HEALTHHELP®

A WNS COMPANY

2025 Defibrillator

Cardiology

CARD-CDEF-HH

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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*.

Preamble: Pediatric Cardiology Preamble

HealthHelp's clinical guidelines for the Cardiology program, are intended to apply to both adults and pediatrics (21 years of age or younger), unless otherwise specified within the criteria.

CRT-D Insertion or Replacement Guideline

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

1. Heart failure is symptomatic (eg, dyspnea, exercise intolerance, fatigue), candidate for implantable cardioverter-defibrillator (ICD) and **ANY** of the following: [7]
    a. Atrial fibrillation is persistent or permanent, left ventricular ejection fraction (LVEF) is 40% or less, heart rate is more than 100 bpm **AND** is a candidate for atrioventricular junction ablation.
    b. Left bundle branch block morphology, despite optimal medical treatment (eg, beta blockers, angiotensin-converting enzyme (ACE) inhibitors, angiotensin receptor blockers, mineral corticoid receptor blockers) and **ALL** of the following:
        i. Left ventricular ejection fraction (LVEF) is 35% or less.
        ii. QRS duration is 130 ms or more.
        iii. Sinus rhythm
    c. Non-left bundle branch block morphology, despite optimal medical treatment (eg, beta blockers, angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, mineral corticoid receptor blockers) and **ALL** of the following:
        i. Left ventricular ejection fraction (LVEF) is 35% or less.
        ii. QRS duration is 150 ms or more.

iii. Sinus rhythm

Reference: [6]

2. Congenital heart disease is known, in a pediatric individual and **ALL** of the following:
   a. Left bundle branch block (LBBB)
   b. LVEF is less than 35%.
   c. New York Heart Association (NYHA) class II to IV
   d. QRS duration is longer than upper limit of normal for age.

   References: [6] [8]

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: [5]
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: [5] [10]
3. Lead fracture or malfunction
   References: [5] [10]
4. National recall of device
   Reference: [5]

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: [10] [2]

2. Infection is related to implant or lead.
   References: [10] [2]
3. Lead fracture, migration or malfunction
   References: [10] [2]

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 **ANY** of the following:

1. Atrial arrhythmias are paroxysmal.
   Reference: [12]
2. Atrioventricular block or sinus node dysfunction is known and symptomatic (eg, dizziness, exercise intolerance, fatigue). []
   Reference: [6]
3. Chronotropic incompetence (eg, failure or delay in reaching heart rate targets during exertion, heart rate instability during sustained exercise, inadequate submaximal heart rate during daily activities) and symptomatic (eg, dizziness, dyspnea, exercise intolerance).
   Reference: [6]
4. Conventional pacemaker or implantable cardioverter defibrillator (ICD) was placed, LVEF is 35% or less despite guideline-directed medical treatment (GDMT) **AND** right ventricle (RV) pacing is more than 20%.
5. Long QT syndrome (QRS duration 120 msec or more)
   Reference: [12]
6. Pacemaker syndrome is suspected.
   Reference: [6]
7. Syncope is recurrent, severe and unpredictable, age is over 40 years old and **ANY** of the following:
   a. Asystolic pauses are more than 3 seconds **AND** spontaneous.
   b. Asystolic pauses are more than 6 seconds **AND** cause is atrioventricular block or sinus arrest.

   Reference: [6]

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*.

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*.

Preamble: Pediatric Cardiology Preamble

HealthHelp's clinical guidelines for the Cardiology program, are intended to apply to both adults and pediatrics (21 years of age or younger), unless otherwise specified within the criteria.

Implantable Cardioverter Defibrillator (ICD) Guideline

A single chamber, dual chamber or subcutaneous implantable cardiac defibrillator (ICD) is considered medically appropriate when the documentation demonstrates **ANY** of the following: (*__NOTE__: *Procedure is preferable in a pregnant individual after the 1st trimester*.)

