Implantable Cardioverter Defibrillator (ICD)

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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:

1) 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.

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

2) Secondary Prevention
   - Patients with a history of a life-threatening clinical event associated with sustained ventricular tachyarrhythmia.

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.
**When Policy Topic is not covered**

The use of a subcutaneous ICD is considered **investigational** for all indications in adult and pediatric patients.

**Adults**
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

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

**Considerations**
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.

Effective in 2013, there are CPT category III codes (0319T-0328T) for subcutaneous implantable defibrillator system reporting (see Code Table).

**Description of Procedure or Service**
The automatic implantable cardioverter defibrillator (ICD) is a device designed to monitor a patient’s heart rate, recognize ventricular fibrillation (VF) or ventricular tachycardia (VT), and deliver an electric shock to terminate these arrhythmias to reduce the risk of sudden death.

Indications for ICD implantation can be broadly subdivided into 1) secondary prevention, i.e., their use in patients who have experienced a potentially life-threatening episode of ventricular tachyarrhythmia (near sudden cardiac death); and 2) primary prevention, i.e., their use in patients who are considered at high risk for sudden cardiac death but who have not yet experienced life-threatening VT or VF.

The standard 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 totally subcutaneous ICD (S-ICD®) has also 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 that is 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 application (PMA) approval process. The FDA-labeled indications generally include patients who have experienced life-threatening ventricular tachyarrhythmia associated with cardiac arrest or ventricular tachyarrhythmia associated with hemodynamic compromise and resistance to pharmacologic treatment. Devices manufactured by Guidant are approved by the FDA for use “in patients at high risk of sudden cardiac death due to ventricular arrhythmias and who have experienced at least 1 of the following: an episode of cardiac arrest (manifested by the loss of consciousness) due to
a ventricular tachyarrhythmia; recurrent, poorly tolerated sustained ventricular tachycardia (VT); or a prior myocardial infarction (MI), left ventricular ejection fraction of less than or equal to 35%, and a documented episode of nonsustained VT, with an inducible ventricular tachyarrhythmia." On July 18, 2002, the FDA expanded the approved indications for the Guidant ICD devices to include the prophylactic use of Guidant ICDs for cardiac patients who have had a previous heart attack and have an ejection fraction that is less than or equal to 30%. This expanded indication is based on the results of the second Multicenter Automatic Defibrillator Implantation Trial (MADIT II trial), which is discussed here. Medtronic devices are approved “to provide ventricular antitachycardia pacing and ventricular defibrillation for automated treatment of life-threatening ventricular arrhythmias.” Other devices have approval language similar to that of Medtronic.

On September 28, 2012, the S-ICD® system by Cameron Health, Inc. was approved by the FDA “to provide defibrillation therapy for the treatment of life-threatening ventricular tachyarrhythmias in patients who do not have symptomatic bradycardia, continual (incessant) ventricular tachycardia, or spontaneous frequently recurring ventricular tachycardia that is reliably terminated with anti-tachycardia pacing.”

NOTE: ICDs may be combined with other pacing devices, such as pacemakers for atrial fibrillation, or biventricular pacemakers designed to treat congestive heart failure. This policy addresses ICDs alone, when used solely to treat patients at risk for ventricular arrhythmias. Policy No. 2.02.10 addresses the use of biventricular pacemakers for the treatment of congestive heart failure alone.

**Rationale**

This policy was created in 1996 and updated periodically with literature review. The most recent update with literature review covers the period of June 2011 through August 2012.

Automatic implantable cardiac defibrillators (ICDs) were first used in survivors of near sudden cardiac death. There has been ongoing interest in using ICDs as primary preventive therapy in patients with risk factors for sudden cardiac death. The first ICD TEC Assessment, published in 2002, addressed this indication. (1) The Assessment 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 prior history of myocardial infarction (MI) and a current history of 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% but also ventricular tachyarrhythmia, as evidenced on an electrophysiologic study. In the subsequent, MADIT II, trial, patients were required to have a lower ejection fraction, less than 30%, but no electrophysiologic study was required. Therefore, the patient selection criteria of the MADIT II trial potentially identify a much larger number of candidates for ICD implantation.

