Medical Coverage Policy

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Implantable Cardioverter Defibrillator (ICD)

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Overview

This Coverage Policy addresses the use of implantable transvenous, subcutaneous cardioverter-defibrillator and substernal cardioverter-defibrillator to monitor heart rhythm and deliver an electrical shock when a life-threatening ventricular arrhythmia is detected.

Coverage Policy

Secondary Prevention of Sudden Cardiac Death (SCD)

A transvenous implantable cardioverter defibrillator (ICD) is considered medically necessary for the secondary prevention of sudden cardiac death for EITHER of the following indications:

- **Individual with cardiac arrest due to ventricular fibrillation (VF) or hemodynamically unstable sustained ventricular tachycardia (VT) after reversable causes (e.g., myocardial ischemia (MI), electrolyte disorder) have been excluded.**

- **Individual with structural heart disease (e.g., prior MI, cardiomyopathy, valvular heart disease, adult congenital heart disease) and spontaneous sustained VT, whether hemodynamically stable or unstable.**

- **Individual with genetic conditions associated with sustained VT/VF (i.e., congenital long QT, short QT, catecholaminergic polymorphic VT, Brugada syndrome, arrhythmogenic right ventricular cardiomyopathy, hypertrophic cardiomyopathy).**

- **Individual without structural heart disease (left ventricular ejection fraction [LVEF] > 50%) or known genetic causes of sustained VT/VF and EITHER of the following:**
  - Bradycardia dependent VT/VF
  - Idiopathic VF/VT with normal ventricular function

- **Individual with unexplained syncope due to ANY of the following:**
  - Cardiac sarcoidosis with documented spontaneous sustained ventricular tachycardia
  - Ischemic heart disease with inducible sustained monomorphic VT on electrophysiological study.
  - Left ventricular non-compaction
  - Nonischemic dilated cardiomyopathy, LVEF ≤ 49%
  - Structural heart disease (e.g. prior MI) with LVEF ≤ 35%
  - Structural heart disease (e.g. prior MI) with LVEF 36%–49% and inducible sustained VT/VF on electrophysiological study.
  - Tetralogy of Fallot with prior corrective surgery

- **Individual with syncope of suspected arrhythmic cause and ANY of the following:**
  - Arrhythmogenic right ventricular cardiomyopathy (ARVC)
  - Brugada ECG pattern
  - Cardiac amyloidosis
  - Catecholaminergic polymorphic VT (CPVT)
  - Hypertrophic Cardiomyopathy (HCM)
  - Long QT Syndrome (LQTS) and EITHER of the following:

- syncope while receiving beta-blockers
- beta-blockers are contraindicated

Primary Prevention of Sudden Cardiac Death

A transvenous implantable cardioverter defibrillator (ICD) is considered medically necessary for the primary prevention of sudden cardiac death for ANY of the following indications:

- **In an individual that is post-acute myocardial infarction (MI) (> 48 hours and < 40 days) and/or revascularization (< 90 days), with LVEF ≤ 40% and BOTH of the following:**
  - **Nonsustained ventricular tachycardia (NSVT)**
  - **Inducible sustained VT at electrophysiological (EP) study**

- **In an individual that is post-MI (≤ 40 Days) and need guideline-directed pacemaker therapy post-MI (e.g., sick sinus syndrome (SSS), complete heart block (CHB), or other indications for permanent pacemaker**), **with LVEF ≤ 40%**

- **In an individual that is post-MI (≥ 40 days) with ischemic cardiomyopathy, no recent percutaneous coronary intervention (PCI) or coronary artery bypass graft (CABG) (≥ 90 days) and ANY of the following:**
  - LVEF ≤ 30% NYHA class I (despite guideline-directed medical therapy)
  - LVEF ≤ 35% NYHA class II or III (despite guideline-directed medical therapy)
  - LVEF ≤ 40% NSVT with EPS showing inducible sustained VT/VF

- **Individual with nonischemic cardiomyopathy, at least 3 months on guideline-directed medical therapy, with LVEF ≤ 35%, NYHA Class II-III**

- **Individual with cardiac sarcoidosis and ANY of the following:**
  - Sustained VT
  - Survivors of SCA
  - LVEF ≤ 35%
  - LVEF > 35% with syncope and/or evidence of myocardial scar by cardiac MRI or positron emission tomographic (PET) scan
  - LVEF > 35%, with inducible sustained VA

- **Individual with ANY of the following conditions:**
  - Myotonic dystrophy
  - Chagas disease
  - Acute lymphocytic myocarditis, newly diagnosed (< 3 months)
  - Giant cell myocarditis
  - Peripartum cardiomyopathy, persists > 3 months postpartum, LVEF ≤ 35%

- **Individual with ANY of the following genetic conditions (excludes syncope and sustained VT, addressed above)**
  - Hypertrophic cardiomyopathy (HCM) with 1 or more risk factors:
    - Prior cardiac arrest or spontaneous nonsustained VT
    - Family history of SCD from HCM
    - LV thickness greater than or equal to 30 mm by echocardiography or cardiovascular magnetic resonance (CMR) imaging
    - abnormal blood pressure response to exercise
    - NSVT episodes on continuous ambulatory electrocardiographic monitoring

    - LV apical aneurysm, independent of size
    - LV systolic dysfunction (EF < 50%) by echocardiography or CMR imaging.
    - Extensive late gadolinium enhancement (LGE) on CMR imaging.
- Arrhythmogenic right ventricular dysplasia/cardiomyopathy with no symptoms due to arrhythmia
- Congenital long QT Syndrome with 1 or more risk factors (e.g., sudden cardiac arrest, family history of SCD, compliance/intolerance to drugs is a concern)
- Catecholaminergic polymorphic VT with nonsustained VT (without syncope)
- Incidentally discovered Brugada by ECG (type I ECG pattern) in the absence of symptoms or family history of sudden cardiac death, with inducible VT or VF at EPS
- Familial dilated nonischemic cardiomyopathy (RV/LV) associated with sudden cardiac death, and ANY of the following:
    - Evidence of structural cardiac disease, but LVEF > 35%
    - Normal ECG and echo, but carrying the implicated gene
    - LV non-compaction with LVEF > 35%
- Nonischemic cardiomyopathy (NICM) due to a Lamin A/C mutation with 2 or more risk factors (e.g., NSVT, LVEF <45%, non-missense mutation, male sex)

A transvenous ICD is considered medically necessary in a child who is receiving optimal medical therapy and has survived cardiac arrest when evaluation fails to identify a reversible cause.

A transvenous ICD is considered medically necessary in a child with hypertrophic cardiomyopathy and unexplained syncope, massive left ventricular hypertrophy, or family history of sudden cardiac death.

A transvenous ICD is considered experimental, investigational or unproven for ANY other indication.

Replacement of a transvenous ICD pulse generator and/or leads is considered medically necessary.

A subcutaneous implantable cardioverter defibrillator (S-ICD) system is considered medically necessary when an individual has met the criteria for a transvenous ICD and has NONE of the following:

- symptomatic bradycardia
- incessant ventricular tachycardia (VT)
- spontaneous frequent recurring VT reliably terminated with anti-tachycardia pacing

A subcutaneous implantable cardioverter defibrillator (S-ICD) system is considered experimental, investigational or unproven for ANY other indication.

A substernal implantable cardioverter-defibrillator is considered experimental, investigational or unproven for ANY indication.

General Background

Transvenous Implantable Cardioverter Defibrillator (ICD)

Sudden cardiac arrest (SCA) and sudden cardiac death (SCD) refer to the sudden stopping of cardiac activity with hemodynamic collapse which is frequently due to sustained ventricular tachycardia/ventricular fibrillation. These events frequently occur in patients with structural heart

disease (that may not have been previously diagnosed), particularly coronary heart disease (CHD). Additionally, there is a high incidence of sudden cardiac death (SCD) in patients with heart failure and diminished left ventricular ejection fraction (LVEF) and in patients who are recovering from acute myocardial infarction (MI). Although the risk of SCD increases in proportion to the severity of cardiac disease in an individual patient, most events occur in patients with no known cardiac history and with few or no risk factors. The risk factors for CHD are also risk factors for SCA. These include dyslipidemia, hypertension, cigarette smoking, physical inactivity, obesity, diabetes mellitus, and a family history of premature CHD or myocardial infarction (Podrid, 2022; Podrid, 2020; Kusmirek and Gold, 2007; Zipes, et al., 2006).

In the United States, SCD is responsible for an estimated 350,000 cardiac deaths per year. Epidemiologic studies suggest that men, Blacks and individuals from socioeconomically disadvantaged backgrounds experience higher rates of cardiac arrest (Podrid, 2020). Banerjee et al. (2021) reported that Blacks and Hispanics tend to reside in neighborhoods that have lower rates of bystander cardiopulmonary resuscitation and automatic external defibrillator (AED) use and, should they happen to survive a cardiac arrest, are less likely to subsequently receive an implantable cardioverter-defibrillator (ICD).

Although a number of studies have investigated the electrophysiologic (EP) mechanisms responsible for the onset of ventricular tachycardia and ventricular fibrillation, antiarrhythmic agents have not been shown to be effective in preventing SCD. Rather, it is the drugs that have no direct EP actions on cardiac muscle or specialized conducting tissue that have been demonstrated to be effective in preventing SCD. Such drugs include beta blockers, ACE inhibitors, angiotensin receptor-blocking agents, lipid-lowering agents, spironolactone, and fibrinolytic and anti-thrombotic agents (Al-Khatib et al., 2017; Zipes, et al., 2006).

SCD, a direct result of cardiac arrest, may be preventable if the arrest is responded to promptly. The implantable cardioverter defibrillator (ICD) is a surgically implanted device designed to constantly monitor an individual's heart rate, recognize VF or VT and deliver an electric shock to terminate these arrhythmias in order to reduce the risk of sudden death. The device is connected to leads positioned inside the heart or on its surface. These leads sense the cardiac rhythm, deliver electrical shocks, and sometimes pace the heart, as needed. The leads are tunneled to a pulse generator, which is implanted in a pouch beneath the skin of the chest or abdomen. Progressive improvements in design and miniaturization have allowed transvenous placement of ICDs to become routine. An epicardial rather than transvenous approach may be required in children, and less commonly in adults. In this surgical procedure one end of the lead is attached to the heart and the other end of the lead is attached to the pulse generator and placed in a pocket created under the skin of the abdomen.

Procedural complication rates range from three to six percent, with up to on-half of these considered serious. Complications include bleeding infections, lead dislodgement, pneumothorax, cardiac perforation, and rarely death. Perioperative mortality with transvenous ICD implantation has ranged from 0.2 to 0.4 percent. Lead-related complications, in addition to infection and dislodgement, include fracture and insulation defects. Most lead dislodgements and infections occur in the first three months following implantation, while lead fractures continue to occur during follow-up. Reported lead failure rates vary from one to nine percent at two years, two to fifteen percent and five years and five to forty percent at eight to ten years. Deaths related to lead failure have been reported but are exceedingly rare. The overall complication rate has decreased over the period from 2006 to 2010, a period that correlates with the introduction of an ICD registry in the US. In an observational study of 367,153 ICD recipients between April 2006 and March 2010, in-hospital complications and mortality significantly decreased from 3.7% during year one to 2.8% during year four (Piccini, 2021).

Additional problems associated with ICDs include inappropriate shock discharge, defibrillator storm with appropriate recurrent ICD discharge for recurrent ventricular tachyarrhythmias, inappropriate discharge for multiple reasons, infections related to implantation and exacerbation of heart failure when a high percentage of the heartbeats are paced from the right ventricle apex and ventricular function is already compromised.

When an ICD nears the end of battery life it is replaced. A pulse generator will last for five or more years in most patients. One study suggested that devices implanted after 2002 have significantly longer battery lives (5.6 versus 4.9 years), and single chamber ICDs implanted since 2002 had the longest battery life (mean 6.7 years).

Two categories of trials have investigated the use of ICDs for prevention of SCD. ICDs have been evaluated for primary (i.e., prophylactic) prevention of SCD in patients who have not experienced a life-threatening ventricular arrhythmia (or a symptomatic equivalent). Secondary prevention trials have evaluated the use of ICDs in patients who have had an abortive cardiac arrest, a life-threatening VT, or unexplained syncope with high probability that a ventricular tachyarrhythmia was the cause (Priori, et al., 2015; Zipes, et al., 2006).

Patel et al. (2016) reported on the gender, racial and health insurance differences in implantable cardioverter-defibrillator (ICD) utilization. The study used a hospitalization database to determine the trend of ICD utilization over the last decade and if disparities in gender, race, and insurance-payer changed over the last decade. The majority of ICDs were implanted in men age ≥ 65 years. Implantation of ICDs was 2.5x more common in men than in women (402 per million vs 163 per million). Approximately 95% of the ICDs were implanted in insured patients, and 5% were used in the uninsured population.

Several reviews have reported on the gender and racial disparities in clinical presentation, management, and outcome of hypertrophic cardiomyopathy (HCM) and heart failure. Black patients with HCM are more likely to present with heart failure but are less commonly referred for symptom management, sudden cardiac death stratification, surgical septal myectomy, or for implantable cardioverter-defibrillators. However, there were no significant differences in clinical outcome between Black and White patient groups for rate of adverse HCM events (including SCD, HCM mortality, heart transplant, and all-cause mortality). Prevalence of bystander cardiopulmonary resuscitation is lower for Black patients than for White patients. Finally, Black patients with HCM have decreased survival after hospital discharge following out-of-hospital cardiac arrest. Women presented with more comorbidities and more severe HF and more frequently non-ischemic cardiomyopathies but they were less likely to be referred for ICD therapy despite current guideline recommendations. ICD devices are underused in women and racial minorities independent of demographics, hospital characteristics, and comorbidities. Women and racial minorities also had higher rates of complications and greater resource use compared with men and those belonging to the White race (Chahine, et al., 2022; Patlolla, et al., 2022; Banerjee, et al., 2021; Ntusi and Sliwa, 2021; Regitz-Zagrosek, 2020; Zhao, et al., 2019; Patel, et al., 2016).

