Implantable Cardioverter Defibrillator (ICD)

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Next Review: 11/2017

Policy
Blue Cross and Blue Shield of Kansas City (Blue KC) will provide coverage for automatic implantable cardioverter defibrillator when it is determined to be medically necessary because the criteria shown below are met.

When Policy Topic is covered
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 "Considerations"):
  o congenital long QT syndrome; OR
Brugada syndrome; OR  
short QT syndrome; OR  
catecholaminergic polymorphic ventricular tachycardia.

Note: Symptomatic heart failure is defined as the presence of dyspnea on exertion, angina, palpitations, or fatigue.

Secondary Prevention

- Patients with a history of a life-threatening clinical event associated with sustained ventricular tachyarrhythmia, after reversible causes (eg, acute ischemia) have been excluded.

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”):
  - congenital long QT syndrome; OR
  - Brugada syndrome; OR
  - short QT syndrome; OR
  - catecholaminergic polymorphic ventricular tachycardia.

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.

**When Policy Topic is not covered**
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 cardiac revascularization procedure in past 3 months (coronary artery bypass graft [CABG] or percutaneous transluminal coronary angioplasty [PTCA]) or are candidates for a cardiac revascularization procedure
- have noncardiac disease that would be associated with life expectancy less than 1 year

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

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

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

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). 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 syncpe 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.
Effective in 2015, the CPT coding for these devices was updated to include insertion of subcutaneous ICD devices (see Code Table)

### Description of Procedure or Service

<table>
<thead>
<tr>
<th>Populations</th>
<th>Interventions</th>
<th>Comparators</th>
<th>Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individuals: <strong>With a high risk of sudden cardiac death due to ischemic cardiomyopathy in adulthood</strong></td>
<td>Interventions of interest are: • Transvenous implantable cardioverter defibrillator placement</td>
<td>Comparators of interest are: • Medical management without implantable cardioverter defibrillator placement</td>
<td>Relevant outcomes include: • Overall survival • Morbid events • Quality of life • Treatment-related morbidity • Treatment-related mortality</td>
</tr>
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<td>Individuals: <strong>With a high risk of sudden cardiac death due to nonischemic dilated cardiomyopathy in adulthood</strong></td>
<td>Interventions of interest are: • Transvenous implantable cardioverter defibrillator placement</td>
<td>Comparators of interest are: • Medical management without implantable cardioverter defibrillator placement</td>
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<td>Interventions of interest are: • Transvenous implantable cardioverter defibrillator placement</td>
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</tr>
<tr>
<td>Individuals: <strong>With high risk of sudden cardiac death due to an inherited cardiac ion channelopathy</strong></td>
<td>Interventions of interest are: • Transvenous implantable cardioverter defibrillator placement</td>
<td>Comparators of interest are: • Medical management without implantable cardioverter defibrillator placement</td>
<td>Relevant outcomes include: • Overall survival • Morbid events • Quality of life • Treatment-related morbidity • Treatment-related mortality</td>
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<tr>
<td>Individuals: <strong>With need for cardioverter defibrillator (no antitachycardia pacer–responsive arrhythmia or need for anti Bradycardia pacer)</strong></td>
<td>Interventions of interest are: • Subcutaneous implantable cardioverter defibrillator placement</td>
<td>Comparators of interest are: • Transvenous implantable cardioverter defibrillator placement</td>
<td>Relevant outcomes include: • Overall survival • Morbid events • Quality of life • Treatment-related morbidity • Treatment-related mortality</td>
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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.

For individuals who have a high risk of sudden cardiac death (SCD) due to ischemic or to nonischemic cardiomyopathy (NICM) in adulthood who receive transvenous ICD (TV-ICD) placement, the evidence includes multiple well-designed, well-conducted randomized controlled trials (RCTs) and systematic reviews of these trials. Relevant outcomes are overall survival, morbid events, quality of life, and treatment-related morbidity and mortality. There is an extensive literature on the use of ICDs in patients with prior arrhythmogenic events and ischemic cardiomyopathy. Earlier trials first demonstrated a benefit in overall mortality for survivors of cardiac arrest and patients with potentially lethal cardiac arrhythmias. Multiple, well-done, RCTs have also shown a benefit in overall mortality for patients with ischemic cardiomyopathy and reduced ejection fraction. RCTs of early ICD implantation following acute myocardial infarction (MI) did not support a benefit for immediate versus delayed implantation for at least 40 days. For NICM, there is less clinical trial data, but the available evidence from a limited number of RCTs enrolling patients with NICM and from subgroup analysis of RCTs with mixed populations supports a survival benefit for this group. There is no high-quality evidence to determine whether early versus delayed implantation improves outcomes for patients with NICM and it is not possible to determine the optimal waiting period for ICD implantation following onset of NICM. At least 1 cohort study has reported that most patients who meet criteria for an ICD at the time of initial NICM diagnosis will no longer meet the criteria several months after initiation of treatment. The evidence is sufficient to determine qualitatively that the technology results in a large improvement in the net health outcome.

For individuals who have a high risk of SCD due to hypertrophic cardiomyopathy (HCM) in adulthood who receive TV-ICD placement, the evidence includes several large registry studies. Relevant outcomes are overall survival, morbid events, quality of life, and treatment-related morbidity and mortality. In these studies, the annual rate of appropriate ICD discharge ranged from 3.6% to 5.3%. Given the long-term high risk of patients with HCM for SCD risk, with the assumption that appropriate shocks are life-saving, these rates are considered adequate evidence for the use of ICDs in patients with HCM. The evidence is sufficient to determine qualitatively that the technology results in a meaningful improvement in the net health outcome.

For individuals who have a high risk of SCD due to an inherited cardiac ion channelopathy who receive TV-ICD placement, the evidence includes small cohort studies of patients with these conditions treated with ICDs. Relevant outcomes are overall survival, morbid events, quality of life, and treatment-related morbidity and mortality. The limited evidence for patients with long QT syndrome (LQTS), catecholaminergic polymorphic ventricular tachycardia (CPVT), and Brugada syndrome (BrS) has reported high rates of appropriate shocks. No studies were identified on the use of ICDs for patients with short QT syndrome (SQTS). 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. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have need for a cardioverter defibrillator but no indications for antibradycardia pacing and no antitachycardia pacing–responsive arrhythmias who receive S-ICD placement, the evidence includes nonrandomized studies and case series. Relevant outcomes are overall survival, morbid events, quality of life, and treatment-related morbidity and mortality. Nonrandomized controlled studies have reported success rates in terminating laboratory-induced ventricular fibrillation that are similar to TV-ICD. However, there is scant evidence on comparative clinical outcomes of both types of ICD over longer periods. Case series have reported high rates of detection and successful conversion of ventricular tachycardia, and inappropriate shock rates in the range reported for TV-ICD. This evidence is not sufficient to determine whether there are small differences in efficacy between the 2 types of devices, which may be clinically important due to the nature to the disorder being treated. Also, adverse event rate is uncertain, with variable rates reported. At least 1 RCT is currently underway comparing S-ICD with TV-ICD. The evidence is insufficient to determine the effects of the technology on health outcomes.

Clinical input was obtained on the use of ICDs in pediatric populations and for primary prevention in patients with cardiac ion channelopathies, and for on the use of the S-ICD. For the use of ICDs in children with HCM or with a history of congenital heart disease, the evidence includes case series. These conditions have a low prevalence and heterogeneous patient populations, creating barriers to trials.

There was consensus that the use of ICDs in certain pediatric populations, consistent with specialty society guidelines, is medically necessary. Indications for the use of ICDs to prevent SCD in HCM in pediatric patients parallel those in adults. There was also consensus that the use of an ICD should be considered medically necessary for primary prevention of ventricular arrhythmias in adults and children with a diagnosis of QTS, BrS, SQTS, or CPVT. Criteria for determining patients at high risk of SCD for the cardiac ion channelopathies was derived from clinical input and specialty society guidelines. There was consensus that the use of an S-ICD should be considered medically necessary, particularly for patients with indications for an ICD but who have difficult vascular access (eg, children or patients undergoing chronic dialysis) or have had TV-ICD lead explantation due to 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 implantable cardioverter defibrillator (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 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 ejection fraction.

**Regulatory Status**

**Transvenous ICDs**

The Food and Drug Administration (FDA) has approved a large number of ICDs through the premarket approval (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. 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>
<thead>
<tr>
<th>Device</th>
<th>Manufacturer</th>
<th>Original PMA Approval Date</th>
<th>Type</th>
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<tr>
<td>Ellipse/ Fortify Assura Family (originally: Cadence Tiered Therapy Defibrillation System)</td>
<td>St. Jude Medical, Inc. (St. Paul, MN)</td>
<td>July 1993</td>
<td>Transvenous</td>
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Subcutaneous ICDs
In September 2012, the FDA approved the Subcutaneous Implantable Defibrillator System (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.

Rationale
This evidence review was created in March 1996 and has been updated periodically with literature searches of the MEDLINE database. The most recent update with literature review covers the period through April 7, 2016.

Transvenous Implantable Cardioverter Defibrillators for Primary Prevention in Adults
Transvenous implantable cardioverter defibrillators (TV-ICDs) have been evaluated for primary prevention in a number of populations considered at high risk of sudden cardiac death (SCD), including those with ischemic cardiomyopathy, nonischemic dilated cardiomyopathy (NIDCM), and hypertrophic cardiomyopathy (HCM). 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.

Overview and Summary of TEC Assessments
Automatic ICDs were first used in survivors of near SCD. There has been ongoing interest in using ICDs as primary preventive therapy in patients with risk factors for SCD. Several BCBSA TEC Assessments have addressed the use of ICDs for primary prevention of SCD. The first TEC Assessment (2002) focused on the Multicenter Automatic Defibrillator Implantation Trials (known as MADIT I and
MADIT II) that compared the use of an ICD with conventional therapy among patients with coronary artery disease with a history of myocardial infarction (MI) and a reduced ejection fraction. The key difference in the 2 trials was the patient selection criteria. In the MADIT I trial, patients were required to have a left ventricular ejection fraction (LVEF) of less than 35% and ventricular tachyarrhythmia, as evidenced on an electrophysiologic study. In MADIT II, patients were required to have a lower ejection fraction (<30%), but no electrophysiologic study was required. Therefore, the patient selection criteria for MADIT II potentially identified a much larger pool of candidates for ICD implantation.