The 2002 TEC Assessment offered the following observations and conclusions:

- 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 October 2004, TEC reassessed ICDs. (2) The 2004 TEC Assessment focused on the results of the 5 randomized clinical 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, and the Cardiomyopathy Trial [CAT]) and 5 additional RCTs:

1. Defibrillator in Acute Myocardial Infarction Trial (DINAMIT);
2. Sudden Cardiac Death in Heart Failure Trial (SCD-HeFT)
3. Comparison of Medical Therapy, Pacing, and Defibrillation in Heart Failure (COMPANION);
4. Defibrillators in Non-Ischemic Cardiomyopathy Treatment Evaluation (DEFINITE); and
5. Amiodarone versus Implantable Defibrillator Randomized Trial (AMIOVIRT).

The 2004 TEC Assessment made the following observations.
Patients Who Have Prior MI and Reduced LVEF. The previous 2002 Assessment concluded that the evidence was sufficient to demonstrate that ICD therapy improves net health outcome in patients with prior MI and reduced LVEF. Both new studies (SCD-HeFT and COMPANION) and the re-analysis of MUSTT findings provide 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 is close to that observed in MADIT II (HR: 0.69, 95% CI: 0.51 to 0.93), and these findings provide additional supportive evidence that ICD therapy reduces mortality. There may be slight but not statistically significantly increased rates of adverse effects associated with ICD therapy; however, serious device-related events are not common. On balance, the significant reductions in mortality associated with ICD therapy outweigh the harms associated with ICD therapy in comparison to conventional treatment. Thus, the available evidence again demonstrates 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 available evidence was insufficient to permit conclusions regarding the effect of ICD therapy on net health outcome for this indication.

Patients Who Have No Prior MI and Reduced LVEF (e.g., Nonischemic Dilated Cardiomyopathy, NIDCM). Results from subjects with NIDCM included in SCD-HeFT and DEFINITE suggest a mortality benefit from ICD therapy, although statistical significance that was not achieved in these studies was likely related to insufficient power. A meta-analysis (3) of 5 trials including nonischemic subjects reports 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 together, the preponderance of evidence suggests 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 adverse events is not well-reported in studies of patients without prior MI, it seems reasonable to expect similar low rates of device-related adverse events as seen in studies of patients with prior MI.

Device-Related Adverse Effects. Device-related adverse effects were inconsistently reported in the available trials, although serious adverse events appear to be uncommon. What is known about device-related adverse effects does not outweigh the significant mortality benefits demonstrated in various studies.

Therefore, the 2004 TEC Assessment made the following conclusions: ICD placement has been performed and investigated in multiple centers throughout the United States, and when performed by similarly experienced personnel, it is reasonable to expect that the improvements observed in the investigational setting will be attainable outside the investigational settings.

Therefore, the use of ICD devices meets the TEC criteria in the prevention of sudden death from ventricular tachyarrhythmia in patients who have:

- Symptomatic* ischemic dilated cardiomyopathy with a history of MI at least 40 days before ICD treatment and LVEF of 35% or less; or
- Symptomatic* 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 (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; or
  - have noncardiac disease that would be associated with life expectancy less than 1 year.
Symptomatic heart failure is defined as the presence of dyspnea on exertion, angina, palpitations, or fatigue.

Further analysis of existing trial data using patient-level meta-analysis may further delineate which subgroups of patients are likely to benefit from ICD placement and those unlikely to benefit who can be spared the morbidity of ICD placement.

Subsequent Evidence and Guidelines
Relevant evidence and most current guidelines identified through Medline published following the 2004 TEC Assessment through October 2009 include the following:
- Reports of BEST-ICD [Beta-blocker Strategy + ICD], and IRIS trials
- Implantation timing placing in NIDCM
- ICD implantation in Hypertrophic Cardiomyopathy
- ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities
- ACC/AHA 2005 Guideline Update for the Diagnosis and Management of Chronic Heart Failure in the Adult

BEST-ICD trial. The BEST-ICD (Beta-blocker Strategy + ICD) trial randomized 143 patients 5–30 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. (4) Entry criteria included an LVEF less than or equal to 35% along with 1 or more non-invasive risk factors (e.g., premature ventricular contractions, heart rate variability, signal-averaged electrocardiography [SAECG]-positive) and be given maximal tolerated beta-blocker (metoprolol) therapy. The authors concluded that using electrophysiology studies to guide ICD placement within 5–30 days after MI did not significantly improve outcomes and survival. This is consistent with the conclusions that ICD placement after early MI does not improve outcomes. The authors also noted that the study 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 less than 35% than expected based on experience reported in the literature.

