

| | | | |
|------------------------------|--|------------------------|--------------|
| 7.01.44 | Implantable Cardioverter Defibrillators | | |
| Original Policy Date: | September 1, 1986 | Effective Date: | July 1, 2023 |
| Section: | 7.0 Surgery | Page: | Page 1 of 59 |

Policy Statement

- I. The use of the automatic implantable cardioverter defibrillator (ICD) may be considered **medically necessary** for primary or secondary prevention for individuals who meets **all** of the following criteria:
 - A. The individual has NOT had an acute myocardial infarction within the past 40 days
 - B. The individual does NOT have New York Heart Association ([NYHA](#)) class IV congestive heart failure
 - C. The individual has NOT had a cardiac revascularization procedure within the past 3 months
 - D. The individual is NOT a candidate for a cardiac revascularization procedure
 - E. The individual does NOT have a life expectancy of less than 1 year
 - F. Reversible causes (e.g., acute ischemia) are NOT present or have been maximally treated
 - G. The use of the implantable cardioverter defibrillator (ICD) is indicated for **one or more** of the following:
 1. Ischemic cardiomyopathy with New York Heart Association ([NYHA](#)) functional class II or III symptoms and left ventricular ejection fraction (LVEF) of 35% or less
 2. Ischemic cardiomyopathy, [NYHA](#) functional class I symptoms and LVEF of 30% or less
 3. Nonischemic dilated cardiomyopathy and **all** of the following:
 - a. LVEF of 35% or less
 - b. Response to [optimal medical therapy](#) has been adequately determined
 4. Hypertrophic cardiomyopathy (HCM) with **both** of the following:
 - a. Judged to be at high risk for sudden cardiac death by a provider experienced in the care of individuals with HCM
 - b. Major risk factors for sudden cardiac death as indicated by **any** of the following:
 - i. History of premature HCM-related sudden death in one or more [first-degree relatives](#) younger than 50 years
 - ii. Left ventricular hypertrophy (left ventricular wall thickness greater than 30 mm)
 - iii. One or more runs of non-sustained ventricular tachycardia at heart rates of 120 beats per minute or greater
 - iv. Prior unexplained syncope inconsistent with neurocardiogenic origin
 5. Considered to be at high risk for sudden cardiac death and diagnosis of [cardiac ion channelopathy](#) as indicated by **any** of the following:
 - a. Congenital Long QT Syndrome (LQTS)
 - b. Brugada Syndrome (BrS)
 - c. Short QT Syndrome (SQTS)
 - d. Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)
 6. Diagnosis of cardiac sarcoid and considered to be at high risk for sudden cardiac death (see Policy Guidelines section)
 7. Individual with a history of a life-threatening clinical event associated with ventricular arrhythmic events such as sustained ventricular tachyarrhythmia or survival of cardiac arrest
 8. Symptomatic, sustained ventricular tachycardia in association with congenital heart disease after hemodynamic and electrophysiologic evaluation
 9. Congenital heart disease with recurrent syncope of undetermined origin in the presence of ventricular dysfunction or inducible ventricular arrhythmias

- II. The use of the ICD is considered **investigational** in primary or secondary prevention individuals who have had **any** of the following risk factors or do not meet the criteria for approval:
 - A. History of an acute myocardial infarction (i.e., less than 40 days before ICD treatment)
 - B. New York Heart Association (NYHA) class IV congestive heart failure (unless individual is eligible to receive a combination cardiac resynchronization therapy ICD device)
 - C. History of a cardiac revascularization procedure in past 3 months (coronary artery bypass graft [CABG] or percutaneous transluminal coronary angioplasty [PTCA])
 - D. Candidates for a cardiac revascularization procedure
 - E. Have noncardiac disease that would be associated with life expectancy less than 1 year

Subcutaneous Implantable Cardioverter Defibrillator

- III. The use of a subcutaneous ICD may be considered **medically necessary** for adult or pediatric individuals who meets criteria for ICD implantation for primary or secondary prevention for any of the above reasons and meet all of the following criteria:
 - A. Have a contraindication to a transvenous ICD due to **any** of the following:
 - 1. Lack of adequate vascular access
 - 2. Compelling reason to preserve existing vascular access (i.e., need for chronic dialysis; younger individual with anticipated long-term need for ICD therapy)
 - 3. History of need for explantation of a transvenous ICD due to a complication, with ongoing need for ICD therapy
 - B. Have no indication for antibradycardia pacing
 - C. No known or anticipated ventricular arrhythmias known or anticipated to respond to antitachycardia pacing
- IV. The use of a subcutaneous ICD is considered **investigational** for individuals who do not meet the criteria outlined above.

NOTE: Refer to [Appendix A](#) to see the policy statement changes (if any) from the previous version.

Policy Guidelines

This evidence review addresses the use of implantable cardioverter defibrillator (ICD) devices as stand-alone interventions, not as combination devices to treat heart failure (i.e., cardiac resynchronization devices) or in combination with pacemakers. Unless specified, the policy statements and rationale refer to transvenous ICDs.

Pediatric Indications

Indications for pediatric ICD use are based on American College of Cardiology (ACC), American Heart Association (AHA), and Heart Rhythm Society (HRS) guidelines published in 2008 (updated in 2012), which acknowledged the lack of primary research on pediatric individuals in this field (see Rationale section). These indications derive from nonrandomized studies, extrapolation from adult clinical trials, and expert consensus.

Criteria for Implantable Cardioverter Defibrillator Implantation in Individuals 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 placement in individuals with cardiac ion channelopathies derive 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 individuals with

inherited primary arrhythmia syndromes, and a report from the HRS and EHRA's Second Consensus Conference on Brugada syndrome.

Indications for consideration for ICD placement for each cardiac ion channelopathy are as follows:

- Long QT syndrome (LQTS):
 - Individuals with a diagnosis of LQTS who are survivors of cardiac arrest
 - Individuals with a diagnosis of LQTS who experience recurrent syncopal events while on β -blocker therapy.
- Brugada syndrome (BrS):
 - Individuals with a diagnosis of BrS who are survivors of cardiac arrest
 - Individuals with a diagnosis of BrS who have documented spontaneous sustained ventricular tachycardia (VT) with or without syncope
 - Individuals with a spontaneous diagnostic type 1 electrocardiogram (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)
 - Individuals with a diagnosis of BrS who develop ventricular fibrillation during programmed electrical stimulation.
- Catecholaminergic polymorphic ventricular tachycardia (CPVT):
 - Individuals with a diagnosis of CPVT who are survivors of cardiac arrest
 - Individuals with a diagnosis of CPVT who experience recurrent syncope or polymorphic/bidirectional VT despite optimal medical management, and/or left cardiac sympathetic denervation.
- Short QT syndrome (SQTS):
 - Individuals with a diagnosis of SQTS who are survivors of cardiac arrest
 - Individuals with a diagnosis of SQTS who are symptomatic and have documented spontaneous VT with or without syncope
 - Individuals with a diagnosis of SQTS who are asymptomatic or symptomatic and have a family history of sudden cardiac death.

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

Criteria for Implantable Cardioverter Defibrillator Implantation in Individuals With Cardiac Sarcoid

Criteria for ICD placement in individuals with cardiac sarcoid derive from a 2014 consensus statement from the HRS and 2017 joint guidelines from the AHA, ACC, and HRS.

Indications for consideration of ICD placement in individuals diagnosed with cardiac sarcoid are as follows:

- Spontaneous sustained ventricular arrhythmias, including prior cardiac arrest, if meaningful survival of greater than 1 year is expected;
- Left ventricular ejection fraction (LVEF) 35% or less, despite optimal medical therapy and a period of immunosuppression (if there is active inflammation), if meaningful survival of greater than 1 year is expected;
- LVEF greater than 35%, if meaningful survival of greater than 1 year is expected;
 - Syncope or near-syncope, felt to be arrhythmic in etiology

- Evidence of myocardial scar by cardiac magnetic resonance imaging (MRI) or positron emission tomographic (PET) scan
- Inducible sustained ventricular arrhythmias (>30 seconds of monomorphic VT or polymorphic VT) or clinically relevant ventricular fibrillation (VF)
- An indication for permanent pacemaker implantation.

Table PG1. New York Heart Association (NYHA) Functional Classification

| Functional Capacity | Objective Assessment |
|--|--|
| Class I. Patients with cardiac disease but without resulting limitation of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain. | A. No objective evidence of cardiovascular disease. |
| Class II. Patients with cardiac disease resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity results in fatigue, palpitation, dyspnea, or anginal pain. | B. Objective evidence of minimal cardiovascular disease. |
| Class III. Patients with cardiac disease resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes fatigue, palpitation, dyspnea, or anginal pain. | C. Objective evidence of moderately severe cardiovascular disease. |
| Class IV. Patients with cardiac disease resulting in inability to carry on any physical activity without discomfort. Symptoms of heart failure or the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort is increased. | D. Objective evidence of severe cardiovascular disease. |

First-Degree Relatives

A first-degree relative is a family member who shares about 50% of their genes with a particular individual in a family. First degree relatives include parents, offspring, and siblings.

Ischemic Cardiomyopathy and Non-ischemic Cardiomyopathy

Ischemic cardiomyopathy (ICM) is defined as left ventricular systolic dysfunction (or disease of the heart muscle) associated with at least 75% stenosis (narrowing) of at least one of the three major coronary arteries, *or* a documented history of myocardial infarction.

Non-ischemic cardiomyopathy (NICM) is defined as left ventricular systolic dysfunction (or disease of the heart muscle) that is not associated with coronary artery disease (CAD) or narrowing of the coronary arteries.

Optimal Medical Therapy

Optimal medical therapy typically includes beta blockers (i.e., carvedilol), angiotensin-converting-enzyme (ACE) inhibitors, angiotensin receptor-blocking (ARB) agents, lipid-lowering agents, fibrinolytic and anti-thrombotic agents, diuretics and spironolactone in therapeutic doses that cause the individual to achieve a blood pressure of less than 120 mmHg systolic.

Primary vs. Secondary Prevention

Primary prevention includes those measures that try to prevent a problem before it occurs.

Secondary prevention includes those measures that are taken early once a problem has already happened to try to prevent it from happening again.

Coding

The following CPT codes describe procedures associated with ICD implants. Depending on the type of ICD implanted, one or a combination of the following codes may be appropriate:

Physician procedure codes: 33216-33217, 33230-33231, 33240-33241, 33249, 33262-33264.

The following CPT code has been **revised**:

- **0577T**: Electrophysiologic evaluation of implantable cardioverter-defibrillator system with substernal electrode (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters)

The following new CPT PLA codes* represent a new procedure for complex tunneling and placement of cardioverter defibrillator leads sub-sternally versus subcutaneous or transvenous placement:

- **0571T**: Insertion or replacement of implantable cardioverter-defibrillator system with substernal electrode(s), including all imaging guidance and electrophysiological evaluation (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters), when performed
- **0572T**: Insertion of substernal implantable defibrillator electrode
- **0573T**: Removal of substernal implantable defibrillator electrode
- **0574T**: Repositioning of previously implanted substernal implantable defibrillator-pacing electrode
- **0575T**: Programming device evaluation (in person) of implantable cardioverter-defibrillator system with substernal electrode, 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
- **0576T**: Interrogation device evaluation (in person) of implantable cardioverter-defibrillator system with substernal electrode, with analysis, review and report by a physician or other qualified health care professional, includes connection, recording and disconnection per patient encounter
- **0578T**: Interrogation device evaluation(s) (remote), up to 90 days, substernal lead implantable cardioverter-defibrillator system with interim analysis, review(s) and report(s) by a physician or other qualified health care professional
- **0579T**: Interrogation device evaluation(s) (remote), up to 90 days, substernal lead implantable cardioverter-defibrillator system, remote data acquisition(s), receipt of transmissions and technician review, technical support and distribution of results
- **0580T**: Removal of substernal implantable defibrillator pulse generator only

***Note:** All imaging guidance (e.g., fluoroscopy) required to complete the substernal implantable defibrillator procedure is included. The extravascular substernal defibrillator-pacing lead is used in patients who have a fragile vasculature which preclude transvenous insertion.

The following category III CPT code is considered an add-on code with code set 0571T-0577T and 0580T:

- **0614T**: Removal and replacement of substernal implantable defibrillator pulse generator

The following HCPCS C-codes relate to the implantation of an ICD. Depending on the type of ICD implanted and the specific components, one or a combination of the following codes may be appropriate: C1721-C1722, C1777, C1882, and C1895-C1896.

The following code is a "Pass Through" HCPCS C-code that represents devices such as the Optimizer Smart Implantable Pulse Generator for Heart Failure, designed to deliver electrical pulses to the right ventricular septum of the heart during the myocardial absolute refractory period:

- **C1824**: Generator, cardiac contractility modulation (implantable)

A subcutaneous implantable defibrillator system is reported with the following CPT codes: 33270-33273, 93260, 93261, and 93644.

Description

An 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), which lacks transvenous leads, is intended to reduce lead-related complications.

Related Policies

- Biventricular Pacemakers (Cardiac Resynchronization Therapy) for the Treatment of Heart Failure
- Wearable Cardioverter Defibrillators

Benefit Application

Benefit determinations should be based in all cases on the applicable contract language. To the extent there are any conflicts between these guidelines and the contract language, the contract language will control. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

Some state or federal mandates (e.g., Federal Employee Program [FEP]) prohibits plans from denying Food and Drug Administration (FDA)-approved technologies as investigational. In these instances, plans may have to consider the coverage eligibility of FDA-approved technologies on the basis of medical necessity alone.

Regulatory Status

Transvenous Implantable Cardioverter Defibrillators

A large number of ICDs have been approved by the FDA through the PMA process (FDA product code: LWS). A 2014 review of the FDA approvals of cardiac implantable devices reported that, between 1979 and 2012, the FDA approved 19 ICDs (7 pulse generators, 3 leads, 9 combined systems) through new PMA applications.¹ Many originally approved ICDs have received multiple supplemental applications. A selective summary of some currently available ICDs is provided in Table 1.

In April 2021, Medtronic issued a recall of the Evera, Viva, Brava, Claria, Amplia, Compia, and Visia ICDs and cardiac resynchronization therapy defibrillators (CRT-Ds) due to an unexpected and rapid decrease in battery life.² The decrease in battery life is caused by a short circuit and will cause some devices to produce a "Recommended Replacement Time" warning earlier than expected. Some devices may progress from this warning to full battery depletion within as little as 1 day. The device may stop functioning if the user does not respond to the first warning. In August 2022, Medtronic issued a recall of the Cobalt XT, Cobalt, and Crome ICDs and CRT-Ds because of risk that the devices may issue a short circuit alert and deliver a reduced energy electric shock instead of delivering a second phase of high voltage therapy.³ The reduced energy electrical shock may fail to correct an arrhythmia or may cause an irregular heartbeat. The FDA identified both events as Class I recalls, the most serious type of recall, indicating a situation in which use of these devices may cause serious injuries or death.

Subcutaneous Implantable Cardioverter Defibrillators

In 2012, the Subcutaneous Implantable Defibrillator (S-ICD™) System was approved by the FDA through the PMA process for the treatment of life-threatening ventricular tachyarrhythmias in patients who do not have symptomatic bradycardia, incessant VT, or spontaneous, frequently recurring VT that is reliably terminated with antitachycardia pacing (Table 1).

In 2015, the Emblem™ S-ICD (Boston Scientific), which is smaller and longer-lasting than the original S-ICD, was approved by the FDA through the PMA supplement process.

In February 2021, Boston Scientific issued a recall of the Emblem S-ICD because of increased risk of device fractures. The FDA designated the recall a Class I event, the most serious type of recall, indicating a situation in which there is a reasonable probability that the use of the device may cause serious injuries or death.⁴

Table 1. Implantable Cardioverter Defibrillators with FDA Approval

| Device | Manufacturer | Original PMA Approval Date |
|--|---|----------------------------|
| <i>Transvenous</i> | | |
| Ellipse™/Fortify Assura™ Family (originally: Cadence Tiered Therapy Defibrillation System) | St. Jude Medical | Jul 1993 |
| Current® Plus ICD (originally: Cadence Tiered Therapy Defibrillation System) | St. Jude Medical | Jul 1993 |
| Dynagen™, Inogen™, Origen™, and Teligen® Family (originally: Ventak, Vitality, Cofient family) | Boston Scientific | Jan 1998 |
| Evera™ Family (originally: Virtuosos/Entrust/Maximo/Intrinsic/Marquis family) | Medtronic | Dec 1998 |
| <i>Subcutaneous</i> | | |
| Subcutaneous Implantable Defibrillator System (S-ICD) | Cameron Health; acquired by Boston Scientific | Sep 2012 |

FDA: Food and Drug Administration; PMA: premarket application.

Rationale

Background

Ventricular Arrhythmia and Sudden Cardiac Death

The risk of ventricular arrhythmia and sudden cardiac death (SCD) may be significantly increased in various cardiac conditions such as ischemic cardiomyopathy, particularly when associated with reduced left ventricular ejection fraction (LVEF) and prior myocardial infarction (MI); nonischemic dilated cardiomyopathy with reduced LVEF; hypertrophic cardiomyopathy and additional risk factors; congenital heart disease, particularly with recurrent syncope; and cardiac ion channelopathies.

Treatment

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

The standard ICD placement surgery 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 ventricular fibrillation shock when a malignant arrhythmia is recognized.

A subcutaneous ICD (S-ICD) has been developed. It does not use transvenous leads and thus avoids the need for venous access and complications associated with the insertion of 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 have been approved by the U.S. Food and Drug Administration (FDA) through the premarket approval (PMA) process. The 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. Also, devices typically have approval in the secondary prevention setting for patients with previous MI and reduced ejection fraction.

Literature Review

Evidence reviews assess the clinical evidence to determine whether the use of technology improves the net health outcome. Broadly defined, health outcomes are the length of life, quality of life, and ability to function including benefits and harms. Every clinical condition has specific outcomes that are important to patients and managing the course of that condition. Validated outcome measures are necessary to ascertain whether a condition improves or worsens; and whether the magnitude of that change is clinically significant. The net health outcome is a balance of benefits and harms. To assess whether the evidence is sufficient to draw conclusions about the net health outcome of a technology, 2 domains are examined: the relevance, and quality and credibility. To be relevant, studies must represent 1 or more intended clinical use of the technology in the intended population and compare an effective and appropriate alternative at a comparable intensity. For some conditions, the alternative will be supportive care or surveillance. The quality and credibility of the evidence depend on study design and conduct, minimizing bias and confounding that can generate incorrect findings. The randomized controlled trial (RCT) is preferred to assess efficacy; however, in some circumstances, nonrandomized studies may be adequate. Randomized controlled trials are rarely large enough or long enough to capture less common adverse events and long-term effects. Other types of studies can be used for these purposes and to assess generalizability to broader clinical populations and settings of clinical practice.

Promotion of greater diversity and inclusion in clinical research of historically marginalized groups (e.g., People of Color [African-American, Asian, Black, Latino and Native American]; LGBTQIA (Lesbian, Gay, Bisexual, Transgender, Queer, Intersex, Asexual); Women; and People with Disabilities [Physical and Invisible]) allows policy populations to be more reflective of and findings more applicable to our diverse members. While we also strive to use inclusive language related to these groups in our policies, use of gender-specific nouns (e.g., women, men, sisters, etc.) will continue when reflective of language used in publications describing study populations.

Transvenous Implantable Cardioverter Defibrillators for Primary Prevention

Clinical Context and Therapy Purpose

The purpose of transvenous implantable cardioverter defibrillator (T-ICD) placement is to provide a treatment option that is an alternative to or an improvement on existing therapies in patients with a high risk of sudden cardiac death (SCD) due to ischemic or non-ischemic cardiomyopathy, inherited cardiac ion channelopathy, or cardiac sarcoid.

The following PICO was used to select literature to inform this review.

Populations

The relevant population of interest is individuals with a high risk of SCD due to ischemic or non-ischemic cardiomyopathy, inherited cardiac ion channelopathy, or cardiac sarcoid.

Interventions

The therapy being considered is T-ICD placement. An 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.

Comparators

Comparators of interest include medical management without ICD placement. Guideline-based medical management for ischemic cardiovascular disease includes antihypertensive therapy and antiarrhythmic medications. Medical management for cardiac sarcoid includes steroid therapy.

Outcomes

The general outcomes of interest are overall survival (OS), morbid events, quality of life, treatment-related mortality, and treatment-related morbidity. Table 2 describes outcomes of interest related to quality of life and treatment-related morbidity for individuals at high risk of SCD due to ischemic or non-ischemic cardiomyopathy.

Table 2. Outcomes of Interest for Individuals at High Risk of Sudden Cardiac Death due to Ischemic or Non-ischemic Cardiomyopathy in Adulthood

| Outcomes | Details | Timing |
|-----------------------------|---|-------------------|
| Quality of life | Can be assessed by patient reported data such as surveys and questionnaires | 1 week to 5 years |
| Treatment-related morbidity | Can be assessed by rates of adverse events, including inappropriate shock, lead failure, infection, and other complications | 1 week to 5 years |

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies;
- To assess long-term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought;
- Studies with duplicative or overlapping populations were excluded.

Review of Evidence**Primary Prevention in Adults**

Transvenous ICDs have been evaluated for primary prevention in a number of populations considered at high risk of 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 RCTs and systematic reviews of these trials, addressing the role of ICDs for primary prevention and identifying specific populations who may benefit.

Ischemic Cardiomyopathy and Nonischemic Dilated Cardiomyopathy**Randomized Controlled Trials**

At least 13 RCTs of ICDs for primary prevention have been conducted. Five were in populations with ischemic cardiomyopathy with prior myocardial infarction (MI; usually ≥ 3 weeks post-MI):

- Multicenter Automatic Defibrillator Implantation Trial (MADIT);
- MADIT II;
- Coronary Artery Bypass Graft (CABG) Patch trial;
- Multicenter Unsustained Tachycardia Trial (MUSTT); and
- Sudden Cardiac Death in Heart Failure (SCD HeFT) trial.

Three trials were conducted in patients implanted with ICD in the first few weeks following MI (recent MI):

- Defibrillator in Acute Myocardial Infarction Trial (DINAMIT);
- Immediate Risk Stratification Improves Survival (IRIS) trial; and
- BETA-blocker Strategy plus ICD (BEST-ICD) trial.

Six trials were conducted in populations with NIDCM:

- Comparison of Medical Therapy, Pacing, and Defibrillation in Heart Failure (COMPANION) trial;
- Amiodarone Versus Implantable Cardioverter-Defibrillator (AMIOVIRT) trial;
- Defibrillators in Non-Ischemic Cardiomyopathy Treatment Evaluation (DEFINITE) trial;
- SCD HeFT trial;
- Cardiomyopathy Trial (CAT); and
- Danish Study to Assess the Efficacy of ICDs in Patients with Non-Ischemic Systolic Heart Failure on Mortality (DANISH).

