Anthem

Clinical UM Guideline

Subject: Cardioverter Defibrillators
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Description

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

For information regarding other technologies for cardiac disease, see:

- MED.00055 Wearable Cardioverter Defibrillators
- CG-SURG-63 Cardiac Resynchronization Therapy with or without an Implantable Cardioverter Defibrillator for the Treatment of Heart Failure

Clinical Indications

Medically Necessary:

Implantable transvenous cardioverter-defibrillator (ICD) therapy is considered **medically necessary** when the criteria below have been met (I , II, **and** III):

I. Indications:
   A. Treatment of ventricular tachyarrhythmias; **or**
   B. Prevention of sudden cardiac death (SCD);
   and
II. Prior Treatment and Prognosis Criteria:
   A. Individual is
      1. On optimal medical therapy; **and**
      2. Has a reasonable expectation of survival with a good functional status for more than 1 year:
      and
III. Clinical criteria (*one* of the following is present [A through K]):
   A. After evaluation to define the cause of the event and to exclude any completely reversible causes in survivors of cardiac arrest due to either ventricular fibrillation (VF) or hemodynamically unstable sustained ventricular tachycardia (VT); **or**
   B. Those with structural heart disease and spontaneous sustained VT, whether hemodynamically stable or unstable; **or**
   C. Those with syncope of undetermined origin with clinically relevant, hemodynamically significant sustained VT; **or**
   D. Those with nonischemic dilated cardiomyopathy (NIDCM) who have an LVEF (left ventricular ejection fraction) less than or equal to 35% after 3 months of Guideline-directed medical therapy (GDMT) and who are in New York Heart Association (NYHA) functional Class II or III Heart Failure (HF); **or**
   E. Those with ischemic cardiomyopathy due to a prior myocardial infarction (MI) who are at least 40 days or more post-MI, with LVEF less than or equal to 30% and are in NYHA functional Class I HF after 3 months of GDMT or with an LVEF less than or equal to 35% and in NYHA Class II or III HF after 3 months of GDMT; **or**
   F. Those with nonsustained VT due to prior MI, LVEF less than 40%, and inducible VF or sustained VT at electrophysiological study; **or**
   G. Those with long-QT syndrome who are experiencing either syncope or VT while receiving beta blockers; **or**
   H. Those with confirmed hypertrophic cardiomyopathy (HCM) with **1 or more** of the following **major risk factors** for sudden cardiac death (SCD):
      1. Personal history of cardiac arrest or sustained ventricular arrhythmias; **or**
      2. Personal history of syncope suspected by clinical history to be arrhythmic within the previous 12 months; **or**
      3. Family history of HCM-related sudden death in one or more 1st or 2nd degree relatives who are less than or equal to 50 years of age or in two or more 3rd degree relatives who are less than or equal to 50 years of age; **or**
      4. LV apical aneurysm, independent of size; **or**
      5. LV systolic dysfunction (LVEF less than 50%); **or**
      6. Nonsustained VT episodes on ECG or continuous ambulatory electrocardiographic monitoring; **or**
      7. Left ventricular (LV) wall thickness greater than or equal to 30 mm in any LV segment **or**
   I. For individuals with symptomatic sustained VT in association with congenital heart disease who have undergone hemodynamic and electrophysiological evaluation; (catheter ablation or surgical repair may offer possible alternatives in carefully selected individuals); **or**
   J. For individuals with congenital heart disease with recurrent syncope of undetermined origin in the presence of either ventricular dysfunction or inducible ventricular arrhythmias at electrophysiological study; **or**
   K. For individuals with cardiac sarcoidosis when **one (1) or more** of the following are met:
      1. Sustained VT **or** survivors of sudden cardiac arrest **or** with an LVEF of 35% or less; **or**
      2. LVEF of greater than 35% with syncope **or** evidence of myocardial scar by cardiac MRI or PET scan; **or**
      3. LVEF of greater than 35% with inducible sustained ventricular arrhythmias.

Implantable transvenous cardioverter-defibrillator (ICD) therapy is considered **medically necessary** for individuals with a confirmed Brugada syndrome diagnosis when either of the following criteria is met (A or B):

A. History of unexplained syncope, documented spontaneous sustained VT with or without syncope, or survivor of a cardiac arrest; **or**
B. Family history of a first- or second-degree relative with sudden cardiac death due to Brugada syndrome or that is unexplained.

Subcutaneous cardioverter-defibrillator (S-ICD) devices are considered **medically necessary** for the following at-risk individuals when the medically necessary criteria above for implantable transvenous cardioverter-defibrillator (ICD) therapy have been met and

the individual does not require cardiac pacing:

A. Individuals with a lack of venous access; **or**
B. Individuals who are immunocompromised; **or**
C. Individuals with prosthetic valves; **or**
D. Individuals with recurrent transvenous lead-related, device-pocket or systemic infections; **or**
E. Individuals with endocarditis; **or**
F. Pediatric individuals.*****

****The FDA interprets pediatrics as individuals who are 21 years of age or younger (that is, up to, but not including, the 22nd birthday). See the Discussion/General Information section for additional information about use of ICD therapy in children.***

Note: *For use of combined ICD/Biventricular pacing (CRT-ICD) devices, in cases of NYHA Class IV heart failure and for other indications, see CG-SURG-63 Cardiac Resynchronization Therapy (CRT), with or without an Implantable Cardioverter Defibrillator (CRT/ICD) for the Treatment of Heart Failure.*

Not Medically Necessary:

The use of an implantable transvenous cardioverter-defibrillator is considered**not medically necessary** when the criteria above are not met and for any other indications.

