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2024 Defibrillator

Cardiology Services

CARD-CDEF-BCBSSC

Last Review Date: 02/29/2024
Previous Review Date: 02/28/2023
Guideline Initiated: 06/30/2019

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Table of Contents

BlueCross and BlueShield of South Carolina

IMPORTANT
To locate the appropriate updated Clinical Policies for BlueCross and BlueShield of South Carolina, please go to: https://www.southcarolinablues.com/web/public/brands/sc/providers/policies-and-authorizations/medical-policies/

TIP
A National Coverage Determination (NCD) or Local Coverage Determination (LCD) may be necessary to review for Medicare participants. Please go to: https://www.cms.gov/medicare-coverage-database/search.aspx for the latest coverage determination information.

Internal Use Only

11231 11235 11233 11238 11241 11234 11240 11474 11224 11229 11225 11226 11274 11269 11267 11268 11264 11263 11236 11237 11239 11242 11228 11646 11275 11262 11265 11266 11270

Cardiac Resynchronization Therapy-Defibrillator (CRT-D)

IMPORTANT
See also, **NCD 20.4**: Implantable Automatic Defibrillators at https://www.cms.gov/medicare-coverage-database/search.aspx *if applicable to individual's healthplan membership*.

CRT-D Insertion or Replacement Guideline

Cardiac resynchronization therapy-defibrillator (CRT-D) is considered medically appropriate when the documentation demonstrates **ALL** of the following:

1. Heart failure (HF) is classified as New York Heart Association (NYHA) class II or III, or ambulatory class IV.

References: [10] [18] [3]

2. Heart failure (HF) is managed on medical therapy.
   References: [10] [18]
3. Left ventricular ejection fraction (LVEF) is 35% or less.
   References: [10] [28] [3]
4. Life expectancy is 1 year or longer and good functional capacity is anticipated.
   References: [10] [1]
5. Sinus rhythm and QRS interval is 130 msec or more. [1]
   References: [10] [28]
6. Ventricular pacing is required (current or anticipated).
   References: [10]

CRT-D Removal Guideline

CRT-D removal (eg, for replacement) is considered medically appropriate when the documentation demonstrates **ANY** of the following:

1. Battery is at end-of-life span, per device manufacturer, or a battery error message occurred, with request for a replacement of the current battery.
   Reference: [8]
2. Complication related to the device including **ANY** of the following:
   a. Cardiac device or implant leakage
   b. Displacement or migration of device
   c. Erosion of device through the skin
   d. Infection is related to implant or is chronic.
   e. Pain is associated with the device.

   References: [8] [15]
3. Lead fracture or malfunction
   References: [8] [15]
4. National recall of device
   Reference: [8]

[1]The European Society of Cardiology (ESC) 2021 cardiac resynchronization therapy updates recommend the QRS threshold be 130 msec or more. This recommendation was based on the Echo-CRT trial, which suggested possible harm from CRT when echocardiographic mechanical dyssynchrony with a QRS duration less than 130 msec were studied.

CRT-D Lead Replacement Guideline

Lead replacement is considered medically appropriate when the documentation demonstrates **ANY** of the following:

1. Cardiac device or implant leakage (other than CRT-D)
   References: [15] [5]
2. Infection is related to implant or lead.
   References: [15] [5]
3. Lead fracture, migration or malfunction
   References: [15] [5]

CRT-D Upgrade from Single Chamber to Dual Chamber Device with Biventricular Pacing Capability Guideline

Replacement with a biventricular pacing device is considered medically appropriate when the documentation demonstrates **ALL** of the following:

1. Condition requires pacing (eg, chronotropic incompetence).
   Reference: [26]
2. Heart failure is progressing to New York Heart Association (NYHA) class of II, III or IV
   References: [26] [11]
3. QRS interval is 120 msec or more.[1]
   Reference: [26] [19] [10]

LCD 39080
See also, **LCD 39080:** Cardiac Resynchronization Therapy at www.cms.gov/medicare-coverage-database/search.aspx *if applicable to individual's healthplan membership*.

CRT-D Procedure Codes

Table 1. Cardiac Resynchronization Therapy-Defibrillator (CRT-D) Associated Procedure Codes

CODE	DESCRIPTION
0695T	Body surface-activation mapping of pacemaker or pacing cardioverter-defibrillator lead(s) to optimize electrical synchrony, cardiac resynchronization therapy device, including connection, recording, disconnection, review, and report; at time of implant or replacement

CODE	DESCRIPTION
33215	Repositioning of previously implanted transvenous pacemaker or implantable defibrillator (right atrial or right ventricular) electrode
33216	Insertion of a single transvenous electrode, permanent pacemaker or implantable defibrillator
33217	Insertion of 2 transvenous electrodes, permanent pacemaker or implantable defibrillator
33218	Repair of single transvenous electrode, permanent pacemaker or implantable defibrillator
33220	Repair of 2 transvenous electrodes for permanent pacemaker or implantable defibrillator
33224	Insertion of pacing electrode, cardiac venous system, for left ventricular pacing, with attachment to previously placed pacemaker or implantable defibrillator pulse generator (including revision of pocket, removal, insertion, and/or replacement of existing generator)
33230	Insertion of pacing implantable defibrillator pulse generator only; with existing dual leads
33231	Insertion of pacing implantable defibrillator pulse generator only; with existing multiple leads
33240	Insertion of implantable defibrillator pulse generator only; with existing single lead
33241	Removal of implantable defibrillator pulse generator only
33244	Removal of single or dual chamber implantable defibrillator electrode(s); by transvenous extraction
33249	Insertion or replacement of permanent implantable defibrillator system with transvenous lead(s), single or dual chamber
33263	Removal of implantable defibrillator pulse generator with replacement of implantable defibrillator pulse generator; dual lead system
33264	Removal of implantable defibrillator pulse generator with replacement of implantable defibrillator pulse generator; multiple lead system
33264	Removal of implantable defibrillator pulse generator with replacement of implantable defibrillator pulse generator; multiple lead system
C1900	Lead, left ventricular coronary venous system
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

Table 2. Cardiac Defibrillator or Pacemaker Device Associated Secondary Codes (Authorization Requirements Depend on Primary Procedure)

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

Implantable Cardioverter Defibrillator (ICD) guideline

IMPORTANT
See also, **NCD 20.4**: Implantable Automatic Defibrillators at https://www.cms.gov/medicare-coverage-database/search.aspx *if applicable to individual's healthplan membership*.

