AAVBC

American Academy of Value Based Care

Cardiomyopathy

Quick Reference Guide

2026

AAVBC Cardiomyopathy Quick Reference Guide

Table of Contents

1. CLINICAL SNAPSHOT

Definition: Cardiomyopathy is a general medical term for diseases of the heart muscle. The word literally translates to "heart muscle disease" (cardio = heart, myo = muscle, pathy = disease). In this condition, the heart muscle becomes enlarged, thickened, or rigid. As the disease progresses, the heart weakens and becomes less able to pump blood throughout the body, which can lead to heart failure, irregular heartbeats (arrhythmias), and valve problems.[1]

ICD-10 Codes:[2]

Code Range	Category	Common Examples
I42.0 – I42.2	Structural	Dilated (I42.0), Hypertrophic (I42.1, I42.2)
I42.3 – I42.5	Tissue-Based	Endomyocardial (I42.3), Restrictive (I42.5)
I42.6 – I42.7	External Causes	Alcoholic (I42.6), Drug-induced (I42.7)
I42.9	Unspecified	Used when the specific type is unknown (Note: Does not map to an HCC)
I43	Secondary	Resulting from other diseases (metabolic/infectious)
I25.5	Ischemic	Specifically caused by heart attacks or coronary artery disease

HCC/RAF V28:

Codes within the I42.- range for cardiomyopathy, including dilated (I42.0), hypertrophic (I42.1, I42.2), and restrictive (I42.4, I42.5) types, map to **HCC 227 with a RAF of 0.189.** This mapping also includes specific causes such as alcoholic cardiomyopathy (I42.6), endocardial fibroelastosis (I42.3), and other specified cardiomyopathies (I42.8). Unspecified cardiomyopathy (I42.9) does not map to an HCC.

Prevalence: Dilated Cardiomyopathy (DCM) is the most common form of the disease and a leading reason for heart transplants. It is estimated that between 0.6 million and 1.3 million Americans are living with DCM. Hypertrophic Cardiomyopathy (HCM) is the most commonly inherited heart disease, affecting approximately 1 in 500 adults (0.2%). Restrictive Cardiomyopathy is the rarest form of the disease in the U.S, accounting for less than 5% of all cardiomyopathy cases.[3,4]

Annual Cost Estimates (Per Member Per Year)[5,6]

Patient Status	Estimated Annual Cost	Primary Drivers
Symptomatic (Chronic Management)	$24,000 – $33,000	Specialist visits, Guideline-Directed Medical Therapy (GDMT), and monitoring
Symptomatic (with Hospitalization)	$35,000 – $60,000+	Emergency department visits and inpatient stays (avg. $15K–$21K per admission)

Patient Status	Estimated Annual Cost	Primary Drivers
Asymptomatic/Subclinical	$3,600 – $5,000	Routine imaging (Echos/MRIs) and preventive medication
Advanced Stage (HF Stage D)	$100,000+	Mechanical assist devices (LVADs) or transplant evaluation

Abbreviations: Echo, Echocardiogram; GDMT, Guideline-Directed Medical Therapy; HF, Heart Failure; LVAD, Left Ventricular Assist Device; MRI, Magnetic Resonance Imaging

2. RECOGNITION AND DIAGNOSIS

Medicare Screening/Diagnostic Workup[7,8]

Medicare coverage for a cardiomyopathy workup follows a "medical necessity" model rather than a broad screening model. While Medicare covers a Cardiovascular Screening (blood tests for lipids) every five years for all members, a diagnostic workup for cardiomyopathy is only covered under Part B when a patient exhibits symptoms or has a high-risk profile.

1. Medicare Diagnostic Coverage (Part B)

For 2026, the diagnostic pathway is covered at 80% of the Medicare-approved amount after you meet your Part B deductible. Key tests include:

- **Echocardiogram (TTE):** The gold standard for initial diagnosis. Medicare covers this when symptoms like chest pain, shortness of breath, or edema are present
- **Cardiac MRI (cMRI):** Covered to distinguish between types of cardiomyopathy (e.g., finding the "late gadolinium enhancement" seen in Hypertrophic Cardiomyopathy)
- **Right Heart Catheterization:** Covered specifically when the physician suspects cardiomyopathy or myocarditis and needs to measure internal heart pressures to guide treatment
- **Heart Failure Management:** If the workup confirms heart failure, Medicare covers the Ambulatory Specialty Model (ASM) — a mandatory five-year program launched in selected areas in 2026 to improve the management of chronic heart failure

Category	Test/Service	Clinical Role	Common CPT Codes
Primary Imaging	**Transthoracic Echocardiogram (TTE)**	First-line evaluation of suspected cardiomyopathy. Assesses EF, wall thickness, chamber size, diastolic function, and valvular disease.	**93306** – Complete TTE with Doppler & color flow **93307** – Complete TTE (no Doppler) **93308** – Limited/follow-up **93356** – Myocardial strain (add-on)
Advanced Imaging	**Cardiac MRI (cMRI)**	Differentiates ischemic vs non-ischemic cardiomyopathy; identifies fibrosis (late gadolinium enhancement); evaluates HCM,	**75557** – MRI, no contrast **75559** – MRI with contrast & stress **75561** – MRI with contrast **75563** – MRI with contrast & flow mapping

Category	Test/Service	Clinical Role	Common CPT Codes
		myocarditis, infiltrative disease	
Hemodynamic Assessment	**Right Heart Catheterization**	Evaluates pulmonary pressures and cardiac output; clarifies severity; differentiates restrictive vs constrictive physiology	**93451** – Right heart cath **93452** – Left heart cath **93453** – Combined right & left heart cath
Heart Failure Evaluation & Ongoing Management	**E/M & Monitoring Services**	Used when cardiomyopathy results in heart failure diagnosis; supports longitudinal management and medication adjustment	**99214–99215** – Established patient E/M **99457** – Remote physiologic monitoring (first 20 min) **99458** – RPM add-on **G2066** – Implantable cardiac monitor interrogation

Abbreviations: TTE, Transthoracic Echocardiogram; EF, Ejection Fraction; f/u, Follow-up; cMRI/MRI, Cardiac Magnetic Resonance Imaging; ICM, Ischemic Cardiomyopathy; NICM, Non-ischemic Cardiomyopathy; LGE, Late Gadolinium Enhancement; HCM, Hypertrophic Cardiomyopathy; RHC, Right Heart Catheterization; LHC, Left Heart Catheterization; PAPs, Pulmonary Artery Pressures; CO, Cardiac Output; E/M, Evaluation and Management; RPM, Remote Physiologic Monitoring; HF, Heart Failure; GDMT, Guideline-Directed Medical Therapy; ICM (In Management context), Implantable Cardiac Monitor

Subtle Early Signs[9–12]

1. The "Unexplained" Fatigue/Decline in Function

- **The Sign:** A progressive decline in METs (Metabolic Equivalents) during ADLs (Activities of Daily Living), characterized by post-exertional malaise following low-intensity aerobic tasks (e.g., shopping or light gardening). Patients may report a change from NYHA Class I (asymptomatic) to Class II (slight limitation) over a six-month trajectory
- **Pathophysiology:** This represents a failure of cardiac reserve. As myocardial contractility or diastolic filling impairs stroke volume (SV), the body cannot meet the increased oxygen demand (VO_2) of skeletal muscle during exertion. To maintain mean arterial pressure (MAP) and cerebral/renal perfusion, the sympathetic nervous system triggers peripheral vasoconstriction, leading to muscular fatigue and the clinical requirement for extended recovery periods

2. Paroxysmal Nocturnal Dyspnea (PND) and Orthopnea

- **The Clinical Sign:** A sudden onset of acute respiratory distress typically occurring 1–3 hours after sleep onset, often requiring the patient to sit upright or stand to achieve relief. This is frequently accompanied by **orthopnea** — a requirement for cephalic elevation (e.g., multiple pillows) to maintain respiratory comfort while supine
- **The Sub-Clinical "Subtle" Presentation:** Patients may exhibit an insidious increase in pillow requirement (e.g., "three-pillow orthopnea") or report a persistent, non-productive **nocturnal cough**. This cough is often a manifestation of pulmonary congestion and bronchial mucosa edema rather than a primary respiratory infection

3. Symptomatic Arrhythmia and Palpitations

The Clinical Sign: Patient reports of subjective awareness of cardiac activity, characterized by "skipped beats" (**Premature Ventricular or Atrial Contractions**), a "flip-flopping" sensation (**Paroxysmal Supraventricular Tachycardia**), or sustained tachydysrhythmias occurring independently of physical or emotional stress. Of particular clinical significance is the onset of these symptoms during periods of physical inactivity or while in a resting state.

Pathophysiology — Electromechanical Remodeling: Structural changes to the myocardium (hypertrophy, dilation, or fibrosis) create a substrate for arrhythmogenesis through several mechanisms:

1. **Mechanical Stretch:** Dilation of the chambers (as seen in DCM) stretches the myocardial fibers, altering the properties of ion channels and increasing **automaticity** in non-pacemaker cells
2. **Myocardial Fibrosis:** As the heart muscle thickens or undergoes remodeling, healthy tissue is replaced by interstitial fibrosis. These fibrotic areas act as barriers to normal electrical conduction, creating circuits for **re-entry**, which can trigger sustained ventricular or atrial arrhythmias
3. **Afterdepolarizations:** Intracellular calcium handling is often impaired in failing myocytes, leading to delayed afterdepolarizations (DADs) that can trigger **triggered activity** (premature beats)

4. Exercise Intolerance and Diminished Metabolic Reserve

The Clinical Sign: A progressive decline in **peak aerobic capacity (VO2 max)** and an abnormal increase in the **Rating of Perceived Exertion (RPE)** relative to submaximal workloads. This often manifests as **chronotropic incompetence** (failure to reach 85% of age-predicted maximal heart rate) or, conversely, an exaggerated tachycardic response to minimal exertion.

Pathophysiology: Impaired Hemodynamic Coupling: The inability to sustain exercise is rarely isolated to a single mechanism; rather, it is a multi-system failure of the "Fick Principle" (Cardiac output = oxygen consumption/arterio-venous oxygen difference)

Systolic/Diastolic Dysfunction: In dilated or restrictive states, the myocardium cannot increase **stroke volume (SV)** via the Frank-Starling mechanism. This leads to a "fixed" cardiac output (CO) that cannot meet the peripheral metabolic demands of working skeletal muscle

Elevated Pulmonary Capillary Wedge Pressure (PCWP): Rapid increases in left ventricular end-diastolic pressure during exertion are transmitted to the pulmonary vasculature, causing **exertional dyspnea** and a sensation of "heaviness" or air hunger

Skeletal Muscle Hypoperfusion: Chronic low-output states trigger a transition from Type I (oxidative) to Type II (glycolytic) muscle fibers, leading to early **lactic acidosis**, premature anaerobic threshold, and rapid muscular fatigue

Autonomic Dysregulation: Increased sympathetic tone and blunted baroreceptor sensitivity prevent efficient heart rate recovery (HRR) post-exertion, a known prognostic marker for adverse cardiac events

Geriatric Risk Factors[13–17]

1. Structural and Physiological Factors (Senescence)

Myocardial Stiffening: Age-related increase in collagen cross-linking and interstitial fibrosis leads to decreased ventricular compliance. This is a primary driver for HFpEF (Heart Failure with preserved Ejection Fraction), the most common form of heart failure in patients over 75

Reduced Beta-Adrenergic Responsiveness: The aging heart has a blunted response to catecholamines, leading to chronotropic incompetence and a reduced ability to increase cardiac output during physical or febrile stress

Transthyretin Amyloidosis (ATTRwt): Formerly known as "senile systemic amyloidosis," this is an increasingly recognized risk factor where wild-type transthyretin proteins misfold and deposit in the myocardium. It is estimated to be present in up to 25% of patients over age 80 with thickened heart walls

2. Comorbidity and Chronic Conditions

Chronic Hypertension: Long-term pressure overload leads to Left Ventricular Hypertrophy (LVH). In geriatrics, this often transitions into a restrictive or dilated phenotype if the hypertension remains poorly controlled

Valvular Heart Disease: Conditions like Calcific Aortic Stenosis create significant afterload, causing compensatory hypertrophy that can eventually lead to secondary cardiomyopathy and heart failure

Diabetes and Metabolic Syndrome: Hyperglycemia promotes the formation of Advanced Glycation End-products (AGEs), which stiffen the myocardium and impair microvascular perfusion, a condition often termed "diabetic cardiomyopathy"

3. Latrogenic and External Risks

Polypharmacy: Elderly patients are frequently on multiple medications that may have negative inotropic effects or cause electrolyte imbalances (e.g., certain calcium channel blockers, NSAIDs, or anti-arrhythmics)

History of Cardiotoxic Therapy: Seniors who are cancer survivors may experience "late-onset" cardiomyopathy decades after exposure to anthracyclines or chest radiation (Radiation-Induced Heart Disease)

Nutritional Deficiencies: Subclinical deficiencies in micronutrients like Thiamine (B1) or Selenium—common in the elderly due to malabsorption or restricted diets — can manifest as high-output heart failure or dilated cardiomyopathy

RED FLAGS - URGENT ACTION[5,18–24]

"Red Flags" represent a transition from chronic compensation to acute decompensation or malignant dysrhythmia. These findings require immediate triage to an Emergency Department or Cardiac Catheterization Lab to prevent sudden cardiac death (SCD) or cardiogenic shock.

