Cardiomyopathy
ICD-10-CM
Clinical overview

Definition
Cardiomyopathy is a disease of the heart muscle that impairs the function of the heart.

Types
Cardiomyopathy can be classified as primary or secondary and ischemic or nonischemic.

- **Primary cardiomyopathy** is a noninflammatory disease of the heart muscle, often of obscure or unknown cause, that occurs in the absence of other cardiac conditions or systemic disease processes.

- **Secondary cardiomyopathy** is caused by a known medical condition (such as hypertension, valve disease, congenital heart disease or coronary artery disease).

- **Ischemic cardiomyopathy** is caused by coronary artery disease and heart attacks, which result in lack of blood flow to the heart muscle, thereby causing damage to the heart muscle.

- **Nonischemic cardiomyopathy** is a type of cardiomyopathy not related to coronary artery disease or poor coronary artery blood flow. There are three main types of nonischemic cardiomyopathy:

  - **Dilated cardiomyopathy (also known as congestive cardiomyopathy)** – This is the most common type of cardiomyopathy. In this disorder, the heart's main pumping chamber – the left ventricle – becomes enlarged (dilated), its pumping ability becomes less forceful and blood doesn't flow as easily through the heart.

  - **Hypertrophic cardiomyopathy** – This type involves abnormal growth or thickening of the heart muscle, particularly affecting the muscle of the left ventricle. As thickening occurs, the heart tends to stiffen and the size of the pumping chamber may shrink, interfering with the heart's ability to deliver blood to the body.

  - **Restrictive cardiomyopathy** – The heart muscle in people with restrictive cardiomyopathy becomes rigid and less elastic, meaning the heart can't properly expand and fill with blood between heartbeats.

Some cardiomyopathies can be reversible. For example:
- Alcoholic cardiomyopathy sometimes can be reversed with complete cessation of alcohol intake. Takotsubo cardiomyopathy is a reversible, stress-induced cardiomyopathy.

Causes
The cause is usually unknown (primary cardiomyopathy), although contributing factors sometimes can be identified. Some of the possible known causes include:
- Long-term high blood pressure
- Coronary artery disease
- Heart valve problems
- Chronic rapid heart rate
- Certain viral infections
- Some chemotherapy drugs
- Pregnancy
- Excessive, long-term use of alcohol
- Heart damage, due to a previous heart attack
- Metabolic disorders (thyroid disease, diabetes, etc.)
- Nutritional deficiencies of essential vitamins and minerals
- Abuse of cocaine or antidepressant medications
- Hemochromatosis – disorder in which iron is not properly metabolized, causing build-up in various organs, including heart muscle (This can lead to a weakening of the heart muscle, resulting in dilated cardiomyopathy.)

Signs and symptoms
There may be no signs or symptoms in the early stages of the disease. But as the condition advances, signs and symptoms usually appear and may include:
- Shortness of breath, especially with physical exertion
- Swelling of lower extremities, abdomen and neck veins
- Fatigue
- Chest pain
- Irregular heartbeats
- Heart murmurs
- Dizziness and lightheadedness
- Fainting

Possible complications
- Heart failure
- Blood clots
- Heart valve problems with associated murmurs
- Cardiac arrest and sudden death

Diagnostic tools

- Medical history and physical exam
- Blood tests, such as measurement of brain natriuretic peptide (BNP) – a protein produced in the heart for which the blood level increases when the heart is under stress from heart failure, a common complication of cardiomyopathy
- Chest X-ray (to check for signs such as enlarged heart or fluid buildup in lungs)
- Electrocardiogram (ECG or EKG)
- Holter monitoring and cardiac event recording
- Echocardiogram
- Cardiac stress testing
- Cardiac magnetic resonance imaging (MRI)
- Cardiac catheterization and heart biopsy

Treatment

- Lifestyle changes:
    - Heart-healthy diet
    - Weight control
    - Stress management
    - Physical activity and exercise
    - Smoking cessation
- Medications:
    - Blood thinners to prevent clots
    - Antiarrhythmics to control heart rate and rhythm
    - Antihypertensives for blood pressure control
    - Diuretics ("water pills") to remove excess sodium and reduce excess fluid in the blood
- Nonsurgical procedure:
    - Alcohol septal ablation, in which a type of alcohol (ethanol) is injected through a tube into the small artery that supplies blood to the thickened area of heart muscle. The alcohol shrinks the thickened heart tissue to a more normal size, allowing blood to flow freely through the ventricle of the heart, which results in improved symptoms.
- Cardiac device implantation:
    - Pacemaker
    - Cardioverter-defibrillator
    - Left ventricular assist device
- Surgical procedures:
    - Heart transplant (a last resort for severe, end-stage cardiomyopathy that cannot be controlled by other means)

LC5407ALL0520-D

Cardiomyopathy
ICD-10-CM
Best documentation practices for physicians

Subjective
- The subjective section of the office note should document current related patient complaints and symptoms. If there are none, the office note should show the patient was screened for current related complaints or symptoms.