IRIS trial. The Immediate Risk Stratification Improves Survival (IRIS) trial evaluated ICD implantation early after MI. (5) Eligible patients were required to have an LVEF 40% or less and either: 1) a heart rate 90 or more beats per minute on initial electrocardiogram (ECG), or 2) nonsustained ventricular tachycardia during Holter monitoring, or both. From 92 centers and 62,944 patients post-MI, 898 were randomized 5 to 31 days following 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 two arms (ICD vs. medical therapy, HR 1.04; 95% CI: 0.81 to 1.35). However, the risk of sudden cardiac death was lower following ICD (HR 0.52; 95% CI: 0.35 to 0.78), but non-sudden cardiac death risk was greater (HR 1.8; 95% CI: 1.0 to 3.2). These results are consistent with guidelines and previous trials.

High-Risk Hypertrophic Cardiomyopathy (HCM). Maron and colleagues (6) reported appropriate ICD discharge rates (terminating either ventricular tachycardia or fibrillation) from an international registry of 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. The mean age of patients was 42 years (standard deviation [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 considered in the study placing patients at 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) one or more runs of nonsustained ventricular tachycardia 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 be 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 nonischemic dilated cardiomyopathy (NIDCM), Ellenbogen and colleagues (7) concluded that approximately one half of arrhythmias terminated by appropriate ICD discharges are not life-threatening. The NNTBs calculated, therefore, represent lower bounds or greatest potential benefit, and the true benefit is likely less (only 6.3% of primary prevention patients had more than one appropriate discharge). Adverse events rates included one or more inappropriate discharges (27%); infections (3.8%); hemorrhage or thrombosis (1.6%); 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.

Al-Khatib and Curtis (8) published an analysis of whether ICD implantations in the U.S. followed evidence-based guidelines using a Medicare ICD registry. There were a total of 111,707 patients who received an ICD between January 2006 and June 2009. Of these, 25,145 (22.5%) did not meet the evidence-based criteria according to ACC/AHA/HRS guidelines. (9) Patients who did not meet evidence-based ICD criteria had a higher mortality than patients who did meet criteria (0.57% vs. 0.18%, respectively; p<0.001) and also had a higher rate of procedural complications (3.2 vs. 2.4%, respectively; p<0.001). Electrophysiologists had a lower rate of non-evidence-based ICD use compared to non-electrophysiologists (20.8% vs. 24.8%, respectively; p<0.001).

**Implantation Timing in Nonischemic Dilated Cardiomyopathy.** For patients with nonischemic cardiomyopathy, the optimal timing of ICD implantation remains uncertain. A substantial percent of patients diagnosed with nonischemic cardiomyopathy (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 prior to ICD implantation arises from the selection criteria of the CAT trial, (10) 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 and Subacius (10) performed a post-hoc analysis of the DEFINITE trial data to examine whether the time from diagnosis of nonischemic dilated cardiomyopathy (NIDCM) was associated with the magnitude of benefit from ICD implantation. Survival benefit was found only for those diagnosed less than 9 months prior to implantation (n=216); no benefit was apparent when NIDCM was diagnosed greater than 9 months prior (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 had 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) Cardiomyopathy (CAT) trial (11) reviewed in the 2002 TEC Assessment. Further evidence is necessary to define when in the natural history of the disease ICD implantation is appropriate.

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 for less than 9 months. Neither of these pieces of evidence represents strong data to support a specific time interval prior to implanting an ICD in patients with NICM.

Zecchin et al. (12) performed a cohort study on 503 consecutive patients diagnosed with idiopathic NICM to determine the extent to which indications for an ICD evolved over the several months following an initial NICM diagnosis. At initial diagnosis, 245 met Sudden Cardiac Death in Heart Failure Trial
(SCD-HeFT) criteria for an ICD, based on an ejection fraction less than 35% and Class II-III heart failure, and 258 did not meet criteria for an ICD. At a mean follow-up of 5.4 months during which patients were treated with angiotensin-converting enzyme inhibitors and beta blockers, there were consistent improvements in ejection fraction and symptoms, such that less than one-third of evaluable patients (31%) 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 prior to optimal treatment and stabilization of patients with newly diagnosed NICM, since the indications for ICD are not stable over time and will change in a substantial numbers of patients following treatment.

