



National Imaging Associates, Inc.	
Clinical guideline IMPLANTABLE CARDIOVERTER DEFIBRILLATOR (ICD)	Original Date: February, 2013 Page 1 of 19
CPT Codes: 33230, 33240, 33249	Last Review Date: March, 2017
Guideline Number: NIA_CG_321	Last Revised Date: July, 2018
Responsible Department: Clinical Operations	Implementation Date: January 2019

INTRODUCTION

(Al-Khatib 2017; Priori 2015; Ganz 2018; Russo 2013;
Epstein 2012; Yancy 2013; Ponikowski 2016; Shen 2017)

- The implantable cardioverter defibrillator (ICD) has become valuable in the management of patients with ventricular arrhythmias (VA) capable of causing syncope, cardiac arrest, and sudden cardiac death (SCD), in either one of two scenarios:
 - Secondary prevention of SCD due to previous ventricular arrhythmic events
 - Primary prevention in patients at high risk for SCD due to ventricular arrhythmia

- Patient eligibility for an ICD presumes all of the following:
 - Anticipated reasonable quality of life for ≥ 1 year post implantation, with consideration of age and comorbidities, although age alone is not a contraindication (Katsumoto 2017)
 - Patient's ability to live with a shock-delivering device that requires management
 - Absence of a *completely* reversible cause that led to VA for which an ICD is being considered (see Additional Information section on reversible causes) (Marine 2018a)
 - Completion of ≥ 3 months of guideline directed medical therapy (GDMT) for heart failure (HF) in most, but not all, cases of primary prevention, unless a supervening indication for non-elective pacemaker implantation or ICD generator replacement arises (to avoid second implantation procedure) (see Additional Information section for definition of GDMT)
 - ICD indications are present in the vast majority of scenarios in which cardiac resynchronization therapy (CRT) is appropriate
 - Sustained VT is defined as having duration > 30 seconds or requiring termination due to hemodynamic compromise in < 30 seconds
 - Elective replacement indicators support generator change if there is anticipated reasonable quality of life for ≥ 1 year (issues pertaining to reduction in VA risk associated with improved LVEF and/or absence of VA

remain unresolved) (Al-Khatib 2017; Russo 2013)

- Guidelines for the pediatric population are extrapolated from the adult population, due to a lack of relevant trials (Brugada 2013; Priori 2015)

INDICATIONS FOR ICD INSERTION

(Al-Khatib 2017; Priori 2015; Ganz 2018; Russo 2013; Epstein 2012; Yancy 2013; Ponikowski 2016; Shen 2017)

General, Secondary Prevention of VT/VT/SCA

(Al-Khatib 2017; Priori 2015; Ganz 2018)

- Patients with ventricular fibrillation (VF) or hemodynamically non-tolerated ventricular tachycardia (VT) after evaluation of etiology of event and exclusion of completely reversible causes (e.g. occurrence of myocardial infarction < 48 hours ago can often be considered a completely reversible cause; also see Ischemic Heart Disease section below) (O’Gara 2013).
- Spontaneous VT lasting > 30 seconds in patients with structural or ischemic heart disease (CAD) or with channelopathies, whether hemodynamically stable or unstable (Ganz 2018).
- Post resuscitation from sudden cardiac arrest due to coronary artery spasm (Montalescot 2013).

Ischemic Heart Disease (CAD)

(Al-Khatib 2017, Priori 2015, Ganz 2018, Russo 2013)

- Sustained VT (> 30 s or with hemodynamic non-tolerance)
- Syncope of undetermined origin, in ischemic heart disease or with prior myocardial infarction (MI), with either one of:
 - hemodynamically significant sustained monomorphic VT induced at electrophysiological study
 - LVEF \leq 35%
- LVEF \leq 35% due to ischemic heart disease or prior MI, NYHA class II or III, despite GDMT, and at least 40 days post-MI and at least 90 days post-revascularization
- LVEF \leq 30% due to ischemic heart disease or prior MI, NYHA class I despite GDMT, and at least 40 days post-MI and at least 90 days post-revascularization (Al-Khatib 2017; Ganz 2018; Russo 2013)
- Non-sustained VT due to prior MI, LVEF \leq 40%, and inducible sustained VT or inducible VF at electrophysiological study. Non-sustained ventricular tachycardia (NSVT) should have been \geq 4 full days post MI or post coronary revascularization (Russo 2013)

- Newly found LVEF < 50% with VF or polymorphic VT < 48 hours post MI, NSVT \geq 4 days later, and inducible VT or VF \geq 4 days post complete coronary revascularization (Russo 2013)
- Newly found LVEF \leq 35% with VF or polymorphic VT < 48 hours post MI, and not amenable to complete coronary revascularization (Russo 2013)
- VF or hemodynamically unstable VT < 48 hours following elective coronary revascularization, without evidence of acute coronary occlusion, provoking infarct, or any other clearly reversible cause (Russo 2013)

Nonischemic cardiomyopathy (NICM) (Al-Khatib 2017)