Implantable Cardioverter Defibrillator (ICD) Guideline

A single or dual chamber implantable cardiac defibrillator (ICD) for the primary or secondary prevention of sudden cardiac death (SCD), is considered medically appropriate when the documentation demonstrates **ANY** of the following:

1. Anti-tachycardia pacing is part of safe/appropriate treatment.[2]
   References: [2] [8] [9]
2. Brugada syndrome, with syncope or ventricular tachycardia
   References: [2] [8] [9]
3. Cardiac sarcoidosis with expected survival of more than 1 year and **ANY** of the following:
   a. Left ventricular ejection fraction (LVEF) is 30% or less.
   b. Sudden cardiac death survivor
   c. Ventricular tachycardia is sustained.

   References: [2] [8] [9]
4. Cardiac transplant is pending (awaiting in outpatient setting).
   References: [2] [8] [9]
5. Chagas disease with an ejection fraction (EF) of 40% or less, as an alternative to medical therapy
   References: [2] [8] [9]
6. EF is 40% or less, with non-sustained ventricular tachycardia **AND** inducible ventricular tachycardia/fibrillation on electrophysiology study (EPS).
   References: [2] [8] [9]
7. Hypertrophic cardiomyopathy and **ANY** of the following:
   a. Blood pressure (BP) response to exercise is abnormal (20 mm Hg decrease in baseline BP or **NO** increase in systolic BP by at least 20 mm Hg during effort).

[2]Indication for dual chamber implantable cardiac defibrillator/pacemaker.

b. Left ventricular hypertrophy is massive (wall thickness of greater than 30mm).

c. Sudden cardiac death family history

d. Syncope

e. Ventricular tachycardia is non-sustained.

f. Ventricular tachycardia **OR** ventricular fibrillation arrest survivor

References: [2] [8] [9] [4] [22] [18]

8. Ischemic cardiomyopathy and **EITHER** of the following:

   a. Myocardial infarction is **MORE** than forty (40) days ago, postrevascularization (eg, coronary artery bypass grafting [CABG], percutaneous coronary intervention [PCI]) is **MORE** than ninety (90) days ago and **ANY** of the following:

      i. Heart failure (HF) New York Heart Association (NYHA) class I with EF 30% or less

      ii. HF NYHA class II or III with EF 35% or less

   b. Ventricular tachycardia is non-sustained, EPS demonstrates inducible ventricular tachycardia or ventricular fibrillation **AND** EF is less than 40%.

   References: [2] [8] [9]

9. Long QT syndrome (QRS duration 120 msec or more) **AND** is experiencing syncope or ventricular tachycardia while on beta-blocker (for primary prevention).
   References: [2] [8] [9]

10. Non-ischemic cardiomyopathy, EF is 35% or less **AND** HF NYHA class II or III.
    References: [2] [8] [9] [23]

11. Right ventricular dysplasia is arrhythmogenic and **ANY** of the following:

    a. Arrhythmogenic right ventricular dysplasia-2 (ARVC2) or ARVC5 genetic testing is positive.

    b. Cardiac arrest history or ventricular tachycardia with hemodynamic compromise

    c. QRS dispersion is increased.

    d. Syncope

    References: [2] [8] [9] [17]

12. Syncope of unknown etiology and **ANY** of the following:

    a. Left ventricular dysfunction with EF of 50% or less **AND** comprehensive evaluation (12-lead electrocardiogram [ECG], ambulatory ECG monitoring, history and physical exam, orthostatic blood pressure measurements, stress testing, tilt-table test) is non-diagnostic for etiology

b. Ventricular fibrillation **OR** sustained ventricular tachycardia is demonstrated on EPS.

References: [2] [8] [9]

13. Ventricular tachycardia is sustained, spontaneous and **NO** reversible cause is identified.
References: [2] [8] [9]

14. Ventricular tachycardia or ventricular fibrillation arrest survivor and **NO** reversible cause is identified.
References: [2] [8] [9]

Subcutaneous Implantable Cardiac Defibrillator (SICD) Guideline

A subcutaneous implantable cardiac defibrillator (S-ICD) insertion for primary or secondary prevention is considered medically appropriate when the documentation demonstrates **ALL** of the following:

1. **NONE** of the following:
    a. Bradycardia is symptomatic.
    b. Ventricular tachycardia and **ANY** of the following:
        i. Incessant
        ii. Spontaneous, frequently occurring and reliably terminated with anti-tachycardia pacing or a unipolar pacemaker.

    References: [2] [14]

2. Clinical condition includes **ANY** of the following:
    a. Anti-tachycardia pacing is part of safe/appropriate treatment.
    b. Atrioventricular (AV) block is 2nd or 3rd degree.
    c. Brugada syndrome, with syncope or ventricular tachycardia
    d. Cardiac arrest survivor from ventricular tachycardia or ventricular fibrillation, when **NO** reversible cause is identified.
    e. Cardiac sarcoidosis, with expected survival of more than 1 year, and **ANY** of the following:
        i. LVEF of 30% or less
        ii. LVEF of 35% or more and **ANY** of the following:
            A. Inducible ventricular arrhythmia is present.