The use of a subcutaneous ICD (S-ICD) is considered **not medically necessary** for all indications when the above criteria are not met.

Coding

The following codes for treatments and procedures applicable to this guideline are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

When services may be Medically Necessary when criteria are met:

CPT	
33202	Insertion of epicardial electrode(s); open incision (eg, thoracotomy, median sternotomy, subxiphoid approach) [when specified as ICD]
33203	Insertion of epicardial electrode(s); endoscopic approach (eg, thoracoscopy, pericardioscopy) [when specified as ICD]
33216	Insertion of a single transvenous electrode, permanent pacemaker or implantable defibrillator [when specified as ICD]
33217	Insertion of 2 transvenous electrodes, permanent pacemaker or implantable defibrillator [when specified as ICD]
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
33249	Insertion or replacement of permanent implantable defibrillator system with transvenous lead(s), single or dual chamber
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
93640	Electrophysiologic evaluation of single or dual chamber pacing cardioverter-defibrillator leads including defibrillation threshold evaluation (induction of arrhythmia, evaluation of sensing and pacing for arrhythmia termination) at time of initial implantation or replacement;
93641	Electrophysiologic evaluation of single or dual chamber pacing cardioverter-defibrillator leads including defibrillation threshold evaluation (induction of arrhythmia, evaluation of sensing and pacing for arrhythmia termination) at time of initial implantation or replacement; with testing of single or dual chamber pacing cardioverter-defibrillator pulse generator

HCPCS	
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)
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

ICD-10 Procedure	
02H40KZ-02H44KZ	Insertion of defibrillator lead into coronary vein [by approach; includes codes 02H40KZ, 02H43KZ, 02H44KZ]
02H60KZ-02H74KZ	Insertion of defibrillator lead into atrium [right or left, by approach; includes codes 02H60KZ, 02H63KZ, 02H64KZ, 02H70KZ, 02H73KZ, 02H74KZ]
02HK0KZ-02HL4KZ	Insertion of defibrillator lead into ventricle [right or left, by approach; includes codes 02HK0KZ, 02HK3KZ, 02HK4KZ, 02HL0KZ, 02HL3KZ, 02HL4KZ]
02HN0KZ-02HN4KZ	Insertion of defibrillator lead into pericardium [by approach; includes codes 02HN0KZ, 02HN3KZ, 02HN4KZ]
0JH608Z-0JH838Z	Insertion of defibrillator generator into subcutaneous tissue and fascia [chest or abdomen, by approach; includes codes 0JH608Z, 0JH638Z, 0JH808Z, 0JH838Z]
0JH60FZ-0JH63FZ	Insertion of subcutaneous defibrillator lead into chest subcutaneous tissue and fascia [by approach; includes codes 0JH60FZ, 0JH63FZ]

ICD-10 Diagnosis	
	All diagnoses including, but not limited to, the following:
D86.85	Sarcoid myocarditis
I21.01-I21.B	Acute myocardial infarction
I22.0-I22.9	Subsequent ST elevation (STEMI) and non-ST elevation (NSTEMI) myocardial infarction
I24.0-I24.9	Other acute ischemic heart disease
I25.10-I25.119	Atherosclerotic heart disease of native coronary artery
I25.2	Old myocardial infarction
I25.5	Ischemic cardiomyopathy
I25.810-I25.9	Other forms of chronic ischemic heart disease
I42.0-I42.9	Cardiomyopathy
I45.81	Long QT syndrome
I46.2-I46.9	Cardiac arrest
I47.0	Re-entry ventricular arrhythmia
I47.20-I47.29	Ventricular tachycardia
I49.01-I49.02	Ventricular fibrillation, ventricular flutter
Q24.8	Other specified congenital malformations of heart [Brugada syndrome]
Q24.9	Congenital malformation of heart, unspecified (congenital disease of heart)
R55	Syncope and collapse
Z82.41	Family history of sudden cardiac death
Z86.74	Personal history of sudden cardiac arrest

When services are Not Medically Necessary:
For the procedure codes listed above when criteria are not met or for situations designated in the Clinical Indications section as not medically necessary.

Discussion/General Information

Implantable cardioverter defibrillators (ICDs) are an important treatment option for individuals with a history of life-threatening ventricular arrhythmias. Randomized clinical trials have shown that ICD use significantly reduces mortality rates for those with coronary artery disease (CAD) and/or a prior myocardial infarction (MI) who have poor ventricular function. Although ICDs for the treatment of atrial fibrillation (AF) have been used in studies, evidence on efficacy and long-term outcomes is limited, and ICD therapy for AF is not considered to be in accordance with generally accepted standards of medical practice at this time.

