

National Imaging Associates, Inc.*	
Clinical guideline IMPLANTABLE CARDIOVERTER DEFIBRILLATOR (ICD)	Original Date: February 2013
CPT Codes: 33230, 33240, 33249	Last Revised Date: March 2021
Guideline Number: NIA_CG_321	Implementation Date: January 2022

GENERAL INFORMATION

It is an expectation that all patients receive care/services from a licensed clinician. All appropriate supporting documentation, including recent pertinent office visit notes, laboratory data, and results of any special testing must be provided. All prior relevant imaging results and the reason that alternative imaging cannot be performed must be included in the documentation submitted.

All indications are predicated on a meaningful life expectancy of greater than one year if the ICD is implanted.

INDICATIONS FOR ICD INSERTION

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

ISCHEMIC HEART DISEASE (CAD)

(Al-Khatib, 2017; Priori, 2015; Russo, 2013)

Primary Prevention of SCD (prophylactic ICD implantation)

- LVEF \leq 35% due to ischemic heart disease and NYHA class II or III, despite guideline-directed medical therapy (GDMT), and at least 40 days post-myocardial infarction (MI) and 90 days post-revascularization
- LVEF \leq 30% due to ischemic heart disease, NYHA class I, GDMT, and at least 40 days post-MI and 90 days post-revascularization
- LVEF \leq 40% with prior MI, NSVT, and inducible sustained ventricular tachycardia (VT) or ventricular fibrillation (VF) at electrophysiological testing

Secondary Prevention of SCD

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- Patients with documented ventricular fibrillation (VF), hemodynamically unstable ventricular tachycardia (VT), or sustained VT, after exclusion of reversible causes
- Syncope of undetermined origin, with Inducible VF or sustained VT at electrophysiological study (EPS)
- Syncope of undetermined origin, with EF \leq 35%

NONISCHEMIC CARDIOMYOPATHY (NICM)

(Al-Khatib, 2017)

Primary Prevention of SCD (prophylactic ICD implantation)

- Lamin A/C gene mutation, with \geq 2 risk factors from the following: NSVT, LVEF $<$ 45%, male sex, nonmissense mutation
- LVEF \leq 35% and NYHA functional Class II or III, despite at least 3 months of GDMT: Recommended
- LVEF \leq 35% and NYHA functional Class I despite at least 3 months of GDMT: May be considered

Secondary Prevention of SCD

- Patients with documented VF, hemodynamically unstable VT, or sustained VT, after exclusion of reversible causes
- LVEF \leq 50% with unexplained syncope presumed to be due to VA and who do not meet indications for primary prevention ICD implantation

ADVANCED HEART FAILURE & TRANSPLANTATION

(Al-Khatib, 2017; Priori, 2015)

- In non-hospitalized patients with NYHA class IV who are candidates for cardiac transplantation or left ventricular assist device (LVAD) (Al-Khatib, 2017; Priori, 2015; Russo, 2013)
- In a patient with an LVAD, sustained ventricular arrhythmias (Al-Khatib, 2017)
- In NYHA ambulatory class IV, with appropriate indications for CRT (see Background Information section for definition of ambulatory NYHA class IV)

MYOCARDIAL DISEASES

Hypertrophic cardiomyopathy

- Previous documented cardiac arrest or sustained ventricular tachycardia
- Adult patients with HCM with at least 1 risk factor for SCD as follows:
 - Sudden death likely attributable to HCM in at least 1 first-degree relative who is \leq 50 years of age
 - LVH \geq 30 mm

- At least 1 recent episode of syncope suspected by history to be arrhythmic (unlikely neurocardiogenic (vasovagal) and especially occurring within 6 months of evaluation (events beyond 5 years do not appear to have relevance))
- LV apical aneurysm
- LV systolic dysfunction (EF < 50%)
- Pediatric patients with HCM with at least 1 risk factor for SCD as follows:
 - Including unexplained syncope
 - LVH ≥ 30 mm
 - Nonsustained ventricular tachycardia
 - Family history of HCM-related SCD (reasonable after consideration of the high complication rate of long-term ICD placement in younger patients)

NOTE: ICD placement for the sole purpose of participation in competitive athletics should not be performed

