical or historical findings other than those outlined above that may, alone or in combination, put them at higher risk for sudden cardiac death. These may include patients with a family history of sudden cardiac death due to LQTS, infants with a diagnosis of LQTS with functional 2:1 atrioventricular block, patients with a diagnosis of LQTS in conjunction with a diagnosis of Jervell and Lange-Nielsen syndrome or Timothy syndrome, and patients a diagnosis of LQTS with profound QT prolongation (>550 msec). These factors should be evaluated on an individualized basis by a clinician with expertise in LQTS in considering the need for an ICD implantation.

**Rationale**

**Transvenous ICDs for Primary Prevention in Adults**

Transvenous implantable cardioverter defibrillators have been evaluated for primary prevention in a number of populations considered at high risk of sudden cardiac death, including those with ischemic cardiomyopathy, nonischemic dilated cardiomyopathy, and hypertrophic cardiomyopathy. There is a large body of evidence, including a number of randomized clinical trials (RCTs) and systematic reviews of these trials addressing the role of ICDs for primary
prevention, and identifying specific populations who may benefit.

Summary:

ICD for Primary Prevention in Adults

A large body of RCTs has addressed the effectiveness of transvenous ICD implantation for primary prevention in patients at high risk of sudden cardiac death due to ischemic cardiomyopathy and nonischemic dilated cardiomyopathy. Evidence from several RCTs demonstrates improvements in outcomes with ICD treatment for patients with symptomatic heart failure due to ischemic or nonischemic cardiomyopathy with LVEF of 35% or less. The notable exception is that data from several RCTs, including the BEST-ICD and IRIS trials, and subanalyses from earlier RCTs, is that outcomes with ICD therapy do not appear to be improved for patients who are treated with an ICD within 40 days of an acute MI. Less evidence is available for the use of ICDs for primary prevention in patients with hypertrophic cardiomyopathy. In several cohort studies, the annual rate of appropriate ICD discharge ranged from 3.6% to 5.3%. Given the long-term high risk of patients with hypertrophic cardiomyopathy for SCD risk, with the assumption that appropriate shocks are life-saving, these rates are considered adequate evidence for the use of SCDs in patients with hypertrophic cardiomyopathy.

ICDs in Patients with Hereditary Arrhythmia Syndromes

ICDs have been used for both primary and secondary prevention in patients with a number of hereditary disorders that predispose to ventricular arrhythmias and sudden cardiac death, including long QT syndrome (LQTS), Brugada syndrome (BrS), short QT syndrome (SQTS), and catecholaminergic polymorphic ventricular tachycardia (CPVT). Some of these conditions are extremely rare, but the use of ICDs has been described in small cohorts of patients with BrS, LQTS, and CPVT.

Summary:

ICD for Patients with Hereditary Arrhythmia Syndromes

The evidence related to the use of ICDs in patients with hereditary arrhythmia syndromes includes primarily single-center cohort studies or registries of patients with LQTS, Brugada syndrome, and CPVT that report on appropriate shock rates. Patient populations typically include a mix of those requiring ICD implantation for primary prevention and those requiring it for secondary prevention. The limited available data for ICDs for LQTS and CPVT report high rates of appropriate shocks. For Brugada syndrome, more data are available and suggest that rates appropriate shocks are similarly high. Studies comparing outcomes between patients treated and untreated with ICDs are not available. However, given the relatively small patient populations and the high risk of cardiac arrhythmias, clinical trials are unlikely.
Use of Automatic Transvenous ICDs in the Pediatric Population

The available evidence for the use of ICDs in pediatric patients is limited and consists primarily of small case series that include patients with mixed indications for ICD placement. Overall, these studies report relatively high rates of appropriate shocks, but also high rates of inappropriate shocks. Pediatric patients may also be eligible ICD implantation if they have hereditary arrhythmias syndromes (see section, “ICDs in Patients with Hereditary Arrhythmia Syndromes.”)

Subcutaneous ICD

The subcutaneous ICD (S-ICD) is intended for patients who do have standard indications for an ICD, but who do not require pacing for bradycardia, or anti-tachycardia overdrive pacing for VT. The S-ICD has been proposed as of particular benefit for patients with limited vascular access, including patients undergoing renal dialysis or children, or those who have had complications with transvenous ICDs requiring device explantation. There were no RCTs identified that compared the performance of a subcutaneous ICD (S-ICD) with transvenous ICDs. Two nonrandomized, comparative studies were identified that compared the efficacy of the two different types of ICDs, and numerous single-arm studies report on outcomes of the S-ICD.