U.S. Food and Drug Administration (FDA): Multiple ICD devices have been approved by the U. S. Food and Drug Administration (FDA) through the Premarket Approval (PMA) process. Manufacturers of ICD devices include Biotronik (Lake Oswego, OR), Boston Scientific (Natick, MA), Sorin Group (Arvada, CO), Medtronic (Minneapolis, MN), and St. Jude Medical (St. Paul, MN).

Professional Societies/Organizations
American Heart Association (AHA)/American College of Cardiology (ACC)/Heart Failure Society of America (HFSA) Guideline for the Management of Heart Failure: The updated AHA/ACC/HFSA guidelines for the management of heart failure (HF) were published in 2022

(Heidenreich, et al., 2022). To develop the guidelines, the committee used the 2019 ACC/AHA evidence-based methodologies to assign each recommendation a Class of Recommendation and a Level of Evidence:

Class (Strength) of Recommendation:

- Class 1 (Strong)
  - Benefit >>>Risk
  - Intervention is recommended; is indicated/useful/effective/beneficial.
- Class 2a (Moderate)
  - Benefit>>Risk
  - Intervention is reasonable; can be useful/effective/beneficial.
- Class 2b (Weak)
  - Benefit ≥ Risk
  - Intervention may be reasonable; may be considered; its usefulness/ effectiveness is unknown/unclear/uncertain or not well-established.
- Class 3 No Benefit (Moderate)
  - Benefit=Risk
  - Intervention is not recommended/indicated/useful/effective/beneficial; it should not be performed/administered.
- Class 3 Harm (Strong)
  - Risk > Benefit
  - Intervention is not recommended/indicated/useful/effective/beneficial; it should not be performed/ administered.

Level (Quality) of Evidence:

- Level A
  - High-quality evidence from more than one RCT.
  - Meta-analyses of high-quality RCTs.
  - One or more RCTs corroborated by high-quality registry studies.
- Level B-R (Randomized)
  - Moderate-quality evidence from one or more RCTs.
  - Meta-analyses of moderate-quality RCTs.
- Level B-NR (Nonrandomized)
  - Moderate-quality evidence from one or more well-designed, well-executed nonrandomized studies, observational studies, or registry studies.
  - Meta-analyses of such studies
- Level C-LD (Limited Data)
  - Randomized or nonrandomized observational or registry studies with design or execution limitations.
  - Meta-analyses of such studies
  - Physiological or mechanistic studies in human subjects
- Level C-EO (Expert Opinion)
  - Consensus of expert opinion based on clinical experience.

The guideline stated that reevaluation of EF (> 40 days after MI, > 90 days after revascularization, > 90 days after GDMT) is useful to determine candidacy for implantable cardioverter defibrillator (ICD) or cardiac resynchronization therapy (CRT). For the primary prevention of SCD in patients who have heart failure with reduced ejection fraction (HFrEF) the guidelines made the following recommendations concerning ICD's (Heidenreich, et al., 2022):

- In patients with nonischemic DCM or ischemic heart disease who are at least 40 days post-MI with LVEF ≤ 35% and a NYHA class II or III symptoms on chronic GDMT, who have reasonable expectation of meaningful survival for >1 year, ICD therapy is recommended

for primary prevention of SCD to reduce total mortality (Class of Recommendation: 1; Level of Evidence: A).
- In patients at least 40 days post-MI with LVEF ≤ 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 (Class of Recommendation: 1; Level of Evidence: B-R)

Heart Rhythm Society (HRS): In 2022 the HRS published an expert consensus statement on evaluation and management of arrhythmic risk in neuromuscular disorders (NMD's). The cardiovascular presentation and management of patients with NMDs is dependent on the specific disorder. This consensus statement focused on the muscular dystrophies exhibiting prominent cardiac and arrhythmic manifestations, including Duchenne muscular dystrophy (DMD), Becker muscular dystrophy (BMD), limb-girdle muscular dystrophy type 2 (LGMD2) and limb-girdle muscular dystrophy type 1B (LGMD1B), myotonic dystrophy type 1 (DM1) and myotonic dystrophy type 2 (DM2), Emery-Dreifuss muscular dystrophy (EDMD), facioscapulohumeral muscular dystrophy (FSHD), and mitochondrial myopathies including Friedreich ataxia (FA) and Kearns-Sayre syndrome (Groh, et al., 2022).

The HRS recommended the following for the use of ICDs to manage arrhythmic risk in neuromuscular disorders (NMD's) using the 2019 ACC/AHA evidence-based methodologies:

Indication	Recommendation for ICD placement	COR/LOE*
Emery-Dreifuss and limb-girdle type 1B muscular dystrophies	In patients with DM1 or DM2 in whom ICD therapy is planned, an ICD system with permanent pacing capability is recommended.	1/B-NR
	In patients with DM1 or DM2 who are survivors of spontaneously occurring hemodynamically significant sustained VT or VF, ICD therapy is indicated if concordant with the patient's goals of care and clinical status.	1/B-NR
	In patients with DM1 or DM2 and an LVEF ≤ 35% despite guideline-directed medical therapy, ICD therapy is indicated if concordant with the patient's goals of care and clinical status.	1/B-NR
	In patients with DM1 or DM2 in whom clinically relevant ventricular arrhythmias are induced during electrophysiological study, ICD therapy is recommended if concordant with the patient's goals of care and clinical status.	1/B-NR
	In patients with DM1 or DM2 in whom permanent pacemaker implantation is indicated, ICD therapy may be considered if concordant with the patient's goals of care and clinical status.	2b/B-NR
Emery-Dreifuss and limb-girdle type 1B muscular dystrophies	In patients with EDMD or LGMD1B in whom ICD therapy is planned, an ICD system with permanent pacing capability is recommended.	1/B-NR
	In patients with EDMD or LGMD1B who are survivors of spontaneously occurring hemodynamically significant sustained VT or VF, ICD therapy is indicated if concordant with the patient's goals of care and clinical status.	1/B-NR

Indication	Recommendation for ICD placement	COR/LOE*
	In patients with EDMD or LGMD1B with at least one of the following: second-degree or third-degree AV block, PR interval ≥ 230 ms, or spontaneous HV ≥ 70 ms, ICD therapy is recommended if concordant with the patient's goals of care and clinical status.	1/B-NR
	In patients with EDMD or LGMD1B with an LVEF ≤ 35% despite guideline-directed medical therapy, ICD therapy is indicated if concordant with the patient's goals of care and clinical status.	1/B-NR
	In patients with EDMD or LGMD1B in whom clinically relevant ventricular arrhythmias are induced during electrophysiological study, ICD therapy is recommended if concordant with the patient's goals of care and clinical status.	1/B-NR
	In patients with EDMD or LGMD1B with LVEF < 45% and nonsustained VT, an ICD is reasonable if concordant with the patient's goals of care and clinical status.	2a/B-NR
	In patients with EDMD or LGMD1B with at least one of the following: LBBB, right bundle branch block (RBBB), or AF or AFL with slow ventricular response (ventricular rate < 50 bpm), ICD therapy is reasonable if concordant with the patient's goals of care and clinical status.	2a/C-LD
	In patients with EDMD or LGMD1B with symptomatic sinus node dysfunction or sinus bradycardia with heart rate < 40 bpm, ICD therapy may be considered if concordant with the patient's goals of care and clinical status	2b/C-LD
Mitochondrial myopathies including Friedreich ataxia	In patients with mitochondrial myopathies including FA with spontaneously occurring VF or sustained hemodynamically significant VT, ICD therapy is indicated if concordant with the patient's goals of care and clinical status.	1/B-NR
	In patients with mitochondrial myopathies including FA with an LVEF ≤ 35% despite guideline-directed medical therapy, ICD therapy is reasonable if concordant with the patient's goals of care and clinical status.	2a/B-NR

*2019 ACC/AHA evidence-based methodologies referenced under: Heidenreich, et al., 2022)

American College of Cardiology Foundation (ACCF)/American Heart Association (AHA): Additional recommendations for patient selection for ICDs in those with hypertrophic cardiomyopathy are included in guidelines for the diagnosis and treatment of hypertrophic cardiomyopathy (Ommen, et al., 2020).

The ACCF/AHA recommended the following for the use of ICDs using the 2019 ACC/AHA evidence-based methodologies that are referenced under: Heidenreich, et al., 2022:

Class I

- The decision to place an ICD in patients with HCM should include application of individual clinical judgment, as well as a thorough discussion of the strength of evidence, benefits, and risks to allow the informed patient's active participation in decision making *(Level of Evidence: C)*

- ICD placement is recommended for patients with HCM with prior documented cardiac arrest, ventricular fibrillation, or hemodynamically significant ventricular tachycardia (VT) *(Level of evidence: B)*

Class IIa

- It is reasonable to recommend an ICD for patients with HCM with:
  - Sudden death presumably caused by HCM in one or more first-degree relatives who are ≤ 50 years of age; *(Level of Evidence: B)*
  - LV wall thickness greater than or equal to 30 mm *(Level of Evidence: B)*
  - One or more recent unexplained syncopal episodes *(Level of Evidence: B)*
  - Any size left ventricular apical aneurysm *(Level of Evidence: B)*
  - Left ventricular systolic dysfunction EF < 50 *(Level of Evidence: B)*

- It is reasonable to recommend an ICD for children with HCM and ≥ 1 conventional risk factor (e.g., unexplained syncope, massive LVH, NSVT, family history of early HCM-related SCD) after considering the relatively high complication rates of long-term ICD placement. *(Level of Evidence: B)*

- It is reasonable to recommend an ICD for patients ≥ 16 years of age with HCM and with ≥ 1 major SCD risk factor after a discussion of the estimated 5-year sudden death risk and mortality rates. (Level of Evidence: B)

Class IIb

- The usefulness of an ICD is uncertain in patients with HCM and no major SCD risk factors *(Level of Evidence: B)*

- ICD may be considered in patients with extensive LGE by contrast-enhanced CMR imaging or NSVT present on ambulatory monitoring. (*Level of Evidence: B)*

- The usefulness of an ICD is uncertain in select pediatric patients with HCM in whom risk stratification is otherwise less certain and it may be useful to consider additional factors such as extensive LGE on contrast-enhanced CMR imaging and systolic dysfunction in risk stratification. (Level of Evidence: C)

Class III: Harm

- ICD placement as a routine strategy in patients with HCM without an indication of increased risk is potentially harmful. *(Level of Evidence: B)*

- ICD placement as a strategy to permit patients with HCM to participate in competitive athletics is potentially harmful. *(Level of Evidence: B)*

As stated above, the guideline includes a recommendation for ICD use in high-risk children with HCM, based on unexplained syncope, massive LV hypertrophy, or family history of SCD, after taking into account the relatively high complication rate of long-term ICD implantation. Although not defined in the guideline, massive LV hypertrophy is generally considered to be a maximal wall thickness approximately three times greater than normal. The authors note that the rate of inappropriate shocks and lead fractures appears to be higher in children than in adults, primarily because their activity level and body growth places continued strain on the leads, which are the weakest link in the system. This is of particular concern, considering the long period of time young patients will have prophylactically implanted devices. Other treatment options that may be considered for children with HCM include pharmacological management and surgical septal myectomy.

Heart Rhythm Society (HRS): In 2019, the HRS published an expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy. Arrhythmogenic cardiomyopathy (ACM) incorporates a broad spectrum of genetic, systemic, infectious, and inflammatory disorders. This designation includes, but is not limited to, arrhythmogenic right/left ventricular cardiomyopathy, cardiac amyloidosis, sarcoidosis, Chagas disease, and left ventricular noncompaction. To develop the guidelines, the committee used the 2016 ACC/AHA evidence-based methodologies to assign each recommendation a Class of Recommendation and a Level of Evidence (Towbin, et al., 2019):

Guideline Class of Recommendation (COR) and Level of Evidence (LOE) are described as follows:

- Class (Strength) of Recommendation:
- Class I (Strong) Benefit >>>Risk
- Class IIa (Moderate) Benefit>>Risk
- Class IIb (Weak) Benefit ≥ Risk
- Class III No Benefit (Moderate) Benefit=Risk
- Class III Harm (Strong) Risk>Benefit

Level (Quality) of Evidence:

- Level A if the data were derived from high-quality evidence from more than one randomized clinical trial(RCT), meta-analyses of high-quality RCTs, or one or more RCTs corroborated by high-quality registry.
- Level B-R when data were derived from moderate quality evidence from one or more RCTs, or meta-analyses of moderate-quality RCTs.
- Level B-NR was used to denote moderate-quality evidence from one or more well-designed, well-executed nonrandomized studies, observational studies, or registry studies. This designation was also used to denote moderate-quality evidence from meta-analyses of such studies.
- Level C-LD when the primary source of the recommendation was randomized or nonrandomized observational or registry studies with limitations of design or execution, meta-analyses of such studies, or physiological or mechanistic studies of human subjects.
- Level C-EO was defined as expert opinion based on the clinical

The consensus statement issued the following recommendations for ICD placement (Towbin, et al., 2019):

Indication	Recommendation for ICD placement	COR/LOE
Arrhythmogenic Cardiomyopathy ACM	In individuals with arrhythmogenic cardiomyopathy ACM who have suffered a cardiac arrest with VT or VF, an ICD is recommended.	I/B-NR
	In individuals with ACM who have sustained VT not hemodynamically tolerated, an ICD is recommended.	I/B-NR
	In individuals with ACM and syncope suspected to be due to a ventricular arrhythmia, an ICD is reasonable.	IIa/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

