American College of Radiology
ACR Appropriateness Criteria®

Nonischemic Myocardial Disease with Clinical Manifestations (Ischemic Cardiomyopathy Already Excluded)

Variant: 1 Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
US echocardiography transthoracic stress	May Be Appropriate	O
CT heart function and morphology with IV contrast	May Be Appropriate	☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
Arteriography coronary	Usually Not Appropriate	☢☢☢
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢
MRI chest without and with IV contrast	Usually Not Appropriate	O
MRI chest without IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without and with IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without IV contrast	Usually Not Appropriate	O
MRI heart with function and vasodilator stress perfusion without and with IV contrast	Usually Not Appropriate	O
CT chest with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without IV contrast	Usually Not Appropriate	☢☢☢
CT coronary calcium	Usually Not Appropriate	☢☢☢
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢

Variant: 2 Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	May Be Appropriate	O
CT heart function and morphology with IV contrast	May Be Appropriate	☢☢☢☢
FDG-PET/CT heart	May Be Appropriate	☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transthoracic stress	Usually Not Appropriate	O
Arteriography coronary	Usually Not Appropriate	☢☢☢
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢
MRI chest without and with IV contrast	Usually Not Appropriate	O
MRI chest without IV contrast	Usually Not Appropriate	O

MRI heart with function and inotropic stress without and with IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without IV contrast	Usually Not Appropriate	O
MRI heart with function and vasodilator stress perfusion without and with IV contrast	Usually Not Appropriate	O
CT chest with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without IV contrast	Usually Not Appropriate	☢☢☢
CT coronary calcium	Usually Not Appropriate	☢☢☢
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢

Variant: 3 Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CT heart function and morphology with IV contrast	May Be Appropriate	☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transthoracic stress	Usually Not Appropriate	O
Arteriography coronary	Usually Not Appropriate	☢☢☢
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢
MRI chest without and with IV contrast	Usually Not Appropriate	O
MRI chest without IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without and with IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without IV contrast	Usually Not Appropriate	O
MRI heart with function and vasodilator stress perfusion without and with IV contrast	Usually Not Appropriate	O
CT chest with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without IV contrast	Usually Not Appropriate	☢☢☢
CT coronary calcium	Usually Not Appropriate	☢☢☢
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢

Variant: 4 Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CT heart function and morphology with IV contrast	May Be Appropriate	☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transthoracic stress	Usually Not Appropriate	O
Arteriography coronary	Usually Not Appropriate	☢☢☢

Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢
MRI chest without and with IV contrast	Usually Not Appropriate	O
MRI chest without IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without and with IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without IV contrast	Usually Not Appropriate	O
MRI heart with function and vasodilator stress perfusion without and with IV contrast	Usually Not Appropriate	O
CT chest with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without IV contrast	Usually Not Appropriate	☢☢☢
CT coronary calcium	Usually Not Appropriate	☢☢☢
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢

Variant: 5 Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	May Be Appropriate	O
CT heart function and morphology with IV contrast	May Be Appropriate	☢☢☢☢
FDG-PET/CT heart	May Be Appropriate	☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transthoracic stress	Usually Not Appropriate	O
Arteriography coronary	Usually Not Appropriate	☢☢☢
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢
MRI chest without and with IV contrast	Usually Not Appropriate	O
MRI chest without IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without and with IV contrast	Usually Not Appropriate	O
MRI heart with function and inotropic stress without IV contrast	Usually Not Appropriate	O
MRI heart with function and vasodilator stress perfusion without and with IV contrast	Usually Not Appropriate	O
CT chest with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without IV contrast	Usually Not Appropriate	☢☢☢
CT coronary calcium	Usually Not Appropriate	☢☢☢
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢

Panel Members

Prabhakar Rajiah, MD[1]; Jacobo Kirsch, MD, MBA[b]; Michael A. Bolen, MD[c]; Juan C. Batlle, MD, MBA[d]; Richard KJ Brown, MD[e]; Christopher J. Francois, MD[f]; Mauricio S. Galizia, MD[g]; Kate Hanneman, MD, MPH[h]; Joao R. Inacio, MD[i]; Amer M. Johri, MD, MSc[j]; Daniel C. Lee, MD, MSc[k]; Satinder P. Singh, MD[l]; Todd C. Villines, MD[m]; Samuel Wann, MD[n]; Stefan L. Zimmerman, MD[o];

Suhny Abbara, MD.[p]

Summary of Literature Review

Introduction/Background

Nonischemic cardiomyopathies (NICMs) encompass a broad spectrum of disorders of the myocardium associated with mechanical or electrical dysfunction leading to inappropriate ventricular hypertrophy or dilation, without evidence of ischemia [1]. Generally, valvular, hypertensive, and congenital diseases are treated separately from the NICM discussed here. The myocardial involvement can be either primary (genetic, acquired, or mixed) or secondary to a systemic disease process [2]. NICM can also be classified into distinct morphological and functional types, each of which can be subclassified as familial or nonfamilial types [3]. In this document, we have adapted this classification with five variants of nonischemic myocardial diseases: 1) hypertrophic cardiomyopathy (HCM); 2) restrictive cardiomyopathy or infiltrative diseases; 3) dilated cardiomyopathy (DCM) or unclassified cardiomyopathy; 4) arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin); and 5) inflammatory cardiomyopathy [2]. With increasing availability and use of genetics, it is now known that cardiomyopathies do not fit into specific morphological and functional phenotypes as discussed above, and there is tremendous genetic heterogeneity. The recently proposed MOGE(S) nosology system provides a more comprehensive classification of cardiomyopathies, describing the morphofunctional phenotype (M), organ (O), genetic inheritance pattern (G), etiological annotation (E), and functional status (S) [4].

NICM has an approximate prevalence of 0.02% with an annual death rate of 25,000 in the United States [2]. In adults, the prevalence of HCM is 1:250 to 500, DCM is 1:250 to 500, and arrhythmogenic right ventricular cardiomyopathy (ARVD) is 1:2,000 to 5,000 [5], whereas these are uncommon in children. Clinical presentation is variable, including heart failure (HF), arrhythmia, sudden death, and acute chest pain. Common presentations include dyspnea, edema, ascites, chest discomfort palpitations, and syncope. In patients with clinical HF, a primary cardiomyopathy is diagnosed in 2% to 15% of patients, whereas in some large-scale trials, patients with nonischemic HF accounted for 18% to 53% of the study population [6]. Acute presentation with chest pain, elevated cardiac enzymes, and abnormal electrocardiogram (ECG) may be seen in inflammatory cardiomyopathies. Unlike ischemic cardiomyopathy, the pathophysiology of NICM is usually unclear and multifactorial, the functional consequences are global, the prognosis is better, and the therapeutic response is different [2].

In patients presenting with HF, imaging is utilized to establish that the symptoms and signs are due to HF, to quantify the ejection fraction (EF), to distinguish patients with reduced EF from those with preserved EF, and to evaluate for ischemia as an etiology. Imaging for HF is discussed in detail in the ACR Appropriateness Criteria® topics on "Suspected New-Onset and Known Nonacute Heart Failure" [7] and "Dyspnea–Suspected Cardiac Origin" [8].

The primary role of imaging in NICM is to characterize the disease and establish the specific etiology, which is essential for determining optimal management. Although patients with NICM require general treatment for HF or arrhythmia, therapy is often tailored, depending on the etiology. For example, iron-overload cardiomyopathy is treated with chelation therapy; cardiac sarcoidosis is treated with high-dose corticosteroids; cardiac amyloidosis is treated with

chemotherapy for light-chain amyloidosis (AL type) and novel therapies for transthyretin type; Fabry disease is treated with enzyme replacement therapy; and severe HCM or endomyocardial fibrosis is treated with surgery [2]. An endomyocardial biopsy may be required for definitive diagnosis in some cases; however, it is an invasive procedure and the yield may be low because of the patchy nature of disease processes. In unexplained cardiomyopathy, the final diagnosis based on biopsy differed from initial diagnosis in 31% of patients, and endomyocardial biopsy made the final diagnosis in 75% of these cases [9]. Imaging is also helpful for quantification of the disease process, risk stratification, prognosis, and monitoring response to therapy.

Special Imaging Considerations

For the purposes of distinguishing between CT and CT angiography (CTA), ACR Appropriateness Criteria topics use the definition in the ACR–NASCI–SIR–SPR Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA) [10]:

> "*CTA uses a thin-section CT acquisition that is timed to coincide with peak arterial or venous enhancement. The resultant volumetric dataset is interpreted using primary transverse reconstructions as well as multiplanar reformations and 3-D renderings.*"

All elements are essential: 1) timing, 2) reconstructions/reformats, and 3) 3-D renderings. Standard CTs with contrast also include timing issues and reconstructions/reformats. Only in CTA, however, is 3-D rendering a **required** element. This corresponds to the definitions that the CMS has applied to the Current Procedural Terminology codes.