Available literature indicates that ICDs are now widely used for the*secondary* prevention of sudden cardiac death (SCD), due to ventricular fibrillation (VF) or ventricular tachycardia (VT). ICD implantation is the generally accepted treatment for those who have experienced an episode of VF not accompanied by an acute MI or other transient or reversible cause. Accepted guidelines prefer this treatment in individuals with sustained VT, causing syncope or hemodynamic compromise. As *primary* prevention, the literature shows that ICD use is superior to conventional antiarrhythmic drug therapy for those who have survived an MI and who have spontaneous, non-sustained VT (NSVT), a low left ventricular ejection fraction (LVEF), and inducible VT at electrophysiological study (EPS).

Several national and international specialty societies have longstanding published guidelines addressing indications for ICD implantation. These include:

- 2002 - American College of Cardiology (ACC)/American Heart Association (AHA)/NASPE (North American Society of Pacing and Electrophysiology) guideline for implantation of cardiac pacemakers and antiarrhythmia devices (Gregoratos, 2002);
- 2003 - ACC/ European Society of Cardiology (ESC) Clinical Expert Consensus Document on Hypertrophic Cardiomyopathy (Maron, 2003);
- 2006 – ACC/AHA/ ESC/European Heart Rhythm Association (EHRA)/ Heart Rhythm Society (HRS) practice guidelines for the management of patients with ventricular arrhythmias and the prevention of SCD (Zipes, 2006);
- 2008 - ACC/AHA/HRS updated guideline for device-based therapy of cardiac rhythm abnormalities (Epstein, 2008);
- 2009 – ACC/AHA Guideline for Management of Heart Failure in Adults (Hunt, 2009);
- 2011 - ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy (Gersh, 2011);
- 2017 AHA/ACC/HRS Guideline for Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death (Al-Khatib, 2017);
- 2019 ACC/AHA Strategy for Prevention of Sudden Cardiac Death in High-Risk Patients with Hypertrophic Cardiomyopathy (Maron, 2019);
- 2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy (Ommen, 2020).

The 2008 ACC/AHA/HRS guideline addressed use of ICDs for children and adolescents with congenital heart disease:

> The indications for ICD implantation in young patients and those with congenital heart disease have evolved over the past 15 years based on data derived primarily from adult randomized clinical trials. Similar to adults, ICD indications (for pediatric patients) have evolved from the secondary prevention of SCD to the treatment of patients with sustained ventricular arrhythmias to the current use of ICDs for primary prevention in patients with an increased risk of SCD. However, in contrast to adults, there are minimal prospective data regarding ICD survival benefit, because fewer than 1% of all ICDs are implanted in pediatric or congenital heart disease (CHD) patients. Considerations, such as the cumulative lifetime risk of SCD in high-risk patients and the need for decades of antiarrhythmic therapy, make the ICD an important treatment option for young patients. Prospective identification and treatment of young patients at risk for sudden death is crucial because compared with adults, a very low percentage of children undergoing resuscitation survive to hospital discharge…Because of concern about drug-induced proarrhythmia and myocardial depression, an ICD (with or without cardiac resynchronization therapy [CRT]) may be preferable to antiarrhythmic drugs in young patients with dilated cardiomyopathy (DCM) or other causes of impaired ventricular function who experience syncope or sustained ventricular arrhythmias…The role of ICDs in primary prevention for children with genetic channelopathies, cardiomyopathies, and congenital heart defects should be defined more precisely and is an area in need of further research (Epstein, 2008).

In 2009, the ACC/AHA published a focused update to the 2005 guidelines for the diagnosis and management of heart failure (HF) in adults which gave a Class IIa recommendation for ICD placement in individuals with ischemic dilated cardiomyopathy (IDCM) who are at least 40 days post-MI, have an LVEF of 30% or less, are in NYHA functional class I HF on chronic optimal medical therapy, and have a reasonable expectation of survival with a good functional status for more than 1 year (Hunt, 2009). This was based, in part, on the findings of the SCD-HeFT trial previously described (Bardy, 2005) and has been incorporated into the medical necessity criteria for

adult indications.

The decision to place an ICD in an individual with IDCM or nonischemic dilated cardiomyopathy (NIDCM) is based, in part, on the measured LVEF. This measurement is subject to change over time based on medical interventions. The placement of an ICD should be reserved for individuals who have received an adequate trial of optimal medical management (Al-Khatib, 2011). In 2012, the term "Guideline-directed medical therapy" (GDMT) was adopted by the writing groups for the major specialty medical societies, (such as found in Tracy, 2012 and Yancy, 2013) to replace "Optimal medical therapy."