The 2002 TEC Assessment concluded: “For patients who have coronary artery disease with prior MI and reduced LVEF and who are similar to those selected in MADIT I and MADIT II, the available evidence demonstrates an improvement in overall mortality associated with ICD treatment compared with conventional therapy.”

In 2004, TEC reassessed ICDs for primary prevention of SCD. The 2004 TEC Assessment focused on the results of the 5 randomized controlled trials (RCTs) included in the 2002 Assessment (including the Multicenter Unsustained Tachycardia Trial [MUSTT], MADIT I, MADIT II, Coronary Artery Bypass Graft [CABG] Patch Trial, the Cardiomyopathy Trial [CAT]) and 5 additional RCTs: Defibrillator in Acute Myocardial Infarction Trial (DINAMIT); Sudden Cardiac Death in Heart Failure Trial (SCD-HeFT); Comparison of Medical Therapy, Pacing, and Defibrillation in Heart Failure (COMPANION); Defibrillators in Non-Ischemic Cardiomyopathy Treatment Evaluation (DEFINITE); and Amiodarone versus Implantable Defibrillator Randomized Trial (AMIOVIRT).

The 2004 Assessment made the following conclusions about the use of ICD devices.

The use of ICD devices meets the TEC criteria in the prevention of sudden death from ventricular tachyarrhythmias in patients who have:

- Symptomatic (defined as the presence of dyspnea on exertion, angina, palpitations, or fatigue) ischemic dilated cardiomyopathy with a history of MI at least 40 days before ICD treatment and LVEF of 35% or less; or
- Symptomatic (defined as the presence of dyspnea on exertion, angina, palpitations, or fatigue) nonischemic dilated cardiomyopathy for more than 9 months’ duration and LVEF of 35% or less.

The use of ICD devices does not meet the TEC criteria in the prevention of sudden death from ventricular tachyarrhythmia in patients who

- have had an acute MI (ie, 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 cardiac revascularization procedure in past 3 months (CABG [coronary artery bypass grafting] or percutaneous transluminal coronary angioplasty [PTCA]) or are candidates for a cardiac revascularization procedure; or
• have noncardiac disease that would be associated with life expectancy less than 1 year.

The 2004 TEC Assessment based its conclusions on the following indication-specific evidence.

**Patients Who Have Prior MI and Reduced LVEF**
The 2002 Assessment concluded the evidence was sufficient to demonstrate that ICD therapy improves net health outcome for primary prevention in patients with prior MI and reduced LVEF. Both new trials (SCD-HeFT, COMPANION) and the reanalysis of MUSTT findings provided additional supportive evidence of improved outcomes in patients with prior MI. The hazard ratio (HR) for all-cause mortality in the ischemic subgroup of SCD-HeFT was 0.79 (95% confidence interval [CI], 0.60 to 1.04), which was close to that observed in MADIT II (HR=0.69; 95% CI, 0.51 to 0.93), and these findings provided additional supportive evidence that ICD therapy reduces mortality. There might be slight but not statistically significantly increased rates of adverse effects (AEs) associated with ICD therapy; however, serious device-related events are not common. On balance, the significant reductions in mortality associated with ICD therapy outweighed the harms associated with ICD therapy compared with conventional treatment. Thus, the available evidence demonstrated that ICD therapy improves health outcomes in patients with coronary artery disease and prior MI and reduced LVEF.

**Patients Who Have Acute MI and Reduced LVEF**
The evidence reviewed in the 2004 TEC Assessment was insufficient to permit conclusions regarding the effect of ICD therapy as primary prevention on the net health outcome for with acute MI and reduced LVEF.

**Patients Who Have No Prior MI and Reduced LVEF (eg, NIDCM)**
Results from subjects with NIDCM included in SCD-HeFT and DEFINITE suggested a mortality benefit from ICD therapy, although lack of statistical significance in these studies was likely related to insufficient power. A meta-analysis of 5 trials including nonischemic subjects reported a statistically significant reduction in mortality associated with ICD therapy. Furthermore, when the body of evidence for ICD therapy in both ischemic and nonischemic populations is considered, the preponderance of evidence suggested that ICD therapy improves health outcomes compared with medical management alone with a relative risk reduction in all-cause mortality between 21% and 35%. While the risk of AEs was not well-reported in studies of patients without prior MI, it seemed reasonable to expect similar low rates of device-related AEs as seen in studies of patients with prior MI.
Subsequent Evidence for the Use of ICDs as Primary Prevention in Adults

Relevant evidence and most current guidelines identified through Medline published after the 2004 TEC Assessment through October 2015 relates to the following subjects:

- Identification of predictors of better or worse outcomes after ICD placement.
- Use of ICD after acute MI: Reports of BEta-blocker STrategy + ICD (BEST-ICD), and Immediate Risk Stratification Improves Survival (IRIS) trials
- Use of ICD in NIDCM, with focus on implantation timing
- Use of ICD in HCM

In 2013, the Agency for Healthcare Research and Quality (AHRQ) Technology Assessment Program published an assessment of the evidence of ICDs for primary prevention of SCD. The review was structured on 3 questions:

- Key Question 1 examined the clinical effectiveness of the ICD versus no ICD, ICD with antitachycardia pacing (ATP) versus ICD alone, or ICD with cardiac resynchronization therapy (CRT) versus ICD alone, and differences among subgroups.
- Key Question 2 examined early and late AEs and inappropriate shocks after ICD implantation, and differences among subgroups.
- Key Question 3 examined eligibility criteria and evaluation methods for patients included in comparative studies and the risk of SCD.

The review included 14 studies comparing ICD with no ICD, 3 studies comparing combined ICD-CRT (CRT-D) with ICD, and 59 articles contributing data on AEs after ICD implantation. Focusing on the evidence reported in the AHRQ report related to the efficacy of ICDs alone, a meta-analysis of 7 RCTs comparing ICD with control was associated with a summary HR of 0.69 (95% CI, 0.60 to 0.79) for death (favoring ICD treatment). A meta-analysis of 5 studies comparing ICD with control was associated with a summary HR of 0.37 (95% CI, 0.26 to 0.52) for reducing SCD. There was low-strength evidence that failed to show a consistent effect of ICDs on quality of life (QOL). One subgroup evaluated was time since MI. The authors concluded:

“An indirect comparison of IRIS and DINAMIT (which included patients with recent MIs, within 31 or 40 days) versus the remaining trials, suggests that patients with recent MIs may have no reduction in all-cause mortality (HR 1.05 [95% CI 0.86, 1.30]) than patients with more distant or no prior MIs (HR 0.69 [95% CI 0.60, 0.79]). By meta-regression, the difference between IRIS and DINAMIT and the other seven RCTs is statistically significant (P = 0.012).”

ICDs for Primary Prevention in Adults After MI

BEST-ICD Trial
The BEST-ICD trial randomized 143 patients 5 to 30 days after acute MI to evaluate whether electrophysiology studies were useful to guide ICD placement.
and improve outcomes in patients at high risk of sudden death. Entry criteria included an LVEF of 35% or less along with 1 or more noninvasive risk factors (eg, premature ventricular contractions, heart rate variability, signal-averaged electrocardiography [SAECG]-positive), and receiving therapy with maximally tolerated β-blockers (metoprolol). The authors concluded that using electrophysiology studies to guide ICD placement within 5 to 30 days after MI did not significantly improve outcomes or survival. This finding is consistent with the conclusions that ICD placement after early MI does not improve outcomes. The authors also noted that the trial screened more than 15,000 patients but ended after randomizing only 12% of the targeted study population, largely because there were far fewer patients with LVEF below 35% than expected based on experience reported in the literature.

**IRIS Trial**
The IRIS trial evaluated ICD implantation early after MI. Eligible patients were required to have an LVEF of 40% or less and either: (1) a heart rate of 90 or more beats per minute on initial electrocardiogram or (2) nonsustained ventricular tachycardia (VT) during Holter monitoring, or both. From 92 centers and 62,944 patients post-MI, 898 were randomized 5 to 31 days after the MI to ICD implantation or medical therapy. Seventy-seven percent had experienced ST elevation MI, 72% of whom underwent PTCA. During a mean 37-month follow-up, overall mortality was similar in the 2 arms (ICD vs medical therapy, HR=1.04; 95% CI, 0.81 to 1.35). However, the risk of SCD was lower following ICD (HR=0.52; 95% CI, 0.35 to 0.78), but non-SCD risk was greater (HR=1.8; 95% CI, 1.0 to 3.2). These results are consistent with guidelines and previous trials.

**ICDs for Primary Prevention in Adults With High-Risk HCM**
Maron et al reported appropriate ICD discharge rates (terminating either VT or ventricular fibrillation [VF]) from an international registry of high-risk HCM patients enrolled at 42 referral and nonreferral institutions. Between 1986 and 2003, ICDs were implanted in 506 patients with HCM—383 for primary prevention and 123 for secondary prevention. Mean age of patients was 42 years (SD=17), and 28% were 30 years of age or younger; 36% were female; mean follow-up was 3.7 years (SD=2.8). Criteria defining patients as high risk and, therefore, candidates for primary prevention included: (1) history of premature HCM-related sudden death in 1 or more first-degree relatives younger than 50 years of age; (2) left ventricular hypertrophy greater than 30 mm; (3) 1 or more runs of nonsustained VT at heart rates of 120 beats per minute or greater on 24-hour Holter monitoring; and (4) prior unexplained syncope inconsistent with neurocardiogenic origin. Abnormal exercise blood pressure was not reported. In the primary prevention group, appropriate discharges occurred at an annual rate of 3.6% (95% CI, 2.7% to 4.8%); in the secondary prevention group, 10.6% (95% CI, 7.9% to 13.9%). Respective 5-year cumulative probabilities of first appropriate discharge were 17% and 39%. If each appropriate discharge was life-saving, 5-year numbers needed to benefit (NNTBs) could have been as low as 5.9 and 2.6 for primary and secondary prevention, respectively, when considering only the first appropriate discharge.
However, when analyzed in NIDCM, Ellenbogen et al concluded that approximately one-half of arrhythmias terminated by appropriate ICD discharges were not life-threatening. The NNTBs calculated in the Maron study, therefore, represent lower bounds or greatest potential benefit, and the true benefit is likely less (only 6.3% of primary prevention patients had >1 appropriate discharge). AE rates included 1 or more inappropriate discharges (27%), infections (3.8%), hemorrhage or thrombosis (1.6%), and lead fractures, dislodgement, and oversensing (6.7%). While the number of risk factors present was not associated with cumulative probability to first appropriate discharge for primary prevention, patient selection for ICD implantation was performed by experienced clinicians. These results, obtained outside the setting of a clinical trial, apply under such conditions.