Nonrandomized studies have suggested that S-ICDs are as effective as TV-ICDs at terminating laboratory-induced ventricular arrhythmias. Data from 2 large patient registries have suggested that S-ICDs are effective at terminating ventricular arrhythmias when they occur. However, no RCTs have directly compared TV- and S-ICDs were identified.

Practice Guidelines and Policy Statements

American College of Cardiology/American Heart Association Heart Failure Management Guidelines

In 2013, the American College of Cardiology (ACC) and American Heart Association (AHA) issued practice guidelines on the management of heart failure, which made the following recommendations about the use of ICD devices as primary prevention

- For patients with stage B heart failure, an ICD is reasonable in patients with asymptomatic ischemic cardiomyopathy who are at least 40 d post-MI, have an LVEF ≤30%, and on guideline-directed medical therapy (GDMT). (class of recommendation: IIa; level of evidence: B).
- For patients with stage C heart failure:
  - ICD therapy is recommended for primary prevention of sudden cardiac death (SCD) in selected patients with heart failure with reduced ejection fraction (HFrEF) at least 40 d post-myocardial infarction (MI) with left ventricular ejection fraction (LVEF) ≤35% and NYHA class II or III symptoms on chronic GDMT, who are expected to live at least 1 year. (Class of recommendation: I; level of evidence: A).
Device-Based Therapy for Cardiac Rhythm Abnormalities Guidelines

In 2012, ACC and AHA, together with the Heart Rhythm Society (HRS), the American Association of Thoracic Surgeons, and the Society of Thoracic Surgeons, issued a focused update to 2008 guidelines for device-based therapy of cardiac rhythm abnormalities. The guidelines make the following recommendations related to ICD therapy in adults, all of which are based on the expectation that patients are receiving optimal medical therapy and have a reasonable expectation of survival with a good functional status for more than a year:

- **Class I recommendations:**
  - ICD therapy is indicated in patients who are survivors of cardiac arrest due to ventricular fibrillation (VF) or hemodynamically unstable sustained ventricular tachycardia (VT) after evaluation to define the cause of the event and to exclude any completely reversible causes. \((\text{Level of Evidence: } A)\)
  - ICD therapy is indicated in patients with structural heart disease and spontaneous sustained VT, whether hemodynamically stable or unstable. \((\text{Level of Evidence: } B)\)
  - ICD therapy is indicated in patients with syncope of undetermined origin with clinically relevant, hemodynamically significant sustained VT or VF induced at electrophysiological study. \((\text{Level of Evidence: } B)\)
  - ICD therapy is indicated in patients with LVEF less than or equal to 35% due to prior MI who are at least 40 days post-MI and are in NYHA functional Class II or III. \((\text{Level of Evidence: } A)\)
  - ICD therapy is indicated in patients with nonischemic dilated cardiomyopathy (DCM) who have an LVEF less than or equal to 35% and who are in NYHA functional Class II or III. \((\text{Level of Evidence: } B)\)
  - ICD therapy is indicated in patients with LV dysfunction due to prior MI who are at least 40 days post-MI, have an LVEF less than or equal to 30%, and are in NYHA functional Class I. \((\text{Level of Evidence: } A)\)
  - ICD therapy is indicated in patients with nonsustained VT due to prior MI, LVEF less than or equal to 40%, and inducible VF or sustained VT at electrophysiological study. \((\text{Level of Evidence: } B)\)

- **Class IIa recommendations:**
  - ICD implantation is reasonable for patients with unexplained syncope, significant LV dysfunction, and nonischemic DCM. \((\text{Level of Evidence: } C)\)
  - ICD implantation is reasonable for patients with sustained VT and normal or near-normal ventricular function. \((\text{Level of Evidence: } C)\)
ICD implantation is reasonable for patients with HCM who have 1 or more major† risk factors for sudden cardiac death (SCD). (*Level of Evidence: C*)

ICD implantation is reasonable for the prevention of SCD in patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy who have 1 or more risk factors for SCD. (*Level of Evidence: C*)