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. The most recent ACC/AHA guidelines (9, 13) do not specifically address the optimal waiting period prior to implantation of an ICD for patients with newly diagnosed NICM.

**Adverse Events**

Ricci et al. (14) 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 of the lead failures (87.5%) were due to lead fracture. The median time until recognition of lead failure, or until an adverse event, was 2.2 days. A total of 22 patients (5.3%) received an inappropriate shock due to lead failure.

Cheng et al. (15) 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 2,628 (1.2%). Factors associated with lead dislodgement were NYHA Class IV heart failure, atrial fibrillation/flutter, a combined ICD-CRT device, and having the procedure performed by a non-electrophysiologist. Lead dislodgement was associated with an increased risk for other cardiac adverse events and death.

Lee et al. (16) evaluated the rate of early complications among patients enrolled in a prospective, multicenter population-based registry of all newly implanted ICDs in Ontario, Canada from February 2007 through May 2009. Of 3,340 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 (cardiac resynchronization therapy) 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).

Several publications have reported on infection rates in patients receiving an ICD. Smit and Schonheyder (17) 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 post-surgical infections. Nery and Nair (18) reported the rate of ICD-associated infections among consecutive patients treated with an ICD at a tertiary referral center. There were a total of 24 infections among 2,417 patients for a rate of 1.0%. Twenty-two of 24 patients with infections (91.7%) required device replacement. Factors associated with infection were device replacement (versus de novo implantation) and use of a complex device (e.g., combined ICD-CRT or dual/triple chamber devices). Sohail et al. (19) performed a case-control study evaluating the risk factors for infection in 68 patients with an ICD infection and 136 matched controls. 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, prolonged hospitalization for >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.

**Use of AICD (Automatic ICD) in the Pediatric Population**

There is limited direct scientific evidence on the efficacy of ICDs in the pediatric population. The majority of published studies in this area are retrospective analyses of small case series. A review of some of the representative publications of this type is summarized below.

The largest published series was a combined series of pediatric patients and patients with congenital heart disease from 4 clinical centers. (20) The median age of this population was 16 years, although some adults were included up to the age of 54 years. A total of 443 patients were included. The most common diagnoses were tetralogy of Fallot and hypertrophic cardiomyopathy. ICD implantation was performed for primary prevention in 52% of patients and for secondary prevention in 48%. Over a 2-year period of follow-up, appropriate shocks occurred in 26% of patients and inappropriate shocks occurred in 21%.

Silka et al. (21) 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 patients (76%), drug-refractory ventricular tachycardia in 13 patients (10%), and syncope with heart disease plus inducible ventricular tachycardia in 13 patients (10%). During a mean follow-up of 31 +/- 23 months, 73 patients (59%) received at least one appropriate shock and 25 patients (20%) received at least one inappropriate shock. The actuarial rates of sudden-death-free survival were 97% at one year, 95% at 2 years, and 90% at 5 years.

Alexander et al. (22) reported on 90 ICD procedures in 76 young patients with a mean age of 16 years (range: 1–30). Indications for placement were 27 patients (36%) with cardiac arrest or sustained ventricular tachycardia, 40 patients (53%) with syncope, 17 patients (22%) with palpitations, 40 patients (53%) with spontaneous ventricular arrhythmias, and 36 patients (47%) with inducible ventricular tachycardia. Numerous patients had more than one indication for ICD in this study. Over a median of 2 years’ follow-up, 28% of patients received an appropriate shock, and 25% of patients received an inappropriate shock. Lewandowski et al. (23) reported on long-term follow-up of 63 patients between the ages of 6-21 years who were treated with an ICD device. After a 10-year follow-up, there were 13 (21%) patients with surgical infections. Fourteen patients (22%) experienced at least one appropriate shock and 17 patients (27%) had at least one inappropriate shock. Serious psychological sequelae developed in 27 patients (43%).