Regarding the timeframe generally considered by consensus as adequate to determine if GDMT has been effective prior to ICD placement is 3 to 6 months. In 2013, a report of the American College of Cardiology Foundation (ACCF) Appropriate Use Criteria Task Force, HRS, 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), and Society for Cardiovascular Magnetic Resonance (SCMR) was issued, in which the following is noted regarding the timeframe for GDMT:

> Patients who are going to receive substantial benefit from medical treatment alone usually show some clinical improvement during the first 3 to 6 months. Medical therapy is also assumed to include adequate rate control for tachyarrhythmias, including atrial fibrillation. Therefore, it is recommended that GDMT be provided for at least 3 months before planned reassessment of LV function to consider device implantation. If LV function improves to the point where primary prevention indications no longer apply, then device implantation is not indicated (Russo, 2013).

Beginning in 2003, a series of clinical expert consensus documents on hypertrophic cardiomyopathy (HCM) have been published by the ACC, ESC, the ACCF Task Force, and the ESC Committee for Practice Guidelines (Maron, 2003; Maron, 2010; Gersh, 2011). These documents have identified major risk factors for SCD in individuals with HCM, including:

- Family history of at least one HCM-related SCD (defined as SCD in at least 1 first-degree relative);
- At least one episode of unexplained recent syncope (defined as 1 or more episodes of unexplained loss of consciousness within the previous 12 months);
- Massive left ventricular (LV) hypertrophy (thickness greater than or equal to 30 mm);
- Nonsustained VT on ambulatory 24-hour (Holter) ECG (defined as a run of 3 or more consecutive ventricular beats at a rate of at least 120 beats/minute, lasting less than 30 seconds);
- Hypotensive or attenuated BP response to exercise (defined as failure of BP to rise, or as a fall in BP) (Elliott, 2000; Maron, 2010).

The 2011 ACCF and AHA guidelines for the diagnosis and treatment of HCM noted that the decision to implant an ICD must be individualized to the unique circumstances of each individual with HCM. They note that each of the identified SCD risk factors has low positive predictive value and propose that a combination of conventional risk factors and other risk modifiers may give the best indication of who should receive an ICD.

In 2020, the ACC/AHA issued an updated guideline for the diagnosis and treatment of patients with HCM, which is considered a full guideline revision intended to replace the former Gersh, 2011 guideline (Ommen, 2020). This document provides a comprehensive guide to the evaluation and management of HCM in adults and children, which is based on the current evidence, including relevant studies and other specialty society guidelines. Numerous modifications were made including recommendations for ICD therapy. The following updated recommendations for ICD therapy and risk stratification for SCD have been incorporated into the criteria for ICD therapy in this document:

> Class I, LOE: B-NR:
> For patients with HCM and previous documented cardiac arrest or sustained VT, ICD placement is recommended.
>
> Class IIa, LOE: B-NR:
> For adult patients with HCM with≥ 1 major risk factors for SCD, it is reasonable to offer an ICD.
> These major risk factors include:
> a. Sudden death judged definitively or likely attributable to HCM in≥ 1 first-degree or close relatives who are≤ 50 years of age;
> b. Massive LVH ≥ 30 mm in any LV segment;
> c. ≥ 1 Recent episodes of syncope suspected by clinical history to be arrhythmic (i.e., unlikely to be of neurocardiogenic [vasovagal] etiology, or related to LVOTO);
> d. LV apical aneurysm, independent of size;
> e. LV systolic dysfunction (EF < 50%).
>
> For children with HCM who have≥ 1 conventional risk factors, including unexplained syncope, massive LVH (left ventricular hypertrophy), NSVT, or family history of early HCM-related SCD, ICD placement is reasonable after considering the relatively high complication rates of long-term ICD placement in younger patients (Bharucha, 2015; Kamp, 2013; Maron, 2013, 2016; Miron, 2020; Moak, 2011; Norrish, 2017; Yetman, 1998).
>
> For patients ≥ 16 years of age with HCM and with≥ 1 major SCD risk factors, discussion of the estimated 5-year sudden death risk and mortality rates can be useful during the shared decision-making process for ICD placement (Ommen, 2020).

The following rationale is excerpted for the above recommendations:

> Over the past several decades, retrospective observational studies have identified a number of noninvasive clinical risk markers associated with increased risk for SCD events in HCM. In association with clinical judgment and shared decision-making, patients with HCM are considered potential candidates for primary prevention ICDs by virtue of ≥ 1 major risk markers, which together have a high sensitivity in predicting those patients with HCM at greatest future risk for SCD events…
> Patients with HCM who have experienced a previous documented cardiac arrest or hemodynamically significant VT/VF remain at significantly increased risk for future life-threatening ventricular tachyarrhythmias and should, therefore, be considered for secondary prevention ICD therapy.
> Identification of adult patients with HCM at high risk for SCD should be guided by the presence of a number of acknowledged noninvasive SCD risk factors. Because each of these major risk factors individually is associated with increased risk, **it would be reasonable to consider primary prevention ICD for patients with ≥ 1 SCD risk factor(s)** (Arnett, 2019; Elliott, 1999; Maron, 2019, 2007; O'Mahony, 2013; Vriesendorp, 2013; Yancy, 2013).