1. Hemodynamic & Perfusion Emergencies

These signs indicate a critical drop in cardiac output (CO) and the failure of systemic perfusion:

Syncope or Near-Syncope:

- Clinical Presentation: Sudden loss of consciousness, particularly during exertion or while seated
- **Urgency**: In cardiomyopathy, syncope is a harbinger of Ventricular Tachycardia (VT) or Ventricular Fibrillation (VF). It is a Tier 1 red flag for sudden cardiac arrest

Hypoperfusion ("Cold and Dry"):

- **Clinical Presentation**: Altered mental status (confusion), cold/clammy extremities, and oliguria (low urine output)
- **Urgency**: Suggests Cardiogenic Shock where the cardiac index (CI) has fallen below the threshold for end-organ viability (<2.2 L/min/m²)

2. Acute Pulmonary Decompensation

These signs indicate a rapid rise in Left Ventricular End-Diastolic Pressure (LVEDP) leading to alveolar flooding:

Acute Respiratory Distress:

- **Clinical Presentation:** Tachypnea (RR >30), use of accessory muscles, and the inability to speak in full sentences
- **Urgency: I**ndicates Acute Decompensated Heart Failure (ADHF) or Flash Pulmonary Edema

Pink, Frothy Sputum:

- **Clinical Presentation:** Hemoptysis-like appearance caused by fluid and red blood cells being forced into the alveoli
- **Urgency:** A classic sign of life-threatening pulmonary edema requiring immediate diuresis or mechanical ventilation (CPAP/BiPAP)

3. Malignant Symptom Clusters

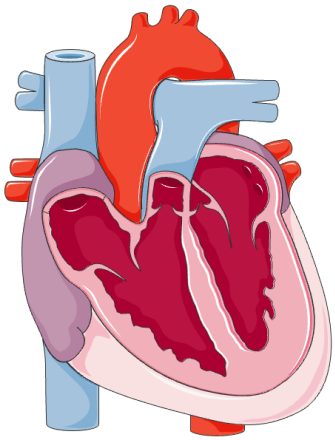
Unstable Angina: Chest pain that is new, worsening, or occurring at rest. In cardiomyopathy, this suggests a severe supply-demand mismatch or concurrent myocardial infarction

"Thunderclap" Palpitations with Dizziness: Rapid, sustained palpitations (HR >150bpm) associated with hypotension. This often indicates a transition into Atrial Fibrillation with RVR or Non-Sustained Ventricular Tachycardia (NSVT)

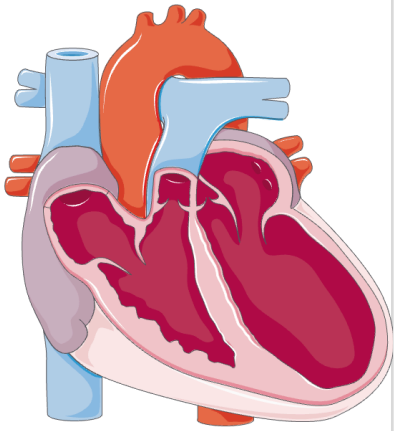
Red Flag	Clinical Suspicion	Immediate Action Required
Syncope	Malignant Arrhythmia (VT/VF)	Stat EKG, Continuous Telemetry, Troponin
Resting Dyspnea	Pulmonary Edema/ADHF	High-flow O2, IV Loop Diuretics
Mental Confusion	Cerebral Hypoperfusion	IV Inotropes, Invasive Hemodynamic Monitoring
Chest Pain at Rest	Ischemia/Myocardial Infarction	12-lead EKG, CXR, Serial Cardiac Enzymes

Abbreviations: ADHF, Acute Decompensated Heart Failure; CXR, Chest X-ray; EKG, Electrocardiogram; IV, Intravenous; O2, Oxygen; VF, Ventricular Fibrillation; VT

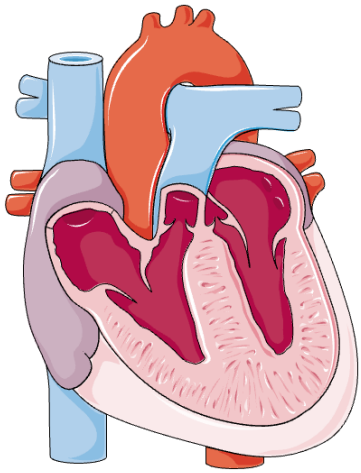
Diagnostic Thresholds



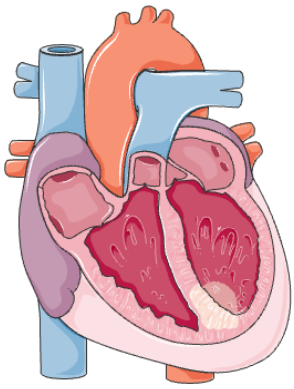
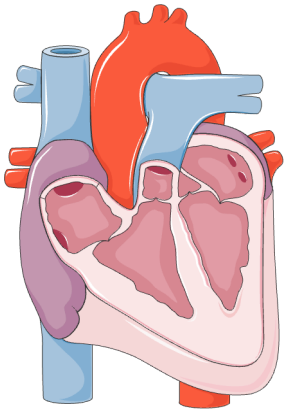
Normal heart



Dilated cardiomyopathy (enlarged ventricle)



Hypertrophic cardiomyopathy (thickened wall)

	
Arrhythmogenic cardiomyopathy (fat and scar tissue)	**Restrictive cardiomyopathy (stiffened wall)**

1. Dilated Cardiomyopathy (DCM)[25,26]

DCM is characterized by ventricular chamber enlargement and systolic dysfunction in the absence of abnormal loading conditions (e.g., severe hypertension or valvular disease). Patients usually present with symptoms of biventricular failure (e.g., fatigue, dyspnoea, orthopnoea, ankle oedema). Associated with a high mortality (2-year survival = 50%) due to progressive cardiogenic shock or ventricular dysrhythmias (sudden cardiac death):

- **Left Ventricular Ejection Fraction (LVEF):** <40-45%
- **Left Ventricular End-Diastolic Dimension (LVEDD):** >117% of the predicted value based on age and Body Surface Area (BSA). (Commonly >58 mm in males or >52 mm in females)
- **Fractional Shortening:** <25%

Common ECG Findings in DCM:

1. **Depolarization/Repolarization Abnormalities:**
   - T-wave inversions (TWI): Very common, especially in lateral or inferior leads
   - Low QRS voltage: Reduced amplitude of the QRS complex, indicating poor electrical signal
   - Abnormal Q waves: Can appear in various leads, mimicking a heart attack
2. **Conduction Defects:**
   - Left Bundle Branch Block (LBBB): A frequent finding, often with right axis deviation (RAD)
   - Intraventricular Conduction Delay (IVCD): General delay in electrical spread within the ventricles
   - Prolonged PR interval/AV block: Indicating slowed conduction between atria and ventricles
3. **Atrial Changes:**

- Left atrial enlargement: Seen as P-wave abnormalities (P mitrale)
- Atrial fibrillation (AFib): A common arrhythmia in DCM, signaling advanced disease

4. **Arrhythmias:**
   - Sinus tachycardia: Heart beating too fast
   - Ventricular ectopics/tachycardia: Premature heartbeats or rapid rhythms from the ventricles

Causes of Dilated Cardiomyopathy:

Can be divided into *ischaemic* and *non-ischaemic*.

1. **Ischaemic**
   - Dilated cardiomyopathy commonly occurs following massive anterior STEMI due to extensive myocardial necrosis and loss of contractility
2. **Non-ischaemic**
   - Most cases are idiopathic
   - Up to 25% are familial (primarily autosomal dominant, some types are X-linked)

A very small proportion may occur with:

- Viral myocarditis (coxsackie B/adenovirus/coronavirus)
- Alcoholism
- Toxins (e.g. doxorubicin)
- Autoimmune disease
- Pregnancy (peripartum cardiomyopathy)

2. Hypertrophic Cardiomyopathy (HCM)[27–29]

Hypertrophic Cardiomyopathy (HCM) is a primary myocardial disorder characterized by left ventricular hypertrophy (LVH) in the absence of abnormal loading conditions (e.g., systemic hypertension or valvular aortic stenosis). Formerly categorized as HOCM or IHSS, the terminology has shifted to HCM to reflect that approximately **75% of patients** do not exhibit outflow obstruction at rest. The hallmark of HCM:

- **Unexplained left ventricular hypertrophy (LVH)**
- Occurring **in the absence of abnormal loading conditions** (e.g., longstanding hypertension or aortic stenosis)

1. **Epidemiology and Genetics**
   - **Prevalence:** ~1:500 in the general population; recognized as the leading cause of **Sudden Cardiac Death (SCD)** in young athletes
   - **Mortality:** Annual attrition rate is estimated at 1-2%
   - **Genetics:** Predominantly an **autosomal dominant** trait with variable penetrance and expressivity
   - **Molecular Basis:** Attributed to >150 identified mutations in genes encoding **sarcomeric proteins**, most commonly the **beta-myosin heavy chain** and **cardiac troponin T**. This genetic heterogeneity accounts for the wide spectrum of clinical phenotypes

2. **Structural and Morphological Characteristics**
    - **Diagnostic Thresholds:** For the general adult, the degree and distribution of LVH is variable: mild hypertrophy (13-15mm) or extreme myocardial thickening (30-60mm) may be seen. Diagnosis is primarily made via **Transthoracic Echocardiogram (TTE)** or **Cardiac MRI (CMR)**.

Patient Profile	Wall Thickness Threshold	Diagnostic Context
Index Patient (General)	≥15 mm	Maximum end-diastolic wall thickness at any segment
Family History/ Genetic +	≥13 mm	Lower threshold used if a 1st-degree relative has HCM or the patient has a known sarcomeric mutation
Pediatric Patients	Z-score ≥2.5	Adjusted for body surface area; ≥2.0 if family history/genetics are positive

    - **Morphological Patterns:**
        - **Asymmetrical Septal Hypertrophy (ASH):** The most prevalent phenotype, involving the anterior interventricular septum. It is frequently associated with **Systolic Anterior Motion (SAM)** of the mitral valve
        - **Concentric Hypertrophy (~20%):** Symmetrical wall thickening
        - **Apical HCM (~10%):** Localized hypertrophy of the LV apex, resulting in a "spade-shaped" LV cavity on ventriculography and "giant" T-wave inversions on ECG (>10 mm) depth in precordial leads)

3. **Pathophysiology and Hemodynamics**
    - **Dynamic LVOT Obstruction:** Defined as a peak instantaneous gradient ≥30 mmHg at rest or with provocative maneuvers (e.g., Valsalva). Obstruction is often caused by the Venturi effect pulling the mitral valve toward the septum during systole (SAM)
    - **Diastolic Dysfunction:** Characterized by impaired myocardial relaxation and increased stiffness, leading to elevated **Left Ventricular End-Diastolic Pressure (LVEDP)** and subsequent retrograde pulmonary venous congestion
    - **Microvascular Ischemia:** Resulting from intramural coronary artery abnormalities (vessel wall thickening/luminal narrowing) and increased myocardial oxygen demand due to high muscle mass
    - **Arrhythmogenic Substrate:** Myocardial **"cellular disarray"** and interstitial fibrosis disrupt normal conduction pathways, facilitating re-entry circuits and increasing the risk of ventricular tachyarrhythmias

4. **Clinical Presentation**
    - **Syncope/Presyncope:** Of the highest clinical concern; typically exertional, suggesting a critical reduction in cardiac output due to dynamic obstruction or the onset of malignant dysrhythmias
    - **Congestive Symptoms:** Exertional dyspnea, PND, and orthopnea resulting from diastolic heart failure

- **Angina Pectoris:** Often occurs in the absence of epicardial coronary artery disease, driven by oxygen supply-demand mismatch
- **Palpitations:** Frequently associated with atrial fibrillation (secondary to left atrial enlargement) or ventricular ectopy

3. Restrictive Cardiomyopathy (RCM)[30–34]

Restrictive cardiomyopathy is the **least common form of cardiomyopathy.** It occurs in the advanced stages of myocardial infiltrative disease (e.g., due to haemochromatosis, amyloidosis or sarcoidosis). RCM is defined by diastolic dysfunction in the presence of normal or near-normal systolic function and wall thickness. Restrictive cardiomyopathy is characterized by:

- **Impaired ventricular filling (diastolic dysfunction)**
- **Normal or near-normal systolic function (preserved EF)**
- **Normal or mildly increased wall thickness**
- **Biatrial enlargement common**

Patients often present with signs of right-sided congestion (edema, ascites) and exercise intolerance despite preserved EF.