In 2015, Magnusson et al reported outcomes for 321 patients with HCM treated with an ICD enrolled in a Swedish registry. Over a mean follow-up of 5.4 years, appropriate ICD discharges in response to VT or VF occurred in 77 (24%) patients, corresponding to an annual rate of appropriate discharges of 5.3%. At least 1 inappropriate shock occurred in 46 (14.3%) patients, corresponding to an annualized event rate of 3.0%. Ninety-two (28.7%) patients required at least 1 surgical intervention for an ICD-related complication, with a total of 150 ICD-related reinterventions. Most reinterventions (n=105 [70%]) were related to lead dysfunction.

**ICDs for Primary Prevention in Adults With NIDCM**

For patients with nonischemic cardiomyopathy (NICM), the optimal timing of ICD implantation remains uncertain. A substantial percentage of patients diagnosed with NICM will improve following initial diagnosis, even when a reversible cause of NICM cannot be identified. Given the current available evidence, it is not possible to predict which patients with idiopathic NICM will improve, nor is it possible to accurately estimate the time course for improvement. The specification of a 9-month waiting period before ICD implantation arises from the selection criteria of the CAT trial, which restricted enrollment to patients with onset of NICM within 9 months. While the results of this trial did not show a benefit for patients with recent onset of NICM, the trial was stopped early due to an unexpectedly low rate of events and was thus underpowered to detect a difference in mortality between groups.

Kadish et al performed a post hoc analysis of the DEFINITE trial data to examine whether the time from diagnosis of NIDCM was associated with the magnitude of benefit from ICD implantation. Survival benefit was found only for those diagnosed less than 9 months before implantation (n=216); no benefit was apparent when NIDCM was diagnosed more than 9 months before (n=242). However, there was a significant discrepancy between arms in the time from diagnosis to randomization—standard therapy patients were randomized a median of 20 months after diagnosis, while those in the ICD arm were randomized at a median of 8 months. The trial was neither designed nor powered to examine a time effect, and the analyses conflict with findings of the smaller (N=104) CAT trial reviewed in the 2002 TEC Assessment. Further evidence is necessary to
define when in the natural history of the disease is appropriate for ICD implantation.

The DEFINITE trial enrolled NICM patients without regard to time since onset, and a post hoc analysis revealed that the benefit was found mainly in patients with onset of NICM less than 9 months. Neither of these pieces of evidence represents strong data to support a specific time interval before implanting an ICD in patients with NICM.

Zecchin et al performed a cohort study on 503 consecutive patients diagnosed with idiopathic NICM to determine the extent to which indications for an ICD evolve over several months after an initial NICM diagnosis. At initial diagnosis, 245 patients met SCD-HeFT criteria for ICD implantation, based on an ejection fraction less than 35% and class II or III heart failure; 258 patients did not meet criteria. At a mean follow-up of 5.4 months, during which patients were treated with angiotensin-converting enzyme inhibitors and β-blockers, there were consistent improvements in ejection fraction and symptoms, such that less than one-third (31%) of evaluable patients still had indications for ICD. Of patients who initially did not have an indication for an ICD, a total of 10% developed indications for an ICD at follow-up. This study highlights the fact that a decision for ICD implantation should not be made before optimal treatment and stabilization of patients with newly diagnosed NICM, because the indications for ICD are not stable over time and will change in a substantial numbers of patients following treatment.

A prospective registry sponsored by the National Heart, Lung, and Blood Institute enrolled 373 patients with recent-onset NICM, and compared mortality in patients receiving an early ICD with those receiving the device at a later time. Forty-three patients received an ICD within 1 month of diagnosis, with a 1-year survival for this group of 97%. Three hundred thirty patients received an ICD between 1 and 6 months, with a 1-year survival of 98%. Seventy-three patients received an ICD at a time after 6 months, with a 1-year survival of 98%. Survival at 2 and 3 years was also similar between groups, with no significant differences.

Some experts consider patients with recently diagnosed NICM and either sustained VT or unexplained syncope to be candidates for earlier ICD implantation due to their higher risk of lethal arrhythmias. However, evidence on this specific population is lacking, and the natural history of patients in this category is not well-characterized. American College of Cardiology and American Heart Association guidelines on device-based therapies do not specifically address the optimal waiting period before implantation of an ICD for patients with newly diagnosed NICM.

Section Summary: ICD for Primary Prevention for in Adults
A large body of RCTs has addressed the effectiveness of TV-ICD implantation for primary prevention in patients at high risk of SCD due to ischemic cardiomyopathy and NIDCM. Evidence from several RCTs has demonstrated improvements in outcomes with ICD treatment for patients with symptomatic heart failure due to
ischemic or NICM with LVEF of 35% or less. The notable exceptions are that data from several RCTs, including the BEST-ICD and IRIS trials and subanalyses from earlier RCTs, show that outcomes with ICD therapy do not appear to improve for patients treated with an ICD within 40 days of acute MI. Less evidence is available for use of ICDs for primary prevention in patients with HCM. In several cohort studies, the annual rates of appropriate ICD discharge ranged from 3.6% to 5.3%. Given the long-term high risk of patients with HCM 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 HCM.

**ICDs in Patients With Hereditary Arrhythmia Syndromes**

ICDs have been used for primary and secondary prevention in patients with a number of hereditary disorders that predispose to ventricular arrhythmias and SCD, 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 use of ICDs has been described in small cohorts of patients with LQTS, BrS, and CPVT.

**Congenital Long QT Syndrome**

In 2010, Horner et al reported on outcomes for 51 patients with genetically confirmed LQTS treated with an ICD from 2000 to 2010 who were included in a single-center retrospective analysis of 459 patients with genetically confirmed LQTS. Of patients treated with ICDs, 43 (84%) received the device as primary prevention. Twelve (24%) patients received appropriate VF or torsades de pointes-terminated ICD shocks. Factors associated with appropriate shocks included secondary prevention indications (p=0.008), QT corrected (QTc) duration greater than 500 ms (p<0.001), non-LQT3 genotype (p=0.02), documented syncope (p=0.05), documented torsades de pointes (p=0.003), and a negative sudden family death history (p<0.001). Inappropriate shocks were delivered in 15 (29%) patients. Patients with the LQT3 genotype only received inappropriate shocks.

**Brugada Syndrome**

Conte et al described outcomes for a cohort of 176 patients with spontaneous or drug-induced Brugada type 1 electrocardiographic (ECG) findings who received an ICD at a single institution and were followed for at least 6 months. Before ICD implantation, 14.2% of subjects had a history of aborted SCD due to sustained spontaneous ventricular arrhythmias, 59.7% had at least 1 episode of syncope, and 25.1% were asymptomatic. Over a mean follow-up of 83.8 months, 30 (17%) patients had spontaneous sustained ventricular arrhythmias detected. Sustained ventricular arrhythmias were terminated by ICD shocks or antitachycardia pacing in 28 (15.9%) patients and 2 (1.1%) patients, respectively. However, 33 (18.7%) patients experienced inappropriate shocks. Eight (4.5%) patients died during follow-up, 3 of cardiac causes.

Dores et al reported results of a Portuguese registry that included 55 patients with BrS, 36 of whom were treated with ICDs for primary or secondary prevention. Before ICD implantation, 52.8% of subjects were asymptomatic, 30.6% had a history of syncope with suspected arrhythmic cause, and 16.7% had a history of
aborted SCD. Over a mean follow-up of 74 months, 7 patients experienced appropriate shocks, corresponding to an incidence rate of 19.4% and an annual event rate of 2.8%. In multivariable analysis, predictors of appropriate shocks were a history of aborted SCD (HR=7.87; 95% CI, 1.27 to 49.6; p=0.027) and nonsustained VT during follow-up (HR=6.73; 95% CI, 1.27 to 35.7; p=0.025).

In data from a U.S. cohort of 33 patients with BrS treated with ICDs, Steven et al reported that two-thirds of patients with a prior history of aborted SCD received appropriate shocks over a mean 7.9 years of follow-up, while none of the 30 patients without a history of aborted SCD had an arrhythmia detected. In a smaller registry that included 25 patients with BrS treated with ICDs, over an average follow-up of 41.2 months, appropriate shocks were delivered in 3 patients, all of whom had prior cardiac arrest.

**Catecholaminergic Polymorphic Ventricular Tachycardia**
Roses-Noguer et al reported results of a small retrospective study of 13 patients with CPVT who received an ICD. The indication for ICD therapy was syncope despite maximal β-blocker therapy in 6 (46%) patients and aborted SCD in 7 (54%) patients. Over a median follow-up of 4.0 years, 10 (77%) patients received a median 4 shocks. For 96 shocks, 87 ECGs were available for review; of those, 63 (72%) were appropriate and 24 (28%) inappropriate. Among appropriate shocks, 20 (32%) restored sinus rhythm.

**AEs in Patients With Hereditary Arrhythmia Syndromes**
In contrast to patients requiring ICDs for primary or secondary prevention after acute MI, patients with hereditary arrhythmia syndromes are more likely to require ICDs for primary prevention.