Regarding ICD therapy in less prevalent cardiac conditions, such as Brugada syndrome (BrS), the 2008 ACC/AHA/HRS guidelines for device-based therapy of cardiac rhythm abnormalities provide a Class IIa recommendation for, "ICD implantation as reasonable for patients with BrS who have had syncope" and for, "ICD implantation as reasonable for patients with BrS who have documented VT that has not resulted in cardiac arrest" (Level of Evidence: C). The following is provided as the basis for these recommendations:

> Primary electrical conditions, (in reference to genetic syndromes that predispose to sustained VT or VF, such as BrS),

> typically exist in the absence of any underlying structural heart disease and predispose to cardiac arrest. Although controversy still exists with regard to risk factors for SCD in these conditions, there is consensus that those with prior cardiac arrest or syncope are at very high risk for recurrent arrhythmic events. On the basis of the absence of any clear or consistent survival benefit of pharmacological therapy for individuals with these genetic arrhythmia syndromes, the ICD is the preferred therapy for those with prior episodes of sustained VT or VF and may also be considered for primary prevention for some with a very strong family history of early mortality… Individuals with syncope and the ECG pattern of spontaneous ST segment elevation (associated with BrS) have a 6-fold higher risk of cardiac arrest than those without syncope and the spontaneous ECG pattern (Epstein, 2008).

In 2017, an updated AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death (Al-Khatib) was published which was based on systematic reviews of the available evidence-based data and additional guidance from specialty societies (Birnie [HRS], 2014; Kron, 2013; Mohsen, 2014; Schuller, 2012; Shen [ACC/AHA/HRS], 2017). This document provided Class I and IIa recommendations for ICD therapy in those with cardiac sarcoidosis and specific clinical indications when there is reasonable expectation of meaningful survival of greater than 1 year. The recommendations with the strongest indications for benefit from ICD therapy have been incorporated into the medically necessary criteria within this document.

HRS collaborated with 12 international cardiology societies in 2019 to publish an expert consensus statement on the evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy (ACM) (Towbin, 2019). The document offers guidance for the diagnosis and treatment of ACM, including arrhythmogenic right ventricular cardiomyopathy (ARVC). Their recommendations include statements that an ICD is reasonable for individuals with mutations of phospholamban, FLNC, or lamin A/C ACM and an LVEF <45%. The authors note that the significance of a positive test for gene mutation is not certain because the penetrance and natural history of these disorders are not well known.

In 2022, the ACC/AHA/HFSA (Heart Failure Society of America) updated its guideline for the management of HF (Heidenreich, 2022). This update retains the 2009 recommendations for ICD therapy and includes a Class IIa Level of Evidence B-R recommendation stating that an ICD is reasonable for individuals with risk factors, LVEF less than or equal to 45%, and genetic arrhythmogenic cardiomyopathy. This is consistent with a IIa B-NR recommendation in the 2017 AHA/ACC/HRS ventricular arrhythmias guideline (Al-Khatib, 2017) stating that ICD implantation can be beneficial for individuals with risk factors, LVEF less than or equal to 45%, and NIDCM due to a Lamin A/C gene mutation. Heidenreich, et al. do not cite RCTs to support their recommendation and Al-Khatib, et al. specifically state that there are no studies that test the effects of ICD implantation on long-term survival for individuals with a Lamin A/C mutation.

Although certain gene mutations may be associated with higher risk for significant ventricular arrhythmias, the penetrance and natural history of these mutations has not been characterized. There are no published studies showing that ICD use improves health outcomes for individuals with these mutations. Additional studies are needed to inform reasonable conclusions about the effects of ICD use in ACM or ARVC, including whether the risk of device-related complications outweighs the benefit of ICD implantation.

On September 28, 2012 the U.S. Food and Drug Administration (FDA) granted clearance for the first subcutaneous ICD device, the S-ICD® System, (developed by Cameron Health®, Inc., now owned by Boston Scientific Corp., San Clemente, CA). According to the FDA press release:

> The S-ICD System is cleared to provide an electric shock to the heart (defibrillation) when the individual's heart is beating at a dangerous level or abnormally fast (ventricular tachyarrhythmias). It is approved only for individuals who do not require a pacemaker or pacing therapy.

This press release went on to say: "The S-ICD System provides an alternative for treating patients with life-threatening heart arrhythmias for whom the routine ICD placement procedure is not ideal," said Christy Foreman, director of the Office of Device Evaluation at the FDA Center for Devices and Radiological Health (CDRH). "Some patients with anatomy that makes it challenging to place one of the implantable defibrillators currently on the market may especially benefit from this device" (CDRH, 2012).