ICD implantation is reasonable to reduce SCD in patients with long-QT syndrome who are experiencing syncope and/or VT while receiving beta blockers. (*Level of Evidence: B*)

ICD implantation is reasonable for nonhospitalized patients awaiting transplantation. (*Level of Evidence: C*)

ICD implantation is reasonable for patients with Brugada syndrome who have had syncope. (*Level of Evidence: C*)

ICD implantation is reasonable for patients with Brugada syndrome who have documented VT that has not resulted in cardiac arrest. (*Level of Evidence: C*)

ICD implantation is reasonable for patients with catecholaminergic polymorphic VT who have syncope and/or documented sustained VT while receiving beta blockers. (*Level of Evidence: C*)

ICD implantation is reasonable for patients with cardiac sarcoidosis, giant cell myocarditis, or Chagas disease. (*Level of Evidence: C*)

- **Class IIb recommendations:**
  
  - 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. (*Level of Evidence: C*)
  
  - ICD therapy may be considered for patients with long-QT syndrome and risk factors for SCD. (*Level of Evidence: B*)
  
  - ICD therapy may be considered in patients with syncope and advanced structural heart disease in whom thorough invasive and noninvasive investigations have failed to define a cause. (*Level of Evidence: C*)
  
  - ICD therapy may be considered in patients with a familial cardiomyopathy associated with sudden death. (*Level of Evidence: C*)
  
  - ICD therapy may be considered in patients with LV noncompaction. (*Level of Evidence: C*)

- **Class III recommendations (not recommended):**
  
  - 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. (*Level of Evidence: C*)
  
  - ICD therapy is not indicated for patients with incessant VT or VF. (*Level of Evidence: C*)
  
  - ICD therapy is not indicated in patients with significant psychiatric illnesses that may be aggravated by device implantation or that may preclude systematic follow-up. (*Level of Evidence: C*)
  
  - ICD therapy is not indicated for NYHA Class IV patients with drug-refractory congestive heart failure who are not candidates for cardiac transplantation or cardiac resynchronization/defibrillator. (*Level of Evidence: C*)
ICD therapy is not indicated for syncope of undetermined cause in a patient without inducible ventricular tachyarrhythmias and without structural heart disease. *(Level of Evidence: C)*

ICD therapy is not indicated when VF or VT is amenable to surgical or catheter ablation (e.g., atrial arrhythmias associated with the Wolff-Parkinson-White syndrome, RV or LV outflow tract VT, idiopathic VT, or fascicular VT in the absence of structural heart disease). *(Level of Evidence: C)*

ICD therapy is not indicated for patients with ventricular tachyarrhythmias due to a completely reversible disorder in the absence of structural heart disease (e.g., electrolyte imbalance, drugs, or trauma). *(Level of Evidence: B)*

The 2012 guidelines make the following recommendations related to ICD therapy in children:

- **Class I recommendations:**
  - 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. *(Level of Evidence: B)*
  - ICD implantation is indicated 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. *(Level of Evidence: C)*

- **Class IIa recommendations:** 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. *(Level of Evidence: B)*

- **Class IIb recommendations:** 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. *(Level of Evidence: C)*

- **Class III recommendations:** All Class III recommendations found in Section 3, “Indications for Implantable Cardioverter-Defibrillator Therapy,” apply to pediatric patients and patients with congenital heart disease, and ICD implantation is not indicated in these patient populations. *(Level of Evidence: C)*

**Expert Consensus Statement on ICD Therapy in Patients Not Well Represented in Clinical Trials**

In 2014, HRS, ACC, and AHA published an expert consensus statement on the use of ICD therapy in patients who were not included or poorly represented in ICD clinical trials, which made a number of consensus-based guidelines on the use of ICDs in selected patient populations.
Guidelines for Hypertrophic Cardiomyopathy

In 2011, ACCF/AHA guidelines were published on the management of patients with hypertrophic cardiomyopathy. These guidelines contained the following statements about the use of ICD in patients with HCM:

**Class I Recommendations**
- The decision to place an ICD in patients with HCM should include application of individual clinical judgment, as well as a thorough discussion of the strength of evidence, benefits, and risks to allow the informed patient’s active participation in decision making. *(Level of Evidence: C)*
- ICD placement is recommended for patients with HCM with prior documented cardiac arrest, ventricular fibrillation, or hemodynamically significant VT. *(Level of Evidence: B)*