Chest Radiography

Chest radiography can provide information on HF and vascular abnormalities; however, there is no specific role for radiography in characterizing the different types of NICM.

Echocardiography

Echocardiography provides information on ventricular function (global/regional, systolic/diastolic), volumes, mass, thickness, as well as valvular function. The morphology can be assessed, although it is limited in the evaluation of the right ventricle (RV). With the use of advanced techniques such as 3-D echocardiography, further subtyping of NICM is possible. Myocardial deformation can be evaluated using tissue Doppler imaging and speckle-tracking (2-D or 3-D). Abnormal global longitudinal strain enables detection of subclinical left ventricle (LV) dysfunction in several disease entities [11]. Doppler metrics are useful in evaluation of diastolic dysfunction, especially in restrictive cardiomyopathies and HCM [12]. However, routine echocardiography does not have tissue characterization capabilities. Contrast echocardiography can be used in the quantification of ventricular volumes and EF as well as regional wall motion when the routine images are suboptimal. It is also used in the evaluation of noncompaction, thrombus aneurysm, and apical lesions such as apical variant HCM, stress-induced cardiomyopathy, and endocardial fibroelastosis [13].

Nuclear Medicine Techniques

Single-photon emission computed tomography (SPECT) and PET myocardial perfusion imaging using thallium-201, Tc-99m-sestamibi/tetrofosmin, and Rb-82 are used to evaluate myocardial ischemia and exclude it as an etiology of the cardiomyopathy. Cardiac function can be quantified using Tc-99m–labeled human albumin serum or red blood cell radionuclide ventriculography, or

SPECT with Tc-99m-sestamibi/tetrofosmin or thallium-201 [14]. Nuclear medicine techniques are also useful in the evaluation of some types of NICM. Gallium-67 (Ga-67), thallium-201, and fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG)-PET are used in the evaluation of cardiac sarcoidosis. Tc-pyrophosphate (PYP), Tc-3,3,-diphosphon-1,2, propanodicarboxylic acid (DPD), I-123 serum amyloid P component, Pittsburgh compound B, and 18F-florbetapir are used in the diagnosis of cardiac amyloidosis [14]. In-111 monoclonal anti-myosin antibody can be used in the diagnosis of acute myocarditis [14]. There are some experimental isotopes that are useful in the evaluation of autonomic innervation and molecular mechanisms of HF, but their applications are still evolving [14].

Cardiac CT

Coronary CTA has a limited role in the evaluation of NICM, predominantly for excluding coronary artery disease (CAD) as the etiology of HF [15]. Cardiac CT can be used in the evaluation of morphology, characterization, and quantification of function in patients when echocardiography is suboptimal because of poor acoustic windows and MRI is suboptimal because of artifacts. The function and volumes obtained from CT correlate with other modalities including MRI [16,17]. With retrospective ECG-gated acquisition, dynamic and functional information can be obtained. First-pass myocardial perfusion can be used to evaluate for ischemia. Delayed iodine-enhancement imaging can show variable patterns of enhancement in NICM, albeit at a lower contrast-to-noise ratio compared with MRI. Similar to MRI, extracellular volume (ECV) can be quantified with CT either using a single- or dual-energy CT technique [18]. CT strain imaging to quantify regional function [19] and CT evaluation of diastolic function are not routinely used in clinical practice [20]. Coronary calcium score is used for risk stratification of CAD in asymptomatic patients and does not have a specific role in evaluation of NICM.

Cardiac MRI

MRI provides information on different facets of NICM using multiple sequences. The balanced steady-state free precession cine sequence is used to evaluate cardiac morphology, which helps in narrowing the etiology of NICM (thickening, thinning, apical ballooning, and prominent trabeculations). MRI is ideal for evaluation of areas that are visually limited in echocardiography such as the LV apex, LV lateral wall, LV basal septum, and the RV [21]. Cardiac function, volumes, and mass can be accurately quantified with high reproducibility. Regional function, which is abnormal in the early stages of several disease processes, can be quantified by several techniques of strain imaging including feature tracking. Real-time cine imaging can be used to exclude other causes such as constrictive pericarditis. Cardiac valvular function can be qualitatively evaluated in cine imaging and quantified in velocity encoded 2-D– or 4-D–phase contrast sequences.

MRI may be helpful in establishing the etiology of NICM. Different patterns of late gadolinium enhancement (LGE) are seen in NICM (linear mid myocardial, patchy mid myocardial, subepicardial, RV insertion point, diffuse subendocardial) [22]. Regardless of etiology, the extent of LGE predicts the risk of developing malignant arrhythmia and HF [21]. MRI can be used to guide endomyocardial biopsy if required. Early gadolinium enhancement (EGE) using T1-weighted spin-echo or fast spin-echo sequences evaluates capillary hyperemia, which is increased in acute inflammatory processes. T2-weighted images are useful in evaluating for myocardial edema. Parametric mapping techniques including T1, T2*, and T2-mapping as well as MR fingerprinting can characterize and quantify fibrosis, edema, iron, deposition, fatty infiltration, and amyloid deposition [23]. T1-mapping can be performed without intravenous (IV) contrast ("native”), which is

useful in patients with renal dysfunction. ECV can be quantified using native and postcontrast T1-mapping along with hematocrit value. ECV is increased in several disorders. T2-mapping is useful in inflammatory processes, whereas T2*-mapping is useful in cases of iron overload [23,24]. These mapping techniques are often more sensitive and reproducible compared with LGE techniques, and they can track changes with therapy [21]. Stress imaging, either with dynamic first-pass perfusion imaging (physiological or pharmacological) or with administration of dobutamine, is used to exclude myocardial ischemia as an etiology. MR angiographic sequences with or without IV contrast can be used to evaluate associated vascular abnormalities. Advanced technologies in cardiac MRI include MR spectroscopy, diffusion tensor imaging, elastography, quantitative myocardial blood flow, and PET/MRI [21]. MRI can now be performed on most pacemakers/implantable cardioverter defibrillators (ICDs) [25-28]. Technical adjustments and use of appropriate sequences are required to obtain good-quality cardiac MRI in patients with indwelling pacemakers/ICDs. For example, use of wide-band inversion recovery sequences can mitigate artifacts expected in an LGE sequence [29,30].

Coronary Arteriography

Coronary arteriography is used to evaluate CAD as a cause of HF, especially in high-risk patients. Right and left heart catheterizations are useful in pulmonary hypertension, providing cardiac hemodynamics and prognostic value. Right heart and simultaneous right and left heart catheterization is useful in distinguishing restrictive cardiomyopathies from constrictive pericarditis [31]. A ventriculogram can be used to evaluate associated regional wall motion abnormalities (RWMA). Endomyocardial biopsy is used to establish etiology in cases that are indeterminate after imaging.

Initial Imaging Definition

Initial imaging is defined as imaging at the beginning of the care episode for the medical condition defined by the variant. More than one procedure can be considered usually appropriate in the initial imaging evaluation when:

- There are procedures that are equivalent alternatives (i.e., only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

- There are complementary procedures (i.e., more than one procedure is ordered as a set or simultaneously wherein each procedure provides unique clinical information to effectively manage the patient's care).

Discussion of Procedures by Variant

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.

HCM is an inherited myocardial hypertrophy with heterogeneous phenotypic expression (asymmetric septal, apical, mid ventricular, lateral wall, mass-like, and concentric types) [32]. Patients with HCM can present with diastolic dysfunction, LV outflow tract (LVOT) obstruction, ischemic chest pain, arrhythmias, or sudden cardiac death [33]. Occasionally, clinical symptoms are produced by papillary muscle abnormalities (anomalous chordal attachment to the base of anterior

leaflet, double bifid muscles, apical displacement, hypermobility, elongated anterior mitral leaflet) without significant myocardial hypertrophy [34]. Asymptomatic family members of HCM often undergo imaging as a screening test.