On March 13, 2015 the EMBLEM™ S-ICD System (Boston Scientific Corp., St. Paul, MN) obtained pre-market clearance from the FDA for indications very similar to the original S-ICD System for individuals who are at risk for SCD but who do not also require a pacemaker or pacing therapy. The FDA required that a post-approval study be conducted by the manufacturer. This prospective cohort study (of 1616 subjects from approximately 50 investigational centers [including up to 140 in the US]), the PRAETORIAN trial, is underway with final 5 year follow/up data planned and interim reports submitted to the FDA at 6 months, 1 year, 18 months, 2, and 3 years. Interim safety data as of Jan. 24, 2018 reported no high impedance alerts and no adverse events, to date, for the 140 enrolled subjects in the U.S (Gold, 2017). At follow-up of 49.1 months, Knops reported interim data that showed primary end-point events occurred in 68 subjects in the S-ICD group and in 68 subjects in the transvenous ICD (TV-ICD) group (48-month Kaplan-Meier estimated cumulative incidence, 15.1% and 15.7%, respectively; hazard ratio [HR], 0.99; 95% confidence interval [CI], 0.71 to 1.39; p=0.01 for noninferiority; p=0.95 for superiority). Device-related complications occurred in 31 subjects in the S-ICD group and in 44 in the TV-ICD group (HR, 0.69; 95% CI, 0.44 to 1.09); inappropriate shocks occurred in 41 and 29 subjects, respectively (HR, 1.43; 95% CI, 0.89 to 2.30). Death occurred in 83 subjects in the S-ICD group and in 68 in the TV-ICD group (HR, 1.23; 95% CI, 0.89 to 1.70); appropriate shocks occurred in 83 and 57 subjects, respectively (HR, 1.52; 95% CI, 1.08 to 2.12) (Knops, 2020). Additional information is available at: https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfpma/pma_pas.cfm?t_id=612348&c_id=4601. Accessed on January 26, 2023.

On August 9, 2016 the FDA cleared the Emblem MRI S-ICD System, which allows individuals with this subcutaneous device to safely undergo magnetic resonance (MR) imaging. The FDA has also allowed for MR conditional labeling for all previously implanted Emblem S-ICD Systems. According to the manufacturer, the company is also actively pursuing MRI compatibility for their currently approved ICD and CRT (cardiac resynchronization therapy) systems via the global ENABLE MRI study.

The criteria within this document for use of ICD therapy are consistent with generally accepted standards of medical practice and are clinically appropriate to treat the cardiac conditions described in the Clinical Indications section of this document.

ICD THERAPY IN CHILDREN

There remains limited data on the clinical utility of ICD in children for many conditions which would commonly be treated with ICD in adults. As such, the potential risks and evidence of benefit from ICD therapy in children is not as well described in the medical literature. Studies have shown higher inappropriate shock rates and lead failure (Berul, 2008) and reduced quality of life (Sears, 2011) in children who have received an ICD. Based on specialty society input and opinions from the practice community, it was determined that ICD therapy should be made available to children based on the currently accepted adult indications for ICD, subject to the expert evaluations and treatment discretion of the treating physician in collaboration with the child's parents or legal guardians (Alexander, 2004; Berul, 2008; Decker, 2009; Dimitrow, 2010; Epstein, 2008; Gersh, 2011; Monserrat, 2003; Silka, 1993). Prior to ICD implantation in a child, the informed consent discussion that the treating provider conducts with the family/guardian should include documentation of a thorough discussion of the probability of a life threatening event, based on the underlying condition, as well as the

potential benefits and harms of the ICD and the family's/guardian's understanding of the information provided.

Definitions

Abnormal blood pressure (BP) response during upright exercise testing: Failure of BP to rise by more than 25 mmHg (flat) or a fall in BP more than 15 mmHg (considered to be a hypotensive response) that occurs during upright exercise stress testing.

Arrhythmia (or dysrhythmia): Problems that affect the electrical system of the heart muscle, producing abnormal heart rhythms and may be classified as either atrial or ventricular, depending on which part of the heart they originate from.

Atrial Fibrillation: A condition in which the atrium (the heart's two upper chambers) produce uncoordinated electrical signals.

Brugada syndrome (BrS): An autosomal-dominant inherited arrhythmic disorder characterized by ST elevations with successive negative T waves in the right precordial leads without structural cardiac abnormalities. Individuals with BrS are at risk for sudden cardiac death (SCD) due to ventricular fibrillation (VF). Mutations in the SCN5A gene represent the most common genotype responsible for BrS but mutations in additional genes have also been associated with BrS and risk for SCD.

Cardiac Sarcoidosis: A multisystem granulomatous disease of unknown etiology. Myocardial involvement occurs in at least 25% of individuals with sarcoidosis who are at increased risk for sudden death from ventricular arrhythmias.