B. Permanent pacing is indicated.

C. Scar is present per cardiac magnetic resonance imaging (MRI) or positron emission tomography (PET).

D. Syncope is present.

iii. Sudden cardiac death survivor

iv. Ventricular tachycardia is sustained.

f. Cardiac transplant is pending (awaiting in outpatient setting).

g. Chagas disease with an EF of 40% or less, as an alternative to medical therapy

h. Hypertrophic cardiomyopathy and **ANY** of the following:

i. Blood pressure response to exercise is abnormal (20 mm Hg decrease in baseline BP or **NO** increase in systolic BP by at least 20 mm Hg during effort).

ii. Left ventricular hypertrophy is massive (wall thickness of greater than 30mm).

iii. Sudden cardiac death family history

iv. Syncope

v. Ventricular tachycardia is non-sustained.

vi. Ventricular tachycardia or ventricular fibrillation arrest survivor

i. Ischemic cardiomyopathy and **ANY** of the following:

i. Myocardial infarction is **MORE** than forty (40) days ago, postrevascularization (eg, coronary artery bypass grafting [CABG], percutaneous coronary intervention [PCI]) is **MORE** than ninety (90) days ago and **ANY** of the following:

A. Heart failure (HF) NYHA class I with EF 30% or less

B. HF NYHA class II or III with EF 35% or less

ii. Ventricular tachycardia is non-sustained with EPS demonstrating inducible ventricular tachycardia or ventricular fibrillation **AND** an EF of less than 40%.

j. Long QT syndrome (QRS duration of 120 msec or more) **AND** syncope or ventricular tachycardia while on beta-blocker (for primary prevention)

k. Non-ischemic cardiomyopathy, EF is 35% or less **AND** HF NYHA class II or III.

l. Right ventricular dysplasia is arrhythmogenic and **ANY** of the following:

i. Age is 50 years or less.

ii. ARVC2 or ARVC5 genetic testing is positive.

iii. Cardiac arrest history **OR** ventricular tachycardia with hemodynamic compromise

iv. QRS dispersion is increased.

v. Syncope

vi. Ventricular involvement

m. Syncope of unknown etiology and **ANY** of the following:

i. Comprehensive evaluation (ambulatory ECG monitoring, 12-lead ECG, history and physical exam, orthostatic blood pressure measurements, stress testing, tilt-table test) is non-diagnostic for etiology **AND** left ventricular dysfunction when EF is 50% or less.

ii. EPS demonstrates ventricular fibrillation or sustained ventricular tachycardia.

n. Ventricular tachycardia is non-sustained, EPS demonstrates inducible ventricular tachycardia or ventricular fibrillation **AND** EF is less than 40%.

o. Ventricular tachycardia is sustained, spontaneous and **NO** reversible cause is identified.

References: [2] [14] [9] [13] [23] [4] [22] [18] [17]

ICD or SICD Removal or Repositioning Guideline

Removal (eg, for replacement) or repositioning of a defibrillator is considered medically appropriate when the documentation demonstrates **ANY** of the following:

1. Device replacement is needed earlier than replacement interval and **ANY** of the following:
   a. Erosion of device through skin
   b. Error of device
   c. Excessive external manipulation
   d. Infection is implantation-related or chronic.
   e. Lead fracture
   f. Leakage of other cardiac devices/implants
   g. National recall on device by the manufacturer
   h. Pain is related to the device.

Reference: [8]

2. End-of-life span as advised by device manufacturer
Reference: [8]

ICD Procedure Codes

Table 1. Automatic Implantable Cardioverter Defibrillator (AICD) Associated Procedure Codes

CODE	DESCRIPTION
33215	Repositioning of previously implanted transvenous pacemaker or implantable defibrillator (right atrial or right ventricular) electrode
33216	Insertion of a single transvenous electrode, permanent pacemaker or implantable defibrillator
33217	Insertion of 2 transvenous electrodes, permanent pacemaker or implantable defibrillator
33218	Repair of single transvenous electrode, permanent pacemaker or implantable defibrillator
33220	Repair of 2 transvenous electrodes for permanent pacemaker or implantable defibrillator
33223	Surgery to relocate pocket of skin that holds heart-assist device. The device emits electrical pulses to train the heart, improving its function.
33230	Insertion of pacing implantable defibrillator pulse generator only; with existing dual leads
33240	Insertion of implantable defibrillator pulse generator only; with existing single lead
33241	Removal of implantable defibrillator pulse generator only
33243	Surgery to remove one or more pacing wires for a cardioverter-defibrillator device.
33244	Removal of single or dual chamber implantable defibrillator electrode(s); by transvenous extraction
33249	Insertion or replacement of permanent implantable defibrillator system with transvenous lead(s), single or dual chamber
33262	Removal of implantable defibrillator pulse generator with replacement of implantable defibrillator pulse generator; single lead system
33263	Removal of implantable defibrillator pulse generator with replacement of implantable defibrillator pulse generator; dual lead system
33270	Insertion or replacement of permanent subcutaneous implantable defibrillator system, with subcutaneous electrode, including defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters, when performed
33271	Insertion of subcutaneous implantable defibrillator electrode
33272	Removal of subcutaneous implantable defibrillator electrode
33273	Repositioning of previously implanted subcutaneous implantable defibrillator electrode
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)

CODE	DESCRIPTION
C1899	Lead, pacemaker/cardioverter-defibrillator combination (implantable)
0571T	Insertion or replacement of implantable cardioverter-defibrillator system with substernal electrode(s), including all imaging guidance and electrophysiological evaluation (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters), when performed
0572T	Insertion of substernal implantable defibrillator electrode
0573T	Removal of substernal implantable defibrillator electrode
0574T	Repositioning of previously implanted substernal implantable defibrillator-pacing electrode
0580T	Removal of substernal implantable defibrillator pulse generator only
0614T	Removal and replacement of substernal implantable defibrillator pulse generator

Wearable Cardiac Defibrillator (WCD)

Wearable Cardiac Defibrillator (WCD) Guideline

A wearable cardiac defibrillator (WCD) is considered medically appropriate when the documentation demonstrates **ALL** of the following:

1. Adherence is demonstrated with medical treatments (eg, appointment maintenance, medication regimen).
   References: [1] [6] [20] [27] [7]
2. Clinical condition includes **ANY** of the following:
   a. Cardiomyopathy with **ALL** of the following:
      i. **ANY** of the following:
         A. Ischemic cardiomyopathy with sustained ventricular tachyarrhythmia (VT) (more than 30 seconds long) and **ANY** of the following:
            I. Myocardial infarction (MI) is within the past 40 days.
            II. VT is induced by electrophysiologic study (EPS).
            III. Spontaneous VT is **NOT** due to transient or irreversible causes **OR** spontaneously occurred more than 48 hours after a myocardial infarction.
         B. Peri-partum cardiomyopathy
      ii. Ejection fraction (EF) is 35% or less (from prior cardiac catheterization, cardiac magnetic resonance imaging [MRI], coronary computed tomography

angiography [CCTA], multigated acquisition [MUGA] scan, transthoracic echocardiogram [TTE]).

iii. Implantable cardioverter defibrillator (ICD) is clinically appropriate and planned to be placed in the next 60 days with **NO** contraindications for ICD placement. [3]

b. Myocardial infarction (MI) occurred within past 40 days and **ANY** of the following:

i. EF is 35% or less.

ii. ICD is planned 40 days after the MI.

c. Peri-operative when **ANY** of the following:

i. Post-operative ICD failure or local infection, with planned reimplant ICD, when **ANY** of the following:

A. After 60 days from the device failure

B. Antibiotic therapy is complete.

C. Infection is resolved (within 30 days to 60 days).

ii. Post revascularization (eg, coronary artery bypass graft [CABG], percutaneous coronary intervention [PCI]) within the past 90 days and **ALL** of the following:

A. ICD is clinically appropriate and planned to be placed more than 90 days post-operative with **NO** contraindications for ICD placement.

B. Left ventricular ejection fraction (LVEF) persists at 35% or less.

d. Sudden cardiac death (SCD) risk and **ALL** of the following:

i. Clinical condition includes **ANY** of the following:

A. Genetic/familial risk with **ANY** of the following:

I. Cardiac arrest survival

II. Hypertrophic obstructive cardiomyopathy (HOCM) family history, **AND** left ventricular septal thickness more is than 30 mm (from prior cardiac catheterization, cardiac MRI, CCTA, MUGA, TTE).

III. Long QT syndrome (QRS duration 120 msec or more) by family history

[3] **Contraindications to ICD placement** include cancer, uremia, liver failure or chronic end-stage disease with life expectancy of 1 year or less.

IV. Pre-syncope is unexplained **OR** syncope with 2 or more episodes and **ALL** of the following:

1. Blood pressure response to exercise is abnormal (20 mm Hg decrease in baseline BP or **NO** increase in systolic BP by at least 20 mm Hg during effort).
2. SCD occurred in a first degree relative (child, parent, sibling)

B. Ventricular fibrillation cardiac arrest

C. Ventricular fibrillation, with no reversible cause

ii. EF is 35% or less.

iii. ICD and **ANY** of the following:

A. ICD is clinically appropriate and planned to be placed in the next 60 days with **NO** contraindications for ICD placement.

B. ICD is inappropriate or refused.

References: [1] [6] [20] [7] [25] [16] [27] [4] [21]

3. **NO** contraindications for WCD (eg, comorbidity exacerbation, infection)
References: [1] [6] [20] [27] [7]

WCD Procedure Codes

Table 1. Wearable Cardiac Device (WCD) Associated Procedure Codes

CODE	DESCRIPTION
93745	Initial set-up and programming by a physician of wearable cardioverter-defibrillator includes initial programming of system, establishing baseline electronic ECG, transmission of data to data repository, patient instruction in wearing system and patient reporting of problems or events
K0606	Automatic external defibrillator, with integrated electrocardiogram analysis, garment type
K0607	Replacement battery for automated external defibrillator, garment type only, each
K0608	Replacement garment for use with automated external defibrillator, each
K0609	Replacement electrodes for use with automated external defibrillator, garment type only, each

Defibrillator Summary of Changes

The Defibrillator guidelines from 2023 to 2024 had the following changes:

- Cardiac Resynchronization Therapy-Defibrillator (CRT-D) clinical guidelines from 2023 to 2024 had the following version changes:

- Added chronotropic incompetence, defibrillator and QRS interval to the definition section.
- "Migration of device" indication under CRT-D removal guideline was removed as it was a duplicate found under ll B.
- "Replacement with a biventricular pacing device (from a single or dual chamber) is considered medically appropriate when the documentation demonstrates ALL of the following" was updated to "Replacement with a biventricular pacing device is considered medically appropriate when the documentation demonstrates ALL of the following".
- Reviewed and updated references to ensure they are within the 5 year time frame or they are sentinel articles.

- Implantable Cardioverter Defibrillator (ICD) clinical guidelines from 2023 to 2024 had the following version changes:
  - Added beta blocker, bradycardia, incessant, MRI, PET and subcutaneous to the definition section.
  - "EF is 40% or less, with non-sustained ventricular tachycardia AND inducible ventricular tachycardia/fibrillation on electrophysiology study (EPS)" indication was updated to "Ventricular tachycardia is non-sustained, electrophysiology study (EPS) demonstrates inducible ventricular tachycardia or ventricular fibrillation AND EF is less than 40%".
  - "**MORE** than forty (40) days since a myocardial infarction, **MORE** than ninety (90) days postrevascularization (eg, coronary artery bypass grafting [CABG], percutaneous coronary intervention [PCI]) and **ANY** of the following:" was updated to "Myocardial infarction is MORE than forty (40) days ago, postrevascularization (eg, coronary artery bypass grafting [CABG], percutaneous coronary intervention [PCI]) is MORE than ninety (90) days ago and ANY of the following:"
  - Reviewed and updated references to ensure they are within the 5 year time frame or they are sentinel articles.
- Wearable Cardiac Defibrillator (WCD) clinical guidelines from 2023 to 2024 had the following version changes:
  - Added CABG, CCTA, MUGA, myocardial infarction and PCI to the definition section.
  - Reviewed and updated references to ensure they are within the 5 year time frame or they are sentinel articles.
- Pediatric indication additions:

- Cardiac Resynchronization Therapy-Defibrillator (CRT-D)
  - "Congenital heart disease is known" indication
- Implantable Cardioverter Defibrillator

Defibrillator Definitions

Angiotensin-converting enzyme inhibitor (ACE inhibitors) are medications that block the conversion of angiotensin I to angiotensin II, leading to decreased blood pressure and reduced workload on the heart.