The concentric type of HCM can be challenging to distinguish from concentric hypertrophy (caused by hypertension, aortic stenosis, and/or coarctation), infiltrative disorders, and athlete's heart. "Phenocopy" conditions mimic HCM, including Anderson-Fabry disease, glycogen storage diseases, lysosomal storage diseases, and mitochondrial diseases [33]. Anderson-Fabry disease is an X-linked storage disorder of glycosphingolipid metabolism due to α-galactosidase deficiency that manifests as LV thickening, diastolic dysfunction, RWMA, and myocardial fibrosis. Danon disease is an X-linked dominant lysosomal storage disorder due to mutation of lysosomal associated protein-2. Danon disease manifests as concentric LV thickening, cardiac failure, and arrhythmia. Of those patients diagnosed with HCM, Fabry disease was ultimately shown to be the etiology in 6% to 12% of patients [35], and Danon disease was ultimately shown to be the etiology in 4% of patients [33]. Athlete's heart is an adaptive hypertrophy of the heart. HCM is generally evaluated with history, clinical examination, ECG, and imaging tests.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
A. Arteriography Coronary

There is no relevant literature to support the use of coronary arteriography for the evaluation of HCM.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
B. Arteriography Coronary with Ventriculography

There is no relevant literature to support the use of coronary arteriography with ventriculography for the evaluation of HCM.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
C. CT Chest

There is no relevant literature to support the use of CT chest for the evaluation of HCM.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
D. CT Coronary Calcium

There is no relevant literature to support the use of CT coronary calcium for the evaluation of HCM.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
E. CT Heart Function and Morphology

Cardiac CT can be used in the evaluation of morphology and function in patients with suboptimal echocardiography. CT can provide accurate measurements of myocardial thickness. Myocardial fibrosis can be demonstrated and quantified in delayed-enhancement images with substantial agreement with MRI [36-38].

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
F. CTA Coronary

There is no relevant literature to support the use of CTA in the evaluation of HCM when ischemic cardiomyopathy has already been excluded.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
G. FDG-PET/CT Heart

There is no relevant literature to support the use of FDG-PET/CT heart for the evaluation of HCM.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
H. MRI Chest

There is no relevant literature to support the use of MRI chest for the evaluation of HCM.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
I. MRI Heart Function and Morphology

MRI provides comprehensive information for the evaluation of HCM, including the morphology, location, distribution, and extent of hypertrophy and fibrosis [39]. MRI is superior to echocardiography in recognizing areas of segmental hypertrophy, which may be missed or underestimated with echocardiography, particularly the LV apex, the RV anterior free wall, and the LV inferoseptum [39,40]. RV hypertrophy is seen in a third of patients [40]. MRI is more accurate than echocardiography in quantifying the myocardial thickness, which is an important prognostic indicator for myectomy [39]. LVOT obstruction (seen in one-third of patients with HCM and provocable in another third), systolic anterior motion of the mitral valve and mitral regurgitation may be seen in asymmetric basal septal type of HCM [33], although the quantification of flow acceleration due to LVOT obstruction is inferior when using MRI compared with echocardiography. MRI also helps in risk stratification and identification of patients who will benefit from primary prevention with ICD, primarily by the use of LGE. LGE is seen in up to 50% to 80% of HCM patients, with the extent of LGE correlating directly with adverse prognosis [39]. HCM patients with LGE have a 7-fold risk for nonsustained ventricular tachycardia, and extensive LGE >15% of LV mass is a marker for sudden death [39]. Apical aneurysm and massive hypertrophy >30 mm are also high-risk factors for sudden cardiac death [39]. Elevated native T1 and ECV measurements may be seen in HCM. One study showed that native T1 has 100% sensitivity, 96% specificity, and 98% accuracy in distinguishing healthy from diseased myocardium, including HCM [41,42]. Another advantage of MRI is its ability to evaluate papillary muscle abnormalities, which require different surgical management [34,43]. MRI is also useful in follow-up after treatment such as myectomy or septal ablation. MRI is used to screen family members with myocardial crypts, elongated mitral leaflets, delayed relaxation, high EF, and LGE seen in gene-positive, phenotype-negative patients [40].

MRI can distinguish HCM from its mimics. Compared with concentric-type HCM, hypertension has milder thickening (<1.6 cm), lower EF (HCM often produces a hyperdynamic high EF), dilated LV (normal or small chamber size in HCM), absent or minimal LGE, lower T1 and ECV, increased LV wall stress (lower LV wall stress in HCM), and lower anteroseptal systolic strain (lower longitudinal systolic strain in HCM) [41]. Myocardial crypts, elongated mitral leaflets, and LGE are seen in gene-positive, phenotype-negative patients [40]. Compared with HCM, athlete's heart has mild concentric hypertrophy, mild LV dilation (<6.5 cm), normal EF, and lacks other findings typical of HCM such as LGE, systolic anterior motion of the mitral valve, and diastolic dysfunction. Athlete's heart usually improves following deconditioning for 3 months. Normal perfusion and normal

movement of myocardial grids with myocardial tagging distinguishes mass-like HCM from neoplasms.

Anderson-Fabry disease presents with concentric LV thickening but may occasionally be asymmetric. Mid myocardial or subepicardial LGE is seen in the basal inferolateral segment of the LV, unlike HCM, wherein LGE is seen anywhere. There is no systolic anterior motion of the mitral valve or LVOT obstruction in Anderson-Fabry disease [35]. Low native T1 values are seen in Fabry disease because of sphingolipid deposition, often prior to the onset of structural and functional abnormalities [44]. With development of fibrosis, long native T1 and elevated ECV values can be seen. High T2 values, RV involvement, valve thickening, and lower global longitudinal strain can also be seen [45]. Danon disease also shows concentric LV thickening with edema and stress perfusion defect. LGE is usually in a mid myocardial distribution, less often in a subendocardial and transmural pattern in anterolateral and inferior segments of the LV, often with sparing of septum [46,47].

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
J. MRI Heart Inotropic Stress

There is no relevant literature to support the use of MRI heart inotropic stress for the evaluation of HCM.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
K. MRI Heart Vasodilator Stress

Reduced myocardial perfusion due to microvascular dysfunction is a poor prognostic factor in HCM. This may be seen even in areas without LGE, both in adults and children [48,49]. There is no evidence to support the use of MRI heart vasodilator stress for the evaluation of HCM when ischemic cardiomyopathy has already been excluded.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
L. Echocardiography Transesophageal

There is no relevant literature to support the use of transesophageal echocardiography for the evaluation of HCM.

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
M. Echocardiography Transthoracic Resting

Echocardiography is usually the initial imaging test in most patients with HCM. It is used in the evaluation of morphology, distribution, and quantification of HCM. Contrast echocardiography improves characterization of apical type of HCM [50]. Echocardiography is the preferred technique for the quantification of LVOT pressure gradient, which is a factor in selecting patients for myomectomy, as well as the assessment of systolic anterior motion, mitral regurgitation, and papillary muscle abnormalities. It can quantify LV systolic function, diastolic function, and left atrial volume. Decreased myocardial strain can be identified using speckle-tracking echocardiography [50]. Stress maneuvers, including a Valsalva maneuver in sitting, semisupine, and standing positions can be used to provoke LVOT gradients that may not be seen in resting states [51]. It is also the first-line test for screening, such as in the risk assessment of sudden cardiac death in competitive athletes [50].

Variant 1: Suspected hypertrophic cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
N. Echocardiography Transthoracic Stress

Exercise stress echo is used to assess provocable LVOT gradient if resting gradient is not severe. It is also useful in the assessment of worsening mitral regurgitation [51].

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.

Infiltrative disease is characterized by deposition of abnormal substances in the myocardium, resulting in myocardial thickening or dilation and restricted ventricular filling. Amyloidosis, Anderson-Fabry disease, acute sarcoidosis, Danon disease, endomyocardial fibrosis, oxalosis, mucopolysaccharidoses, and Friedrich ataxia result in myocardial thickening, whereas chronic sarcoidosis, scleroderma, and iron overload result in myocardial thinning [52]. Cardiac amyloidosis is usually of AL type or transthyretin-related amyloidosis (ATTR type), resulting in myocardial and valvular thickening and presenting with HF or arrhythmia. Myocardial involvement occurs in 25% of patients with systemic sarcoidosis in the United States [53]. Cardiac sarcoidosis is characterized by myocardial infiltration with noncaseating granulomas and presents with conduction abnormalities, arrhythmias, sudden cardiac death, HF, pericardial effusion, or ventricular aneurysms [2]. Diagnosis is based on the Japanese Ministry of Health and Welfare guidelines [54] or expert consensus recommendations [55]. Siderotic cardiomyopathy is characterized by iron deposition from frequent blood transfusions and altered iron hemostasis in hemoglobinopathy patients. Siderotic cardiomyopathy presents in advanced stages with HF, conduction abnormalities, or sudden death [2]. Scleroderma involves the heart in 80% (in autopsy) of cases, manifesting as HF, arrhythmia, CAD, peripheral vascular disease, or sudden death [2]. Endomyocardial fibrosis (Loeffler endocarditis in nontropical regions) is a spectrum of hyperesoinophlic syndrome (eosinophils >1,500/mm^3; >6 months), with cardiac involvement seen in 50% of these patients [56]. There is an early necrotic phase followed by thrombotic and fibrotic phases. Myocardial oxalosis presents with LV thickening, heart block, and conduction abnormalities. Friedreich ataxia is characterized by mitochondrial iron accumulation, with cardiomyopathy seen in 63% of these patients [52]. Mucopolysaccharidoses has variable phenotypic expression. Infiltrative disease is generally evaluated with history, clinical examination, ECG, serology, and imaging tests [52]. Endomyocardial biopsy may be ultimately required for definitive diagnosis.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
A. Arteriography Coronary