Etiology of Restrictive Cardiomyopathy

1. **Familial (Inherited)**

   These conditions are typically driven by genetic mutations affecting sarcomeric proteins, storage, or metabolic pathways:

   - Familial, unknown gene
   - Sarcomeric protein mutations:
   - Troponin I (May present as RCM ± HCM)
   - Essential light chain of myosin
   - Familial Amyloidosis:
   - Transthyretin (Associated with RCM + neuropathy)
   - Apolipoprotein (Associated with RCM + nephropathy)
   - Desminopathy
   - Pseudoxanthoma elasticum
   - Haemochromatosis (Can be hereditary or acquired)
   - Anderson–Fabry disease
   - Glycogen storage disease

2. **Non-familial (Acquired)**

   These conditions result from systemic diseases, external agents, or idiopathic processes:

   - Amyloid (AL/prealbumin)
   - Scleroderma
   - Endomyocardial fibrosis
   - Hypereosinophilic syndrome

- Chromosomal cause
- Carcinoid heart disease
- Metastatic cancers
- Radiation therapy
- Drug-induced: Specific agents: Serotonin, methysergide, ergotamine, mercurial agents, busulfan. Anthracyclines (e.g., Doxorubicin)
- Idiopathic (Unknown acquired cause)

Pathophysiology and Hemodynamics:

The primary defect in RCM is a significant increase in myocardial stiffness, which leads to a precipitous rise in ventricular pressure with even small increases in volume:

- **Diastolic Dysfunction:** Severe impairment of ventricular relaxation and filling (Grade III restrictive pattern). The ventricles are non-compliant, leading to a rapid rise in Left Ventricular End-Diastolic Pressure (LVEDP)
- **Atrial Remodeling:** Because the ventricles cannot accommodate blood flow, pressure is transmitted retrogradely. This results in severe bi-atrial enlargement, which serves as a substrate for atrial fibrillation and mural thrombi
- **The "Square Root" Sign:** In invasive hemodynamics, the ventricular pressure tracing often shows a "dip-and-plateau" pattern (the square root sign), indicating rapid early diastolic filling followed by an abrupt halt due to the rigid myocardium

Diagnostic Thresholds and Imaging

RCM is often a diagnosis of exclusion, requiring the differentiation from Constrictive Pericarditis, which shares similar hemodynamic profiles:

1. **Echocardiography:**
   - **Diastolic Parameters:** Grade II or III (restrictive) diastolic dysfunction on Doppler imaging (E/A ratio >2, DT <160 ms)
   - **Atrial Volume Index:** Massive bi-atrial enlargement (indexed Left Atrial Volume >40–50mL/m^2, often significantly higher in RCM)
   - **Wall Thickness:** Typically normal, though mildly increased in infiltrative types like Amyloidosis.
2. **Cardiac MRI (cMRI):**
   - **T1 Mapping/ECV:** Increased Extracellular Volume (ECV >30%) is a hallmark of interstitial expansion
   - **LGE Pattern:** Diffuse, subendocardial late gadolinium enhancement is highly suggestive of Amyloidosis
3. **Brain Natriuretic Peptide (BNP):** Usually significantly elevated (>400 pg/mL) in RCM, whereas it is often normal or only mildly elevated in Constrictive Pericarditis

Clinical Manifestations:

- **Right-Sided Heart Failure:** Predominant symptoms include peripheral edema, ascites, and painful hepatomegaly due to venous congestion
- **Kussmaul's Sign:** An abnormal paradoxical rise in Jugular Venous Pressure (JVP) during inspiration
- **Exercise Intolerance:** Resulting from a "fixed" stroke volume; the heart cannot increase cardiac output in response to demand
- **Arrhythmias:** High incidence of Atrial Fibrillation and high-grade AV block (especially in Sarcoidosis or Amyloidosis)

4. Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)[7,35,36]

ARVC is characterized by the fibro-fatty replacement of the right ventricular (RV) myocardium. Diagnosis follows **The Padua Criteria**, requiring a combination of major and minor criteria across structural, histological, ECG, and genetic categories:

The Padua Criteria: Diagnostic Standards for Arrhythmogenic Cardiomyopathy (ACM)

Category	Right Ventricular (RV) Criteria	Left Ventricular (LV) Criteria
I. Morpho-functional Abnormalities	**Major**: Regional RV akinesia, dyskinesia, or localized bulging accompanied by global RV dilatation or significant systolic dysfunction (reduced RVEF) **Minor**: Isolated regional RV akinesia, dyskinesia, or localized free wall aneurysm	**Minor**: General LV systolic impairment (depressed LVEF or reduced GLS) with or without chamber dilatation (increased LVEDV) **Minor**: Regional hypokinesia or akinesia localized to the LV free wall or septum
II. Structural Myocardial Abnormalities	**Major**: Transmural Late Gadolinium Enhancement (LGE) stria pattern in at least one RV region (inlet, outlet, or apex) **Major** (Biopsy): Histological evidence of myocardial fibrous replacement (with/without fat) in one or more samples	**Major**: Mid-myocardial or subepicardial LGE (stria pattern) in at least one Bull's Eye segment of the LV free wall or septum (excluding isolated junctional LGE)
III. ECG Repolarization Changes	**Major**: T-wave inversion (TWI) in right precordial leads (V1 through V3) or further in post-pubertal adults (without complete RBBB) **Minor**: TWI in V1–V2 (without RBBB) or TWI in V1–V4 specifically in the presence of complete RBBB	**Minor**: T-wave inversion in the left precordial leads (V4 through V6) in the absence of complete LBBB
IV. ECG Depolarization Changes	**Minor**: Epsilon waves (low-amplitude signals between QRS and T-wave) in leads V1–V3. **Minor**: Terminal activation delay (≥55 ms) from S-wave nadir to end of QRS in V1, V2, or V3 (without RBBB)	**Minor**: Low QRS voltage (<0.5 mV peak-to-peak) in the limb leads, provided obesity or effusion are excluded.
V. Arrhythmia Morphology	**Major**: Sustained/non-sustained VT or frequent PVCs (>500/24h) showing an	**Minor**: Sustained/non-sustained VT or frequent PVCs (>500/24h) showing a RBBB

Category	Right Ventricular (RV) Criteria	Left Ventricular (LV) Criteria
	LBBB morphology with a non-inferior axis. **Minor**: PVCs or VT with an LBBB morphology and an inferior axis (RVOT-like pattern)	morphology (excluding fascicular patterns)
VI. Family History & Genetics	**Major**: Confirmed ACM in a 1st-degree relative or detection of a pathogenic ACM genetic mutation in the patient	**Minor**: Suspected ACM/premature sudden death (<35 years) in a 1st-degree relative, or confirmed ACM in a 2nd-degree relative

Abbreviations: ACM, Arrhythmogenic Cardiomyopathy; BSA, Body Surface Area; CE-CMR, Contrast-Enhanced Cardiac Magnetic Resonance; CMR, Cardiac Magnetic Resonance; EDV, End Diastolic Volume; EF, Ejection Fraction; EMB, Endomyocardial Biopsy; LBBB, Left Bundle Branch Block; LGE, Late Gadolinium Enhancement; LV, Left Ventricle; RBBB, Right Bundle Branch Block; RV, Right Ventricle; RVOT, Right Ventricular Outflow Tract; TWI, T-Wave Inversion; TAD, Terminal Activation Duration; QRS, Ventricular Depolarization Complex; PVC, Premature Ventricular Contraction; NSVT, Non-sustained Ventricular Tachycardia; GLS, Global Longitudinal Strain.

Core Components of ARVC Diagnosis:

- **Imaging (Preferred: Cardiac MRI):** Identifies regional right ventricular (RV) akinesia, dyskinesia, aneurysms, or significant dilation. Cardiac MRI is preferred for detecting fibro-fatty infiltration and wall motion abnormalities
- **Electrocardiogram (ECG/SAECG):** Evaluates for depolarization/conduction abnormalities (e.g., epsilon waves, QRS prolongation) and repolarization abnormalities (e.g., T-wave inversion in V1-V3)
- **Arrhythmia Monitoring:** 24-hour Holter or exercise testing to detect ventricular arrhythmias (premature ventricular contractions)
- **Genetic Testing & Family History:** Identifies pathogenic mutations (often in desmosomal genes) and assesses family history of the disease or sudden death
- **Histology (Biopsy):** Rarely used due to its invasive nature, endomyocardial biopsy can identify myocardial atrophy, fibrosis, and fibro-fatty replacement

5. Unclassified Cardiomyopathies[37–39]

1. Left Ventricular Non-compaction (LVNC)

Left Ventricular Non-compaction (LVNC), or spongiform cardiomyopathy, is a rare heart muscle disorder resulting from a failure in the embryologic process of myocardial compaction, usually occurring in the first trimester.

Key Characteristics of LVNC:

- **Two-layered Myocardial Substrate:** The ventricular wall is characterized by two distinct layers: a thin, compacted epicardial layer and a thick, hyper-trabeculated (spongy) endocardial layer
- **Deep Intertrabecular Recesses:** The non-compacted layer contains deep recesses that fill with blood directly from the left ventricular cavity
- **Morphological Location:** The anomaly is typically most pronounced in the apical and mid-lateral regions of the left ventricle

- **Pathophysiology:** The failure to complete the compaction process prevents the spongy, embryonic meshwork of fibers from maturing, resulting in decreased heart function, potential for thromboembolism due to stagnant blood in the recesses, and ventricular arrhythmias

Diagnostic Thresholds:

- Jenni Criteria (ECHO): A non-compacted to compacted (NC/C) ratio >2.0 measured in end-systole (short-axis view)
- Petersen Criteria (CMR): An NC/C ratio >2.3 in end-diastole

Clinical Significance:

- Patients are at high risk for the "triple threat": progressive systolic heart failure, malignant ventricular arrhythmias (secondary to the arrhythmogenic substrate of the trabeculations), and systemic thromboembolism (due to blood stasis within deep intertrabecular recesses)

2. Takotsubo Syndrome (TTS)

TTS is a temporary, reversible heart condition often triggered by intense emotional or physical stress, causing sudden weakness of the heart muscle. Often triggered by a "catecholamine storm," TTS mimics an acute coronary syndrome (ACS) but typically lacks obstructive coronary artery disease.

- **Morphological Variants:** While apical ballooning is classic (80%), midventricular, basal (inverted), and focal variants are recognized
- **Hemodynamics:**

↓Contractility ↔	**↓Systolic volume**	**↓Stroke Work**	**↓Efficiency**
↑LVEDV	**↑LVESV**	**↑Relaxation time**	**↑ Potential energy**
↑LVEDP	**↔LVSV**	**↔Compliance**	

Takotsubo syndrome is associated with a severely impaired cardiac contractility and a shortened systolic period, which is balanced by increasing left ventricular end-diastolic volume (LVEDV) to preserve the left ventricular stroke volume (LVSV). Diastolic properties are characterized by prolonged active relaxation, but unaltered passive elastic properties and myocardial energetics reveal an inefficient system with increased potential and decreased kinetic energy (stroke work). EDPVR = end-diastolic pressure–volume relationship; ESPVR = end-systolic pressure–volume relationship; LV = left ventricular; LVEDP = left ventricular end-diastolic pressure; LVESV = left ventricular end-systolic volume

- **Physician Note:** Acute management focuses on identifying Left Ventricular Outflow Tract Obstruction (LVOTO). In LVOTO-positive shock, inotropes are contraindicated as they exacerbate the gradient; pure alpha-agonists (e.g., phenylephrine) and beta-blockers are preferred once hemodynamics allow

3. Endocardial Fibroelastosis (EFE)

Endocardial fibroelastosis (EFE) refers to a pronounced, diffuse thickening of the ventricular endocardium and presents as unexplained heart failure in infants and children.

- **Primary vs. Secondary:** Primary EFE is idiopathic; secondary EFE is often seen in the context of left-sided obstructive lesions, notably Hypoplastic Left Heart Syndrome (HLHS)
- **Imaging Hallmarks:** Transthoracic Echocardiogram (TTE) reveals a "bright," hyperechoic endocardial rim
- CMR provides superior characterization with a distinct hypointense rim on perfusion sequences and a hyperintense rim on Late Gadolinium Enhancement (LGE)

4. Mitochondrial Cardiomyopathy

Mitochondrial cardiomyopathy (MCM) is a rare, inherited heart muscle disease caused by genetic defects in mitochondrial DNA (mtDNA) or nuclear DNA that impair mitochondrial energy production, specifically affecting the oxidative phosphorylation (OXPHOS) chain.

- **Phenotypic Diversity:** Typically manifests as a Hypertrophic (HCM) or Dilated (DCM) phenotype, often with a rapid transition to systolic failure during metabolic crises (e.g., febrile illness)
- **Clinical Red Flags:** Suspicion should be high in patients presenting with cardiomyopathy alongside multisystemic "clues": sensorineural hearing loss, ptosis, ophthalmoplegia, or diabetes at a young age

5. Hypokinetic Non-dilated Cardiomyopathy (H-NDLVC)

Previously termed "systolic dysfunction with minimal dilatation," this is now recognized as a distinct subset of the DCM spectrum.

- **Definition:** Reduced Left Ventricular Ejection Fraction (LVEF <45-50%) in the absence of significant LV chamber enlargement (LVEDV <2 SD from mean)
- **Prognostic Insight:** Despite the "normal" heart size on chest X-ray, these patients often carry a high genetic burden (notably LMNA or DSP mutations) and may have a disproportionately high risk of sudden cardiac death relative to their degree of LV dilatation

Summary Table for Key Findings in Cardiomyopathy

Metric	Dilated (DCM)	Hypertrophic (HCM)	Restrictive (RCM)
LVEF	Reduced (<45%)	Preserved or Hyperdynamic	Usually Preserved (>50%)
Wall Thickness	Normal or Thinned	Thickened (≥15 mm)	Normal or Mildly Thickened
LV Cavity Size	Increased (Dilation)	Small or Obliterated	Normal to Small
Diastolic Function	Variable	Impaired (Grade I-II)	Severely Impaired (Grade III)
Global Longitudinal Strain (GLS)	Reduced (typically >−12%)	Reduced (esp. at apex/septum)	Severely reduced (>−10%)

Abbreviations: DCM, Dilated Cardiomyopathy; HCM, Hypertrophic Cardiomyopathy; RCM, Restrictive Cardiomyopathy; LVEF, Left Ventricular Ejection Fraction; LV, Left Ventricle; GLS, Global Longitudinal Strain

Clues to Dig Deeper[40–43]

1. The "Low Voltage" Paradox (EKG vs. Echo)

This is one of the most significant clues for **Infiltrative Cardiomyopathy** (e.g., Amyloidosis)

- **The Clue:** The Echocardiogram shows significant ventricular wall thickening (pseudohypertrophy), but the EKG shows **low QRS voltage** or a **pseudo-infarction pattern** (Q waves in the absence of CAD)
- **Clinical Significance:** Thick walls should produce large electrical signals. If the signals are small, the "thickening" is likely not muscle, but protein (amyloid) or iron (hemochromatosis) displacing healthy myocytes

2. Elevated Biomarkers at Baseline

In a stable patient, biomarkers should be negligible.