Indication	Recommendation for ICD placement	COR/LOE
	In individuals with ACM (other than ARVC) and hemodynamically tolerated VT, an ICD is recommended.	I/B-NR
Arrhythmogenic Right Ventricular Cardiomyopathy	In individuals with arrhythmogenic right ventricular cardiomyopathy (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
Phospholamban Cardiomyopathy	In individuals with phospholamban cardiomyopathy and LVEF 45%, or NSVT, an ICD is reasonable.	IIa/B-NR
Lamin A/C ACM	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 lamin A/C ACM and an indication for pacing, an ICD with pacing capabilities is reasonable.	IIa C-LD
Secondary Prevention: Cardiac Amyloidosis	In individuals with cardiac amyloidosis who have survived a cardiac arrest, an ICD is recommended if meaningful survival greater than 1 year is expected.	I/C-EO
Primary Prevention: Cardiac Amyloidosis	In individuals with AL-type cardiac amyloidosis with nonsustained ventricular arrhythmias, a prophylactic ICD may be considered if meaningful survival greater than 1 year is expected. Guideline noted: Primary prevention ICD implantation remains controversial, and there are conflicting data on the prevention of SCD in cardiac Amyloidosis	IIb/B-NR
Left Ventricular Non-Compaction (LVNC)	ICD implantation is recommended in individuals with LVNC and evidence of ventricular tachyarrhythmias associated with syncope or resuscitated sudden death if meaningful survival greater than 1 year is expected.	I/B-NR
	ICD implantation is reasonable in individuals with LVNC and evidence of nonsustained VT associated with a reduced ejection fraction.	IIa/B-NR

American Heart Association (AHA)/American College of Cardiology (ACC)/Health Rhythm Society (HRS): The AHA/ACC/HRS 2017 guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death recommended the following for ICD placement using the Class of Recommendation (COR) and LOE system mentioned above by Towbin, et al. (2019) (Al-Khatib, et al., 2017):

Indication	Recommendation for ICD placement	COR/LOE
Adult Congenital Heart Disease	In patients with adult congenital heart disease and hemodynamically unstable VT, an ICD is recommended after evaluation and appropriate treatment for residual lesions/ventricular dysfunction if meaningful survival of greater than 1 year is expected.	I/B-NR

Indication	Recommendation for ICD placement	COR/LOE
	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 adults with repaired tetralogy of Fallot physiology and inducible VT/VF or spontaneous sustained VT, implantation of an ICD is reasonable if meaningful survival greater than 1 year is expected.	IIa/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 implantation for inducible sustained VA is reasonable if meaningful survival of greater than 1 year is expected.	IIa/B-NR
	In patients with adult congenital heart disease and severe ventricular dysfunction (LVEF <35%) and symptoms of heart failure despite GDMT or additional risk factors, ICD implantation may be considered if meaningful survival of greater than 1 year is expected.	IIb/B-NR
Arrhythmogenic Right Ventricular Cardiomyopathy	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 ≤35%), an ICD is recommended if meaningful survival 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 greater than 1 year is expected.	IIa/B-NR
Brugada Syndrome	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
Cardiac Channelopathies	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
Cardiac Sarcoidosis	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, 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.	IIa/B-NR

Indication	Recommendation for ICD placement	COR/LOE
	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
Catecholaminergic Polymorphic Ventricular Tachycardia	In patients with catecholaminergic polymorphic ventricular tachycardia and recurrent sustained VT or syncope, while receiving adequate or maximally tolerated beta blocker, treatment intensification with either combination medication therapy (eg, beta blocker, flecainide), left cardiac sympathetic denervation, and/or an ICD is recommended.	I/B-NR
Congenital Long QT Syndrome	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 asymptomatic patients with long QT syndrome and a resting QTc greater than 500 ms while receiving a beta blocker, intensification of therapy with medications (guided by consideration of the particular long QT syndrome type), left cardiac sympathetic denervation or an ICD may be considered.	IIb/B-NR
Coronary Artery Spasm	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
Early Repolarization "J-wave" Syndrome	In patients with early repolarization pattern on ECG and cardiac arrest or sustained VA, an ICD is recommended if meaningful survival greater than 1 year is expected.	I/B-NR
Heart Failue	In patients with HFrEF who are awaiting heart transplant and who otherwise would not qualify for an ICD (eg, NYHA class IV and/or use of inotropes) with a plan to discharge home, an ICD is reasonable.	IIa/B-NR
Heart Transplant	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
Hypertrophic Cardiomyopathy (HCM)	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 greater than 1 year is expected.	I/B-NR

Indication	Recommendation for ICD placement	COR/LOE
	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: a. Maximum LV wall thickness ≥30 mm	IIa/B=NR
	b. SCD in 1 or more first-degree relatives presumably caused by HCM	IIa/C-LD
	c. 1 or more episodes of unexplained syncope within the preceding 6 months	IIa/C-LD
	In patients with HCM who have spontaneous NSVT 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 greater than 1 year is expected.	IIa/B-NR IIa/C-LD
	In patients with HCM who have NSVT or an abnormal blood pressure response with exercise but do not have any other SCD risk modifiers, an ICD may be considered, but its benefit is uncertain.	IIb/B-NR IIb/B-NR
	In patients with an identified HCM genotype in the absence of SCD risk factors, an ICD should not be implanted	III/B-NR
Idiopathic Polymorphic VT/VF	In patients resuscitated from SCA due to idiopathic polymorphic VT or VF, an ICD is recommended if meaningful survival greater than 1 year is expected.	I/B-NR
Left Ventricular Assist Device	In patients with an LVAD and sustained VA, an ICD can be beneficial.	IIa/C-LD
Myocarditis	In patients with giant cell myocarditis with VF or hemodynamically unstable VT treated according to GDMT, an ICD and/or an antiarrhythmic medication may be considered if meaningful survival of greater than 1 year is expected.	IIb/C-LD
Neuromuscular Disorders	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 limbgirdle 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
Pregnancy	In pregnant patients needing an ICD or VT ablation, it is reasonable to undergo these procedures during pregnancy, preferably after the first trimester.	IIa/B-NR

Indication	Recommendation for ICD placement	COR/LOE
Primary Prevention of SCD in Patients with Ischemic Heart Disease	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
	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
	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/C-EO
Primary Prevention of SCD in Patients with Nonischemic Cardiomyopathy (NICM)	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 Lamin A/C 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.	Ia/B-NR
	In patients with NICM, HF with NYHA classI 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/C-EO
Secondary Prevention of SCD in Patients with Ischemic Heart Disease	In patients with ischemic heart disease, who either survive SCA due to VT/VF or experience hemodynamically unstable VT	I/B-R
	or stable sustained VT not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.	I/B-NR
	In patients with ischemic heart disease and unexplained syncope who have inducible sustained monomorphic VT	I/B-NR

Indication	Recommendation for ICD placement	COR/LOE
	on electrophysiological study, an ICD is recommended if meaningful survival of greater than 1 year is expected.	
Secondary Prevention of SCD in Patients with Nonischemic Cardiomyopathy (NICM)	In patients with NICM who either survive SCA due to VT/VF or experience hemodynamically unstable VT	I/B-R
	or stable sustained VT not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.	I/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 greater than 1 year is expected.	IIa/B-NR
Short QT Syndrome	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
Ventricular Arrhythmias (VA)	In patients with ischemic cardiomyopathy, NICM, or adult congenital heart disease who have syncope or other VA symptoms and who do not meet indications for a primary prevention ICD, an electrophysiological study can be useful for assessing the risk of sustained VT.	IIa/B-R

American Heart Association (AHA)/American College of Cardiology (ACC)/Health Rhythm Society (HRS): Using the same 2016 evidence guidelines for class of recommendation (COR) and level of evidence (LOE) mentioned by Towbin, et al., (2019) the AHA/ACC/HRS 2017 guideline for the evaluation and management of patients with syncope recommended the following for ICD placement (Shen, et al., 2017):

Indication	Recommendation for ICD placement	COR/LOE
Syncope	ICD implantation is recommended in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC) who present with syncope and have a documented sustained VA.	I/B-NR
	ICD implantation is recommended in patients with cardiac sarcoidosis presenting with syncope and documented spontaneous sustained VA.	I/B-NR
Unexplained Syncope	An ICD is recommended in patients with syncope of undetermined origin with clinically relevant and significant VA induced at the time of an EPS.	NA
	ICD therapy is reasonable for patients with unexplained syncope and nonischemic dilated cardiomyopathy with significant LV dysfunction.	NA
Syncope of suspected arrhythmic cause	ICD implantation is reasonable in patients with HCM presenting with ≥ 1 recent episodes of syncope suspected to be of arrhythmic nature.	NA
	ICD implantation is reasonable in patients with ARVC who present with syncope of suspected arrhythmic etiology.	IIa/B-NR

Indication	Recommendation for ICD placement	COR/LOE
	ICD implantation is reasonable in patients with cardiac sarcoidosis and syncope of suspected arrhythmic origin, particularly with LV dysfunction or pacing indication.	IIa/B-NR
	ICD implantation is reasonable in patients with Brugada ECG pattern and syncope of suspected arrhythmic etiology.	IIa/B-NR
	ICD implantation may be considered in patients with short-QT pattern and syncope of suspected arrhythmic etiology.	IIb/C-EO
	ICD implantation is reasonable in patients with LQTS and suspected arrhythmic syncope who are on beta-blocker therapy or are intolerant to beta-blocker therapy.	IIa/B-NR
	ICD implantation may be considered in patients with early repolarization pattern and suspected arrhythmic syncope in the presence of a family history of early repolarization pattern with cardiac arrest.	IIb/C-EO
Exercise or stress-induced syncope	ICD therapy is reasonable in patients with catecholaminergic polymorphic ventricular tachycardia (CPVT) and a history of exercise- or stress-induced syncope despite use of optimal medical therapy or left cardiac sympathetic denervation (LCSD).	IIa/B-NR

American College of Cardiology Foundation (ACCF)/Heart Rhythm Society (HRS)/American Heart Association (AHA)/American Society of Echocardiography (ASE)/Heart Failure Society of America (HFSA)/Society for Cardiovascular Angiography and Interventions (SCAI)/Society of Cardiovascular Computed Tomography (SCCT)/Society for Cardiovascular Magnetic Resonance (SCMR): The 2013 appropriate use criteria for implantable cardioverter-defibrillators and cardiac resynchronization therapy described the appropriate use of these devices for selected patient populations (Russo, et al., 2013). The authors stated that the appropriate use criteria should be used in conjunction with the ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities (Epstein et al., 2008) and the 2012 focused update of that guideline (Tracy, et al., 2012).

The appropriateness scores for each indication reflect the median score of the 17 technical panel members. The authors state that "The relationship of these criteria to existing guidelines was provided to the technical panel. In addition, extensive links to clinical trials and other literature regarding the role of ICD and CRT in each clinical scenario were provided to technical panel members. This document represents the current understanding of the clinical utility of ICD and CRT implantation in clinical practice as measured by physicians with a variety of backgrounds and areas of expertise. It is the goal that these criteria will help provide a guide to inform medical decisions and help clinicians and stakeholders understand areas of consensus as well as uncertainty, while identifying areas where there are gaps in knowledge that warrant additional investigation".

Recommendations are provided based on the following scoring method:

- **Median score 7–9:** Appropriate care: An appropriate option for management of patients in this population due to benefits generally outweighing risks; effective option for individual care plans, although not always necessary, depending on

physician judgment and patient-specific preferences (i.e., procedure is generally acceptable and is generally reasonable for the indication).

- **Median score 4–6:** May be appropriate for care: At times an appropriate option for management of patients in this population due to variable evidence or agreement regarding the benefit/risk ratio, potential benefit based on practice experience in the absence of evidence, and/or variability in the population; effectiveness for individual care must be determined by a patient's physician in consultation with the patient based on additional clinical variables and judgment along with patient preferences (i.e., procedure may be acceptable and may be reasonable for the indication).
- **Median score 1–3:** Rarely appropriate care: Rarely an appropriate option for management of patients in this population due to the lack of a clear benefit/risk advantage; rarely an effective option for individual care plans; exceptions should have documentation of the clinical reasons for proceeding with this care option (i.e., procedure is not generally acceptable and is not generally reasonable for the indication).

Generally, criteria that have been deemed Appropriate or May Be Appropriate in these scenarios often meet Class I, IIa, or IIb criteria in guideline documents, are supported by a critical mass of existing data, or were deemed by the technical panel to meet sufficient clinical judgment to be reasonable and appropriate.

Indications rated as Appropriate are detailed below; indications rated as May be Appropriate and Rarely Appropriate are outlined in the appropriate use criteria document described above.

The following indications were rated as Appropriate Care (median score 7-9):
Secondary Prevention

Coronary artery disease (CAD): ventricular fibrillation (VF) or hemodynamically unstable ventricular tachycardia (VT) associated with acute (< 48 hours) myocardial infarction (MI) (newly diagnosed, no prior assessment of left ventricular ejection fraction (LVEF)

- Total Revascularization Completed After Cardiac Arrest
  - VF or polymorphic VT during acute (< 48 hours) MI, NSVT 4 days post MI, Inducible VT/VF at EPS ≥ 4 days after revascularization, LVEF 36–49% (7)
  - VF or polymorphic VT during acute (< 48 hours) MI, LVEF ≤ 35% (8)
- Obstructive CAD with coronary anatomy not amenable to revascularization
  - VF or polymorphic VT during acute (< 48 hours) MI, no electrophysiologic study (EPS) done (7)

CAD: VF or Hemodynamically Unstable VT < 48 h (Acute) Post-Elective Revascularization

- No evidence for acute coronary occlusion, restenosis, preceding infarct, or other clearly reversible cause, LVEF ≤ 35% (7)

CAD: VF or Hemodynamically Unstable VT (No Recent MI [< 40 Days] Prior to VF/VT and/or No Recent Revascularization [3 Months] Prior to VF/VT)

- No identifiable transient and completely reversible causes. No need for revascularization identified by catheterization performed following VF/VT (9)
- No revascularization performed (significant CAD present at catheterization performed following VF/VT, but coronary anatomy not amenable to revascularization (9)
- Significant CAD identified at catheterization performed following VF/VT. Complete revascularization performed after cardiac arrest. LVEF ≤ 49% (7)
- Significant CAD identified at catheterization performed following VF/VT. Incomplete revascularization performed after cardiac arrest LVEF ≥ 50% (7)
- Significant CAD identified at catheterization performed following VF/VT. Incomplete revascularization performed after cardiac arrest. LVEF 36–49% (8)

- Significant CAD identified at catheterization performed following VF/VT. Incomplete revascularization performed after cardiac arrest. LVEF ≤ 35% (9)