The characteristics and mortality results for these 3 groups of trials are shown in Table 3.

Most trials for both ischemic and nonischemic cardiomyopathy have reported results consistent with a mortality benefit for ICD in patients with left ventricular systolic dysfunction or with heart failure and reduced ejection fraction, although not all trials were powered for the mortality outcome and some findings were not statistically significant. However, the DINAMIT, IRIS, and BEST-ICD trials did not support a mortality benefit for ICD in the early weeks following MI, and CABG Patch showed no benefit in patients having recently undergone coronary revascularization. Another notable exception is the 2016 DANISH trial, which enrolled primarily outpatients with nonischemic cardiomyopathy (NICM) in stable condition who were almost all receiving β -blockers or angiotensin-converting enzyme inhibitors, with the majority also receiving mineralocorticoid-receptor antagonists. While overall mortality did not differ significantly between the ICD and medical therapy groups in DANISH, SCD was significantly reduced in the ICD group (4% vs. 8%; hazard ratio [HR], 0.50; 95% confidence interval [CI], 0.31 to 0.82).

Table 3. Characteristics and Results of RCTs of ICDs for Primary Prevention

| Trial | Participants | Treatment Groups | | Mean Follow-Up | Mortality Results | |
|--------------------------------------|--|--|--|-------------------------------------|---|--|
| | | Group | n | | Hazard Ratio | 95% CI |
| <i>ICM with prior MI</i> | | | | | | |
| MADIT (1996)⁵ | <ul style="list-style-type: none"> • LVEF \leq35% • Asymptomatic unsustained VT • MI \geq3 wk prior • Inducible VT • NYHA class I to III | <ul style="list-style-type: none"> • ICD • Standard therapy | <ul style="list-style-type: none"> • 95 • 101 | 27 mo (trial stopped early by DSMB) | 0.46 | 0.26 to 0.82 |
| MADIT II (2002)⁶ | <ul style="list-style-type: none"> • LVEF \leq30% • No history of VT • MI \geq1 mo prior • NYHA class I to III | <ul style="list-style-type: none"> • ICD • Standard therapy | <ul style="list-style-type: none"> • 742 • 490 | 20 mo (trial stopped early by DSMB) | 0.69 | 0.51 to 0.93 |
| CABG Patch (1997)⁷ | <ul style="list-style-type: none"> • Scheduled for CABG • LVEF \leq35% • No sustained VT or VF • Signal-averaged ECG abnormalities • 82% had prior MI, time since MI not reported | <ul style="list-style-type: none"> • ICD during CABG • No ICD | <ul style="list-style-type: none"> • 446 • 454 | 32 mo | 1.07 | 0.81 to 1.42 |
| MUSTT (1999)⁸ | <ul style="list-style-type: none"> • LVEF \leq40% • Asymptomatic unsustained VT • Inducible VT • MI \geq4 d prior (median, \gg3 y prior) • No sustained VT or VF | <ul style="list-style-type: none"> • EPS-guided therapy (AAD with or without ICD) (202 got ICD) • Standard therapy | <ul style="list-style-type: none"> • 351 • 353 | 39 mo | <ul style="list-style-type: none"> • 5-y outcomes^b: • EPS-guided vs standard therapy: 0.80 • ICD vs AAD alone: 0.42 | <ul style="list-style-type: none"> • 0.64 to 1.01 • 0.29 to 0.61 |

| Trial | Participants | Treatment Groups | Mean Follow-Up | Mortality Results |
|--|--|---|---|---|
| SCD HeFT (2005)⁹ | <ul style="list-style-type: none"> • LVEF \leq35% • NYHA class II to III • 52% received ICM • Treated with ACE inhibitors and β-blockers | Ischemic patients: <ul style="list-style-type: none"> • ICD • Amiodarone • Placebo | <ul style="list-style-type: none"> • 431 • 426 • 453 45 mo | • ICD vs. placebo • Ischemic: 0.79 ^a • Overall: 0.77 ^a • 0.60 to 1.04 • 0.62 to 0.96 |
| <i>ICM with recent MI</i> | | | | |
| DINAMIT (2004)¹⁰ | <ul style="list-style-type: none"> • LVEF \leq35% • NYHA class I to III • MI in preceding 6 to 40 d (mean, 18 d) • No sustained VT or VF for >48 h after index MI • Reduced HR variability or elevated resting HR | <ul style="list-style-type: none"> • ICD • Standard therapy | <ul style="list-style-type: none"> • 332 • 342 30 mo | 1.08 0.76 to 1.55 |
| IRIS (2009)¹¹ | <ul style="list-style-type: none"> • MI in preceding 5 to 31 d • At least 1 of the following: <ul style="list-style-type: none"> ○ LVEF \leq40% and resting HR \geq90 or unsustained VT | <ul style="list-style-type: none"> • ICD • Standard therapy | <ul style="list-style-type: none"> • 445 • 453 37 mo | 1.04 0.81 to 1.35 |
| BEST-ICD (2005)¹² | <ul style="list-style-type: none"> • LVEF \leq35% • NYHA class I to III • No unsustained VT or sustained ventricular arrhythmias (except primary VF) • MI in preceding 5 to 30 d • At least 1 other risk factor | <ul style="list-style-type: none"> • EPS-guided therapy (24 got ICD) • Standard therapy | <ul style="list-style-type: none"> • 79 • 59 540 d | 1-year mortality ^d <ul style="list-style-type: none"> • EPS-guided therapy: 14% • Conventional therapy: 18% 2-year mortality ^d <ul style="list-style-type: none"> • EPS-guided therapy: 20% • Conventional therapy: 29.5% |
| <i>Nonischemic cardiomyopathy</i> | | | | |
| DEFINITE (2004)¹³ | <ul style="list-style-type: none"> • LVEF \leq35% • NYHA class II to IV | <ul style="list-style-type: none"> • ICD and medical therapy • Medical therapy alone | <ul style="list-style-type: none"> • 229 • 229 29 mo | • 0.65 (0.40 to 1.06) |
| SCD HeFT (2005)⁹ | <ul style="list-style-type: none"> • LVEF \leq35% • NYHA class II to III • 48% with non-ICM • Treated with ACE inhibitors and β-blockers | Nonischemic patients: <ul style="list-style-type: none"> • ICD • Amiodarone • Placebo | <ul style="list-style-type: none"> • 398 • 419 • 394 45 mo | • ICD vs. placebo • Nonischemic: 0.73 ^a • Overall: 0.77 ^a • 0.50 to 1.07 • 0.62 to 0.96 |
| COMPANION (2004)¹⁴ | <ul style="list-style-type: none"> • LVEF \leq35% • NYHA class III to IV • DCM | Nonischemic patients: <ul style="list-style-type: none"> • CRT-D • Medical therapy • CRT | <ul style="list-style-type: none"> • 270 • 127 • 285 16 mo | • CRT-D vs. medical therapy • Nonischemic: 0.50 • Overall: 0.64 • 0.29 to 0.88 • 0.48 to 0.86 |

| Trial | Participants | Treatment Groups | Mean Follow-Up | Mortality Results |
|-------------------------------------|---|--|--|--|
| AMIOVIRT (2003)¹⁵ | <ul style="list-style-type: none"> • LVEF \leq35% • NYHA class I to III • DCM • Asymptomatic unsustained VT | <ul style="list-style-type: none"> • ICD • Amiodarone | <ul style="list-style-type: none"> • 51 • 52 | 2 years 1-year survival ^d <ul style="list-style-type: none"> • ICD: 96% • Amiodarone: 90% 2-year survival ^d <ul style="list-style-type: none"> • ICD: 88% • Amiodarone: 87% |
| CAT (2002)¹⁶ | <ul style="list-style-type: none"> • LVEF \leq30% • NYHA class II to III • No symptomatic VT, VF, or bradycardia • Recent-onset DCM | <ul style="list-style-type: none"> • ICD • Control | <ul style="list-style-type: none"> • 50 • 54 | 23 mo (trial stopped early due to low event rates) <ul style="list-style-type: none"> • ICD: 4 deaths (8%)^d • Control: 2 deaths (3.7%) |
| DANISH (2016)¹⁷ | <ul style="list-style-type: none"> • LVEF \leq35% • NYHA class II to IV • 58% received CRT • Almost all patients on ACE inhibitors or β-blockers; <ul style="list-style-type: none"> ◦ 60% treated with mineralocorticoid-receptor antagonist | <ul style="list-style-type: none"> • ICD and medical therapy • Medical therapy | <ul style="list-style-type: none"> • 556 • 560 | 5.6 years 0.87 0.68 to 1.12 |

AAD: antiarrhythmic drugs; ACE: angiotensin-converting enzyme; CABG: coronary artery bypass grafting; CI: confidence interval; CRT: cardiac resynchronization therapy; CRT-D: cardiac resynchronization therapy implantable cardioverter defibrillator; DCM: dilated cardiomyopathy; DSMB: Data Safety Monitoring Board; ECG: electrocardiogram; EPS: electrophysiologic study; HR: heart rate; ICD: implantable cardioverter defibrillator; ICM: ischemic cardiomyopathy; LVEF: left ventricular ejection fraction; MI: myocardial infarction; NYHA: New York Heart Association; RCT: randomized controlled trial; VF: ventricular fibrillation; VT: ventricular tachycardia.

^a 97.5% CI.

^b Relative risk.

^c Median.

^d Hazard ratio not given, no significant differences.

Systematic Reviews

Characteristics and results of systematic reviews of primary prevention ICD trials are described in Tables 4 and 5. Woods et al (2015) published an individual patient data network meta-analysis of primary prevention RCTs evaluating implantable cardiac devices, including studies of patients with heart failure and reduced ejection fraction and excluding studies of patients with recent MI or coronary revascularization.¹⁸ The COMPANION, DEFINITE, MADIT, MADIT II, SCD HeFT, AMIOVIRT, and CAT trials were included, representing 6134 patients for the direct ICD comparisons and 12638 patients overall.

Subsequent systematic reviews and meta-analyses of ICD trials in NICM incorporated the 2016 DANISH trial results.^{19,20,21,22,23} Two reviews published in 2017 included the CAT, AMIOVIRT, DEFINITE, SCD HeFT, COMPANION, and DANISH trials; one review published in 2021 included the CAT, AMIOVIRT, DEFINITE, and DANISH trials; other reviews included all but the COMPANION trial. The majority of the reviews concluded that there was a statistically significant overall reduction in mortality for ICD versus medical therapy, ranging from 20% to 23%, even with the inclusion of the null DANISH results.

The risk for death varies by age, sex, and clinical characteristics such as left ventricular ejection fraction (LVEF) and time since revascularization and comorbid conditions (e.g., diabetes, kidney

disease). Meta-analyses have examined whether there is a beneficial effect on mortality of ICD in these subgroups. Earley et al (2014) conducted a review of evidence for the Agency for Healthcare Research and Quality on use of ICD across important clinical subgroups.²⁴ Reviewers included 10 studies that provided subgroup analyses. Subgroup data were available from at least 4 studies for sex, age (<65 years vs. ≥65 years), and QRS interval (<120 ms vs. ≥120 ms); they were combined to calculate a relative odds ratio (OR) using random-effects meta-analyses. Other comparisons of subgroups were not meta-analyzed because too few studies compared them; however, no consistent differences between subgroups were found across studies for diabetes. The Woods et al (2015) individual patient data network meta-analysis (described previously) also examined ICD and medical therapy in various subgroups, and similarly concluded that ICD reduced mortality in patients with heart failure and reduced ejection fraction for QRS intervals less than 120 ms, 120 to 149 ms, and 150 ms or higher, ages less than 60 and 60 and older, and for men.¹⁸ However, the effect on mortality in women was not statistically significant (HR, 0.93; 95% CI, 0.73 to 1.18).

Table 4. Characteristics of Systematic Reviews & Meta-Analysis of ICDs for Primary Prevention

| Study | Dates | Trials | Participants | N (Range) | Design | Duration |
|-----------------------------|-----------|--------|---|----------------------|--|----------|
| Woods (2015) ¹⁸ | 1990-2010 | 13 | Patients with heart failure who received ICD | 12,638 (17 to 2,521) | RCT | NR |
| Earley (2014) ²⁴ | 1996-2010 | 14 | Adults eligible to receive an ICD for primary prevention of SCD | NR | RCT, Nonrandomized comparative studies | NR |

ICD: implantable cardioverter defibrillator; NR: not reported; RCT: randomized controlled trial; SCD: sudden cardiac death.

Table 5. Results of Systematic Reviews & Meta-Analysis of ICDs for Primary Prevention

| Study | Mortality |
|-----------------------------|---|
| Woods (2015) ¹⁸ | Estimated Effect of ICD on Mortality Compared with MT 0.71 (95% CI, 0.63 to 0.80) |
| Earley (2014) ²⁴ | Mortality Benefit of Variables (ROR) |
| Sex | 0.95 (95% CI, 0.75 to 1.27) |
| Age | 0.93 (95% CI, 0.73 to 1.20) |
| QRS interval | 1.13 (95% CI, 0.82 to 1.54) |

CI: confidence interval; ICD: implantable cardioverter defibrillator; MT: medical therapy; ROR: relative odds ratio.

Registry Studies

Fontenla et al (2016) reported on results from the Spanish UMBRELLA Registry, a multicenter, observational, prospective nationwide registry of 1514 patients implanted with Medtronic ICDs equipped with remote monitoring who were enrolled between 2012 and 2013.²⁵ The mean age of enrollees was 64 years; 82% of the patients were men; and 65% received an ICD for primary prevention. Fifty-one percent of the patients had ischemic heart disease, 30% had NICM, 7% had HCM, 3% had Brugada syndrome (BrS), and 1.4% had long QT syndrome (LQTS). Mean follow-up was 26 months. The cumulative incidence of sustained ventricular arrhythmias was 15% (95% CI, 13% to 16%) at 1 year, 23% (95% CI, 21% to 25%) at 2 years, and 31% (95% CI, 28% to 34%) at 3 years. Thirteen percent of the episodes of sustained ventricular arrhythmias self-terminated and did not require shocks. One hundred seventy-five (12%) patients had 482 appropriate shocks, and 76 (5%) patients had 190 inappropriate shocks.

High-Risk Hypertrophic Cardiomyopathy

Schinkel et al (2012) conducted a systematic review and meta-analysis of 27 observational studies (16 cohorts, 2190 patients) reporting outcomes after ICD therapy for HCM.²⁶ Most patients (83%) received an ICD for primary prevention of SCD. The mean age was 42, 38% of patients were women, and patients had a mean of 1.8 risk factors for SCD. With a mean follow-up of 3.7 years, 14% of patients had an appropriate ICD intervention with an annualized rate of 3.3%. Twenty percent of patients had an inappropriate ICD intervention, for an annualized rate of 4.8%. The annualized

cardiac mortality rate was 0.6%, the noncardiac mortality rate was 0.4%, and heart transplantation rate was 0.5%.

Magnusson et al (2015) reported on outcomes for 321 patients with HCM treated with an ICD and enrolled in a Swedish registry.²⁷ 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.

Inherited Cardiac Ion Channelopathy

Implantable cardioverter defibrillators have been used for primary and secondary prevention in patients with a number of hereditary disorders (also called cardiac ion channelopathies) that predispose to ventricular arrhythmias and SCD, including LQTS, BrS, short QT syndrome, and catecholaminergic polymorphic ventricular tachycardia (CPVT). Some of these conditions are extremely rare. Use of ICDs has been described in small cohorts of patients with LQTS, BrS, and CPVT.

Long QT Syndrome

Horner et al (2010) 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.²⁸ 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 ($p=.008$), QT corrected duration greater than 500 ms ($p<.001$), non-*LQT3* genotype ($p=.02$), documented syncope ($p=.05$), documented torsades de pointes ($p=.003$), and a negative sudden family death history ($p<.001$). Inappropriate shocks were delivered in 15 (29%) patients. Patients with the *LQT3* genotype only received inappropriate shocks.

Brugada Syndrome

Hernandez-Ojeda et al (2017) reported on results from a single-center registry of 104 patients with BrS who were treated with ICDs.²⁹ Ten (9.6%) patients received an ICD for secondary prevention and 94 (90.4%) patients received an ICD for primary prevention. During an average 9.3-year follow-up, 21 (20.2%) patients received a total of 81 appropriate shocks. In multivariate analysis, type 1 electrocardiogram (ECG) with syncope and secondary prevention indication were significant predictors of appropriate therapy. Nine (8.7%) patients received 37 inappropriate shocks. Twenty-one (20.2%) patients had other ICD-related complications.

Conte et al (2015) described outcomes for a cohort of 176 patients with spontaneous or drug-induced Brugada type 1 ECG findings who received an ICD at a single institution and were followed for at least 6 months.³⁰ 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 in 28 (15.9%) patients and antitachycardia pacing in 2 (1.1%) patients. However, 33 (18.7%) patients experienced inappropriate shocks.

Dores et al (2015) reported on results of a Portuguese registry that included 55 patients with BrS, 36 of whom were treated with ICDs for primary or secondary prevention.³¹ Before ICD placement, 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; $p=.027$) and nonsustained VT during follow-up (HR, 6.73; 95% CI, 1.27 to 35.7; $p=.025$).

Catecholaminergic Polymorphic Ventricular Tachycardia

Roses-Noguer et al (2014) reported on results of a small retrospective study of 13 patients with CPVT who received an ICD.³² 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 of 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.

Cardiac Sarcoid

Sarcoidosis is a systemic granulomatous disease of unknown etiology, with a worldwide prevalence of about 4.7 to 64 in 100,000.³³ The annual incidence of sarcoidosis in the United States has been estimated at 10.9 per 100,000 in White individuals and 35.5 per 100,000 in Black individuals. Cardiac involvement occurs in about 5% of systemic sarcoidosis cases. Steroid therapy is recommended as first-line treatment based on small cohort studies showing benefit, although there is conflicting evidence about its efficacy on long-term disease outcomes.³⁴

Mantini et al (2012) published a review on the diagnosis and management of cardiac sarcoid, including a treatment algorithm.³⁵ Limited evidence from small cohort studies suggested that an ICD could prevent dangerous arrhythmias or SCD even in patients with a relatively preserved LVEF. Evidence from case series also suggested that programmed electrical stimulation could identify patients with cardiac sarcoid with electrical instability and help to determine who should get ICD.

Section Summary: Transvenous Implantable Cardioverter Defibrillator for Primary Prevention in Adults

Ischemic Cardiomyopathy and Nonischemic Dilated Cardiomyopathy

A large body of RCTs has addressed the effectiveness of T-ICD implantation for primary prevention in patients at high risk of SCD due to ischemic cardiomyopathy and NICM. Evidence from several RCTs has demonstrated improvements in outcomes with ICD treatment for patients with symptomatic heart failure due to ischemic or NICM with an LVEF of 35% or less. The notable exceptions are that data from several RCTs, including the BEST-ICD, DINAMIT, and IRIS trials and subgroup analyses from earlier RCTs, have shown that outcomes with ICD therapy do not appear to improve for patients treated with an ICD within 40 days of recent MI and the CABG Patch trial did not find a benefit for patients undergoing coronary revascularization.

Hypertrophic Cardiomyopathy

Less evidence is available for the use of ICDs for primary prevention in patients with HCM. In a meta-analysis of cohort studies, the annual rates of appropriate ICD discharge were 3.3%, and the mortality rate was 1%. Given the long-term high risk of SCD in patients with HCM, with the assumption that appropriate shocks are life-saving, these rates are considered adequate evidence for the use of T-ICDs in patients with HCM.

Inherited Cardiac Ion Channelopathy

The evidence related to the use of ICDs in patients with inherited cardiac ion channelopathy includes primarily single-center cohort studies or registries of patients with LQTS, BrS, and CPVT that have reported on appropriate shock rates. Patient populations typically include a mix of those requiring ICD placement for primary or secondary prevention. The limited available data for ICDs for LQTS and CPVT have indicated high rates of appropriate shocks. For BrS, more data are available and have suggested that rates of 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. Given the long-term high risk of SCD in patients with inherited cardiac ion channelopathy, with the assumption that appropriate shocks are life-saving, these studies are considered adequate evidence for the use of T-ICDs in patients with inherited cardiac ion channelopathy.

Cardiac Sarcoid

The evidence related to the use of ICDs in patients with cardiac sarcoid includes small cohort studies of patients with cardiac sarcoid treated with ICDs who received appropriate shocks. Studies comparing outcomes between patients treated and untreated with ICDs are not available. However, given the relatively small number of patients with cardiac sarcoid (5% of those with systemic sarcoidosis), clinical trials are unlikely. Given the long-term high risk of SCD in patients with cardiac sarcoid, with the assumption that appropriate shocks are life-saving, these studies are considered adequate evidence to support the use of T-ICDs in patients with cardiac sarcoid who have not responded to optimal medical therapy.

Primary Prevention in Pediatric Populations

There is limited direct evidence on the efficacy of ICDs in the pediatric population. Most published studies have retrospectively analyzed small case series that included mixed populations with mixed indications for device placement. Some representative series are reviewed next.

The largest published series, by Berul et al (2008), combined pediatric patients and patients with congenital heart disease from 4 clinical centers.³⁶ The median age was 16 years, although some adults included were as old as 54 years. A total of 443 patients were included. The most common diagnoses were tetralogy of Fallot and HCM. Defibrillator placement was performed for primary prevention in 52% of patients and 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 (1993) compiled a database of 125 pediatric patients treated with an ICD through a query of the manufacturers of commercially available devices.³⁷ Indications for ICD placement were survivors of cardiac arrest (95 [76%] patients), drug-refractory VT (13 [10%] patients), and syncope with heart disease and inducible VT (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. Actual rates of SCD-free survival were 97% at 1 year, 95% at 2 years, and 90% at 5 years. Alexander et al (2004) reported on 90 ICD procedures in 76 young patients (mean age, 16 years; range, 1 to 30 years).³⁸ Indications for placement were 27 (36%) patients with cardiac arrest or sustained VT, 40 (53%) with syncope, 17 (22%) with palpitations, 40 (53%) with spontaneous ventricular arrhythmias, and 36 (47%) 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% received an inappropriate shock. Lewandowski et al (2010) reported on long-term follow-up for 63 patients, between the ages of 6 and 21 years, who were treated with an ICD device.³⁹ 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: Primary Prevention in Pediatric Populations

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 device placement. Overall, these studies have reported both relatively high rates of appropriate and inappropriate shocks. Pediatric patients may be eligible for ICD placement if they have inherited cardiac ion channelopathy (see Inherited Cardiac Ion Channelopathy section).

Transvenous Implantable Cardioverter Defibrillators for Secondary Prevention Clinical Context and Therapy Purpose

The purpose of T-ICD placement is to provide a treatment option that is an alternative to or an improvement on existing therapies in patients with life-threatening ventricular tachyarrhythmia or fibrillation or who have been resuscitated from sudden cardiac arrest.

The following PICO was used to select literature to inform this review.