- **Cardiac Sarcoidosis** with one of the following (Al-Khatib, 2017; Priori, 2015; Shen, 2017):
 - Cardiac arrest or documented sustained VT
 - LVEF ≤ 35%
 - LVEF > 35% with inducible sustained ventricular arrhythmia at EPS
 - Syncope and/or scar on CMR or positron emission tomography (PET)
 - Requires a permanent pacemaker
- **Neuromuscular Disorders (including but not limited to Duchenne, Becker, Limb-girdle type 1B, Limb-girdle type 2C-2F, Limb-girdle type 2I, Myotonic type 1, Myotonic type 2, Emery-Dreifuss, Facioscapulohumeral)** with one of the following (Al-Khatib, 2017):
 - Primary and secondary prevention, with same indications as for NICM (Priori, 2015)
 - Emery-Dreifuss or limb-girdle type I-B muscular dystrophy with progressive cardiac involvement
- **Arrhythmogenic right ventricular cardiomyopathy** and **at least 1** of the following risk factors for SCD: (Al-Khatib, 2017; Calkins, 2017; Corrado, 2015; Epstein, 2012; Shen, 2017)
 - Resuscitated sudden cardiac arrest
 - Sustained VT
 - Right or left ventricular systolic dysfunction with an ejection fraction ≤ 35%
 - Syncope with documented or presumed ventricular arrhythmia

CHANNELOPATHIES

- **Congenital long QT syndrome** with **one** of the following (Al-Khatib, 2017; Epstein, 2012;

Goldenberg, 2008; Priori, 2015; Schwartz, 2012)

- Sudden cardiac arrest
 - Sustained VT or recurrent syncope when beta blocker is ineffective or not tolerated
 - QTc > 500 ms on a beta blocker (Al-Khatib, 2017)
 - Strong family history of SCD
 - High risk genotype
- **Brugada syndrome and spontaneous type 1 Brugada electrocardiographic pattern with one of the following:**
(Al-Khatib, 2017; Epstein, 2012; Katsumoto, 2018; Priori, 2015)
 - Cardiac arrest
 - Documented sustained ventricular arrhythmia
 - Syncope presumed to be due to ventricular arrhythmia
- **Catecholaminergic polymorphic VT with one of the following** (Al-Khatib, 2017; Priori, 2013, Epstein, 2012; Russo, 2013):
 - Sudden cardiac arrest
 - Syncope or sustained VT
 - Inducible VT or VF
- **Early Repolarization (“J-wave Syndrome”) 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

ADULT & PEDIATRIC CONGENITAL HEART DISEASE (CHD)

(Al-Khatib, 2017; Brugada, 2013; Hernandez-Madrid, 2018; Khairy, 2014; Priori, 2015; Shen, 2017)

- Cardiac arrest due to VF or VT, or unstable VT, after exclusion of a reversible etiology
- Systemic LVEF ≤ 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 VF or sustained VT
 - ≥ 1 risk from the following list:
 - Prior palliative systemic to pulmonary shunts
 - Unexplained syncope
 - Frequent PVCs
 - Atrial tachycardia

- Left ventricular dysfunction or diastolic dysfunction
 - NSVT
 - QRS duration ≥ 180 ms
 - Dilated right ventricle
 - Residual pulmonary regurgitation or stenosis
 - RV Hypertension
- 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 ≥ 140 ms
 - Severe systemic AV valve regurgitation
- Syncope of unknown origin in the presence of either at least moderate ventricular dysfunction or marked hypertrophy or inducible sustained VT or VF (Al-Khatib, 2017; Shen, 2017)
- Syncope and moderate or severe complexity CHD, with high clinical suspicion of ventricular arrhythmias
- Non-hospitalized patients with CHD awaiting heart transplantation
- Left ventricular non-compaction that meets same indications as NICM, including a familial history of SCD (Biagini, 2006; Russo, 2018)

EXEMPTIONS

Indications for ICD with an Appropriate Pacing Modality in Special Situations

(Katsumoto, 2014; Russo, 2013)*

- ICD criteria met, and elevated troponin is deemed not due to a myocardial infarction (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**
- 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)

*** With these ICD indications, CRT would sometimes be the appropriate pacing modality. CRT is likely to be the appropriate modality with anticipated requirement for significant (> 40%) ventricular pacing**

**** These indications avoid a second implantation procedure within less than 3 months**

BACKGROUND

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

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).