- Sustained VT (> 30 s or with hemodynamic non-tolerance)
- Syncope in NICM (nonischemic cardiomyopathy) that is presumed to be due to ventricular arrhythmia, given the weak correlation between VT or VF inducibility and mortality
- NICM with LVEF \leq 35% and NYHA functional Class I, II, or III, despite GDMT, and at least 90 days (or 3 months) after diagnosis of dilated cardiomyopathy (DCM).
- NICM due to a *Lamin A/C* mutation, who have \geq 2 risk factors from the following list:
 - NSVT
 - LVEF < 45%
 - Nonmissense mutation
 - male sex
- In borderline uncertain indications for ICD in NICM, late gadolinium enhancement on cardiovascular magnetic resonance (CMR) provides evidence of mid wall fibrosis, indicating a higher risk for cardiac arrest and SCD. (Al-Khatib 2017; Halliday 2017; Kuruvilla 2014)
- Peripartum cardiomyopathy with LVEF \leq 35% that persists > 3 months postpartum despite 3 months of GDMT. (Russo 2013)
- Familial dilated cardiomyopathy with LVEF > 35%, with family history associated with SCD **OR** with a LMNA mutation (Russo 2013; Hershberger 2018)

Advanced Heart Failure & Transplantation (Al-Khatib 2017; Priori 2015)

- **In NYHA class IV and/or using inotropes**, awaiting transplantation or an LVAD, either non-hospitalized or planning imminent discharge, without other qualifying ICD criteria met (Al-Khatib 2017; Priori 2015; Russo 2013)
- **In a patient with an LVAD**, sustained ventricular arrhythmias (Al-Khatib 2017)
- **Severe allograft vasculopathy**, with severe LV dysfunction, with expected survival \geq 1 year (Al-Khatib 2017)
- In NYHA ambulatory class IV, with appropriately indicated CRT implantation (see Additional Information section for definition of ambulatory NYHA class IV)

Myocardial Diseases

- **Giant cell myocarditis** with (Al-Khatib 2017; Priori 2015):
 - VF or hemodynamically unstable VT, even if early in the course, OR
 - Requires a pacemaker
- **Chronic Chagas cardiomyopathy** for one of the following (Priori 2015; Marin-Neto 2018):
 - Cardiac arrest or sustained VT
 - Ejection fraction < 40%
- **Cardiac Sarcoidosis** for one of the following (Al-Khatib 2017; Shen 2017; Priori 2015):
 - Sustained VT or sudden cardiac arrest, even if early in the course
 - LVEF \leq 35%
 - LVEF > 35% with inducible sustained ventricular arrhythmia
 - Syncope
 - Scar on CMR or positron emission tomography (PET)
 - Require a permanent pacemaker, even if transient (Blankstein 2018)
- **Neuromuscular Disorders** for one of the following (Al-Khatib 2017):
 - Primary and secondary prevention as for NICM (Priori 2016)
 - Emery-Dreifuss or limb-girdle type I-B muscular dystrophy with progressive cardiac muscle involvement
 - Type 1 myotonic dystrophy (Steinert Disease) with an indication for a permanent pacemaker
- **Hypertrophic cardiomyopathy (HCM)** with \geq 1 major risk factors for SCD (Al-Khatib 2017; Maron 2018; Shen 2017; Epstein 2012):
 - Prior sudden cardiac arrest (SCA) due to VT or VF
 - Sustained VT with syncope or hemodynamic compromise
 - Maximum LV wall thickness \geq 30 mm
 - SCD \geq 1 first degree relatives, presumably caused by HCM
 - \geq 1 episodes of unexplained syncope within the preceding 6 months
 - NSVT
 - Abnormal BP response to exercise in patients < 40 yr
 - BP rise < 20 mm Hg or fall of > 20 from exercise peak during ongoing exercise
 - Borderline evidence of the above risk factors plus one of the following:
 - End stage HCM with LVEF < 50%
 - Left ventricular apical aneurysm
 - age < 30 years old
 - Late gadolinium enhancement on CMR \geq 15%
 - Marked left ventricular outflow tract (LVOT) gradient (at least \geq 50 mm Hg peak at rest)
 - Syncope > 5 years ago
 - Only if the above method is not helpful, the ESC HCM Risk-SCD Calculator can be used, according to the limitations on the web page (O'Mahoney 2017):

Available at: <http://www.doc2do.com/hcm/webHCM.html>

- **Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C)** and ≥ 1 of the following risk factors for SCD:
(Al-Khatib 2017; Shen 2017; McKenna 2018; Calkins 2017; Epstein 2012)
 - Resuscitated cardiac arrest
 - Sustained VT
 - Right or left ventricular ejection fraction $\leq 35\%$
 - Syncope with documented or presumed/suspected ventricular arrhythmia
 - Electrocardiographic abnormalities such as any one of the following:
 - T wave inversion in V1 to V3 during sinus rhythm
 - Frequent premature ventricular contractions (PVCs) (e.g. $> 30/\text{hour}$)
 - Nonsustained VT
 - Positive electrophysiologic study (EPS) for sustained VT
 - High risk genotypes or multiple mutations
 - Borderline evidence of the above risk factors plus one of the following:
 - male sex
 - extensive/severe involvement/dilation of the right or left ventricle
 - continued vigorous exertion