Cardiomyopathy (CM): A disease in which the heart muscle becomes inflamed affecting cardiac function. There are multiple types of CM, (with the three main types being dilated, hypertrophic, and restrictive [see below]):

- Dilated - This is the most common form, in which the heart cavity is enlarged and stretched (cardiac dilation). The heart is weak and doesn't pump normally, and most individuals develop congestive heart failure. Abnormal heart rhythms and disturbances in the heart's electrical conduction may also occur.
- Hypertrophic (HCM) - In this condition, the muscle mass of the left ventricle enlarges or "hypertrophies." In one form of the disease, the wall between the two pumping chambers becomes enlarged and obstructs the blood flow from the left ventricle. In the other form of the disease, non-obstructive hypertrophic cardiomyopathy, the enlarged muscle doesn't obstruct blood flow.
- Ischemic Dilated (IDCM) - Left ventricular systolic dysfunction (or disease of the heart muscle) associated with at least 75% narrowing of at least one of the three major coronary arteries (marked stenosis) or a documented history of MI.
- Nonischemic Dilated (NIDCM) - Left ventricular systolic dysfunction (or disease of the heart muscle) that is not associated with Coronary Artery Disease (CAD) or narrowing of the coronary arteries. There are a few different types of NIDCM but all involve thickening (abnormal enlargement) of the walls of the heart and progressive weakening of the pumping efficiency of the heart.
- Restrictive - This is the least common type in the United States. The myocardium (heart muscle) of the ventricles becomes excessively "rigid," making it more difficult for the ventricles to fill with blood between heartbeats. This type of cardiomyopathy is usually due to another disease process.

Congestive Heart Failure (CHF), also referred to as Heart Failure (HF): This is a condition in which the heart can't pump enough blood to the body's other organs. The "failing" heart keeps working but not as efficiently as it should. As blood flow out of the heart slows, blood returning to the heart through the veins backs up, causing congestion in the tissues.

Coronary Artery Disease (CAD): Heart problems caused by narrowed heart arteries. When arteries are narrowed, less blood and oxygen reaches the heart which can ultimately lead to a heart attack (myocardial infarction - MI).

Defibrillation: A process in which an electronic device (a defibrillator) gives the heart an electric shock, helping to re-establish normal contraction rhythms in a heart that is not properly beating. This may be done using an external device or by a device implanted in the body.

Ejection Fraction (EF) or Left Ventricular Ejection Fraction (LVEF): The percentage of blood ejected from the left ventricle with each heartbeat. Normal readings would be in the 58-70% range and lower values would indicate ventricular dysfunction.

Electrophysiology Studies (EPS): These studies evaluate the electrophysiological properties of the heart, such as automaticity, conduction, and whether the condition is refractory to management with medications. Additional capabilities of this testing include: ability to initiate and terminate tachycardia to map activation sequences and to evaluate individuals for various forms of therapy and to judge response to therapy.

Myocardial Infarction (MI): This is the medical term for "heart attack." A MI occurs when the blood supply to part of the heart muscle (the myocardium) is severely reduced or blocked (stenosed).

According to the 2012 European Society of Cardiology, American College of Cardiology Foundation, American Heart Association, and the World Heart Federation (ESC/ACCF/AHA/WHF) Expert Consensus Document: Third Universal Definition of Myocardial Infarction, the following definitions are provided for acute evolving MI and prior MI:

Acute MI is defined as – Detection of a rise and/or fall of cardiac biomarker values (preferably cardiac troponin [cTn]) with at least one value above the 99th percentile upper reference limit (URL) and with at least one of the following:

- Symptoms of ischemia;
- New, or presumed new, significant ST-segment-T wave (ST-T) changes or new left bundle branch block (LBBB);
- Development of pathological Q waves in the EKG;
- Imaging evidence of new loss of viable myocardium or new regional wall motion abnormality;
- Identification of an intracoronary thrombus by angiography or autopsy.

Prior MI is defined as – Any one of the following:

- Pathological Q waves with or without symptoms in the absence of non-ischemic causes;
- Imaging evidence of a region of loss of viable myocardium that is thinned and fails to contract in the absence of a non-ischemic cause;
- Pathological findings of a prior MI (Thygesen, 2012).

New York Heart Association (NYHA) Definitions:
The NYHA classification of heart failure is a 4-tier system that categorizes based on subjective impression of the degree of functional compromise. The four NYHA functional classes are as follows:

- Class I - individuals with cardiac disease but without resulting limitation of physical activity; ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain; symptoms only occur on severe exertion;
- Class II - individuals with cardiac disease resulting in slight limitation of physical activity; they are comfortable at rest; ordinary

physical activity, (e.g., moderate physical exertion, such as carrying shopping bags up several flights of stairs) results in fatigue, palpitation, dyspnea, or anginal pain;
- Class III - individuals with cardiac disease resulting in marked limitation of physical activity; they are comfortable at rest; less than ordinary activity causes fatigue, palpitation, dyspnea or anginal pain;
- Class IV - individuals with cardiac disease resulting in inability to carry on any physical activity without discomfort; symptoms of heart failure or the anginal syndrome may be present even at rest; if any physical activity is undertaken, discomfort is increased.

Non-sustained/Sustained Ventricular Tachycardia: Ventricular tachycardia is considered non-sustained (NSVT) when 3 or more consecutive ventricular beats occur at a rate of at least 120 beats/minute which lasts less than 30 seconds. If the rhythm lasts more than 30 seconds, it is known as a sustained ventricular tachycardia (even if it terminates on its own, [that is, without medical intervention] after 30 seconds).