There is no relevant literature to support the use of coronary arteriography for the evaluation of restrictive cardiomyopathy.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
B. Arteriography Coronary with Ventriculography

There is no relevant literature to support the use of coronary arteriography with ventriculography for infiltrative cardiac diseases. Right heart catheterization is used for the evaluation of hemodynamics, which is useful in the diagnosis of pulmonary hypertension and has prognostic value. In addition, right and left heart catheterization can be used to evaluate for constrictive pericarditis, which often has to be distinguished from restrictive cardiomyopathy.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic

cardiomyopathy already excluded. Initial imaging.
C. CT Chest

There is no relevant literature to support the use of CT chest for evaluation of restrictive cardiomyopathy. CT chest may show mediastinal lymphadenopathy and lung changes in systemic sarcoidosis. Pericardial calcification points toward a pericardial constriction rather than restrictive cardiomyopathy.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
D. CT Coronary Calcium

There is no relevant literature to support the use of CT coronary calcium for the evaluation of restrictive cardiomyopathy. Incidental pericardial calcification points toward a pericardial constriction rather than restrictive cardiomyopathy.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
E. CT Heart Function and Morphology

Abnormal first-pass perfusion, delayed iodine enhancement, and high ECV values have been shown with CT in cardiac amyloidosis [57,58]. Subepicardial or mid myocardial delayed iodine enhancement has also been shown to identify cardiac sarcoidosis [59]. Pericardial calcification points toward a pericardial constriction rather than restrictive cardiomyopathy.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
F. CTA Coronary

There is no relevant literature to support the use of coronary CTA for the evaluation of restrictive cardiomyopathy when ischemic cardiomyopathy has already been excluded.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
G. Nuclear Medicine

Tc-DPD and Tc-PYP have high specificity in the diagnosis of cardiac amyloidosis. Ga-67 scintigraphy shows high uptake in cardiac sarcoidosis, with the intensity correlating with degree of inflammation [55]; however, it has low sensitivity [55]. Perfusion defects seen in thallium-201 and Tc-99m myocardial scintigraphy, as well as Rb-82, can be distinguished from ischemia by using PET/CT [55].

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
H. FDG-PET/CT Heart

FDG-PET performed after suppressing normal glucose metabolism shows high uptake in cardiac sarcoidosis, with reverse distribution in thallium-201 scans. FDG-PET had an 82% to 100% specificity and 39% to 91% specificity in the diagnosis of cardiac sarcoidosis [54]. FDG has higher sensitivity than Ga-67 scintigraphy, although Ga-67 scintigraphy is included in the imaging criteria [54]. A meta-analysis showed 89% sensitivity, 78% specificity, and area under the receiver operator characteristic curve of 93% for diagnosis of cardiac sarcoidosis [60]. The FDG activity can be quantified to improve the diagnostic accuracy, assess disease activity, and evaluate prognosis [60]. Simultaneous PET/MRI has been shown to be feasible with diagnostic image quality to evaluate cardiac sarcoidosis [61].

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
I. MRI Chest

There is no relevant literature to support the use of MRI chest for the evaluation of restrictive cardiomyopathy. MRI chest may show mediastinal lymphadenopathy and lung changes in systemic sarcoidosis.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
J. MRI Heart Function and Morphology

MRI has distinctive appearances in several infiltrative disorders and restrictive cardiomyopathies. Cardiac amyloidosis produces concentric thickening of ventricles, atria, interatrial septum, and valves, with low signal in T2-weighted images [62]. Diffuse subendocardial LGE is seen in early stages, which progresses to transmural LGE. Abnormal LGE has a pooled specificity of 92% and sensitivity of 85% in the diagnosis of cardiac amyloidosis [63]. Dark blood pool and earlier nulling of myocardium is also seen in cardiac amyloidosis. High native T1 and ECV values are more sensitive than LGE and reliably distinguish amyloidosis from HCM [64]. MRI can distinguish AL and ATTR types, with ATTR amyloidosis showing more LV thickening and mass, lower left ventricular ejection fraction (LVEF), greater LGE, more transmural LGE, and lower T1 values than the AL type [64]. An LGE-based scoring system was shown to have 87% sensitivity and 96% specificity in distinguishing AL and ATTR amyloidosis [64]. LGE, T1, and ECV abnormalities all correlate with prognosis in cardiac amyloidosis [65]. Transmural LGE is a predictor of adverse events including death [66]. Postcontrast difference in T1 between subepicardium and subendocardium of >23 ms predicts mortality with high accuracy [67]. Low T2 value and short T1 in >50% of myocardium T1 scout image are also poor prognostic factors [2].

MRI has sensitivity of 75% to 100% and specificity of 75% to 77% in the diagnosis of cardiac sarcoidosis [54,68,69]. In the acute stage, MRI shows wall thickening, high T2 signal (due to edema), high native T1 and T2 values, RWMA, and LGE. LGE is more common in the basal septal and lateral walls of the LV in a subepicardial or mid myocardial distribution. In the chronic stage, wall thinning, aneurysms, RWMA, and LGE may be seen (mid myocardial, subepicardial, and/or transmural) [70]. LGE correlates with prognosis with a hazard ratio of 32 for lethal events [71]. There is a good response to steroids in patients with lower LGE at initiation of therapy [72]. High native T1 and T2 values provide higher discriminatory accuracy compared with traditional criteria and help in evaluating the response to treatment [73].

Myocardial iron deposition can be reliably quantified using T2* techniques. Myocardial T2* <20 ms indicates significant iron deposition and <10 ms indicates advanced iron deposition with high accuracy [74]. T1-mapping is more reproducible and sensitive, with low T1 values seen in 32% of patients with normal T2* [75]. With appropriate use of chelation therapy, improvements in T2* and LVEF has been reported [76]. The use of MRI has resulted in improved outcomes with death rates declining to 2.3 per 1,000 compared with 7.9 per 1,000 prior to the use of MRI [76].

Linear mid myocardial LGE is seen in 66% of patients with scleroderma, either in the ventricular septum or the LV free wall at the basal and mid levels [77]. Patchy RV insertion enhancement can be seen in 17% of patients (76). LGE is more severe in patients with longer duration of Raynaud disease [78]. High native T1 and ECV values are seen in asymptomatic patients with no known

cardiac involvement, due to inflammation, and are associated with low diastolic and systolic strain rates [78]. Focal edema and fibrosis are also seen. Pericarditis, pericardial effusion, and adhesions may be seen.

In endomyocardial fibrosis, the apical wall is thickened and has a high T2 signal. A characteristic 3-layered pattern of LGE is seen with an inner layer of dark nonenhancing thrombus, middle layer of subendocardial LGE due to diffuse fibrosis (from LVOT to apex), and outer layer of nonenhancing normal myocardium [56,79]. LGE was associated with poor functional class and higher chance of surgery [56]. In Churg Strauss syndrome, LGE is seen in apical and mid segments and in anterior and anteroseptal segments in a subendocardial distribution. In myocardial oxalosis, concentric LV thickening and diastolic dysfunction are seen. In Friedreich ataxia, concentric or asymmetric LV thickening, diastolic dysfunction, and fibrosis may be seen. Mucopolysaccharidoses have variable expression, including asymmetric septal thickening, mitral or aortic valve pathologies, and normal EF [52].

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
K. MRI Heart Inotropic Stress

There is no relevant literature to support the use of MRI heart inotropic stress for the evaluation of restrictive cardiomyopathy. Ischemia has already been excluded.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
L. MRI Heart Vasodilator Stress

There is no relevant literature to support the use of MRI heart vasodilator stress for the evaluation of restrictive cardiomyopathy. Ischemia has already been excluded.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
M. Echocardiography Transesophageal

There is no relevant literature to support the use of transesophageal echo for the evaluation of restrictive cardiomyopathy.