Channelopathies

- **Congenital long QT syndrome** with one of the following (Al-Khatib 2017; Zimetbaum 2018; Priori 2015; Goldenberg 2008; Schwartz 2012; Epstein 2012; Schwartz 2012)
[Diagnosis based upon the Schwartz score, if in question (see Additional Information section) (Schwartz 2011; Schwartz 2018)]:
 - Cardiac arrest
 - Sustained VT or recurrent syncope despite optimal beta blocker (or with beta blocker intolerance/noncompliance)
 - QTc > 500 ms on a beta blocker (Al-Khatib 2017)
 - Jervell and Lange-Nielson syndrome
 - Strong family history of SCD
 - High risk genotype
- **Brugada syndrome** with one of the following:
(Al-Khatib 2017; Priori 2015; Katsumoto 2017; Epstein 2012)
 - Cardiac arrest
 - Sustained ventricular arrhythmia
 - Syncope with a spontaneous Brugada type I electrocardiogram (ECG)
- **Catecholaminergic polymorphic VT** with one of the following (Al-Khatib 2017; Buxton 2018; Epstein 2012; Russo 2013):
 - Cardiac arrest
 - Syncope or sustained VT while receiving optimal dosing of beta blockers
 - Inducible VT or VF

- **Early Repolarization or Short QT Syndrome** with one of the following (Al-Khatib 2017; Priori 2015):
 - Cardiac arrest
 - Sustained ventricular arrhythmia
- **Idiopathic Polymorphic VT/VF** with one of the following (Al-Khatib 2017):
 - Cardiac arrest due to polymorphic VT or VF
 - Idiopathic VF (Russo 2013; Priori 2015)
 - First degree relative with SCD (Russo 2013)

Miscellaneous

- **Unexplained syncope following appropriate thorough evaluation** with one of the following:
 - Advanced structural heart disease (Epstein 2012)
 - Hypertensive heart disease with LVH and LVEF \leq 35% (Russo 2013)

Adult & Pediatric Congenital (Structural) Heart Disease (ACHD)

Main references: (Khairy 2014; Hernandez-Madrid 2018)

Secondary references: (Al-Khatib 2017; Priori 2015; Brugada 2013; Shen 2017)

- Cardiac arrest due to VF or hemodynamically unstable VT after evaluation to define the cause of the event and exclusion of a completely reversible etiology.
- Spontaneous symptomatic sustained VT, after undergoing hemodynamic and EP evaluation (hemodynamics impact arrhythmia risk and require optimization). Catheter ablation or surgery may offer a reasonable alternative or adjunct to ICD.
- Systemic LVEF \leq 35%, biventricular physiology, and NYHA class II or III on GDMT.
- Tetralogy of Fallot with one of the following (Al-Khatib 2017; Shen 2017):
 - Spontaneous sustained VT
 - Inducible monomorphic or polymorphic sustained VT or VF
 - Multiple risks from the following list:
 - Left ventricular dysfunction
 - NSVT
 - QRS duration \geq 180 ms
 - Extensive right ventricular scarring
- Single or systemic right ventricular ejection fraction (RVEF) $<$ 35%, in the presence of an additional risk factor such as:
 - NSVT
 - Unexplained syncope
 - NYHA class II or III, despite GDMT (Al-Khatib 2017; Priori 2015)
 - QRS duration \geq 140 ms
 - Severe systemic AV valve regurgitation

- Syncope of unknown origin in the presence of either advanced ventricular dysfunction (EF < 35%) or marked hypertrophy or inducible sustained VT or VF (Al-Khatib 2017; Shen 2017)
- Syncope and moderate or complex congenital heart disease (CHD), with high clinical suspicion of ventricular arrhythmia despite thorough invasive and non-invasive evaluation not defining a cause
- Non-hospitalized patients with CHD awaiting heart transplantation
- Left ventricular noncompaction that meets same indications as NICM, including a familial history of SCD (Connolly 2018; Russo 2018).

ICD NOT Recommended in CHD

- Patients with less than 1 year of expected survival, even if they otherwise meet ICD implantation criteria.
- Incessant VT or VF.
- Significant psychiatric illnesses that may be aggravated by device implantation or that may preclude systematic follow-up.
- NYHA Class IV symptoms with drug-refractory congestive heart failure and who are not eligible for cardiac transplantation, ventricular assist device, or cardiac resynchronization therapy defibrillator (CRT-D).
- Advanced pulmonary vascular disease/Eisenmenger syndrome - generally not considered candidates for ICD therapy.

Exemptions

Indications for ICD with an Appropriate Pacing Modality in Special Situations (Marine 2018b; Katsumoto 2014; Russo 2013) *

- ICD criteria met, and elevated troponin is deemed to not be due to a myocardial infarction (although troponin elevation can also be secondary to VT or VF, requiring judgment) (Al-Khatib 2017).
- ICD criteria met, except for myocardial infarction within 40 days or revascularization within 3 months, but a non-elective permanent pacemaker (new or replacement) is required, and recovery of left ventricular function to LVEF > 35% is uncertain or not expected (Russo 2013).**
- ICD criteria met, except NICM or ischemic cardiomyopathy has not had 3 months' time for LVEF to improve on medical therapy, a non-elective permanent pacemaker is required, and recovery of LVEF is uncertain or not expected.**
- Pre-existing ICD (with or without pre-existing CRT) requiring non-elective generator replacement within <40 days post myocardial infarction or < 3 months post revascularization restrictions.**
- Patient met primary prevention criteria for an ICD prior to coronary revascularization, and it is unlikely that LVEF will recover to > 35% despite a 90 day wait (Katsumoto 2014).
- Listed for transplantation or received a LVAD within 3 months of revascularization, but *not* within 40 days of myocardial infarction (Katsumoto 2014).