In 2016, Olde Nordkamp et al reported on a systematic review and meta-analysis of studies reporting on ICD complications in individuals with inherited arrhythmia syndromes. The review included 63 cohort studies with a total of 4916 patients (710 [10%] with arrhythmogenic right VT; 1037 [21%] with BrS; 28 [0.6%] with CPVT; 2466 [50%] with HCM; 162 [3.3%] with lamin A/C gene mutations; 462 [9.4%] with LQTS; 51 [1.0%] with SQTS). Overall, inappropriate shocks occurred in 20% over a mean follow-up of 51 months, corresponding to an inappropriate shock rate of 4.7% per year (95% CI, 4.2% to 5.3%). Over a mean follow up of 55 months, ICD-related complications occurred in 22%, most commonly lead malfunction (10.3% of patients). The pooled rate of ICD-related complications was 4.4% per year (95% CI, 3.6% to 5.2%).

**Section 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, BrS, and CPVT that report on appropriate shock rates. Patient populations typically include a mix of those requiring ICD implantation for primary or secondary prevention. The limited available data for ICDs for LQTS and CPVT have reported high rates of appropriate shocks. For BrS, more data are available.
and have suggested 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.

**TV-ICDs in Pediatric Population**  
There is limited direct scientific evidence on the efficacy of ICDs in the pediatric population. Most published studies retrospectively analyze small case series. A review of some representative series is provided next.

The largest published series combined pediatric patients and patients with congenital heart disease from 4 clinical centers. The median age was 16 years, although some adults included were to the age of 54 years. A total of 443 patients were included. The most common diagnoses were tetralogy of Fallot and HCM. ICD implantation was performed for primary prevention in 52% of patients and for secondary prevention in 48%. Over a 2-year follow-up, appropriate shocks occurred in 26% of patients and inappropriate shocks occurred in 21%.

Silka et al compiled a database of 125 pediatric patients treated with an ICD, through query of the manufacturers of commercially available devices. Indications for ICD placement were survivors of cardiac arrest in 95 (76%) patients, drug-refractory VT in 13 (10%) patients, and syncope with heart disease plus inducible VT in 13 (10%) patients. During a mean follow-up of 31 months, 73 (59%) patients received at least 1 appropriate shock and 25 (20%) received at least 1 inappropriate shock. Actuarial rates of SCD-free survival were 97% at 1 year, 95% at 2 years, and 90% at 5 years.

Alexander et al reported on 90 ICD procedures in 76 young patients with a mean age of 16 years (range, 1-30 years). Indications for placement were 27 (36%) patients with cardiac arrest or sustained VT, 40 (53%) patients with syncope, 17 (22%) patients with palpitations, 40 (53%) patients with spontaneous ventricular arrhythmias, and 36 (47%) patients with inducible VT. Numerous patients had more than 1 indication for ICD in this study. Over a median follow-up of 2 years, 28% of patients received an appropriate shock and 25% of patients received an inappropriate shock. Lewandowski et al reported on long-term follow-up of 63 patients, between the ages 6 and 21 years, who were treated with an ICD device. At 10-year follow-up, 13 (21%) patients had surgical infections. Fourteen (22%) patients experienced at least 1 appropriate shock and 17 (27%) had at least 1 inappropriate shock. Serious psychological sequelae developed in 27 (43%) patients.

**Section Summary: TV-ICDs in Pediatric Patients**  
The available evidence for the use of ICDs in pediatric patients is limited and consists primarily of small case series that include mixed populations with mixed indications for ICD placement. Overall, these studies reported both relatively high rates of appropriate and inappropriate shocks. Pediatric patients may also be eligible ICD implantation if they have hereditary arrhythmias syndromes (see ICDs in Patients With Hereditary Arrhythmia Syndromes section).
Adverse Events Associated With TV-ICDs

Perrson et al conducted a systematic review of AEs following ICD implantation. They included data from 35 cohort studies, reported in 53 articles. In-hospital serious AE rates ranged from 1.2% to 1.4%, most frequently pneumothorax (0.4%-0.5%) and cardiac arrest (0.3%). Posthospitalization complication rates varied: device-related complications occurred in 0.1% to 6.4%; lead-related complications in 0.1% to 3.9%; infection in 0.2% to 3.7%; thrombosis in 0.2% to 2.9%; and inappropriate shock in 3% to 21%.

In another systematic review of AEs following ICD implantation, Ezzat et al compared rates of AEs reported in clinical trials of ICDs with those reported in the U.S. National Cardiovascular Data Registry. The review included 18 RCTs (total N=6796 patients). In pooled analysis, the overall AE rate was 9.1% (95% CI, 6.4% to 12.6%). Rates of access-related complications, lead-related complications, generator-related complications, and infection were 2.1% (91% CI, 1.3% to 3.3%), 5.8% (95% CI, 3.3% to 9.8%), 2.7% (95% CI, 1.3% to 5.7%), and 1.5% (95% CI, 0.8% to 2.6%), respectively. Complication rates in the RCTs were higher than those in the U.S. registry, which reports only in-hospital complications (9.1% in the RCTs vs 3.08% in U.S. registry data, p<0.01). The overall complication rate was similar to that reported by Kirkfelt et al, in a population-based cohort study including all Danish patients who underwent a cardiac implantable electronic device procedure from 2010 to 2011 (562/5918 patients [9.5%] with at least 1 complication).

In 2011, van Rees et al reported results of a systematic review of implantation-related complications in RCTs of ICDs and cardiac resynchronization therapy (CRT) devices. The review included 18 trials and 3 subgroup analyses. Twelve trials assessed ICDs, 4 of which used both thoracotomy and nonthoracotomy ICDs (n=951) and 8 of which used nonthoracotomy ICDs (n=3828). For nonthoracotomy ICD implantations, the rates for in-hospital and 30-day mortality were 0.2% and 0.6%, respectively, and pneumothorax was reported in 0.9% of cases. For thoracotomy ICD implantations, the average in-hospital mortality rate was 2.7%. For nonthoracotomy ICD implantations, the overall lead-dislodgement rate was 1.8%.

In a large retrospective, single-center study including 1043 transvenous lead extraction procedures in 985 patients, Maytin et al reported a cumulative mortality rates of 2.1%, 4.2%, 8.4%, and 46.8% at 30 days, 3 months, 1 year, and 10 years postprocedure, respectively. Most lead extractions were due to infection (50%) or lead malfunction (30%). Of the 21 patients with an initial extraction due to infection, 10 required another extraction procedure for infection. Lead extraction due to infection was associated with a significantly increased mortality risk. However, it is unclear whether this mortality risk was related to the ICD lead extraction or underlying patient morbidity.
Lead Failure
The failure of leads in several specific ICD devices led the U.S. Food and Drug Administration (FDA) to require St. Jude Medical to conduct 3-year postmarket surveillance studies to address concerns related to premature insulation failure and to address important questions related to follow-up of affected patients. A 2010 report had found that 57 deaths and 48 serious cardiovascular injuries associated with device-assisted ICD or pacemaker lead extraction were reported to the FDA’s Manufacturers and User Defined Experience (MAUDE) database.

In 2015, Providencia et al reported on a meta-analysis of 17 observational studies evaluating lead performance, including a total of 49,871 leads (5538 Durata, 10,605 Endotak Reliance, 16119 Sprint Quattro, 11,709 Sprint Fidelis, 5900 Riata). Overall, the incidence of lead failure was 0.93 per 100 lead-years (95% CI, 0.88 to 0.98). In analysis of studies restricted to head-to-head comparisons of leads, there was no significant difference in the lead failure rate among nonrecalled leads (Endotak Reliance, Durata, Sprint Quattro).

Birnie et al reported clinical predictors of failure for 3169 Sprint Fidelis leads implanted from 2003 to 2007 at 11 of 23 centers participating in the Canadian Heart Rhythm Society Device Committee study. A total of 251 lead failures occurred, corresponding to a 5-year lead failure rate of 16.8%. Factors associated with higher failure rates included female sex (HR=1.51; 95% CI, 1.14 to 2.04; p=0.005), axillary vein access (HR=1.94; 95% CI, 1.23 to 3.04), and subclavian vein access (HR=1.63; 95% CI, 1.08 to 2.46). In a previous study from 3 centers reporting on predictors of Fidelis lead failures, compared with Quattro lead failures, Hauser et al reported a failure rate for the Fidelis lead of 2.81% per year (vs 0.42% per year for Quattro leads; p<0.001).

In an earlier study from 12 Canadian centers, Gould et al reported outcomes from ICD replacements due to ICD advisories from 2004 to 2005, which included 451 replacements (of 2635 advisory ICD devices). Over 355 days of follow-up, 41 (9.1%) complications occurred, including 27 (5.9%) requiring surgical reintervention and 2 deaths.

In another multicenter study, Eckstein et al reported rates of lead failure among 1317 consecutive patients with an ICD implanted at 3 European centers from 1993 to 2004. The end point of lead failure, defined as a case requiring surgical revision to correct the lead-related problem, occurred in 38 patients. Lead malfunction resulted in inappropriate ICD therapies in 29 (76%) of the failures, while the remaining lead malfunctions were detected during routine follow-up. Over a median follow-up of 3.1 years, among the 38 patients with lead malfunction, the cumulative incidences of lead failure recurrence were 4.4%, 14.1%, and 19.8% at years 2, 3, and 4, respectively. A study reporting on another single-center European registry which included 990 patients who underwent first implantation of an ICD from 1992 to 2005, found an estimated lead survival rate of 85% and 60% after 5 and 8 years, respectively.
In a large prospective multicenter study, Poole et al reported complications rates associated with generator replacements and/or upgrade procedures of pacemaker or ICD devices, which included 1031 patients without a planned transvenous lead replacement (cohort 1) and 713 with a planned transvenous lead replacement (cohort 2). A total of 9.8% and 21.9% of cohort 1 and 19.2% and 25.7% of cohort 2 had a single-chamber ICD and a dual-chamber ICD, respectively, at baseline. The overall periprocedural complication rates for those with a planned transvenous lead replacement were cardiac perforation in 0.7%, pneumothorax or hemothorax in 0.8%, cardiac arrest in 0.3%, and, most commonly, need to reoperate because of lead dislodgement or malfunction in 7.9%. Although rates were not specifically reported for ICD replacements, complication rates were higher for ICDs and CRT devices than pacemakers.