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
N. Echocardiography Transthoracic Resting

Echocardiography is often the initial imaging test that identifies possible restrictive cardiomyopathy. Echocardiography can evaluate diastolic function with high accuracy, which is often impaired in early stages of several restrictive cardiomyopathies. Decreased systolic and diastolic mitral annular velocities, restrictive pattern in mitral valve such as high velocity (E wave), short deceleration time and low late diastolic filling (A wave), and elevated filling pressures (E/e ratio) are seen [12,80]. Before the widespread use of harmonic imaging, the myocardium was described as having a characteristic speckled (starry sky) appearance [64], a finding now considered obsolete with the widespread use of harmonic imaging techniques. A relative apical sparing of longitudinal strain of 1.0 (average apical longitudinal strain/average of basal and mid longitudinal strain) has a high sensitivity (93%) and specificity (82%) in distinguishing cardiac amyloidosis from controls [81]. Apical thickening may be seen in endomyocardial fibrosis. In sarcoidosis, echocardiography shows ventricular septal thickening and diastolic dysfunction in the

acute phase but thinning in the chronic phase with associated RWMA, aneurysm, and global systolic dysfunction [55].

Variant 2: Suspected restrictive cardiomyopathy or infiltrative disease. Ischemic cardiomyopathy already excluded. Initial imaging.
O. Echocardiography Transthoracic Stress

There is no relevant literature to support the use of echocardiography transthoracic stress for the evaluation of restrictive cardiomyopathy. Ischemia has already been excluded.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.

DCM is characterized by a dilated ventricle and global systolic dysfunction. Ischemia is the most common cause of DCM and is excluded as discussed above. Approximately 50% of nonischemic DCM is idiopathic and is usually seen in a younger age group [2]. Other etiologies include toxins, familial inheritance, infections, infiltrative disorders, autoimmune conditions, metabolic derangements, and arrhythmias. Alcoholic cardiomyopathy is seen in heavy drinkers with probable genetic susceptibility, more common in men 30 to 55 years of age [2,82]. Chemotherapeutic agents such as anthracyclines, tyrosine kinase inhibitors, trastuzumab, and interferons induce cardiomyopathy. There is a higher risk for cardiomyopathy with higher cumulative dose of chemotherapy, combination with other chemotherapeutic agents, associated radiation, and higher age. Acute cardiac changes can be seen as early as in a few hours after initiation, whereas late changes may be seen over decades with LV dilation and EF decrease, which limits the aggressive use of chemotherapy [83].

Peripartum cardiomyopathy is an idiopathic cardiomyopathy seen either in the late stage of pregnancy or in the first 5 months after delivery [84]. It is seen in 1 in 2,500 to 4,000 births in the United States [84]. Risk factors for peripartum cardiomyopathy include age >30 years, nonwhite background, multiparity, poor socioeconomic status, prolonged tocolytic therapy, hypertension, preeclampsia, and cocaine use [85]. These patients are evaluated with ECG, serological biomarkers, and imaging tests. Endomyocardial biopsy may be needed to exclude myocarditis. Several types of inherited muscular dystrophies can also produce DCM. These muscular dystrophies present with HF, arrhythmia, or sudden death by thromboembolism.

There are several unclassified NICMs. LV noncompaction is characterized by prominent trabeculations due to persistent embryonic sinusoids, leading to LV failure, thromboembolism, and arrhythmias [86]. Stress-induced cardiomyopathy (also known as Takotsubo cardiomyopathy) is characterized by transient LV systolic dysfunction attributed to catecholamine release, possibly following a stressful event. It presents similarly to acute myocardial infarction with chest pain, ST-segment elevation on ECG, and elevated cardiac enzymes. It accounts for 2% of myocardial infarction with nonobstructive coronary arteries (MINOCA) [87]. One study found that incidental CAD was found in 10% of patients with stress-induced cardiomyopathy [88]. Diagnosis is made based on the Mayo Clinic or InterTAK diagnostic criteria [89]. LVOT obstruction, arrhythmia, shock, ventricular rupture, thrombus, and death may also be seen [2]. Cardiomyopathy can be seen in cirrhotic patients, independent of alcohol exposure. Patients with DCM are evaluated with history, clinical examination, lab tests, ECG, coronary angiography, and imaging.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
A. Arteriography Coronary

There is no literature to support the use of coronary arteriography for the evaluation of nonischemic dilated or unclassified cardiomyopathy when ischemia has already been excluded. Stress-induced cardiomyopathy has been reported to be triggered by acute myocardial ischemia [90].

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
B. Arteriography Coronary with Ventriculography

There is no relevant literature to support the use of coronary arteriography with ventriculography for the evaluation of nonischemic dilated or unclassified cardiomyopathy when ischemia has already been excluded. If performed, RWMA not explained by a culprit lesion may be seen in left ventriculography [90]. With LV apical ballooning patterns and normal coronaries on CTA or coronary angiography, stress-induced cardiomyopathy can be confirmed, except in patients with red flags for acute myocarditis, in which MRI is indicated.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
C. CT Chest

There is no relevant literature to support the use of CT chest for the evaluation of nonischemic dilated or unclassified cardiomyopathy.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
D. CT Coronary Calcium

There is no relevant literature to support the use of CT coronary calcium for the evaluation of nonischemic DCM or unclassified cardiomyopathy.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
E. CT Heart Function and Morphology

CT can be used for morphological and functional evaluation in patients in whom echocardiogram is suboptimal. CT is accurate in distinguishing idiopathic from ischemic DCM [91]. CT has been shown to be accurate in the diagnosis and characterization of LV noncompaction using the standard MRI criteria of end-diastolic noncompacted LV myocardial thickness to compacted LV myocardial thickness ratio of >2.3 [92]. CT can show the abnormalities of stress-induced cardiomyopathy, including absence of delayed enhancement [93,94].

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
F. CTA Coronary

There is no relevant literature to support the use of CTA coronary for the evaluation of nonischemic DCM or unclassified cardiomyopathy when ischemic cardiomyopathy has already been excluded.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
G. Nuclear Medicine

A multiple-uptake gated acquisition scan can be used to measure LV dysfunction in patients with chemotherapy cardiomyopathy [83]. In the classic clinical setting, there is no need for nuclear

medicine techniques in stress-induced cardiomyopathy. Perfusion imaging shows mild diminished perfusion. Metabolic imaging using FDG-PET and SPECT I-123-β-methyl-iodophenyl pentadecanoic acid show reduced metabolism, and I-123-metaiodobenzylguanidine shows reduced sympathetic innervation [95].

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
H. FDG-PET/CT Heart

There is no relevant literature to support the use of FDG-PET/CT as the first-line imaging modality in the evaluation of nonischemic or unclassified cardiomyopathy. One study found that nearly 50% of patients with unexplained cardiomyopathy and arrhythmia demonstrate focal inflammation in FDG-PET/CT, which is indicative of inflammatory cardiomyopathy [96].

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
I. MRI Chest

There is no relevant literature to support the use of MRI chest for the evaluation of nonischemic DCM or unclassified cardiomyopathy.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
J. MRI Heart Function and Morphology

In DCM, MRI helps in establishing the etiology and quantifying the abnormalities. Dilated ventricles, secondary tricuspid and/or mitral regurgitation due to annular dilation, regional ventricular dysfunction, ventricular wall thinning, and eccentric remodeling are seen. Myocardial infarct is diagnosed if there is subendocardial or transmural pattern of LGE in a vascular distribution. In idiopathic DCM with nonobstructed coronary arteries, linear or patchy mid myocardial LGE, primarily at the base and mid septum, is seen in 28% of patients. No LGE is seen in 59%. Subendocardial LGE is seen in 13% of these patients that is either due to atypical nonischemic fibrosis or silent ischemia from coronary embolus or recanalized plaque rupture [97]. Using LGE, 19% of additional patients gained an indication for ICD, and 11% avoided a previously planned ICD compared with standard of care [98]. High T1 and ECV values show more sensitivity than LGE [68]. Native T1 value, ECV value, presence and extent of LGE, and EF correlate with adverse prognosis [99].