**Subcutaneous ICD**

The first study on outcomes of an entirely subcutaneous ICD was published in 2010. (24) This study described the development and testing of the device, including empiric evidence for the optimal placement of the subcutaneous electrode. In addition, 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 ventricular fibrillation was induced in 53/55. All episodes were correctly detected by the subcutaneous ICD. In 52/53 patients, 2 consecutive episodes of ventricular arrhythmia were successfully terminated. In the final patient, the arrhythmia was terminated on one occasion but not on the other. In the cohort portion of this study, 54/55 patients were alive at last follow-up. The one death was due to renal failure, and this patient requested removal of the subcutaneous ICD prior to death. An infection at the generator site occurred in 2 patients, necessitating a revision procedure. Another 3 patients had lead dislodgement requiring repositioning. There were a total of 12 episodes of ventricular tachycardia that were detected by the subcutaneous ICD; all 12 episodes were successfully terminated by countershock.

The Subcutaneous versus Transvenous Arrhythmia Recognition Testing (START) study compared the performance of a subcutaneous ICD with a transvenous ICD for detecting arrhythmias in the
electrophysiology lab. (25) The patient population included 64 patients who were scheduled for ICD implantation. All patients had a transvenous ICD placed, as well as subcutaneous electrodes attached to a subcutaneous ICD. Arrhythmias were induced and the sensitivity and specificity of detection by each device was compared. For ventricular arrhythmias, sensitivity of detection was 100% for the subcutaneous ICD and 99% for the transvenous ICD. Specificity was 98.0% for the subcutaneous ICD device compared to 76.7% for the transvenous device (p<0.001).

Clinical Input Received through Physician Specialty Societies and Academic Medical Centers
In response to requests, input was received from no physician specialty societies and 6 academic medical centers while this policy was under review in 2011. 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.

For most policy indications, including pediatric indications, there was 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 selected 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. Specialty society 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.

Summary
There is an extensive literature base 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 demonstrated a benefit in overall mortality for patients with ischemic cardiomyopathy and reduced ejection fraction. The indications for ICDs in these groups of patients parallel the inclusion criteria for the major trials and the recommendations from major specialty society guidelines. RCTs of early ICD implantation following acute MI do not support a benefit for immediate ICD implantation versus delayed implantation for at least 40 days.

For NICM, there is less clinical trial evidence available, 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 not high-quality evidence available 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 one cohort study reports that the majority of patients who meet criteria for an ICD at the time of initial NICM diagnosis will no longer meet the criteria for an ICD several months after initiation of treatment.

For pediatric patients, there is no direct evidence on the benefit of ICD implantation from high-quality clinical trials. Indications for pediatric patients are based on specialty society guidelines and from specialty society clinical input, both of which extrapolate findings from adult populations to the pediatric population.

A subcutaneous ICD (S-ICD®) has been developed that does not employ transvenous leads. A small amount of literature has been published on the subcutaneous ICD, with results so far indicating that the subcutaneous ICD may approximate the performance of a transvenous ICD. Due to the limited evidence, the subcutaneous ICD is considered investigational.

Clinical Practice Guidelines and Position Statements
The ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities updates the 2002 Guideline for Implantation of Cardiac Pacemakers and Antiarrhythmic Devices. Guideline recommendations are classified into three levels: Classes I, II, and III. Class I is defined as “conditions for which there is evidence for and/or general agreement that the procedure or treatment is useful and effective.” Only Class I recommendations are listed here. Each recommendation is further classified as either A, B, or C, based on the weight of the evidence available. Level A is applied when data are from multiple, randomized clinical trials; level B is when data are from a limited number of randomized trials; and level C is when the recommendation is primarily based on expert consensus. The 2008 guidelines of the ACC/AHA/HRS for implantation of cardiac pacemakers and antiarrhythmia devices include the following Class I indications for ICDs:

1. 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)
2. ICD therapy is indicated in patients with structural heart disease and spontaneous sustained VT, whether hemodynamically stable or unstable. (Level of Evidence: B)
3. ICD therapy is indicated in patients with syncpe of undetermined origin with clinically relevant, hemodynamically significant sustained VT or VF induced at electrophysiological study. (Level of Evidence: B)
4. ICD therapy is indicated in patients with left ventricular ejection fraction (LVEF) less than 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)
5. ICD therapy is indicated in patients with NIDCM who have an LVEF less than or equal to 35% and who are in NYHA functional Class II or III. (Level of Evidence: B)
6. 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 30%, and are in NYHA functional Class I. (Level of Evidence: A)
7. ICD therapy is indicated in patients with nonsustained VT due to prior MI, LVEF less than 40%, and inducible VF or sustained VT at electrophysiological study. (Level of Evidence: B)

Notable is revision of LVEF from less than 40% to less than 35% for use as criteria for primary prevention—consistent with both clinical trial entry criteria and this policy.

