ICD-10-CM Documentation and Coding Best Practices
Cardiomyopathy

Cardiomyopathy refers to diseases of the heart muscle, which can become enlarged, thick or rigid. In rare cases, cardiac muscle tissue can be replaced with scar tissue. As the condition worsens, the heart becomes weaker and less able to pump blood through the body or to maintain a normal electrical rhythm.

Causes

Risk factors that can increase the possibility of developing cardiomyopathy include: coronary artery disease, a history of heart attack(s), viral infections that cause heart inflammation, long-term hypertension or alcoholism, obesity, and diabetes to name a few. However, in most cases the exact cause is usually unknown (called *primary* or *idiopathic cardiomyopathy*).

Symptoms

Some patients will never have symptoms; others will not develop them until later in the disease. Symptoms can include:

- Fatigue
- Dizziness, lightheadedness or fainting
- Shortness of breath or trouble breathing (dyspnea)
- Swelling in the ankles, feet, legs, abdomen and neck veins

Treatment

Treatment depends on the type of cardiomyopathy, the severity of symptoms, and the patient's age and overall health.

- Lifestyle changes can help manage condition(s) that may be causing cardiomyopathy. Recommendations include:
  - Consuming a heart healthy diet, engaging in physical activity, losing excess weight, giving up smoking, avoiding alcohol and illegal drugs, getting enough sleep, and reducing stress
- Medicines may be prescribed to:
  - Lower blood pressure (ACE inhibitors, angiotensin II receptor blockers, beta blockers, calcium channel blockers)
  - Slow the heart rate (beta blockers, calcium channel blockers, digoxin)
  - Prevent arrhythmias (antiarrhythmics)
  - Remove excess fluid and sodium (diuretics)
  - Prevent blood clots (anticoagulants)
- Alcohol septal ablation
- Surgery
  - Septal myectomy – option for severe cases of obstructive hypertrophic cardiomyopathy
  - Surgically implanted devices
    - Pacemaker
    - Left ventricular assist device (LVAD)
    - Cardiac resynchronization therapy (CRT) device
    - Implantable cardioverter defibrillator (ICD)
  - Heart transplant – last resort treatment when all other options have failed

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Types of Cardiomyopathy

- ***Dilated Cardiomyopathy*** (DCM) – The heart muscle stretches and becomes thinner, and the inner chambers of the heart enlarge (dilate). The heart muscle does not contract normally and does not pump blood very well.
- ***Hypertrophic Cardiomyopathy*** *(HCM)* – Heart muscle cells enlarge causing the walls of the ventricles (usually the left ventricle) to thicken. This may block the flow of blood out of the ventricle, a condition called obstructive hypertrophic cardiomyopathy.
- ***Restrictive Cardiomyopathy*** – The heart's ventricles become rigid because abnormal tissue, such as scar tissue, replaces normal heart muscle. The ventricles cannot relax and fill with blood normally, and blood flow through the heart is reduced.
- ***Arrhythmogenic Right Ventricular Dysplasia***–The muscle tissue in the right ventricle dies and is replaced with scar tissue. This disrupts the heart's electrical signals and causes arrhythmias.
- ***Ischemic Cardiomyopathy***–A lack of blood supply to the heart muscle caused by coronary artery disease and heart attacks results in diffuse fibrosis of heart muscle tissue, leading to heart failure with left ventricle dilation.
- ***Stress-induced Cardiomyopathy*** – Also known as ***takotsubo cardiomyopathy*** or ***broken heart syndrome***. A sudden surge of stress hormones causes one part of the heart to enlarge temporarily while the rest of the heart functions normally. Can lead to severe, short-term muscle failure. Stress-induced cardiomyopathy is reversible.

Cardiomyopathy codes include:

- I25.5 Ischemic cardiomyopathy
- I42.0 Dilated cardiomyopathy
- I42.1 Obstructive hypertrophic cardiomyopathy
- I42.2 Other hypertrophic cardiomyopathy
- I42.3 Endomyocardial (eosinophilic) disease
- I42.4 Endocardial fibroelastosis
- I42.5 Other restrictive cardiomyopathy
- I42.6 Alcoholic cardiomyopathy *(Code also: Alcoholism)*
- I42.7 Cardiomyopathy due to drug and external agent *(Code also: Poisoning due to drug or toxin, or adverse effect of drug)*
- I42.8 Other cardiomyopathies
- I42.9 Cardiomyopathy (primary)(secondary), unspecified
- I43 Cardiomyopathy in diseases classified elsewhere *(Code first: Underlying disease)*
- I51.81 Takotsubo syndrome

Documentation Guidance

When documenting cardiomyopathy, include the following:

- ✓ Type – dilated, congestive, etc.
- ✓ Cause – congenital, alcohol, etc.
- ✓ Disease status - stable, improved, etc.
- ✓ Treatment plan - medicines, lifestyle changes, etc.

Coding Guidance

- ***Dilated Ischemic Cardiomyopathy*** – For patients with "dilated ischemic cardiomyopathy" or documentation of both ischemic and dilated cardiomyopathy, code *I25.5, Ischemic cardiomyopathy*, is advised. Dilated cardiomyopathy is most commonly the result of ischemic cardiomyopathy; the underlying disease should be reported.
- ***Congestive Cardiomyopathy*** - Coding guidelines state that congestive cardiomyopathy, which can also be called "congestive dilated cardiomyopathy," should be reported with *I42.0, Dilated cardiomyopathy.*
- ***Hypertensive Cardiomyopathy*** - Hypertensive cardiomyopathy requires the assignment of two diagnosis codes:
  1) *I11.9, Hypertensive heart disease*
  2) *I43, Cardiomyopathy in diseases classified elsewhere*

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