* With these ICD indications, CRT would sometimes be the appropriate pacing modality. CRT is highly likely to be the appropriate modality when > 40% rhythm requires pacing.

** These indications enable avoidance of a second implantation procedure within less than 3 months.

ADDITIONAL INFORMATION

General

(Al-Khatib 2017; Priori 2015; Ganz 2018; Russo 2013; Epstein 2012; Yancy 2013; Ponikowski 2016; Shen 2017)

Implantable cardioverter defibrillators (ICDs) are indicated for the treatment of life-threatening ventricular tachycardia and ventricular fibrillation. An ICD system includes a pulse generator and one or more leads. ICDs are indicated both for patients who have survived life threatening rhythm disturbances (secondary prevention) and for those who are at risk for them (primary prevention).

- An ICD continually monitors heart rhythm. If a rapid rhythm is detected, the device delivers electrical therapy directly to the heart muscle in order to terminate the rapid rhythm and restore a normal heart rhythm. There are two types of therapy that can be delivered:
 - Rapid pacing, which is painless, is often effective in terminating ventricular tachycardia.
 - High-voltage shocks, which are painful to the patient, are necessary for ventricular fibrillation and also for instances where rapid pacing has failed to correct ventricular tachycardia.
- In addition, all ICDs have pacing capability, and they deliver pacing therapy for slow heart rhythms (bradycardia).
- The parameters defining limits for pacing therapy and for tachycardia therapy are programmable using noninvasive radio signals on all available ICDs.
- **Waiting Period** is an important issue in the timing of ICD insertion for primary prevention. This has resulted from guidelines and payment policies, predominantly on the part of CMS, which mirror the inclusion criteria of published primary and secondary prevention trials. For example, most primary prevention trials have excluded patients with recent coronary revascularization (under 3 months or 90 days) or recent myocardial infarction (under 40 days). Studies of patients who have received ICDs early after myocardial infarction have not demonstrated a mortality benefit.
 - Most guidelines recommend waiting periods that are reasonable and appropriate, but there are certain clinical scenarios in which exemptions might be required. For example, a patient with a longstanding cardiomyopathy, who is a candidate for an ICD, might have a small non-revascularized non-ST-elevation myocardial infarction (STEMI). This patient's LVEF will certainly not improve over the next 40 days, and withholding an ICD makes little sense.
 - This scenario would be rendered even more problematic if the patient required a non-elective pacemaker, since waiting 40 days post myocardial infarction to upgrade

a pacemaker to an ICD would subject the patient (and payer) to two procedures instead of one. Therefore, these guidelines adhere to the current waiting periods but also provide an opportunity to request exemptions where patient benefit is clearly documented (see “Exemptions” section above).

Reversible Causes of Ventricular Arrhythmia (Marine 2018a)

In some survivors of SCA or sustained VT, a transient or reversible cause (e.g. acute myocardial ischemia, electrolyte disturbances, medication-related proarrhythmia) can be identified which is thought to have been the cause. Initial treatment should be directed at the underlying disorder. However, prior to concluding that the VA was entirely due to a reversible cause, a thorough evaluation should be performed, for which electrophysiologic consultation might be required. As opposed to completely reversible causes, a reversible condition might be only a precipitant of ventricular arrhythmia in a patient who is otherwise predisposed and therefore considered high risk for recurrence, especially in the context of possible recurrence of precipitating factors. A prime example is a patient who presents with VF and is found to have mild hypokalemia, in which case it is generally not appropriate to assign the entirety of the cause of the ventricular arrhythmia to the low potassium level alone.

Correction of a reversible cause of SCA or sustained VT is most likely to be adequate in one of several settings:

- **Polymorphic VT or VF** that is preceded by clear evidence of myocardial ischemia or acute myocardial infarction within the past 48 hours - In such cases, revascularization is often adequate for the purpose of reducing the risk of SCD. However, some of these patients will later qualify for a primary prevention ICD due to severe left ventricular systolic dysfunction or HF. Guideline-directed medical therapy should be applied, and follow-up evaluation with a cardiologist soon after discharge should be arranged for additional risk stratification. A repeat evaluation of LV function is recommended >40 days post-MI and >90 days after revascularization to determine if the patient qualifies for ICD implantation based on consideration for primary prevention indications.

An important caveat is that **sustained monomorphic VT** in the setting of prior myocardial infarction is typically due to scar-related (substrate) re-entry and is not due to the occurrence of ischemia. Thus, in patients with stable CAD and sustained monomorphic VT, coronary *revascularization alone is considered an ineffective therapy* to prevent such recurrent VT.

- **Polymorphic VT in the setting of acquired QT prolongation** - Withdrawal of the offending drug and avoidance of other QT prolonging medications may be adequate to reduce the risk of SCD.

- **VF in the setting of Wolff-Parkinson-White syndrome in patients with a structurally normal heart** – These patients are adequately treated with catheter ablation of the accessory pathway.

- **Idiopathic monomorphic VT in the setting of a structurally normal heart** – Such patients are usually adequately treated with medical therapy or catheter ablation (see VA in Structurally Normal Hearts, below).

- **VT/VF occurring in the setting of drug overdose** – Examples include cocaine, amphetamines, digoxin, tricyclic antidepressants, and antiarrhythmic drugs.