**Class IIa Recommendations**
- It is reasonable to recommend an ICD for patients with HCM with:
  - Sudden death presumably caused by HCM in 1 or more first-degree relatives. *(Level of Evidence: C)*
  - A maximum LV wall thickness greater than or equal to 30 mm. *(Level of Evidence: C)*
  - One or more recent, unexplained syncopal episodes. *(Level of Evidence: C)*
- An ICD can be useful in select patients with NSVT [nonsustained VT] (particularly those <30 years of age) in the presence of other SCD risk factors or modifiers. *(Level of Evidence: C)*
- An ICD can be useful in select patients with HCM with an abnormal blood pressure response with exercise in the presence of other SCD risk factors or modifiers. *(Level of Evidence: C)*

It is reasonable to recommend an ICD for high-risk children with HCM, based on unexplained syncope, massive LV hypertrophy, or family history of SCD, after taking into account the relatively high complication rate of long-term ICD implantation. *(Level of Evidence: C)*.

**Class IIb Recommendations**
- The usefulness of an ICD is uncertain in patients with HCM with isolated bursts of NSVT when in the absence of any other SCD risk factors or modifiers. *(Level of Evidence: C)*
- The usefulness of an ICD is uncertain in patients with HCM with an abnormal blood pressure response with exercise when in the absence of any other SCD risk factors or modifiers, particularly in the presence of significant outflow obstruction. *(Level of Evidence: C)*

**Class III Recommendations: Harm**
- ICD placement as a routine strategy in patients with HCM without an indication of increased risk is potentially harmful. *(Level of Evidence: C)*
- ICD placement as a strategy to permit patients with HCM to participate in competitive athletics is potentially harmful. *(Level of Evidence: C)*
ICD placement in patients who have an identified HCM genotype in the absence of clinical manifestations of HCM is potentially harmful. *(Level of Evidence: C)*

**American Heart Association/Heart Rhythm Society**

In 2010, the AHA issued a scientific statement, endorsed by the HRS, on cardiovascular implantable electronic device (CIED) infections and their management. This statement makes the following class I recommendations about removal of infected CIEDs:

- Complete device and lead removal is recommended for all patients with definite CIED infection, as evidenced by valvular and/or lead endocarditis or sepsis. *(Level of Evidence: A)*
- Complete device and lead removal is recommended for all patients with CIED pocket infection as evidenced by abscess formation, device erosion, skin adherence, or chronic draining sinus without clinically evident involvement of the transvenous portion of the lead system. *(Level of Evidence: B)*
- Complete device and lead removal is recommended for all patients with valvular endocarditis without definite involvement of the lead(s) and/or device. *(Level of Evidence: B)*
- Complete device and lead removal is recommended for patients with occult staphylococcal bacteremia. *(Level of Evidence: B)*

**European Society of Cardiology et al**

In 2015, the European Society of Cardiology (ESC) and the Association for European Paediatric and Congenital Cardiology (AEPC) issued guidelines on the management of patients with ventricular arrhythmias and the prevention of SCD. These guidelines make the following statements on use of device-based therapy for ventricular arrhythmia and prevention of SCD:

**Class I Recommendations**

- “ICD implantation is recommended in patients with documented VF or haemodynamically not tolerated VT in the absence of reversible causes or within 48 h after myocardial infarction who are receiving chronic optimal medical therapy and have a reasonable expectation of survival with a good functional status >1 year.” *(Level of Evidence: A)*

**Class IIa Recommendations**

- “ICD implantation should be considered in patients with recurrent sustained VT (not within 48 h after myocardial infarction) who are receiving chronic optimal medical therapy, have a normal LVEF and have a reasonable expectation of survival with good functional status for > 1 year.” *(Level of Evidence: C)*
- “Subcutaneous defibrillators should be considered as an alternative to transvenous defibrillators in patients with an indication for an ICD when pacing therapy for bradycardia support, cardiac resynchronization or antitachycardia pacing is not needed.” *(Level of Evidence: C)*
Class IIb Recommendations

- “The subcutaneous ICD may be considered as a useful alternative to the transvenous ICD system when venous access is difficult, after the removal of a transvenous ICD for infections or in young patients with a long-term need for ICD therapy.” (Level of Evidence: C)

Heart Rhythm Society/European Heart Rhythm Association/Asia-Pacific Heart Rhythm Society

In 2013, the Heart Rhythm Society (HRS), European Heart Rhythm Association (EHRA), and the Asia-Pacific Heart Rhythm Society (APHRS) issued a consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes, which included a number of recommendations related to ICD use in patients with long QT syndrome (LQTS), Brugada syndrome (BrS), catecholaminergic polymorphic ventricular tachycardia (CPVT), and short QT syndrome (SQTS).