Ricci et al evaluated the incidence of lead failure in a cohort study of 414 patients implanted with an ICD with Sprint Fidelis leads. Patients were followed for a median of 35 months. Lead failures occurred in 9.7% (40/414) of patients, for an annual rate of 3.2% per patient-year. Most lead failures (87.5%) were due to lead fracture. Median time until recognition of lead failure, or until an AE, was 2.2 days. A total of 22 (5.3%) patients received an inappropriate shock due to lead failure.

Cheng et al examined the rate of lead dislodgements in patients enrolled in a national cardiovascular registry. Of 226,764 patients treated with an ICD between April 2006 and September 2008, lead dislodgement occurred in 2628 (1.2%). Factors associated with lead dislodgement were NYHA class IV heart failure, AF or atrial flutter, a combined ICD-CRT device, and having the procedure performed by a nonelectrophysiologist. Lead dislodgement was associated with an increased risk for other cardiac AEs and death.

In a single-center study, Borleffs et al reported on the risk of transvenous lead failure among 2068 ICD patients with 2161 defibrillation leads. Over a mean follow-up of 885 days, 146 (6.8%) leads were removed or capped in 139 patients. In 64 patients, the cause of removal or capping was not lead failure. Eighty-two cases of lead failure were identified, with a median time to failure of 1187 days (interquartile range, 597-1783 days). The cumulative incidences of lead failure-free follow-up at 1, 2, 5, and 10 years were 99.4%, 98.6%, 93.5%, and 83.6%, respectively. In another single-center study, Faulknier et al reported on the time-dependent hazard of failure of the Sprint Fidelis leads for 426 leads implanted at a single center. Over an average follow-up of 2.3 years, 38 (8.92%) leads failed. There was a 3-year survival of 90.8% (95% CI, 87.4% to 94.3%), with a hazard of fracture increasing exponentially over time by a power of 2.13 (95% CI, 1.98 to 2.27; p<0.001).

**Infection Rates**
Several publications have reported infection rates in patients receiving an ICD. Smit et al published a retrospective, descriptive analysis of the types and distribution of infections associated with ICDs over a 10-year period in Denmark. Of 91 total infections identified, 39 (42.8%) were localized pocket infections, 26 (28.6%) were endocarditis, 17 (18.7%) were ICD-associated bacteremic
infections, and 9 (9.9%) were acute postsurgical infections. Nery et al reported the rate of ICD-associated infections among consecutive patients treated with an ICD at a tertiary referral center.\textsuperscript{47} Twenty-four of 2417 patients had infections, for a rate of 1.0%. Twenty-two (91.7%) of the 24 patients with infections required device replacement. Factors associated with infection were device replacement (vs de novo implantation) and use of a complex device (eg, combined ICD-CRT or dual-/triple-chamber devices). Sohail et al performed a case-control study evaluating the risk factors for infection in 68 patients with an ICD infection and 136 matched controls.\textsuperscript{48} On multivariate analysis, the presence of epicardial leads (odds ratio [OR], 9.7; \(p=0.03\)) and postoperative complications at the insertion site (OR=27.2, \(p<0.001\)) were significant risk factors for early infection. For late-onset infections, hospitalization for more than 3 days (OR=33.1, \(p<0.001\) for 2 days vs 1 day) and chronic obstructive pulmonary disease (OR=9.8, \(p=0.02\)) were significant risk factors.

Chua et al described the diagnosis and management of ICD infections in a retrospective case series that included 123 patients, 36 of whom were treated for ICD infections.\textsuperscript{49} Most (\(n=117\) [95%]) patients required removal of the device and all lead material. Of those who had all hardware removed, 1 patient experienced a relapse, while 3 of the 6 patients who did not undergo hardware removal experienced a relapse.

Borleffs et al also reported on complications after ICD replacement for pocket-related complications, including infection or hematoma, in a single-center study.\textsuperscript{50} Of 3161 ICDs included, 145 surgical reinterventions were required in 122 ICDs in 114 patients. Ninety-five (66%) reinterventions were due to infection and the remaining 50 (34%) were due to other causes. Compared with first-implanted ICDs, the occurrence of surgical reintervention in replacements was 2.5 (95% CI, 1.6 to 3.7) times higher for infectious and 1.7 (95% CI, 0.9 to 3.0) times higher for noninfectious causes.

**Inappropriate Shocks**

Inappropriate shocks may occur with ICDs due to faulty sensing or sensing of atrial arrhythmias with rapid ventricular conduction; they may lead to reduced QOL and risk of ventricular arrhythmias. In the MADIT II trial (described above), 1 or more inappropriate shocks occurred in 11.5% of ICD subjects and were associated with a greater likelihood of mortality (HR=2.29; 95% CI, 1.11 to 4.71; \(p=0.02\)).\textsuperscript{51}

Tan et al conducted a systematic review to identify outcomes and AEs associated with ICDs with built-in therapy-reduction programming.\textsuperscript{52} Six randomized trials and 2 nonrandomized cohort studies (total \(N=7687\) patients) were included (3598 with conventional ICDs, 4089 therapy-reduction programming). A total of 267 (4.9%) patients received inappropriate ICD shocks, 99 (3.4%) in the therapy reduction group and 168 (6.9%) in the conventional programming group (relative risk [RR], 50%; 95% CI, 37% to 61%; \(p<0.001\)). Therapy-reduction programming was associated with a significantly lower risk of death than conventional programming (RR=30%; 95% CI, 16% to 41%; \(p<0.001\)).
Sterns et al (2016) reported results of an RCT comparing a strategy using a prolonged VF detection time to reduce inappropriate shocks with a standard strategy among secondary prevention patients. The present study is a prespecified subgroup analysis of the PainFree SST trial, which compared standard and prolonged detection in patients receiving an ICD for secondary prevention. Patients who were treated for secondary prevention indications were randomized to a prolonged VF detection period (“Number of Intervals to Detect” VF 30/40; n=352) or a standard detection period (“Number of Intervals to Detect” VF 18/24; n=353). At 1 year, arrhythmic syncope-free rates were 96.9% in the 30/40 (intervention) group and 97.7% in the 18/24 (control) group (rate difference, -1.1%; 90% lower confidence limit, -3.5%; above the prespecified noninferiority margin of -5%; noninferiority p=0.003).

An earlier (2015) nonrandomized prospective trial by Auricchio et al evaluated newer-generation ICD programming strategies for reducing inappropriate shocks. The study included 2790 patients with an indication for ICD implantation who were given a device programmed with a SmartShock Technology designed to differentiate between ventricular arrhythmias and other rhythms. The inappropriate shock incidence for dual-/triple-chamber ICDs was 1.5% at 1 year (95% CI, 1.0% to 2.1%), 2.8% at 2 years (95% CI, 2.1% to 3.8%), and 3.9% at 3 years (95% CI, 2.8% to 5.4%).

Other Complications
Lee et al evaluated the rate of early complications among patients enrolled in a prospective, multicenter population-based registry of all newly implanted ICDs in Ontario, from February 2007 through May 2009. Of 3340 patients receiving an ICD, major complications (lead dislodgement requiring intervention, myocardial perforation, tamponade, pneumothorax, infection, skin erosion, hematoma requiring intervention) within 45 days of implantation occurred in 4.1% of new implants. Major complications were more common in women, in patients who received a combined ICD-CRT device, and in patients with a left ventricular end-systolic size of larger than 45 mm. Direct implant-related complications were associated with a major increase in early death (HR=24.9; p<0.01).

Furniss et al prospectively evaluated changes in high-sensitivity troponin T (hs-TnT) level and ECG that occur during ICD implantation alone, ICD implantation with testing, and ICD testing alone. The 13 subjects undergoing ICD implantation alone had a median increase in hs-TnT of 95% (p=0.005) while the 13 undergoing implantation and testing had a median increase of 161% (p=0.005). Those undergoing testing alone demonstrated no significant change in hs-TnT levels.

Subcutaneous ICDs
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 antitachycardia overdrive pacing for VT. The S-ICD has been proposed as of particular benefit to patients with limited vascular access, including patients undergoing renal dialysis or children, or those who have had complications.
requiring TV-ICDs explantation. No RCTs were identified comparing the performance of an S-ICD with TV-ICDs. Two nonrandomized, comparative studies were identified that assessed the efficacy of the 2 types of ICDs, and numerous single-arm studies have reported on outcomes of the S-ICD.

S-ICD Efficacy

Nonrandomized Comparative Studies
Kobe et al compared the efficacy of the S-ICD and the TV-ICD in terminating laboratory-induced VF.57 Sixty-nine patients from 3 centers in Germany treated with an S-ICD were matched by age and sex with 69 patients treated with a TV-ICD. One patient in the TV-ICD group developed a pericardial effusion requiring pericardiocentesis. Termination of induced VF was successful in 89.5% of the patients in the S-ICD group and 90.8% of patients with a TV-ICD (p=0.815). Patients in both groups were followed for a mean of 217 days. One patient in the S-ICD group had the device explanted at 8 weeks due to local infection, and a second patient had the S-ICD changed to a TV-ICD because of the need for antitachycardia overdrive pacing due to frequent episodes of VT. Three patients in the S-ICD group received appropriate shocks for ventricular arrhythmias compared with 9 patients in the TV-group (p=0.05). Inappropriate shocks occurred in 5 patients in the S-ICD group and 3 patients in the TV-ICD group (p=0.75).

The Subcutaneous versus Transvenous Arrhythmia Recognition Testing (START) study compared the performance of an S-ICD with a TV-CD for detecting arrhythmias in the electrophysiology lab.58 The population included 64 patients scheduled for ICD implantation. All patients had a TV-ICD placed as well as subcutaneous electrodes attached to an S-ICD. Arrhythmias were induced and the sensitivity and specificity of detection by each device were compared. For ventricular arrhythmias, sensitivity of detection was 100% for the S-ICD and 99% for the TV-ICD. Specificity was 98.0% for the S-ICD device and 76.7% for the transvenous device (p<0.001).