1. Arrhythmogenic right ventricular cardiomyopathy is known, life expectancy is more than 1 year and **ANY** of the following:
    a. Sudden cardiac arrest (SCA) history
    b. Syncope **AND** ventricular arrhythmia is suspected cause.
    c. Ventricular dysfunction is significant (right ventricular ejection fraction [RVEF] or left ventricular ejection fraction [LVEF] is 35% or less).
    d. Ventricular tachycardia (VT) is sustained (Persists for at least 30 seconds or is hemodynamically unstable).

    References: [1] [5] [14]
2. Brugada syndrome is known, electrocardiogram (ECG) demonstrates a spontaneous type 1 Brugada pattern, life expectancy is more than 1 year and **ANY** of the following:

a. Sudden cardiac arrest history

b. Syncope history and ventricular arrhythmia is suspected.

c. Ventricular arrhythmia is sustained.

References: [1] [5] [14]

3. Cardiac sarcoidosis is known, life expectancy is more than 1 year and **ANY** of the following:

   a. LVEF is 35% or less.

   b. LVEF is more than 35% and **ANY** of the following:

      i. Myocardial scar is suspected based on magnetic resonance imaging (MRI) or positron emission tomography (PET) of the heart

      ii. Permanent pacemaker is indicated.

      iii. Syncope

   c. Sudden cardiac arrest history

   d. VT is sustained (Persists for at least 30 seconds or is hemodynamically unstable).

   References: [1] [5] [14]

4. Cardiac transplant or left ventricular assist device (LVAD) is pending (awaiting in outpatient setting).
   References: [1] [5] [14]

5. Catecholaminergic polymorphic ventricular tachycardia (CPVT) and **EITHER** of the following:

   a. Cardiac arrest or syncope of arrhythmic origin is known, in a pediatric individual, **DESPITE** beta blocker use plus flecainide (Class IC antiarrhythmic) and/or history of cardiac sympathetic denervation.

   b. Recurrent, sustained VT or syncope is known **AND** beta blocker is adequate or maximally tolerated.

   References: [1] [5] [14] [13]

6. Chagas disease is known in a pediatric individual, with advanced 2nd or 3rd degree AV block **AND** spontaneous resolution (eg, electrolyte imbalances, increased vagal tone, infection, medication effects) is **NOT** likely.
   References: [1] [5] [14] [13]

7. Congenital heart disease (CHD) is known and **ANY** of the following:,

a. Complexity is moderate or severe (aortic coarctation, aortic stenosis, Ebstein anomaly of the tricuspid valve), is repaired, syncope with **NO** known etiology and **EITHER** of the following;
   i. Ventricular dysfunction is moderate (LVEF is 50% or less).
   ii. Ventricular hypertrophy is marked (wall thickness of 15 mm or more).
b. Sudden cardiac arrest history where VF or VT are suspected **AND NO** etiology or reversible causes are determined on evaluation.
c. Tetralogy of Fallot is repaired and **EITHER** of the following:
   i. VT or VF are inducible.
   ii. VT is spontaneous and sustained (Persists for at least 30 seconds or is hemodynamically unstable).
d. VT is hemodynamically unstable.
e. VT is sustained (Persists for at least 30 seconds or is hemodynamically unstable) or syncope due to arrhythmogenic syncope **AND** LVEF is less than 35%, in a pediatric individual.

References: [1] [5] [14] [13]

8. Coronary artery spasms with sudden cardiac arrest history and multiple (more than 1) vasodilator medications are attempted **WITHOUT** relief.
   References: [1] [5] [14]
9. Early repolarization pattern or short QT syndrome is demonstrated on ECG, cardiac arrest history or sustained ventricular arrhythmia is known.
   References: [1] [5] [14]
10. Emery-Dreifuss or limb-girdle type IB muscular dystrophy is known **AND** cardiac involvement is progressive.
    References: [1] [5] [14]
11. Hypertrophic cardiomyopathy (HCM) is known, life expectancy is more than 1 year and **ANY** of the following:
    a. Left ventricular (LV) wall thickness is 30 mm or more.
    b. Non-sustained VT (NSVT) is spontaneous or exercise produces an abnormal blood pressure response (*See **Definitions** section*) and **ANY** of the following:
       i. NSVT is 3 beats or more.
       ii. Sudden cardiac arrest family history
    c. Sudden cardiac death in 1st degree relative (child, parent, sibling) thought to be caused by HCM

d. Syncope is known with 1 or more episodes in last 6 months.

e. VF or VT sudden cardiac arrest history

f. VT is spontaneous and sustained **AND** causes hemodynamic instability or syncope.