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

**Class I recommendations**
- The decision to place an ICD in patients with HCM should include application of individual clinical judgment, as well as a thorough discussion of the strength of evidence, benefits, and risks to allow the informed patient’s active participation in decision making. (Level of Evidence: C)
- ICD placement is recommended for patients with HCM with:
  - 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 [non-sustained VT] (particularly those <30 years of age) in the presence of other SCD risk factors or modifiers. (Level of Evidence: C)
- An ICD can be useful in select patients with HCM with an abnormal blood pressure response with exercise in the presence of other SCD risk factors or modifiers. (Level of Evidence: C)
- It is reasonable to recommend an ICD for high-risk children with HCM, based on unexplained syncope, massive LV hypertrophy, or family history of SCD, after taking into account the relatively high complication rate of long-term ICD implantation. (Level of Evidence: C)
Class IIb recommendations
- The usefulness of an ICD is uncertain in patients with HCM with isolated bursts of NSVT when in the absence of any other SCD risk factors or modifiers. (Level of Evidence: C)
- The usefulness of an ICD is uncertain in patients with HCM with an abnormal blood pressure response with exercise when in the absence of any other SCD risk factors or modifiers, particularly in the presence of significant outflow obstruction. (Level of Evidence: C)

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

In April 2009, the ACC/AHA published updated guidelines on the management of chronic heart failure in collaboration with the American College of Chest Physicians and the International Society for Heart and Lung Transplantation. (13) The guidelines follow the evidence criteria listed here for ICD placement, and only Class I recommendations are listed as follows:

1. An ICD is recommended as secondary prevention to prolong survival in patients with current or prior symptoms of HF [heart failure] and reduced LVEF who have a history of cardiac arrest, ventricular fibrillation, or hemodynamically destabilizing ventricular tachycardia. (Level of Evidence: A)
2. ICD therapy is recommended for primary prevention of sudden cardiac death to reduce total mortality in patients with nonischemic dilated cardiomyopathy or ischemic heart disease at least 40 days post-MI, an LVEF less than or equal to 35%, and NYHA functional Class II or III symptoms while receiving chronic optimal medical therapy, and who have reasonable expectation of survival with a good functional status for more than 1 year. (Level of Evidence: A)

Guidelines concerning ICD use in pediatric populations have been published. These are derived from nonrandomized studies, extrapolation from adult clinical trials, and expert consensus. (9) The ACC/AHA/HRS published the following indications for ICD use in pediatric patients in 2008 (9):

Class I indications
- Survivors of cardiac arrest, after reversible causes have been excluded (Level of evidence B)
- Symptomatic, sustained ventricular tachycardia in association with congenital heart disease in patients who have undergone hemodynamic and electrophysiologic evaluation (Level of evidence C)

Class IIa indications
- Reasonable for patients with congenital heart disease with recurrent syncope of undetermined origin in the presence of either ventricular dysfunction or inducible ventricular arrhythmias (Level of evidence B)

Class IIb indications
- May be considered for patients with recurrent syncope associated with complex congenital heart disease and advanced systemic ventricular dysfunction when thorough invasive and non-invasive investigations have failed to reveal a cause (Level of evidence C)

Medicare National Coverage Policy
In January 2005, Medicare issued the following revised national coverage guideline for the use of ICDs. (27)