In **most other cases**, life-threatening ventricular arrhythmias should **not** be attributed solely to a reversible disorder, and patients should be managed according to guidelines for secondary prevention.

Ventricular Arrhythmias in Structurally Normal Hearts

Sustained VT is uncommon in patients with structurally normal hearts. In patients with structurally normal hearts, an ICD is generally *not* recommended in patients with monomorphic VT. When there are substantial symptoms (or adverse effect on LVEF or sustained VT) these patients can be treated with medical therapy or, more often, catheter ablation. An ICD would not be required since the risk of sudden cardiac death is typically low, especially following successful catheter ablation (e.g. atrial arrhythmias associated with the Wolff-Parkinson-White syndrome, RV or LV outflow tract VT, idiopathic VT, or fascicular VT in the absence of structural heart disease) (Ganz 2018; Al-Khatib 2017; Dukkipati 2017a; Callans 2018).

Reduced LVEF deemed solely due to a high burden of ventricular ectopy (PVCs, VT), comprising > 10% of QRS complexes, is considered a reversible cause for the reduced LVEF. When ablation can eradicate the ventricular ectopy and restore LVEF > 35%, an ICD would not be indicated; it should be considered if the LVEF remains ≤ 35% (Dukkipati a&b 2017).

Some polymorphic VT or VF in *structurally normal* hearts can be treated with ablation of the triggering PVC, e.g. catecholaminergic polymorphic VT/VF, idiopathic VF, Congenital Long QT Syndrome, Brugada Syndrome, and Early Repolarization Syndrome. In Brugada patients, the substrate of the VF can often be ablated by an epicardial approach. Generally, *an ICD is still required* in this group of patients with polymorphic VT or VF (given the complexity, any decision to avoid an ICD implant following such potential VA cures by ablation in structurally normal hearts is beyond the scope of this guideline) (Dukkipati a&b 2017).

Wearable Cardioverter Defibrillator (WCD)

The WCD will require additional clinical trials to determine its optimal place in the treatment of ventricular arrhythmia, since to date, only one randomized clinical trial, VEST, had been presented orally at the American College of Cardiology meeting in March, 2018. The WCD is addressed in a separate document.

Unanswered Questions

Additional issues need to be addressed with respect to vulnerability to SCD. While ventricular arrhythmia within the first 48 hours post myocardial infarction might not qualify for longer term risk that warrants an ICD, the presence of sustained monomorphic

VT (as opposed to polymorphic VT or VF) in that time frame bespeaks underlying substrate for chronic risk of recurrence of the sustained monomorphic VT, in which case either an ICD versus WCD plus watchful waiting and/or EPS testing/ablation might prove helpful. Further study is necessary (Liang 2014).

NYHA Class Definitions (Russo 2013; Colucci 2018)

- Class I: No limitation of functional activity or only at levels of exertion that would limit normal individuals (patient can carry 24 pounds up 8 stairs, play basketball, and shovel soil).
- Class II: Slight limitation of activity. Fatigue, palpitation, or dyspnea with moderate exercise (patient able to dance, garden, walk 4 MPH on level ground, and have sexual intercourse).
- Class III: Marked limitation of activity. Fatigue, palpitation, or dyspnea with minimal activity (patient able to shower, make bed, bowl or golf, dress, and walk 2.5 MPH on level ground).
- Class IV: Severe limitation of activity. Symptoms even at rest, worse with activity (patient unable to comfortably perform any significant activity).
- Ambulatory Class IV: Class IV heart failure with: 1) no active acute coronary syndrome; 2) no inotropes; and 3) on GDMT.

Heart Block Definitions (Epstein 2012)

- First Degree: All atrial beats are conducted to the ventricles, but with a delay of > 200ms.
- Second Degree: Intermittent failure of conduction of single beats from atrium to ventricles.
 - Type I: Conducted beats have variable conduction times from atrium to ventricles.
 - Type II: Conducted beats have uniform conduction times from atrium to ventricles.
 - Advanced: Two or more consecutive non-conducted beats (premature atrial beats might not normally be conducted).
- Third Degree: No atrial beats are conducted from atrium to ventricle

Guideline Directed (or Optimal) Medical Therapy for Heart Failure (Yancy 2013; Yancy 2017)

- Angiotensin converting enzyme (ACE-I), angiotensin receptor blockers (ARB), or combined angiotensin receptor inhibitor and neprilysin inhibitor (ARNI)
- Beta blocker (might be less critical in permanent atrial fibrillation, still recommended) (Kotecha 2017).

- Addition of loop diuretic for all NYHA class II – IV patients
- Addition of hydralazine and nitrate for persistently symptomatic African Americans
- Addition of an aldosterone antagonist, provided eGFR is > 30 ml/mi
- n and K+ < 5.0
- Not required for consideration of ICD: Ivabradine for NYHA class II – III, when a beta blocker has failed to reduce a sinus rate to < 70 bpm. Ivabradine listed as a class IIa recommendation, while others are class I recommendations. CRT trials antedated routine use of ivabradine.