CAD: VF or Hemodynamically Unstable VT During Exercise Testing Associated with Significant CAD
- No revascularization performed (significant CAD present at catheterization performed following VF/VT, but coronary anatomy not amenable to revascularization) (9)
- Significant CAD identified at catheterization performed following VF/VT. Complete revascularization performed after cardiac arrest. LVEF ≤ 35% (7)
- Significant CAD identified at catheterization performed following VF/VT. Incomplete revascularization performed after cardiac arrest LVEF ≥ 36% (7)
- Significant CAD identified at catheterization performed following VF/VT. Incomplete revascularization performed after cardiac arrest LVEF ≤ 35% (8)

No CAD, VF or Hemodynamically Unstable VT
- Dilated nonischemic cardiomyopathy (9)
- VF/Hemodynamically Unstable VT Associated With Other Structural Heart Disease
    - Myocardial Sarcoidosis (9)
    - Giant cell myocarditis (8)

Genetic Diseases with Sustained VT, VF
- Congenital long QT (9)
- Short QT (9)
- Catecholaminergic polymorphic VT (9)
- Brugada Syndrome (9)
- ARVC with successful ablation of all inducible monomorphic VTs (9)
- ARVC with unsuccessful attempt to ablate an inducible VT (9)
- ARVC without attempted ablation (9)
- Hypertrophic cardiomyopathy (9)

No Structural Heart Disease (LVEF > 50%) or Known Genetic Causes of Sustained VT/VF
- Idiopathic VF With Normal Ventricular Function
    - No family history of sudden cardiac death (9)
    - First degree relative with sudden cardiac death (9)

Syncope in Patients without Structural Heart Disease
- Unexplained Syncope in a Patient With Long QT Syndrome
    - While on treatment with beta blockers (9)
    - Not being treated with beta blockers (7)
- Unexplained Syncope in a Patient with Brugada ECG Pattern
    - No EPS performed (8)
    - EPS performed. No ventricular arrhythmia induced (8)
    - EPS performed. Sustained VT/VF induced (9)
- Unexplained Syncope in a Patient with Catecholaminergic Polymorphic VT
    - While on treatment with beta blockers (8)
    - Not being treated with beta blockers (8)

Syncope in Patients with Coronary Artery Disease
- Unexplained Syncope With Prior MI and No Acute MI, LVEF 36%–49%
    - EPS revealed inducible sustained VT/VF (9)

Unexplained Syncope with Prior MI and no Acute MI. LVEF ≤ 35%
- EPS not performed (9)
- Inducible VT/VF on EPS (9)

- Not inducible at EPS (8)

Syncope in Patients with Nonischemic Structural Heart Disease
- Unexplained Syncope in a Patient with Left Ventricular Hypertrophy, Without Criteria for Hypertrophic Cardiomyopathy
  - Left ventricular hypertrophy/hypertensive heart disease, LVEF ≤ 35% (8)
- Unexplained Syncope in a Patient with Nonischemic Cardiomyopathy
  - Nonischemic dilated cardiomyopathy, LVEF ≤ 35% (8)
  - Left ventricular non-compaction, LVEF 36%–49% (7)
  - Left ventricular non-compaction, ≤ 35% (8)
  - Hypertrophic cardiomyopathy (8)
  - Tetralogy of Fallot with prior corrective surgery (7)
- Unexplained syncope in a Patient With Arrhythmogenic Right Ventricular Cardiomyopathy
  - No EPS performed (7)
  - No induction of VT/VF at EPS (7)
  - Inducible VT/VF at EPS. All inducible VTs successfully ablated (7)
  - Inducible VT/VF at EPS. Ablation unsuccessful (8)

Sustained Hemodynamically Stable Monomorphic VT Associated with Structural Heart Disease
- CAD and prior MI
  - LVEF ≥ 36% (7)
  - LVEF ≤ 35% (9)
- CAD and prior MI. All inducible VTs successfully ablated. LVEF ≤ 35% (9)
- CAD and prior MI. Troponin elevation thought to be secondary to VT. All inducible VTs successfully ablated. LVEF 36%–49% (7)
- CAD and prior MI. Troponin elevation thought to be secondary to VT. All inducible VTs successfully ablated. LVEF ≤ 35% (8)
- Nonischemic dilated cardiomyopathy. LVEF ≥ 50% (7)
- Nonischemic dilated cardiomyopathy. LVEF 36%–49% (7)
- Nonischemic dilated cardiomyopathy LVEF ≤ 35% (9)
- Nonischemic dilated cardiomyopathy. All inducible VTs successfully ablated. LVEF 36%–49% (7)
- Nonischemic dilated cardiomyopathy. All inducible VTs successfully ablated. LVEF ≤ 35% (8)
- Bundle branch re-entry successfully ablated in a patient with nonischemic cardiomyopathy. LVEF 36%–49% (7)
- Bundle branch re-entry successfully ablated in a patient with nonischemic cardiomyopathy. LVEF ≤ 35% (8)

Primary Prevention

Post-Acute Myocardial Infarction (MI) (< 40 days) LVEF ≤ 30%
- Revascularized after Acute MI
  - Asymptomatic nonsustained ventricular tachycardia (NSVT) (> 4 days post MI). EPS with inducible sustained VT (EPS performed after revascularization, within 30 days of MI) (7)
  - Asymptomatic NSVT (> 4 days post MI). EPS with inducible sustained VT (EPS performed after revascularization, between 30 and 40 days after MI) (8)

- Not Revascularized. Obstructive CAD With Coronary Anatomy Not Amenable to Revascularization
  - Asymptomatic NSVT (> 4 days post MI). EPS with inducible sustained VT (EPS performed within 30 days of MI) (7)

- Asymptomatic NSVT (> 4 days post MI) EPS with inducible sustained VT (EPS performed between 30 and 40 days after MI) (8)

Post-Acute MI (≤ 40 days) LVEF 31%–40%

- Revascularized for acute MI
  - Asymptomatic NSVT (> 4 days post MI). EPS with inducible sustained VT (EPS performed after revascularization, within 30 days of MI) (7)
  - Asymptomatic NSVT (> 4 days post MI) EPS with inducible sustained VT (EPS performed after revascularization, between 30 and 40 days after MI) (7)

Post-Acute MI (≤ 40 days) and Pre-Existing Chronic Cardiomyopathy (≥ 3 Months)

- LVEF < 30% due to old infarction. NYHA class I (8)
- LVEF < 35% due to old infarction. NYHA class II–III (9)
- LVEF < 35% due to nonischemic causes. NYHA class II–III (8)

Post-MI (≤ 40 Days) and Need for Guideline-Directed Pacemaker Therapy Post-MI (e.g., Sick Sinus Syndrome (SSS), Complete Heart Block (CHB), or Other Indications for Permanent Pacemaker)

- LVEF ≤ 35% (7)

Post-Myocardial Infarction (> 40 Days) With Ischemic Cardiomyopathy

- No Recent Percutaneous Coronary Intervention (PCI) or Coronary Artery Bypass Graft (CABG)
  - LVEF < 30%, New York Heart Association (NYHA) Class I (8)
  - LVEF < 30%, New York Heart Association (NYHA) Class II or III (9)
  - LVEF 31%–35%. NYHA Class I (7)
  - LVEF 31%–35%. NYHA Class II or III (9)
  - LVEF 36%–40%. Asymptomatic NSVT. EPS with inducible sustained VT/VF (8)
- Recent PCI or CABG (≤ 3 months)
  - Pre-existing documented cardiomyopathy. LVEF ≤ 35% on guideline-directed medical therapy > 3 months before PCI/CABG (8)
  - LVEF ≤ 35%. Need for permanent pacemaker post-revascularization (e.g., SSS, CHB, or other guideline-directed indications for permanent pacemaker) (8)

Duration of Guideline-Directed Medical Therapy for Ischemic Cardiomyopathy without Recent MI (Revascularization Not Indicated)

- LVEF ≤ 35%. On guideline-directed medical therapy for < 3 months, NSVT, EPS with inducible sustained VT (8)
- LVEF ≤ 35%. On guideline-directed medical therapy ≥ 3 months (9)

Nonischemic Cardiomyopathy

- At Least 3 Months on Guideline-Directed Medical Therapy
  - LVEF < 30%, NYHA Class I (7)
  - LVEF < 30%, NYHA Class II or III (9)
  - LVEF 31%–35%, NYHA Class I (7)
  - LVEF 31–35%, NYHA Class II or III (9)

Specific Etiologies

- Sarcoid heart disease, myotonic dystrophy, or Chagas disease, with LVEF ≤ 35% (8)
- Giant cell myocarditis, LVEF ≤ 35% (8)
- Giant cell myocarditis, LVEF > 35% (7)
- Peripartum cardiomyopathy, persists > 3 months postpartum (8)

Genetic Conditions (Excludes Syncope and Sustained VT, addressed above)

- Hypertrophic cardiomyopathy with 1 or more risk factors (7)
  - Major risk factors:
    - prior cardiac arrest, spontaneous nonsustained VT, family history of SCD, LV thickness greater than or equal to 30 mm, and an abnormal blood pressure response to exercise
  - Possible risk factors
    - AF, myocardial ischemia, LV outflow obstruction, high-risk mutations, and intense (competitive) physical exertion
- Arrhythmogenic right ventricular dysplasia/cardiomyopathy with no symptoms due to arrhythmia (7)
- Congenital long QT Syndrome with 1 or more risk factors, receiving guideline-directed medical therapy (7)
  - Risk factors:
    - sudden cardiac arrest, strong family history of SCD or when compliance or intolerance to drugs is a concern
- Catacholiminergic polymorphic VT with nonsustained VT (without syncope)
  - Not receiving beta-blockers, flecainide, or propafenone (7)
  - Receiving beta-blockers (7)
  - Not tolerating or breakthrough nonsustained ventricular arrhythmias on beta-blockers (8)
- Incidentally discovered Brugada by ECG (type I ECG pattern) in the absence of symptoms or family history of sudden cardiac death, with inducible VT or VF at EPS (7)
- Familial dilated nonischemic cardiomyopathy (RV/LV) associated with sudden cardiac death
  - Evidence of structural cardiac disease, but LVEF > 35% (7)
  - LV non-compaction with LVEF > 35% (7)

Use Outside the U.S.

In 2022, the European Society of Cardiology (ESC) updated the 2015 guidelines for the management of patients with ventricular arrhythmias (VA) and the prevention of sudden cardiac death (SCD). The ESC stated that ICD is an integral part of treating patients surviving a CA due to a VA or those deemed to be at high risk of SCA (Zeppenfeld, et al., 2022).

The ESC also noted that while working up a patient for ICD therapy, it is of extremely important to consider the patient's life expectancy, quality of life, and comorbidities, and to reassess and discuss these issues with the patient at the time of generator change. Additionally, there is evidence that patients with end-stage renal disease, with diabetes, and elderly patients benefit less or not at all from a primary prevention ICD. Women have been underrepresented in all primary prevention trials and data suggest that they may benefit less. A general guideline to follow is that the SCD risk needs to be weighed against the individual's competing risk of a non-arrhythmic death.

This guideline provides practical, evidence-based recommendations which are classified as Class I, Class IIa, Class IIb, and Class III.

The classification system is described as follows:

- Class I: Evidence and/or general agreement that a given treatment or procedure is beneficial, useful, effective.
- Class II: Conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of the given treatment or procedure.
  - Class IIa: Weight of evidence/opinion is in favor of usefulness/efficacy.
  - Class IIb: Usefulness/efficacy is less well established by evidence/opinion.

- Class III: Evidence or general agreement that the given treatment or procedure is not useful/effective, and in some cases may be harmful.

The weight of evidence supporting each recommendation is classified as follows:

- Level of evidence A: Data derived from multiple randomized clinical trials or meta-analyses.
- Level of evidence B: Data derived from a single randomized clinical trial or large non-randomized clinical trials.
- Level of evidence C: Consensus of opinion of the experts and/or small studies, retrospective studies, registries.

The guidelines issued the following recommendations for ICD placement for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death:

Indication	Recommendation for ICD placement	COR/LOE
Arrhythmogenic Right Ventricular Cardiomyopathy (AVRC)	ICD implantation should be considered in symptomatic patients with definite ARVC, moderate right or left ventricular dysfunction, and either NSVT or inducibility of SMVT at PES.	IIa/C
	ICD implantation is recommended in ARVC patients with hemodynamically not-tolerated VT or VF.	I/C
	ICD implantation should be considered in patients with definite ARVC and severe RV or LV systolic dysfunction.	IIa/C
	ICD implantation should be considered in patients with definite ARVC and an arrhythmic syncope.	IIa/B
	ICD implantation should be considered in ARVC patients with a hemodynamically tolerated SMVT.	IIa/C
Coronary Artery Disease	ICD therapy should be considered in patients with CAD, NYHA class I, and LVEF ≤30% despite ≥3 months of OMT	IIa/B
	ICD therapy is recommended in patients with CAD, symptomatic heart failure (NYHA class II–III), and LVEF ≤35% despite ≥ 3 months of OMT	Ia
	ICD implantation should be considered in patients with CAD, LVEF ≤ 40% despite ≥ 3 months of OMT, and NSVT, if they are inducible for SMVT by PES.	IIa/B
Cardiac Sarcoidosis	ICD implantation is recommended in patients with cardiac sarcoidosis who have an LVEF ≤35%.	I/B
	ICD implantation is recommended in patients with cardiac sarcoidosis who (1) have documented sustained VT, or (2) aborted CA.	I/B
	In patients with cardiac sarcoidosis who have an indication for permanent cardiac pacing related to high-degree AV block, ICD implantation should be considered, regardless of LVEF.	IIa/C
	In patients with cardiac sarcoidosis who have an LVEF ≤ 35% but significant LGE at CMR after resolution of acute inflammation, ICD implantation should be considered.	IIa/B