Pettit et al reported on a small study comparing the S-ICD with TV-ICDs in children treated at 2 Scottish centers.59 The study included 15 patients, 9 treated with S-ICDs and 6 treated with 8 TV-ICDs. There were no deaths in either group over at least 1 year of follow-up. For the study’s secondary outcome (survival free of inappropriate ICD therapy or surgical reintervention), rates were higher for the S-ICD group (89%) than for the transvenous group (25%; p=0.024). In another small study, Jarman et al reported on 16 patients with median age 20 years (range, 10-48 years) treated with S-ICDs.60 All procedures were completed without acute complications, but 3 children required surgical reintervention. Four patients experienced inappropriate shocks, all due to T-wave oversensing.

Noncomparative Studies
The largest single-arm study reporting on outcomes with S-ICDs (Lambiase et al) described patients in the EFFORTLESS S-ICD Registry, a multicenter European registry to report outcomes for patients treated with S-ICD.61 At the time of analysis, the registry included 472 patients, 241 (51%) of whom were enrolled
prospectively, at a median follow-up of 498 days. Nine (2%) patients died during the reported period, none of which occurred in the perioperative period, although the cause of death was unknown for 1 patient. A total of 317 spontaneous episodes in 85 patients were recorded during the follow-up, of which 169 episodes received therapy (59 patients). Of the 145 classified untreated episodes, 93 were adjudicated as inappropriate sensing, 37 were nonsustained VT or VF, 12 were nonsustained sustained VT above the discrimination zone, and 3 were unclassified. Of the VT or VF episodes, the first shock conversion efficacy was 88%, with 100% overall successful clinical conversion after a maximum of 5 shocks. A total of 73 inappropriate shocks were recorded in 32 patients over an average follow-up of 18 months (360-day inappropriate shock rate, 7%).

Olde Nordkamp et al used data from the EFFORTLESS S-ICD Registry to evaluate rates of inappropriate shocks associated with the S-ICD. The patient population included 581 S-ICD recipients, 48 (8.3%) of whom experienced a total of 101 inappropriate shocks over a follow-up of 21.4 months. Most inappropriate shocks (73%) were related to T-wave oversensing.

A second large series was a multicenter study of 330 patients from several countries, the S-ICD System Clinical Investigation (S-ICD IDE Study). An S-ICD was successfully implanted in 314 (95.1%) of 330 patients. Laboratory-induced VF was successfully terminated in more than 90% of patients, which was one of the primary outcomes of the study. The second primary outcome (>99% freedom from complications at 180 days) was also met. Patients were followed for a mean of 11 months. There were 38 spontaneous episodes of VT in 21 (6.7%) patients, and all were successfully terminated. Inappropriate shocks were received by 41 (13.1%) patients.

Gold et al published a subanalysis of patients in the S-ICD IDE Study to evaluate a discrimination algorithm to reduce inappropriate shocks. Patients could receive 1 of 2 shock detection algorithms, a single- or double-zone configuration. In the single-zone configuration, shocks were delivered for detected heart rates above the programmed rate threshold. In the dual-zone configuration, arrhythmia discrimination algorithms were active in a lower rate zone up to a shockable heart rate threshold. At hospital discharge, dual-zone programming was used in 226 (72%) subjects and single-zone programming was used in the remaining 88 (28%) subjects. Inappropriate shocks occurred on 23 (10.2%) of 226 subjects with dual-zone programming and 23 (26.1%) of 88 subjects with single-zone programming. Freedom from appropriate shocks did not differ between groups (p<0.001).

In 2015, Burke et al published a pooled analysis of patients from the S-ICD IDE Study and the EFFORTLESS Registry, which included 882 patients. The poolability of data across the 2 studies was assessed by analysis of complications, appropriate and inappropriate shocks, conversion efficacy, and mortality by study, with additional analyses for outcomes that differed by study. Patients were followed for a mean of 651 (±345) days. Most (63%) patients presented with a history of previous TV-ICDs that required extraction due to infection. Within 30
days of the procedure, 4.5% of subjects experienced a complication, while 11.1% of subjects experienced a complication within 3 years of the procedure. The most common complication was infection requiring device removal/revision (17 events in 14 patients [1.7%]). Mortality was low: the annual mortality rate was 1.6% and the 2-year mortality rate was 3.2%. The Kaplan-Meier incidence of time to first therapy for VT or VF was 5.3% at 1 year, 7.9% at 2 years, and 10.5% at 3 years. Excluding VT or VF storms, 111 discrete VT or VF events were treated, with 100 (90.1%) terminated with the first shock, and 109 (98.2%) terminated within the 5 shocks available. The Kaplan-Meier incidence of time to first inappropriate shock was 13.1% at 3 years. In patients with dual-zone programming at the index procedure, the Kaplan-Meier incidence of inappropriate shock at 3 years was 11.7% compared with 20.5% with single-zone programming. A significant study effect was observed for inappropriate shocks (p=0.021), with a smaller proportion of inappropriate shocks in the EFFORTLESS group; but this effect was negated after correction for initially programmed number of zones, shock zone rate, and conditional zone rate.

In 2016, Boersma et al reported outcomes for patients in the S-ICD IDE Study and the EFFORTLESS Registry stratified by whether patients had been previously treated with a TV-ICD. At the time of analysis, 866 patients were available for inclusion. Of those, 75 (8.7%) were implanted with an S-ICD following TV-ICD extraction for a system-related infection and 44 (5.1%) were implanted following TV-ICD extraction for reasons other than system-related infection; the remaining 747 (86.3%) were de novo implants. Patients explanted for infection were older than patients whose TV-ICD was explanted for non-infection-related events and those with de novo implants (55.5, 47.8, and 49.9 years, respectively; p=0.01) were more likely to have an ICD for secondary prevention (42.7%, 37.2%, and 25.6%, respectively; p<0.001) and had a higher incidence of comorbidities. There were no significant differences in the rates of system- or procedure-related complications between patients whose TV-ICDs were explanted for infection, those whose TV-ICDs were explanted for noninfectious reasons, and de novo S-ICD patients (10.7%, 6.8%, and 9.6%, respectively; p=0.078).

Another subanalysis of the pooled S-ICD IDE Study and EFFORTLESS Registry data, which included 882 patients at analysis, evaluated the effect of surgical learning curves on implant time, procedure complications, and inappropriate shocks. Rates of complications were significantly lower in patients treated by the least experienced providers (9.8%) than those treated with the most experienced (5.4%; p=0.02).

In 2016, Lambiase et al evaluated use of the S-ICD in patients with HCM in the S-ICD IDE Study and the EFFORTLESS Registry. Lambiase reported on 99 patients with HCM, who were compared with 773 non–HCM patients. At the time of reporting, 3 episodes of ventricular arrhythmias had been identified in the HCM cohort, all of which were successfully terminated. In the HCM group, 12.5% of subjects had experienced an inappropriate shock at a mean follow-up of 22.0 months, which did not differ significantly from the rate in non–HCM patients (10.7%; p=NS).
Bardy et al described the development and testing of the device, including empirical evidence for the optimal placement of the subcutaneous electrode.69 A total of 55 patients were tested in the electrophysiology lab for termination of induced arrhythmias and subsequently followed for a mean of 10.1 months for successful termination of detected arrhythmias and clinical outcomes. In the electrophysiology lab study, intraoperative VF was induced in 53 of 55 patients. All episodes were correctly detected by the S-ICD. In 52 of 53 patients, 2 consecutive episodes of ventricular arrhythmia were successfully terminated. In the final patient, the arrhythmia was terminated only once. In the cohort portion of this study, 54 of 55 patients were alive at last follow-up. The 1 death was due to renal failure, and this patient requested removal of the S-ICD before death. An infection at the generator site occurred in 2 patients, necessitating a revision procedure. Another 3 patients had lead dislodgement requiring repositioning. Twelve episodes of VT were detected by the S-ICD, all of which were successfully terminated by countershock. In 2015, Theuns et al reported long-term follow-up of what appears to be the same cohort.70 Over a median follow-up of 5.8 years, 26 (47%) devices were replaced and 5 (9%) were explanted. Four (7%) patients required S-ICD explantation and replacement with a transvenous system, 2 due to a requirement for cardiac resynchronization therapy, 1 due to a requirement for bradycardia pacing, and 1 due to ineffective defibrillation testing. Most devices (81%) were replaced due to an elective replacement indication, at a median time to replacement of 5.0 years. Event-free rates for device replacement after 2, 4, and 6 years were 94%, 89%, and 30%, respectively. A total of 119 delivered shocks in 16 patients (29%) were recorded.

A series of 118 patients from 4 centers in the Netherlands was published in 2012. Patients were followed for a mean of 18 months.71 Device-related complications occurred in 14% of patients, including infection (5.9%), dislodgement of the device or leads (3.3%), skin erosion (1.7%), and battery failure (1.7%). In 1 patient, the S-ICD was replaced with a TV-ICD because of the need for antitachycardia pacing. Over the entire follow-up period, 8 patients experienced 45 appropriate shocks, with a first-shock conversion efficacy of 98%. Fifteen (13%) patients received a total of 33 inappropriate shocks. Two patients died, 1 due to cancer and 1 to progressive heart failure.

In another smaller series, Aydin et al reported outcomes for 40 consecutive patients implanted with S-ICDs at 3 German centers.72 Patients were considered for S-ICD if they met criteria for ICD implantation for primary or secondary prevention as specified by the American College of Cardiology/American Heart Association/European Society of Cardiology; did not have symptomatic bradycardia, incessant VT, or documented spontaneous, frequently recurring VT that was reliably terminated with antitachycardia pacing; and did not have pacemakers. Of the cohort, 25.0% had a prior TV-ICD, and 57.5% received the S-ICD for secondary prevention. Over a median follow-up of 229 days, S-ICD activity was recorded in 10.0% of patients, for whom 25 episodes were retrieved. Of these, 21 shock episodes were correctly identified as ventricular tachyarrhythmia. The overall S-ICD shock efficacy was 96.4% (95% CI, 12.8% to 100%).
El-Chami et al reported on single-center outcomes after S-ICD placement in patients with end-stage renal disease (ESRD) undergoing chronic dialysis, which included 79 patients who underwent S-ICD placement, 27 of whom were on chronic dialysis. This research was prompted by prior studies that suggested higher mortality rates for ESRD patients implanted with TV-ICDs. The composite outcome (frequency of death, heart failure hospitalization, or appropriate S-ICD shocks) was nonsignificantly higher in the ESRD group (23.8% per year vs 10.9% per year, \( p=0.317 \)), a difference primarily driven by a significantly higher incidence of appropriate S-ICD shocks in the ESRD group (17.9% per year vs 1.4% per year, \( p=0.021 \)). Koman et al found no significant differences in appropriate or inappropriate shocks between patients with on chronic hemodialysis (n=18) and nonhemodialysis patients (n=68) treated with S-ICD at a single institution.