Populations

The relevant population of interest is individuals with life-threatening ventricular tachyarrhythmia or fibrillation or who have been resuscitated from sudden cardiac arrest.

Interventions

The therapy being considered is T-ICD placement. An ICD is a device designed to monitor a patient's heart rate, recognize VF or VT, and deliver an electric shock to terminate these arrhythmias to reduce the risk of sudden death.

Comparators

Comparators of interest include medical management without ICD placement.

Outcomes

The general outcomes of interest are OS, morbid events, quality of life, treatment-related mortality, and treatment-related morbidity.

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies;
- To assess long-term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought;
- Studies with duplicative or overlapping populations were excluded.

Review of Evidence**Secondary Prevention in Adults**

At least 5 trials comparing ICD plus medical therapy with medical therapy alone have been conducted in the secondary prevention setting: the Antiarrhythmics Versus Implantable Defibrillators (AVID) trial⁴⁰, (N=1016), Cardiac Arrest Survival in Hamburg (CASH) trial⁴¹, (N=288), Canadian Implantable Defibrillator Study (CIDS)⁴², (N=659), Defibrillator Versus Beta-Blockers for Unexplained Death in Thailand (DEBUT)⁴³, trial (N=66; pilot, n=20; main study, n=46), and Wever et al (1995)⁴⁴, (N=60). The trials are shown in Table 6. The mean length of follow-up varied from 18 to 57 months across trials. Lee et al (2003) combined the AVID, CASH, CIDS, and Wever et al (1995) trials in a meta-analysis of secondary prevention trials.⁴⁵ The mortality analysis included 2023 participants and 518 events. In combined estimates, the ICD group had a significant reduction in both mortality (HR, 0.75; 95% CI, 0.64 to 0.87) and SCD (HR, 0.50; 95% CI, 0.34 to 0.62) compared with the group receiving medical therapy alone. To support National Institute for Health and Care Excellence guidance on the use of ICDs, AVID, CASH, CIDS, and the pilot DEBUT participants were combined in a meta-analysis.⁴⁶ The results were similar, indicating a reduction in mortality for ICDs compared with medical therapy alone (relative risk [RR], 0.75; 95% CI, 0.61 to 0.93). Two other meta-analyses that included AVID, CIDS, and CASH reached similar conclusions.^{47,48}

Table 6. RCTs of ICDs for Secondary Prevention

| Trials | Participants | Treatment Groups | | Mortality Results | |
|---------------------------|---|------------------|-------|-------------------|--------------|
| | | Group | N | RR | 95% CI |
| AVID (1997) ⁴⁰ | Patients resuscitated from near-fatal VT/VF, sustained VT with syncope, or sustained VT with LVEF \leq 40% and symptoms | • ICD | • 507 | 0.66 | 0.51 to 0.85 |
| | | • AAD | • 509 | | |
| CASH (2000) ⁴¹ | Patients resuscitated from cardiac arrest due to sustained ventricular arrhythmia | • ICD | • 99 | 0.82 | 0.60 to 1.11 |
| | | • Amiodarone | • 92 | | |
| | | • Metoprolol | • 97 | | |

| Trials | Participants | Treatment Groups | Mortality Results |
|------------------------------------|--|--|---|
| CIDS (2000) ⁴² . | Patients with VF, out-of-hospital cardiac arrest requiring defibrillation, VT with syncope, VT with rate ≥ 150 /min causing presyncope or angina in patients with LVEF $\leq 35\%$ or syncope with inducible VT | <ul style="list-style-type: none"> • ICD • 328 • Amiodarone • 331 | 0.85 0.67 to 1.10 |
| Wever et al (1995) ⁴⁴ . | Patients with previous MI and resuscitated cardiac arrest due to VT or VF and inducible VT | <ul style="list-style-type: none"> • ICD • 29 • AAD • 31 | 0.39 0.14 to 1.08 |
| DEBUT (2003) ⁴³ . | Patients with SUDS or probable SUDS survivors with ECG abnormalities showing a RBBB-like pattern with ST elevation in the right precordial leads and inducible VT/VF | Pilot <ul style="list-style-type: none"> • ICD • 10 • β-blocker therapy • 10 Main trial <ul style="list-style-type: none"> • β-blocker therapy • 37 • ICD • 29 | <ul style="list-style-type: none"> • RR not calculable (DSMB stopped trial early due to efficacy of ICD) • 7 deaths in β-blockers vs. 0 in ICD |

AAD: antiarrhythmic drugs; CI: confidence interval; DSMB: Data Safety Monitoring Board; ECG: electrocardiogram; ICD: implantable cardioverter defibrillator; LVEF: left ventricular ejection fraction; MI: myocardial infarction; RBBB: right bundle-branch block; RCT: randomized controlled trial; RR: relative risk; SUDS: sudden unexplained death syndrome ; VF: ventricular fibrillation; VT: ventricular tachycardia.

An analysis by Chan and Hayward (2005) using the National Veterans Administration database previously confirmed that this mortality benefit is generalizable to the clinical setting.⁴⁹ A cohort of 6996 patients in the National Veterans Administration database, from 1995 to 1999, who had new-onset ventricular arrhythmia and preexisting ischemic heart disease and congestive heart failure were included. Of those, 1442 patients had received an ICD. Mortality was determined through the National Death Index at 3 years from the hospital discharge date. The cohort was stratified by quintiles of a multivariable propensity score created using many demographic and clinical confounders. The propensity score-adjusted mortality reduction for ICD compared with no ICD was an RR of 0.72 (95% CI, 0.69 to 0.79) for all-cause mortality and an RR of 0.70 (95% CI, 0.63 to 0.78) for cardiovascular mortality.

Section Summary: Secondary Prevention in Adults

Systematic reviews of RCTs in patients who have experienced symptomatic life-threatening sustained VT or VF or have been successfully resuscitated from sudden cardiac arrest have shown a 25% reduction in mortality for ICD compared with medical therapy. Analysis of data from a large administrative database has confirmed that this mortality benefit is generalizable to the clinical setting.

Secondary Prevention in Pediatric Populations

There is limited direct evidence on the efficacy of ICDs in the pediatric population. Most published studies have retrospectively analyzed small case series that included mixed populations with mixed indications for device placement. Some representative series were reviewed above (see Primary Prevention in Pediatric Populations section).

Section Summary: Secondary Prevention in Pediatric Populations

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 device placement. Overall, these studies have reported both relatively high rates of appropriate and inappropriate shocks. Pediatric patients may be eligible for ICD placement if they have inherited cardiac ion channelopathy (see Inherited Cardiac Ion Channelopathy section).

Adverse Events Associated With Transvenous Implantable Cardioverter Defibrillators

Systematic Reviews: Mixed Adverse Events

Characteristics and results of systematic reviews of adverse events associated with transvenous ICDs are described in Tables 7 and 8. Persson et al (2014) conducted a systematic review of adverse events following ICD placement.⁵⁰ In-hospital serious adverse event rates ranged from 1.2% to 1.4%, most frequently pneumothorax (0.4% to 0.5%) and cardiac arrest (0.3%).

In another systematic review of adverse events following ICD placement, Ezzat et al (2015) compared event rates reported in clinical trials of ICDs with those reported in the U.S. National Cardiovascular Data Registry.⁵¹ 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 the U.S. registry, $p < .01$). The overall complication rate was similar to that reported by Kirkfelt et al (2014), in a population-based cohort study including all Danish patients who underwent a cardiac implantable electronic device procedure from 2010 to 2011 (562 [9.5%] of 5918 patients with at least 1 complication).⁵²

Van Rees et al (2011) reported on results of a systematic review of RCTs assessing implant-related complications of ICDs and cardiac resynchronization therapy (CRT) devices.⁵³ Reviewers 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 placement, 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 placement, the average in-hospital mortality rate was 2.7%. For nonthoracotomy ICD placement, the overall lead dislodgement rate was 1.8%.

Olde Nordkamp et al (2016) reported on a systematic review and meta-analysis of studies reporting on ICD complications in individuals with inherited arrhythmia syndromes.⁵⁴ Reviewers 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 variants; 462 [9.4%] with LQTS; 51 [1.0%] with short QT syndrome).

Table 7. Systematic Reviews & Meta-Analysis Characteristics for Adverse Events Associated With T-ICDs

| Study | Dates | Trials | Participants | N (Range) | Design | Duration |
|------------------------------------|-----------|--|--|-------------------|----------------|----------|
| Persson (2014) ⁵⁰ | 2005-2012 | <ul style="list-style-type: none"> 53 trials; 35 cohorts | Patients receiving ICD placement | NR | Cohort studies | NR |
| Ezzat (2015) ⁵¹ | 2001-2011 | 18 | Patients receiving ICD placement | 6796 (16 to 1530) | RCTs | NR |
| Olde Nordkamp (2016) ⁵⁴ | 1997-2014 | 63 | Patients with inherited arrhythmia syndromes receiving ICD placement | 4916 (NR) | Cohort studies | NR |

ICD: implantable cardioverter defibrillator; NR: not reported; RCT: randomized controlled trials; T-ICD: transvenous implantable cardioverter defibrillator.

Table 8. Systematic Reviews & Meta-Analysis Results for Adverse Events Associated With T-ICDs

| Study | Rate of Adverse Events | Rates of Specific Complications |
|------------------------------|-----------------------------|---|
| Persson (2014) ⁵⁰ | | |
| Range | 1.2% to 1.4% ¹ | <ul style="list-style-type: none"> Device-related: <0.1% to 6.4% Lead-related: <0.1% to 3.9% Infection: 0.2% to 3.7% Inappropriate shock: 3% to 21% |
| Ezzat (2015) ⁵¹ | 9.1 (95% CI, 6.4% to 12.6%) | <ul style="list-style-type: none"> Access-related: 2.1% (95% CI, 1.3% to 3.3%) Lead-related: 5.8% (95% CI, 3.3% to 9.8%) |

| Study | Rate of Adverse Events | Rates of Specific Complications |
|--|---|--|
| Olde Nordkamp (2016)⁵⁴ | 22% (4.4% per year; 95% CI, 3.6% to 5.2%; p<.001) | <ul style="list-style-type: none"> • Generator-related: 2.7% (95% CI, 1.3% to 5.7%) • Infection: 1.5% (95% CI, 0.8% to 2.6%) • Lead malfunction: 10.3% • Infection: 3.0% (0.53% per year) • Inappropriate shock: 20% (4.7% per year; 95% CI, 4.2% to 5.3%; p<.001) |

CI: confidence interval; T-ICD: transvenous implantable cardioverter defibrillator.

¹Only serious adverse events, which included cardiac arrest, cardiac perforation, cardiac valve injury, coronary venous dissection, hemothorax, pneumothorax, deep phlebitis, transient ischemic attack, stroke, myocardial infarction, pericardial tamponade, arteriovenous fistula, and, in 1 study, lead dislodgement.

Systematic Reviews: Specific Complications

Lead Failure

The failure of leads in 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 important questions related to follow-up of affected patients.⁵⁵ An evaluation by Hauser et al (2010) 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 database.⁵⁶

Providencia et al (2015) reported on a meta-analysis of 17 observational studies evaluating the performance of 49871 leads (5538 Durata, 10605 Endotak Reliance, 16119 Sprint Quattro, 11709 Sprint Fidelis, 5900 Riata).⁵⁷ Overall, the incidence of lead failure was 0.93 per 100 lead-years (95% CI, 0.88 to 0.98). In an analysis of studies restricted to head-to-head comparisons of leads, there were no significant differences in lead failure rates among nonrecalled leads (Endotak Reliance, Durata, Sprint Quattro).

Birnie et al (2012) reported on clinical predictors of failure for 3169 Sprint Fidelis leads implanted from 2003 to 2007 at 11 centers participating in the Canadian Heart Rhythm Society study.⁵⁸ 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=.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 study from 3 centers reporting on predictors of Fidelis lead failures, compared with Quattro lead failures, Hauser et al (2011) reported a failure rate for the Fidelis lead of 2.81% per year (vs. 0.42% per year for Quattro leads; p<.001).⁵⁹

In a large prospective multicenter study, Poole et al (2010) reported on 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).⁶⁰ 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. Overall periprocedural complication rates for those with a planned transvenous lead replacement were a 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 (2012) evaluated the incidence of lead failure in a cohort of 414 patients given an ICD with Sprint Fidelis leads.⁶¹ 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. The median time until recognition of lead failure, or until an adverse event, was 2.2 days. A total of 22 (5.3%) patients received an inappropriate shock due to lead failure.

Cheng et al (2010) examined the rate of lead dislodgements in patients enrolled in a national cardiovascular registry.⁶² Of 226764 patients treated with an ICD between 2006 and 2008, lead dislodgement occurred in 2628 (1.2%). Factors associated with lead dislodgement were New York Heart Association (NYHA) class IV heart failure, atrial fibrillation or atrial flutter, a combined ICD and 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.

In another single-center study, Faulkner et al (2010) reported on the time-dependent hazard of failure of Sprint Fidelis leads.⁶³ Over an average follow-up of 2.3 years, 38 (8.9%) of 426 leads failed. There was a 3-year lead survival rate 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; $p < .001$).

Infection Rates

Several publications have reported on infection rates in patients receiving an ICD. Smit et al (2010) published a retrospective, descriptive analysis of the types and distribution of infections associated with ICDs over a 10-year period in Denmark.⁶⁴ 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 (2010) reported on the rate of ICD-associated infections among consecutive patients treated with an ICD at a tertiary referral center.⁶⁵ 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 (e.g., combined ICD plus CRT or dual-/triple-chamber devices). Sohail et al (2011) performed a case-control study evaluating the risk factors for an ICD-related infection in 68 patients and 136 matched controls.⁶⁶ On multivariate analysis, the presence of epicardial leads (OR, 9.7; $p = .03$) and postoperative complications at the insertion site (OR, 27.2, $p < .001$) were significant risk factors for early infection. For late-onset infections, hospitalization for more than 3 days (OR, 33.1, $p < .001$ for 2 days vs. 1 day) and chronic obstructive pulmonary disease (OR, 9.8, $p = .02$) were significant risk factors.

Borleffs et al (2010) also reported on complications after ICD replacement for pocket-related complications, including infection or hematoma, in a single-center study.⁶⁷ Of 3161 ICDs included, 145 surgical reinterventions were required for 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 infection and 1.7 (95% CI, 0.9 to 3.0) times higher for non-infection-related causes.

Inappropriate Shocks

Inappropriate shocks may occur with ICDs due to faulty sensing or sensing of atrial arrhythmias with rapid ventricular conduction. These shocks may lead to reduced quality of life 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; $p = .02$).⁶⁸

Tan et al (2014) conducted a systematic review to identify outcomes and adverse events associated with ICDs with built-in therapy-reduction programming.⁶⁹ Six randomized trials and 2 nonrandomized cohort studies (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 (RR, 0.50; 95% CI, 0.37 to 0.61; $p < .001$). Therapy-reduction programming was associated with a significantly lower risk of death than conventional programming (RR, 0.30; 95% CI, 0.16 to 0.41; $p < .001$).

Sterns et al (2016) reported on 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.⁷⁰ This trial reported on a prespecified subgroup analysis of the PainFree SST trial, which compared standard with prolonged detection in patients receiving an ICD for secondary prevention. Patients treated for secondary prevention indications were randomized to a prolonged VF detection period (n=352) or a standard detection period (n=353). At 1 year, arrhythmic syncope-free rates were 96.9% in the intervention group, and 97.7% in the control group (rate difference, -1.1%; 90% lower confidence limit, -3.5%; above the prespecified noninferiority margin of -5%; p=.003 for noninferiority).

Auricchio et al (2015) assessed data from the PainFree SST trial, specifically newer ICD programming strategies for reducing inappropriate shocks.⁷¹ A total of 2790 patients with an indication for ICD placement 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 (2010) evaluated rates of early complications among patients enrolled in a prospective, multicenter population-based registry of all newly implanted ICDs in Ontario, from 2007 through 2009.⁷² 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<.01).

Furniss et al (2015) prospectively evaluated changes in high-sensitivity troponin T levels and ECG results that occur during ICD placement alone, ICD placement with testing, and ICD testing alone.⁷³ The 13 subjects undergoing ICD placement alone had a median increase in high-sensitivity troponin T level of 95% (p=.005) while the 13 undergoing implantation and testing had a median increase of 161% (p=.005). Those undergoing testing alone demonstrated no significant change in high-sensitivity troponin T levels.

Subcutaneous Implantable Cardioverter Defibrillators in Individuals with a Contraindication to a Transvenous Implantable Cardioverter Defibrillator

Clinical Context and Therapy Purpose

The purpose of subcutaneous implantable cardioverter defibrillators (S-ICD) placement in patients with a contraindication to transvenous T-ICD is to provide a treatment option that is an alternative to or an improvement on existing therapies such as medical management without ICD placement. The following PICO was used to select literature to inform this review.

Populations

The population of interest is individuals who need an ICD and have a contraindication to a T-ICD. There are no defined guidelines for the selection of S-ICD versus T-ICD. Currently, S-ICDs are generally considered in the following situations:

- Individuals at high risk of infection, inadequate venous access, and any individuals without a pacing indication.
- Younger individuals due to the expected longevity of the implanted leads and a desire to avoid chronic transvenous leads (e.g., individuals with HCM, congenital cardiomyopathies, or inherited channelopathies).
- Individuals at high risk for bacteremia, such as individuals on hemodialysis or with chronic indwelling endovascular catheters.
- Individuals with challenging vascular access or prior complications with T-ICDs

Interventions

The therapy being considered is S-ICD. An ICD is a device designed to monitor an individual's heart rate, recognize VF or VT, and deliver an electric shock to terminate these arrhythmias to reduce the risk of sudden death. A S-ICD, which lacks transvenous leads, is intended to reduce lead-related complications. The S-ICD is intended for individuals who have standard indications for an ICD, but who do not require pacing for bradycardia or antitachycardia overdrive pacing for VT. The S-ICD is proposed to benefit individuals with limited vascular access (including individuals undergoing renal dialysis or children) or those who have had complications requiring T-ICDs explantation.

The S-ICD is comprised of a pulse generator and single shocking coil running along the left parasternal margin. These are both implanted subcutaneously without endovascular access. The electrode is designed to be implanted using anatomical landmarks only without the need for fluoroscopy or other medical imaging systems during the surgical implant procedure.

Comparators

The comparator of interest is medical management without ICD placement.

Outcomes

The general outcomes of interest are OS, morbid events, quality of life, treatment-related mortality, and treatment-related morbidity. Table 9 describes outcomes of interest related to quality of life and treatment-related morbidity for individuals who need an ICD and have a contraindication to a T-ICD.

Table 9. Outcomes of Interest for Individuals Who Need an ICD and Have a Contraindication to a T-ICD

| Outcomes | Details | Timing |
|-----------------------------|---|-------------------|
| Quality of life | Can be assessed by patient reported data such as surveys and questionnaires | 1 week to 5 years |
| Treatment-related morbidity | Can be assessed by rates of adverse events, including inappropriate shock, lead failure, infection, and other complications | 1 week to 5 years |

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies;
- To assess long-term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought;
- Studies with duplicative or overlapping populations were excluded.

Review of Evidence**Randomized Trials**

Healey et al (2022) published 2.5 year interim results of the randomized, multicenter Avoid Transvenous Leads in Appropriate Subjects (ATLAS S-ICD) trial.⁷⁴ This trial included 544 individuals (141 female) with a primary or secondary prevention indication for an ICD who were younger than 60 years, had a cardiogenic phenotype, or had prespecified risk factors for lead complications. Of those, 503 were randomized to S-ICD (n=251) or T-ICD (n=252). Mean age of included patients was 49 years. The primary outcome focused on perioperative complications that are lead-related. Within 6 months of implantation, perioperative, lead-related complications occurred in 1 patient (0.4%) with an S-ICD and in 12 patients (4.8%) with T-ICD (risk difference, -4.4%; 95% CI, -6.9 to -1.9; p=.001). Overall, complications between groups were similar at 6 months, including device-related infection requiring surgery (S-ICD, 11 patients vs T-ICD, 14 patients; risk difference, -1.2; 95% CI, -2.4 to 0.1). More patients in the S-ICD group experienced ICD site pain on the day of implant (p<.001) and 1 month later

($p=.035$) compared to T-ICD patients. There were no differences in pain scores at 6 months. After a follow-up of 2.5 years, there was a trend for more inappropriate shocks with S-ICD (S-ICD, 16 patients vs T-ICD, 7 patients; HR, 2.37; 95% CI, 0.98 to 5.77), but no increase in failed appropriate ICD shocks (HR, 0.61; 95% CI, 0.15 to 2.57), however, this trial was not powered to detect differences in clinical shock outcomes. Although the ATLAS trial found a decreased risk of lead-related perioperative complications, it was underpowered to detect differences in clinical shock outcomes, however, extended follow-up is ongoing.

Nonrandomized Trials

Several nonrandomized trials and registry studies have reported outcomes for patients receiving a S-ICD, with follow up periods up to 5.8 years (Table 10). The Implant and Midterm Outcomes of the Subcutaneous Implantable Cardioverter-Defibrillator Registry (EFFORTLESS) is a multicenter European registry reporting outcomes for patients treated with S-ICD. Several publications from EFFORTLESS (Evaluation of Factors Impacting Clinical Outcome and Cost Effectiveness of the S-ICD), the pivotal trial submitted to the FDA for the investigational device exemption, and other studies are summarized in Table 10. In the EFFORTLESS registry, among 472 enrolled patients, the complication-free rate was 94% at 360 days and there was a 13.1% inappropriate shock rate at 3 years' follow-up. Gold et al (2021) reported 18-month data from the UNTOUCHED study, a multinational, prospective trial designed to assess the performance of the S-ICD in primary prevention patients with a low LVEF and NYHA II/III heart failure or coronary artery disease.⁷⁵ At 18 months, the complication-free rate was 92.7% and the inappropriate shock-free rate was 95.9%. One-year data from the S-ICD Post Approval Study and 18-month data from the UNTOUCHED study have been published; these studies are ongoing. The S-ICD System Post-Approval Study (PAS) is a nonrandomized, standard-of-care registry in the United States that has prospectively enrolled and followed S-ICD recipients.⁷⁶ Over the first 1 year postimplantation, complications were observed in 119 patients, with a complication-free rate at 1 year of 92.5%. The most common complication was device system infection in 44 of 1637 patients. Gold et al (2022) reported on the 3-year postimplantation follow-up data of the S-ICD PAS.⁷⁷ Within 3 years, infection was observed in 55 patients (3.3%) with 69% of infections occurring within 90 days of implantation and the majority (92.7%) within 1 year of implantation. No patient included in the registry had more than 1 infection and no infections occurred after 2 years in the cohort. The annual post-infection mortality rate was 0.6%. Based on their findings, the authors developed a risk score for likelihood of developing an infection, with diabetes, age ≥ 55 years, previous ICD implant, or LVEF $\leq 30\%$ all identified as contributing risk to S-ICD related infection. This risk score has not been externally validated. The S-ICD PAS study has been completed (NCT01736618) but 5-year results have yet to be published. Five-year data from the PAS should provide more information on longer-term adverse events such as lead failure and need for device replacement.