In chemotherapy cardiomyopathy, MRI helps in arbitrating discrepancies between imaging modalities, which may affect management. A reduction of EF by >10% or a reduction of EF >5% in symptomatic individuals is diagnostic of this entity (as long as the resultant EF is <53%) [83,100]. Early markers of cardiac involvement include elevated LV end-systolic volume (seen within 1 month); increased LV mass (due to edema); RWMA (decreased mid wall circumferential strain); high T1, T2, and ECV values; high signal in T2-weighted images (edema); and EGE [83]. Patients with edema are more likely to have right ventricular ejection fraction (RVEF) reduction at follow-up [100]. LGE can be seen in 0% to 100% of patients, either in mid myocardial or subepicardial distribution and rarely diffuse, indicating irreversible damage [101]. In late-onset cardiomyopathy in cancer survivors, abnormal or subnormal LVEF and RVEF, as well as high LV volumes without LGE, were seen at a median of 7.8 years after anthracycline therapy [102]. Increased ECV has been shown in cancer survivors [100]. LGE has been shown in 9% to 18% in mid myocardial, subepicardial, or RV insertion point distributions [101].

In peripartum cardiomyopathy, MRI provides additional information pertaining to diagnosis and prognosis, which are not obtained in echocardiography. Gadolinium contrast is avoided until after delivery. LV dilation, global LV systolic dysfunction, RV dysfunction, and LGE are seen [84]. LGE is seen in 40% in a subepicardial or mid myocardial distribution in the anterior and anterolateral LV segments (occasionally subendocardial or transmural), more commonly in scans taken >7 days after the acute phase [84]. High T1 and T2 values and EGE are also seen in the acute stages [103]. Patients with LGE showed higher decompensation and did not regain LVEF [84].

Muscular dystrophies may present with ventricular dilation, systolic dysfunction, and mid myocardial/subepicardial pattern of LGE, with occasional noncompacted myocardium. LGE may be present when echocardiography is still normal [104] and is an adverse prognostic determinant [84]. A higher amount of LGE is associated with lower LVEF, but LGE has a variable association with arrhythmia [104]. With longer duration of steroid treatment, lower increase in fibrosis burden was seen over time [105]. T1 and ECV values are also abnormal, which were associated with arrhythmia. Strain imaging shows abnormalities in earlier stages, before onset of overt HF, shows better serial decline in LV function, and provides reliable monitoring of progression of dystrophy [106]. LGE is also seen in mutation carriers [104].

In noncompaction, MRI shows a 2-layered structure of outer compacted and inner noncompacted myocardium, with the ratio of noncompacted to compacted myocardial thickness >2.3 in end diastole [107]. In borderline patients, additional metrics—such as trabecular mass >15 g/m^2, ratio of trabecular to total LV mass >20% to 25% [107], involvement of basal segments (with ratio >2), and at least one segment with ratio of >3.0—are helpful in diagnosing LV noncompaction with sensitivities and specificities up to 100% [86]. A poor prognosis with development of HF and arrhythmia can be expected with higher ratios and with LGE. LGE may be seen in the trabeculations as well as subendocardium [108,109]. On direct comparison with echocardiography, both at end diastole and end systole, MRI was shown to evaluate all the LV segments, provide a more accurate and reliable assessment of extent of noncompacted myocardium, and provide supplemental morphological information beyond that obtained from 2-D echocardiography [110]. There is better correlation of end-diastolic than end-systolic ratio between echocardiography and MRI [110], but the end-systolic ratio in MRI had stronger association with events, HF, and systolic dysfunction than end-diastolic measurements [111]. Recent studies have shown that a significant number (15%–43%) of asymptomatic subjects who are free from cardiovascular diseases satisfy the currently used MRI diagnostic criteria for noncompaction, indicating that these criteria have poor specificity. This may, therefore, represent a variant anatomical phenotype than cardiomyopathy [112,113].

MRI in stress-induced cardiomyopathy shows reversible global systolic dysfunction and LV apical ballooning with normal or hyperkinetic basal segments and akinetic/hypokinetic apical segments. There are also reverse and mid ventricular variants. RV is involved in 40% of cases, which is associated with a worse prognosis [114]. Myocardial edema may be present, leading to a high signal in T2-weighted images, high native T1, and high T2 values, typically confined to the abnormal segment [115]. Edema diffuses more than myocardial ischemia and decreases within a few weeks unlike myocardial ischemia, which may take up to 3 months to diminish [115]. Typically, there is no LGE. However, recent studies have shown that LGE may be present in up to 40% of patients, typically in the areas of RWMA. This is usually of lower signal intensity (<5 SD above

remote normal myocardium) than the LGE of myocardial infarction [87]. These patients may have irreversible damage with worse prognosis and longer recovery time [116,117]. MR diagnosis of stress-induced cardiomyopathy is made based on a typical pattern of LV dysfunction in a noncoronary pattern, myocardial edema corresponding to areas with RWMA, absence or insignificant LGE (<5 SD above remote normal myocardium), and markers for myocardial inflammation (EGE ratio > 4.0) [114]. MRI is superior to echocardiography in evaluating the RV involvement and complications [114]. Functional improvement occurs usually in 3 to 4 months but may take up to 12 months in 5% of patients; it may recur in 5% to 11% of patients [87]. Although stress-induced cardiomyopathy was typically thought to be completely reversible, recent literature indicates long-term clinical consequences [118].

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
K. MRI Heart Inotropic Stress

There is no relevant literature to support the use of MRI heart inotropic stress for the evaluation of nonischemic DCM or unclassified cardiomyopathy when ischemia has already been excluded.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
L. MRI Heart Vasodilator Stress

There is no relevant literature to support the use of MRI vasodilator stress for the evaluation of nonischemic DCM or unclassified cardiomyopathy when ischemia has already been excluded.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
M. Echocardiography Transesophageal

There is no relevant literature to support the use of transesophageal echocardiography for the evaluation of nonischemic DCM or unclassified cardiomyopathy.

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
N. Echocardiography Transthoracic Resting

Echocardiography can evaluate the function in several types of nonischemic DCM. It does not provide tissue characterization to identify the specific cause of cardiomyopathy, but the presence of systolic dysfunction in patients on chemotherapy, postpartum, and alcoholic patients is suggestive of cardiomyopathy.

In chemotherapy, a reduction of EF by >10% or a reduction of EF >5% in symptomatic individuals is diagnostic of this entity (as long as the resultant EF is <53%). Calculation of LVEF by 3-D echocardiography is more reproducible and accurate than by 2-D echocardiography and is preferred for the evaluation and longitudinal assessment of patients treated with chemotherapy [100]. A 10% to 15% reduction of peak systolic global longitudinal strain by speckle-tracking echocardiography is the most useful parameter to predict cardiotoxicity [119]. Global radial and circumferential strains are abnormal in late survivors, but their clinical value is less proven [119]. Decreased global longitudinal strain with preserved EF is the most common echocardiographic abnormality in cancer survivors [100]. Echocardiography can confirm, quantify, and detect associated abnormalities and complications, as well as risk stratify patients [120].

In LV noncompaction, echocardiography shows a 2-layered structure with prominent LV trabeculations (end-systolic ratio >2) and deep perfused intertrabecular recesses in color Doppler [86,121]. The sensitivity and reproducibility of echocardiography is improved by using LV contrast [122]. LV strain is decreased in noncompacted as well as compacted segments [122]. Transient reversible global systolic dysfunction as well as RWMA (apical, mid ventricular, basal, or focal in anterolateral segment) are seen in stress-induced cardiomyopathy [95]. Wall motion abnormalities show circular pattern in speckle echocardiography with improved detection using IV contrast [95].

Variant 3: Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
O. Echocardiography Transthoracic Stress

There is no relevant literature to support the use of echocardiography transthoracic stress for the evaluation of nonischemic DCM or idiopathic cardiomyopathy when ischemia has already been excluded.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.

ARVD is an inherited cardiomyopathy that primarily affects the RV, is characterized by fibro-fatty replacement of the myocardium, and may result in arrhythmias, biventricular dysfunction, and sudden cardiac death. This should be distinguished from a benign entity, RV outflow tract-ventricular tachycardia, which is associated with a structurally normal heart. Diagnosis of ARVD is made using the 2010 criteria, which includes investigation of family history, pathological tissue characterization, ECG depolarization abnormalities, ECG repolarization abnormalities, and RV wall motion abnormalities [123]. In diagnosed patients, family members should be screened [124]. Arrhythmia-induced cardiomyopathy refers to reversible HF and LV dysfunction in patients with tachycardias, atrial fibrillation, and premature ventricular contractions without an underlying heart disease [125]. This is a diagnosis of exclusion, made when the EF is low (<50%), with improvement of >15% following treatment for arrhythmia [126]. Arrhythmia-induced cardiomyopathy should be suspected in patients with a mean heart rate >100 beats/min, atrial fibrillation with rapid ventricular rate, and/or premature ventricular contractions ≥10% [125]. There is a correlation between the LV systolic dysfunction and the rate as well as duration of arrhythmia [126]. Patients with suspected arrhythmogenic cardiomyopathies are initially evaluated with medical history, family history, clinical examination, 12-lead ECG, signal-averaged ECG, exercise stress test, 24-hour Holter monitor, and imaging (echocardiography, MRI, or CT).