References: [1] [5] [14] [11]

12. Long QT syndrome (QRS duration 120 msec or more) and **EITHER** of the following:

    a. Sudden cardiac arrest history, in a pediatric individual.

    b. Syncope or ventricular tachycardia is known **DESPITE** treatment with a beta-blocker.

    References: [1] [5] [14] [13]

13. Sudden cardiac death prevention, life expectancy is more than 1 year and **EITHER** of the following:

    a. Primary prevention of SCD, with ischemic heart disease and **ANY** of the following:

        i. Myocardial infarction (MI) is 40 days ago or more, and **ANY** of the following:

            A. HF NYHA class I, **DESPITE** GDMT **AND** left ventricular ejection fraction (LVEF) is 30% or less.

            B. HF NYHA class II or III, **DESPITE** GDMT **AND** LVEF is 35% or less.

        ii. Non-ischemic cardiomyopathy (NICM) or neuromuscular disorder (eg, Becker dystrophy, Duchenne dystrophy, myotonic dystrophy) is known and **ANY** of the following:

            A. HF NYHA class II to III and LVEF is 35% or less, **DESPITE** guideline-directed medical therapy (GDMT).

            B. *Lamin A/C* mutation is known and **ANY 2** of the following:

                I. LVEF is less than 45%.

                II. Male

                III. Non-missense mutation

                IV. Non-sustained ventricular tachycardia (VT)

        iii. Non-sustained ventricular tachycardia (VT) is caused by prior MI and **ALL** of the following:

            A. Electrophysiology study (EPS) demonstrates inducible, sustained VT or ventricular fibrillation.

B. LVEF is 40% or less.

iv. Revascularization (eg, coronary artery bypass grafting [CABG], percutaneous coronary intervention [PCI]) is 90 days ago or more and **ANY** of the following:

A. HF NYHA class I, **DESPITE** GDMT **AND** LVEF is 30% or less.

B. HF NYHA class II or III, **DESPITE** GDMT **AND** LVEF is 35% or less.

b. Secondary prevention of SCD and **ANY** of the following:

i. Ischemic heart disease is known and **ANY** of the following:

A. **NO** reversible causes ventricular fibrillation (VF) or ventricular tachycardia (VT) (eg, anemia, coronary vasospasm, infection, transient thrombosis)

B. Syncope is unexplained **AND** electrophysiology study demonstrates inducible monomorphic VT.

ii. NICM or neuromuscular disorder (eg, Becker dystrophy, Duchenne dystrophy, myotonic dystrophy) is known and **ANY** of the following:

A. **NO** reversible causes ventricular fibrillation (VF) or ventricular tachycardia (VT) (eg, anemia, coronary vasospasm, infection, transient thrombosis)

B. Syncope is known and **ALL** of the following:

I. Do **NOT** meet indications for primary prevention ICD

II. Ventricular arrhythmia is suspected cause.

References: [1] [5] [14]

14. Tachycardia/bradycardia (Tachy-brady) syndrome is known, in a pediatric individual.
References: [1] [5] [14] [13]

15. VF or VT is idiopathic and polymorphic, with a resuscitated sudden cardiac arrest history.
References: [1] [5] [14]

Subcutaneous Implantable Cardiac Defibrillator (SICD) Guideline

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

1. Pacing is **NOT** required (eg, bradycardia, cardiac resynchronization, VT).
   References: [1] [9]
2. Vascular access is inadequate, infection risk is high (eg, immunocompromised) or simple lead extraction is anticipated **AND** meets appropriate indication for placement of ICD.
   References:[1] [9]

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: [5]
2. End-of-life span as advised by device manufacturer
   Reference: [5]

Wearable Cardiac Defibrillator (WCD)

Preamble: Pediatric Cardiology Preamble

HealthHelp's clinical guidelines for the Cardiology program, are intended to apply to both adults and pediatrics (21 years of age or younger), unless otherwise specified within the criteria.