Table 10. Summary of Nonrandomized Trials of S-ICD

| Study; Trial | Countries | N | Mean FU | Results | Values |
|---|-----------------------|------|-----------|--|---------|
| Burke et al (2020) ⁷⁶ S-ICD PAS NCT01736618 | US | 1637 | 1 y | Outcomes | |
| | | | | • Complication-free rate at 1 y | • 92.5% |
| | | | | • Appropriate shock rate at 1 y | • 5.3% |
| | | | | • Inappropriate shocks at 1 y | • 6.5% |
| | | | | • Death at 1 y | • 5.4% |
| Gold et al (2021) ⁷⁵ UNTOUCHED | US, Canada, Europe | 1111 | 18 months | • Inappropriate shock-free rate at 18 months | • 94.8% |
| | | | | • Appropriate shock-free rate at 18 months | • 94.3% |
| | | | | • Complication-free rate at 18 months | • 92.7% |
| | | | | • Overall survival rate at 18 months | • 94.9% |
| | | | | • e at 18 months | |

| Study; Trial | Countries | N | Mean FU | Results |
|--|---|---|--|---|
| Lambiase et al (2014) ⁷⁸ ; Olde Nordkamp et al (2015) ⁷⁹ ; Boersma et al (2017) ⁸⁰ , EFFORTLESS S- ICD Registry | 10 European countries | • 985 • 928 • 697 • 498 • 300 • 82 | • 3.1 y • 1 y • 2 y • 3 y • 4 y • 5 y | <ul style="list-style-type: none"> • Complication-rates by 360 d • 8.4% • Inappropriate shocks by 360 d • 8.1% • Complication rates through follow-up • 11.7% • Inappropriate shocks through follow-up • 11.7% • Appropriate shocks through follow-up • 13.5% |
| Weiss et al (2013) ⁸¹ , IDE study | U.S., U.K., New Zealand, Netherlands | 330 | 11 mo | <ul style="list-style-type: none"> • Implanted successfully • 95% • Complication-free at 180 d • 99% • Inappropriate shocks • 13% • Episodes of discrete spontaneous VT or VF, all successfully converted • 38 |
| Burke et al (2015) ⁷⁶ ; Boersma et al (2016) ⁸² ; Lambiase et al (2016) ⁸³ , EFFORTLESS and IDE studies | Multiple European countries, U.S., New Zealand | 882 | 651 d | <ul style="list-style-type: none"> • Complications within 3 y • 11% • Infections requiring device removal or revision • 1.7% • Annual mortality rate • 1.6% • 2-y cumulative mortality • 3.2% • Incidence of therapy for VT or VF: • 5.3% ○ 1 year • 7.9% ○ 2 years • 10.5% ○ 3 years • 13.1% • Incidence of inappropriate shock at 3 y |
| Bardy et al (2010) ⁸⁴ ; Theuns et al (2015) ⁸⁵ . | Europe, New Zealand | 55 | 5.8 y | <ul style="list-style-type: none"> • Devices replaced • 26 (47%) • Devices explanted • 5 (9%) • Replaced with T-ICD • 4 (7%) • Shocks recorded in 16 (29%) patients • 119 |
| Olde-Nordkamp et al (2012) ⁸⁶ . | Netherlands | 118 | 18 mo | <ul style="list-style-type: none"> • All device-related complications • 14% • Infections • 5.9% • Dislodgements of device/leads • 3.3% • Skin erosion • 1.7% • Battery failure • 1.7% • Replaced with T-ICD • 1 (0.8%) • Appropriate shocks experienced in 8 patients • 45 • Total inappropriate shocks delivered to 15 (13%) patients • 33 • Deaths (cancer, progressive heart failure) • 2 |

FU: follow-up; S-ICD: subcutaneous implantable cardioverter defibrillator; T-ICD: transvenous implantable cardioverter defibrillator; VF: ventricular fibrillation; VT: ventricular tachycardia.

Section Summary: Subcutaneous-Implantable Cardioverter Defibrillators in Individuals with a Contraindication to a Transvenous Implantable Cardioverter Defibrillator

An RCT found that S-ICD significantly decreases the risk of lead-related perioperative complications compared to T-ICD. However, this study was not powered to detect differences in the rates of failed shocks or inappropriate shocks and an extension study is ongoing. Nonrandomized studies have suggested that S-ICDs are as effective as T-ICDs at terminating laboratory-induced ventricular arrhythmias. Data from large patient registries have suggested that S-ICDs are effective at

terminating ventricular arrhythmias when they occur. Given the need for cardioverter defibrillation for SCD risk in this population, with the assumption that appropriate shocks are life-saving, these studies suggest S-ICDs, in patients with contraindication to T-ICD, are likely improvements over medical management alone.

Subcutaneous Implantable Cardioverter Defibrillators in Individuals with No Contraindication to a Transvenous Implantable Cardioverter Defibrillator

Clinical Context and Therapy Purpose

The purpose of S-ICD placement in individuals with no contraindication to a T-ICD is to provide a treatment option that is an alternative to or an improvement on existing therapies.

The following PICO was used to select literature to inform this review.

Populations

The population of interest is individuals who need an ICD and have no contraindication to a T-ICD. There are no defined guidelines for the selection of S-ICD versus T-ICD. Currently, S-ICDs are generally considered in the following situations:

- Individuals at high risk of infection, inadequate venous access, and any patient without a pacing indication.
- Younger individuals due to the expected longevity of the implanted leads and a desire to avoid chronic transvenous leads (e.g., patients with HCM, congenital cardiomyopathies, or inherited channelopathies).
- Individuals at high risk for bacteremia, such as individuals on hemodialysis or with chronic indwelling endovascular catheters.
- Individuals with challenging vascular access or prior complications with T-ICDs.

Interventions

The therapy being considered is S-ICD. An ICD is a device designed to monitor an individual's heart rate, recognize VF or VT, and deliver an electric shock to terminate these arrhythmias to reduce the risk of sudden death. An S-ICD, which lacks transvenous leads, is intended as an alternative to T-ICD to reduce lead-related complications. The S-ICD is comprised of a pulse generator and single shocking coil running along the left parasternal margin. These are both implanted subcutaneously without endovascular access. The electrode is designed to be implanted using anatomical landmarks only without the need for fluoroscopy or other medical imaging systems during the surgical implant procedure.

Comparators

The comparator of interest is T-ICD placement.

Outcomes

The general outcomes of interest are OS, morbid events, quality of life, treatment-related mortality, and treatment-related morbidity. Outcomes should be assessed from 1 week to 5 years or longer. Specific outcomes include the following:

- Sudden cardiac death
- All-cause mortality
- Adverse events including nonlead-related complications (device infection, hematoma, pneumothorax, pericardial effusion), inappropriate shocks, device failure; and lead-related complications
- Cardiovascular mortality
- Health-related quality of life
- Hospital re-admission

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies;
- To assess long-term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought;
- Studies with duplicative or overlapping populations were excluded.

Review of Evidence

Randomized Controlled Trials

The Prospective, Randomized Comparison of Subcutaneous and Transvenous Implantable Cardioverter Defibrillator Therapy (PRAETORIAN) trial was a noninferiority RCT that compared S-ICD to T-ICD in 849 patients with an indication for ICD but no indication for pacing (Table 11).⁸⁷ The trial is the only RCT on the effect of an S-ICD with health outcomes. Patients were eligible if they were 18 years and older with a class I or IIa indication for ICD therapy for primary or secondary prevention, according to professional society guidelines, and no indication for pacing. The median age of enrolled patients was 63 years (interquartile range, 55 to 70). Most enrolled patients were diagnosed with ischemic and nonischemic cardiomyopathy and 19.7% were women. The median LVEF was 30%. The primary endpoint in PRAETORIAN was the composite of device-related complications and inappropriate shocks (see Table 11 for outcome definitions). The trial was designed to test the hypothesis of noninferiority of the S-ICD as compared with the T-ICD with respect to the time from device implantation to the first occurrence of a primary endpoint event. The primary analysis was the modified intention-to-treat (ITT) cohort (i.e. patients were analyzed in accordance to the treatment group to which they were originally assigned, regardless of withdrawals, losses to follow-up, or crossovers). Patients who did not receive a device and patients who proved ineligible for 1 of the treatments due to incomplete or inadequate screening were excluded from this analysis. In the as-treated cohort, patients were analyzed in the group of the specific ICD type which they received at initial implantation regardless of randomization result, withdrawals, losses to follow-up, or crossovers. The noninferiority margin for the upper boundary of the 95% CI for the HR was set at 1.45. The trial's main results are summarized in Tables 12 to 14. The S-ICD was noninferior to the T-ICD on the composite endpoint of device-related complications and inappropriate shocks. The HR for the primary endpoint was 0.99 (95% CI, 0.71 to 1.39; noninferiority margin, 1.45; $p=.01$ for noninferiority; $p=.95$ for superiority). Results for the modified ITT analysis and as-treated analysis did not differ. There were more device-related complications in the T-ICD group and more inappropriate shocks in the S-ICD group, but the trial was not powered for these endpoints. Secondary endpoints and mortality results are summarized in Table 13. There were more deaths from any cause in the S-ICD group than in the T-ICD group (16.4% vs. 13.1%; HR, 1.23; 95% CI, 0.89 to 1.70), but the number of SCDs did not differ between groups (18 in each group). There were more appropriate shocks in the S-ICD group (19.2% vs. 11.5%; HR, 1.52; 95% CI, 1.08 to 2.12). Other secondary endpoints did not differ between the groups.

While the rate of SCD in the PRAETORIAN trial was low (18 patients in each group), the number of overall deaths was 151, and actually occurred more frequently than the composite outcome (Table 13). The HR for all-cause mortality was 1.23 (95% CI, 0.89 to 1.70). The PRAETORIAN trial investigators conducted competing risks analyses to account for discontinuation of follow-up before the primary endpoint had occurred in (1) the modified ITT population with competing risk of death, and (2) the true ITT population with competing risk of death and discontinuation of follow-up. These analyses led to consistent estimates of the HR (and 95% CI) for the primary endpoint.

Device and lead complications occurred more frequently in the T-ICD group (Table 14).

Table 11. PRAETORIAN Trial Characteristics

| Study | Countries | Sites | Dates | Participants | Interventions | Primary Endpoint Definitions |
|---|-----------------------|-------|---------------------------------|--|---------------------------------|--|
| PRAETORIAN | | | | | Active Comparator | |
| Knops et al (2020)⁸⁷, | | | | | | |
| | Europe (92.4%) and US | 39 | March 2011 through January 2017 | Eligibility: 18 years and older Class I or IIa indication for primary or secondary prevention, according to professional society guidelines. Exclusions: Previous ICD implantation, unsuitability for S-ICD therapy according to QRS-T-wave sensing analysis, and indications for either bradycardia pacing or biventricular pacing. | S-ICD (n = 426) T-ICD (n = 423) | Composite of device-related complications and inappropriate shocks. Inappropriate shocks were defined as shock therapy for anything else but VF or VT. For example, supraventricular tachycardia with fast ventricle response (including sinus tachycardia and atrial fibrillation), T-wave oversensing, detection of physiological- or other non-cardiac activity and lead- or device failure. Complications included: <ul style="list-style-type: none"> • device infection that led to the extraction of the lead or generator; • pocket hematoma that led to drainage, blood transfusion, or prolongation of hospitalization; • device-related thrombotic events; • pneumothorax or hemothorax that led to intervention or prolongation of hospitalization; • cardiac perforation or tamponade; • lead repositioning or replacement; • other complications related to the lead or generator that led to medical or surgical intervention. |

ICD: implantable cardioverter defibrillator; PRAETORIAN: Prospective, Randomized Comparison of Subcutaneous and Transvenous Implantable Cardioverter Defibrillator Therapy; S-ICD: subcutaneous implantable cardioverter defibrillator; T-ICD: transvenous implantable cardioverter defibrillator; VF: ventricular fibrillation; VT: ventricular tachycardia.

Table 12. PRAETORIAN Trial Results - Primary Composite Endpoint and Components

| Study | Endpoint (4-year cumulative incidence) | S-ICD (n = 426) | T-ICD (n = 423) | Hazard Ratio (95% CI) |
|---|--|-----------------|-----------------|---|
| PRAETORIAN | Primary composite endpoint (modified ITT analysis) | 68 (15.1%) | 68 (15.7%) | 0.99 (0.71 to 1.39) p = .01 for noninferiority; p = .95 for superiority |
| Knops et al (2020)⁸⁷, | Device-related complication | 31 (5.9%) | 44 (9.8%) | 0.69 (0.44 to 1.09) |
| | Inappropriate shock | 41 (9.7%) | 29 (7.3%) | 1.43 (0.89 to 2.30) |

| Study | Endpoint (4-year cumulative incidence) | S-ICD (n = 426) | T-ICD (n = 423) | Hazard Ratio (95% CI) |
|-------|--|-----------------|-----------------|-----------------------|
| | Primary composite endpoint (as-treated analysis) | 68/428 (15.9%) | 68/421 (16.2%) | 0.98 (0.70 to 1.37) |

CI: confidence interval; ITT: intention-to-treat; PRAETORIAN: Prospective, Randomized Comparison of Subcutaneous and Transvenous Implantable Cardioverter Defibrillator Therapy; S-ICD: subcutaneous implantable cardioverter defibrillator; T-ICD: transvenous implantable cardioverter defibrillator.

Table 13. PRAETORIAN Trial Results - Secondary Endpoints

| Study | End Point | S-ICD (N=426) | T-ICD (N=423) | Hazard Ratio (95% CI) |
|--|--|---------------|---------------|-----------------------|
| PRAETORIAN | Death from any cause | 83 (16.4%) | 68 (13.1%) | 1.23 (0.89 to 1.70) |
| Knops et al (2020)⁸⁷ | Sudden cardiac death | 18 (4.2%) | 18 (4.3%) | |
| | Other cardiovascular death | 34 (8.0%) | 28 (6.6%) | |
| | Noncardiovascular death | 31 (7.3%) | 22 (5.2%) | |
| | Appropriate shock therapy | 83 (19.2%) | 57 (11.5%) | 1.52 (1.08 to 2.12) |
| | Antitachycardia pacing (appropriate) | 6 (0.6%) | 54 (12.9%) | |
| | Antitachycardia pacing (inappropriate) | 1 (0.3%) | 30 (7.2%) | |
| | Major adverse cardiac event | 64 (13.3%) | 80 (16.4%) | 0.80 (0.57 to 1.11) |
| | Hospitalization for heart failure | 79 (17.4%) | 74 (16.1%) | 1.08 (0.79 to 1.49) |
| | Crossover to other study device | 18 (4.3%) | 11 (2.7%) | 1.64 (0.77 to 3.47) |

CI: confidence interval; PRAETORIAN: Prospective, Randomized Comparison of Subcutaneous and Transvenous Implantable Cardioverter Defibrillator Therapy; S-ICD: subcutaneous implantable cardioverter defibrillator; T-ICD: transvenous implantable cardioverter defibrillator.

Table 14. PRAETORIAN Trial Results - Specific Complications

| Study | Endpoint | S-ICD (N=426) | T-ICD (N=423) |
|--|--|--------------------|--------------------|
| PRAETORIAN | Complications within the first 30 days | 3.8% | 4.7% |
| Knops et al (2020)⁸⁷ | Lead-related complications | 1.4% | 6.6% |
| | Device-related complications | 31 (5.9%) | 44 (9.8%) |
| | Infection | 4 (1 lead-related) | 8 (5 lead-related) |
| | Bleeding | 8 | 2 |
| | Thrombotic event | 1 | 2 |
| | Pneumothorax | 0 | 4 |
| | Lead perforation | 0 | 4 |
| | Tamponade | 0 | 2 |
| | Lead repositioning | 2 | 7 |
| | Other lead or device complication | 19 | 20 |
| | Lead replacement | 3 | 9 |
| | Device malfunction | 4 | 6 |
| | Sensing issues | 4 | 0 |
| | Pacing indication | 5 | 1 |
| | Implantation failure | 0 | 3 |
| | Defibrillation test failure | 3 | 0 |
| | Pain or discomfort | 2 | 3 |

ICD: implantable cardioverter defibrillator; PRAETORIAN: Prospective, Randomized Comparison of Subcutaneous and Transvenous Implantable Cardioverter Defibrillator Therapy; S-ICD: subcutaneous implantable cardioverter defibrillator; T-ICD: transvenous implantable cardioverter defibrillator.

Study relevance, design, and conduct limitations of PRAETORIAN are summarized in Tables 15 and 16. The choice of a composite primary endpoint poses several challenges to interpreting the results of PRAETORIAN. In PRAETORIAN, the components of the composite endpoint were discordant; device-related complications were expected to favor S-ICD and inappropriate shocks were expected to favor T-ICD. The timing of the components of the composite outcome assessment is important in interpreting the study results and explaining expected treatment results to patients. Early benefit could favor 1 treatment over another, and results could change with longer follow-up. This is an important point to consider when assessing complications such as lead failure, which continue to increase over the life of the device. Additionally, because the composite was not used in earlier trials of the active comparator, there is no historical data on which to derive the expected performance of the active control. The inappropriate shock rate was based on results from the MADIT-RT trial, which compared programmed high-rate or delayed T-ICD therapy, and the expected rate of complications was based on results from MADIT-RT and the SCD-HeFT trial, which compared amiodarone to T-ICD. To estimate the expected event rate in PRAETORIAN, the researchers combined these 2 endpoints to arrive at the expected 17.2% event rate for the composite primary outcome. The study authors do not cite any previous RCTs that used the composite endpoint of complications and inappropriate shocks. All-cause mortality was a primary endpoint in several previous RCTs of T-ICD. However, the PRAETORIAN trial protocol (2012) noted that all-cause mortality was not chosen as the primary endpoint because “mortality event rates in both groups are presumed to be low, leading to an extremely large trial size if this would serve as a primary endpoint.” The protocol also states that safety and efficacy of the S-ICD have been demonstrated in earlier trials and that the composite endpoint was “preferred above all-cause mortality, as practical, reasonably achievable, and pertinent to most cardiologists.”

Another major limitation of PRAETORIAN was that the median 48-month follow-up was not long enough to determine complications over the life of the device. In fact, the PRAETORIAN study authors note in their discussion, “longer-term follow-up of this cohort will be important because the incidence of lead-related complications increases over time with the transvenous ICD and because battery longevity is a limiting factor for the subcutaneous ICD.” Five-year data from the S-ICD PAS should provide more information on longer-term adverse events such as lead failure and need for device replacement.

Quality of life data from PRAETORIAN were collected but have not yet been published. These data could shed light on the relative importance to patients of adverse events such as inappropriate shocks and device replacement, especially if quality of life data were reported by subgroups of patients who experienced shocks. For example, these data might indicate that inappropriate shocks are so distressing to patients that they outweigh any potential benefits of S-ICDs.

Finally, the underenrollment of women in the trial (19.7%) potentially limits the applicability of its results, although a subgroup analysis by sex was consistent with the primary analysis on the composite endpoint (HR in women 0.65; 95% CI, 0.28 to 1.47).

Table 15. Study Relevance Limitations

| Study | Population ^a | Intervention ^b | Comparator ^c | Outcomes ^d | Duration of Follow-up ^e |
|----------------------------------|-------------------------|---------------------------|-------------------------|--|---|
| PRAETORIAN | 4. Women underenrolled | | | 6. Composite endpoint with discordant outcomes | 2. 4-year median follow-up not sufficient to assess complications over the life of the device |
| Knops et al (2020) ⁸⁷ | (19.7%) | | | | |

The study limitations stated in this table are those notable in the current review; this is not a comprehensive gaps assessment.

^a Population key: 1. Intended use population unclear; 2. Study population is unclear; 3. Study population not representative of intended use; 4. Enrolled populations do not reflect relevant diversity; 5. Other.

^b Intervention key: 1. Not clearly defined; 2. Version used unclear; 3. Delivery not similar intensity as comparator; 4. Not the intervention of interest (e.g., proposed as an adjunct but not tested as such); 5. Other.

^c Comparator key: 1. Not clearly defined; 2. Not standard or optimal; 3. Delivery not similar intensity as

intervention; 4. Not delivered effectively; 5. Other.

^d Outcomes key: 1. Key health outcomes not addressed; 2. Physiologic measures, not validated surrogates; 3. Incomplete reporting of harms; 4. Not establish and validated measurements; 5. Clinically significant difference not prespecified; 6. Clinically significant difference not supported; 7. Other.

^e Follow-Up key: 1. Not sufficient duration for benefit; 2. Not sufficient duration for harms; 3. Other.

Table 16. Study Design and Conduct Limitations

| Study | Allocation ^a | Blinding ^b | Selective Reporting ^c | Data Completeness ^d | Power ^e | Statistical ^f |
|----------------------------------|-------------------------|--|--|--------------------------------|--------------------|--|
| PRAETORIAN | | | | | | |
| Knops et al (2020) ⁸⁷ | | 2. Clinical-events committee was not blinded to treatment assignment | 2. Quality of life data collected but not yet published. | | | 5. Rationale for choice of noninferiority margin unclear |

The study limitations stated in this table are those notable in the current review; this is not a comprehensive gaps assessment.

^a Allocation key: 1. Participants not randomly allocated; 2. Allocation not concealed; 3. Allocation concealment unclear; 4. Inadequate control for selection bias; 5. Other.

^b Blinding key: 1. Participants or study staff not blinded; 2. Outcome assessors not blinded; 3. Outcome assessed by treating physician; 4. Other.

^c Selective Reporting key: 1. Not registered; 2. Evidence of selective reporting; 3. Evidence of selective publication; 4. Other.

^d Data Completeness key: 1. High loss to follow-up or missing data; 2. Inadequate handling of missing data; 3. High number of crossovers; 4. Inadequate handling of crossovers; 5. Inappropriate exclusions; 6. Not intent to treat analysis (per protocol for noninferiority trials); 7. Other.

^e Power key: 1. Power calculations not reported; 2. Power not calculated for primary outcome; 3. Power not based on clinically important difference; 4. Other.

^f Statistical key: 1. Analysis is not appropriate for outcome type: (a) continuous; (b) binary; (c) time to event; 2. Analysis is not appropriate for multiple observations per patient; 3. Confidence intervals and/or p values not reported; 4. Comparative treatment effects not calculated; 5. Other.

Comparative Observational Studies

Several observational studies have directly compared T-ICD to S-ICD. These studies are briefly described in Table 17. All studies were performed in the U.S. and/or Europe. Nonrandomized controlled studies have reported success rates in terminating laboratory-induced VF that are similar to T-ICD. However, there is scant evidence on comparative clinical outcomes of both types of ICD over longer periods. Adverse event rates are uncertain, with variable rates reported.