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
A. Arteriography Coronary

There is no relevant literature to support the use of coronary arteriography for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
B. Arteriography Coronary with Ventriculography

Imaging is initially performed with noninvasive tests such as MRI or CT. However, the 2010 criteria specifies RV angiographic criteria for ARVD, including regional RV akinesia, RV dyskinesia, or RV aneurysm [123].

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin).

Ischemic cardiomyopathy already excluded. Initial imaging.
C. CT Chest

There is no relevant literature to support the use of CT chest for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
D. CT Coronary Calcium

There is no relevant literature to support the use of CT coronary calcium for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
E. CT Heart Function and Morphology

ECG-gated cardiac CT shows wall motion abnormalities and allows quantification of ventricular volumes and function. RV myocardial fat may be seen but is nonspecific. A single study showed that a CT-based scoring system based on fatty tissue, bulging appearance, and dilation of RV had 87% sensitivity, 94.4% specificity, positive predictive value of 87%, negative predictive value of 94.4%, and accuracy of 92.2% for diagnosis of definitive ARVD [127].

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
F. CTA Coronary

There is no relevant literature to support the use of coronary CTA for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
G. FDG-PET/CT Heart

There is no relevant literature to support the use of FDG-PET/CT for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
H. MRI Chest

There is no relevant literature to support the use of MRI chest for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
I. MRI Heart Function and Morphology

On MRI, major RV wall motion abnormality (aneurysm, akinesis, dyskinesis, asynchronous contraction) with either low RVEF (<40%) or dilated RV (end-diastolic volume index >100 mL/m^2 in men; >100 mL/m^2 in women) is a major criterion for ARVD. Although RVEF in the 40% to 45% range and mildly dilated RV (end-diastolic volume index 100–110 mL/m^2 in men, 90–100 mL/m^2 in women) is a minor criterion for ARVD, according to the revised task force criteria [123,124]. The major criteria have 95% specificity, whereas the minor criteria have 85% to 97% specificity for diagnosis of ARVD [124]. Use of the new criteria has shown lower yield but higher positive predictive value [124]. Fat as well as LGE may be seen in the RV myocardium in up to 88% of

patients, reflecting fibro-fatty infiltration [124]. LV changes may also be seen, demonstrating higher association with ventricular arrhythmias. LV involvement is seen in 76% of ARVD, with some of them having LV-dominant disease. In LV-dominant disease, LGE is more common in the septum like RV-dominant disease, in which LGE is more common in the inferior and lateral LV walls [124]. RV strain by MRI can quantitatively identify regional dysfunction in ARVD and may detect preclinical disease [128,129]. MRI can distinguish ARVD from the benign RV outflow tract tachycardia by demonstrating larger RV diameter, more dispersed RV contraction, and lower RV function [130]. Tachycardia-induced cardiomyopathy shows LV systolic dysfunction that correlates with the rate and duration of tachycardia, with LGE seen in 5% of these patients [126]. MR studies have shown that two typical scar patterns—anteroseptal and inferolateral—account for 89% of arrhythmogenic substrates in NICM, with three distinct ventricular tachycardia morphologies [131]. MRI along with electrophysiological voltage mapping provides a roadmap for an atrial or pulmonary vein ablation procedure, and MRI identifies areas of nontransmural scar and gray zone not detected by traditional voltage mapping.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
J. MRI Heart Inotropic Stress

There is no relevant literature to support the use of MRI heart inotropic for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
K. MRI Heart Vasodilator Stress

There is no relevant literature to support the use of MRI heart vasodilator stress for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
L. Echocardiography Transesophageal

There is no relevant literature to support the use of transesophageal echocardiography for the evaluation of arrhythmogenic cardiomyopathies.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
M. Echocardiography Transthoracic Resting

Echocardiography is an initial imaging tool in patients with suspected ARVD and is used for frequent follow-up, particularly in patients with devices. ARVD is diagnosed based on the 2010 criteria of major wall motion abnormalities along with enlarged RV outflow tract (parasternal long axis ≥32 mm or parasternal short axis ≥36 mm) or decreased fractional area change (≤33%) [123,124]. Evaluation of the entire RV and quantification of function are challenging with 2-D echocardiography. Additional techniques include RV myocardial performance index, IV echocardiographic contrast, tricuspid annular plane systolic excursion (M-mode or tissue Doppler imaging), strain imaging, speckle-tracking, and 3-D echocardiography.

Variant 4: Suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin). Ischemic cardiomyopathy already excluded. Initial imaging.
N. Echocardiography Transthoracic Stress

There is no relevant literature to support the use of echocardiographic transthoracic stress for the

evaluation of arrhythmogenic cardiomyopathies.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.

Inflammatory myocardial disease can present either in acute or subacute fashion. Acute myocarditis is due to infections (viral, bacterial, fungal, or tuberculosis), toxins, drugs, injuries, or idiopathic etiology. It can present with acute chest pain, elevated cardiac enzymes, and ECG changes that may mimic acute coronary syndrome (MINOCA). Other presentations include LV dysfunction, arrhythmias, and sudden cardiac death. Acute myocarditis accounts for up to 75% of patients who present with MINOCA, 12% of those with sudden death, and 9% of DCM [132]. Patients may recover or progress to DCM. Sarcoidosis may occasionally present in an acute fashion similar to acute myocarditis. Myocarditis can also be seen in rheumatological diseases, such as rheumatoid arthritis, systemic lupus erythematosus, and systemic sclerosis [133].

Chagas diseases are caused by a parasite, *Trypanosoma cruzi*, which is endemic in Central and South America, with 13% of the population at risk and 11% affected [134]. Chagas disease has an acute, a long indeterminate, and a chronic cardiac phase, with one-third of seropositive individuals developing chronic heart disease [135]. Cardiac Chagas presents as HF, arrhythmia, heart block, sudden death, and thromboembolic events [2]. Human immunodeficiency virus can cause cardiomyopathy in 8% of asymptomatic individuals [2]. There is no single test that can accurately diagnose inflammatory cardiomyopathy. Patients with suspected inflammatory cardiomyopathy are evaluated using history, clinical examination, serology, ECG, and noninvasive imaging tests. Endomyocardial biopsy with histopathology, immunohistology, and molecular techniques may be necessary for diagnosis.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
A. Arteriography Coronary

There is no relevant literature to support the use of coronary arteriography for the evaluation of inflammatory myocardial disorders when ischemia has already been excluded.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
B. Arteriography Coronary with Ventriculography

There is no relevant literature to support the use of coronary arteriography for the evaluation of inflammatory myocardial disorders.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
C. CT Chest

There is no relevant literature to support the use of CT chest for the evaluation of inflammatory myocardial disorders.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
D. CT Coronary Calcium

There is no relevant literature to support the use of CT coronary calcium for the evaluation of inflammatory myocardial disorders.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already

excluded. Initial imaging.
E. CT Heart Function and Morphology

CT has been shown to display focal or multifocal enhancement and absence of coronary stenosis correlating with MRI [136].

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
F. CTA Coronary

There is no relevant literature to support the use of CTA coronary arteries for the evaluation of inflammatory myocardial disorders.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
G. FDG-PET/CT Heart

FDG-PET/CT may be useful in the evaluation of inflammatory cardiomyopathies, particularly in the evaluation of acute presentation of cardiac sarcoidosis [54,60]. FDG-PET/CT is not commonly used in the diagnosis of myocarditis. However, if performed, high uptake may be seen in FDG-PET/CT. In 111-antimyosin antibody can be used to identify myocarditis [14].