Aortic Coarctation is a birth defect in which a part of the aorta is narrower than usual.

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a form of heart disease that usually appears in adulthood. ARVC is a disorder of the myocardium, which is the muscular wall of the heart. This condition causes part of the myocardium to break down over time, increasing the risk of an abnormal heartbeat (arrhythmia) and sudden death.

Arrhythmogenic right ventricular dysplasia (ARVD) (also called arrhythmogenic right ventricular cardiomyopathy, right ventricular cardiomyopathy or right ventricular dysplasia) is a rare type of cardiomyopathy that occurs when the muscle tissue in the right ventricle dies and is replaced with scar tissue. This disrupts the heart's electrical signals and causes arrhythmias. Symptoms include palpitations and fainting after physical activity. ARVD usually affects teens or young adults and can cause sudden cardiac arrest (SCA) in young athletes. Researchers believe that arrhythmogenic right ventricular dysplasia is an inherited disease.

Atrioventricular (AV) Block is an interruption or delay of electrical conduction from the atria to the ventricles due to conduction system abnormalities in the AV node or the His-Purkinje system (electrical conduction fibers and cells in the ventricles). Conduction delay or block can be physiologic if the atrial rate is abnormally fast or pathologic at normal atrial rates.

Beta blocker is a type of drug that blocks the action of substances, such as adrenaline, on nerve cells and causes blood vessels to relax and dilate (widen). This allows blood to flow more easily and lowers blood pressure and the heart rate.

Bradycardia is a heart rate that is too slow. What is considered too slow can depends on age and physical condition. In general, for adults, a resting heart rate of fewer than 60 beats per minute (BPM) qualifies as bradycardia. Causes for bradycardia may include: problems with the sinoatrial (SA) node, sometimes called the heart’s natural pacemaker, problems in the conduction pathways of the heart that do not allow electrical impulses to pass properly from the atria to the ventricles, metabolic problems (eg, hypothyroidism), damage to the heart from heart disease or heart attack, and certain heart medications that can cause the side effect of bradycardia.

Brugada syndrome is a rare inherited cardiovascular disorder characterized by disturbances affecting the electrical system of the heart. The main symptom is irregular heartbeat and, without treatment, may result in sudden death.

Cardiomyopathy is a disease of the heart muscle that makes it harder for the heart to pump blood to the rest of the body. Cardiomyopathy can lead to heart failure. The main types of cardiomyopathy include dilated, hypertrophic and restrictive cardiomyopathy.

Cardiac resynchronization therapy is a procedure to implant a device in the chest to make the heart's chambers contract in a more organized and efficient way. Cardiac resynchronization therapy (CRT) uses a device called a biventricular pacemaker (also called a cardiac resynchronization device) that sends electrical signals to both ventricles. The signals trigger the ventricles to contract in a more coordinated way, which improves the pumping of blood out of the heart. Sometimes the device also contains an implantable cardioverter-defibrillator (ICD), which can deliver an electrical shock to reset the heart if the heart rhythm becomes dangerously erratic.

Cardiac sarcoidosis is an inflammatory granulomatous disease that can affect the heart. Up to one-quarter of the population with systemic sarcoidosis may have evidence of cardiac involvement. The clinical manifestations of cardiac sarcoidosis (CS) include heart block, atrial arrhythmias, ventricular arrhythmias and heart failure.

Cardiac sympathetic denervation (CSD) is a surgical procedure aimed at reducing ventricular arrhythmias by removing or interrupting the sympathetic nerves supplying the heart. It is particularly used in cases refractory to other treatments.

Catecholamenergic polymorphic ventricular tachycardia (CPVT) is a rare, inherited arrhythmia syndrome characterized by exercise or emotion-induced ventricular arrhythmias in individuals with structurally normal hearts and a normal resting electrocardiogram (ECG).

Chagas disease is caused by the tropical parasite Trypanosoma cruzi, which is transmitted to animals and people by insect vector and is also referred to as American trypanosomiasis. There is an acute and, if untreated, chronic phase. Complications of chronic Chagas may include arrhythmia, congestive heart failure and/or a dilated esophagus or colon.

Chronotropic incompetence is the inability to increase and maintain heart rate appropriately in the setting of increased physiologic demand.

Class 1C antiarrhythmic drugs (AADs) are effective first-line agents for atrial fibrillation (AF) treatment. These agents commonly are avoided in patients with known coronary artery disease (CAD), due to known increased risk in the postmyocardial infarction population.

Coronary artery bypass graft (CABG) is a surgical procedure performed to shunt blood around a narrowing or blockage in the coronary artery of the heart. This procedure involves attaching one end of a segment of blood vessel (eg, a vein of the leg) that was removed from another part of the body into the aorta, and the other end of the segment into the coronary artery beyond the obstructed area, to increase blood flow.

Coronary computed tomography angiography (CCTA) is a non-invasive test that uses a computed tomography (CT) scanner to obtain a 3-dimensional image of the heart, including blood vessels that supply blood to the heart muscle (coronary arteries). During the CCTA, contrast dye is injected into the vein so that the coronary arteries can be seen. CCTA provides images to

identify a narrowing or blockage of the coronary arteries caused by plaque and allows for accurate visualization of the 3-dimensional heart structure (to include the valves of the heart).