- **The Clue:** Persistent, low-level elevation of **High-Sensitivity Troponin (hs-cTn)** or **NT-proBNP** in a patient who is not in acute distress
- **Clinical Significance:** This suggests chronic, ongoing myocardial stress or "smoldering" myocyte death. Even if the Ejection Fraction is currently preserved, these patients are at high risk for structural remodeling

3. "Relative" Hypotension

For patients with a known history of chronic hypertension, a sudden "improvement" in blood pressure can be a deceptive clue.

- **The Clue:** A patient who was previously on three antihypertensives now has "perfect" blood pressure (e.g., 110/70mmHg) without medication
- **Clinical Significance:** This often indicates a failing heart that can no longer generate enough systemic pressure — a sign of transitioning into a dilated or low-output state

4. Abnormal Heart Rate Recovery (HRR)

This is a powerful sub-clinical clue often found during routine stress testing.

- **The Clue:** The heart rate fails to drop by at least **12 beats in the first minute** after stopping exercise
- **Clinical Significance:** This indicates **autonomic dysregulation** and is often an early sign that the heart's compensatory mechanisms are exhausted, preceding overt heart failure symptoms by months or years

5. The "Cherry on Top" (Strain Imaging)

When standard 2D Echo measurements (Ejection Fraction (EF) and Left Ventricular End-Diastolic Diameter (LVEDD)) are borderline, **Global Longitudinal Strain (GLS)** is the clinician's best tool for "digging deeper."

- **The Clue:** An "Apical Sparing" pattern on a strain map (often called a "cherry on top" appearance)
- **Clinical Significance:** The base and mid-sections of the heart are struggling, while the apex is still contracting well. This pattern is **pathognomonic for Cardiac Amyloidosis**

Common Oversights[12,44–47]

1. Attribution Error: "Deconditioning" vs. Heart Failure

One of the most frequent oversights is attributing exertional dyspnea and fatigue solely to age, obesity, or lack of exercise.

- **The Oversight:** Failing to pursue an echocardiogram in patients with a gradual decline in functional capacity
- **Clinical Clue:** If a patient's exercise tolerance has shifted significantly over a 6–12 month period, it should be treated as a **hemodynamic failure** until proven otherwise
- **Impact:** Delays the initiation of **Guideline-Directed Medical Therapy (GDMT)**, which is most effective when started early in the remodeling process

2. Missing the "Amyloid Mimic" in Hypertrophic States

In older patients, a thickened heart wall is often reflexively diagnosed as "hypertensive heart disease."

- **The Oversight:** Assuming all LVH is due to pressure overload (hypertension)
- **Clinical Clue:** The **Voltage-to-Mass Discrepancy**. If the Echo shows a wall thickness of $\geq$15mm but the EKG shows low QRS voltage, the diagnosis is likely **Cardiac Amyloidosis**, not hypertension
- **Impact:** Hypertensive treatments like ACE inhibitors or Beta-blockers can actually worsen symptoms in Amyloid patients by reducing necessary preload

3. Underestimating "Recovered" Ejection Fraction

When a patient's EF improves from 30% to 50% following treatment, there is a temptation to discontinue medications.

- **The Oversight:** Considering the patient "cured" and tapering GDMT (Guideline-Directed Medical Therapy)
- **Clinical Reality:** This is **Heart Failure with Improved Ejection Fraction (HFimpEF)**. The underlying molecular and structural substrate remains; withdrawing therapy often leads to rapid relapse and "recurrent" dilated cardiomyopathy
- **Impact:** High rate of readmission and accelerated myocardial fibrosis

4. Diagnostic "Tunnel Vision" on Ischemia

Because Coronary Artery Disease (CAD) is so prevalent, many clinicians stop investigating once a stress test or angiogram comes back "clean."

- **The Oversight:** Failing to investigate **Non-Ischemic Cardiomyopathy** (NICM) once obstructive CAD is ruled out

- **Clinical Clue:** If the patient has heart failure symptoms but clear arteries, you must look for "niche" causes: viral myocarditis, sarcoidosis, or genetic mutations (e.g., *LMNA* or *TTN*)
- **Impact:** Missing potentially treatable inflammatory or metabolic causes of heart muscle disease

5. Coding Specificity (The ICD-10/HCC Gap)

As discussed in the coding section, a significant administrative oversight is the use of non-specific codes.

- **The Oversight:** Using **I42.9** (Cardiomyopathy, unspecified)
- **The Consequence:** This code **does not map to an HCC**, resulting in an inaccurate representation of the patient's risk profile and potentially lower reimbursement for complex care coordination
- **Best Practice:** Always code the phenotype (Dilated, Hypertrophic, Restrictive) or the etiology (Ischemic, Alcoholic)

Key Differentials[3,34,48–51]

1. Differentials for the Dilated Phenotype (DCM)

The primary goal is to determine if the ventricular dilation is "idiopathic" or a secondary result of chronic stress.

- **Ischemic Heart Disease (CAD):** The most common mimic. Chronic multivessel disease leads to "hibernating myocardium" and remodeling.
  - *Differentiator:* Stress testing or coronary angiography; Cardiac MRI showing subendocardial (vs. mid-wall) Late Gadolinium Enhancement (LGE)
- **Valvular Heart Disease:** Severe aortic regurgitation or chronic mitral regurgitation can cause volume overload and secondary dilation
  - *Differentiator:* Echocardiographic evidence of primary valvular pathology
- **Tachycardia-Induced Cardiomyopathy:** Persistent rapid ventricular response (e.g., Atrial Fibrillation) can weaken the muscle
  - *Differentiator:* Reversibility of LV dysfunction after rate or rhythm control
- **High-Output Heart Failure:** Conditions like severe anemia, thyrotoxicosis, or AV fistulas
  - *Differentiator:* Clinical history and elevated Cardiac Index (CI) on right heart catheterization

2. Differentials for the Hypertrophic Phenotype (HCM)

The clinical focus here is distinguishing between "true" HCM (sarcomeric) and "pseudohypertrophy."

- **Hypertensive Heart Disease:** Chronic afterload causes concentric hypertrophy
  - *Differentiator:* History of poorly controlled BP; hypertrophy is usually symmetrical and rarely exceeds 15mm
- **"Athlete's Heart":** Physiological remodeling due to intense athletic training
  - *Differentiator:* LV cavity is usually enlarged (unlike HCM); wall thickness regresses with deconditioning
- **Cardiac Amyloidosis:** Infiltration of amyloid protein mimics thickened walls

- *Differentiator:* **Voltage-to-mass discrepancy** (Low EKG voltage vs. thick Echo walls); Apical sparing on strain imaging
- **Aortic Stenosis:** Pressure overload causes compensatory LVH
  - *Differentiator:* Presence of a systolic ejection murmur and calcified aortic valve on Echo

3. Differentials for the Restrictive Phenotype (RCM)

The most critical differential in this category is distinguishing between a stiff muscle and a stiff "container" (the pericardium)

- **Constrictive Pericarditis:** Scarring and calcification of the pericardium limit heart expansion
  - *Differentiator:* **Respiratory variation** in mitral inflow (>25%) and "septal bounce" on Echo; normal BNP levels
- **Cardiac Sarcoidosis:** Granulomatous infiltration
  - *Differentiator:* Often involves conduction blocks (AV block) and patchy LGE on Cardiac MRI or FDG-PET uptake
- **Hemochromatosis:** Iron deposition in the myocytes
  - *Differentiator:* Elevated serum ferritin and transferrin saturation; specific T2* sequences on Cardiac MRI

Summary of Key Differentials

Phenotype	Top Differential	Key Diagnostic Test
Dilated	Ischemic Cardiomyopathy	Coronary Angiography/cMRI
Hypertrophic	Hypertensive Heart Disease	Ambulatory BP Monitoring
Restrictive	Constrictive Pericarditis	Cardiac MRI/Right Heart Cath
Arrhythmogenic	Right Ventricular Infarction	ECG (ST changes)/Cardiac Enzymes/holter monitor

Abbreviations: ICM, Ischemic Cardiomyopathy; cMRI, Cardiac Magnetic Resonance Imaging; CAD, Coronary Artery Disease; HHD, Hypertensive Heart Disease; ABPM, Ambulatory Blood Pressure Monitoring; LVH, Left Ventricular Hypertrophy; RHC, Right Heart Catheterization; RVI, Right Ventricular Infarction; ECG, Electrocardiogram; ST, ST-segment; AEM, Ambulatory Electrocardiographic Monitoring

Comorbidity Screening[46,52–57]

In 2026, routine comorbidity screening is considered a foundational component of **Guideline-Directed Medical Therapy (GDMT)**. Because cardiomyopathy is often a progressive syndrome exacerbated by systemic factors, standardized screening protocols are used to identify "disease multipliers" that accelerate heart failure.

The following screening guidelines are standard for any patient with a new or established diagnosis of cardiomyopathy:

1. Metabolic & Endocrine Screening

Metabolic disorders are primary drivers of myocardial remodeling and diastolic dysfunction.

- **Diabetes Mellitus (HbA1c):** Screened annually. The presence of diabetes often dictates the early initiation of **SGLT2 inhibitors**, which provide both glycemic control and significant cardioprotection
- **Thyroid Function (TSH/Free T4):** Both hyper- and hypothyroidism can exacerbate arrhythmias (especially Atrial Fibrillation) and impair myocardial contractility
- **Iron Deficiency (Ferritin & TSAT):** Even in the absence of anemia, absolute or functional iron deficiency is screened for in all cardiomyopathy patients
    - *Threshold:* Ferritin <100 ng/mL or Ferritin 100-300 ng/mL with TSAT <20%

2. Sleep & Respiratory Screening

Sleep-disordered breathing increases sympathetic drive and pulmonary pressures, placing significant strain on the right and left ventricles.

- **Obstructive Sleep Apnea (OSA):** Routine screening using the **STOP-BANG** questionnaire. If positive, a formal sleep study (polysomnography) is required
- **Chronic Obstructive Pulmonary Disease (COPD):** Pulmonary function tests (PFTs) are used to differentiate between cardiac dyspnea and primary lung disease, especially in patients with a history of tobacco use

3. Renal & Electrolyte Monitoring

The "Cardiorenal Syndrome" describes the bidirectional failure of the heart and kidneys.

- **Chronic Kidney Disease (CKD):** Serial monitoring of **eGFR** and **Cystatin C**. Renal impairment significantly complicates the use of RAAS inhibitors and diuretics
- **Electrolyte Homeostasis:** Frequent monitoring of potassium and magnesium, as imbalances in these ions are highly pro-arrhythmic in a remodeled heart

4. Systemic Infiltrative & Inflammatory Screening

As discussed in the geriatric and restrictive sections, specific "clues" prompt deeper screening for systemic disease:

- **Monoclonal Gammopathy (Amyloid Screening):** Serum/Urine Immunofixation Electrophoresis (SIFE/UIFE) and Serum Free Light Chains (sFLC) to rule out **AL Amyloidosis**
- **Connective Tissue Disease (CTD):** ANA or RF titers if the patient has symptoms of lupus, scleroderma, or sarcoidosis

Comorbidity	Screening Tool	Recommended Frequency
Renal Function	eGFR/Creatinine	Every 3–6 months (depending on GDMT)
Iron Status	Ferritin/TSAT	Annually
Diabetes	HbA1c	Annually
Sleep Apnea	STOP-BANG / Sleep Study	At diagnosis; then as symptoms dictate
Depression/Anxiety	PHQ-9/GAD-7	Annually (due to high prevalence in HF)

Staging/Severity Matrix

In clinical practice, the severity of cardiomyopathy is assessed using two complementary frameworks: the **AHA/ACC Stages**, which track the structural progression of the disease, and the **NYHA Functional Class**, which measures the patient's symptomatic burden.

Beginning in 2025–2026, the integration of **Global Longitudinal Strain (GLS)** and **NT-proBNP** thresholds has become standard for refining these stages.

1. Structural Progression: AHA/ACC Stages

This matrix focuses on the development of the disease. Once a patient moves to a higher stage, they cannot "regress" to a previous stage, even if symptoms improve.