Long QT Syndrome

Class I Recommendations

- ICD implantation is recommended for patients with a diagnosis of LQTS who are survivors of a cardiac arrest.

Class IIa Recommendations

- ICD implantation can be useful in patients with a diagnosis of LQTS who experience recurrent syncopal events while on beta-blocker therapy.

Class III Recommendations: Harm

- Except under special circumstances, ICD implantation is not indicated in asymptomatic LQTS patients who have not been tried on beta-blocker therapy.

Brugada Syndrome

Class I Recommendations:

- ICD implantation is recommended in patients with a diagnosis of BrS who:
  - Are survivors of a cardiac arrest and/or
  - Have documented spontaneous sustained VT with or without syncope.

Class IIa Recommendations:

- ICD implantation can be useful in patients with a spontaneous diagnostic type I ECG who have a history of syncope judged to be likely caused by ventricular arrhythmias.

Class IIb Recommendations:

- ICD implantation may be considered in patients with a diagnosis of BrS who develop VF during programmed electrical stimulation (inducible patients).

Class III Recommendations: Harm

- ICD implantation is not indicated in asymptomatic BrS patients with a drug-induced type I ECG and on the basis of a family history of SCD alone.
Catecholaminergic Polymorphic Ventricular Tachycardia

Class I Recommendations:
- ICD implantation is recommended for patients with a diagnosis of CPVT who experience cardiac arrest, recurrent syncope or polymorphic/bidirectional VT despite optimal medical management, and/or left cardiac sympathetic denervation.

Class III Recommendations: Harm
- ICD as a standalone therapy is not indicated in an asymptomatic patient with a diagnosis of CPVT

Short QT Syndrome

Class I Recommendations:
- ICD implantation is recommended in symptomatic patients with a diagnosis of SQTS who:
  - Are survivors of cardiac arrest and/or
  - Have documented spontaneous VT with or without syncope.

Class IIb Recommendations:
- ICD implantation may be considered in asymptomatic patients with a diagnosis of SQTS and a family history of sudden cardiac death.

Heart Rhythm Society and European Heart Rhythm Association

In a 2005 consensus report from the second consensus conference on BrS, HRS and EHRA addressed diagnostic criteria, risk-stratification schemes, and device- and pharmacologic-based therapy for BrS. This report makes the following recommendations for ICD implantation in BrS:

Symptomatic patients displaying the type 1 Brugada ECG (either spontaneously or after sodium channel blockade) who present with aborted sudden death should receive an ICD without additional need for electrophysiologic studies.

- Symptomatic patients displaying the type 1 Brugada ECG (either spontaneously or after sodium channel blockade) who present with syncope, seizure, or nocturnal agonal respiration should undergo ICD implantation after noncardiac causes of these symptoms have been ruled out.
- Asymptomatic patients displaying a type 1 Brugada ECG (either spontaneously or after sodium channel blockade) should undergo EPS [electrophysiology studies] if a family history of sudden cardiac death is suspected to be the result of Brugada syndrome. EPS is justified when the family history is negative for sudden cardiac death if the type 1 ECG occurs spontaneously. If inducible for ventricular arrhythmia, then the patient should receive an ICD.
- Asymptomatic patients who have no family history and who develop a type 1 ECG only after sodium channel blockade should be closely followed up.
Pediatric and Congenital Electrophysiology Society (PACES)/HRS

In 2014, PACES and HRS issued an expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease (CHD) which made the following recommendations on the use of ICD therapy in adults with CHD:

**Class I Recommendations:**
- ICD therapy is indicated in adults with CHD who are survivors of cardiac arrest due to ventricular fibrillation or hemodynamically unstable ventricular tachycardia after evaluation to define the cause of the event and exclude any completely reversible etiology (Level of evidence: B).
- ICD therapy is indicated in adults with CHD and spontaneous sustained ventricular tachycardia who have undergone hemodynamic and electrophysiologic evaluation (Level of evidence: B).
- ICD therapy is indicated in adults with CHD and a systemic left ventricular ejection fraction <35%, biventricular physiology, and New York Heart Association (NYHA) class II or III symptoms (Level of evidence: B).