Defibrillator is a device that provides an electric shock to the heart to allow it to get out of a potentially fatal abnormal heart rhythm, or arrhythmia.

Ebstein anomaly is a congenital heart defect characterized by the abnormal development and displacement of the tricuspid valve, leading to tricuspid regurgitation and right ventricular dysfunction.

Ejection fraction (EF) is a measurement of how much blood the left ventricle pumps out with each contraction. It is measured in percentages with a normal measurement usually between 50 and 70%.

Electrocardiogram (ECG or EKG) is a test that measures and records the electrical activity of the heart. The ECG electrical activity is divided into the P wave, PR interval, QRS complex, QT interval, ST segment, T wave and U wave. An ECG is useful in establishing many cardiac diagnoses.

Emery-Dreifuss muscular dystrophy (EDMD) is a genetic condition characterized by muscle weakness, joint contractures, and cardiac complications.

Electrophysiology study (EPS) is a minimally invasive procedure that evaluates the electrical conduction system of the heart to assess the electrical activity, conduction pathways and abnormal heart beats. During an EPS, the sinus rhythm, and supraventricular and ventricular arrhythmias of baseline cardiac intervals, are recorded. The study is indicated to investigate the cause, location of origin and best treatment (drug therapy, catheter ablation or implantable cardioverter-defibrillator), for various abnormal heart rhythms.

Guideline-directed medical therapy (GDMT) refers to the optimal course of treatment for each stage of a chronic cardiac condition (eg, angina, heart failure), including those at high risk of disease progression but without structural heart disease or symptoms. The goal is titration of medications to maximum tolerated doses.

Heart block also called atrioventricular (AV) block, is partial or complete interruption of impulse transmission from the atria to the ventricles. [4]

Normally, electrical signals travel from the upper chambers of the heart (atria) to the lower chambers (ventricles). The AV node is a cluster of cells that connect the electrical activity from the top chambers of your heart (atria) to the bottom chambers (ventricles). A heart block occurs when the electrical signal does not travel through the AV node to the ventricles. The result is a heart that does not function effectively, and it cannot pump blood through its chambers and out to the body as a normal heart would. Heart block can be first, second or third degree, depending on the extent of electrical signal impairment.

[4]Merck & Co., Inc., "Atrioventricular Block." [Online]. Available: www.merckmanuals.com

First-degree heart block: The electrical impulse still reaches the ventricles, but moves more slowly than normal through the AV node. The impulses are delayed. This is the mildest type of heart block.

Second-degree heart block is classified into two categories: Type I and Type II. In second-degree heart block, the impulses are intermittently blocked. Type I, also called Mobitz Type I or Wenckebach's AV block: This is a less serious form of second-degree heart block. The electrical signal gets slower and slower until the heart actually skips a beat. Type II, also called Mobitz Type II: While most of the electrical signals reach the ventricles every so often, some do not and the heartbeat becomes irregular and slower than normal.

Third-degree heart block: The electrical signal from the atria to the ventricles is completely blocked. To make up for this, the ventricle usually starts to beat on its own acting as a substitute pacemaker but the heartbeat is slower and often irregular and not reliable. Third-degree block seriously affects the heart's ability to pump blood out to the body.

Heart failure (HF) (also known as **congestive heart failure [CHF]**) is a condition that develops when the heart is unable to pump enough blood for the body's needs. HF occurs when the heart cannot fill with enough blood or is too weak to pump properly. Decompensated heart failure is sudden worsening (exacerbation) of heart failure symptoms (eg, difficulty breathing, lower extremity edema, fatigue) to where the heart can no longer continue to compensate for its full function.

Hemodynamic instability is a condition caused by abnormal or unstable blood pressure that results in improper circulation and organs of the body do not receive adequate blood flow. It is characterized by chest pain, confusion, abnormal heart rate, loss of consciousness, restlessness, shortness of breath, cold hands, arms, legs or feet, etc.

Hypertrophic cardiomyopathy (HCM) is a congenital or acquired disorder, characterized by marked ventricular hypertrophy with diastolic dysfunction but without increased afterload (eg, due to valvular aortic stenosis, coarctation of the aorta, systemic hypertension). In obstructive HCM, the wall (septum) between the two bottom chambers of the heart thickens. The walls of the pumping chamber can also become stiff. It may block or reduce the blood flow from the left ventricle to the aorta. **Left ventricular outflow tract (LVOT)** obstruction is a common feature of HCM and a cause of symptoms and exercise limitation. LVOT obstruction is defined as a peak LVOT gradient of more than 30 mmHg at rest or more than 50 mmHg with provocation. Most people with HCM have LVOT. In non-obstructive HCM, the heart's main pumping chamber still becomes stiff. This limits how much blood the ventricle can take in and pump out, but blood flow is not blocked. [5,6]

Implantable cardiac defibrillator (ICD) is a mechanical device that is placed within the body and is designed to recognize certain types of arrhythmias such as ventricular tachycardia and

[5]Merck & Co., Inc., "Hypertrophic Cardiomyopathy." [Online]. Available:www.merckmanuals.com

[6]American Heart Association (AHA). "Health Topics." [Online]. Available: www.heart.org

ventricular fibrillation. The defibrillator corrects the heart rhythm when needed by delivering precisely calibrated and timed electrical shocks to restore a normal heartbeat.

Incessant is something that continues without interruption.

Ischemic cardiomyopathy is a type of dilated cardiomyopathy. It is a term that is used when the heart muscle is weakened as a result of coronary artery disease or myocardial infarction.

Lamin A/C mutation refers to genetic alterations in the LMNA gene, which encodes the nuclear envelope proteins lamin A and C. These mutations can lead to a variety of diseases known as laminopathies, affecting multiple systems including skeletal muscle, cardiac muscle, adipose tissue, and others.

Left bundle branch block (LBBB) is a delay or obstruction along the electrical pathway to the heart's left ventricle, which can be caused by underlying heart problems. There are often no symptoms involved, however, symptomatic persons can experience syncope or pre-syncope, fatigue and shortness of breath.