Stage	Clinical Definition	Management Focus
Stage A (At Risk)	High risk (HTN, Diabetes, Family History) but no structural disease or symptoms	Risk factor modification (ACEi/SGLT2i in select patients)
Stage B (Pre-HF)	Structural heart disease (e.g., LVEF <50%, LVH) but no current or prior symptoms	Initiation of GDMT to prevent remodeling
Stage C (Symptomatic)	Structural heart disease with current or prior symptoms of heart failure	Full GDMT (Quadruple therapy), diuretics, and device evaluation
Stage D (Advanced)	Refractory symptoms at rest despite maximal medical therapy	Mechanical Circulatory Support (LVAD), Transplant, or Palliative care

Abbreviations: ACEi, Angiotensin-Converting Enzyme Inhibitor; AF, Atrial Fibrillation; AHA/ACC, American Heart Association/American College of Cardiology; ARNI, Angiotensin Receptor-Neprilysin Inhibitor; BB, Beta-Blocker; BID, Twice a Day; DM, Diabetes Mellitus; GDMT, Guideline-Directed Medical Therapy; HF, Heart Failure; HTN, Hypertension; LVAD, Left Ventricular Assist Device; LVEF, Left Ventricular Ejection Fraction; LVH, Left Ventricular Hypertrophy; MCS, Mechanical Circulatory Support; MRA, Mineralocorticoid Receptor Antagonist; NYHA, New York Heart Association; QD, Once a Day; SGLT2i, Sodium-Glucose Cotransporter-2 Inhibitor

2. Functional Severity: NYHA Classification

The New York Heart Association (NYHA) scale is fluid; a patient's class can change frequently based on their response to treatment:

- **Class I:** No limitation of physical activity. Ordinary physical activity does not cause undue fatigue or dyspnea
- **Class II:** Slight limitation. Comfortable at rest, but ordinary activity results in fatigue, palpitations, or dyspnea
- **Class III:** Marked limitation. Comfortable at rest, but less than ordinary activity causes symptoms
- **Class IV:** Unable to carry out any physical activity without discomfort. Symptoms present at rest

3. Physiological Severity Matrix (Biomarkers & Imaging)

For medical professionals, staging is further quantified by objective hemodynamic and laboratory thresholds:

Severity Level	LVEF (%)	NT-proBNP (pg/mL)	GLS (Strain)
Mild	40% – 50%	125 – 450	−14% to −16%
Moderate	30% – 39%	450 – 900	−10% to −13%
Severe	<30%	>900	>−10%

Abbreviations: GLS, Global Longitudinal Strain; LVEF, Left Ventricular Ejection Fraction; NT-proBNP, N-Terminal Pro-B-Type Natriuretic Peptide

3. MEAT DOCUMENTATION ESSENTIALS

MONITOR: "Echo 03/2025 LVEF 30% (↓ from 35% 6mo ago), LVEDD 6.2 cm, NT-proBNP 520 (baseline 480), weight stable at 182lb dry weight, BP 112/68 on GDMT"

EVALUATE: "NYHA class II → III with decreased walking tolerance (2 blocks → ½ block); mild bilateral edema; no new ischemic changes; labs show K 4.3, Cr 1.2 — tolerating ARNI therapy"

ASSESS: "Non-ischemic dilated cardiomyopathy (I42.0) with stage C HFrEF (LVEF 30%), symptomatic progression likely due to ongoing LV remodeling; high risk for decompensation given rising BNP and functional decline"

TREAT: "Increased sacubitril/valsartan to 97/103 mg BID; continued carvedilol 25 mg BID and empagliflozin 10 mg daily; reinforced sodium restriction; scheduled repeat echo in 6 months; cardiology follow-up in 4 weeks"

Clinical Documentation Elements

Reflect phenotype, severity, and longitudinal management.

Link causal relationships: Link symptoms and complications directly to the cardiomyopathy phenotype "Exertional dyspnea and NYHA Class III fatigue **due** to progressive left ventricular remodeling and wall thinning in the setting of non-ischemic dilated cardiomyopathy"

Include current data: Include dated imaging and biomarkers reviewed during the encounter. “Latest Echocardiogram [Date] confirms HFrEF with an LVEF of 28% and significant global longitudinal strain (GLS) impairment at -9%”

Specify stage precisely: Specify cardiomyopathy subtype and structural features. “Hypertrophic Obstructive Cardiomyopathy (I42.1) involving asymmetrical septal hypertrophy (19mm) with a resting LVOT gradient of 35mmHg”

Document chronicity: Document stability or progression with ongoing GDMT and titration. “Stable chronic dilated cardiomyopathy (I42.0); continuing long-term GDMT including Sacubitril/Valsartan 97/103mg BID and Empagliflozin 10mg QD for symptom stabilization”

Reframing Common Documentation Shortcuts

Instead of…	Prefer documenting…	Why this supports care
“Stable CM”	“Non-ischemic dilated cardiomyopathy stable; LVEF 35% unchanged from prior echo (date); NT-proBNP 420 at dry-weight baseline.”	Anchors stability to objective comparison
“Worsening HF”	“NYHA class II → III; stage C HFrEF progression; LVEDD 6.4 cm on echo (date).”	Quantifies functional and structural change
“CM and swelling”	“Dependent edema secondary to restrictive cardiomyopathy; JVP 12 cm; furosemide increased to 40mg BID.”	Links symptom causally and justifies management
“Refilled meds”	“Sacubitril/valsartan titrated to 97/103 mg BID; potassium 4.2 and creatinine 1.1 reviewed (date).”	Demonstrates active titration and safety monitoring
“Short of breath”	“Paroxysmal nocturnal dyspnea and three-pillow orthopnea consistent with HFrEF and elevated LV filling pressures.”	Connects symptom to pathophysiology

4. TREATMENT AND REFERRAL QUICK GUIDE

Therapy Escalation Criteria

The treatment of cardiomyopathy is focused on three main goals: slowing disease progression, managing symptoms, and preventing sudden cardiac death (SCD).

In 2026, the standard of care is defined by Guideline-Directed Medical Therapy (GDMT), which has shifted toward "Quadruple Therapy" for patients with reduced function.

1. Pharmacological Management (The "Four Pillars")

For patients with Dilated or Ischemic cardiomyopathy (HFrEF), these four classes of drugs are initiated simultaneously to reduce mortality and hospitalization:

- **ARNI (Angiotensin Receptor-Neprilysin Inhibitor):** Such as Sacubitril/Valsartan. This has replaced ACE inhibitors as the preferred first-line agent for its superior ability to reduce cardiac remodeling
- **Beta-Blockers:** (e.g., Carvedilol, Metoprolol Succinate).These block the toxic effects of chronic sympathetic nervous system activation, slowing the heart rate and allowing the muscle to recover
- **MRA (Mineralocorticoid Receptor Antagonists):** (e.g., Spironolactone). These prevent fibrosis (scarring) of the heart muscle
- **SGLT2 Inhibitors:** (e.g., Empagliflozin, Dapagliflozin). Originally diabetes drugs, these are now a cornerstone of cardiomyopathy treatment for their unique ability to reduce "preload" and "afterload" while protecting renal function

2. Device and Surgical Interventions

When medications alone are insufficient, mechanical or electrical interventions may be necessary:

- **ICD (Implantable Cardioverter Defibrillator):** Recommended for patients with an LVEF <35% for greater than 90 days. It monitors for lethal arrhythmias (VT/VF) and delivers a shock to restore normal rhythm
- **CRT (Cardiac Resynchronization Therapy):** A specialized pacemaker used when the left and right ventricles do not beat in sync (often seen with a Left Bundle Branch Block)
- **Septal Myectomy/Alcohol Septal Ablation:** Used specifically for Hypertrophic Obstructive Cardiomyopathy (HOCM) to remove or shrink the thickened part of the heart wall that blocks blood flow
- **LVAD (Left Ventricular Assist Device):** A mechanical pump used for Stage D patients as a "bridge to transplant" or as permanent "destination therapy"

3. Emerging Targeted Therapies (2026 Standards)

Newer drugs target the specific molecular cause of the cardiomyopathy rather than just the symptoms:

- **Mavacamten:** A first-in-class cardiac myosin inhibitor used specifically for Obstructive HCM. It reduces the excessive "cross-bridging" of heart muscle fibers that causes thickening and obstruction
- **Tafamidis:** Used for Transthyretin Amyloid Cardiomyopathy (ATTR-CM). It stabilizes the TTR protein to prevent it from misfolding and depositing in the heart muscle

4. Lifestyle & Management Thresholds

To maintain stability, patients must manage "loading conditions" on the heart:

- **Sodium Restriction:** Typically <2000mg per day to prevent fluid overload
- **Fluid Management:** Daily weight monitoring. A gain of >2 to 3 lbs in 24 hours often triggers an "action plan" (e.g., a temporary increase in diuretics)
- **Exercise:** Cardiac rehabilitation is strongly recommended to improve peak VO2 and functional capacity

Phenotype	Primary Treatment Goal	Key Therapy
Dilated (DCM)	Improve Contractility	Quadruple Therapy (GDMT)
Hypertrophic (HCM)	Reduce Obstruction	Mavacamten/Septal Myectomy
Restrictive (RCM)	Manage Fill Pressures	Diuretics/Amyloid-specific drugs
Arrhythmogenic (ARVC)	Prevent SCD	Exercise restriction/ICD placement

Abbreviations: ARVC, Arrhythmogenic Right Ventricular Cardiomyopathy; DCM, Dilated Cardiomyopathy; GDMT, Guideline-Directed Medical Therapy; HCM, Hypertrophic Cardiomyopathy; ICD, Implantable Cardioverter-Defibrillator; RCM, Restrictive Cardiomyopathy; SCD, Sudden Cardiac Death

Treatment Options

This list categorizes common and advanced pharmacological interventions by their primary application across the three major cardiomyopathy phenotypes: **Dilated (DCM)**, **Hypertrophic (HCM)**, and **Restrictive (RCM)**.

Pharmacological Management of Cardiomyopathy

1. Dilated Cardiomyopathy (DCM)/HFrEF

The focus here is on "Quadruple Therapy" to improve systolic function and prevent remodeling.

- **Beta-Blockers (Metoprolol, Carvedilol, Nebivolol):** First-line for all DCM patients to reduce sympathetic drive and improve survival.
- **ACE Inhibitors/ARBs (Lisinopril, Losartan, Valsartan):** Used to reduce afterload and block the Renin-Angiotensin system; increasingly replaced by ARNI therapy
- **Sacubitril/Valsartan (Entresto):** The preferred agent for DCM; reduces hospitalization and mortality more effectively than ACE inhibitors alone
- **SGLT2 Inhibitors (e.g., Empagliflozin, Dapagliflozin):** Now standard of care for DCM to reduce preload and protect renal function
- **MRA's (Spironolactone, Eplerenone):** Used to block aldosterone-mediated fibrosis and manage fluid
- **Diuretics (Furosemide, Bumetanide):** Critical for symptomatic relief of pulmonary and peripheral edema
- **Ivabradine (Corlanor):** Specifically for patients in sinus rhythm with a heart rate $\geq$70bpm despite maximal beta-blocker therapy
- **Digoxin:** Often added for refractory symptoms or to control heart rate in concurrent Atrial Fibrillation

2. Hypertrophic Cardiomyopathy (HCM)

The focus is on relieving Left Ventricular Outflow Tract (LVOT) obstruction and improving diastolic filling.

- **Mavacamten (Camzyos):** Specifically indicated for **Obstructive HCM**. It reduces the excessive actin-myosin cross-bridging that drives hypertrophy and obstruction

- **Beta-Blockers:** First-line for obstructive HCM to slow the heart rate and increase diastolic filling time
- **Antiarrhythmics (Disopyramide):** Specifically used in HCM for its **negative inotropic** effect to reduce the LVOT gradient
- **Calcium Channel Blockers (Verapamil/Diltiazem):** *Note: Not in your list, but often used as an alternative to Beta-blockers in HCM*
- **Diuretics:** Used cautiously; over-diuresis can decrease preload and **worsen** obstruction in HCM

3. Restrictive Cardiomyopathy (RC)

The focus is primarily on volume management and rhythm control, as the ventricles are stiff and non-compliant.

- **Diuretics (Furosemide, Bumetanide):** The mainstay of treatment to manage the severe right-sided heart failure (ascites, edema) typical of RCM
- **Blood Thinners (Apixaban, Warfarin):** High priority in RCM because the massive bi-atrial enlargement significantly increases the risk of atrial thrombi and stroke
- **Beta-Blockers/Digoxin:** Used primarily for rate control if Atrial Fibrillation is present
- **ACE Inhibitors/ARBs:** Used with extreme caution; RCM patients are often sensitive to blood pressure drops (hypotension)

Drug Class	Dilated (DCM)	Hypertrophic (HCM)	Restrictive (RCM)
Beta-Blockers	Primary (Survival)	Primary (Gradient)	Rate Control
ARNI (Entresto)	Primary (Remodeling)	N/A	Rare
SGLT2 Inhibitors	Primary (Fluid/Renal)	Research ongoing	Symptom Management
Mavacamten	N/A	Specific to HCM	N/A
Diuretics	Congestion Relief	Use with Caution	Essential (Edema)
Blood Thinners	If AFib present	If AFib present	High Priority

Abbreviations: AF, Atrial Fibrillation; ARNI, Angiotensin Receptor-Neprilysin Inhibitor; BB, Beta-Blocker; HCM, Hypertrophic Cardiomyopathy; MRA, Mineralocorticoid Receptor Antagonist; N/A, Not Applicable; SGLT2i

Non-Rx Treatment Documentation

Implantable Electrical Devices

These devices address the electrical instabilities and mechanical dyssynchrony common in remodeled hearts.

- **Implantable Cardioverter Defibrillator (ICD):** * **Indication:** Primary prevention for patients with an LVEF ≤35% for greater than 90 days or secondary prevention for survivors of cardiac arrest
  - **Function:** Continuously monitors the cardiac rhythm and delivers a high-energy shock to terminate lethal ventricular tachycardia (VT) or fibrillation (VF)

- **Cardiac Resynchronization Therapy (CRT/Biventricular Pacing):**
  - **Indication:** Patients with an LVEF <35% and evidence of electrical dyssynchrony (typically a Left Bundle Branch Block with QRS duration >150ms
  - **Function:** Paces both ventricles simultaneously to restore a coordinated contraction, improving cardiac output and reversing remodeling

Therapeutic and Lifestyle Management

Non-procedural interventions are essential for maintaining hemodynamic stability and improving quality of life.