QRS Complex: The portion of an electrocardiogram (EKG) reading, which represents the spread of the electrical impulse through the ventricles.

Structural Heart Disease: A general or umbrella term that encompasses the full scope of conditions caused by defects or abnormalities in the heart's valves, walls and/or muscle. Heart valve conditions are either congenital (present at birth) or can form later in life, due to aging, infection or a correlated underlying condition, and affect the cardiac structure and proper pumping function of the myocardium. The term, structural heart disease, has also been described as, “Non-coronary cardiovascular disease processes and related interventions” (DeMaria, 2014). This refers to heart disease for which some therapy, surgical or percutaneous, exists. Examples include aortic stenosis, hypertrophic cardiomyopathy, some arrhythmias and coronary artery disease where an abnormality of the cardiac structure interferes with normal function (Portions excerpted from UChicago Medicine Valvular & Structural Heart Disease 2019; available at: https://www.uchicagomedicine.org/conditions-services/heart-vascular/valve-disease. Accessed on January 17, 2023).

Sudden Cardiac Arrest (SCA): Refers to a sudden cessation of cardiac activity such that the victim becomes unresponsive with no normal breathing and no signs of circulation. If corrective measures are not taken rapidly, this condition progresses to SCD.

Sudden Cardiac Death (SCD also called sudden death): Death resulting from an abrupt loss of heart function (cardiac arrest).

Syncope: An episode where the individual experiences loss of consciousness lasting at least several seconds. If the person only experiences extreme dizziness but with no actual loss of consciousness, this is termed “Pre-Syncope.”

Ventricular Fibrillation (Vfib or VF): This is a condition in which the heart's electrical activity becomes disordered, resulting in the heart's lower (pumping) chambers contract in a rapid, unsynchronized fashion, (that is, the ventricles "flutter" rather than beat), and the heart pumps little or no blood.

Ventricular Tachyarrhythmias: This medical term refers to a rapid heartbeat that may be regular or irregular and arises from the ventricle or pumping chamber of the heart. Two common tachyarrhythmias are ventricular tachycardia and ventricular fibrillation.

Ventricular Tachycardia (Vtach or VT): This is a fast regular heart rate (usually of 100 or more beats per minute) that starts in the lower chambers (ventricles) and may result from serious heart disease that usually requires prompt treatment.

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Index

The use of specific product names is illustrative only. It is not intended to be a recommendation of one product over another, and is not intended to represent a complete listing of all products available.

History

Status	Date	Action
Reviewed	08/10/2023	Medical Policy & Technology Assessment Committee (MPTAC) review. Revised References and Websites sections. Updated Coding section with 10/01/2023 ICD-10-CM changes; added I21.B to end of range.
Revised	05/11/2023	MPTAC review. Revised formatting in MN section. Updated References section.
Reviewed	02/16/2023	MPTAC review. The Discussion and References sections were updated. Updated Coding section, removed CPT 00534 associated anesthesia.
	9/28/2022	Updated Coding section with 10/01/2022 ICD-10-CM changes; added I47.20-I47.29 replacing I47.2.
Reviewed	02/17/2022	MPTAC review. References were updated.

Revised	02/11/2021	MPTAC review. The MN criteria regarding major risk factors in HCM were revised consistent with updated ACC/AHA guideline recommendations (Ommen, 2020) to require 1 or more risk factors as follows: 1. Personal history of cardiac arrest or sustained ventricular arrhythmias; 2. Personal history of syncope suspected by clinical history to be arrhythmic within the previous 12 months; 3. Family history of HCM-related sudden death in one or more 1st or 2nd degree relatives who are less than or equal to 50 years of age or in two or more 3rd degree relatives who are less than or equal to 50 years of age; 4. LV apical aneurysm, independent of size; 5. LV systolic dysfunction (LVEF less than 50%); 6. Nonsustained VT episodes on ECG or continuous ambulatory electrocardiographic monitoring; 7. Left ventricular (LV) wall thickness greater than or equal to 30 mm in any LV segment; Removed “Abnormal BP response during exercise.” Language was added to the Clinical Indications for the S-ICD for clarification to say: “When the individual does not require cardiac pacing.” Minor additional edits were made to the Clinical Indications section for clarification. The Discussion and References sections were updated. Reformatted and updated Coding section.
Reviewed	02/20/2020	MPTAC review. References were updated.
	10/01/2019	Updated Coding section with 10/01/2019 ICD-10-PCS changes; added 0JH60FZ, 0JH63FZ. A definition for structural heart disease was added to the Definitions section and References section was updated.
New	03/21/2019	MPTAC review. Moved content of SURG.00033 Cardioverter Defibrillators to a new clinical utilization management guideline document with the same title. The Note about ICD therapy in children was moved from the Clinical Indications section to the Discussion/Background section. The Discussion and References sections were updated.