The Centers for Medicare and Medicaid Services (CMS) determined that the evidence is adequate to conclude that an ICD is reasonable and necessary for the following:
- Patients with ischemic dilated cardiomyopathy (IDCM), documented prior MI, NYHA Class II and III heart failure, and measured LVEF of 35% or less;
- Patients with nonischemic dilated cardiomyopathy (NIDCM) >9 months, NYHA Class II and III heart failure, and measured LVEF of 35% or less;
- Patients who meet all current CMS coverage requirements for a cardiac resynchronization therapy (CRT) device and have NYHA Class IV heart failure;
- For each of these groups, patients must not have:
  - Cardiogenic shock or symptomatic hypotension while in a stable baseline rhythm;
  - Had a coronary artery bypass graft (CABG) or PTCA within the past 3 months;
  - Had an acute MI within the past 40 days;
  - Clinical symptoms or findings that would make them a candidate for coronary revascularization;
  - Irreversible brain damage from pre-existing cerebral disease;
  - Any disease, other than cardiac disease (e.g., cancer, uremia, liver failure), associated with a likelihood of survival less than 1 year;

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

The Medicare policy for ischemic and nonischemic dilated cardiomyopathy is consistent with this policy.

References
1. Blue Cross and Blue Shield Association Technology Evaluation Center (TEC). Use of implantable cardioverter-defibrillators for prevention of sudden death in patients at high risk for ventricular arrhythmia. TEC Assessments 2002; 17(Tab 10).
27. CMS. Medicare Policy. Available online at: https://www.cms.hhs.gov/mcd/viewdecisionmemo.asp?id=148

**Billing Coding/Physician Documentation Information**

33216 Insertion of a transvenous electrode; single chamber (1 electrode) permanent pacemaker or single chamber pacing cardioverter-defibrillator
33217 Insertion of a transvenous electrode; dual chamber (2 electrodes) permanent pacemaker or dual chamber pacing cardioverter-defibrillator
33218 Repair of single transvenous electrode for a single chamber, permanent pacemaker or single chamber pacing cardioverter-defibrillator
33220 Repair of 2 transvenous electrodes for a dual chamber permanent pacemaker or dual
chamber pacing cardioverter-defibrillator

33223 Revision of skin pocket for cardioverter-defibrillator

33240 Insertion of pacing cardioverter-defibrillator pulse generator only; with existing single lead

33230 Insertion of pacing cardioverter-defibrillator pulse generator only; with existing dual lead

33231 Insertion of pacing cardioverter-defibrillator pulse generator only; with existing multiple leads

33241 Subcutaneous removal of single or dual chamber pacing cardioverter-defibrillator pulse generator

33262 Removal of pacing cardioverter-defibrillator pulse generator with replacement of pacing cardioverter-defibrillator pulse generator; single lead system

33263 Removal of pacing cardioverter-defibrillator pulse generator with replacement of pacing cardioverter-defibrillator pulse generator; dual lead system

33264 Removal of pacing cardioverter-defibrillator pulse generator with replacement of pacing cardioverter-defibrillator pulse generator; multiple lead system

33243 Removal of single or dual chamber pacing cardioverter-defibrillator electrode(s); by thoracotomy

33244 Removal of single or dual chamber pacing cardioverter-defibrillator electrode(s); by transvenous extraction

33249 Insertion or replacement of permanent pacing cardioverter-defibrillator system with transvenous lead(s), single or dual chamber

0319T Insertion or replacement of subcutaneous implantable defibrillator system with subcutaneous electrode

0320T Insertion of subcutaneous defibrillator electrode

0321T Insertion of subcutaneous implantable defibrillator pulse generator only with existing subcutaneous electrode

0323T Removal of subcutaneous implantable defibrillator pulse generator only

0323T Removal of subcutaneous implantable defibrillator pulse generator with replacement of subcutaneous implantable defibrillator pulse generator only

0324T Removal of subcutaneous defibrillator electrode

0325T Repositioning of subcutaneous implantable defibrillator electrode and/or pulse generator

0326T 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)

0327T Interrogation device evaluation (in person) with analysis, review and report, includes connection, recording and disconnection per patient encounter; implantable subcutaneous lead defibrillator system

0328T 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; implantable subcutaneous lead defibrillator system

C1721 Cardioverter-defibrillator, dual chamber (implantable)

C1722 Cardioverter-defibrillator, single chamber (implantable)

C1882 Cardioverter-defibrillator, other than single or dual chamber (implantable)

G0448 Insertion or replacement of a permanent pacing cardioverter-defibrillator system with transvenous lead(s), single or dual chamber with insertion of pacing electrode, cardiac venous system, for left ventricular pacing

**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.

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.