Objective
- In the objective section, include any current associated physical exam findings (such as edema/swelling of the lower extremities, abdomen or neck veins) and related diagnostic testing results.

Assessment
Specificity:
- The term "cardiomyopathy" is broad and nonspecific. It is important to describe the particular type of cardiomyopathy to the highest level of specificity.
- Document the current status of cardiomyopathy (stable, improved, worsening, etc.).

Abbreviations:
- A good rule of thumb for any medical record is to limit – or avoid altogether – the use of abbreviations. There are several commonly used medical abbreviations for different types of cardiomyopathy (CM, CMP, HCM, HOCM, etc.), but some of these abbreviations have other meanings. The meaning of an abbreviation can often be determined based on context, but this is not always true.
- Best practice is to always document the specific type of cardiomyopathy by spelling it out in full.

Associated conditions:
- Clearly link secondary cardiomyopathy to the underlying causative condition by using terms such as "due to," "secondary to," "associated with," "related to," etc.

Current versus historical/transient:
- Do not use the descriptor "history of" to describe current cardiomyopathy. In diagnosis coding, "history of" means the condition occurred in the past and is no longer a current problem.
- Temporary or transient cardiomyopathy that occurred in the past and is no longer present should not be documented as if it is current.

Plan
- Document a specific and concise treatment plan.
- Clearly link the cardiomyopathy diagnosis to any medications being used to treat the condition.
- Document referrals to specialists or other providers.
- Include the date of the patient's next appointment.

Electronic medical record (EMR) issues
Some electronic medical records insert ICD-10-CM code descriptions into the medical record to represent the final diagnosis.
- For example: "I42.8, Other cardiomyopathies."
- This diagnosis description by itself is vague and incomplete. The medical record should clearly describe the particular "other" type of cardiomyopathy.

Per the ICD-10-CM Official Guidelines for Coding and Reporting:
- Alphabetic Index entries in the ICD-10 coding manual with NEC (not elsewhere classified) designation represent "Other" codes in the Tabular List.
- These Alphabetic Index entries represent specific disease entities for which no specific code exists so the term is included within an "other" code.
- Codes from the Tabular List titled Other" or "Other specified" are for use when the information in the medical record provides a specific description of the condition but for which a specific code does not exist.
- The "Other" code with description should not be used, by itself, as a final diagnosis without clear documentation of the actual condition that describes the particular "other" condition.

Another scenario that causes confusion is one in which the assessment section documents a provider-stated diagnosis *PLUS* an EMR-inserted diagnosis code with description that does not match – or may even contradict – the stated diagnosis. Example:

Assessment: Cardiomyopathy, unspecified
I42.Ø Dilated cardiomyopathy

In this scenario, the provider's final diagnostic statement in bold is Cardiomyopathy, unspecified, which codes to I42.9. The EMR-inserted diagnosis code with description – I42.Ø Dilated cardiomyopathy – does not match the stated diagnosis. This leads to confusion regarding which diagnostic statement is correct. To avoid confusion and

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ensure accurate diagnosis code assignment, the provider's stated final diagnosis must either

a) match the code with description; OR
b) it must classify in ICD-10-CM to the EMR-inserted diagnosis code with description.

Note: ICD-10-CM is a statistical classification; it is not a substitute for a healthcare provider's final diagnostic statement. It is the healthcare provider's responsibility to provider legible, clear, concise and specific documentation of a final diagnosis described to the highest level of specificity, which is then translated to a code for reporting purposes. It is not appropriate for healthcare providers to simply list a code number or select a code number from a list of codes in place of a written final diagnosis.

Reference: AHA Coding Clinic, Code Number in Lieu of a Diagnosis, Fourth quarter 2015, pages 34-35

LC5407ALL0520-D

Cardiomyopathy
ICD-10-CM
Tips and resources for coders

Coding basics

- ICD-10-CM code assignment is based on the specific diagnosis description and details documented in the individual medical record. To ensure accurate and specific diagnosis code assignment, the coder must review the entire medical record and then, in accordance with ICD-10-CM official coding conventions and guidelines:
- Search the alphabetic index for that specific description and the corresponding code.
- Verify the code in the tabular list, carefully following all instructional notes.

Coding cardiomyopathy

Many of the most common cardiomyopathies classify to category I42, Cardiomyopathy.

- A fourth character is required to specify the particular type of cardiomyopathy.
- The broad and nonspecific final diagnosis of "cardiomyopathy" leads to the broad and nonspecific diagnosis code I42.9, Cardiomyopathy, unspecified.
- Code I42.9 should be assigned only when no information in the medical record identifies the particular type of cardiomyopathy.