**S-ICD Safety: Inappropriate Shocks**

Although Kobe et al reported no differences between inappropriate shock rates in patients treated TV-ICD or S-ICD, noncomparative studies have reported relatively high rates of inappropriate shocks with S-ICD. Inappropriate shocks from S-ICDs often result from T-wave oversensing. Because the sensing algorithm and the discrimination algorithm for arrhythmia detection are fixed in the S-ICD, management to reduce inappropriate shocks for an S-ICD differs from that for a TV-ICD. Kooiman et al reported inappropriate shock rates among 69 patients treated at a single center with an S-ICD between February 2009 and July 2012 who were not enrolled in 1 of 2 other concurrent trials. Over a total follow-up of 1316 months (median per patient, 21 months), the annual incidence of inappropriate shocks was 10.8%. In 8 patients, inappropriate shocks were related to T-wave oversensing. After patients underwent adjustment of the sensing vector, no further inappropriate shocks occurred in 87.5% of patients with T-wave oversensing.

Brisben et al (2015) described the development of an algorithm designed to reduce T-wave oversensing by S-ICDs. The algorithm was developed using 133 episodes of T-wave oversensing and 70 episodes of appropriately treated VT or VF collected from S-ICD logfiles and 174 VT or VF recordings from an ECG signal library. It was validated using 164 episodes of T-wave oversensing from S-ICD logfiles and 137 and 328 recorded episodes, respectively, of VT or VF and supraventricular tachycardia from an ECG signal library. The revised algorithm was associated with a reduction in T-wave oversensing of 39.8% (95% CI, 28.4% to 51.2%; \( p=0.001 \) vs baseline). Patient outcomes after the use of this algorithm have not been reported yet.

Groh et al evaluated an ECG screening test to identify potential S-ICD candidates at risk for T-wave oversensing. One hundred patients who had previously undergone TV-ICD implantation, who were not receiving bradycardia pacing and did not have an indication for pacing, were included. ECGs were obtained with lead placement to mimic the sensing vectors available on the S-ICD, and a patient was considered to qualify for S-ICD if the screening ECG template passed in any same
lead supine and standing, at any gain, and without significant morphologic changes in QRS complexes. Of the included subjects potentially eligible for S-ICD, 8% were failed based the ECG screening.

**Section Summary: Subcutaneous ICDs**

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.

**Ongoing and Unpublished Clinical Trials**

Some currently unpublished trials that might influence this review are listed in Table 2.

**Table 2. Summary of Key Trials**

<table>
<thead>
<tr>
<th>NCT No.</th>
<th>Trial Name</th>
<th>Planned Enrollment</th>
<th>Completion Date</th>
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<td>NCT01296022a</td>
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<td>Dec 2019</td>
</tr>
<tr>
<td>Unpublished</td>
<td></td>
<td></td>
<td></td>
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<td>NCT00673842a</td>
<td>Efficacy of Implantable Defibrillator Therapy After a Myocardial Infarction (REFINE-ICD)</td>
<td>1400</td>
<td>Dec 2019 (suspended)</td>
</tr>
</tbody>
</table>

NCT: national clinical trial.  
*a* Denotes industry-sponsored or cosponsored trial.

**Summary of Evidence**

For individuals who have a high risk of sudden cardiac death (SCD) due to ischemic or to nonischemic cardiomyopathy (NICM) in adulthood who receive transvenous implantable cardioverter defibrillator (ICD) (TV-ICD) placement, the evidence includes multiple well-designed, well-conducted randomized controlled trials (RCTs) and systematic reviews of these trials. Relevant outcomes are overall survival, morbid events, quality of life, and treatment-related morbidity and mortality. There is an extensive literature on the use of ICDs in patients with prior arrhythmogenic events and ischemic cardiomyopathy. Earlier trials first demonstrated a benefit in overall mortality for survivors of cardiac arrest and patients with potentially lethal cardiac arrhythmias. Multiple, well-done, RCTs have also shown a benefit in overall mortality for patients with ischemic cardiomyopathy and reduced ejection fraction. RCTs of early ICD implantation following acute myocardial infarction (MI) did not support a benefit for immediate versus delayed implantation for at least 40 days. For NICM, there is less clinical trial data, but the available evidence from a limited number of RCTs enrolling patients with NICM and from subgroup analysis of RCTs with mixed populations supports a survival benefit for this group. There is no high-quality evidence to determine whether early versus delayed implantation improves outcomes for patients with NICM and it is not possible to determine the optimal waiting period for ICD implantation following onset of NICM. At least 1 cohort study has reported that most patients who meet
criteria for an ICD at the time of initial NICM diagnosis will no longer meet the criteria several months after initiation of treatment. The evidence is sufficient to determine qualitatively that the technology results in a large improvement in the net health outcome.

For individuals who have a high risk of SCD due to hypertrophic cardiomyopathy (HCM) in adulthood who receive TV-ICD placement, the evidence includes several large registry studies. Relevant outcomes are overall survival, morbid events, quality of life, and treatment-related morbidity and mortality. In these studies, the annual rate of appropriate ICD discharge ranged from 3.6% to 5.3%. Given the long-term high risk of patients with HCM for SCD risk, with the assumption that appropriate shocks are life-saving, these rates are considered adequate evidence for the use of ICDs in patients with HCM. The evidence is sufficient to determine qualitatively that the technology results in a meaningful improvement in the net health outcome.

For individuals who have a high risk of SCD due to an inherited cardiac ion channelopathy who receive TV-ICD placement, the evidence includes small cohort studies of patients with these conditions treated with ICDs. Relevant outcomes are overall survival, morbid events, quality of life, and treatment-related morbidity and mortality. The limited evidence for patients with long QT syndrome (LQTS), catecholaminergic polymorphic ventricular tachycardia (CPVT), and Brugada syndrome (BrS) has reported high rates of appropriate shocks. No studies were identified on the use of ICDs for patients with short QT syndrome (SQTS). 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. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have need for a cardioverter defibrillator but no indications for antibradycardia pacing and no antitachycardia pacing–responsive arrhythmias who receive subcutaneous ICD (S-ICD) placement, the evidence includes nonrandomized studies and case series. Relevant outcomes are overall survival, morbid events, quality of life, and treatment-related morbidity and mortality. Nonrandomized controlled studies have reported success rates in terminating laboratory-induced ventricular fibrillation that are similar to TV-ICD. However, there is scant evidence on comparative clinical outcomes of both types of ICD over longer periods. Case series have reported high rates of detection and successful conversion of ventricular tachycardia, and inappropriate shock rates in the range reported for TV-ICD. This evidence is not sufficient to determine whether there are small differences in efficacy between the 2 types of devices, which may be clinically important due to the nature to the disorder being treated. Also, adverse event rate is uncertain, with variable rates reported. At least 1 RCT is currently underway comparing S-ICD with TV-ICD. The evidence is insufficient to determine the effects of the technology on health outcomes.
Supplemental Information

Clinical Input Received From Physician Specialty Societies and Academic Medical Centers
While the various physician specialty societies and academic medical centers may collaborate with and make recommendations during this process through the provision of appropriate reviewers, input received does not represent an endorsement or position statement by the physician specialty societies or academic medical centers, unless otherwise noted.

2015 Input
In response to requests, input was received from 5 academic medical centers and 1 physician specialty society (4 responses), for a total of 9 responses, while this policy was under review in 2015. Input focused on use of ICDs as primary prevention for cardiac ion channelopathies and on use of the S-ICD. Reviewers generally indicated that an ICD should be considered medically necessary for primary prevention of ventricular arrhythmias in both adults and children with a diagnosis of long QT syndrome (LQTS), Brugada syndrome (BrS), short QT syndrome (SQTS), and catecholaminergic polymorphic ventricular tachycardia (CPVT). Reviewers generally indicated that the S-ICD should be considered medically necessary particularly for patients with indications for an ICD but who have difficult vascular access or have had TV-ICD lead explantation due to complications.

2011 Input
In response to requests, input was received from 6 academic medical centers and no physician specialty societies while this policy was under review in 2011. For most policy indications, including pediatric, there was general agreement from those providing input. On the question of timing of ICD implantation, input was mixed, with some commenting about the potential role of early implantation in select patients. Reviewers indicated that a waiting period of 9 months for patients with nonischemic cardiomyopathy was not supported by the available evidence or consistent with the prevailing practice patterns in academic medical centers. Clinical input emphasized the difficulty of prescribing strict timeframes given the uncertainty of establishing the onset of cardiomyopathy and the inability to risk-stratify patients based on time since onset of cardiomyopathy.

Practice Guidelines and Position Statements

American College of Cardiology, American Heart Association, et al
Heart Failure
In 2013, the American College of Cardiology Foundation (ACCF) and American Heart Association (AHA) issued practice guidelines on the management of heart failure. These guidelines made the following recommendations on use of ICD devices as primary prevention:

- For patients with stage B heart failure, “placement of an ICD is reasonable in patients with asymptomatic ischemic cardiomyopathy who are at least 40 days
post-MI, have an LVEF 30%, or less, are on guideline-directed medical therapy (GDMT), and have reasonable expectation of survival with a good functional status for more than 1 year.” (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).
  - ICD therapy is recommended for primary prevention of SCD in selected patients with HFrEF at least 40 d post-MI with LVEF ≤330% and NYHA class I symptoms while receiving GDMT, who are expected to live at least 1 year.” (Class of recommendation: I; level of evidence: B).
  - An ICD “is of uncertain benefit to prolong meaningful survival in patients with a high risk of nonsudden death such as frequent hospitalizations, frailty, or severe comorbidities.” (Class of recommendation: IIb; level of evidence: B).