Schwartz score diagnostic criteria for long QT syndrome (LQTS) (Schwartz 2011)

Criteria	Points
Electrocardiographic findings*	
QTc [¶]	
▪ ≥480 ms	3
▪ 460 to 479 ms	2
▪ 450 to 459 ms (in males)	1
QTc [¶] fourth minute of recovery from exercise stress test ≥480 ms	1
Torsades de pointes ^Δ	2
T wave alternans	1
Notched T wave in 3 leads	1
Low heart rate for age [◊]	0.5
Clinical history	
Syncope ^Δ	
▪ With stress	2
▪ Without stress	1
Congenital deafness	0.5

Family history	
Family members with definite LQTS§	1
Unexplained sudden cardiac death below age 30 among immediate family members§	0.5

SCORE:

- ≤1 point = low probability of long QT syndrome (LQTS).
- 1.5 to 3 points = intermediate probability of LQTS, requires addition of genotyping to further classify risk as low or high.
- ≥3.5 points = high probability of LQTS.

* In the absence of medications or disorders known to affect these electrocardiographic features.

¶ QTc calculated by Bazett's formula where $QTc = QT/\sqrt{RR}$.

Δ Mutually exclusive.

◇ Resting heart rate below the second percentile for age.

§ The same family member cannot be counted in A and B.

Abbreviations

ACE-I	Angiotensin converting enzyme inhibitor
ACHD	Adult congenital heart disease
ARNI	Combined angiotensin receptor inhibitor and neprilysin inhibitor
ARVD/C	Arrhythmogenic right ventricular dysplasia/cardiomyopathy
AV	Atrioventricular
CAD	Coronary artery disease, same as ischemic heart disease
CHD	Congenital heart disease
CHF	Congestive heart failure
CRT	Cardiac resynchronization therapy
CRT-D	Cardiac resynchronization therapy ICD system
DCM	Dilated cardiomyopathy
ECG	Electrocardiogram
EPS	Electrophysiologic Study
GDMT	Guideline-Directed Medical Therapy
HCM	Hypertrophic cardiomyopathy
HF	Heart failure
HRS	Heart Rhythm Society
HV	His-ventricle
ICD	Implantable cardioverter-defibrillator
LBBB	Left bundle-branch block
LV	Left ventricular/left ventricle
LVAD	Left ventricular assist device, mechanical heart
LVEF	Left ventricular ejection fraction
MI	Myocardial infarction

ms	Milliseconds
NICM	Nonischemic cardiomyopathy
NYHA	New York Heart Association
RV	Right ventricular/right ventricle
STEMI	ST-elevation myocardial infarction
SND	Sinus node dysfunction
VT	Ventricular tachycardia
VF	Ventricular fibrillation

REFERENCES

Al-Khatib SM, Stevenson WG, Ackerman MUJ, et al. 2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death, Accepted Manuscript in Press. *JACC*, Available at http://www.onlinejacc.org/content/accj/early/2017/10/19/j.jacc.2017.10.054.full.pdf?_ga=2.14551095.1842511958.1523473576-1847200754.1521829021 Retrieved April 11, 2018

Blankstein R, Cooper LT. Management and prognosis of cardiac sarcoidosis. UpToDate. Waltham, MA; June, 2018. Available at: https://www.uptodate.com/contents/management-and-prognosis-of-cardiac-sarcoidosis?search=heart%20block%20in%20sarcoidosis§ionRank=1&usage_type=default&anchor=H2076470966&source=machineLearning&selectedTitle=1~150&display_rank=1#H2076470966 Retrieved July 3, 2018

Buxton A. Catecholaminergic polymorphic ventricular tachycardia and other polymorphic ventricular tachycardias with a normal QT interval. UpToDate. Waltham, MA; May, 2018. Available at: https://www.uptodate.com/contents/catecholaminergic-polymorphic-ventricular-tachycardia-and-other-polymorphic-ventricular-tachycardias-with-a-normal-qt-interval?search=catecholaminergic%20polymorphic%20ventricular%20tachycardia&source=search_result&selectedTitle=1~39&usage_type=default&display_rank=1#H6471335 Retrieved Jun 8, 2018

Brugada J, Blom N, Sarquella-Brugada, et al. Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group Joint Consensus Statement. *Europace*. 2013;15:1337-1382.

Calkins H, Corrado D, Marcus F. Risk Stratification in Arrhythmogenic Right Ventricular Cardiomyopathy. 2017. *Circulation*. 2017;136(21):2068-2082.

Callans DJ. Monomorphic ventricular tachycardia in the absence of apparent structural heart disease. UpToDate; June, 2018. Available at: https://www.uptodate.com/contents/monomorphic-ventricular-tachycardia-in-the-absence-of-apparent-structural-heart-disease?topicRef=921&source=see_link#H21 Retrieved July 25, 2018

Colucci WS. Predictors of survival in heart failure with reduced ejection fraction. UpToDate. Waltham, MA; June, 2018. Available at: https://www.uptodate.com/contents/predictors-of-survival-in-heart-failure-with-reduced-ejection-fraction?search=nyha%20classes§ionRank=1&usage_type=default&anchor=H3&source=machineLearning&selectedTitle=1~150&display_rank=1#H3 Retrieved July 2, 2018, 2018

Connolly HM, Attenhofer-Jost C. Isolated left ventricular noncompaction, UpToDate. Waltham, MA; June, 2018. Available at: <https://www.uptodate.com/contents/isolated-left-ventricular>

[noncompaction?search=left%20ventricular%20noncompaction§ionRank=1&usage_type=default&anchor=H19&source=machineLearning&selectedTitle=1~23&display_rank=1#H1860083297](https://www.uptodate.com/contents/noncompaction?search=left%20ventricular%20noncompaction§ionRank=1&usage_type=default&anchor=H19&source=machineLearning&selectedTitle=1~23&display_rank=1#H1860083297) Retrieved July 10, 2018

Dukkipati, SR, Choudry S, Koruth JS, et al. Catheter ablation of ventricular tachycardia in structurally normal hearts; Indications, strategies, and outcomes – Part 1. *JACC*. 2017a;70:2909–23.