Wearable Cardiac Defibrillator (WCD) Guideline

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

1. Definitive procedure is planned (eg, cardiac transplant, ICD implant) in the next 60 days **AND** ventricular arrhythmia risk is elevated (eg, electrolyte imbalance, ischemia, Lamin A/C variant, structural heart disease).
   References: [1] [3] [14] [4]
2. **ANY** of the following:
   a. Secondary prevention of sudden cardiac death (SCD) and **ANY** of the following:
      i. Ischemic heart disease is known and **ANY** of the following:
         A. **NO** reversible causes of ventricular fibrillation (VF) or ventricular tachycardia (VT) (eg, anemia, coronary vasospasm, infection, transient thrombosis)
         B. Syncope is unexplained **AND** electrophysiology study demonstrates inducible monomorphic VT.
      ii. Non-ischemic cardiomyopathy (NICM) or neuromuscular disorder (eg, Becker dystrophy, Duchenne dystrophy, myotonic dystrophy) is known and **ANY** of the following:
         A. **NO** reversible causes of ventricular fibrillation (VF) or ventricular tachycardia (VT) (eg, anemia, coronary vasospasm, infection, transient thrombosis)
         B. Syncope is known and **ALL** of the following:
            I. Do **NOT** meet indications for primary prevention ICD
            II. Ventricular arrhythmia is suspected cause.
   b. Sudden cardiac arrest history or ventricular arrhythmia is sustained **AND** removal of ICD is required (eg, infection).
      References: [1] [3] [14] [4]

Defibrillator Summary of Changes

The Defibrillator guidelines from 2024 to 2025 had the following version changes:

- Added the following to keep in line with current evidence:
  - Cardiac Resynchronization Therapy-Defibrillator
    - Indications under "Heart failure"
    - "Congenital heart disease" indication
  - CRT-D Upgrade from Single Chamber to Dual Chamber Device with Biventricular Pacing Capability

        - "Atrial arrhythmias" indication
        - "Pacemaker syndrome" indication
    - Implantable Cardioverter Defibrillator
        - "Catecholaminergic polymorphic ventricular tachycardia (CPVT)" indication
        - "Congenital heart disease" indication
        - "Coronary artery spasms" indication
        - "Early repolarization pattern" indication
        - "Emery-Dreifuss" indication
        - Indications under "Brugada syndrome"
        - "Sudden cardiac death" indication
    - Wearable Cardiac Defibrillator
        - "Definitive procedure" indication
        - "Secondary prevention of sudden cardiac death (SCD)" indication
- Removed the following as evidence does not support the indication:
    - CRT-D Upgrade from Single Chamber to Dual Chamber Device with Biventricular Pacing Capability
        - "Condition requires permanent dual chamber pacing (eg, chronotropic incompetence)." indication
    - Implantable Cardioverter Defibrillator (ICD)
        - "Anti-tachycardia pacing" indication
    - Subcutaneous Implantable Cardiac Defibrillator (SICD)
        - Indications as the guideline aligns with ICD
    - Wearable Cardiac Defibrillator
        - "Clinical condition includes" indication
        - "**NO contraindications** for WCD" indication
- Mid-cycle update: 4/17/2025
    - Changed verbiage from "maximally tolerated beta blocker" to "beta blocker use" from the Catecholaminergic polymorphic ventricular tachycardia (CPVT) indication.