**Device-Based Therapy for Cardiac Rhythm Abnormalities**

In 2013, ACCF and AHA, 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 for 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. (Level of Evidence: A).
- ICD therapy is indicated in patients with structural heart disease and spontaneous sustained VT, whether hemodynamically stable or unstable. (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. (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. (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. (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. (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. (Level of Evidence: B)

**Class IIa Recommendations**

- ICD implantation is reasonable for patients with unexplained syncope, significant LV dysfunction, and nonischemic DCM. (Level of Evidence: C)
- ICD implantation is reasonable for patients with sustained VT and normal or near-normal ventricular function. (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 [right ventricle] 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 2013 update to the 2008 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*)

**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 not included or poorly represented in ICD clinical trials. The statement made a number of consensus-based guidelines on the use of ICDs in selected patient populations.  

**Hypertrophic Cardiomyopathy**

In 2011, ACCF and AHA guidelines were published on the management of patients with HCM. These guidelines contained the following statements on use of ICDs 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. (*Level of Evidence: C*)
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 and Heart Rhythm Society**

In 2010, AHA issued a scientific statement, endorsed by 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." \( (\text{Level of Evidence: } C) \)

- “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.” \( (\text{Level of Evidence: } C) \)

**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.” \( (\text{Level of Evidence: } C) \)

**Heart Rhythm Society et al**
In 2013, HRS, the European Heart Rhythm Association, and the Asia-Pacific Heart Rhythm Society 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 LQTS, BrS, CPVT, and SQTS.\(^8^4\)

**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 and Heart Rhythm Society**

In 2014, Pediatric and Congenital Electrophysiology Society and HRS issued an expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease (CHD). The statement 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
Not applicable.

Medicare National Coverage
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 preexisting cerebral disease;
- Any disease, other than cardiac disease (eg, 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 an Food and Drug Administration–approved category B Investigational Device Exemption clinical trial (42 CFR §405.201), a trial under the CMS Clinical Trial Policy (National Coverage Determination 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 evidence review.

References


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 (CHRHS), and the International Society for Adult Congenital Heart Disease (ISACHD). Can J Cardiol. Oct 2014;30(10):e1-e63. PMID 25262867

### Billing Coding/Physician Documentation Information

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>33216</td>
<td>Insertion of a transvenous electrode; permanent pacemaker or cardioverter-defibrillator</td>
</tr>
<tr>
<td>33217</td>
<td>Insertion of 2 transvenous electrode; permanent pacemaker or cardioverter-defibrillator</td>
</tr>
<tr>
<td>33218</td>
<td>Repair of single transvenous electrode, permanent pacemaker or pacing cardioverter-defibrillator</td>
</tr>
<tr>
<td>33220</td>
<td>Repair of 2 transvenous electrodes for permanent pacemaker or pacing cardioverter-defibrillator</td>
</tr>
<tr>
<td>33223</td>
<td>Relocation of skin pocket for cardioverter-defibrillator</td>
</tr>
<tr>
<td>33240</td>
<td>Insertion of pacing cardioverter-defibrillator pulse generator only; with existing single lead</td>
</tr>
<tr>
<td>33241</td>
<td>Insertion of pacing cardioverter-defibrillator pulse generator only; with existing dual leads</td>
</tr>
<tr>
<td>33243</td>
<td>Insertion of pacing cardioverter-defibrillator pulse generator only; with existing multiple leads</td>
</tr>
<tr>
<td>33240</td>
<td>Insertion of implantable defibrillator pulse generator only; with existing single lead</td>
</tr>
<tr>
<td>33241</td>
<td>Insertion of implantable defibrillator pulse generator only; with existing dual leads</td>
</tr>
<tr>
<td>33243</td>
<td>Insertion of implantable defibrillator pulse generator only; with existing multiple leads</td>
</tr>
<tr>
<td>93260</td>
<td>Programming device evaluation (in person) with iterative adjustment of the implantable device to test the function of the device and select optimal permanent programmed values with analysis, review and report</td>
</tr>
</tbody>
</table>
by a physician or other qualified health care professional; implantable subcutaneous lead defibrillator system

93261 Interrogation device evaluation (in person) with analysis, review and report by a physician or other qualified health care professional, includes connection, recording and disconnection per patient encounter; implantable subcutaneous lead defibrillator system

93282 Programming device evaluation (in person) with iterative adjustment of the implantable device to test the function of the device and select optimal permanent programmed values with analysis, review and report by a physician or other qualified health care professional; single lead implantable cardioverter-defibrillator system

93283 Programming device evaluation (in person) with iterative adjustment of the implantable device to test the function of the device and select optimal permanent programmed values with analysis, review and report by a physician or other qualified health care professional; dual lead implantable cardioverter-defibrillator system

93284 Programming device evaluation (in person) with iterative adjustment of the implantable device to test the function of the device and select optimal permanent programmed values with analysis, review and report by a physician or other qualified health care professional; multiple lead implantable cardioverter-defibrillator system

93289 Interrogation device evaluation (in person) with analysis, review and report by a physician or other qualified health care professional, includes connection, recording and disconnection per patient encounter; single, dual, or multiple lead implantable cardioverter-defibrillator system, including analysis of heart rhythm derived data elements

93640 Electrophysiologic evaluation of single or dual chamber pacing cardioverter-defibrillator leads including defibrillation threshold evaluation (induction of arrhythmia, evaluation of sensing and pacing for arrhythmia termination) at time of initial implantation or replacement;

93641 Electrophysiologic evaluation of single or dual chamber pacing cardioverter-defibrillator leads including defibrillation threshold evaluation (induction of arrhythmia, evaluation of sensing and pacing for arrhythmia termination) at time of initial implantation or replacement; with testing of single or dual chamber pacing cardioverter-defibrillator pulse generator

93642 Electrophysiologic evaluation of single or dual chamber pacing cardioverter-defibrillator (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing and pacing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters)

93644 Electrophysiologic evaluation of subcutaneous implantable defibrillator (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters)

C1721 Cardioverter-defibrillator, dual chamber (implantable)
C1722 Cardioverter-defibrillator, single chamber (implantable)
C1882 Cardioverter-defibrillator, other than single or dual chamber
Implantable Cardioverter Defibrillator (ICD) 7.01.44

(implantable)

**ICD-10 Codes**
- **I42.1**- Hypertrophic cardiomyopathy code range.
- **I42.2**
- **I45.81** Long QT syndrome
- **I45.89** Other specified conduction disorders
- **I46.2, I46.8, I46.9** Cardiac arrest code range
- **I47.2** Ventricular tachycardia
- **I49.01** Ventricular fibrillation
- **I49.9** Cardiac arrhythmia, unspecified
- **Q20.0-** Congenital malformations of cardiac chambers and connections code range
- **Q20.9**
- **Q21.0-** Congenital malformations of cardiac septa code range
- **Q21.9**
- **Q22.0-** Congenital malformations of pulmonary and tricuspid valves code range
- **Q22.9**
- **Q23.0-** Congenital malformations of aortic and mitral valves code range
- **Q23.9**
- **Q24.0-** Other congenital malformations of heart code range
- **Q24.9**

Deleted Codes: (as of 1/1/2015) 0319T, 0320T, 0321T, 0322T, 0323T, 0324T, 0325T, 0326T, 0327T, 0328T.

**Additional Policy Key Words**

N/A

**Policy Implementation/Update Information**

10/1/88 New policy added to the Surgery section.
6/1/00 No policy statement changes.
6/1/01 Policy archived.
8/1/05 Policy removed from Archives. Policy statement revised to include the following investigational indications:

In patients who:
- have had an acute myocardial infarction (i.e., less than 40 days before AICD 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 cardiac revascularization procedure in past 3 months (coronary artery bypass graft [CABG] or percutaneous transluminal coronary angioplasty [PTCA]) or are candidates for a cardiac revascularization procedure
- have noncardiac disease that would be associated with life expectancy less than 1 year

8/1/06  No policy statement changes.
8/1/07  No policy statement changes.
8/1/08  Policy statement revised to indicate that use of ICD in certain high-risk patients with hypertrophic cardiomyopathy may be considered medically necessary.
8/1/09  No policy statement changes.
8/1/10  No policy statement changes.
8/1/11  Policy statements specific to ICD indications in pediatric patients added to policy statements and rationale. Policy statement revised to clarify the indications in ischemic cardiomyopathy with separate indications for class II/III and class I patients. Policy statement with waiting time in nonischemic cardiomyopathy was revised based on additional clinical input.
1/1/12  Coding updated.
8/1/12  No policy statement changes.
11/1/12  Policy statement added on the use of subcutaneous ICD, considered investigational for all indications. ACCF/AHA guidelines on management of patients with HCM added to policy.
11/1/13  No policy statement changes.
11/1/14  A clause “…after reversible causes (eg, acute ischemia) have been excluded” added to current statement on secondary prevention in adults. Updated CPT definitions and added new 2015 CPT codes.
10/15/15  Subcutaneous ICD may be considered medically necessary.
11/1/15  Added: Diagnosis of cardiac ion channelopathies and considered to be at high risk for sudden cardiac death to primary prevention medically necessary statement. Added: Hypertrophic cardiomyopathy and diagnosis of cardiac ion channelopathies to pediatrics medically necessary statement.
11/1/16  No policy statement changes.

State and Federal mandates and health plan contract language, including specific provisions/exclusions, take precedence over Medical Policy and must be considered first in determining eligibility for coverage. The medical policies contained herein are for informational purposes. The medical policies do not constitute medical advice or medical care. Treating health care providers are independent contractors and are neither employees nor agents Blue KC and are solely responsible for diagnosis, treatment and medical advice. No part of this publication may be reproduced, stored in a retrieval system or transmitted, in any form or by any means, electronic, photocopying, or otherwise, without permission from Blue KC.