Table 17. Summary of Observational Comparative Studies of S-ICD and T-ICD

| Study | Study Type | N | Follow-Up | Results | | | |
|---------------------------------------|---|------------------------|--------------------|---|--|--|----------|
| | | | | Outcomes | T-ICD | S-ICD | DC T-ICD |
| Mithani et al (2018) ⁸⁸ | Matching based on dialysis status, sex, age | 182 (91 matched pairs) | 180 d | <ul style="list-style-type: none"> Inappropriate shocks Infection requiring explant Death from all causes Total with adverse event or death | <ul style="list-style-type: none"> 2.2% 1.1% 2.2% 7.7% | <ul style="list-style-type: none"> 1.1% 3.3% 2.2% 5.5% | |
| Honarbaksh et al (2017) ⁸⁹ | Propensity matched case-control | 138 (69 matched pairs) | 32 mo ^a | <ul style="list-style-type: none"> Total device-related complications Infections Inappropriate shocks Failure to cardiovert VA | <ul style="list-style-type: none"> 29% 5.8% 8.7% 1.4% | <ul style="list-style-type: none"> 9% 1.4% 4.3% 1.4% | |
| Kobe et al (2017) ⁹⁰ | Sex- and age-matched case-control | 120 (60 pairs); 84 | 942 d vs. 622 d | <ul style="list-style-type: none"> Posttraumatic stress disorder Major depression | <ul style="list-style-type: none"> 14.3% 9.5% | <ul style="list-style-type: none"> 14.3% 4.8% | |

| Study | Study Type | N | Follow-Up | Results |
|---|---|----------------------------|--------------------|--|
| | | pairs analyzed | | <ul style="list-style-type: none"> SF-12 physical well-being score <ul style="list-style-type: none"> • 40 • 52 SF-12 mental well-being score <ul style="list-style-type: none"> • 47 • 52 |
| Pedersen et al (2016)⁹¹ | Retrospective analysis of propensity-matched cohort | 334 (167 matched pairs) | 6 mo | <ul style="list-style-type: none"> SF-12 physical well-being score <ul style="list-style-type: none"> • 43 • 45 SF-12 mental well-being score <ul style="list-style-type: none"> • 44 • 45 |
| Brouwer et al (2016)⁹² | Retrospective analysis of propensity-matched cohort | 280 (140 matched pairs) | 5 y | <ul style="list-style-type: none"> Overall complications <ul style="list-style-type: none"> • 18% Lead complications <ul style="list-style-type: none"> • 11.5% Non-lead complications <ul style="list-style-type: none"> • 2.2% • 3.6% Infections <ul style="list-style-type: none"> • 31% Appropriate ICD intervention (HR, 2.4; 95% CI, NR; p=.01) <ul style="list-style-type: none"> • 95% Inappropriate ICD intervention (HR, 1.3; 95% CI, NR; p=.42) <ul style="list-style-type: none"> • 21% • 96% Survival <ul style="list-style-type: none"> • 14% • 0.8% • 9.9% • 4.1% |
| Friedman et al (2016)⁹³ | Retrospective analysis of propensity-matched cohort from NCDR for ICD | 5760 (1920 matched groups) | NR | <ul style="list-style-type: none"> Any in-hospital complication <ul style="list-style-type: none"> • 0.6% • 0.1% Deaths <ul style="list-style-type: none"> • 0.9% • 0.2% Infections <ul style="list-style-type: none"> • 0.05% • 0.1% Lead dislodgements <ul style="list-style-type: none"> • 0.1% • 0.6% Pneumothorax <ul style="list-style-type: none"> • 0.2% • 0% • 0.3% |
| Kobe et al (2013)⁹⁴ | Sex- and age-matched case-control | 138 (69 matched pairs) | 217 d ^a | <ul style="list-style-type: none"> Pericardial effusion <ul style="list-style-type: none"> • 1 • 0 Successful termination of induced VF <ul style="list-style-type: none"> • 91% • 9 • 3 Appropriate shocks <ul style="list-style-type: none"> • 3 • 5 Inappropriate shocks <ul style="list-style-type: none"> • 0 • 3 • 5 |

CI: confidence interval; DC: dual chamber; HR: hazard ratio; ICD: implantable cardioverter defibrillator; NCDR: National Cardiovascular Data Registry; NR: not reported; SF-12: 12-Item Short-Form Health Survey; S-ICD: subcutaneous implantable cardioverter defibrillator; T-ICD: transvenous implantable cardioverter defibrillator; VA: ventricular arrhythmia; VF: ventricular fibrillation.

^a Mean.

Section Summary: Subcutaneous Implantable Cardioverter Defibrillators In Patients With No Contraindications to a Transvenous Implantable Cardioverter Defibrillator

The PRAETORIAN trial is the only RCT on the effect of an S-ICD with health outcomes. PRAETORIAN found that S-ICD was noninferior to T-ICD on a composite outcome of complications and inappropriate shock at 48 months (HR, 0.99; 95% CI, 0.71 to 1.39; noninferiority margin, 1.45; p=.01 for noninferiority; p=.95 for superiority). There were more device related complications in the T-ICD group and more inappropriate shocks in the S-ICD group, but the trial was not powered for these endpoints. There is uncertainty over the applicability and interpretation of PRAETORIAN based on the choice of a composite outcome with discordant results, unclear rationale for choice of the noninferiority margin, inadequate length of follow up to determine rates of complications, and lack of reporting of quality of life data. Comparative observational studies are insufficient to draw conclusions on whether there are small differences in efficacy between the 2 types of devices, and reported variable adverse event rates. Ongoing studies could provide additional evidence on complications and device safety over the longer term.

Supplemental Information

The purpose of the following information is to provide reference material. Inclusion does not imply endorsement or alignment with the evidence review conclusions.

Clinical Input 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.

2020 Medical Advisory Panel

In October 2020, the BCBSA Medical Advisory Panel (MAP) reviewed the evidence for individuals who need an implantable cardioverter defibrillator (ICD) and have no contraindication to transvenous ICD placement and agreed that for this indication, the evidence is insufficient to determine that the technology results in an improvement in the net health outcome.

2015 Input

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

2011 Input

In response to requests, input was received from 6 academic medical centers 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 placement, 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. Input emphasized the difficulty of prescribing strict time frames 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

Guidelines or position statements will be considered for inclusion in 'Supplemental Information' if they were issued by, or jointly by, a US professional society, an international society with US representation, or National Institute for Health and Care Excellence (NICE). Priority will be given to guidelines that are informed by a systematic review, include strength of evidence ratings, and include a description of management of conflict of interest.

American Heart Association/American College of Cardiology et al - Heart Failure (2022)

In 2022, the American Heart Association (AHA), American College of Cardiology (ACC), and the Heart Failure Society of America released a guideline for the management of heart failure.⁹⁵ This guideline includes ICD recommendations which are summarized in Table 18.

Table 18. Guideline for the Management of Heart Failure - Recommendations for ICDs

| Recommendation | COR | LOE |
|---|-----|-----|
| "In patients with nonischemic DCM or ischemic heart disease at least 40 days post-MI with LVEF \leq 35% and NYHA class I or II symptoms on chronic GDMT, who have | 1 | A |

| Recommendation | COR | LOE |
|--|------------|------|
| reasonable expectation of meaningful survival for >1 year, ICD therapy is recommended for primary prevention of SCD to reduce total mortality." | | |
| "A transvenous ICD provides high economic value in the primary prevention of SCD particularly when the patient's risk of death caused by ventricular arrhythmia is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient's burden of comorbidities and functional status." | | A |
| "In patients at least 40 days post-MI with LVEF \leq 30% and NYHA class I symptoms while receiving GDMT, who have reasonable expectation of meaningful survival for >1 year, ICD therapy is recommended for primary prevention of SCD to reduce total mortality." | 1 | B-R |
| "In patients with genetic arrhythmogenic cardiomyopathy with high-risk features of sudden death, with EF \leq 45%, implantation of ICD is reasonable to decrease sudden death." | 2a | B-NR |
| "For patients whose comorbidities or frailty limit survival with good functional capacity to <1 year, ICD and CRT-D are not indicated." | No benefit | C-LD |

A: high; B-NR: moderate, non-randomized; B-R: moderate, randomized; C-LD: limited data; COR: class of recommendation; CRT-D: cardiac resynchronization therapy with defibrillation; DCM: dilated cardiomyopathy; EF: ejection fraction; GDMT: guideline-directed management and therapy; ICD: implantable cardioverter defibrillator; LOE: level of evidence; LVEF: left ventricular ejection fraction; MI: myocardial infarction; NYHA: New York Heart Association; SCD: sudden cardiac death.

American Heart Association/American College of Cardiology et al - Hypertrophic Cardiomyopathy (2020)

In 2020, the AHA and ACC published a joint Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy.⁹⁶ Recommendations relevant to this review are summarized in Table 19.

Table 19. Patient Selection for ICD Placement in High-Risk Patients With Hypertrophic Cardiomyopathy

| Recommendation | COR | LOE |
|--|---------|------|
| For patients with HCM, and previous documented cardiac arrest or sustained ventricular tachycardia, ICD placement is recommended. | I | B-NR |
| For adult patients with HCM with 1 or more major risk factors for SCD, it is reasonable to offer an ICD. | 2a | B-NR |
| For children with HCM who have 1 or more conventional risk factors, ICD placement is reasonable after considering the relatively high complication rates of long-term ICD placement in younger patients. | 2a | B-NR |
| For patients 16 years and older with HCM and 1 or more major SCD risk factors, discussion of the estimated 5-year sudden death risk and mortality rates can be useful during the shared decision-making process for ICD placement. | 2a | B-NR |
| In patients with HCM without risk factors, ICD placement should not be performed. | 3: Harm | B-NR |
| In patients with HCM, ICD placement for the sole purpose of participation in competitive athletics should not be performed. | 3: Harm | B-NR |
| In patients with HCM who are receiving an ICD, either a single chamber transvenous ICD or a subcutaneous ICD is recommended after a shared decision-making discussion that takes into consideration patient preferences, lifestyle, and expected potential need for pacing for bradycardia or ventricular tachycardia termination. | I | B-NR |

B-NR: moderate, non-randomized; COR: class of recommendation; HCM: hypertrophic cardiomyopathy; ICD: implantable cardioverter defibrillator; LOE: level of evidence; SCD: sudden cardiac death.

American Heart Association/American College of Cardiology et al - Ventricular Arrhythmias and Prevention of Sudden Cardiac Death (2017)

The AHA, ACC, and Heart Rhythm Society (2017) published joint guidelines on the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death.⁹⁷ This guideline supersedes the 2008 guideline for device-based therapy of cardiac rhythm abnormalities⁹⁸, and the subsequent 2012 focused update.⁹⁹ The most up-to-date recommendations on the use of

transvenous ICD devices from the 2017 guidelines are presented in Tables 20 to 24. Table 25 summarizes the most up-to-date recommendations regarding S-ICDs.

Table 20. Recommendations on Use of ICDs as Secondary Prevention of SCD of Ischemic Heart Disease or Nonischemic Cardiomyopathy

| Recommendation | COR | LOE |
|--|-----|-------------|
| "In patients with ischemic heart disease, who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R) or stable sustained VT (LOE: B-NR) not due to reversible causes, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-R B-NR |
| "A transvenous ICD provides intermediate value in the secondary prevention of SCD particularly when the patient's risk of death due to a VA is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient's burden of comorbidities and functional status." | | B-R |
| "In patients with ischemic heart disease and unexplained syncope who have inducible sustained monomorphic VT on electrophysiological study, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-NR |
| "In patients resuscitated from SCA due to coronary artery spasm in whom medical therapy is ineffective or not tolerated, an ICD is reasonable if meaningful survival of greater than 1 year is expected." | IIa | B-NR |
| "In patients resuscitated from SCA due to coronary artery spasm, an ICD in addition to medical therapy may be reasonable if meaningful survival of greater than 1 year is expected." | IIb | B-NR |
| "In patients with NICM who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R) (1-4) or stable VT (LOE: B-NR) (5) not due to reversible causes, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-R B-NR |
| "In patients with NICM who experience syncope presumed to be due to VA and who do not meet indications for a primary prevention ICD, an ICD or an electrophysiological study for risk stratification for SCD can be beneficial if meaningful survival of greater than 1 year is expected." | IIa | B-NR |
| "In patients with arrhythmogenic right ventricular cardiomyopathy and an additional marker of increased risk of SCD (resuscitated SCA, sustained VT, significant ventricular dysfunction with RVEF or LVEF \leq 35%), an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-NR |
| "In patients with arrhythmogenic right ventricular cardiomyopathy and syncope presumed due to VA, an ICD can be useful if meaningful survival of greater than 1 year is expected." | IIa | B-NR |

B-NR: moderate, non-randomized; B-R: moderate, randomized; COR: class of recommendation; ICD: implantable cardioverter defibrillator; LOE: level of evidence; LVEF: left ventricular ejection fraction; NICM: nonischemic cardiomyopathy; RVEF: right ventricular ejection fraction; SCA: sudden cardiac arrest; SCD: sudden cardiac death; VA: ventricular arrhythmia; VF: ventricular fibrillation; VT: ventricular tachycardia.

Table 21. Recommendations on Use of ICDs as a Primary Prevention of Ischemic Heart Disease or Nonischemic Cardiomyopathy

| Recommendation | COR | LOE |
|--|-----|-----|
| "In patients with LVEF of 35% or less that is due to ischemic heart disease who are at least 40 days' post-MI and at least 90 days postrevascularization, and with NYHA class II or III HF despite GDMT, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | A |
| "In patients with LVEF of 30% or less that is due to ischemic heart disease who are at least 40 days' post-MI and at least 90 days postrevascularization, and with NYHA class I HF despite GDMT, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | A |
| "A transvenous ICD provides high value in the primary prevention of SCD particularly when the patient's risk of death due to a VA is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient's burden of comorbidities and functional status." | | B-R |
| "In patients with NSVT due to prior MI, LVEF of 40% or less and inducible sustained VT or VF at electrophysiological study, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-R |

| Recommendation | COR | LOE |
|---|------------------|------|
| "In nonhospitalized patients with NYHA class IV symptoms who are candidates for cardiac transplantation or an LVAD, an ICD is reasonable if meaningful survival of greater than 1 year is expected." | IIa | B-NR |
| "An ICD is not indicated for NYHA class IV patients with medication-refractory HF who are not also candidates for cardiac transplantation, an LVAD, or a CRT defibrillator that incorporates both pacing and defibrillation capabilities." | III ^a | C-EO |
| "In patients with NICM, HF with NYHA class II-III symptoms and an LVEF of 35% or less, despite GDMT, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | A |
| "In patients with NICM due to a <i>Lamc A/C</i> mutation who have 2 or more risk factors (NSVT, LVEF <45%, nonmissense mutation, and male sex), an ICD can be beneficial if meaningful survival of greater than 1 year is expected." | IIa | B-NR |
| "In patients with NICM, HF with NYHA class I symptoms and an LVEF of 35% or less, despite GDMT, an ICD may be considered if meaningful survival of greater than 1 year is expected." | IIb | B-R |
| "In patients with medication-refractory NYHA class IV HF who are not also candidates for cardiac transplantation, an LVAD, or a CRT defibrillator that incorporates both pacing and defibrillation capabilities, an ICD should not be implanted." | III ^a | C-EO |

A: high; B-NR: moderate, non-randomized; B-R: moderate, randomized; C-EO: consensus of expert opinion; CRT: cardiac resynchronization therapy; COR: class of recommendation; GDMT: guideline-directed management and therapy; HF: heart failure; ICD: implantable cardioverter defibrillator; LOE: level of evidence; LVAD: left ventricular assist device; LVEF: left ventricular ejection fraction; MI: myocardial infarction; NICM: nonischemic cardiomyopathy; NSVT: nonsustained ventricular tachycardia; NYHA: New York Heart Association; SCD: sudden cardiac death; VA: ventricular arrhythmia; VF: ventricular fibrillation; VT: ventricular tachycardia.
^a No benefit.

Table 22. Recommendations on Use of ICDs for HCM

| Recommendation | COR | LOE |
|---|------------------|----------------------|
| "In patients with HCM who have survived an SCA due to VT or VF, or have spontaneous sustained VT causing syncope or hemodynamic compromise, an ICD is recommended if meaningful survival of greater than 1 year is expected" | I | B-NR |
| "In patients with HCM and 1 or more of the following risk factors, an ICD is reasonable if meaningful survival of greater than 1 year is expected: | | |
| <ul style="list-style-type: none"> • Maximum LV wall thickness ≥ 30 mm (LOE: B-NR). • SCD in 1 or more first-degree relatives presumably caused by HCM (LOE: C-LD). • 1 or more episodes of unexplained syncope within the preceding 6 months (LOE: C-LD)" | IIa | B-NR C-LD C-LD |
| "In patients with HCM who have spontaneous NSVT (LOE: C-LD) or an abnormal blood pressure response with exercise (LOE: B-NR), who also have additional SCD risk modifiers or high risk features an ICD is reasonable if meaningful survival of greater than 1 year is expected" | IIa | B-NR C-LD |
| "In patients with HCM who have NSVT (LOE: B-NR) or an abnormal blood pressure response with exercise (LOE: B-NR) but do not have any other SCD risk modifiers, an ICD may be considered, but its benefit is uncertain." | IIb | B-NR B-NR |
| "In patients with an identified HCM genotype in the absence of SCD risk factors, an ICD should not be implanted" | III ^a | B-NR |

B-NR: moderate, non-randomized; C-LD: limited data; COR: class of recommendation; HCM: hypertrophic cardiomyopathy; ICD: implantable cardioverter defibrillator; LOE: level of evidence; LV: left ventricular; NSVT: nonsustained ventricular tachycardia; SCA: sudden cardiac arrest; SCD: sudden cardiac death; VF: ventricular fibrillation; VT: ventricular tachycardia.

^a No benefit.

Table 23. Recommendations on Use of ICDs for Cardiac Sarcoidosis

| Recommendation | COR | LOE |
|--|-----|------|
| "In patients with cardiac sarcoidosis who have sustained VT or are survivors of SCA or have an LVEF of 35% or less, an ICD is recommended, if meaningful survival of greater than 1 year is expected." | I | B-NR |
| "In patients with cardiac sarcoidosis and LVEF greater than 35% who have syncope and/or evidence of myocardial scar by cardiac MRI or positron emission tomographic (PET) scan, | IIa | B-NR |

| Recommendation | COR | LOE |
|--|-----|------|
| and/or have an indication for permanent pacing, implantation of an ICD is reasonable, provided that meaningful survival of greater than 1 year is expected." | | |
| "In patients with cardiac sarcoidosis and LVEF greater than 35%, it is reasonable to perform an electrophysiological study and to implant an ICD, if sustained VA is inducible, provided that meaningful survival of greater than 1 year is expected." | IIa | C-LD |
| "In patients with cardiac sarcoidosis who have an indication for permanent pacing, implantation of an ICD can be beneficial." | IIa | C-LD |

B-NR: moderate, non-randomized; C-LD: limited data; COR: class of recommendation; ICD: implantable cardioverter defibrillator; LOE: level of evidence; LVEF: left ventricular ejection fraction; MRI: magnetic resonance imaging; SCA: sudden cardiac arrest; VA: ventricular arrhythmia; VT: ventricular tachycardia.

Table 24. Recommendations on Use of ICDs for Other Conditions

| Recommendation | COR | LOE |
|---|-----|------|
| "In patients with HFrEF who are awaiting heart transplant and who otherwise would not qualify for an ICD (e.g., NYHA class IV and/or use of inotropes) with a plan to discharge home, an ICD is reasonable." | IIa | B-NR |
| "In patients with an LVAD and sustained VA, an ICD can be beneficial." | IIa | C-LD |
| "In patients with a heart transplant and severe allograft vasculopathy with LV dysfunction, an ICD may be reasonable if meaningful survival of greater than 1 year is expected." | IIb | B-NR |
| "In patients with neuromuscular disorders, primary and secondary prevention ICDs are recommended for the same indications as for patients with NICM if meaningful survival of greater than 1 year is expected" | I | B-NR |
| "In patients with Emery-Dreifuss and limb-girdle type IB muscular dystrophies with progressive cardiac involvement, an ICD is reasonable if meaningful survival of greater than 1 year is expected." | IIa | B-NR |
| "In patients with myotonic dystrophy type 1 with an indication for a permanent pacemaker, an ICD may be considered to minimize the risk of SCA from VT if meaningful survival of greater than 1 year is expected." | IIb | B-NR |
| "In patients with a cardiac channelopathy and SCA, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-NR |
| "In high-risk patients with symptomatic long QT syndrome in whom a beta blocker is ineffective or not tolerated, intensification of therapy with additional medications (guided by consideration of the particular long QT syndrome type), left cardiac sympathetic denervation, and/or an ICD is recommended." | I | B-NR |
| "In patients with catecholaminergic polymorphic VT and recurrent sustained VT or syncope, while receiving adequate or maximally tolerated beta blocker, treatment intensification with either combination medication therapy, left cardiac sympathetic denervation, and/or an ICD is recommended." | I | B-NR |
| "In patients with Brugada syndrome with spontaneous type 1 Brugada electrocardiographic pattern and cardiac arrest, sustained VA or a recent history of syncope presumed due to VA, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-NR |
| "In patients with early repolarization pattern on ECG and cardiac arrest or sustained VA, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-NR |
| "In patients with short QT syndrome who have a cardiac arrest or sustained VA, an ICD is recommended if meaningful survival greater than 1 year is expected." | I | B-NR |
| "In patients resuscitated from SCA due to idiopathic polymorphic VT or VF, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-NR |
| "For older patients and those with significant comorbidities, who meet indications for a primary prevention ICD, an ICD is reasonable if meaningful survival of greater than 1 year is expected." | IIa | B-NR |
| "In patients with adult congenital heart disease with SCA due to VT or VF in the absence of reversible causes, an ICD is recommended if meaningful survival of greater than 1 year is expected." | I | B-NR |
| "In patients with repaired moderate or severe complexity adult congenital heart disease with unexplained syncope and at least moderate ventricular dysfunction or marked hypertrophy, either ICD implantation or an electrophysiological study with ICD | IIa | B-NR |

| Recommendation | COR | LOE |
|--|-----|-----|
| implantation for inducible sustained VA is reasonable if meaningful survival of greater than 1 year is expected." | | |

B-NR: moderate, non-randomized; C-LD: limited data; COR: class of recommendation; ECG: electrocardiogram; HFrEF; heart failure with reduced ejection fraction; ICD: implantable cardioverter defibrillator; LOE: level of evidence; LV: left ventricle; LVAD: left ventricular assist device; NICM: nonischemic cardiomyopathy; NYHA: New York Heart Association; SCA: sudden cardiac arrest; VA: ventricular arrhythmia; VF: ventricular fibrillation; VT: ventricular tachycardia.