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
H. MRI Chest

There is no relevant literature to support the use of MRI chest for the evaluation of inflammatory myocardial disorders.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
I. MRI Heart Function and Morphology

In acute myocarditis, MRI is performed in patients with symptoms of myocarditis, evidence of myocardial injury, and suspected viral etiology. MRI has been shown to have an impact on making a decision in >50% of patients and provides a new diagnosis in 11% of patients [115]. One study showed that using MRI at a lower threshold in patients with MINOCA (ie, using MRI independent of clinical likelihood of myocarditis) led to a 6.3-fold increase in the incidence of myocarditis with doubling of MRIs positive for myocarditis, indicating that myocarditis is currently an underdiagnosed entity [137]. MRI shows functional abnormalities (global systolic dysfunction or focal wall motion abnormalities), capillary hyperemia (high signal in EGE), edema (high signal in T2-weighted images, high native T1 and T2 values, increased ECV), necrosis/fibrosis (LGE in mid myocardial/subepicardial; high T1 and ECV), and pericardial effusion. The Lake Louise criteria, which were used in the diagnosis of acute myocarditis, required two out of the three criteria (edema, EGE, and/or LGE) to be positive [138]. A combination of all three is required if high positive predictive value is desired (positive likelihood ratio of 7.7, accuracy of 80%, specificity of 90%, sensitivity of 77%, positive predictive value of 96%, and negative predictive value of 53%), whereas T2 or LGE criteria are adequate for high sensitivity (91% sensitivity, 84% accuracy) [138]. Removing EGE as a criterion does not change the accuracy (80% with, 84% without) but reduces sensitivity (90% with, 60% without) [138]. Native T1-mapping is useful in detecting subtle, focal disease with sensitivity of 90%, specificity of 91%, and accuracy of 91%, which is superior to T2-weighted MRI and LGE techniques [115]. The updated Lake Louise criteria requires at least one T2-based criterion (global/regional elevation of myocardial T2 or increased T2 signal of myocardium)

with at least one T1-based criterion (elevated myocardial T1, elevated ECV, or LGE) for diagnosing acute myocarditis with high specificity [139]. Having only one criterion will still support a diagnosis of acute myocarditis but has lower specificity than with two criteria [139]. Different LGE patterns have been reported based on the viral etiology, with parvovirus B19 showing subepicardial or mid myocardial distribution LGE in the LV inferolateral wall and recovering without serious damage, whereas HHV-6 infection involves the LV basilar septum in linear mid myocardial LGE pattern, often rapidly progressing to HF [140]. Myocardial edema without fibrosis indicates good potential for recovery, whereas a high amount of EGE and LGE indicate adverse prognosis, particularly if LGE is persistent at 4 weeks after onset [2]. LGE may not correlate with the clinical and lab markers, indicating it is an independent risk assessment tool [141]. A normal MRI in patients with suspected myocarditis indicates a good long-term prognosis, independent of clinical and other findings [142].

In Chagas disease, patients are typically not imaged in the acute phase, but the indeterminate phase may show changes including RWMA and diastolic dysfunction without overt systolic dysfunction. The chronic phase shows global systolic dysfunction, apical aneurysm, and thrombus. LGE is seen in up to 72% of patients [135] and in 100% of those with arrhythmias, more common in apical and basal inferolateral segments. LGE has been reported in all the phases, including early indeterminate [135]. LGE is subendocardial in 27%, transmural in 36%, mid myocardial in 14%, and subepicardial in 23% of patients [143]. Hence, the pattern is not specific, with contributions possibly from myocarditis and microvascular dysfunction. The diagnosis is therefore made in the context of appropriate epidemiological history [143]. EGE and myocardial edema similar to that of acute myocarditis can also be seen in all phases [135]. All these parameters correlated directly with disease severity [143].

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
J. MRI Heart Inotropic Stress

There is no relevant literature to support the use of MRI heart inotropic stress for the evaluation of inflammatory myocardial disorders. Ischemia has already been excluded.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
K. MRI Heart Vasodilator Stress

There is no relevant literature to support the use of MRI heart vasodilator stress for the evaluation of inflammatory myocardial disorders. Ischemia has already been excluded.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
L. Echocardiography Transesophageal

There is no relevant literature to support the use of echocardiography transesophageal for the evaluation of inflammatory myocardial disorders.

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
M. Echocardiography Transthoracic Resting

Echocardiography shows global and regional functional abnormalities in acute myocarditis. Pericardial effusion may also be seen. Echocardiography is a first-line imaging modality in the evaluation of Chagas disease. It may present with hypokinetic dilated LV with diminished LVEF or

biventricular dilation. Aneurysms, thrombus, and valvular disease (mitral and tricuspid regurgitation) may be seen [144]. Global longitudinal strain correlates with the amount of myocardial fibrosis in MRI [145].

Variant 5: Suspected inflammatory cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.
N. Echocardiography Transthoracic Stress

There is no relevant literature to support the use of echocardiography transthoracic stress for the evaluation of inflammatory myocardial disorders. Ischemia has already been excluded.

Summary of Recommendations

- **Variant 1**: MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, or ultrasound (US) echocardiography transthoracic resting, is usually appropriate for the initial imaging of patients with suspected HCM when ischemic cardiomyopathy has already been excluded. US echocardiography transthoracic resting is the initial imaging test for morphology, quantification, and hemodynamics. MRI with or without IV contrast provides accurate evaluation of morphology and quantification and assessment of papillary muscle abnormalities. MRI with IV contrast is used for tissue characterization and risk stratification based on fibrosis.
- **Variant 2**: MRI heart function and morphology without and with IV contrast or US echocardiography transthoracic resting is usually appropriate for the initial imaging of patients with suspected restrictive cardiomyopathy or infiltrative disease when ischemic cardiomyopathy has already been excluded. US echocardiography transthoracic resting is the first-line imaging modality that can detect infiltrative disease and quantitate diastolic function. MRI with IV contrast is used for tissue characterization and risk stratification.
- **Variant 3**: MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, or US echocardiography transthoracic resting, is usually appropriate for the initial imaging of suspected nonischemic dilated and unclassified cardiomyopathy when ischemic cardiomyopathy has already been excluded. US echocardiography transthoracic resting is the initial imaging modality for morphology and function. MRI with or without IV contrast also provides information on morphology and function. MRI with IV contrast is used for tissue characterization and risk stratification.
- **Variant 4**: MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, or US echocardiography transthoracic resting is usually appropriate for the initial imaging of suspected arrhythmogenic cardiomyopathy (arrhythmia of ventricular origin) when ischemic cardiomyopathy has already been excluded. US echocardiography transthoracic resting is the initial imaging modality for morphology and function. MRI with or without IV contrast also provides information on morphology and function. MRI with IV contrast is used for tissue characterization and risk stratification.
- **Variant 5**: MRI heart function and morphology without and with IV contrast or US echocardiography transthoracic resting is usually appropriate for the initial imaging of suspected inflammatory cardiomyopathy when ischemic cardiomyopathy has already been excluded. US echocardiography transthoracic resting is the initial imaging modality used for determination of morphology and function. MRI with or without IV contrast also provides information on morphology and function. MRI with IV contrast is used for tissue characterization and risk stratification.

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at https://acsearch.acr.org/list. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria.

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® Radiation Dose Assessment Introduction document.

Relative Radiation Level Designations

Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range

O	0 mSv	0 mSv
☢	<0.1 mSv	<0.03 mSv
☢☢	0.1-1 mSv	0.03-0.3 mSv
☢☢☢	1-10 mSv	0.3-3 mSv
☢☢☢☢	10-30 mSv	3-10 mSv
☢☢☢☢☢	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (e.g., region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies."

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The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of

this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.

[a]Mayo Clinic, Rochester, Minnesota. [b]Panel Chair, Cleveland Clinic Florida, Weston, Florida. [c]Panel Vice-Chair, Cleveland Clinic, Cleveland, Ohio. [d]Miami Cardiac and Vascular Institute and Baptist Health of South Florida, Miami, Florida. [e]University of Utah, Department of Radiology and Imaging Sciences, Salt Lake City, Utah; Commission on Nuclear Medicine and Molecular Imaging. [f]University of Wisconsin, Madison, Wisconsin. [g]University of Iowa Hospitals and Clinics, Iowa City, Iowa. [h]Toronto General Hospital, University of Toronto, Toronto, Ontario, Canada. [i]The Ottawa Hospital, University of Ottawa, Ottawa, Ontario, Canada. [j]Queen's University, Kingston, Ontario, Canada; American Society of Echocardiography. [k]Northwestern University Feinberg School of Medicine, Chicago, Illinois; Society for Cardiovascular Magnetic Resonance. [l]University of Alabama at Birmingham, Birmingham, Alabama. [m]University of Virginia Health System, Charlottesville, Virginia; Society of Cardiovascular Computed Tomography. [n]Wisconsin Heart Hospital, Milwaukee, Wisconsin; American Society of Nuclear Cardiology. [o]Johns Hopkins Medical Institute, Baltimore, Maryland. [p]Specialty Chair, UT Southwestern Medical Center, Dallas, Texas.