Indication	Recommendation for ICD placement	COR/LOE
	Patients with cardiac sarcoidosis, LVEF 35–50%, and inducible SMVT at PES, ICD implantation should be considered.	IIa/C
	In patients with cardiac sarcoidosis who have a LVEF 35–50% and minor LGE at CMR, after resolution of acute inflammation, PES for risk stratification should be considered.	IIa C
Chagas' Cardiomyopathy	In patients with Chagas' cardiomyopathy and symptomatic VT in whom AADs (amiodarone and beta-blockers) are ineffective or not tolerated, ICD implantation may be considered.	IIb/C
Coronary Heart Disease	In adults with CHD with biventricular physiology and a left systemic ventricle presenting with symptomatic heart failure (NYHA II/III) and EF ≤ 35% despite ≥3 months of OMT, ICD implantation is indicated.	I C
	In patients with CHD with presumed arrhythmic syncope and with either at least moderate ventricular dysfunction or inducible SMVT on PES, ICD implantation should be considered.	IIa C
	In patients with advanced single ventricle or systemic RV dysfunction with additional risk factors, ICD implantation may be considered.	IIb C
	In patients with CHD with not tolerated VT/aborted CA due to VF, ICD implantation is indicated after exclusion of reversible causes.	I/C
Dilated cardiomyopathy (DCM)/Hypokinetic non-dilated cardiomyopathy (HNDCM)	ICD implantation should be considered in DCM/HNDCM patients with a pathogenic mutation in LMNA gene, if the estimated 5-year risk of life-threatening VA is ≥ 10% and in the presence of NSVT or LVEF < 50% or AV conduction delay.	IIa/B
	ICD implantation should be considered in DCM/HNDCM patients with an LVEF ≤ 50% and ≥2 risk factors (syncope, LGE on CMR, inducible SMVT at PES, pathogenic mutations in LMNA, PLN, FLNC, and RBM20 genes).	IIa/C
	ICD implantation should be considered in patients with DCM/HNDCM and hemodynamically tolerated SMVT.	IIa/C
	ICD implantation should be considered in patients with DCM/HNDCM, symptomatic heart failure (NYHA class II–III) and LVEF ≤ 35% after ≥3 months of OMT.	IIa/A
	ICD implantation is recommended in patients with DCM/HNDCM, who survive SCA due to VT/VF or experience hemodynamically not-tolerated SMVT.	I/B
Hypertrophic cardiomyopathy (HCM)	ICD implantation should be considered in HCM patients aged 16 years or more with an intermediate 5-year risk of SCD (≥ 4 to < 6%), and with (a) significant LGE at CMR (usually ≥15% of LV mass); or (b) LVEF < 50%; or (c) abnormal blood pressure	IIa/B

Indication	Recommendation for ICD placement	COR/LOE
	response during exercise test; or (d) LV apical aneurysm; or (e) presence of sarcomeric pathogenic mutation.	
	In children less than16 years of age with HCM and an estimated 5-year risk of SCD ≥ 6% (based on HCM Risk-Kids score), ICD implantation should be considered.	IIa/B
	ICD implantation may be considered in HCM patients aged 16 years or more with an estimated 5-year risk of SCD of ≥4 to < 6%.	IIb B
	ICD implantation is recommended in HCM patients with hemodynamically not-tolerated VT or VF.	I/B
	ICD implantation may be considered in HCM patients aged 16 years or more with a low estimated 5-year risk of SCD (< 4%) and with (a) significant LGE at CMR (usually ≥15% of LV mass); or (b) LVEF < 50%; or (c) LV apical aneurysm.	IIb/B
Brugada Syndrome	ICD implantation is recommended in patients with BrS who: (a) Are survivors of an aborted CA and/or (b) Have documented spontaneous sustained VT.	I C
	ICD implantation should be considered in patients with type 1 Brugada pattern and an arrhythmic syncope.	IIa C
	ICD implantation may be considered in selected asymptomatic BrS patients with inducible VF during PES using up to 2 extra stimuli.	IIb C
Left ventricular non-compaction (LVNC)	In patients with an LVNC cardiomyopathy phenotype based on CMR or echocardiography, implantation of an ICD for primary prevention of SCD should be considered to follow DCM/HNDCM recommendations.	IIa/C
Myotonic Dystrophy	ICD implantation is recommended in patients with myotonic dystrophy and SMVT or aborted CA not caused by BBR-VT.	I/C
	In myotonic dystrophy patients without AV conduction delay and a syncope highly suspicious for VA, ICD implantation should be considered.	IIa/C
	In myotonic dystrophy patients with palpitations highly suspicious for VA and induction of a non-BBR-VT, ICD implantation should be considered.	IIa/C
	In patients with myotonic dystrophy undergoing ablation for BBR-VT, pacemaker/ICD implantation is recommended.	I C
	In patients with limb-girdle type 1B or Emery–Dreifuss muscular dystrophies and indication for pacing, ICD implantation should be considered.	IIa/C
	Implantation of an ICD may be considered in patients with Duchenne/Becker muscular dystrophy and significant LGE at CMR.	IIb/C

Indication	Recommendation for ICD placement	COR/LOE
	Implantation of an ICD over a permanent pacemaker may be considered in myotonic dystrophy patients with additional risk factors for VA and SCD.	IIb/C
Myocarditis	In patients with hemodynamically not-tolerated sustained VT or VF during the acute phase of myocarditis, ICD implantation before hospital discharge should be considered.	IIa/C
	In patients with hemodynamically not-tolerated SMVT occurring in the chronic phase of myocarditis, an ICD implantation is recommended.	I/C
Sustained monomorphic ventricular tachycardia (SMVT)	In patients with hemodynamically tolerated SMVT occurring in the chronic phase of myocarditis, ICD implantation should be considered.	IIa/C
	In patients with hemodynamically well-tolerated SMVT occurring in the chronic phase of myocarditis, preserved LV function and a limited scar amenable to ablation, catheter ablation may be considered as an alternative to ICD therapy, after discussion with the patient and provided that established endpoints have been reached.	IIb/C
	ICD implantation should be considered in patients with a hemodynamically tolerated SMVT and an LVEF ≥ 40% if VT ablation fails, is not available, or is not desired.	IIa/C
Light-chain amyloidosis or Transthyretin-associated cardiac amyloidosis	An ICD should be considered in patients with light-chain amyloidosis or transthyretin-associated cardiac amyloidosis and hemodynamically not-tolerated VT.	IIa/C
Andersen–Tawil syndrome	ICD implantation is recommended in patients with Andersen–Tawil syndrome after aborted CA or not-tolerated sustained VT.	I/C
	ICD implantation may be considered in patients with Andersen–Tawil syndrome who have a history of unexplained syncope or suffer from tolerated sustained VT.	IIb/C
Early repolarization syndrome/Early repolarization pattern	ICD implantation is recommended in patients with a diagnosis of ERS who have survived a CA.	I/B
	ICD implantation is not recommended in asymptomatic patients with an isolated ERP.	III C
	ICD implantation or quinidine may be considered in individuals with ERP and arrhythmic syncope and additional risk features.	IIb/C
	ICD implantation or quinidine may be considered in asymptomatic individuals who demonstrate a high-risk ERP in the presence of a family history of unexplained juvenile SD.	IIb/C

Indication	Recommendation for ICD placement	COR/LOE
Short QT syndrome	ICD implantation is recommended in patients with a diagnosis of SQTS who: (a) are survivors of an aborted CA and/or (b) have documented spontaneous sustained VT.	I/C
	ICD implantation should be considered in SQTS patients with arrhythmic syncope.	IIa/C
Transplant	In patients awaiting heart transplantation, ICD implantation for primary prevention should be considered.	IIa C
	In selected transplanted patients with cardiac allograft vasculopathy or treated rejection, ICD implantation may be considered.	IIb/C
Catecholaminergic polymorphic ventricular tachycardia	ICD implantation should be considered in patients with CPVT who experience arrhythmic syncope and/or documented bidirectional/PVT while on the highest tolerated beta-blocker dose and on flecainide.	IIa/C
Long QT syndrome	ICD implantation in addition to beta-blockers is recommended in LQTS patients with CA.	I/B
	ICD implantation is recommended in patients with LQTS who are symptomatic while receiving beta-blockers and genotype-specific therapies.	I/C
	Either ICD implantation or LCSD should be considered in patients with symptomatic LQTS, when beta-blockers and genotype-specific therapies are not tolerated or contraindicated at the therapeutic dose.	IIa/C
	ICD implantation may be considered in asymptomatic LQTS patients with high-risk profile CA (according to the 1-2-3 LQTS Risk calculator) in addition to genotype-specific medical therapies (mexiletine in LQT3 patients).	IIb
Ventricular Arrhythmias	ICD implantation is recommended in idiopathic VF.	I/B
	ICD implantation is recommended in patients with documented VF or hemodynamically not-tolerated VT in the absence of reversible causes.	I/A
	ICD implantation is recommended in patients without ongoing ischemia with documented VF or hemodynamically not-tolerated VT occurring later than 48 h after MI.	I/A
	ICD implantation should be considered in LVAD recipients with symptomatic sustained VAs.	IIa/B
Tetralogy of Fallot	In patients with repaired TOF who present with SMVT or recurrent, symptomatic appropriate ICD therapy for SMVT, catheter ablation performed in specialized centers is recommended.	I/C
	In patients after repair of TOF with arrhythmia symptoms and a positive PES, or a combination of other risk factors and a positive PES, ICD implantation should be considered.	IIa/C
Pregnancy	If ICD implantation is indicated during pregnancy, implantation is recommended with optimal radiation protection.	I/C

Indication	Recommendation for ICD placement	COR/LOE
Coronary artery spasm	In SCA survivors with coronary artery spasm implantation of an ICD should be considered.	IIa/C

In 2021 the European Society of Cardiology (ESC) updated the guidelines for the diagnosis and treatment of acute and chronic heart failure (McDonagh, et al., 2021). The plan of the guidelines is to help health professionals manage people with heart failure (HF) according to the best available evidence. This guideline used the evidence-based recommendations listed above in the guidelines for the management of patients with ventricular arrhythmias (VA) and the prevention of sudden cardiac death (SCD) (Zeppenfeld, et al., 2022).

The ESC explained that ICDs are effective for treating and preventing potentially lethal ventricular arrhythmias, and if using a transvenous systems, can also prevent bradycardia. There are some antiarrhythmic drugs that have the potential to reduce the rate of tachyarrhythmias and sudden death, however they do not reduce overall mortality.

The guidelines issued the following recommendations for ICD placement to reduce the risk of sudden death and all-cause mortality in heart failure:

- In patients with symptomatic HF (NYHA class II–III) of an ischaemic aetiology (unless they have had a MI in the prior 40 days), and an LVEF ≤ 35% despite ≥ 3 months of OMT, provided they are expected to survive substantially longer than 1 year with good functional status (Class of Recommendation: I; Level of Evidence: A).
- An ICD should be considered to reduce the risk of sudden death and all-cause mortality in patients with symptomatic HF (NYHA class II–III) of a non-ischaemic aetiology, and an LVEF ≤ 35% despite ≥ 3 months of OMT, provided they are expected to survive substantially longer than 1 year with good functional status (Class of Recommendation: II; Level of Evidence: A).
- In patients who have recovered from a ventricular arrhythmia causing haemodynamic instability, and who are expected to survive for >1 year with good functional status, in the absence of reversible causes or unless the ventricular arrhythmia has occurred < 48 h after a MI (Class of Recommendation: I; Level of Evidence: A).
- ICD implantation is not recommended within 40 days of a MI as implantation at this time does not improve prognosis (Class of Recommendation: III; Level of Evidence: A).
- ICD therapy is not recommended in patients in NYHA class IV with severe symptoms refractory to pharmacological therapy unless they are candidates for CRT, a VAD, or cardiac transplantation (Class of Recommendation: III; Level of Evidence: C).

A NICE Technology Appraisal Guidance on the Implantable Cardioverter Defibrillators and Cardiac Resynchronization Therapy for Arrhythmias and Heart Failure, updated in 2014, states that ICDs are recommended as options for:

- treating people with previous serious ventricular arrhythmia, that is, people who, without a treatable cause:
  - have survived a cardiac arrest caused by either ventricular tachycardia (VT) or ventricular fibrillation or
  - have spontaneous sustained VT causing syncope or significant hemodynamic compromise or have sustained VT without syncope or cardiac arrest, and also have an associated reduction in left ventricular ejection fraction (LVEF) of 35% or less but their symptoms are no worse than class III of the New York Heart Association (NYHA) functional classification of heart failure
- treating people who:

- have a familial cardiac condition with a high risk of sudden death, such as long QT syndrome, hypertrophic cardiomyopathy, Brugada syndrome or arrhythmogenic right ventricular dysplasia or
- have undergone surgical repair of congenital heart disease

Other Indications: Transvenous Implantable Cardioverter Defibrillator (ICD):
ICDs are indicated for primary and secondary prophylaxis of sudden cardiac death in selected patients which has been described above. There is insufficient evidence in the published peer-reviewed scientific literature to support the use of an ICD for any other indication, including but not limited to mitral annulus disjunction (MAD).

Mitral annular disjunction (MAD) is a structural abnormality where there is a separation between the mitral valve annulus and the left atrial wall which is not well understood. Mitral annular disjunction appears to be common in myxomatous mitral valve disease and mitral valve prolapse which can be detected on cardiac imaging. It is proposed that MAD can cause ventricular arrhythmias and sudden cardiac death. Treatment options have not been established.