**Class IIa Recommendations:**
- ICD therapy is reasonable in selected adults with tetralogy of Fallot and multiple risk factors for sudden cardiac death, such as left ventricular systolic or diastolic dysfunction, nonsustained ventricular tachycardia, QRS duration >180 ms, extensive right ventricular scarring, or inducible sustained ventricular tachycardia at electrophysiologic study (Level of evidence: B).

**Class IIb Recommendations:**
- ICD therapy may be reasonable in adults with a single or systemic right ventricular ejection fraction <35%, particularly in the presence of additional risk factors such as complex ventricular arrhythmias, unexplained syncope, NYHA functional class II or III symptoms, QRS duration >140 ms, or severe systemic AV valve regurgitation (Level of evidence: C).
- ICD therapy may be considered in adults with CHD and a systemic ventricular ejection fraction <35% in the absence of overt symptoms (NYHA class I) or other known risk factors (Level of evidence: C).
- ICD therapy may be considered in adults with CHD and syncope of unknown origin with hemodynamically significant sustained ventricular tachycardia or fibrillation inducible at electrophysiologic study (Level of evidence: B).
- ICD therapy may be considered for nonhospitalized adults with CHD awaiting heart transplantation (Level of evidence: C).
- ICD therapy may be considered for adults with syncope and moderate or complex CHD in whom there is a high clinical suspicion of ventricular arrhythmia and in whom thorough invasive and noninvasive investigations have failed to define a cause (Level of evidence: C).
Class III Recommendations:

- All Class III recommendations listed in current ACC/AHA/HRS guidelines apply to adults with CHD (Level of evidence: C).
- Adults with CHD and advanced pulmonary vascular disease (Eisenmenger syndrome) are generally not considered candidates for ICD therapy (Level of evidence: B).
- Endocardial leads are generally avoided in adults with CHD and intracardiac shunts. Risk assessment regarding hemodynamic circumstances, concomitant anticoagulation, shunt closure prior to endocardial lead placement, or alternative approaches for lead access should be individualized (Level of Evidence: B).

U.S. Preventive Services Task Force Recommendations

Use of implantable cardioverter defibrillators is not a preventive service.

Medicare Policy

In January 2005, Medicare issued the following revised national coverage guideline for the use of ICDs. The Centers for Medicare and Medicaid Services (CMS) determined that the evidence is adequate to conclude that an ICD is reasonable and necessary for the following:

- Patients with ischemic dilated cardiomyopathy (IDCM), documented prior MI, NYHA Class II and III heart failure, and measured LVEF of 35% or less;
- Patients with nonischemic dilated cardiomyopathy (NIDCM) >9 months, NYHA Class II and III heart failure, and measured LVEF of 35% or less;
- Patients who meet all current CMS coverage requirements for a cardiac resynchronization therapy (CRT) device and have NYHA Class IV heart failure;

For each of these groups, patients must not have:

- Cardiogenic shock or symptomatic hypotension while in a stable baseline rhythm;
- Had a coronary artery bypass graft (CABG) or PTCA within the past 3 months;
- Had an acute MI within the past 40 days;
- Clinical symptoms or findings that would make them a candidate for coronary revascularization;
- Irreversible brain damage from pre-existing cerebral disease;
- Any disease, other than cardiac disease (e.g., cancer, uremia, liver failure), associated with a likelihood of survival less than 1 year;

In addition, CMS specifies that the beneficiary receiving the ICD implantation for primary prevention must be enrolled in either a U.S. Food and Drug Administration (FDA)-approved category B Investigational Device Exemption clinical trial (42 CFR §405.201), a trial under the CMS Clinical Trial Policy (NCD Manual §310.1), or a qualifying data collection system including approved clinical trials and registries.
The Medicare policy for ischemic and nonischemic dilated cardiomyopathy is consistent with this policy.

References


83. Priori SG, Blomstrom-Lundqvist C, Mazzanti A, et al. 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac


86. Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). Can J Cardiol. Oct 2014;30(10):e1-e63. PMID 25262867

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Signature on File
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