- Removed "adherence is demonstrated with medical treatments (eg, appointment maintenance, medication regimen)" from under "Wearable cardiac defibrillator" as it is not a necessary indication for the approval of the guideline

Defibrillator Definitions

Abnormal blood pressure response during exercise includes a hypertensive response (systolic BP ≥210 mm Hg in men and ≥190 mm Hg in women), a diastolic BP increase >10 mm Hg or ≥90 mm Hg, a failure to increase systolic BP by at least 20 mm Hg, or a fall in systolic BP below baseline or by more than 20 mm Hg from peak exercise.
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.
Atrial arrhythmias are irregular heartbeats that occur when the upper chambers of the heart (atria) beat out of rhythm with the lower chambers (ventricles). This can cause the heart to beat too fast, too slow, or in an irregular way. Atrial arrhythmias can be harmless or life-threatening.
Atrial fibrillation (AF) is a cardiac rhythm disorder characterized by uncontrolled atrial activation without effective atrial contraction. On the electrocardiogram (ECG), P waves are absent. AF is characterized by rapid oscillations or fibrillatory waves that vary in amplitude, shape and timing associated with an irregular ventricular response.

- **Paroxysmal AF** terminates spontaneously or with intervention within 7 days of onset. Episodes typically convert back to sinus rhythm within 48 hours.
- **Persistent AF** is continuous AF sustained beyond 7 days.

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.
Atrioventricular (AV) junctional ablation is a medical procedure that uses radiofrequency energy to destroy the atrioventricular (AV) node, the area of tissue that connects the upper and lower chambers of the heart. The procedure is used to treat atrial fibrillation (AFib) by blocking the faulty electrical signals that cause it.
Becker muscular dystrophy is a genetic disorder that gradually makes the body's muscles weaker and smaller.
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.

Biventricular pacing device, also known as a cardiac resynchronization therapy (CRT) device, is a type of pacemaker that coordinates the contractions of the heart's left and right ventricles. It's used to treat heart failure and improve the quality of life for patients with severe symptoms that aren't controlled by medication.

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.

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare genetic heart condition that causes an irregular heart rhythm, or arrhythmia, that can lead to sudden cardiac death. CPVT is caused by mutations in genes that control calcium levels in the heart muscle cells, which can lead to irregular electrical signals.

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.

Channelopathies are genetically determined disorders characterized by abnormal function of ion channels, leading to various clinical manifestations including muscle, cardiac, and neurological disorders.

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

Congenital heart disease (CHD) is a term for a variety of birth defects that affect heart anatomy and function. Congenital is defined as present since birth. CHD occurs when the heart, or blood vessels near the heart, do not develop normally. Common heart defects include: atrial septal defect, coarctation of the aorta, d-transposition of the great arteries, Ebstein's anomoly, patent ductus arteriosus, tetralogy of fallot, total anomalous pulmonary venous connection and ventricular septal defect.

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 artery spasm is a brief, sudden narrowing of one or more of the arteries that supply blood to the heart. This causes a temporary reduction in blood flow to the heart, which can lead to chest pain, also known as angina.

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.

Duchenne muscular dystrophy is a severe progressive X-linked muscular dystrophy of males marked by early childhood onset and absence of the protein dystrophin.

Dyspnea is difficult, painful breathing or shortness of breath.

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%.

Emery-Dreifuss muscular dystrophy is a condition that primarily affects muscles used for movement (skeletal muscles) and the heart (cardiac muscle). Among the earliest features of this disorder are joint deformities called contractures.

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.

Guideline directed medical therapy (GDMT) for heart failure are specific treatments (eg, medications, implantable cardiac defibrillators [ICD]) used by health care providers as a standardized treatment for heart failure.