Table 25. Recommendations on Use of S-ICDs

| Recommendation | COR | LOE |
|--|------------------|------|
| "In patients who meet criteria for an ICD who have inadequate vascular access or are at high risk for infection, and in whom pacing for bradycardia or VT termination or as part of CRT is neither needed nor anticipated, a subcutaneous implantable cardioverter-defibrillator is recommended." | I | B-NR |
| "In patients who meet indication for an ICD, implantation of a subcutaneous implantable cardioverter-defibrillator is reasonable if pacing for bradycardia or VT termination or as part of CRT is neither needed nor anticipated." | IIa | B-NR |
| "In patients with an indication for bradycardia pacing or CRT, or for whom antitachycardia pacing for VT termination is required, a subcutaneous implantable cardioverter-defibrillator should not be implanted." | III ^a | B-NR |

B-NR: moderate, non-randomized; COR: class of recommendation; CRT: cardiac resynchronization therapy; ICD: implantable cardioverter defibrillator; LOE: level of evidence; VT: ventricular tachycardia.

^a Harm.

Heart Rhythm Society- Arrhythmogenic Cardiomyopathy (2019)

In 2019, the Heart Rhythm Society published a consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy.¹⁰⁰ Recommendations related to ICD risk stratification and placement decisions are shown in Table 26.

Table 26. Recommendations on Risk Stratification and ICD Decisions

| Recommendation | COR ¹ | LOE ² |
|--|------------------|------------------|
| In individuals with ARVC with hemodynamically tolerated sustained VT, an ICD is reasonable. | IIa | B-NR |
| ICD implantation is reasonable for individuals with ARVC and three major, two major and two minor, or one major and four minor risk factors for ventricular arrhythmia. | IIa | B-NR |
| ICD implantation may be reasonable for individuals with ARVC and two major, one major and two minor, or four minor risk factors for ventricular arrhythmia. | IIb | B-NR |
| In individuals with ACM with LVEF 35% or lower and NYHA class II-III symptoms and an expected meaningful survival of greater than 1 year, an ICD is recommended. | I | B-R |
| In individuals with ACM with LVEF 35% or lower and NYHA class I symptoms and an expected meaningful survival of greater than 1 year, an ICD is reasonable. | IIa | B-R |
| In individuals with ACM (other than ARVC) and hemodynamically tolerated VT, an ICD is recommended. | I | B-NR |
| In individuals with phospholamban cardiomyopathy and LVEF <45% or NSVT, an ICD is reasonable. | IIa | B-NR |
| In individuals with lamin A/C ACM and two or more of the following: LVEF <45%, NSVT, male sex, an ICD is reasonable. | IIa | B-NR |
| In individuals with FLNC ACM and an LVEF <45%, an ICD is reasonable. | IIa | C-LD |
| In individuals with lamin A/C ACM and an indication for pacing, an ICD with pacing capabilities is reasonable. | IIa | C-LD |

ACM: arrhythmogenic cardiomyopathy; ARVC: arrhythmogenic right ventricular cardiomyopathy; COR: Class of Recommendation; FLNC: filamin-C; HRS: Heart Rhythm Society; ICD: Implantable cardioverter defibrillator; LOE: Level of Evidence; LVEF: left ventricular ejection fraction; NSVT: nonsustained ventricular tachycardia; NYHA: New York Heart Association; VT: ventricular tachycardia.

¹ Class I: Strong; Class IIa: Moderate; Class IIb: Weak. ² B-R: Randomized; B-NR: nonrandomized; C-LD: limited data.

Heart Rhythm Society et al - Inherited Primary Arrhythmia Syndromes (2013)

The Heart Rhythm Society, the European Heart Rhythm Association, and the Asia-Pacific Heart Rhythm Society (2013) issued a consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes, which included recommendations on ICD use in patients with long QT syndrome, Brugada syndrome, catecholaminergic polymorphic ventricular tachycardia, and short QT syndrome (Table 27).¹⁰¹

Table 27. Recommendations on ICDs in Inherited Primary Arrhythmia Syndromes

| Recommendation | COR |
|---|------------------|
| <i>Long QT syndrome</i> | |
| ICD implantation is recommended for patients with a diagnosis of LQTS who are survivors of a cardiac arrest. | I |
| ICD implantation can be useful in patients with a diagnosis of LQTS who experience recurrent syncope events while on beta-blocker therapy. | IIa |
| Except under special circumstances, ICD implantation is not indicated in asymptomatic LQTS patients who have not been tried on beta-blocker therapy. | III ^a |
| <i>Brugada syndrome</i> | |
| ICD implantation is recommended in patients with a diagnosis of BrS who: | |
| • Are survivors of a cardiac arrest and/or | I |
| • Have documented spontaneous sustained VT with or without syncope. | |
| 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. | IIa |
| ICD implantation may be considered in patients with a diagnosis of BrS who develop VF during programmed electrical stimulation (inducible patients). | IIb |
| 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. | III ^a |
| <i>Catecholaminergic polymorphic ventricular tachycardia</i> | |
| 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. | I |
| ICD as a stand alone therapy is not indicated in an asymptomatic patient with a diagnosis of CPVT. | III ^a |
| <i>Short QT syndrome</i> | |
| 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. | I |
| ICD implantation may be considered in asymptomatic patients with a diagnosis of SQTS and a family history of sudden cardiac death. | IIb |

BrS: Brugada syndrome; COR: class of recommendation; CPVT: catecholaminergic polymorphic ventricular tachycardia; ECG: electrocardiogram; HRS: Heart Rhythm Society; ICD: implantable cardioverter defibrillator; LQTS: long QT syndrome; SCD: sudden cardiac death; SQTS: short QT syndrome; VF: ventricular fibrillation; VT: ventricular tachycardia.

^a Not recommended.

Heart Rhythm Society - Cardiac Sarcoidosis (2014)

In 2014, the Heart Rhythm Society published a consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis, including recommendations for ICD implantation in patients with cardiac sarcoidosis (Table 28).³³ The writing group concluded that although there are few data specific to ICD use in patients with cardiac sarcoidosis, data from the major primary and secondary prevention ICD trials were relevant to this population and recommendations from the general device guideline documents apply to this population.

Table 28. Recommendations for ICD Implantation in Patients with Cardiac Sarcoidosis

| Recommendation | COR ¹ |
|--|------------------|
| ICD implantation is recommended in patients with cardiac sarcoidosis and one or more of the following: | I |

- Spontaneous sustained ventricular arrhythmias, including prior cardiac arrest
- LVEF $\leq 35\%$, despite optimal medical therapy and a period of immunosuppression (if there is active inflammation).

ICD implantation can be useful in patients with cardiac sarcoidosis, independent of ventricular function, and one or more of the following:

- An indication for permanent pacemaker implantation; IIa
- Unexplained syncope or near-syncope, felt to be arrhythmic in etiology;
- Inducible sustained ventricular arrhythmias (>30 seconds of monomorphic VT or polymorphic VT) or clinically relevant VF.

ICD implantation may be considered in patients with LVEF in the range of 36%–49% and/or an RV ejection fraction <40%, despite optimal medical therapy for heart failure and a period of immunosuppression (if there is active inflammation). IIb

ICD implantation is not recommended in patients with no history of syncope, normal LVEF/RV ejection fraction, no LGE on CMR, a negative EP study, and no indication for permanent pacing. However, these patients should be closely followed for deterioration in ventricular function. ICD implantation is not recommended in patients with one or more of the following: III

- Incessant ventricular arrhythmias;
- Severe New York Heart Association class IV heart failure.

COR: Class of Recommendation; EP: electrophysiologic; ICD: implantable cardioverter defibrillator; LGE-CMR: late gadolinium-enhanced cardiovascular magnetic resonance; LOE: Level of Evidence; LVEF: left ventricular ejection fraction; RV: right ventricular.

¹Class I: Strong; Class IIa: Moderate; Class IIb: Weak.

Pediatric and Congenital Electrophysiology Society et al

The Pediatric and Congenital Electrophysiology Society and Heart Rhythm Society (2014) issued an expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease.¹⁰² The statement made the following recommendations on the use of ICD therapy in adults with congenital heart disease (Table 29).

Table 29. Recommendations on ICDs in the Management of CHD

| Recommendation | COR | LOE |
|---|-----|-----|
| 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. | I | B |
| ICD therapy is indicated in adults with CHD and spontaneous sustained ventricular tachycardia who have undergone hemodynamic and electrophysiologic evaluation. | I | B |
| ICD therapy is indicated in adults with CHD and a systemic left ventricular ejection fraction <35%, biventricular physiology, and NYHA class II or III symptoms. | I | B |
| 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. | IIa | B |
| 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. | IIb | 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. | Ib | 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. | Ib | B |
| ICD therapy may be considered for nonhospitalized adults with CHD awaiting heart transplantation. | Ib | 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. | Ib | C |

| Recommendation | COR | LOE |
|--|------------------|-----|
| Adults with CHD and advanced pulmonary vascular disease (Eisenmenger syndrome) are generally not considered candidates for ICD therapy. | III ^a | |
| 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. | III ^a | |

AV: atrioventricular ; CHD: congenital heart disease; COR: class of recommendation; ICD: implantable cardioverter defibrillator; LOE: level of evidence; NYHA: New York Heart Association.

^a Not recommended.

In 2021, the Pediatric and Congenital Electrophysiology Society and Heart Rhythm Society also issued an expert consensus statement on the indications and management of cardiovascular implantable electronic devices in pediatric patients.¹⁰³ Table 30 summarizes recommendations for ICD therapy from this statement.

Table 30. Recommendations for ICD Therapy in Pediatric Patients

| Recommendation | COR | LOE |
|--|---------------|------|
| ICD implantation is indicated for survivors of SCA due to VT/VF if completely reversible causes have been excluded and an ICD is considered to be more beneficial than alternative treatments that may significantly reduce the risk of SCA. | I | B-NR |
| ICD implantation may be considered for patients with sustained VT that cannot be adequately controlled with medication and/or catheter ablation. | 2b | C-EO |
| ICD therapy may be considered for primary prevention of SCD in patients with genetic cardiovascular diseases and risk factors for SCA or pathogenic mutations and family history of recurrent SCA. | 2b | C-EO |
| ICD therapy is not indicated for patients with incessant ventricular tachyarrhythmias due to risk of ICD storm. | 3: Harm | C-EO |
| ICD therapy is not indicated for patients with ventricular arrhythmias that are adequately treated with medication and/or catheter ablation. | 3: Harm | C-LD |
| ICD therapy is not indicated for patients who have an expected survival <1 year, even if they meet ICD implantation criteria specified in the above recommendations. | 3: Harm | C-EO |
| ICD implantation along with the use of beta-blockade is indicated for patients with a diagnosis of LQTS who are survivors of SCA. | I | B-NR |
| ICD implantation is indicated in LQTS patients with symptoms in whom beta-blockade is either ineffective or not tolerated and cardiac sympathetic denervation or other medications are not considered effective alternatives. | I | B-NR |
| ICD therapy may be considered for primary prevention in LQTS patients with established clinical risk factors and/or pathogenic mutations. | 2b | C-LD |
| ICD implantation is not indicated in asymptomatic LQTS patients who are deemed to be at low risk of SCA and have not been tried on beta-blocker therapy. | 3: Harm | C-LD |
| ICD implantation is indicated in patients with a diagnosis of CPVT who experience cardiac arrest or arrhythmic syncope despite maximally tolerated beta-blocker plus flecainide and/or cardiac sympathetic denervation. | I | C-LD |
| ICD implantation is reasonable in combination with pharmacologic therapy with or without cardiac sympathetic denervation when aborted SCA is the initial presentation of CPVT. Pharmacologic therapy and/or cardiac sympathetic denervation without ICD may be considered as an alternative. | 2a | C-LD |
| ICD therapy may be considered in CPVT patients with polymorphic/bidirectional VT despite optimal pharmacologic therapy with or without cardiac sympathetic denervation. | 2b | C-LD |
| ICD implantation is not indicated in asymptomatic patients with a diagnosis of CPVT. | 3: Harm | C-EO |
| ICD implantation is indicated in patients with a diagnosis of BrS who are survivors of SCA or have documented spontaneous sustained VT. | I | B-NR |
| ICD implantation is reasonable for patients with BrS with a spontaneous type I Brugada ECG pattern and recent syncope presumed due to ventricular arrhythmias. | 2a | B-NR |
| ICD implantation may be considered in patients with syncope presumed due to ventricular arrhythmias with a type I Brugada ECG pattern only with provocative medications. | 2b | C-EO |
| ICD implantation is not indicated in asymptomatic BrS patients in the absence of risk factors. | 3: No benefit | C-EO |

| Recommendation | COR | LOE |
|---|---------------|------|
| ICD implantation is indicated in patients with HCM who are survivors of SCA or have spontaneous sustained VT. | I | B-NR |
| For children with HCM who have ≥ 1 primary risk factors, including unexplained syncope, massive left ventricular hypertrophy, nonsustained VT, or family history of early HCM-related SCD, ICD placement is reasonable after considering the potential complications of long-term ICD placement. | 2a | B-NR |
| ICD implantation may be considered in patients with HCM without the above risk factors but with secondary risk factors for SCA such as extensive LGE cardiac MRI or systolic dysfunction. | 2b | B-NR |
| ICD implantation is not indicated in patients with an identified HCM genotype in the absence of known pediatric SCA risk factors. | 3: Harm | C-LD |
| ICD implantation is indicated in patients with ACM who have been resuscitated from SCA or sustained VT that is not hemodynamically tolerated. | I | B-NR |
| ICD implantation is reasonable in patients with ACM with hemodynamically tolerated sustained VT, syncope presumed due to ventricular arrhythmia, or an LVEF $\leq 35\%$. | 2a | B-NR |
| ICD implantation may be considered in patients with inherited ACM associated with increased risk of SCD based on an assessment of additional risk factors. | 2b | C-LD |
| ICD implantation is indicated in patients with NIDCM who either survive SCA or experience sustained VT not due to completely reversible causes. | I | B-NR |
| ICD implantation may be considered in patients with NIDCM and syncope or an LVEF $\leq 35\%$, despite optimal medical therapy. | 2b | C-LD |
| ICD implantation is not recommended in patients with medication-refractory advanced heart failure who are not cardiac transplantation or left ventricular assist device candidates. | 3: Harm | C-EO |
| ICD therapy is not indicated for patients with advanced heart failure who are urgently listed for cardiac transplantation and will remain in the hospital until transplantation, even if they meet ICD implantation criteria specified in the above recommendations. | 3: No benefit | C-EO |
| ICD implantation is indicated for CHD patients who are survivors of SCA after evaluation to define the cause of the event and exclude any completely reversible causes. | I | B-NR |
| ICD implantation is indicated for CHD patients with hemodynamically unstable sustained VT who have undergone hemodynamics and EP evaluation. | I | C-LD |
| ICD implantation is reasonable for CHD patients with systemic LVEF $< 35\%$ and sustained VT or presumed arrhythmogenic syncope. | 2a | C-LD |
| ICD implantation may be considered for CHD patients with spontaneous hemodynamically stable sustained VT who have undergone hemodynamic and EP evaluation. | 2b | C-EO |
| ICD implantation may be considered for CHD patients with unexplained syncope in the presence of ventricular dysfunction, nonsustained VT, or inducible ventricular arrhythmias at EP study. | 2b | C-LD |
| ICD implantation may be considered for CHD patients with a single or systemic right ventricular ejection fraction $\leq 35\%$, particularly in the presence of additional risk factors such as VT, arrhythmic syncope, or severe systemic AV valve insufficiency. | 2b | C-EO |

ACM: arrhythmogenic cardiomyopathy; AV: atrioventricular; B-NR: moderate, non-randomized; BrS: Brugada syndrome; C-EO: consensus of expert opinion; CHD: congenital heart disease; C-LD: limited data; COR: class of recommendation; CPVT: catecholaminergic polymorphic ventricular tachycardia; ECG: electrocardiogram; EP: electrophysiology; HCM: hypertrophic cardiomyopathy; ICD: implantable cardioverter defibrillator; LGE: late gadolinium-enhanced; LOE: level of evidence; LQTS: long QT syndrome; LVEF: left ventricular ejection fraction; MRI: magnetic resonance imaging; NIDCM: non-ischemic dilated cardiomyopathy; SCA: sudden cardiac arrest; SCD: sudden cardiac death; VF: ventricular fibrillation; VT: ventricular tachycardia.

U.S. Preventive Services Task Force Recommendations

Not applicable.

Medicare National Coverage

There is a National Coverage Determination for ICDs.¹⁰⁴ According to the most recent publication (effective February 15, 2018), Centers for Medicare and Medicaid Services will cover ICDs for the following patient indications:

1. Patients with a personal history of sustained ventricular tachycardia (VT) or cardiac arrest due to ventricular fibrillation (VF).
2. Patients with a prior myocardial infarction (MI) and a measured left ventricular ejection fraction (LVEF) ≤ 0.30 .
3. Patients who have severe ischemic dilated cardiomyopathy but no personal history of sustained VT or cardiac arrest due to VF, and have New York Heart Association (NYHA) Class II or III heart failure, LVEF $\leq 35\%$.
4. Patients who have severe non-ischemic dilated cardiomyopathy but no personal history of cardiac arrest or sustained VT, NYHA Class II or III heart failure, LVEF $\leq 35\%$, and been on optimal medical therapy for at least three (3) months.
5. Patients with documented familial, or genetic disorders with a high risk of life-threatening tachyarrhythmias (sustained VT or VF), to include, but not limited to, long QT syndrome or hypertrophic cardiomyopathy.
6. Patients with an existing ICD may receive an ICD replacement if it is required due to the end of battery life, Elective Replacement Indicator (ERI), or device/lead malfunction.

For each group:

1. Patients must be clinically stable (e.g., not in shock, from any etiology);
2. LVEF must be measured by echocardiography, radionuclide (nuclear medicine) imaging, cardiac magnetic resonance imaging (MRI), or catheter angiography;
3. Patients must not have:
 - Significant, irreversible brain damage; or,
 - Any disease, other than cardiac disease (e.g., cancer, renal failure, liver failure) associated with a likelihood of survival less than one (1) year; or,
 - Supraventricular tachycardia such as atrial fibrillation with a poorly controlled ventricular rate.

Ongoing and Unpublished Clinical Trials

Some unpublished trials that may influence this review are listed in Table 31.

Table 31. Summary of Key Trials

| NCT No. | Trial Name | Planned Enrollment | Completion Date |
|--------------------------|---|--------------------|----------------------------------|
| <i>Ongoing</i> | | | |
| NCT02845531 | Implantable Cardioverter Defibrillator Versus Optimal Medical Therapy In Patients With Variant Angina Manifesting as Aborted Sudden Cardiac Death (VARIANT ICD) | 140 | Jun 2023 |
| NCT01296022 ^a | Randomized Trial to Study the Efficacy and Adverse Effects of the Subcutaneous and Transvenous Implantable Cardioverter Defibrillator (ICD) in Patients With a Class I or IIa Indication for ICD Without an Indication for Pacing | 850 | Dec 2023 (extended follow-up) |
| NCT01085435 ^a | Evaluation of Factors Impacting Clinical Outcome and Cost Effectiveness of the S-ICD (The EFFORTLESS S-ICD Registry) | 994 | Dec 2023 |
| NCT02787785 ^a | Multicenter Automatic Defibrillator Implantation Trial With Subcutaneous Implantable Cardioverter Defibrillator (MADIT S-ICD) | 40 | Oct 2023 |
| <i>Unpublished</i> | | | |
| NCT01736618 ^a | Subcutaneous Implantable Cardioverter Defibrillator System Post Approval Study (UNTOUCHED) | 1766 | Oct 2021 |
| NCT00673842 ^a | Risk Estimation Following Infarction Noninvasive Evaluation - ICD Efficacy | 1000 | Dec 2021 |

NCT: national clinical trial.

^a Denotes industry-sponsored or cosponsored trial.