Left ventricular assist device (LVAD) is a device that helps pump blood from the lower chambers of the heart (ventricles) to the rest of the body.

Left ventricular hypertrophy (LVH) is a term for a heart's left pumping chamber that has thickened and may not be pumping efficiently. In response to this pressure overload, the inner walls of the heart may respond by getting thicker. These thickened walls can cause the left ventricle to weaken, stiffen and lose elasticity, which may prevent healthy blood flow.

Limb-girdle muscular dystrophy type 1B (LGMD1B) is an autosomal dominant form of muscular dystrophy characterized by progressive muscle weakness primarily affecting the shoulder and pelvic girdles, with a high prevalence of cardiac arrhythmias and late-onset dilated cardiomyopathy due to mutations in the lamin A/C gene.

Long QT syndrome (LQTS) is an abnormal feature of the heart's electrical system that can lead to a potentially life-threatening arrhythmia called torsades de pointes. Torsades de pointes may result in syncope or sudden cardiac death.

Magnetic resonance imaging (MRI) is a non-invasive diagnostic technique that produces computerized images of internal body tissues and is based on nuclear magnetic resonance of atoms within the body induced by the application of radio waves.

Multigated acquisition (MUGA) scan is a noninvasvie nuclear imaging test also known as radionucleotide ventriculography (RVG) and gated equilibrium radionucleotide angiography (ERNA). that uses a radioactive isotope called technetium tagged to red blood cells (RBC) to evaluate the filling and pumping properties of the heart and physical structures by comparing the illuminated blood pool to the darkened walls on the image. Single or multiple measurements of left and/or right ventricular function are obtained. The method can be used to assess regional and global wall motion; cardiac chamber size and morphology; and ventricular systolic and diastolic function, including left and right ventricular ejection fractions.

Myocardial infarction (MI), also called a heart attack, occurs when the blood flow that brings oxygen to the heart muscle is severely reduced or cut off completely. The coronary arteries that

supply the heart muscle with blood flow can become narrowed from a buildup of fat, cholesterol and other substances that together are called plaque. This process is known as atherosclerosis. When plaque within a coronary artery breaks, a blood clot forms around the plaque and can block the flow of blood through the artery to the heart muscle. Ischemia results when there is an inadequate blood supply to the heart muscle causing damage or death of part of the heart muscle, resulting in an MI.

Table 1. New York Heart Association (NYHA) Functional Classification for Heart Failure

CLASS	SYMPTOMS EXPERIENCED
Class I (Mild)	Cardiac disease, but no symptoms and no limitation in ordinary physical activity (eg, shortness of breath when walking, climbing stairs).
Class II (Mild)	Mild symptoms (eg, mild shortness of breath and/or angina) and slight limitation during ordinary activity.
Class III (Moder-ate)	Marked limitation in activity due to symptoms, even during less-than-ordinary activity, (eg, walking short distances [20–100 m]). Comfortable only at rest. Class IIIa: no dyspnea at rest. Class IIIb: recent dyspnea at rest.
Class IV (Severe)	Severe limitations. Experience symptoms while at rest. Unable to carry on any physical activity with-out discomfort.

Non-missense mutation refers to any mutation that does not result in a simple amino acid substitution (missense mutation). These can include nonsense mutations, frameshift mutations, splice site mutations, and noncoding mutations.

Pacemaker syndrome is an array of cardiovascular and neurologic signs and symptoms resulting from disruption of appropriate AV synchrony (AV dyssynchrony) caused by suboptimal pacing, inappropriate programming of pacing parameters or upper-limit behavior of AV synchronous pacing systems.

Percutaneous coronary intervention (PCI) is a non-surgical procedure that uses a catheter (a thin flexible tube) to place a small structure called a stent to open up blood vessels in the heart that have been narrowed by plaque buildup, a condition known as atherosclerosis.

Peripartum cardiomyopathy is a weakness of the heart muscle that begins sometime during the final month of pregnancy through about five months after delivery, without any other known cause. Most commonly it occurs right after delivery. It is a rare condition that can carry mild or severe symptoms.

Positron emission tomography (PET) scan is a procedure in which a small amount of radioactive glucose (sugar) is injected into a vein, and a scanner is used to make detailed, computerized pictures of areas inside the body where the glucose is taken up. It is a medical imaging test that shows the metabolic or biochemical function of organs and tissues.

QRS interval is the time required for a stimulus to spread through the ventricles (ventricular depolarization), and is normally 0.11 sec or less when measured by a computer (eg, electrocardiogram).

Short QT syndrome (SQTS) is a genetically inherited disorder characterized by a shortened QT interval on the ECG, leading to an increased risk of atrial and ventricular arrhythmias and sudden cardiac death.

Sinus node dysfunction, previously known as sick sinus syndrome, is an abnormal function in the sinoatrial (SA) node (also called sinus node). The sinus node is the natural pacemaker of the heart and is responsible for the regular, rhythmic heartbeat. When the sinus node malfunctions, abnormalities may result (eg, bradycardia, tachycardia, tachycardia-bradycardia syndrome, sinus pauses or arrest). Clinical symptoms result from hypoperfusion of end organs. Symptoms of sinus node dysfunction can include palpitations, syncope, pre-syncope, chest pain, weakness or decreased physical activity tolerance.

Stenosis is a narrowing or constriction of the diameter of a bodily passage or orifice.

Subcutaneous means beneath, or under, all the layers of the skin.

Sudden Cardiac Arrest (SCA) is a sudden cessation of cardiac activity resulting in unresponsiveness with no normal breathing and no signs of circulation. If corrected measures are not performed, the condition progresses to sudden cardiac death (SCD). [7]

Sudden cardiac death (SCD) occurs when the heart malfunctions and unexpectedly and suddenly stops beating due to electrical impulse problems. Myocardial infarction increases the risk of SCD. Conditions associated with SCD include arrhythmogenic right ventricular dysplasia (ARVD), long QT syndrome, hypertrophic obstructive cardiomyopathy (HOCM) or Brugada syndrome.