These medications include:

- Angiotensin converting enzyme inhibitors (ACE-I) (eg, enalapril, lisinopril)
- Angiotensin receptor blockers (ARB) (eg, losartan, olmesartan)
- Angiotensin receptor-Neprilysin inhibitors (ARNI) (eg, Entresto)
- Aldosterone antagonists (eg, eplerenone, spirololactone)
- Beta blockers (eg, atenolol, metoprolol)
- Hydralazine/isosorbide dinitrate (HYD/ISDN)
- Hyperpolarization activated cyclic nucleotide-gated (HCN) channel blockers (eg, Ivabradine).
- Loop diuretics (eg, hydrochlorothiazide, metolazone)

Heart block also called atrioventricular (AV) block, is partial or complete interruption of impulse transmission from the atria to the ventricles. [1]

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.

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.

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

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.

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. [2,3]

Inducible monomorphic ventricular tachycardia (MVT) is a fast, abnormal heart rhythm originating in the ventricles where the heart beats at a rate of 100 or more per minute, with each beat having the same shape (monomorphic) and can be induced during an electrocardiogram (ECG). It's characterized by wide complex QRS waves and can be dangerous if not treated, potentially leading to ventricular fibrillation and cardiac arrest.

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

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

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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.
Lead fracture is a break in the conductor coil within the lead of a pacemaker, and can lead to pacemaker failure. Symptoms can include dizziness, syncope, chest discomfort, palpitations or less commonly with extracardiac symptoms.
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 ejection fraction (LVEF), also known as ejection fraction (EF), is defined as the percentage of blood ejected from the left ventricle during each contraction.
Left ventricular hypertrophy (LVH) is an increase in the thickness of the left ventricular wall, often due to chronic pressure or volume overload, and can lead to heart failure.
Limb-girdle muscular dystrophy (LGMD) is a group of genetic diseases that cause muscle weakness and wasting in the arms and legs. The muscles closest to the body, like the shoulders, upper arms, pelvic area, and thighs, are usually the first to be affected.
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.
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.
Neuromuscular disorder is a wide-range of diseases affecting the peripheral nervous system, which consists of all the motor and sensory nerves that connect the brain and spinal cord to the rest of the body.

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	SYMPTOMS EXPERIENCED
Class II (Mild)	Mild symptoms (eg, mild shortness of breath and/or angina) and slight limitation during ordinary activity.
Class III (Moderate)	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 without discomfort.

Myotonic dystrophy is a form of muscular dystrophy that affects muscles and many other organs in the body.

Normal sinus rhythm (NSR) is the rhythm that originates from the sinus node (the heart's natural pacemaker, which is located in the superior right atrium) and describes the characteristic rhythm of the healthy human heart.

Pacemaker is a small, battery-powered device that's surgically implanted in the chest or abdomen to monitor and regulate the heart's rhythm and rate. A pacemaker sends electrical impulses to the heart to help it beat at a normal rhythm and rate. The device has long, thin wires that connect it to the heart through a large vein.

Paroxysmal is a sudden attack or increase of symptoms of a disease (such as pain, coughing, shaking, etc.) that often occurs again and again.

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.

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).

QRS durations by age are as follows:

- Adults (age is over 16 years old): QRS interval is up to 110 ms.
- Age is 4 years to 16 years: QRS interval is up to 100 ms.
- Age is less than 4 years old: QRS interval is up to 90 ms.

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.

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). [4]

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.

Tachycardia-bradycardia syndrome occurs when the heart fluctuates between beating too quickly (tachycardia) and too slowly (bradycardia).

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.

Ventricular arrhythmia is an abnormal heart rhythm that makes the lower chambers of the heart twitch instead of pump, which can limit or stop your heart from supplying blood to your body.

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, 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 hypertrophy (VH) is a condition in which the walls of the heart's lower chambers, or ventricles, thicken. The most common type of VH is left ventricular hypertrophy (LVH), which affects the heart's main pumping chamber, the left ventricle.

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).

[4]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.

- **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.

For Internal Use Only:

11248 11249 11253 11282 11325 11328 11333 11349 11350 11351 11352 11354 11355 11356 11358 11359 11360 11361 11362 11365 11366 11367 11368 11369 11370 11374 11375 11394 11395 11396 11565