- **Cardiac Rehabilitation:** A structured, supervised exercise program designed to increase **peak VO_2** and improve skeletal muscle oxygen extraction, which reduces the workload on the heart
- **Sodium and Fluid Restriction:**
  - **Sodium:** Generally limited to <2,000 mg/day to prevent fluid retention
  - **Fluid:** Often limited to 1.5 to 2 L/day in symptomatic patients to prevent pulmonary congestion
- **Sleep Apnea Treatment (CPAP/BiPAP):** Treating obstructive sleep apnea is a non-medication "must" because it reduces nocturnal hypoxia and the sympathetic surges that drive heart failure progression

Follow-up Timing

In 2026, the follow-up cadence for cardiomyopathy is determined by the patient's AHA/ACC Stage, the stability of their NYHA Functional Class, and the timing of medication titration. A "one-size-fits-all" annual check-up is no longer the standard of care for patients on Guideline-Directed Medical Therapy (GDMT).

1. Post-Diagnosis & Titration Phase (Months 1-6)

- During the initial "stabilization" phase, follow-up is frequent to ensure the patient reaches target doses of the "Four Pillars" (ARNI, Beta-blocker, MRA, SGLT2i)
- **Clinical Visits:** Every 2-4 weeks until target medication doses are achieved or symptoms stabilize
- **Laboratory Monitoring:** 1-2 weeks after starting or increasing an ARNI or MRA to check Potassium and Creatinine/eGFR
- **Repeat Imaging:** A follow-up Echocardiogram is typically performed 3 months after reaching maximal tolerated GDMT to reassess the Ejection Fraction (EF) for potential ICD/CRT eligibility

2. Chronic Maintenance Phase (Stable Stage C)

Once a patient is stable on maximal therapy, the interval extends, but monitoring remains proactive to catch sub-clinical decompensation.

- **Clinical Visits:** Every 3-6 months
- **Biomarker Surveillance:** Serial NT-proBNP testing at each visit to establish a "dry" baseline
- **Echocardiography:** Annually, or sooner if there is a change in clinical status (e.g., new-onset edema or decreased exercise tolerance)

- **Device Checks:** For patients with an ICD or CRT, remote monitoring occurs continuously, with in-person interrogation every 6-12 months

4. Red Flag "Urgent" Follow-up

The standard schedule is bypassed if a patient experiences "breakthrough" symptoms. A clinic visit or telehealth assessment should occur within 24-48 hours for:

- Weight gain of >3 lbs in 24 hours
- Increased requirement for pillows at night (orthopnea)
- New-onset palpitations or lightheadedness

Phenotype	Key Follow-up Focus	Recommended Interval
Hypertrophic (HCM)	LVOT Gradient & SCD Risk	Annual Echo + 24-48 h Holter monitoring
Restrictive (RCM)	Volume & Atrial Fibrillation	Every 3 months (high risk for sudden fluid shifts)
Amyloidosis (ATTR)	Protein load & Extracardiac signs	Every 3–6 months; annual Technetium Pyrophosphate (PYP) is usually not required once diagnosed
ARVC	Ventricular Arrhythmia Burden	Every 6 months with EKG and Holter

Abbreviations: AF, Atrial Fibrillation; ARVC, Arrhythmogenic Right Ventricular Cardiomyopathy; ATTR, Transthyretin Amyloidosis; EKG, Electrocardiogram; HCM, Hypertrophic Cardiomyopathy; Holter, Ambulatory Electrocardiography Monitor; LVOT, Left Ventricular Outflow Tract; PYP, Technetium Pyrophosphate Scan; RCM, Restrictive Cardiomyopathy; SCD, Sudden Cardiac Death

Test	Frequency (Stable Patient)	Rationale
EKG	Every 6–12 months	Monitor for new blocks or Atrial Fib
Comprehensive Metabolic Panel	Every 3–6 months	Monitor renal function on diuretics/ARNI
Echocardiogram	Annually	Monitor for reverse remodeling or decline
6-Minute Walk Test	Annually	Objective assessment of functional capacity

Abbreviations: 6MWT, 6-Minute Walk Test; AF, Atrial Fibrillation; ARNI, Angiotensin Receptor-Neprilysin Inhibitor; CMP, Comprehensive Metabolic Panel; EKG, Electrocardiogram; GDMT, Guideline-Directed Medical Therapy; HF, Heart Failure; K+, Potassium; LVEF, Left Ventricular Ejection Fraction; SCr, Serum Creatinine

Patient Education and Adherence

1. The "Heart Failure Zones" (Self-Monitoring)

Patients are taught to use a "Stoplight" tool to categorize their daily symptoms. This enables early intervention before a full decompensation occurs.

Zone	Clinical Status	Required Action
Green (All Clear)	No shortness of breath; stable weight; no swelling	Continue usual medications; monitor weight daily
Yellow (Caution)	Weight gain of >3 lbs in 24 hrs; increased edema; needing more pillows to sleep	Call the Heart Clinic; potential adjustment of diuretics (water pills)
Red (Emergency)	Struggling to breathe at rest; chest pain; fainting or confusion	Call 911/Go to Emergency

Abbreviations: BID, Twice a Day; ED, Emergency Department; HF, Heart Failure; JVP, Jugular Venous Pressure; LVEDP, Left Ventricular End-Diastolic Pressure; NYHA, New York Heart Association; PND, Paroxysmal Nocturnal Dyspnea; QD, Once a Day; SOB, Shortness of Breath

2. Medication Literacy: The "Pillars" Rationale

A common cause of non-adherence is the "treatment burden" of taking 5-10 different pills. Education focuses on the specific protective role of each drug:

"The Shield" (Beta-Blockers): Protects the heart from toxic stress hormones that cause it to weaken over time

"The Remodeler" (ARNI/Entresto): Helps the heart muscle regain its shape and pumping strength

"The Filter" (SGLT2i/Empagliflozin): Originally for diabetes, this "flushes" extra sugar and salt to reduce pressure on the heart and kidneys

"The Scars-Preventer" (MRA/Spironolactone): Stops the heart from developing stiff scar tissue (fibrosis)

3. The "SADMANS" Rule for Sick Days

In 2026, patients are educated on when to temporarily pause medications to prevent kidney injury during acute illness (e.g., fever, vomiting, or diarrhea):

- S - SGLT2 inhibitors
- A - ACE inhibitors
- D - Diuretics
- M - Metformin
- A - ARBs
- N - NSAIDs (e.g., Ibuprofen) — Note: Cardiomyopathy patients should generally avoid NSAIDs entirely
- S - Statins (some protocols)

4. Lifestyle & Dietary Adherence Strategies

- **The "Rule of Two" (Sodium/Fluid):** Aim for <2,000mg of sodium and ~2L of fluid daily. Patients are taught to read labels for "Hidden Sodium" in processed foods

- **Daily Weight Tracker:** The most critical non-drug tool. Patients weigh themselves at the same time every morning (after voiding, before eating)
- **Cardiac Rehab:** Patients are educated that exercise is "prescribed" just like a pill. It improves the body's ability to use oxygen, directly reducing the heart's workload

Tool	Purpose	Frequency
Pill Organizer	Reduces missed doses in complex regimens	Weekly setup
Weight Log	Early detection of fluid retention	Daily (Morning)
Heart Failure Diary	Tracks symptoms, blood pressure, and heart rate	Daily
Home BP Monitor	Ensures medications aren't dropping pressure too low	Twice Daily (or as directed)

Abbreviations: BP, Blood Pressure; DBP, Diastolic Blood Pressure; GDMT, Guideline-Directed Medical Therapy; HF, Heart Failure; HR, Heart Rate; MAP, Mean Arterial Pressure; SBP, Systolic Blood Pressure

Comorbidity Management

1. The Cardiorenal-Metabolic (CRM) Triad

The most critical comorbidities involve the bidirectional relationship between the heart, kidneys, and metabolic system.

Chronic Kidney Disease (CKD):

- **Management:** Use of **SGLT2 inhibitors** and **Finerenone** (a non-steroidal MRA) to provide dual cardio-renal protection
- **Clinical Pivot:** Dose adjustments of diuretics and ARNI therapy are required as eGFR fluctuates. In 2026, the "permissiveness" for slight creatinine rises (up to 30%) is standard when initiating GDMT

Type 2 Diabetes (T2D):

- **Management:** SGLT2 inhibitors are now first-line therapy regardless of HbA1c, specifically to reduce heart failure hospitalizations
- **Avoidance:** Thiazolidinediones (e.g., Pioglitazone) are generally contraindicated due to fluid retention risks

Iron Deficiency:

- **Management:** Routine screening with Ferritin and TSAT
- **Intervention:** IV Iron (e.g., **Ferric Carboxymaltose**) is indicated for symptomatic patients even in the absence of anemia, as it improves functional capacity and reduces hospitalizations

2. Respiratory & Sleep Comorbidities

Pulmonary pressures and oxygenation levels directly impact ventricular wall stress.

Obstructive Sleep Apnea (OSA):

- **Management:** CPAP or BiPAP therapy is mandatory for symptomatic patients. OSA causes nocturnal sympathetic surges and increased negative intrathoracic pressure, which increases left ventricular afterload

COPD/Asthma:

- **Management:** Use of **cardioselective beta-blockers** (e.g., Bisoprolol or Metoprolol Succinate) is generally safe and necessary
- **Clinical Pivot:** Ensuring dyspnea is correctly attributed to either the lungs (airflow obstruction) or the heart (congestion) via NT-proBNP and PFTs

3. Arrhythmia & Rate Management

Structural changes in cardiomyopathy provide the perfect substrate for electrical instability.

Atrial Fibrillation (AFib):

- **Management:** In 2026, an **"Early Rhythm Control"** strategy (including catheter ablation) is preferred over simple rate control for cardiomyopathy patients to preserve atrial contribution to ventricular filling
- **Anticoagulation:** Direct Oral Anticoagulants (DOACs) are preferred to prevent thromboembolism.

Hyperkalemia:

- **Management:** Instead of discontinuing life-saving MRAs or ARNIs due to high potassium, clinicians now utilize **potassium binders** (e.g., Patiromer or Sodium Zirconium Cyclosilicate) to maintain patients on GDMT

Comorbidity	Key Monitoring	Primary Intervention Strategy
CKD	eGFR/Cystatin C	SGLT2i + Renoprotective GDMT
Iron Deficiency	Ferritin/TSAT	IV Iron Repletion (not oral)
Sleep Apnea	STOP-BANG/Sleep Study	CPAP to reduce sympathetic drive
Atrial Fib	EKG/Holter	Early Ablation + Anticoagulation
Depression	PHQ-9	Cardiac Rehab + Selective SSRIs

Abbreviations: AF, Atrial Fibrillation; CKD, Chronic Kidney Disease; CPAP, Continuous Positive Airway Pressure; Cystatin C, CysC; eGFR, Estimated Glomerular Filtration Rate; EKG, Electrocardiogram; GDMT, Guideline-Directed Medical Therapy; Holter, Ambulatory Electrocardiography Monitor; IV, Intravenous; PHQ-9, Patient Health Questionnaire-9; SGLT2i, Sodium-Glucose

Comorbidity	Key Monitoring	Primary Intervention Strategy
Cotransporter-2 Inhibitor; SSRIs, Selective Serotonin Reuptake Inhibitors; STOP-BANG, Snoring, Tiredness, Observed apnea, high blood Pressure, Body mass index, Age, Neck circumference, and Gender; TSAT, Transferrin Saturation		

Cost-Smart Options

In 2026, the high cost of newer, life-saving therapies like **ARNI** (Entresto), **SGLT2 inhibitors**, and **Cardiac Myosin Inhibitors** (Mavacamten) presents a significant challenge. However, several strategies—from newly negotiated Medicare pricing to "generic-first" protocols — can significantly reduce the financial burden of cardiomyopathy care.

1. The "Generic Foundation" Strategy

While new drugs get the headlines, three of the "Four Pillars" of cardiomyopathy therapy have high-quality, low-cost generic versions that should be maximized first.

Drug Class	Examples (Generic)	Typical Out-of-Pocket Cost
Beta-Blockers	Metoprolol, Carvedilol	$4 – $10 (90-day supply)
ACE-i/ARBs	Lisinopril, Losartan, Valsartan	$4 – $10 (90-day supply)
MRAs	Spironolactone	$4 – $10 (90-day supply)
SGLT2i	Farxiga or Jardiance	$300 (30-day supply for Farxiga) – $600+ (30-day supply for Jardiance)

Abbreviations: ACE-i, Angiotensin-Converting Enzyme Inhibitor; ARB, Angiotensin II Receptor Blocker; MRA, Mineralocorticoid Receptor Antagonist; SGLT2i

2. 2026 Medicare Price Negotiations

Starting in **January 2026**, the first round of Medicare-negotiated drug prices under the Inflation Reduction Act goes into effect. This significantly impacts cardiomyopathy patients on brand-name medications:

- **Entresto (Sacubitril/Valsartan):** One of the primary drugs selected for negotiation. Beneficiaries will see a significant reduction in out-of-pocket costs compared to previous years
- **SGLT2 Inhibitors:** Certain medications in this class (like Farxiga or Jardiance) are also subject to negotiated caps for Medicare Part D enrollees in 2026
- **The $2,000 Cap:** For all Medicare Part D beneficiaries, total out-of-pocket spending on *all* prescription drugs is now capped at **$2,000 per year** as of 2025

3. Patient Assistance Programs (PAPs) for Newer Drugs

For high-cost "specialty" cardiomyopathy drugs where generics do not yet exist, manufacturer programs are the primary cost-saving tool.