Literature Review - Transvenous Implantable Cardioverter Defibrillator (ICD): Dejgaard et al. (2018) conducted a cross-sectional multicenter study that evaluated clinical presentations, morphology, association with mitral valve prolapse and ventricular arrhythmias in patients with mitral annular disjunction (MAD). The study aimed to clinically characterize patients with MAD and to describe the MAD morphology by echocardiography and advanced cardiac magnetic resonance (CMR) imaging. Additionally, the relationship between MAD and mitral valve prolapse (MVP) along with potential markers for ventricular arrhythmias were evaluated. Patients (n=116) were included if MAD was detected in any imaging modality. Echocardiography assessed for the presence of MVP and measured the MAD distance in the parasternal long axis. Cardiac magnetic resonance (CMR) measured circumferential MAD in the annular plane, longitudinal MAD distance, and myocardial fibrosis. Fourteen patients had severe arrhythmic events which were defined as aborted cardiac arrest and sustained ventricular tachycardia. The authors reported that patients with severe arrhythmic events were younger (p=0.001), had lower ejection fraction (p=0.002) and had more frequently papillary muscle fibrosis (p=0.03). Mitral valve prolapse was evident in 90 (78%) patients and was not associated with ventricular arrhythmias. The number of premature ventricular contractions per 24 hours and the prevalence of ventricular arrhythmia did not differ between MAD patients with and without concomitant MVP. Markers of ventricular arrhythmia were younger age, previous syncope, more premature ventricular contractions, papillary muscle fibrosis, and larger longitudinal MAD distance in the posterolateral wall assessed by CMR. Author noted limitations included: the study design, partial retrospective collection of arrhythmic events and only symptomatic patients seeking medical advice were included, which prevented the evaluation of MAD in the general population. Additionally, there was possible selection biases and the 24-h ECG recordings were not performed in all patients; however, there was no difference between frequency of 24 hour ECG recordings in patients with or without arrhythmias. The authors concluded that ventricular arrhythmias were frequent in patients with MAD. A total of 26 (22%) patients with MAD did not have MVP, and MVP was not associated with arrhythmic events. Future studies should address the cause of MAD and the mechanism of arrhythmias to which patients with this condition are prone.

Lee et al. (2017) conducted a prospective study that assessed the functional implication of mitral annular disjunction in mitral valve prolapse using 3D characterization. A total of 156 patients, which included 101 patients with MVP, 25 patients with functional mitral regurgitation (FMR) secondary to heart failure and 30 control subjects were referred for transesophageal echocardiography. The anatomic basis of abnormal annular structure and dynamics in MVP remains unresolved, and thus optimal treatment strategy is uncertain. The study aimed to assess that mitral annular disjunction (MAD) is associated with abnormal annular dynamics due to

decoupling of annular–ventricular function. All patients underwent 2-dimensional and 3D transesophageal echocardiographic examination for the evaluation of the leaflet and annular pathology. The spatial relation between atrial wall, mitral valve (MV), and left ventricle (LV) attachment was examined for MAD. The 3D extent of MAD and annular dynamics were quantitatively assessed. The LV global longitudinal strain and basal circumferential strains were measured by speckle tracking echocardiography. Of the 101 patients with MVP, MAD was seen in 42 patients (MAD + group) and measured 8.9 mm, circumferentially spanning 87 ± 41. The disjunctive annulus displayed paradoxical systolic expansion and flattening ($p<0.0001$), despite preserved and comparable LV strains with normal patients. The 3D extent of MAD correlated significantly with abnormal annular dynamics and larger regurgitant orifice ($p<0.0001$). In MVP patients without MAD, the LV global longitudinal strain correlated inversely with change in height ($p<0.0001$), whereas LV basal circumferential strain correlated with change in area ($p<0.0001$), but not in patients with MAD ($p>0.05$). The authors concluded that MAD is a common anatomic abnormality in MVP. Additional studies are needed to determine the prognostic importance of MAD in terms of the progression of primary mitral regurgitation and treatment strategies.

There is a paucity of well-designed evidence evaluating the standard defined work-up or defined treatment options for MAD. Well-designed studies are needed to assess the role of implantable cardioverter defibrillators (ICDs) in treating arrhythmias associated with MAD.

Subcutaneous ICD

The subcutaneous ICD (S-ICD) is an alternative to transvenous ICDs for selected patients. To implant the device, an incision is made in the left chest along the rib cage to create a pouch beneath the skin. A subcutaneous electrode is connected to the pulse generator, and the system is adjusted using an external programmer prior to closing the incisions. Since no electrodes are placed in or on the heart, investigators expect fewer perioperative and long-term vascular complications, problems with obtaining venous access, and lead complications. Avoiding the intravascular space has inherent limitations; however. The S-ICD cannot provide antitachycardia pacing, advanced diagnostics, or radiofrequency interrogation with remote monitoring. The S-ICD therefore would not be considered for patients with symptomatic bradycardia, incessant ventricular tachycardia, or spontaneous, frequently recurring ventricular tachycardia that is reliably terminated with anti-tachycardia pacing.

The median longevity of the first generation S-ICD system is reported as five years. The majority of devices were replaced because of battery depletion (Theuns, et al., 2015).

In the EFFORTLESS Registry, discussed below Lambiase et al. (2014), the rate of complications requiring reintervention within 360 days was 6.4%. Complication rates among various publications on the S-ICD range from 1.3 to 19%. Inappropriate shocks are one of the most common and concerning complications, with most studies reporting an incidence of 4-16%. The most common cause is over sensing of T-waves. Inappropriate shocks are more likely to occur in younger, physically active patients. Pocket infections have been reported in 1–10% of implantations, and complicated infections requiring device explantation have been reported in 1–4% of patients. Lead dislodgement or migration has been reported in 3–11% of patients and is thought to result from vigorous physical activity without adequate fixation of the parasternal lead. Suture sleeves are currently used to anchor the parasternal lead in order to eliminate lead dislodgement and migration. Less common complications that may require reintervention include skin erosion, premature battery depletion, or explantation due to the need for antitachycardia/bradycardia pacing or a new indication for resynchronization therapy.

U.S. Food and Drug Administration (FDA): The Subcutaneous Implantable Cardioverter Defibrillator (S-ICD™) System (Cameron Health, Inc., San Clemente, CA) received FDA approval through the PMA process on September 28, 2012. Cameron Health was subsequently acquired by

Boston Scientific. The S-ICD System is intended to provide defibrillation therapy for the treatment of life-threatening ventricular tachyarrhythmias in patients who do not have symptomatic bradycardia, incessant ventricular tachycardia, or spontaneous, frequently recurring ventricular tachycardia that is reliably terminated with anti-tachycardia pacing.

FDA approval was based on review of data from a 321-patient non-randomized study conducted at 33 investigational sites, in which 304 patients were successfully implanted with the S-ICD System (Weiss, et al., 2013). The primary safety endpoint was defined as the 180 day S-ICD system Type I complication-free rate. Type I complications were defined as clinical events caused by the device that required invasive intervention. The primary effectiveness endpoint was defined as the acute ventricular fibrillation conversion effectiveness rate of induced episodes. The panel concluded that the primary safety and efficacy endpoints were both met. The data provided reasonable assurance through response to induced and spontaneous episodes that the device functioned as intended, and the incidence of inappropriate shocks was comparable to that of transvenous ICDs.

According to the FDA approval letter, a post-approval trial is required, consisting of continued follow-up of patients who participated in the S-ICD IDE and prospective enrollment of patients with newly implanted devices. Approximately 1616 patients from approximately 50 investigational centers in the US are to be followed annually through 60 months post-implant, with at least 1025 evaluable at 60 months. The primary safety endpoint is the Type 1 complication-free rate at 60 months, and the primary effectiveness endpoint is the first-shock effectiveness in converting spontaneous discrete episodes of ventricular tachycardia/ventricular fibrillation through 60 months, which will be compared to a performance criterion of 94.0%. The S-ICD System post approval study can be found at ClinicalTrials.gov Identifier: NCT01736618.

On March 13, 2015 the EMBLEM™ S-ICD System (Boston Scientific Corp., St. Paul, MN) received FDA approval through the PMA process. FDA approval was based on the original PMA for the S-ICD (P110042/S043). The Emblem MRI S-ICD and the Emblem S-ICD subcutaneous electrode, insertion tool, and software application and programmer were approved (P110042/S058) on August 8, 2016.

Boston Scientific initiated a recall on 12/2/2020 for the EMBLEM S-ICD Subcutaneous Electrode (Model 3501) because of increased risk of fractures at a specific point. If the device fractures during use, this may lead to serious adverse events, including injury or death. Manufacturing Dates: March 2016 to present with Distribution Dates: June 2017 to present.

Boston Scientific initiated a recall on 12/2/2020 for the EMBLEM Subcutaneous Implantable Cardioverter Defibrillator (Model: S-ICD A209 and MRI S-ICD A219) because a manufacturing process may allow moisture to get inside the defibrillator and cause a short-circuit when it tries to deliver high voltage shocks. If this happens during use, this may lead to serious adverse events, including injury or death. Distribution Dates: June 1, 2015–September 30, 2019.

Literature Review - Subcutaneous ICD

Prospective Studies: Gold et al. (2022) reported the three-year results from the S-ICD Post-Approval Study with the 30-day results and one-year results previously reported by Gold et al. (2017) and Burke, et al. (2020). The S-ICD Post Approval Study is a US prospective registry to evaluate the short- and long-term safety and efficacy of the S-ICD system. At three years follow-up, the incidence and predictors of S-ICD–related infection were noted and used to develop an infection risk score. The baseline demographic characteristics and outcomes with 3-year postimplantation follow-up were compared between patients with and without device-related infection. A risk score was derived from multivariable proportional hazards analysis of 22 variables. Infection was observed in 55 patients (3.3%), with 69% of infections occurring within 90 days and a vast majority (92.7%) within 1 year of implantation. Late infections more likely

involved device erosion. There were not any infections noted after two years. The annual mortality rate postinfection was 0.6% per year. There were not any lead extraction complications or bacteremia related to infection. The ICD infection risk score was created with diabetes, age, prior transvenous ICD implant, and ejection fraction as predictors. Patients with a risk score of ≥ 3 had an 8.8 hazard ratio (95% confidence interval 2.8–16.3) of infection compared to a 0 risk score. The study concluded that infection rates in the S-ICD Post Approval Study were similar to other S-ICD populations and not associated with systemic blood-borne infections. The authors reported that late infection (> 1 year) is rare and associated with system erosion. Identifying high-risk subgroups may assist in developing preventive strategies to reduce further infection with this device. S-ICD implantation after TVICD infection is a viable approach that may be preferable to implanting another transvenous device. No health disparities were identified by the investigators.

Gold et al. (2021) conducted a prospective, multinational study to evaluate the outcomes of the S-ICD in primary prevention patients with low ejection fraction (UNTOUCHED trial). The trial was designed to evaluate the inappropriate shock (IAS) rate in a more typical, contemporary ICD patient population implanted with the S-ICD using standardized programming and enhanced discrimination algorithms. Primary prevention patients with an LVEF ≤ 35% (ischemic or nonischemic heart disease) eligible for S-ICD were enrolled in the study. The EMBLEM (Boston Scientific, Marlborough, MA) model A209 (Generation [Gen] 2) or A219 (Gen 3) S-ICD was implanted in 1116 patients with 1111 patients included in the 18 month postimplant follow-up analysis, including 808 patients (72.4%) from the United States. The cohort had a mean age of 55.8±12.4 years, 25.6% were women, 23.4% were Black, 53.5% had ischemic heart disease, 87.7% had symptomatic heart failure, and the mean left ventricular ejection fraction was 26.4±5.8%. The primary end point measured the IAS-free rate at 540 days (18 months) compared to a performance goal of 91.6%. Secondary end points measured freedom from all-cause shock at 18 months and procedure-related complications. Over the course of the study, 67 patients (6.0%) were lost to follow-up. At 18 months, the IAS-free rate is 95.9% meeting the performance goal of 91.6%. The freedom from all-cause shock rate was clinically significant at 90.6%, meeting the performance goal set to 85.8% ($p<0.0001$). Significant predictors of all-cause shock were a history of AF and lower LVEF ($p=0.009$ and $p=0.008$, respectively). The appropriate shock-free rate over 18 months was 94.3%. Conversion success rate for appropriate, discrete episodes was 98.4%. Complication-free rate at 18 months was 92.7%. The study concluded that high efficacy and safety was demonstrated with contemporary S-ICD devices and programming despite the relatively high incidence of comorbidities in comparison with earlier S-ICD trials. The inappropriate shock rate (3.1% at 1 year) is the lowest reported for the S-ICD and lower than many transvenous ICD studies using contemporary programming to reduce IAS.

Burke et al. (2020) reported the one-year results from the S-ICD Post-Approval Study with 30 day results previously reported by Gold et al. (2017). Two-hundred and fifteen patients were lost to follow-up and death was the most common reason (89 of 1,637). The complication-free rate at one year was 92.5%. The appropriate shock (AS) rate was 5.3%. A total of 395 ventricular tachycardia (VT) or fibrillation (VF) episodes were appropriately sensed with 131 of those self-terminating. Efficacy of the first and final shock (up to 5 shocks) for the 127 discrete appropriate shock episodes were 91.3% and 100.0%, respectively. Discrete AS episodes included 67 monomorphic VT (MVT) and 60 polymorphic VT (PVT)/VF, with first shock efficacy of 95.2% and 86.7%, respectively. There were 19 storm events in 18 subjects, with 84.2% conversion success. The authors concluded that during the first year after implantation, a predominantly primary prevention population with low ejection fraction demonstrated a high complication-free rate and spontaneous event shock efficacy for MVT and PVT/VF arrhythmias at rapid ventricular rates.

In a multicenter prospective study, Gold et al. (2017) reported results from the S-ICD Post-Approval Study. The objective of this registry is to evaluate the short- and long-term safety and efficacy of the S-ICD system. Patients deemed appropriate for implantation of an S-ICD system

were eligible for enrollment. Patients were excluded if they had a remaining life expectancy of one year or were ineligible for the S-ICD owing to bradycardia or a history of pace-terminable ventricular tachycardia. The primary and secondary safety end points were S-ICD system complication-free rate and electrode-related complication-free rate at 60 months. A total of 1637 patients underwent S-ICD implantation. The cohort included 68.6% (1123/1637) male patients, and 13.4% (220/1636) were receiving dialysis for endstage renal disease. The mean age was 52 6 15 years, with a mean left ventricular ejection fraction of 32.0%. Induced ventricular tachycardia/ventricular tachycardia was successfully converted in 98.7% (1394/1412) of patients. The 30-day complication-free rate was 96.2%. Predictors of complications included diabetes, younger age, and higher body mass index. Only perioperative outcomes are available at this time. A five-year follow-up of this cohort is planned.