Patient eligibility for an ICD presumes all the following:

- Anticipated reasonable quality of life for \geq 1-year post implantation (Katsumoto, 2018)
- 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 Background Information section on reversible causes)
- Completion of \geq 3 months of guideline-directed medical therapy (GDMT) for heart failure (HF), unless an intervening indication for pacemaker implantation arises (see Background Information section for definition of GDMT)
- ICD indications are present in most 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

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

OVERVIEW

General

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

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 to terminate the rapid rhythm and restore a normal heart rhythm. There are two types of therapy that can be delivered:
 - Rapid pacing OR
 - High-voltage shocks are necessary for ventricular fibrillation and when rapid pacing has failed to correct the abnormal rhythm
- In addition, all ICDs have pacing capability, and 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

NYHA Class Definitions

(Campeau, 1976; Goldman, 1981; Russo, 2013)

- **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, and walk 4 MPH on level ground).
- **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.

Guideline-Directed (or Optimal) Medical Therapy for Heart Failure

(Yancy, 2013, 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) (Kotek, 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

- Normal serum sodium and potassium
- 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.

Abbreviations

ACE-I	Angiotensin converting enzyme inhibitor
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
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
SCD	Sudden Cardiac Death
STEMI	ST-elevation myocardial infarction
SND	Sinus node dysfunction
VT	Ventricular tachycardia
VF	Ventricular fibrillation

POLICY HISTORY

Date	Summary
March 2021	<ul style="list-style-type: none"> • Added section to clarify indications for unexplained syncope • Revised and added reference for hypertrophic cardiomyopathy to include: <ul style="list-style-type: none"> ○ Indication for EF < 50% • Added statement on consideration for ICD in children • Statement on placement of ICD in competitive athletics • Addition: All indications are predicated on an expected life expectancy of greater than one year if the ICD is implanted. • Reorganization: Ischemic Heart Disease (CAD) sections • Addition : indications under ischemic heart disease for NSVT due to prior MI, LVEF ≤ 40%, and inducible VT or VF at EPS • Revision: HCM section as per new HOCM guidelines J Am Coll Cardiol. 2020 Dec, 76 (25) 3022–3055 • Addition: Neuromuscular Disorders section, various included types, expanded criteria • Deletion: Channelopathies section, deleted types from High Risk Genotype • Addition: Brugada syndrome, broadened definition to “Syncope presumed to be due to ventricular arrhythmia” • Addition: added “J-wave Syndrome” to Early Repolarization section • Changes to Adult & Pediatric Congenital Heart Disease (CHD) section: <ul style="list-style-type: none"> • Addition: added “Cardiac arrest due to VF or VT, or unstable VT, after exclusion of a reversible etiology” to Adult & Pediatric Congenital Heart Disease (CHD) section • Revision: Tetralogy of Fallot, changed to “Residual pulmonary regurgitation or stenosis” • Addition: Tetralogy of Fallot, added RV Hypertension • Revision: “Syncope of unknown origin in the presence of either at least moderate ventricular dysfunction or marked hypertrophy or inducible sustained VT or VF” • Revision: “Syncope and moderate or <u>severe</u> complexity CHD, with high clinical suspicion of ventricular arrhythmias”
March 2020	<ul style="list-style-type: none"> • Added general information section as Introduction which outlines requirements for documentation of pertinent office notes by a licensed clinician, and inclusion of laboratory testing and relevant imaging results for case review • Removed the statement regarding waiting period from the Overview section • Updated and added new references

August 2019	<ul style="list-style-type: none"> • Removed indications under ischemic heart disease for NSVT due to prior MI, LVEF \leq 40%, and inducible VT or VF at EPS • Removed indications under ischemic heart disease for VT or VF < 48 hours post MI or elective coronary revascularization • Under NICM, removed indication for peripartum cardiomyopathy with LVEF \leq 35% that persists > 3 months • Under advanced heart failure and transplantation, removed indication for severe allograft vasculopathy • Revision to cardiac sarcoidosis indication to add cardiac arrest • Under hypertrophic cardiomyopathy revised indications for documented NSVT to include an additional SCD risk modifier (age < 30 yr, delayed hyperenhancement on cardiac MRI, LVOT obstruction, or syncope > 5 yr ago) or high risk feature (LV aneurysm or LVEF < 50%) • Removed indications for giant cell myocarditis and chronic Chagas cardiomyopathy • Removed indication for hypertensive heart disease with LVH and LVEF \leq 35% • Under Tetralogy of Fallot added the following indications : <ul style="list-style-type: none"> ○ Prior palliative systemic to pulmonary shunts ○ Unexplained syncope ○ Frequent PVCs ○ Atrial tachycardia ○ Left ventricular diastolic dysfunction ○ Dilated right ventricle

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Reviewed / Approved by NIA Clinical Guideline Committee

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