- **Mavacamten (Camzyos):** For Hypertrophic Cardiomyopathy (HCM). Manufacturers often provide a **$0 Co-pay Card** for commercially insured patients. For those without insurance, a PAP may provide the drug for free if income thresholds are met
- **Tafamidis (Vyndamax):** Used for Amyloidosis. Given its high cost ($225K/year), most patients utilize foundations like the *Patient Access Network (PAN)* or the *HealthWell Foundation* to cover the large copays

4. Tactical Strategies for Patients and Providers

- **90-Day Prescriptions:** Requesting 90-day instead of 30-day supplies can often save one full copay every quarter and reduces dispensing fees
- **The "SGLT2i Switch":** If one SGLT2 inhibitor (e.g., Jardiance) is not covered by a specific insurance formulary, the other (e.g., Farxiga) almost always is. They are generally considered "clinically interchangeable" for heart failure
- **Prior Authorization (PA) Support:** In 2026, many clinics use **automated PA software** or pharmacy technicians to speed up the approval process for Entresto or SGLT2i, ensuring patients don't pay "list price" while waiting for approval

Medication Tier	Typical Monthly Cost	Strategy
Tier 1 (Generics)	<$15	Use 90-day mail order
Tier 2 (Preferred Brand)	$30 – $100	Check for 2026 Medicare Negotiated Price
Tier 3 (Specialty/PAPs)	$100+	Manufacturer Copay Cards/Foundations

Quality Metrics Tie-In

HEDIS/STAR Domain	Specific Cardiomyopathy Metric Tie-In	Documentation Requirement
Safety (PCR)	Plan All-Cause Readmission: Patients with HFrEF have among the highest 30-day readmission rates in Medicare	Document a post-discharge visit within 7–14 days focusing on Medication Reconciliation (MRP) and volume status (dry weight/edema)
Chronic Care (CBP)	Controlling Blood Pressure: Hypertension is a "disease multiplier" for HCM and Dilated CM	Document the latest BP reading <140/90 mmHg. If elevated, document a plan for titration of GDMT (e.g., ARNI or Beta-blockers)
Effectiveness (FMC)	Follow-Up After ER Visit: High rates of "crisis" visits for acute decompensated heart failure (ADHF) or arrhythmias	Document a follow-up service within 7 days of an ED visit. Include an updated "Heart Failure Action Plan" (Green/Yellow/Red zones)
Prevention (CRE)	Cardiac Rehabilitation: Low utilization despite evidence that rehab reduces mortality in cardiomyopathy	Document a referral to Cardiac Rehab following a diagnosis or acute event to improve functional capacity and peak VO2

Abbreviations: ADHF, Acute Decompensated Heart Failure; ARNI, Angiotensin Receptor-Neprilysin Inhibitor; BP, Blood Pressure; CBP, Controlling Blood Pressure; CRE, Cardiac Rehabilitation; ED/ER, Emergency Department/Room; FMC, Follow-Up After Emergency Department Visit for People with Multiple High-Risk Chronic Conditions; GDMT, Guideline-Directed Medical Therapy;

HEDIS/STAR Domain	Specific Cardiomyopathy Metric Tie-In	Documentation Requirement

HCM, Hypertrophic Cardiomyopathy; HEDIS, Healthcare Effectiveness Data and Information Set; HFrEF, Heart Failure with Reduced Ejection Fraction; MRP, Medication Reconciliation Post-Discharge; PCR, Plan All-Cause Readmissions; VO2, Maximal Oxygen Consumption

5. CODING REMINDERS AND CASE EXAMPLES BOX

Critical Element	Clinical Requirement	Example Documentation
Stage & Level	Use the AHA/ACC Stage and NYHA Functional Class to quantify severity	Stage C, NYHA Class III. Documenting the specific stage and functional limitation is vital for Risk Adjustment and GDMT justification
Etiology	Link the cardiomyopathy to its underlying cause (e.g., Ischemic, Genetic, Toxic)	Ischemic Cardiomyopathy (I25.5) secondary to chronic multivessel CAD and prior apical MI. Establishing causality ensures correct ICD-10 mapping
Validation Data	Include objective metrics such as LVEF, GLS, and NT-proBNP levels	Echo (01/2026): LVEF 28%; GLS -9%; Pro-BNP 850 pg/mL. This satisfies the Monitor and Evaluate portions of the MEAT criteria
Complications	Explicitly link secondary conditions (e.g., heart failure, arrhythmias, or renal impact)	HFrEF (I50.22) and Atrial Fibrillation (I48.21) secondary to chronic Dilated Cardiomyopathy (I42.0); manages with Entresto and Apixaban

Abbreviations: ACEi, Angiotensin-Converting Enzyme Inhibitor; ADHF, Acute Decompensated Heart Failure; ARNI, Angiotensin Receptor-Neprilysin Inhibitor; BB, Beta-Blocker; CRT, Cardiac Resynchronization Therapy; DCM, Dilated Cardiomyopathy; EF, Ejection Fraction; GDMT, Guideline-Directed Medical Therapy; GLS, Global Longitudinal Strain; HCC, Hierarchical Condition Category; HCM, Hypertrophic Cardiomyopathy; HFrEF, Heart Failure with Reduced Ejection Fraction; HOCM, Hypertrophic Obstructive Cardiomyopathy; ICD, Implantable Cardioverter Defibrillator; LVEF, Left Ventricular Ejection Fraction; LVOT, Left Ventricular Outflow Tract; MEAT, Monitor, Evaluate, Assess, Treat; MRA, Mineralocorticoid Receptor Antagonist; NT-proBNP, N-Terminal Pro-Brain Natriuretic Peptide; NYHA, New York Heart Association; RCM, Restrictive Cardiomyopathy; SGLT2i, Sodium-Glucose Cotransporter 2 Inhibitor

Annual Clinical Review and Confirmation

Confirm phenotype, stage, and ongoing myocardial management.

- **Annual review**: Cardiomyopathy must be reassessed once per calendar year via face-to-face or synchronous audio-video encounter, with MEAT documented by 12/31
- **Visit modality**: In-person or synchronized audio-video telehealth encounters qualify when phenotype, severity, and management are addressed in accordance with current CMS parity standards
- **Clinical precision**: Documentation should specify cardiomyopathy phenotype (dilated, hypertrophic, restrictive) and clinical stage/severity (HFrEF/HFpEF, NYHA class). Under CMS HCC

V28, risk varies across HF and cardiomyopathy categories (HF ≈0.360; cardiomyopathy ≈0.189), reinforcing accurate characterization of current disease status

Good Documentation is Comprehensive Coding

If documentation shows…	Strengthen it by documenting…
Nonspecific diagnosis	"Chronic dilated cardiomyopathy (I42.0), Stage C, NYHA class III, LVEF 28%"
Unclear chronicity	"Non-ischemic DCM diagnosed (date); symptoms persistent >6 months; stable on GDMT"
Limited objective support	Dated NT-proBNP, LVEF trend, GLS measurement, or cardiac MRI findings
Unclear treatment rationale	Link therapy to functional status and prior response (eg, ARNI justified by persistent class III symptoms on ACEi)
Inconsistent severity signals	Align echo findings, exam (edema, JVP), and symptom reporting (orthopnea, PND) within a single NYHA class and stage

Abbreviations: DCM, Dilated Cardiomyopathy; NICM, Non-ischemic Cardiomyopathy; GDMT, Guideline-Directed Medical Therapy; NYHA, New York Heart Association; LVEF, Left Ventricular Ejection Fraction; GLS, Global Longitudinal Strain; CMR, Cardiac Magnetic Resonance; NT-proBNP, N-terminal pro-B-type Natriuretic Peptide; ARNI, Angiotensin Receptor-Neprilysin Inhibitor; ACEi, Angiotensin-Converting Enzyme inhibitor; TTE, Transthoracic Echocardiogram; JVP, Jugular Venous Pressure; PND, Paroxysmal Nocturnal Dyspnea; MRA, Mineralocorticoid Receptor Antagonist; SGLT2i, Sodium-Glucose Cotransporter-2 Inhibitor; HCC, Hierarchical Condition Category

EHR Tips

Feature	EHR Configuration Tip	Clinical & Quality Impact
Auto-Classification	Phenotype Specificity Tool: Prompts for the specific phenotype (Dilated, HCM, Restrictive) whenever "Cardiomyopathy" is entered	Ensures high-specificity coding for HCC 227 (V28 model), reducing the use of unspecified I42.9
Alert Systems	LVEF Surveillance Alert: Automatically flags if LVEF is ≤35% and no ICD or CRT is documented in the surgical history	Supports CMS Star Ratings and ensures primary prevention of Sudden Cardiac Death
BPA (Best Practice Advisory)	GDMT Gap Analysis: Triggers for Stage C HFrEF patients missing any of the "Four Pillars" (ARNI, BB, MRA, SGLT2i)	Reduces clinical inertia; ensures adherence to 2026 heart failure guidelines
Problem List Prompts	"Chronicity & Status" Hard-Stop: Mandatory selection of "Acute," "Chronic," or "Acute on Chronic" when adding I50.x codes	Satisfies the MEAT (Monitor, Evaluate, Assess, Treat) criteria for RADV risk adjustment
Outcome Tracking	KCCQ-12 Integration: Automated patient portal prompts for the Kansas City Cardiomyopathy Questionnaire prior to visits	Provides objective longitudinal data for MIPS and HEDIS reporting on patient quality of life

Abbreviations: ADHF, Acute Decompensated Heart Failure; ARNI, Angiotensin Receptor-Neprilysin Inhibitor; ATTR, Transthyretin Amyloidosis; BPA, Best Practice Advisory; DCM, Dilated Cardiomyopathy; EF, Ejection Fraction; EHR, Electronic Health Record; GDMT, Guideline-Directed Medical Therapy; GLS, Global Longitudinal Strain; HCC, Hierarchical Condition Category; HCM,

Feature	EHR Configuration Tip	Clinical & Quality Impact

Hypertrophic Cardiomyopathy; HFrEF, Heart Failure with Reduced Ejection Fraction; ICD, Implantable Cardioverter Defibrillator; KCCQ-12, Kansas City Cardiomyopathy Questionnaire; LVEF, Left Ventricular Ejection Fraction; LVOT, Left Ventricular Outflow Tract; MEAT, Monitor, Evaluate, Assess, Treat; MIPS, Merit-based Incentive Payment System; MRA, Mineralocorticoid Receptor Antagonist; NT-proBNP, N-Terminal Pro-Brain Natriuretic Peptide; NYHA, New York Heart Association; RCM, Restrictive Cardiomyopathy; SGLT2i, Sodium-Glucose Cotransporter 2 Inhibitor

Brief Case Examples

SUCCESS: Accurate Capture of Phenotype & Status

- **Case:** "68yo with chronic **Non-Ischemic Dilated Cardiomyopathy (I42.0)**, Stage C, currently **NYHA Class III**. Echo (01/12/2026): **LVEF 32%**, GLS -11%. Patient reports stable exertional dyspnea; denies PND/orthopnea. Continuing maximal **GDMT**: Entresto 97/103mg BID, Metoprolol Succinate 100mg QD, and Empagliflozin 10mg QD."
- **Why it Works:** Captures **HCC 227** (Cardiomyopathy) and **HCC 226** (Chronic Heart Failure)
  - **Estimated RAF (V28):** ~0.189 (CM) + ~0.360 (HF) = 0.549
  - **Financial Impact:** 0.549 x $10,402.34 = **Estimated $5,710.88/year**

PITFALL: The "Unspecified" Diagnostic Trap

- **Case:** "72yo male here for follow-up of **Cardiomyopathy** and heart failure. Symptoms are stable. Refilled heart medications. Will see back in 6 months."
- **Why it Fails:** Uses unspecified code (**I42.9**). In the 2026 V28 model, many unspecified cardiac codes do not map to a payment HCC
  - **Result: No RAF Capture**
  - **Financial Loss:** Loss of approximately **$5,710/year** in risk-adjusted funding due to lack of specificity and MEAT evidence

FIX: Re-establishing Specificity & Causal Linkage

- **Case:** "**Hypertrophic Obstructive Cardiomyopathy (I42.1)** stable; patient currently NYHA Class II. LVOT gradient remains **38 mmHg** per latest Echo. No lightheadedness or syncope reported. Continuing **Mavacamten 5mg QD** for symptom management and **Metoprolol** for rate control. BP 118/74, HR 62 today; K+ 4.1 stable"
- **Why it Works:** Recaptures **HCC 227**. It links the specific drug (Mavacamten) to the obstructive phenotype and provides objective data (LVOT gradient)
- **Estimated RAF (V28):** ~0.189 (CM) + ~0.360 (HF if linked) = **~0.549**
- **Financial Impact:** Adds approx. **$5,710/year** by satisfying the high-specificity requirements of the 2026 model

REFERENCES

1. What Is Cardiomyopathy? www.heart.org. Accessed January 20, 2026. https://www.heart.org/en/health-topics/cardiomyopathy/what-is-cardiomyopathy-in-adults