A prospective case series (S-ICD® System Clinical Investigation [IDE] study) conducted by Weiss et al. (2013) evaluated the safety and effectiveness of a subcutaneous ICD (Cameron Health/Boston [n=330] Scientific) for treatment of life-threatening ventricular arrhythmias. Patients were enrolled if they were aged ≥ 18 years and had a guideline indication for ICD implantation. Patients with a life expectancy of less than one year were not enrolled. Patients with documented spontaneous and frequently recurring VT reliably terminated with antitachycardia pacing were excluded unless the patient was not a candidate for a transvenous ICD system. Patients with existing epicardial patches or subcutaneous electrodes in the left thoracic space were also excluded. Patients with unipolar pacemakers or pacing devices that revert to unipolar pacing could not participate in the study. Patients with an estimated glomerular filtration rate ≤ 29 mL/min per 1.73m^2 were excluded. The primary safety endpoint was 180 day complication-free rate compared with a prespecified performance goal of 79%. The primary effectiveness end-point was the induced VF conversion rate compared with a prespecified performance goal of 88%, with success defined as two consecutive ventricular fibrillation (VF) conversions of four attempts. Of 330 enrolled patients, implantation was attempted in 321 and was successful in 314. The 180 day system complication-free rate was 99%, and sensitivity analysis of the acute VF conversion rate was > 90%. There were 38 episodes of ventricular tachycardia/ventricular fibrillation recorded in 20 patients; all were successfully converted. Inappropriate shocks were received by 41 (13.1%) patients.

Non-Comparative Studies: Lambiase et al. (2022) reported the five year results of the EFFORTLESS study. The five-year results focused on late complications, inappropriate shock (IAS) rates, appropriate shock (AS) rates, shock efficacy, defibrillation testing (DFT) on generator replacement and mortality, along with specific analysis to understand the predictors of later events. Nine hundred and eighty-four of 994 enrolled patients with diverse diagnoses underwent S-ICD implantation. One hundred and seventy-one patients withdrew including 87 (8.8%) with device explanted, and 65 (6.6%) lost to follow-up. Of the explants, only 20 (2.0%) patients needed a transvenous device for pacing indications. At the median follow-up of 5.1 years; 703 patients remained in the study. At five years, all-cause mortality was 9.3% and the complication rate was 15.2%. First and final shock efficacy for discrete ventricular arrhythmias was consistent at 90% and 98%, respectively, with storm episode final shock efficacy at 95.2%. Time to therapy remained unaltered. Early complications did not predict later complications. There were no structural lead failures. Inappropriate shock rate at five years was 16.9% and self-terminating inappropriately sensed episodes predicted late IAS. Predictors of late AS included self-terminating appropriately sensed episodes and earlier AS. In this diverse S-ICD registry population, spontaneous shock efficacy was consistently high over five years. Very few patients underwent S-ICD replacement with a transvenous device for pacing indications. The authors concluded that a high level of VT/VF shock efficacy was maintained over the median 5.1-year follow-up, along with a low complication rate, a very low percentage of conversion to transvenous devices, and few IASs for AF/SVT.

Boersma et al. (2017) reported on the full EFFORTLESS cohort, which is the largest S-ICD database in the world with the longest follow-up. This observational nonrandomized standard of care registry included nearly 1000 patients at 42 clinical centers in 10 countries. Average follow-up was 3.1 years with 82 completing the study protocol 5-year visit. The primary goal of the EFFORTLESS registry is to demonstrate the safety of the S-ICD by evaluating complications and inappropriate shock rate. Patients eligible for implantation of an S-ICD system or with an S-ICD currently implanted at enrollment were eligible for inclusion. Exclusion criteria involved patients with spontaneous, incessant, or frequently recurring ventricular tachycardia (VT) amenable to ATP; patients with an indication for cardiac resynchronization therapy or symptomatic bradycardia, and patients with unipolar pacemakers or implanted systems that revert to unipolar pacing. Average age was 48 years, 28% were women, mean ejection fraction was 43 and 65% had a primary prevention indication. The S-ICD system and procedure complication rate was 4.1% at 30 days and 8.4% at 360 days. Few device extractions occurred due to need for antitachycardia (n=5), or biventricular (n=4) or bradycardia pacing (n=1). Inappropriate shocks occurred in 8.1% at 1 year and 11.7% after 3.1 years. At implant, 99.5% of patients had a successful conversion of induced ventricular tachycardia or ventricular fibrillation. The 1- and 5-year rates of appropriate shock were 5.8% and 13.5%, respectively. Conversion success for discrete spontaneous episodes was 97.4%. Infections requiring device removal occurred in 24 (2.4%) patients over the 3.1-year average follow-up. Infections requiring device removal were most common in the first year.

Burke et al. (2015) reported the 2-year pooled results of the IDE study and EFFORTLESS S-ICD Registry. The study included 882 subjects who underwent implantation of the S-ICD and were followed for 651 ± 345 days with a mean 22-month follow-up. Patients with recurrent VT reliably terminated with antitachycardia pacing and patients in need of pacing were excluded. Patients with end stage renal disease were excluded from the IDE trials. Spontaneous ventricular tachyarrhythmia (VT)/ventricular fibrillation (VF) events (n=111) were treated in 59 subjects; 100 (90.1%) events were terminated with 1 shock, and 109 events (98.2%) were terminated within the 5 available shocks. The estimated 3-year inappropriate shock rate was 13.1%. Estimated 3-year, all-cause mortality was 4.7% (95% confidence Interval [CI]: 0.9% to 8.5%), with 26 deaths (2.9%). Device-related complications occurred in 11.1% at 3 years. There were no electrode failures, and no S-ICD–related endocarditis or bacteremia occurred. Three devices (0.3%) were replaced for right ventricular pacing. The 6-month complication rate decreased by quartile of enrollment (Q1: 8.9%; Q4: 5.5%), and there was a trend toward a reduction in inappropriate shocks (Q1: 6.9%; Q4: 4.5%). The authors concluded that the S-ICD system demonstrated high efficacy for VT/VF. Complications and inappropriate shock rates were reduced consistently with strategic programming and as operator experience increased. The outcomes data from these trials, reported thus far, is reported as sufficient to demonstrate the safety and efficacy of the S-ICD devices for a limited subset of individuals who do not have a pacing requirement.

Lambiase et al. (2014) reported clinical, system, and patient related outcome data from S-ICD patients implanted since the commercial release of the S-ICD (n=472). The EFFORTLESS S-ICD Registry is an observational non-randomized standard of care evaluation conducted outside the US, where the S-ICD has been available since 2009. Of 471 patients, 241 were enrolled prospectively. The mean follow-up duration was 558 days (range 13-1342). The inclusion criteria included patients receiving an S-ICD. Specific contraindications included class I indications for permanent pacing, pace-terminable VT, and previously implanted functional unipolar pacing system. Seventy-two percent of patients were male, the mean age was 49 ± 18 years (range 9-88 years), with a mean ejection fraction of 42%. A total of 317 spontaneous episodes were recorded in 86 patients during the follow-up period; 169 of these (53%) received therapy (93 for VT/VF). One patient died of recurrent VF and severe bradycardia. First shock conversion efficacy was 88%, with 100% overall successful clinical conversion after a maximum of 5 shocks. A total of 73 inappropriate shocks were recorded in 32 patients over an average follow-up of 18 months (360-day inappropriate shock rate of 7%). The majority were due to oversensing of cardiac signals.

Procedure-related complication requiring intervention occurred in 29 (6.4%) patients. The most frequent complication was system infection (2.4%), with serious infection requiring implant removal in 10 patients), suboptimal electrode position/electrode movement (1.1%) and erosion or extrusion of the implanted electrode or pulse generator (0.9%).

Retrospective Studies: A retrospective analysis of 5760 patients from the National Cardiovascular Data Registry ICD Registry was performed to compare in-hospital outcomes among patients with a subcutaneous implantable cardioverter defibrillator (S-ICD) with those of patients with a single-chamber (SC)–ICD and dual-chamber (DC)–ICD (Friedman, et al., 2016). For the comparative analysis the population was restricted to individuals who were admitted for ICD implantation and were eligible for an S-ICD, single-chamber (SC)–ICD, or dual-chamber (DC)–ICD. The study excluded individuals with a previous ICD as well as those with bradycardia or resynchronization indication for permanent pacing or patients undergoing implantation during an acute hospitalization. The main outcomes measures were analysis of trends in S-ICD adoption as a function of total ICD implants and comparison of in-hospital outcomes (death, complications, and defibrillation threshold [DFT] testing) among S-ICD and transvenous (TV)-ICD recipients. A total of 3717 received S-ICDs. A total of 27.8% of the patients were female; the mean age was 67.03 years. Compared with SC-ICD and DC-ICD recipients, those with S-ICDs were more often younger, female, black, undergoing dialysis, and had experienced prior cardiac arrest. Among 2791 patients with S-ICD who underwent DFT testing, 2588 (92.7%), 2629 (94.2%), 2635 (94.4%), and 2784 (99.7%) were successfully defibrillated. The in-hospital complication rates associated with S-ICDs (0.9%) were comparable to those of SC-ICDs (0.6%) and DC-ICD rates (1.5%). Mean length of stay after S-ICD implantation was comparable to that after SC-ICD implantation and less than after DC-ICD implantation.

Olde Nordkamp et al. (2012) conducted a retrospective study to evaluate the efficacy and safety of the S-ICD in the first 118 patients implanted with the device at four high-volume ICD implantation centers in the Netherlands. Patients with a Class I or IIa indication for ICD therapy (according to the AHA/ACC/ESC 2006 guidelines for prevention of sudden cardiac death) were eligible for the device. The S-ICD was implanted without fluoroscopy using only anatomical landmarks. All patients were evaluated within two months after implantation and at six month intervals thereafter. Individual follow-up visits occurred if indicated (e.g., following shock therapy or complications). At 18 months of follow-up, 8 patients experienced 45 successful, appropriate shocks (98% first shock conversion efficacy). Fifteen patients (13%) received inappropriate shocks, primarily due to T-wave over-sensing. This issue was largely solved by a software upgrade and changing S-ICD settings. Sixteen patients (14%) experienced complications; the most frequent complications were infection (7, 5.9%) and lead dislodgement (3, 2.5%). Inappropriate shocks and complications were more common in the first 15 implantations per center, reflecting an apparent learning curve. The authors concluded that the S-ICD is a viable alternative to conventional ICD systems in selected patients. Randomized controlled trials with the S-ICD and transvenous ICD will further define the role of the S-ICD as an adjunctive or primary therapy in patients at risk for sudden cardiac death.

The safety and efficacy of implantable transvenous ICDs (T-ICD) in diverse patient populations has been demonstrated during three decades of use in over one million patients. The S-ICD has not been shown to be safe and effective in a diverse patient population, nor has it been shown to be non-inferior to the T-ICD. Although the primary safety and efficacy endpoints were met in the investigational device exemption (IDE) study on which FDA approval of the S-ICD was based, the study did not test the ability of the S-ICD to terminate spontaneous ventricular fibrillation. The IDE efficacy endpoint was based on detection and termination of induced VF, and did not demonstrate the efficacy of the S-ICD in ambulatory patients. Although the S-ICD is a promising technology, additional well-designed trials are needed to determine the long-term safety and efficacy of S-ICDs and to define patient selection criteria. An S-ICD may be indicated, however,

for selected patients at increased risk for bacteremia, patients with indwelling intravascular hardware at risk for endovascular infection, or in patients with compromised venous access.

In a discussion of the S-ICD, Knight et al. (UpToDate, 2021) notes that the S-ICD system obviates some of the mechanical complications associated with transvenous lead implantation, and the solid core design and lack of exposure to the repeated mechanical stresses of myocardial contraction may improve lead durability when compared to transvenous leads. The S-ICD system does have its own potential complications, including inappropriate shocks, pocked infection and lead dislodgement or migration. The authors note that limited data directly comparing the efficacy of the S-ICD with traditional ICDs, and that patient selection criteria are continuing to evolve. There are no guidelines for the selection of an S-ICD over a transvenous-ICD (TV-ICD). The authors consider several clinical factors in choosing a device for a patient with an indication for an ICD for primary or secondary prevention. The authors suggest that if there is no indication for transvenous pacing, cardiac resynchronization therapy, or antiachycardia pacing. An S-ICD may be considered in a patient less than age 45, or a patient with an indwelling central venous catheter, high risk for systemic infection, complex congenital heart disease or challenging vascular access, multiple prior transvenous endocardial leads, or a TV-ICD complication.

Professional Societies/Organizations
American Heart Association (AHA)/American College of Cardiology (ACC): The 2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy issued the following recommendation for a subcutaneous ICD using using the 2019 ACC/AHA evidence-based methodologies previously mentioned by Heidenreich, et al., 2022 (Ommen, et al., 2022).

- 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 VT termination (Class of Recommendation (COR): 1; Level of Evidence: B-NR)

The subtest of the guideline discussed the advantages and disadvantages of the subcutaneous ICD. The advantages included the lack of a transvenous lead, potentially fewer lead failures, and ease of removal. Disadvantages included the larger size of the device, the shorter battery longevity, potentially increased inappropriate shocks because of T-wave oversensing and myopotentials, and shorter history of use.

American Heart Association (AHA)/American College of Cardiology (ACC)/Heart Rhythm Society (HRS): The 2017 AHA/ACC/HRS Guideline for Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death (Al-Khatib et al.) provided the following recommendations using the Class of Recommendation (COR) and LOE system mentioned previously by Towbin, et al. (2019) for a subcutaneous implantable cardioverter-defibrillator:

Class 1

- 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 (Burke, et al., 2015; El-Chami, et al., 2015; Lambiase, et al., 2014; Weiss, et al., 2013; Bardy, et al., 2010). (Level of Evidence: B-NR).

The recommendation supportive text in the guideline states that difficulties in achieving venous access can prolong the implantation procedure and occasionally result in failed ICD implantation. These difficulties are likely to be encountered in patients with limited venous access such as

patients with ESRD. The risk of infection appears to be lower with subcutaneous implantable cardioverter-defibrillators than with transvenous ICDs. Therefore, a subcutaneous implantable cardioverter-defibrillator may be preferred in patients who are at high risk of infection, such as those with a prior device infection, ESRD, diabetes mellitus, or who are chronically immunosuppressed.