Hypertensive cardiomyopathy classifies to category I11, Hypertensive heart disease, with an additional code of I43, Cardiomyopathy in diseases classified elsewhere.

Congestive cardiomyopathy is also known as dilated cardiomyopathy. Both of these descriptions classify to code I42.Ø, Dilated cardiomyopathy.

- Congestive cardiomyopathy often is associated with congestive heart failure and basically has the same symptoms.
- Treatment typically focuses on management of the congestive heart failure; therefore, heart failure (category I5Ø.-) is reported as the principal diagnosis with an additional code for the cardiomyopathy.

Hypertrophic cardiomyopathy can be obstructive or nonobstructive.

- I42.1 Obstructive hypertrophic cardiomyopathy
- I42.2 Other hypertrophic cardiomyopathy
  Includes nonobstructive hypertrophic cardiomyopathy

Reminders

- Use caution when coding cardiomyopathy from abbreviations (CM, HCM, HOCM, etc.). A code should not be assigned unless the meaning of the abbreviation is clear based on review of the entire medical record.
- Watch for modifying descriptors that affect code assignment (secondary, alcoholic, nutritional, metabolic or cardiomyopathy due to other diseases).
- Some secondary cardiomyopathies are coded with a single combination code, while other secondary cardiomyopathies require the use of two codes. See coding examples on page 5.
- Takotsubo cardiomyopathy is a reversible form of cardiomyopathy that classifies to code I51.81, Takotsubo syndrome. This code includes the following conditions:
  - Reversible left ventricular dysfunction following sudden emotional stress
  - Stress-induced cardiomyopathy
  - Takotsubo cardiomyopathy
  - Transient left ventricular apical ballooning syndrome
- The term "ischemic cardiomyopathy" is sometimes used to refer to a condition in which ischemic heart disease causes diffuse fibrosis or multiple infarction, leading to heart failure with left ventricular dilation. This is not a true cardiomyopathy. When no further clarification is available, this condition is coded to I25.5, Ischemic cardiomyopathy.
- In ICD-10-CM, dilated cardiomyopathy and ischemic cardiomyopathy are classified to different codes. When a medical record documents a current diagnosis of ischemic dilated cardiomyopathy, both codes are needed to fully capture this condition:
  - I25.5 Ischemic cardiomyopathy
  - I42.Ø Dilated cardiomyopathy
- Non-ischemic cardiomyopathy with no other description and no mention of cause codes to I42.8.

ALPHABETIC INDEX
Cardiomyopathy (familial) (idiopathic)
non-ischemic – *see also* by cause I42.8

TABULAR LIST
I42.8 Other cardiomyopathies

Cardiomyopathy
ICD-10-CM
Tips and resources for coders

Coding examples

Example 1	
Final diagnosis	Cardiomyopathy (nonspecific)
ICD-10-CM code	I42.9 Cardiomyopathy, unspecified

Example 2	
Final diagnosis	Ischemic dilated cardiomyopathy
ICD-10-CM code(s)	I25.5 Ischemic cardiomyopathy I42.Ø Dilated cardiomyopathy

Example 3	
Final diagnosis	Hypertensive cardiomyopathy
ICD-10-CM codes(s)	I11.9 Hypertensive heart disease without heart failure I43 Cardiomyopathy in disease classified elsewhere

Example 4	
Final diagnosis	Cardiomyopathy due to sarcoidosis
ICD-10-CM code	D86.85 Sarcoid myocarditis

Example 5	
Final diagnosis	Metabolic cardiomyopathy
ICD-10-CM code(s)	E88.9 Metabolic disorder, unspecified I43 Cardiomyopathy in diseases classified elsewhere

Example 6	
Final diagnosis	Rheumatic cardiomyopathy
ICD-10-CM code	IØ9.Ø Rheumatic myocarditis

Example 7	
Final diagnosis	Stress-induced cardiomyopathy
ICD-10-CM code	I51.81 Takotsubo syndrome

Example 8	
Final diagnosis	Restrictive cardiomyopathy
ICD-10-CM code	I42.5 Other restrictive cardiomyopathy

Example 9	
Final diagnosis	End-stage dilated cardiomyopathy
ICD-10-CM code	I42.Ø Dilated cardiomyopathy

Example 10	
Final diagnosis	Alcoholism in remission with alcoholic cardiomyopathy
ICD-10-CM codes(s)	F1Ø.21 Alcohol dependence, in remission I42.6 Alcoholic cardiomyopathy

References: American Hospital Association Coding Clinic; ICD-10-CM Official Guidelines for Coding and Reporting; ICD-10-CM and ICD-10-PCS Coding Handbook; Mayo Clinic; National Heart, Lung, and Blood Institute

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