Syncope is a transient loss of consciousness and postural tone (ability to maintain or change position intentionally) due to insufficient cerebral perfusion. The loss of consciousness is associated with prompt recovery, not needing resuscitation.

Tetralogy of Fallot is a congenital abnormality of the heart characterized by pulmonary stenosis, an opening in the interventricular septum, malposition of the aorta over both ventricles and hypertrophy of the right ventricle.

Transthoracic echocardiogram (TTE) involves placing a device called a transducer on the chest. The device sends ultrasound waves through the chest wall to the heart. As the ultrasound waves bounce off the structures of the heart, a computer converts them into pictures on the computer screen. A TTE uses sound waves to create pictures of the heart chambers, valves, walls and the blood vessels attached to your heart. The test is also called echocardiography or diagnostic cardiac ultrasound.

Ventricular fibrillation (VF) also called V-fib, is a serious cardiac rhythm disorder in which disordered electrical activity causes the heart's lower chambers (ventricles) to quiver or fibrillate,

[7]Al-Khatib, S.M., Stevenson, W.G., et al. 2017 AHA/ACC/HRS Guideline for Management of Patients WithVentricular Arrhythmias and the Prevention of Sudden Cardiac Death. A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. Circulation, 138(13), 2018.

instead of contracting (beating) normally. This prohibits the heart from pumping blood, causing collapse and cardiac arrest. This type of arrhythmia is a life-threatening medical emergency.
Ventricular tachyarrhythmia is a term designated to the types of tachycardias, or fast heart rhythms, that originate from the lower ventricles. These rhythm disturbances are varied and are most often linked to structural heart disease with or without coronary artery disease.
Ventricular tachycardia (VT) is a rhythm disorder caused by abnormal electrical signals in the ventricles of the heart.

- **Monomorphic ventricular tachycardia** is ventricular tachycardia with stable QRS morphology.
- **Non-sustained ventricular tachycardia (NSVT)** is defined as 3 or more consecutive beats originating from the ventricle, lasting less than 30 seconds, at a rate more than 100 beats per minute (bpm).
- **Polymorphic ventricular tachycardia** is a ventricular rhythm, with a rate greater than 100 bpm with a varying QRS pattern that terminates spontaneously (causing syncope if lasting more than a few seconds) or will deteriorate into ventricular fibrillation, causing cardiac arrest.
- **Sustained ventricular tachycardia (SVT)** is defined as a ventricular rhythm more than 100 bpm (widened QRS complex with duration greater than 120 ms) lasting more than 30 seconds or requiring termination due to hemodynamic instability.

Wearable cardioverter defibrillator (WCD) is a treatment option when the risk of sudden cardiac death (SCD) is high. Unlike an implantable cardioverter defibrillator (ICD), a WCD is worn outside the body rather than implanted in the chest. A WCD is designed to continuously monitor cardiac rhythm, detect life-threatening rapid heart rhythms and automatically deliver a treatment shock to restore normal heart rhythm.

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Disclaimer section

Purpose

The purpose of the HealthHelp's clinical guidelines is to assist healthcare professionals in selecting the medical service that may be appropriate and supported by evidence to safely improve outcomes. Medical information is constantly evolving, and HealthHelp reserves the right to review and update these clinical guidelines periodically. HealthHelp reserves the right to include in these guidelines the clinical indications as appropriate for the organization's program objectives. Therefore the guidelines are not a list of all the clinical indications for a stated procedure, and associated Procedure Code Tables may not represent all codes available for that state procedure or that are managed by a specific client-organization.

Clinician Review

These clinical guidelines neither preempt clinical judgment of trained professionals nor advise anyone on how to practice medicine. Healthcare professionals using these clinical guidelines are responsible for all clinical decisions based on their assessment. All Clinical Reviewers are instructed to apply clinical indications based on individual patient assessment and documentation, within the scope of their clinical license.

Payment

The use of these clinical guidelines does not provide authorization, certification, explanation of benefits, or guarantee of payment; nor do the guidelines substitute for, or constitute, medical advice. Federal and State law, as well as member benefit contract language (including definitions and specific contract provisions/exclusions) take precedence over clinical guidelines and must be considered first when determining eligibility for coverage. All final determinations on coverage and payment are the responsibility of the health plan. Nothing contained within this document can be interpreted to mean otherwise.

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National and Local Coverage Determination (NCD and LCD)

NOTICE
To ensure appropriate review occurs to the most current NCD and/or LCD, always defer to https://www.cms.gov/medicare-coverage-database/search.aspx.

Background

National Coverage Determinations (NCD) and Local Coverage Determinations (LCD) are payment policy documents outlined by the Centers for Medicare and Medicaid Services (CMS) and the government's delegated Medicare Audit Contractors (MACs) that operate regionally in jurisdictions.
CMS introduced variation between different jurisdictions/Medicare Audit Contractors (MACs) and their associated covered code lists with the transition to ICD 10. The variation resulted in jurisdictions independently defining how codes are applied for exclusions, limitations, groupings, ranges, etc. for the medical necessity indications outlined in the NCD and LCD. Due to this variation, there is an inconsistent use/application of codes and coverage determinations across the United States between the different MACs.
In addition, **WITHOUT** notice, CMS can change the codes that indicate medical necessity and the format of the coverage determinations/associated documents (eg, Articles). This is an additional challenge for organizations to keep up with ongoing, unplanned changes in covered codes and medical necessity indications.

Medical Necessity Codes

Due to the variation in code application between jurisdictions/MACs and that updates can happen without notification, HealthHelp is not able to guarantee full accuracy of the codes listed for any Coverage Determination, and advises that prior to use, the associated Coverage Determination Articles are reviewed to ensure applicability to HealthHelp's programs and any associated NCDs and LCDs.

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