Dukkipati SR, Koruth JS, Choudry S, et al. Catheter ablation of ventricular tachycardia in structural heart disease; Indications, strategies, and outcomes – Part 2. *JACC*. 2017b;70:2924-41.

Epstein AE, DiMarco JP, Ellenbogen KA, et al. 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. *Circulation*. 2013; 127:e283-e352. Available at: <http://circ.ahajournals.org/content/127/3/e283>

Ganz LI. Implantable cardioverter-defibrillators: Overview of indications, components, and functions. UpToDate. Waltham MA; June, 2018. Available at: https://www.uptodate.com/contents/implantable-cardioverter-defibrillators-overview-of-indications-components-and-functions?search=ICD%20indications%20for%20secondary%20prevention§ionRank=1&usage_type=default&anchor=H2&source=machineLearning&selectedTitle=1~150&display_rank=1#H2 Retrieved June 6, 2018

Goldenberg I, Moss AJ. Long QT Syndrome. *JACC*. 2008;51(24):2291-2300.

Halliday BP, Gulati A, Ali A, et al. Association between mid-wall late gadolinium enhancement and sudden cardiac death in patients with dilated cardiomyopathy and mild and moderate left ventricular systolic dysfunction. *Circulation*. 2017;135(22):2106-2115. Available at: <http://circ.ahajournals.org/content/135/22/2106> Retrieved June 6, 2018

Hernandez-Madrid A, Paul T, Abrams D, et al. Arrhythmias in congenital heart disease: A position paper of the European Heart Rhythm Association (EHRA), Association for European Paediatric and Congenital Cardiology (AEPC), and the European Society of Cardiology (ESC) Working Group on Grown-up Congenital Heart Disease, endorsed by HRS, PACES, APHRS, and SOLAECE. *Europace*. 2018;0:1-35. Available at: <https://academic.oup.com/europace/advance-article-abstract/doi/10.1093/europace/eux380/4944677>

Hershberger RE. Familial dilated cardiomyopathy: Prevalence diagnosis and treatment. UpToDate. Waltham, MA; June, 2008. Available at: https://www.uptodate.com/contents/familial-dilated-cardiomyopathy-prevalence-diagnosis-and-treatment?search=familial%20dilated%20cardiomyopathy&source=search_result&selectedTitle=1~19&usage_type=default&display_rank=1 Retrieved July 10, 2018

Katsumoto FM, Calkins H, Boehmer J, et al. HRS/ACC/AHA Expert Consensus Statement on the Use of Implantable Cardioverter-Defibrillator Therapy in Patients Who Are Not Included or Not Well represented in Clinical Trials. *Heart Rhythm*. 2014;11(7):1270-1303.

Katsumoto FM, Bailey Kr, Chaouki AS, et al. Systematic review for the 2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death. *JACC*. 2017. (in press) Available at: http://www.onlinejacc.org/content/early/2017/10/19/j.jacc.2017.10.052?_ga=2.219363033.1711384757.1528142205-1917600455.1527897917 Retrieved June 6, 2018

Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS Expert consensus statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease. *Heart Rhythm*. 2014;11: e102-e165.

Kuruville S, Adenaw N, Katwal AB, et al. Late gadolinium enhancement on cardiac magnetic resonance predicts adverse cardiovascular outcomes in nonischemic cardiomyopathy: a systematic review and meta-analysis. *Circ Cardiovasc Imaging*. 2014; 7:250-8.

Liang JJ, Hodge DO, Mehta RA, et al. Outcome in patients with sustained ventricular tachyarrhythmias occurring within 48 h of acute myocardial infarction: when is ICD appropriate? *Europace*. 2014;16:759-1766.

Marine JE, Russo AM. Secondary prevention of sudden cardiac death in heart failure and cardiomyopathy. UpToDate. Waltham, MA; May, 2018a. Available at: https://www.uptodate.com/contents/secondary-prevention-of-sudden-cardiac-death-in-heart-failure-and-cardiomyopathy?search=reversible%20causes%20of%20ventricular%20arrhythmia§ionRank=1&usage_type=default&anchor=H1075672&source=machineLearning&selectedTitle=2~150&display_rank=2#H1075672 Retrieved June 12, 2018

Marine JE, Russo AM. Primary prevention of sudden cardiac death in heart failure and cardiomyopathy. UpToDate. Waltham, MA; May, 2018b. Available at: https://www.uptodate.com/contents/primary-prevention-of-sudden-cardiac-death-in-heart-failure-and-cardiomyopathy?search=ICD%20indications§ionRank=1&usage_type=default&anchor=H957895585&source=machineLearning&selectedTitle=2~150&display_rank=2#H957895585 Retrieved June 11, 2018