References

1. Rome BN, Kramer DB, Kesselheim AS. FDA approval of cardiac implantable electronic devices via original and supplement premarket approval pathways, 1979–2012. *JAMA*. Jan 2014; 311(4): 385–91. PMID 24449317
2. Food and Drug Administration. Medtronic Recalls Evera, Viva, Brava, Claria, Amplia, Compia, and Visia Implantable Cardioverter Defibrillators (ICDs) and Cardiac Resynchronization Therapy (CRT-Ds) Due to Risk of Shortened Battery Life. April 12, 2021. <https://www.fda.gov/medical-devices/medical-device-recalls/medtronic-recalls-evera-viva-brava-claria-amplia-compia-and-visia-implantable-cardioverter>. Accessed April 1, 2023.
3. Food and Drug Administration. Medtronic Recalls Cobalt XT, Cobalt and Crome ICDs and CRT-Ds for Risk that Devices May Issue a Short Circuit Alert and Deliver Reduced Energy Shock During High Voltage Therapy. August 19, 2022. <https://www.fda.gov/medical-devices/medical-device-recalls/medtronic-recalls-cobalt-xt-cobalt-and-crome-icds-and-crt-ds-risk-devices-may-issue-short-circuit>. Accessed April 2, 2023.
4. Food and Drug Administration. Boston Scientific Recalls EMBLEM S-ICD Subcutaneous Electrode (Model 3501) Due to Risk of Fractures. February 10, 2021. <https://www.fda.gov/medical-devices/medical-device-recalls/boston-scientific-recalls-emblem-s-icd-subcutaneous-electrode-model-3501-due-risk-fractures>. Accessed April 3, 2023.
5. Moss AJ, Hall WJ, Cannom DS, et al. Improved survival with an implanted defibrillator in patients with coronary disease at high risk for ventricular arrhythmia. Multicenter Automatic Defibrillator Implantation Trial Investigators. *N Engl J Med*. Dec 26 1996; 335(26): 1933–40. PMID 8960472
6. Moss AJ, Zareba W, Hall WJ, et al. Prophylactic implantation of a defibrillator in patients with myocardial infarction and reduced ejection fraction. *N Engl J Med*. Mar 21 2002; 346(12): 877–83. PMID 11907286
7. Bigger JT. Prophylactic use of implanted cardiac defibrillators in patients at high risk for ventricular arrhythmias after coronary-artery bypass graft surgery. Coronary Artery Bypass Graft (CABG) Patch Trial Investigators. *N Engl J Med*. Nov 27 1997; 337(22): 1569–75. PMID 9371853
8. Buxton AE, Lee KL, Fisher JD, et al. A randomized study of the prevention of sudden death in patients with coronary artery disease. Multicenter Unsustained Tachycardia Trial Investigators. *N Engl J Med*. Dec 16 1999; 341(25): 1882–90. PMID 10601507
9. Bardy GH, Lee KL, Mark DB, et al. Amiodarone or an implantable cardioverter-defibrillator for congestive heart failure. *N Engl J Med*. Jan 20 2005; 352(3): 225–37. PMID 15659722
10. Hohnloser SH, Kuck KH, Dorian P, et al. Prophylactic use of an implantable cardioverter-defibrillator after acute myocardial infarction. *N Engl J Med*. Dec 09 2004; 351(24): 2481–8. PMID 15590950
11. Steinbeck G, Andresen D, Seidl K, et al. Defibrillator implantation early after myocardial infarction. *N Engl J Med*. Oct 08 2009; 361(15): 1427–36. PMID 19812399
12. Raviele A, Bongioni MG, Brignole M, et al. Early EPS/ICD strategy in survivors of acute myocardial infarction with severe left ventricular dysfunction on optimal beta-blocker treatment. The BEta-blocker STRategy plus ICD trial. *Europace*. Jul 2005; 7(4): 327–37. PMID 16028343
13. Kadish A, Dyer A, Daubert JP, et al. Prophylactic defibrillator implantation in patients with nonischemic dilated cardiomyopathy. *N Engl J Med*. May 20 2004; 350(21): 2151–8. PMID 15152060
14. Bristow MR, Saxon LA, Boehmer J, et al. Cardiac-resynchronization therapy with or without an implantable defibrillator in advanced chronic heart failure. *N Engl J Med*. May 20 2004; 350(21): 2140–50. PMID 15152059

15. Strickberger SA, Hummel JD, Bartlett TG, et al. Amiodarone versus implantable cardioverter-defibrillator: randomized trial in patients with nonischemic dilated cardiomyopathy and asymptomatic nonsustained ventricular tachycardia--AMIOVIRT. *J Am Coll Cardiol*. May 21 2003; 41(10): 1707-12. PMID 12767651
16. Bänsch D, Antz M, Boczor S, et al. Primary prevention of sudden cardiac death in idiopathic dilated cardiomyopathy: the Cardiomyopathy Trial (CAT). *Circulation*. Mar 26 2002; 105(12): 1453-8. PMID 11914254
17. Køber L, Thune JJ, Nielsen JC, et al. Defibrillator Implantation in Patients with Nonischemic Systolic Heart Failure. *N Engl J Med*. Sep 29 2016; 375(13): 1221-30. PMID 27571011
18. Woods B, Hawkins N, Mealing S, et al. Individual patient data network meta-analysis of mortality effects of implantable cardiac devices. *Heart*. Nov 2015; 101(22): 1800-6. PMID 26269413
19. Wolff G, Lin Y, Karathanos A, et al. Implantable cardioverter/defibrillators for primary prevention in dilated cardiomyopathy post-DANISH: an updated meta-analysis and systematic review of randomized controlled trials. *Clin Res Cardiol*. Jul 2017; 106(7): 501-513. PMID 28213711
20. Stavrakis S, Asad Z, Reynolds D. Implantable Cardioverter Defibrillators for Primary Prevention of Mortality in Patients With Nonischemic Cardiomyopathy: A Meta-Analysis of Randomized Controlled Trials. *J Cardiovasc Electrophysiol*. Jun 2017; 28(6): 659-665. PMID 28316104
21. Akel T, Lafferty J. Implantable cardioverter defibrillators for primary prevention in patients with nonischemic cardiomyopathy: A systematic review and meta-analysis. *Cardiovasc Ther*. Jun 2017; 35(3). PMID 28129469
22. Golwala H, Bajaj NS, Arora G, et al. Implantable Cardioverter-Defibrillator for Nonischemic Cardiomyopathy: An Updated Meta-Analysis. *Circulation*. Jan 10 2017; 135(2): 201-203. PMID 27993908
23. Wasiak M, Tajstra M, Kosior D, et al. An implantable cardioverter-defibrillator for primary prevention in non-ischemic cardiomyopathy: A systematic review and meta-analysis. *Cardiol J*. 2023; 30(1): 117-124. PMID 33843044
24. Earley A, Persson R, Garlitski AC, et al. Effectiveness of implantable cardioverter defibrillators for primary prevention of sudden cardiac death in subgroups a systematic review. *Ann Intern Med*. Jan 21 2014; 160(2): 111-21. PMID 24592496
25. Fontenla A, Martínez-Ferrer JB, Alzueta J, et al. Incidence of arrhythmias in a large cohort of patients with current implantable cardioverter-defibrillators in Spain: results from the UMBRELLA Registry. *Europace*. Nov 2016; 18(11): 1726-1734. PMID 26705555
26. Schinkel AF, Vriesendorp PA, Sijbrands EJ, et al. Outcome and complications after implantable cardioverter defibrillator therapy in hypertrophic cardiomyopathy: systematic review and meta-analysis. *Circ Heart Fail*. Sep 01 2012; 5(5): 552-9. PMID 22821634
27. Magnusson P, Gadler F, Liv P, et al. Hypertrophic Cardiomyopathy and Implantable Defibrillators in Sweden: Inappropriate Shocks and Complications Requiring Surgery. *J Cardiovasc Electrophysiol*. Oct 2015; 26(10): 1088-94. PMID 26178879
28. Horner JM, Kinoshita M, Webster TL, et al. Implantable cardioverter defibrillator therapy for congenital long QT syndrome: a single-center experience. *Heart Rhythm*. Nov 2010; 7(11): 1616-22. PMID 20816872
29. Hernandez-Ojeda J, Arbelo E, Borrás R, et al. Patients With Brugada Syndrome and Implanted Cardioverter-Defibrillators: Long-Term Follow-Up. *J Am Coll Cardiol*. Oct 17 2017; 70(16): 1991-2002. PMID 29025556
30. Conte G, Sieira J, Ciconte G, et al. Implantable cardioverter-defibrillator therapy in Brugada syndrome: a 20-year single-center experience. *J Am Coll Cardiol*. Mar 10 2015; 65(9): 879-88. PMID 25744005
31. Dores H, Reis Santos K, Adragão P, et al. Long-term prognosis of patients with Brugada syndrome and an implanted cardioverter-defibrillator. *Rev Port Cardiol*. Jun 2015; 34(6): 395-402. PMID 26028488

32. Roses-Noguer F, Jarman JW, Clague JR, et al. Outcomes of defibrillator therapy in catecholaminergic polymorphic ventricular tachycardia. *Heart Rhythm*. Jan 2014; 11(1): 58-66. PMID 24120999
33. Birnie DH, Sauer WH, Bogun F, et al. HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. *Heart Rhythm*. Jul 2014; 11(7): 1305-23. PMID 24819193
34. Plitt A, Dorbala S, Albert MA, et al. Cardiac sarcoidosis: case report, workup, and review of the literature. *Cardiol Ther*. Dec 2013; 2(2): 181-97. PMID 25135396
35. Mantini N, Williams B, Stewart J, et al. Cardiac sarcoid: a clinician's review on how to approach the patient with cardiac sarcoid. *Clin Cardiol*. 2012; 35(7): 410-5. PMID 22499155
36. Berul CI, Van Hare GF, Kertesz NJ, et al. Results of a multicenter retrospective implantable cardioverter-defibrillator registry of pediatric and congenital heart disease patients. *J Am Coll Cardiol*. Apr 29 2008; 51(17): 1685-91. PMID 18436121
37. Silka MJ, Kron J, Dunnigan A, et al. Sudden cardiac death and the use of implantable cardioverter-defibrillators in pediatric patients. *The Pediatric Electrophysiology Society. Circulation*. Mar 1993; 87(3): 800-7. PMID 8443901
38. Alexander ME, Cecchin F, Walsh EP, et al. Implications of implantable cardioverter defibrillator therapy in congenital heart disease and pediatrics. *J Cardiovasc Electrophysiol*. Jan 2004; 15(1): 72-6. PMID 15028076
39. Lewandowski M, Sterlinski M, Maciag A, et al. Long-term follow-up of children and young adults treated with implantable cardioverter-defibrillator: the authors' own experience with optimal implantable cardioverter-defibrillator programming. *Europace*. Sep 2010; 12(9): 1245-50. PMID 20650939
40. Antiarrhythmics versus Implantable Defibrillators (AVID) Investigators. A comparison of antiarrhythmic-drug therapy with implantable defibrillators in patients resuscitated from near-fatal ventricular arrhythmias. *N Engl J Med*. Nov 27 1997; 337(22): 1576-83. PMID 9411221
41. Kuck KH, Cappato R, Siebels J, et al. Randomized comparison of antiarrhythmic drug therapy with implantable defibrillators in patients resuscitated from cardiac arrest : the Cardiac Arrest Study Hamburg (CASH). *Circulation*. Aug 15 2000; 102(7): 748-54. PMID 10942742
42. Connolly SJ, Gent M, Roberts RS, et al. Canadian implantable defibrillator study (CIDS) : a randomized trial of the implantable cardioverter defibrillator against amiodarone. *Circulation*. Mar 21 2000; 101(11): 1297-302. PMID 10725290
43. Nademanee K, Veerakul G, Mower M, et al. Defibrillator Versus beta-Blockers for Unexplained Death in Thailand (DEBUT): a randomized clinical trial. *Circulation*. May 06 2003; 107(17): 2221-6. PMID 12695290
44. Wever EF, Hauer RN, van Capelle FL, et al. Randomized study of implantable defibrillator as first-choice therapy versus conventional strategy in postinfarct sudden death survivors. *Circulation*. Apr 15 1995; 91(8): 2195-203. PMID 7697849
45. Lee DS, Green LD, Liu PP, et al. Effectiveness of implantable defibrillators for preventing arrhythmic events and death: a meta-analysis. *J Am Coll Cardiol*. May 07 2003; 41(9): 1573-82. PMID 12742300
46. National Institute for Health and Care Excellence (NICE). Implantable cardioverter defibrillators and cardiac resynchronisation therapy for arrhythmias and heart failure (Review of TA95 and TA120). 2014; <https://www.nice.org.uk/guidance/ta314/documents/arrhythmias-icds-heart-failure-cardiac-resynchronisation-fad-document2>. Accessed April 3, 2023.
47. Connolly SJ, Hallstrom AP, Cappato R, et al. Meta-analysis of the implantable cardioverter defibrillator secondary prevention trials. AVID, CASH and CIDS studies. Antiarrhythmics vs Implantable Defibrillator study. Cardiac Arrest Study Hamburg. Canadian Implantable Defibrillator Study. *Eur Heart J*. Dec 2000; 21(24): 2071-8. PMID 11102258
48. Betts TR, Sadarmin PP, Tomlinson DR, et al. Absolute risk reduction in total mortality with implantable cardioverter defibrillators: analysis of primary and secondary prevention trial data to aid risk/benefit analysis. *Europace*. Jun 2013; 15(6): 813-9. PMID 23365069

49. Chan PS, Hayward RA. Mortality reduction by implantable cardioverter-defibrillators in high-risk patients with heart failure, ischemic heart disease, and new-onset ventricular arrhythmia: an effectiveness study. *J Am Coll Cardiol*. May 03 2005; 45(9): 1474-81. PMID 15862422
50. Persson R, Earley A, Garlitski AC, et al. Adverse events following implantable cardioverter defibrillator implantation: a systematic review. *J Interv Card Electrophysiol*. Aug 2014; 40(2): 191-205. PMID 24948126
51. Ezzat VA, Lee V, Ahsan S, et al. A systematic review of ICD complications in randomised controlled trials versus registries: is our 'real-world' data an underestimation?. *Open Heart*. 2015; 2(1): e000198. PMID 25745566
52. Kirkfeldt RE, Johansen JB, Nohr EA, et al. Complications after cardiac implantable electronic device implantations: an analysis of a complete, nationwide cohort in Denmark. *Eur Heart J*. May 2014; 35(18): 1186-94. PMID 24347317
53. van Rees JB, de Bie MK, Thijssen J, et al. Implantation-related complications of implantable cardioverter-defibrillators and cardiac resynchronization therapy devices: a systematic review of randomized clinical trials. *J Am Coll Cardiol*. Aug 30 2011; 58(10): 995-1000. PMID 21867832
54. Olde Nordkamp LR, Postema PG, Knops RE, et al. Implantable cardioverter-defibrillator harm in young patients with inherited arrhythmia syndromes: A systematic review and meta-analysis of inappropriate shocks and complications. *Heart Rhythm*. Feb 2016; 13(2): 443-54. PMID 26385533
55. Food and Drug Administration. Premature Insulation Failure in Recalled Riata Implantable Cardioverter Defibrillator (ICD) Leads Manufactured by St. Jude Medical, Inc.: FDA Safety Communication. 2014; <https://wayback.archive-it.org/7993/20170722215745/https://www.fda.gov/MedicalDevices/Safety/AlertsandNotices/ucm314930.htm>. Accessed April 4, 2023.
56. Hauser RG, Katsiyannis WT, Gornick CC, et al. Deaths and cardiovascular injuries due to device-assisted implantable cardioverter-defibrillator and pacemaker lead extraction. *Europace*. Mar 2010; 12(3): 395-401. PMID 19946113
57. Providência R, Kramer DB, Pimenta D, et al. Transvenous Implantable Cardioverter-Defibrillator (ICD) Lead Performance: A Meta-Analysis of Observational Studies. *J Am Heart Assoc*. Oct 30 2015; 4(11). PMID 26518666
58. Birnie DH, Parkash R, Exner DV, et al. Clinical predictors of Fidelis lead failure: report from the Canadian Heart Rhythm Society Device Committee. *Circulation*. Mar 13 2012; 125(10): 1217-25. PMID 22311781
59. Hauser RG, Maisel WH, Friedman PA, et al. Longevity of Sprint Fidelis implantable cardioverter-defibrillator leads and risk factors for failure: implications for patient management. *Circulation*. Feb 01 2011; 123(4): 358-63. PMID 21242478
60. Poole JE, Gleva MJ, Mela T, et al. Complication rates associated with pacemaker or implantable cardioverter-defibrillator generator replacements and upgrade procedures: results from the REPLACE registry. *Circulation*. Oct 19 2010; 122(16): 1553-61. PMID 20921437
61. Ricci RP, Pignalberi C, Magris B, et al. Can we predict and prevent adverse events related to high-voltage implantable cardioverter defibrillator lead failure?. *J Interv Card Electrophysiol*. Jan 2012; 33(1): 113-21. PMID 21882010
62. Cheng A, Wang Y, Curtis JP, et al. Acute lead dislodgements and in-hospital mortality in patients enrolled in the national cardiovascular data registry implantable cardioverter defibrillator registry. *J Am Coll Cardiol*. Nov 09 2010; 56(20): 1651-6. PMID 21050975
63. Faulkner BA, Traub DM, Aktas MK, et al. Time-dependent risk of Fidelis lead failure. *Am J Cardiol*. Jan 01 2010; 105(1): 95-9. PMID 20102898
64. Smit J, Korup E, Schønheyder HC. Infections associated with permanent pacemakers and implanted cardioverter-defibrillator devices. A 10-year regional study in Denmark. *Scand J Infect Dis*. Sep 2010; 42(9): 658-64. PMID 20465488
65. Nery PB, Fernandes R, Nair GM, et al. Device-related infection among patients with pacemakers and implantable defibrillators: incidence, risk factors, and consequences. *J Cardiovasc Electrophysiol*. Jul 2010; 21(7): 786-90. PMID 20102431

66. Sohail MR, Hussain S, Le KY, et al. Risk factors associated with early- versus late-onset implantable cardioverter-defibrillator infections. *J Interv Card Electrophysiol*. Aug 2011; 31(2): 171-83. PMID 21365264
67. Borleffs CJ, Thijssen J, de Bie MK, et al. Recurrent implantable cardioverter-defibrillator replacement is associated with an increasing risk of pocket-related complications. *Pacing Clin Electrophysiol*. Aug 2010; 33(8): 1013-9. PMID 20456647
68. Daubert JP, Zareba W, Cannom DS, et al. Inappropriate implantable cardioverter-defibrillator shocks in MADIT II: frequency, mechanisms, predictors, and survival impact. *J Am Coll Cardiol*. Apr 08 2008; 51(14): 1357-65. PMID 18387436
69. Tan VH, Wilton SB, Kuriachan V, et al. Impact of programming strategies aimed at reducing nonessential implantable cardioverter defibrillator therapies on mortality: a systematic review and meta-analysis. *Circ Arrhythm Electrophysiol*. Feb 2014; 7(1): 164-70. PMID 24446023
70. Sterns LD, Meine M, Kurita T, et al. Extended detection time to reduce shocks is safe in secondary prevention patients: The secondary prevention substudy of PainFree SST. *Heart Rhythm*. Jul 2016; 13(7): 1489-96. PMID 26988379
71. Auricchio A, Schloss EJ, Kurita T, et al. Low inappropriate shock rates in patients with single- and dual/triple-chamber implantable cardioverter-defibrillators using a novel suite of detection algorithms: PainFree SST trial primary results. *Heart Rhythm*. May 2015; 12(5): 926-36. PMID 25637563
72. Lee DS, Krahn AD, Healey JS, et al. Evaluation of early complications related to De Novo cardioverter defibrillator implantation insights from the Ontario ICD database. *J Am Coll Cardiol*. Feb 23 2010; 55(8): 774-82. PMID 20170816
73. Furniss G, Shi B, Jimenez A, et al. Cardiac troponin levels following implantable cardioverter defibrillation implantation and testing. *Europace*. Feb 2015; 17(2): 262-6. PMID 25414480
74. Healey JS, Krahn AD, Bashir J, et al. Perioperative Safety and Early Patient and Device Outcomes Among Subcutaneous Versus Transvenous Implantable Cardioverter Defibrillator Implantations : A Randomized, Multicenter Trial. *Ann Intern Med*. Dec 2022; 175(12): 1658-1665. PMID 36343346
75. Gold MR, Lambiase PD, El-Chami MF, et al. Primary Results From the Understanding Outcomes With the S-ICD in Primary Prevention Patients With Low Ejection Fraction (UNTOUCHED) Trial. *Circulation*. Jan 05 2021; 143(1): 7-17. PMID 33073614
76. Burke MC, Gold MR, Knight BP, et al. Safety and Efficacy of the Totally Subcutaneous Implantable Defibrillator: 2-Year Results From a Pooled Analysis of the IDE Study and EFFORTLESS Registry. *J Am Coll Cardiol*. Apr 28 2015; 65(16): 1605-1615. PMID 25908064
77. Gold MR, Aasbo JD, Weiss R, et al. Infection in patients with subcutaneous implantable cardioverter-defibrillator: Results of the S-ICD Post Approval Study. *Heart Rhythm*. Dec 2022; 19(12): 1993-2001. PMID 35944889
78. Lambiase PD, Barr C, Theuns DA, et al. Worldwide experience with a totally subcutaneous implantable defibrillator: early results from the EFFORTLESS S-ICD Registry. *Eur Heart J*. Jul 01 2014; 35(25): 1657-65. PMID 24670710
79. Olde Nordkamp LR, Brouwer TF, Barr C, et al. Inappropriate shocks in the subcutaneous ICD: Incidence, predictors and management. *Int J Cardiol*. Sep 15 2015; 195: 126-33. PMID 26026928
80. Boersma L, Barr C, Knops R, et al. Implant and Midterm Outcomes of the Subcutaneous Implantable Cardioverter-Defibrillator Registry: The EFFORTLESS Study. *J Am Coll Cardiol*. Aug 15 2017; 70(7): 830-841. PMID 28797351
81. Weiss R, Knight BP, Gold MR, et al. Safety and efficacy of a totally subcutaneous implantable-cardioverter defibrillator. *Circulation*. Aug 27 2013; 128(9): 944-53. PMID 23979626
82. Boersma L, Burke MC, Neuzil P, et al. Infection and mortality after implantation of a subcutaneous ICD after transvenous ICD extraction. *Heart Rhythm*. Jan 2016; 13(1): 157-64. PMID 26341604

83. Lambiase PD, Gold MR, Hood M, et al. Evaluation of subcutaneous ICD early performance in hypertrophic cardiomyopathy from the pooled EFFORTLESS and IDE cohorts. *Heart Rhythm*. May 2016; 13(5): 1066-1074. PMID 26767422
84. Bardy GH, Smith WM, Hood MA, et al. An entirely subcutaneous implantable cardioverter-defibrillator. *N Engl J Med*. Jul 01 2010; 363(1): 36-44. PMID 20463331
85. Theuns DA, Crozier IG, Barr CS, et al. Longevity of the Subcutaneous Implantable Defibrillator: Long-Term Follow-Up of the European Regulatory Trial Cohort. *Circ Arrhythm Electrophysiol*. Oct 2015; 8(5): 1159-63. PMID 26148819
86. Olde Nordkamp LR, Dabiri Abkenari L, Boersma LV, et al. The entirely subcutaneous implantable cardioverter-defibrillator: initial clinical experience in a large Dutch cohort. *J Am Coll Cardiol*. Nov 06 2012; 60(19): 1933-9. PMID 23062537
87. Knops RE, Olde Nordkamp LRA, Delnoy PHM, et al. Subcutaneous or Transvenous Defibrillator Therapy. *N Engl J Med*. Aug 06 2020; 383(6): 526-536. PMID 32757521
88. Mithani AA, Kath H, Hunter K, et al. Characteristics and early clinical outcomes of patients undergoing totally subcutaneous vs. transvenous single chamber implantable cardioverter defibrillator placement. *Europace*. Feb 01 2018; 20(2): 308-314. PMID 28383717
89. Honarbakhsh S, Providencia R, Srinivasan N, et al. A propensity matched case-control study comparing efficacy, safety and costs of the subcutaneous vs. transvenous implantable cardioverter defibrillator. *Int J Cardiol*. Feb 01 2017; 228: 280-285. PMID 27865198
90. Köbe J, Hucklenbroich K, Geisendörfer N, et al. Posttraumatic stress and quality of life with the totally subcutaneous compared to conventional cardioverter-defibrillator systems. *Clin Res Cardiol*. May 2017; 106(5): 317-321. PMID 27878381
91. Pedersen SS, Mastenbroek MH, Carter N, et al. A Comparison of the Quality of Life of Patients With an Entirely Subcutaneous Implantable Defibrillator System Versus a Transvenous System (from the EFFORTLESS S-ICD Quality of Life Substudy). *Am J Cardiol*. Aug 15 2016; 118(4): 520-6. PMID 27353211
92. Brouwer TF, Yilmaz D, Lindeboom R, et al. Long-Term Clinical Outcomes of Subcutaneous Versus Transvenous Implantable Defibrillator Therapy. *J Am Coll Cardiol*. Nov 08 2016; 68(19): 2047-2055. PMID 27810043
93. Friedman DJ, Parzynski CS, Varosy PD, et al. Trends and In-Hospital Outcomes Associated With Adoption of the Subcutaneous Implantable Cardioverter Defibrillator in the United States. *JAMA Cardiol*. Nov 01 2016; 1(8): 900-911. PMID 27603935
94. Köbe J, Reinke F, Meyer C, et al. Implantation and follow-up of totally subcutaneous versus conventional implantable cardioverter-defibrillators: a multicenter case-control study. *Heart Rhythm*. Jan 2013; 10(1): 29-36. PMID 23032867
95. Heidenreich PA, Bozkurt B, Aguilar D, et al. 2022 AHA/ACC/HFSA Guideline for the Management of Heart Failure: Executive Summary: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. May 03 2022; 145(18): e876-e894. PMID 35363500
96. Ommen SR, Mital S, Burke MA, et al. 2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: Executive Summary: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. Dec 22 2020; 142(25): e533-e557. PMID 33215938
97. Al-Khatib SM, Stevenson WG, Ackerman MJ, et al. 2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death: Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *J Am Coll Cardiol*. Oct 02 2018; 72(14): 1677-1749. PMID 29097294
98. Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the ACC/AHA/NASPE 2002 Guideline Update for Implantation of Cardiac Pacemakers and Antiarrhythmia Devices): developed in collaboration with the

- American Association for Thoracic Surgery and Society of Thoracic Surgeons. *Circulation*. May 27 2008; 117(21): e350-408. PMID 18483207
99. Epstein AE, DiMarco JP, Ellenbogen KA, et al. 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. *J Am Coll Cardiol*. Jan 22 2013; 61(3): e6-75. PMID 23265327
100. Towbin JA, McKenna WJ, Abrams DJ, et al. 2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy. *Heart Rhythm*. Nov 2019; 16(11): e301-e372. PMID 31078652
101. Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes: document endorsed by HRS, EHRA, and APHRS in May 2013 and by ACCF, AHA, PACES, and AEPC in June 2013. *Heart Rhythm*. Dec 2013; 10(12): 1932-63. PMID 24011539
102. Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). *Can J Cardiol*. Oct 2014; 30(10): e1-e63. PMID 25262867
103. Shah MJ, Silka MJ, Silva JNA, et al. 2021 PACES Expert Consensus Statement on the Indications and Management of Cardiovascular Implantable Electronic Devices in Pediatric Patients: Developed in collaboration with and endorsed by the Heart Rhythm Society (HRS), the American College of Cardiology (ACC), the American Heart Association (AHA), and the Association for European Paediatric and Congenital Cardiology (AEPC) Endorsed by the Asia Pacific Heart Rhythm Society (APHRS), the Indian Heart Rhythm Society (IHRS), and the Latin American Heart Rhythm Society (LAHRS). *JACC Clin Electrophysiol*. Nov 2021; 7(11): 1437-1472. PMID 34794667
104. Centers for Medicare & Medicaid Services. National Coverage Determination (NCD) for Implantable Automatic Defibrillators (20.4). 2018; <https://www.cms.gov/Medicare-Coverage-Database/view/ncacal-decision-memo.aspx?proposed=N&NCAId=288>. Accessed April 3, 2023.