2. ICD-10 Version:2019. Accessed January 20, 2026. https://icd.who.int/browse10/2019/en#I42.4
3. Ommen SR, Ho CY, Asif IM, et al. 2024 AHA/ACC/AMSSM/HRS/PACES/SCMR Guideline for the Management of Hypertrophic Cardiomyopathy: A Report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines. *Circulation*. 2024;149(23). doi:10.1161/CIR.0000000000001250
4. Myers MC, Berge A, Zhong Y, et al. Prevalence and Incidence of Dilated Cardiomyopathy in the United States and Western Europe: A Systematic Review. *Cardiol Res*. 2025;16(4):295-305. doi:10.14740/cr2071
5. Writing Committee Members*, Kazi DS, Abdullah AR, et al. 2025 AHA/ACC Statement on Cost/Value Methodology in Clinical Practice Guidelines (Update From 2014 Statement): A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. 2025;152(18). doi:10.1161/CIR.0000000000001377
6. Wei C, Heidenreich PA, Sandhu AT. The economics of heart failure care. *Prog Cardiovasc Dis*. 2024;82:90-101.
7. Graziano F, Zorzi A, Cipriani A, et al. Contemporary diagnostic approach to arrhythmogenic cardiomyopathy: The three-step work-up. *Trends Cardiovasc Med*. 2025;35(2):107-113.
8. Brieler J, Breeden MA, Tucker J. Cardiomyopathy: an overview. *Am Fam Physician*. 2017;96(10):640-646.
9. Zipes DP. *Braunwald's Heart Disease e-Book: A Textbook of Cardiovascular Medicine*. Elsevier Health Sciences; 2018.
10. Hall JE, Hall ME, Guyton AC. Textbook of medical physiology. *No Title*. Published online 2011.
11. Fletcher GF, Ades PA, Kligfield P, et al. Exercise standards for testing and training: a scientific statement from the American Heart Association. *Circulation*. 2013;128(8):873-934.
12. Heidenreich PA, Bozkurt B, Aguilar D, et al. 2022 AHA/ACC/HFSA guideline for the management of heart failure: executive summary: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2022;79(17):1757-1780.
13. Darby SC, Ewertz M, McGale P, et al. Risk of ischemic heart disease in women after radiotherapy for breast cancer. *N Engl J Med*. 2013;368(11):987-998.
14. Lakatta EG, Levy D. Arterial and cardiac aging: major shareholders in cardiovascular disease enterprises: Part I: aging arteries: a "set up" for vascular disease. *Circulation*. 2003;107(1):139-146.
15. Kannel WB. Blood pressure as a cardiovascular risk factor: prevention and treatment. *Jama*. 1996;275(20):1571-1576.
16. Jia G, Hill MA, Sowers JR. Diabetic cardiomyopathy: an update of mechanisms contributing to this clinical entity. *Circ Res*. 2018;122(4):624-638.
17. Page RL. Drugs that may cause or exacerbate heart failure: a scientific statement from the American Heart Association (vol 134, pg e32, 2016). *Circulation*. 2016;134(12):E261-E261.
18. Endorsed by the European Society of Intensive Care Medicine (ESICM), Authors/Task Force Members, Nieminen MS, et al. Executive summary of the guidelines on the diagnosis and treatment of acute heart failure: the Task Force on Acute Heart Failure of the European Society of Cardiology. *Eur Heart J*. 2005;26(4):384-416.
19. Nieminen MS, Böhm M, Cowie MR, et al. Executive summary of the guidelines on the diagnosis and treatment of acute heart failure: the Task Force on Acute Heart Failure of the European Society of Cardiology. *Eur Heart J*. 2005;26(4):384-416. doi:10.1093/eurheartj/ehi044
20. January CT, Wann LS, Calkins H, et al. 2019 AHA/ACC/HRS Focused Update of the 2014 AHA/ACC/HRS Guideline for the Management of Patients With Atrial Fibrillation. *J Am Coll Cardiol*. 2019;74(1):104-132. doi:10.1016/j.jacc.2019.01.011
21. Shen WK, Sheldon RS, Benditt DG, et al. 2017 ACC/AHA/HRS guideline for the evaluation and management of patients with syncope: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *J Am Coll Cardiol*. 2017;70(5):620-663.
22. Yancy CW, Jessup M, Bozkurt B, et al. 2017 ACC/AHA/HFSA focused update of the 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology/American

Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. *J Am Coll Cardiol*. 2017;70(6):776-803.
23. Priori SG, Blomström-Lundqvist C, Mazzanti A, et al. 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC). Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). *Eur Heart J*. 2015;36(41):2793-2867. doi:10.1093/eurheartj/ehv316
24. Amsterdam EA, Wenger NK, Brindis RG, et al. 2014 AHA/ACC Guideline for the Management of Patients With Non–ST-Elevation Acute Coronary Syndromes: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation*. 2014;130(25). doi:10.1161/CIR.0000000000000134
25. Burns E, Buttner R, Buttner EB and R. Dilated Cardiomyopathy (DCM). Life in the Fast Lane • LITFL. August 1, 2018. Accessed January 14, 2026. https://litfl.com/dilated-cardiomyopathy-dcm-ecg-library/
26. Pogodina VV. Elizaveta Nilolaevna Levkovich-75th birthday. *Acta Virol*. 1975;19(6):509.
27. Basit H, Alahmadi MH, Rout P, Sharma S. [Figure, Hypertrophic Cardiomyopathy Treatment Mapping. The...]. June 7, 2024. Accessed January 20, 2026. https://www.ncbi.nlm.nih.gov/books/NBK430788/figure/article-18919.image.f2/
28. Gruner C, Care M, Siminovitch K, et al. Sarcomere protein gene mutations in patients with apical hypertrophic cardiomyopathy. *Circ Cardiovasc Genet*. 2011;4(3):288-295. doi:10.1161/CIRCGENETICS.110.958835
29. Guigui SA, Torres C, Escolar E, Mihos CG. Systolic anterior motion of the mitral valve in hypertrophic cardiomyopathy: a narrative review. *J Thorac Dis*. 2022;14(6):2309-2325. doi:10.21037/jtd-22-182
30. Burns E, Buttner R, Buttner EB and R. Restrictive Cardiomyopathy. Life in the Fast Lane • LITFL. August 1, 2018. Accessed January 14, 2026. https://litfl.com/restrictive-cardiomyopathy-ecg-library/
31. Muchtar E, Blauwet LA, Gertz MA. Restrictive Cardiomyopathy: Genetics, Pathogenesis, Clinical Manifestations, Diagnosis, and Therapy. *Circ Res*. 2017;121(7):819-837. doi:10.1161/CIRCRESAHA.117.310982
32. Restrictive Cardiomyopathy: Practice Essentials, Background, Pathophysiology. Published online August 26, 2025. Accessed January 20, 2026. https://emedicine.medscape.com/article/153062-overview?form=fpf
33. Rapezzi C, Aimo A, Barison A, et al. Restrictive cardiomyopathy: definition and diagnosis. *Eur Heart J*. 2022;43(45):4679-4693. doi:10.1093/eurheartj/ehac543
34. Parakh N, Mehrotra S, Seth S, et al. NT pro B type natriuretic peptide levels in constrictive pericarditis and restrictive cardiomyopathy. *Indian Heart J*. 2015;67(1):40-44. doi:10.1016/j.ihj.2015.02.008
35. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the Task Force Criteria. *Eur Heart J*. 2010;31(7):806-814. doi:10.1093/eurheartj/ehq025
36. Malik N, Mukherjee M, Wu KC, et al. Multimodality Imaging in Arrhythmogenic Right Ventricular Cardiomyopathy. *Circ Cardiovasc Imaging*. 2022;15(2):e013725. doi:10.1161/CIRCIMAGING.121.013725
37. Doshi S, Ramakrishnan S, Gupta SK. Invasive hemodynamics of constrictive pericarditis. *Indian Heart J*. 2015;67(2):175-182. doi:10.1016/j.ihj.2015.04.011
38. Petersen SE, Selvanayagam JB, Wiesmann F, et al. Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. *J Am Coll Cardiol*. 2005;46(1):101-105. doi:10.1016/j.jacc.2005.03.045
39. Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. *Heart Br Card Soc*. 2001;86(6):666-671. doi:10.1136/heart.86.6.666
40. Cole CR, Blackstone EH, Pashkow FJ, Snader CE, Lauer MS. Heart-Rate Recovery Immediately after Exercise as a Predictor of Mortality. *N Engl J Med*. 1999;341(18):1351-1357. doi:10.1056/NEJM199910283411804
41. Phelan D, Collier P, Thavendiranathan P, et al. Relative apical sparing of longitudinal strain using two-dimensional speckle-tracking echocardiography is both sensitive and specific for the diagnosis of cardiac amyloidosis. *Heart Br Card Soc*. 2012;98(19):1442-1448. doi:10.1136/heartjnl-2012-302353

42. Gertz MA, Benson MD, Dyck PJ, et al. Diagnosis, Prognosis, and Therapy of Transthyretin Amyloidosis. *J Am Coll Cardiol*. 2015;66(21):2451-2466. doi:10.1016/j.jacc.2015.09.075
43. Dispenzieri A, Gertz MA, Kyle RA, et al. Serum Cardiac Troponins and N-Terminal Pro-Brain Natriuretic Peptide: A Staging System for Primary Systemic Amyloidosis. *J Clin Oncol*. 2004;22(18):3751-3757. doi:10.1200/JCO.2004.03.029
44. Halliday BP, Wassall R, Lota AS, et al. Withdrawal of pharmacological treatment for heart failure in patients with recovered dilated cardiomyopathy (TRED-HF): an open-label, pilot, randomised trial. *Lancet Lond Engl*. 2019;393(10166):61-73. doi:10.1016/S0140-6736(18)32484-X
45. Wilcox JE, Fang JC, Margulies KB, Mann DL. Heart Failure With Recovered Left Ventricular Ejection Fraction: JACC Scientific Expert Panel. *J Am Coll Cardiol*. 2020;76(6):719-734. doi:10.1016/j.jacc.2020.05.075
46. Maurer MS, Schwartz JH, Gundapaneni B, et al. Tafamidis treatment for patients with transthyretin amyloid cardiomyopathy. *N Engl J Med*. 2018;379(11):1007-1016.
47. Garcia-Pavia P, Rapezzi C, Adler Y, et al. Diagnosis and treatment of cardiac amyloidosis: a position statement of the ESC Working Group on Myocardial and Pericardial Diseases. *Eur Heart J*. 2021;42(16):1554-1568. doi:10.1093/eurheartj/ehab072
48. Pinto YM, Elliott PM, Arbustini E, et al. Proposal for a revised definition of dilated cardiomyopathy, hypokinetic non-dilated cardiomyopathy, and its implications for clinical practice: a position statement of the ESC working group on myocardial and pericardial diseases. *Eur Heart J*. 2016;37(23):1850-1858. doi:10.1093/eurheartj/ehv727
49. Maron BJ, Pelliccia A. The heart of trained athletes: cardiac remodeling and the risks of sports, including sudden death. *Circulation*. 2006;114(15):1633-1644. doi:10.1161/CIRCULATIONAHA.106.613562
50. Vogel-Claussen J, Rochitte CE, Wu KC, et al. Delayed enhancement MR imaging: utility in myocardial assessment. *Radiogr Rev Publ Radiol Soc N Am Inc*. 2006;26(3):795-810. doi:10.1148/rg.263055047
51. Birnie DH, Sauer WH, Bogun F, et al. HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. *Heart Rhythm*. 2014;11(7):1305-1323. doi:10.1016/j.hrthm.2014.03.043
52. McMurray JJV, Solomon SD, Inzucchi SE, et al. Dapagliflozin in Patients with Heart Failure and Reduced Ejection Fraction. *N Engl J Med*. 2019;381(21):1995-2008. doi:10.1056/NEJMoa1911303
53. Ponikowski P, Kirwan BA, Anker SD, et al. Ferric carboxymaltose for iron deficiency at discharge after acute heart failure: a multicentre, double-blind, randomised, controlled trial. *Lancet Lond Engl*. 2020;396(10266):1895-1904. doi:10.1016/S0140-6736(20)32339-4
54. Biondi B. Mechanisms in endocrinology: Heart failure and thyroid dysfunction. *Eur J Endocrinol*. 2012;167(5):609-618. doi:10.1530/EJE-12-0627
55. Chung F, Yegneswaran B, Liao P, et al. STOP questionnaire: a tool to screen patients for obstructive sleep apnea. *Anesthesiology*. 2008;108(5):812-821. doi:10.1097/ALN.0b013e31816d83e4
56. Pitt B, Zannad F, Remme WJ, et al. The Effect of Spironolactone on Morbidity and Mortality in Patients with Severe Heart Failure. *N Engl J Med*. 1999;341(10):709-717. doi:10.1056/NEJM199909023411001
57. Kouranos V, Sharma R. Cardiac sarcoidosis: state-of-the-art review. *Heart Br Card Soc*. 2021;107(19):1591-1599. doi:10.1136/heartjnl-2019-316442

Medical Disclaimer

The information provided by the American Academy of Value-Based Care (AAVBC) in this document, including but not limited to coding guidance, and related content, is intended for educational and informational purposes only. This content is designed to assist healthcare providers, organizations, and professionals in understanding and applying Value-Based Care principles, practices, and regulatory compliance standards.

AAVBC does not provide legal, clinical, or medical advice. The content presented should not be interpreted or relied upon as specific legal, medical, clinical, or professional guidance. While efforts are made to ensure the accuracy and

currency of the information provided, AAVBC does not guarantee that the materials presented are complete, comprehensive, or without error.

Providers and healthcare professionals must independently evaluate all content and guidance presented herein in the context of applicable laws, regulations, clinical guidelines, payer policies, patient-specific conditions, and contraindications. Any treatments, procedures, diagnostic approaches, medications, or strategies discussed are illustrative and should not be directly applied without a thorough and independent review of current, evidence-based clinical resources, and regulatory requirements.

For coding, documentation, utilization management, quality measures (including STAR ratings), and compliance matters, users are responsible for consulting official guidelines issued by regulatory bodies, including but not limited to the Centers for Medicare & Medicaid Services (CMS), the American Medical Association (AMA), relevant state agencies, and other authoritative entities.

AAVBC expressly disclaims liability for any loss, claim, or damages arising directly or indirectly from the use or reliance on information provided in this document. Users should consult their organization's legal counsel, compliance officer, or qualified professional advisor for advice specific to their situation.

By accessing and using this document, you agree to AAVBC's terms and acknowledge that the use of the content is at your own risk and discretion.