Marin-Neto JA, Simoes MV, Schmidt A, et al. Chagas heart disease: Treatment and prognosis. UpToDate. Waltham, MA; May, 2018. Available at: https://www.uptodate.com/contents/chagas-heart-disease-treatment-and-prognosis?search=icd%20in%20chagas%20heart%20disease§ionRank=2&usage_type=default&anchor=H372251625&source=machineLearning&selectedTitle=1~150&display_rank=1# Retrieved June 7, 2018

Maron MS. Hypertrophic cardiomyopathy: Assessment and management of ventricular arrhythmias and sudden cardiac death risk. UpToDate. Waltham, MA; May, 2018. Available at: [https://www.uptodate.com/contents/hypertrophic-cardiomyopathy-assessment-and-](https://www.uptodate.com/contents/hypertrophic-cardiomyopathy-assessment-and)

[management-of-ventricular-arrhythmias-and-sudden-cardiac-death-risk?search=icd%20in%20hcm§ionRank=1&usage_type=default&anchor=H99765267&source=machineLearning&selectedTitle=1~150&display_rank=1#H99765267](https://www.uptodate.com/contents/management-of-ventricular-arrhythmias-and-sudden-cardiac-death-risk?search=icd%20in%20hcm§ionRank=1&usage_type=default&anchor=H99765267&source=machineLearning&selectedTitle=1~150&display_rank=1#H99765267) Retrieved June 7, 2018

McKenna WJ. Arrhythmogenic right ventricular cardiomyopathy: Treatment and prognosis. UpToDate. Waltham, MA; May, 2018. Available at: https://www.uptodate.com/contents/arrhythmogenic-right-ventricular-cardiomyopathy-treatment-and-prognosis?search=icd%20in%20arvd§ionRank=1&usage_type=default&anchor=H1717857965&source=machineLearning&selectedTitle=1~150&display_rank=1#H1717857965 Retrieved June 7, 2018

Montalescot G, Sechtem U, Achenbach S, et al. 2013 ESC guidelines on the management of stable coronary artery disease: The Task Force on the management of stable coronary artery disease of the European Society of Cardiology. *European Heart Journal*. 2013;34(38): 2949–3003 . Available at: <https://academic.oup.com/eurheartj/article/34/38/2949/442952>

O’Gara PT, Kushner FG, Ascheim DD, et al. ACCF/AHA guideline for the management of ST-elevation myocardial infarction: A report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *JACC*. 2013; 61:e78 –140.

O’Mahony C, Jichi F, Ommen SR, et al. An international external validation study of the 2014 European Society of Cardiology guideline on sudden cardiac death prevention in hypertrophic cardiomyopathy (evidence from HCM). *Circulation*. 2018;137(10):1015-1023.

Ponikowski P, Voors AA, Anker SD, et al. 2016 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure, The Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC), Developed with the special contribution of the Heart Failure Association (HFA) of the ESC, *European Heart Journal*. 2016;37:2129–2200.

Priori SG, Blomstrom-Lundqvist C, Mazzanti A, et al. 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death, The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC). *European Heart Journal*. 2015;36:2793–2867.

Russo, AM, Stainback RF, Bailey SR, et al. ACCF/HRS/AHA/ASE/HFSA/SCAI/SCCT/SCMR 2013 appropriate use criteria for implantable cardioverter-defibrillators and cardiac resynchronization therapy: A report of the American College of Cardiology Foundation Appropriate Use Criteria Task Force, Heart Rhythm Society, American Heart Association, American Society of Echocardiography, Heart Failure Society of America, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, and Society for Cardiovascular Magnetic Resonance (endorsed by the American Geriatrics Society). *JACC*. 2013;61(12):1318–1368.

Schwartz PJ, Crotti L, Insolia R. Long QT syndrome, from genetics to management, *Circ*

Arrhythm Electrophysiol. 2012;5:868-877.

Schwartz PJ, Crotti L. QTc behavior during exercise and genetic testing for the long-QT syndrome. *Circulation.* 2011; 124:2181-2184. Available at: <http://circ.ahajournals.org/content/124/20/2181>

Schwartz PJ, Ackerman MJ. Congenital long QT syndrome: Diagnosis. UpToDate, Waltham, MA; May, 2018. Available at: https://www.uptodate.com/contents/congenital-long-qt-syndrome-diagnosis?topicRef=988&source=related_link#H3346885055 Retrieved June 8, 2018

Yancy CW, Jessup M, Bozkurt B, et al. 2013 ACCF/AHA guideline for the management of heart failure: A report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *Circulation.* 2013;128:e240–e327.

Yancy CW, Jessup M, Bozkurt B, et al. 2017 ACC/AHA/HFSA focused update of the 2013 ACCF/AHA guideline for the management of heart failure: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. *JACC.* 2017;70:776–803.

Shen W-K, Sheldon RS, Benditt DG, et al. 2017 ACC/AHA/HRS guideline for the evaluation and management of patients with syncope: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *JACC.* 2017;70:e39–110.

Zimetbaum PJ, Berul CI. Prognosis and management of congenital long QT syndrome. UpToDate. Waltham, MA; May, 2018. Available at: https://www.uptodate.com/contents/prognosis-and-management-of-congenital-long-qt-syndrome?search=long%20and%20short%20qt%20syndromes&source=search_result&selectedTitle=8~150&usage_type=default&display_rank=8# Retrieved June 8, 2018

Reviewed / Approved by  Caroline Carney, MD, Chief Medical Officer