Documentation for Clinical Review

Please provide the following documentation:

- History and physical and/or cardiology consultation report including:
 - Clinical justification for ICD placement including major risk factors for sudden cardiac death
 - Date ICD procedure is planned and type of ICD requested (automatic or subcutaneous)
 - Past medical treatment and response(s)
 - Myocardial infarction history including date (if applicable)
 - NYHA Functional Classification
 - Past cardiac surgical history (e.g., ICD placement or explantation, revascularization procedures) and dates associated (if applicable)
 - Estimated life expectancy based on medical history (non-cardiac)
 - Specific family history of sudden cardiac death (including generation)
 - Cardiac monitoring result(s) (e.g., EKG, Holter, echocardiogram, hemodynamic or EP studies)
 - Echocardiogram report within the past six months including Left Ventricular Ejection Fraction (LVEF) if applicable

Post Service (in addition to the above, please include the following):

- Operative procedure report(s) relating to an ICD (if applicable)

Coding

This Policy relates only to the services or supplies described herein. Benefits may vary according to product design; therefore, contract language should be reviewed before applying the terms of the Policy.

The following codes are included below for informational purposes. Inclusion or exclusion of a code(s) does not constitute or imply member coverage or provider reimbursement policy. Policy Statements are intended to provide member coverage information and may include the use of some codes for clarity. The Policy Guidelines section may also provide additional information for how to interpret the Policy Statements and to provide coding guidance in some cases.

| Type | Code | Description |
|------|-------|---|
| CPT® | 0571T | Insertion or replacement of implantable cardioverter-defibrillator system with substernal electrode(s), including all imaging guidance and electrophysiological evaluation (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters), when performed |
| | 0572T | Insertion of substernal implantable defibrillator electrode |
| | 0573T | Removal of substernal implantable defibrillator electrode |
| | 0574T | Repositioning of previously implanted substernal implantable defibrillator-pacing electrode |
| | 0575T | Programming device evaluation (in person) of implantable cardioverter-defibrillator system with substernal electrode, 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 |
| | 0576T | Interrogation device evaluation (in person) of implantable cardioverter-defibrillator system with substernal electrode, with analysis, review and report by a physician or other qualified health care professional, includes connection, recording and disconnection per patient encounter |
| | 0577T | Electrophysiologic evaluation of implantable cardioverter-defibrillator system with substernal electrode (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters) |
| | 0578T | Interrogation device evaluation(s) (remote), up to 90 days, substernal lead implantable cardioverter-defibrillator system with interim analysis, review(s) and report(s) by a physician or other qualified health care professional |
| | 0579T | Interrogation device evaluation(s) (remote), up to 90 days, substernal lead implantable cardioverter-defibrillator system, remote data acquisition(s), receipt of transmissions and technician review, technical support and distribution of results |
| | 0580T | Removal of substernal implantable defibrillator pulse generator only |

| Type | Code | Description |
|------|-------|---|
| | 0614T | Removal and replacement of substernal implantable defibrillator pulse generator |
| | 33216 | Insertion of a single transvenous electrode, permanent pacemaker or implantable defibrillator |
| | 33217 | Insertion of 2 transvenous electrodes, permanent pacemaker or implantable defibrillator |
| | 33218 | Repair of single transvenous electrode, permanent pacemaker or implantable defibrillator |
| | 33220 | Repair of 2 transvenous electrodes for permanent pacemaker or implantable defibrillator |
| | 33223 | Relocation of skin pocket for implantable defibrillator |
| | 33230 | Insertion of implantable defibrillator pulse generator only; with existing dual leads |
| | 33231 | Insertion of implantable defibrillator pulse generator only; with existing multiple leads |
| | 33240 | Insertion of implantable defibrillator pulse generator only; with existing single lead |
| | 33241 | Removal of implantable defibrillator pulse generator only |
| | 33243 | Removal of single or dual chamber implantable defibrillator electrode(s); by thoracotomy |
| | 33244 | Removal of single or dual chamber implantable defibrillator electrode(s); by transvenous extraction |
| | 33249 | Insertion or replacement of permanent implantable defibrillator system, with transvenous lead(s), single or dual chamber |
| | 33262 | Removal of implantable defibrillator pulse generator with replacement of implantable defibrillator pulse generator; single lead system |
| | 33263 | Removal of implantable defibrillator pulse generator with replacement of implantable defibrillator pulse generator; dual lead system |
| | 33264 | Removal of implantable defibrillator pulse generator with replacement of implantable defibrillator pulse generator; multiple lead system |
| | 33270 | Insertion or replacement of permanent subcutaneous implantable defibrillator system, with subcutaneous electrode, including defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters, when performed |
| | 33271 | Insertion of subcutaneous implantable defibrillator electrode |
| | 33272 | Removal of subcutaneous implantable defibrillator electrode |
| | 33273 | Repositioning of previously implanted subcutaneous implantable defibrillator electrode |
| | 93260 | 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; 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 |

| Type | Code | Description |
|------|-------|---|
| | 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 transvenous implantable 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 transvenous implantable 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 transvenous implantable defibrillator system |
| | 93287 | Peri-procedural device evaluation (in person) and programming of device system parameters before or after a surgery, procedure, or test with analysis, review and report by a physician or other qualified health care professional; single, dual, or multiple lead implantable 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 transvenous implantable defibrillator system, including analysis of heart rhythm derived data elements |
| | 93295 | Interrogation device evaluation(s) (remote), up to 90 days; single, dual, or multiple lead implantable defibrillator system with interim analysis, review(s) and report(s) by a physician or other qualified health care professional |
| | 93296 | Interrogation device evaluation(s) (remote), up to 90 days; single, dual, or multiple lead pacemaker system, leadless pacemaker system, or implantable defibrillator system, remote data acquisition(s), receipt of transmissions and technician review, technical support and distribution of results |
| | 93297 | Interrogation device evaluation(s), (remote) up to 30 days; implantable cardiovascular physiologic monitor system, including analysis of 1 or more recorded physiologic cardiovascular data elements from all internal and external sensors, analysis, review(s) and report(s) by a physician or other qualified health care professional |
| | 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 |

| Type | Code | Description |
|-------|-------|---|
| | 93642 | Electrophysiologic evaluation of single or dual chamber transvenous 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) |
| HCPCS | C1721 | Cardioverter-defibrillator, dual chamber (implantable) |
| | C1722 | Cardioverter-defibrillator, single chamber (implantable) |
| | C1777 | Lead, cardioverter-defibrillator, endocardial single coil (implantable) |
| | C1824 | Generator, cardiac contractility modulation (implantable) |
| | C1882 | Cardioverter-defibrillator, other than single or dual chamber (implantable) |
| | C1895 | Lead, cardioverter-defibrillator, endocardial dual coil (implantable) |
| | C1896 | Lead, cardioverter-defibrillator, other than endocardial single or dual coil (implantable) |
| | C1899 | Lead, pacemaker/cardioverter-defibrillator combination (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 |

Policy History

This section provides a chronological history of the activities, updates and changes that have occurred with this Medical Policy.

| Effective Date | Action |
|----------------|---|
| 09/01/1986 | BCBSA Medical Policy adoption |
| 06/01/1999 | Policy moved into Archive status |
| 10/01/2002 | BCBSA Medical Policy adoption |
| 08/01/2005 | Policy Revision |
| 05/01/2006 | Policy Revision |
| 12/18/2009 | Policy Revision with title change from Automatic Implantable Cardioverter Defibrillators (A-IDC) for Prevention of Sudden Death |
| 08/04/2010 | Administrative Review |
| 03/13/2012 | Coding Update |
| 01/11/2013 | Policy revision with position change |
| 01/21/2013 | Policy documentation clarification |
| 01/30/2015 | Coding update |
| 07/31/2015 | Coding update |
| 01/01/2016 | Policy revision with position change |
| 08/01/2016 | Policy revision without position change |
| 07/01/2017 | Policy revision without position change |
| 07/01/2018 | Policy revision without position change |
| 07/01/2019 | Policy title change from Implantable Cardioverter Defibrillator Policy revision without position change |

| Effective Date | Action |
|----------------|--|
| 03/01/2020 | Coding update |
| 05/01/2020 | Administrative update. Policy statement and guidelines updated. |
| 08/01/2020 | Annual review. Coding update. Policy statement, guidelines and literature updated. |
| 01/01/2021 | Coding update |
| 07/01/2021 | Annual review. No change to policy statement. Literature review updated. |
| 07/01/2022 | Annual review. Policy statement, guidelines and literature updated. |
| 07/01/2023 | Annual review. Policy statement, guidelines and literature updated. |

Definitions of Decision Determinations

Medically Necessary: Services that are Medically Necessary include only those which have been established as safe and effective, are furnished under generally accepted professional standards to treat illness, injury or medical condition, and which, as determined by Blue Shield, are: (a) consistent with Blue Shield medical policy; (b) consistent with the symptoms or diagnosis; (c) not furnished primarily for the convenience of the patient, the attending Physician or other provider; (d) furnished at the most appropriate level which can be provided safely and effectively to the patient; and (e) not more costly than an alternative service or sequence of services at least as likely to produce equivalent therapeutic or diagnostic results as to the diagnosis or treatment of the Member's illness, injury, or disease.

Investigational/Experimental: A treatment, procedure, or drug is investigational when it has not been recognized as safe and effective for use in treating the particular condition in accordance with generally accepted professional medical standards. This includes services where approval by the federal or state governmental is required prior to use, but has not yet been granted.

Split Evaluation: Blue Shield of California/Blue Shield of California Life & Health Insurance Company (Blue Shield) policy review can result in a split evaluation, where a treatment, procedure, or drug will be considered to be investigational for certain indications or conditions, but will be deemed safe and effective for other indications or conditions, and therefore potentially medically necessary in those instances.

Prior Authorization Requirements and Feedback (as applicable to your plan)

Within five days before the actual date of service, the provider must confirm with Blue Shield that the member's health plan coverage is still in effect. Blue Shield reserves the right to revoke an authorization prior to services being rendered based on cancellation of the member's eligibility. Final determination of benefits will be made after review of the claim for limitations or exclusions.

Questions regarding the applicability of this policy should be directed to the Prior Authorization Department at (800) 541-6652, or the Transplant Case Management Department at (800) 637-2066 ext. 3507708 or visit the provider portal at www.blueshieldca.com/provider.

We are interested in receiving feedback relative to developing, adopting, and reviewing criteria for medical policy. Any licensed practitioner who is contracted with Blue Shield of California or Blue Shield of California Promise Health Plan is welcome to provide comments, suggestions, or concerns. Our internal policy committees will receive and take your comments into consideration.

For utilization and medical policy feedback, please send comments to: MedPolicy@blueshieldca.com

Disclaimer: This medical policy is a guide in evaluating the medical necessity of a particular service or treatment. Blue Shield of California may consider published peer-reviewed scientific literature, national guidelines, and local

7.01.44 Implantable Cardioverter Defibrillators

Page 56 of 59

standards of practice in developing its medical policy. Federal and state law, as well as contract language, including definitions and specific contract provisions/exclusions, take precedence over medical policy and must be considered first in determining covered services. Member contracts may differ in their benefits. Blue Shield reserves the right to review and update policies as appropriate.

Appendix A

| POLICY STATEMENT | |
|--|--|
| BEFORE <u>Red font: Verbiage removed</u> | AFTER <u>Blue font: Verbiage Changes/Additions</u> |
| <p>Implantable Cardioverter Defibrillators 7.01.44</p> <p>Policy Statement:</p> <ol style="list-style-type: none"> I. The use of the automatic implantable cardioverter defibrillator (ICD) may be considered medically necessary for primary or secondary prevention for a patient who meets all of the following: <ol style="list-style-type: none"> A. The patient has NOT had an acute myocardial infarction within the past 40 days B. The patient does NOT have New York Heart Association (NYHA) class IV congestive heart failure C. The patient has NOT had a cardiac revascularization procedure within the past 3 months D. The patient is NOT a candidate for a cardiac revascularization procedure E. The patient does NOT have a life expectancy of less than 1 year F. Reversible causes (e.g., acute ischemia) are NOT present or have been maximally treated G. The use of the implantable cardioverter defibrillator (ICD) is indicated for one or more of the following: <ol style="list-style-type: none"> 1. Ischemic cardiomyopathy, New York Heart Association (NYHA) functional class II or III symptoms and left ventricular ejection fraction of 35% or less 2. Ischemic cardiomyopathy, NYHA functional class I symptoms and left ventricular ejection fraction of 30% or less 3. Nonischemic dilated cardiomyopathy and all of the following: <ol style="list-style-type: none"> a. Left ventricular ejection fraction of 35% or less b. Response to optimal medical therapy has been adequately determined 4. Hypertrophic cardiomyopathy (HCM) with both of the following: | <p>Implantable Cardioverter Defibrillators 7.01.44</p> <p>Policy Statement:</p> <ol style="list-style-type: none"> I. The use of the automatic implantable cardioverter defibrillator (ICD) may be considered medically necessary for primary or secondary prevention for individuals who meets all of the following criteria: <ol style="list-style-type: none"> A. The individual has NOT had an acute myocardial infarction within the past 40 days B. The individual does NOT have New York Heart Association (NYHA) class IV congestive heart failure C. The individual has NOT had a cardiac revascularization procedure within the past 3 months D. The individual is NOT a candidate for a cardiac revascularization procedure E. The individual does NOT have a life expectancy of less than 1 year F. Reversible causes (e.g., acute ischemia) are NOT present or have been maximally treated G. The use of the implantable cardioverter defibrillator (ICD) is indicated for one or more of the following: <ol style="list-style-type: none"> 1. Ischemic cardiomyopathy with New York Heart Association (NYHA) functional class II or III symptoms and left ventricular ejection fraction (LVEF) of 35% or less 2. Ischemic cardiomyopathy, NYHA functional class I symptoms and LVEF of 30% or less 3. Nonischemic dilated cardiomyopathy and all of the following: <ol style="list-style-type: none"> a. LVEF of 35% or less b. Response to optimal medical therapy has been adequately determined 4. Hypertrophic cardiomyopathy (HCM) with both of the following: |

POLICY STATEMENT

| BEFORE Red font: Verbiage removed | AFTER Blue font: Verbiage Changes/Additions |
|---|--|
| <ul style="list-style-type: none"> a. Judged to be at high risk for sudden cardiac death by a physician experienced in the care of patients with HCM b. Major risk factors for sudden cardiac death as indicated by any of the following: <ul style="list-style-type: none"> i. History of premature HCM-related sudden death in one or more first-degree relatives younger than 50 years ii. Left ventricular hypertrophy (left ventricular wall thickness greater than 30 mm) iii. One or more runs of non-sustained ventricular tachycardia at heart rates of 120 beats per minute or greater iv. Prior unexplained syncope inconsistent with neurocardiogenic origin 5. Considered to be at high risk for sudden cardiac death and diagnosis of cardiac ion channelopathy as indicated by any of the following: <ul style="list-style-type: none"> a. Congenital Long QT Syndrome (LQTS) b. Brugada Syndrome (BrS) c. Short QT Syndrome (SQTS) d. Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) 6. Diagnosis of cardiac sarcoid and considered to be at high risk for sudden cardiac death (see Policy Guidelines section) 7. Patient with a history of a life-threatening clinical event associated with ventricular arrhythmic events such as sustained ventricular tachyarrhythmia or survival of cardiac arrest 8. Symptomatic, sustained ventricular tachycardia in association with congenital heart disease after hemodynamic and electrophysiologic evaluation 9. Congenital heart disease with recurrent syncope of undetermined origin in the presence of ventricular dysfunction or inducible ventricular arrhythmias | <ul style="list-style-type: none"> a. Judged to be at high risk for sudden cardiac death by a provider experienced in the care of individuals with HCM b. Major risk factors for sudden cardiac death as indicated by any of the following: <ul style="list-style-type: none"> i. History of premature HCM-related sudden death in one or more first-degree relatives younger than 50 years ii. Left ventricular hypertrophy (left ventricular wall thickness greater than 30 mm) iii. One or more runs of non-sustained ventricular tachycardia at heart rates of 120 beats per minute or greater iv. Prior unexplained syncope inconsistent with neurocardiogenic origin 5. Considered to be at high risk for sudden cardiac death and diagnosis of cardiac ion channelopathy as indicated by any of the following: <ul style="list-style-type: none"> a. Congenital Long QT Syndrome (LQTS) b. Brugada Syndrome (BrS) c. Short QT Syndrome (SQTS) d. Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) 6. Diagnosis of cardiac sarcoid and considered to be at high risk for sudden cardiac death (see Policy Guidelines section) 7. Individual with a history of a life-threatening clinical event associated with ventricular arrhythmic events such as sustained ventricular tachyarrhythmia or survival of cardiac arrest 8. Symptomatic, sustained ventricular tachycardia in association with congenital heart disease after hemodynamic and electrophysiologic evaluation 9. Congenital heart disease with recurrent syncope of undetermined origin in the presence of ventricular dysfunction or inducible ventricular arrhythmias |

POLICY STATEMENT

| BEFORE Red font: Verbiage removed | AFTER Blue font: Verbiage Changes/Additions |
|---|---|
| <p>II. The use of the ICD is considered not medically necessary in primary or secondary prevention patients who have had any of the following risk factors or do not meet the criteria for approval:</p> <ul style="list-style-type: none"> A. History of an acute myocardial infarction (i.e., less than 40 days before ICD treatment) B. New York Heart Association (NYHA) class IV congestive heart failure (unless patient is eligible to receive a combination cardiac resynchronization therapy ICD device) C. History of a cardiac revascularization procedure in past 3 months (coronary artery bypass graft [CABG] or percutaneous transluminal coronary angioplasty [PTCA]) D. Candidates for a cardiac revascularization procedure E. Have noncardiac disease that would be associated with life expectancy less than 1 year <p>Subcutaneous ICD</p> <ul style="list-style-type: none"> I. The use of a subcutaneous ICD may be considered medically necessary for an adult or pediatric patient who meets criteria for ICD implantation for primary or secondary prevention for any of the above reasons and meet all of the following criteria: <ul style="list-style-type: none"> A. Have a contraindication to a transvenous ICD due to any of the following: <ul style="list-style-type: none"> 1. Lack of adequate vascular access 2. Compelling reason to preserve existing vascular access (i.e., need for chronic dialysis; younger patient with anticipated long-term need for ICD therapy) 3. History of need for explantation of a transvenous ICD due to a complication, with ongoing need for ICD therapy B. Have no indication for antibradycardia pacing C. No known or anticipated ventricular arrhythmias that would respond to anti-tachycardia pacing II. The use of a subcutaneous ICD is considered investigational for individuals who do not meet the criteria outlined above. | <p>II. The use of the ICD is considered investigational in primary or secondary prevention individuals who have had any of the following risk factors or do not meet the criteria for approval:</p> <ul style="list-style-type: none"> A. History of an acute myocardial infarction (i.e., less than 40 days before ICD treatment) B. New York Heart Association (NYHA) class IV congestive heart failure (unless individual is eligible to receive a combination cardiac resynchronization therapy ICD device) C. History of a cardiac revascularization procedure in past 3 months (coronary artery bypass graft [CABG] or percutaneous transluminal coronary angioplasty [PTCA]) D. Candidates for a cardiac revascularization procedure E. Have noncardiac disease that would be associated with life expectancy less than 1 year <p>Subcutaneous Implantable Cardioverter Defibrillator</p> <ul style="list-style-type: none"> III. The use of a subcutaneous ICD may be considered medically necessary for adult or pediatric individuals who meets criteria for ICD implantation for primary or secondary prevention for any of the above reasons and meet all of the following criteria: <ul style="list-style-type: none"> A. Have a contraindication to a transvenous ICD due to any of the following: <ul style="list-style-type: none"> 1. Lack of adequate vascular access 2. Compelling reason to preserve existing vascular access (i.e., need for chronic dialysis; younger individual with anticipated long-term need for ICD therapy) 3. History of need for explantation of a transvenous ICD due to a complication, with ongoing need for ICD therapy B. Have no indication for antibradycardia pacing C. No known or anticipated ventricular arrhythmias known or anticipated to respond to antitachycardia pacing IV. The use of a subcutaneous ICD is considered investigational for individuals who do not meet the criteria outlined above. |