Class IIa

- 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 (Burke, et al., 2015; Lambiase, et al., 2014; Weiss, et al., 2013; Bardy, et al., 2010). (Level of Evidence: B-NR).

The recommendation supportive text in the guideline states that nonrandomized studies show that the subcutaneous implantable cardioverter-defibrillator reliably detects and converts VF during defibrillation threshold testing and successfully terminates spontaneous sustained VT that occurs during follow-up. An ongoing trial will compare the effect of the subcutaneous implantable cardioverter-defibrillator with that of the transvenous ICD on the outcomes of inappropriate shocks, complications, shock efficacy, and mortality (Olde Nordkamp, et al., 2012).

Class III: Harm

- 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 (Burke, et al., 2015; Lambiase, et al., 2014; Weiss, et al., 2013; de Bie, et al., 2013; Köbe, et al., 2013; Olde Nordkamp, et al., 2012; Bardy, et al., 2010). (Level of Evidence: B-NR).

The recommendation supportive text in the guideline states that the subcutaneous implantable cardioverter-defibrillator is incapable of bradycardia pacing, biventricular pacing, or antitachycardia pacing. Patients who need any of these types of pacing from an ICD should not be offered a subcutaneous implantable cardioverter-defibrillator. Some clinical scenarios may come up in which a transvenous pacemaker for bradycardia pacing in a patient with a subcutaneous implantable cardioverter-defibrillator- which is needed; this can be performed as long as the pacing is not unipolar. Leadless pacing devices for patients who require bradycardia pacing will be evaluated with the subcutaneous implantable cardioverter-defibrillator in the near future.

Use Outside the U.S.

In 2022, the European Society of Cardiology (ESC) updated the 2015 guidelines for the management of patients with ventricular arrhythmias (VA) and the prevention of sudden cardiac death (SCD). The ESC stated that ICD is an integral part of treating patients surviving a CA due to a VA or those deemed to be at high risk of SCA (Zeppenfeld, et al., 2022).

The ESC provided the following recommendation for a subcutaneous implantable cardioverter-defibrillator using the Class of Recommendation (COR) and LOE system for ICD's which was previously mentioned:

Class IIa

- Subcutaneous defibrillators should be considered as an alternative to transvenous defibrillators in patients with an indication for an ICD when pacing therapy for bradycardia support, cardiac resynchronization or antitachycardia pacing is not needed (Level of evidence: B).

The Canadian Cardiovascular Society/Canadian Heart Rhythm Society 2016 Implantable Cardioverter-Defibrillator Guidelines recommend an S-ICD be considered in patients with limited

vascular access or pocket sites in whom an ICD is recommended (Strong Recommendation; Low-Quality Evidence) (Bennett, et al., 2017).

The NICE guidance issued in December 2017, stated that current evidence on the safety and efficacy of subcutaneous implantable cardioverter defibrillator insertion for preventing sudden cardiac death is adequate to support the use of this procedure provided that standard arrangements are in place for clinical governance, consent and audit.

Substernal implantable cardioverter-defibrillator

The substernal ICD system, also known as extravascular ICD (EV ICD) with substernal lead placement, provides defibrillation and pacing therapies and has been proposed as an alternative to the available ICD systems. The substernal ICD system is an investigational device and not currently available. Evidence published to date evaluating the substernal ICD system is limited and studies that further evaluate safety and efficacy are currently in progress.

U.S. Food and Drug Administration (FDA): Currently, there is no FDA approval for the substernal ICD system.

Literature Review: Friedman et al. (2022) conducted a prospective, single-group, nonrandomized, premarket global clinical study that evaluated the safety and efficacy of the extravascular ICD system. The study included patients (n=316) with a class I or IIa indication for an ICD for primary or secondary prevention. The primary efficacy outcome measured the successful defibrillation at implantation. This outcome would be met if the lower boundary of the one-sided 97.5% confidence interval for the percentage of patients with successful defibrillation was greater than 88%. The primary safety outcome measured the freedom from major system- or procedure-related complications at six months. The safety outcome would be met if the lower boundary of the one-sided 97.5% confidence interval for the percentage of patients free from such complications was greater than 79%. Of the 356 patients were enrolled, 316 had an implantation attempt. Among the 302 patients in whom ventricular arrhythmia could be induced and who completed the defibrillation testing protocol, the percentage of patients with successful defibrillation was 98.7% (lower boundary of the one-sided 97.5% confidence interval [CI], 96.6%; $p<0.001$ for the comparison to the performance goal of 88%); 299 of 316 patients (94.6%) were discharged with a working ICD system. The estimate of the percentage of patients free from major system- or procedure-related complications at six months was 92.6% (lower boundary of the one-sided 97.5% CI, 89.0%; $p<0.001$ for the comparison to the performance goal of 79%). There were no major intraprocedural complications were reported. At six months, 25 major complications were observed, in 23 of 316 patients (7.3%). The success rate of antitachycardia pacing, as assessed with generalized estimating equations, was 50.8% (95% CI, 23.3 to 77.8). A total of 29 patients received 118 inappropriate shocks for 81 arrhythmic episodes. Eight systems were explanted without extravascular ICD replacement over the 10.6-month mean follow-up period. Limitations of the study included the lack of a comparison group and implantation was performed at expert centers, with a prespecified follow-up and testing plan. Additionally, the number of episodes of spontaneous arrhythmia was modest, and defibrillation testing may not be a good indicator of clinical shock efficacy. The authors reported that the study population was younger than typical ICD recipients and had a high frequency of hypertrophic cardiomyopathy, and may not be applicable to an older, sicker population and should be performed with caution. Testing at 6 months was performed in a subgroup of patients and was designed to assess maintained shock efficacy for ventricular arrhythmia and not the defibrillation threshold. Therefore, these data do not provide information on threshold changes over time. Observations regarding pause-prevention pacing are limited. The authors noted that women may have been slightly underrepresented in the trial, comprising 25.3% of enrolled patients compared to the estimate that women represent 30-40% of sudden cardiac deaths. No information on gender identity was collected in our study. For geographical representation, patients were enrolled at 46

sites in 17 countries across Australia, New Zealand, Canada, Europe, the Middle East, Hong Kong, and the United States in the Extravascular ICD Pivotal Study. Additional long term randomized control trials with large patient populations are needed to validate the outcomes of this study and establish the efficacy and safety of the extravascular ICD system.

Crozier et al. (2020) conducted a prospective, nonrandomized, pilot study at four centers in Australia and New Zealand that evaluated the safety and performance of a substernal implantable cardioverter-defibrillator (ICD). Eligible patients (n=21) were referred for ICD implantation with a Class I or IIa indication on the basis of current clinical practice guidelines. Among the 21 patients undergoing attempted implantation, 81% were men aged 22–77 years and 86% had primary ICD indications. Patients (n=21) received a substernal ICD system but one patient had to have the device explanted. The primary efficacy outcome measured the success of defibrillation testing during implantation. Ventricular fibrillation (VF) was induced via the device at implantation and defibrillation efficacy was tested by inducing, detecting, and converting VF episodes. Implantation required termination of VF with either a single 20-J shock or on two consecutive episodes with a 30-J shock. If the patient was successfully defibrillated at 20 J, defibrillation efficacy was assessed at 15 J. The primary safety outcome measured any complication related to the substernal ICD system or procedure that resulted in death, system revision, hospitalization, prolongation of a hospitalization, or permanent loss of defibrillation function due to device dysfunction. Patients received follow-up at two weeks, 4–6 weeks and three months after implantation. At the three-month follow-up, devices were interrogated, sensing and pacing tolerability testing performed, and chest radiography (day one, week two, weeks 4–6, and three months) and chest computed tomography (three months) performed. Among the 20 patients who completed defibrillation testing, 18 (90%) were able to be converted to sinus rhythm with 15 J (n=11), 20 J (n=4), or 30 J in two consecutive terminations (n=3) as required per protocol. The two patients who were successfully defibrillated at 15 J were tested at 10 J, and both were successful at 10 J. The two patients who did not pass defibrillation testing underwent explantation, with subsequent implantation of transvenous defibrillators. Among 20 patients who underwent successful implantation, the median defibrillation threshold was 15 J, and pacing was successful in 95% at ≥ 10 J. There were no intraprocedural complications. There were six adverse events that occurred within three months. One patient experienced an inappropriate shock 78 days post-implantation because of P-wave oversensing that occurred when the lead tip deflected toward the right atrial appendage. The system was subsequently explanted at 85 days post-implantation. The 90-day rate of freedom from systemic or procedural major complication was 94.1%. In addition to the single instance of inappropriate shock, two patients reported inspiratory discomfort post-operatively, and three had minor wound issues (two with swelling or impaired healing and one with superficial wound infection at the xiphoid incision site with minor purulent discharge, which resolved with an antibiotic course and a change of dressing), all of which resolved without sequelae. Fifteen patients remain under follow-up to date. Author noted limitations included short-term follow-up and the small patient cohort of predominantly male patients from a single geographic region. The study concluded that larger, longer-term evaluation will be needed to address the long-term sensing performance of the system and detection algorithms, whether predictors exist to ascertain probable defibrillation efficacy prior to implantation, how effectively ATP from a lead in this configuration performs relative to transvenous systems, and the extractability of the EV ICD system.

Boersma et al (2019) conducted the Acute Extravascular Defibrillation, Pacing, and Electrogram (ASD2) study which was a prospective multicenter, worldwide, nonrandomized, acute, proof-of-concept clinical trial. The study evaluated the feasibility of sensing, pacing, and defibrillation from an investigational lead designed specifically for the substernal space. An investigational lead was inserted into the substernal space via a minimally invasive subxiphoid access, and a cutaneous defibrillation patch or subcutaneous active can emulator was placed on the left mid-axillary line. Pacing thresholds and extracardiac stimulation were evaluated. Up to two episodes of ventricular

fibrillation were induced to test defibrillation efficacy. Eighty-seven patients were enrolled across 16 sites in Europe (n=54), the United States (n=19), New Zealand (n=10), Hong Kong (n=3), and Australia (n=1). Following data collection, the ASD2 research system was removed before the planned procedure of the patient. The investigational lead was placed in 79 patients. The investigational lead deployed successfully during the first insertion attempt in 66 patients (83.5%) and was redeployed (in 1–4 attempts) to achieve the preferred orientation in all remaining patients. Ventricular pacing was successful in at least one vector in 76 of 78 patients (97.4%), and 72 of 78 (92.3%) patients had capture in ≥ 1 vector with no extracardiac stimulation. A 30-J shock successfully terminated 104 of 128 episodes (81.3%) of ventricular fibrillation in 69 patients. Of the 79 patients who underwent the ASD2 study, there were seven adverse events in six patients adjudicated as causally (n=5) or as possibly (n=2) related to the ASD2 procedure. Four of the five adverse events adjudicated as being causally related to the ASD2 procedure resolved with no lasting effect on the patient; these included bleeding at the incision site, mild erythema at the incision, an episode of transient atrial fibrillation that occurred during VF induction, and reaction to anesthesia that resulted in low oxygen saturation. The fifth event was a pericardial effusion with tamponade. The authors concluded that the study demonstrated the ability to pace, sense, and defibrillate using a lead designed specifically for the substernal space. However, further evaluation is needed to assess the impacts of pacing and defibrillation on lead stability, patient movement or posture, and chronic tissue encapsulation, as well as long-term system management issues related to infection, system modification, or extraction.

Professional Societies/Organizations
Clinical guidelines that recommend use of a substernal implantable cardioverter-defibrillator are lacking.

Medicare Coverage Determinations

	Contractor	Determination Name/Number	Revision Effective Date
NCD	National	Implantable Automatic Defibrillators (20.4)	3/26/2019
LCD		No Determination found	

Note: Please review the current Medicare Policy for the most up-to-date information.
(NCD = National Coverage Determination; LCD = Local Coverage Determination)

Coding Information

Notes:
1. This list of codes may not be all-inclusive.
2. Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Transvenous Implantable Cardioverter Defibrillator (ICD)

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

CPT®* Codes	Description
33202	Insertion of epicardial electrode(s); open incision (eg, thoracotomy, median sternotomy, subxiphoid approach)
33203	Insertion of epicardial electrode(s); endoscopic approach (eg, thoracoscopy, pericardioscopy)

CPT®* Codes	Description
33216	Insertion of a single transvenous electrode, permanent pacemaker or implantable defibrillator
33217	Insertion of 2 transvenous electrodes, permanent pacemaker or implantable defibrillator
33224	Insertion of pacing electrode, cardiac venous system, for left ventricular pacing, with attachment to previously placed pacemaker or implantable defibrillator pulse generator (including revision of pocket, removal, insertion and/or replacement of existing generator)
33225	Insertion of pacing electrode, cardiac venous system, for left ventricular pacing, at time of insertion of implantable defibrillator or pacemaker pulse generator (eg, for upgrade to dual chamber system) (List separately in addition to code for primary procedure)
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 electrodes(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

HCPCS Codes	Description
C1721	Cardioverter-defibrillator, dual chamber (implantable)
C1722	Cardioverter-defibrillator, single chamber (implantable)
C1777	Lead, cardioverter-defibrillator, endocardial single coil (implantable)
C1882	Cardioverter-defibrillator, other than single or dual chamber (implantable)
C1883	Adaptor/extension, pacing lead or neurostimulator lead (implantable)
C1895	Lead, cardioverter-defibrillator, endocardial dual coil (implantable)
C1896	Lead, cardioverter-defibrillator, other than endocardial single or dual coil (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

Subcutaneous Implantable Cardioverter Defibrillator (S-ICD)

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

CPT®* Codes	Description
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
33999†	Unlisted procedure, cardiac surgery
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
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)

†Note: Considered medically necessary when used to report implantation of subcutaneous implantable cardioverter defibrillator (S-ICD).

Substernal implantable cardioverter-defibrillator

Considered Experimental/Investigational/Unproven:

CPT®* Codes	Description
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

CPT®* Codes	Description
	or other qualified health care professional, includes connection, recording and disconnection per patient encounter
0577T	Electrophysiological 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
0614T	Removal and replacement of a substernal implantable defibrillator pulse generator

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