

**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
MRI heart function and morphology without and with IV contrast	Usually Appropriate	○
US echocardiography transthoracic resting	Usually Appropriate	○
MRI heart function and morphology without IV contrast	Usually Appropriate	○
CT heart function and morphology with IV contrast	May Be Appropriate	⊕⊕⊕⊕
US echocardiography transthoracic stress	May Be Appropriate	○
MRI heart with function and inotropic stress without and with IV contrast	Usually Not Appropriate	○
MRI heart with function and vasodilator stress perfusion without and with IV contrast	Usually Not Appropriate	○
US echocardiography transesophageal	Usually Not Appropriate	○
Arteriography coronary	Usually Not Appropriate	⊕⊕⊕
Arteriography coronary with ventriculography	Usually Not Appropriate	⊕⊕⊕
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	⊕⊕⊕
CTA coronary arteries with IV contrast	Usually Not Appropriate	⊕⊕⊕
FDG-PET/CT heart	Usually Not Appropriate	⊕⊕⊕⊕
MRI chest without and with IV contrast	Usually Not Appropriate	○
MRI chest without IV contrast	Usually Not Appropriate	○
MRI heart with function and inotropic stress without IV contrast	Usually Not Appropriate	○
CT coronary calcium	Usually Not Appropriate	⊕⊕⊕

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

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

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

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

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

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

NONISCHEMIC MYOCARDIAL DISEASE WITH CLINICAL MANIFESTATIONS (ISCHEMIC CARDIOMYOPATHY ALREADY EXCLUDED)

Expert Panel on Cardiac Imaging: Prabhakar Rajiah, MD^a; 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, MDⁱ; Amer M. Johri, MD, MSc^j; Daniel C. Lee, MD, MSc^k; Satinder P. Singh, MD^l; Todd C. Villines, MD^m; Samuel Wann, MDⁿ; 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

^aMayo Clinic, Rochester, Minnesota. ^bPanel Chair, Cleveland Clinic Florida, Weston, Florida. ^cPanel Vice-Chair, Cleveland Clinic, Cleveland, Ohio. ^dMiami Cardiac and Vascular Institute and Baptist Health of South Florida, Miami, Florida. ^eUniversity of Utah, Department of Radiology and Imaging Sciences, Salt Lake City, Utah. ^fUniversity of Wisconsin, Madison, Wisconsin. ^gUniversity of Iowa Hospitals and Clinics, Iowa City, Iowa. ^hToronto General Hospital, University of Toronto, Toronto, Ontario, Canada. ⁱThe Ottawa Hospital, University of Ottawa, Ottawa, Ontario, Canada. ^jQueen's University, Kingston, Ontario, Canada; Cardiology expert. ^kNorthwestern University Feinberg School of Medicine, Chicago, Illinois; Society for Cardiovascular Magnetic Resonance. ^lUniversity of Alabama at Birmingham, Birmingham, Alabama. ^mUniversity of Virginia Health System, Charlottesville, Virginia; Society of Cardiovascular Computed Tomography. ⁿWisconsin Heart Hospital, Milwaukee, Wisconsin; Nuclear cardiology expert. ^oJohns Hopkins Medical Institute, Baltimore, Maryland. ^pSpecialty Chair, UT Southwestern Medical Center, Dallas, Texas.

The American College of Radiology seeks and encourages collaboration with other organizations on the development of the ACR Appropriateness Criteria through representation of such organizations on expert panels. Participation on the expert panel does not necessarily imply endorsement of the final document by individual contributors or their respective organization.

Reprint requests to: publications@acr.org

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 (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient’s care)

OR

- There are complementary procedures (ie, more than one procedure is ordered as a set or simultaneously in which 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.

Arteriography Coronary

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

Arteriography Coronary with Ventriculography

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

CT Chest

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

CT Coronary Calcium

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

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].

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.

FDG-PET/CT Heart

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

MRI Chest

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

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 provokable 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].

MRI Heart Inotropic Stress

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

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.

Echocardiography Transesophageal

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

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].

Echocardiography Transthoracic Stress

Exercise stress echo is used to assess provokable 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 hyperesoinophilic syndrome (eosinophils $>1,500/\text{mm}^3$; >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. Friedrich 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.

Arteriography Coronary

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

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.

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.

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.

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.

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.

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].

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].

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.

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].

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.

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.

Echocardiography Transesophageal

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

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].

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.

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].

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.

CT Chest

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

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.

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].

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.

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].

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].

MRI Chest

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

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/m², 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].

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.

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.

Echocardiography Transesophageal

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

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].

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).

Arteriography Coronary

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

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].

CT Chest

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

CT Coronary Calcium

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

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].

CTA Coronary

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

FDG-PET/CT Heart

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

MRI Chest

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

MRI Heart Function and Morphology

On MRI, major RV wall motion abnormality (aneurysm, akinesia, dyskinesia, asynchronous contraction) with either low RVEF (<40%) or dilated RV (end-diastolic volume index >100 mL/m² in men; >100 mL/m² 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/m² in men, 90–100 mL/m² 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.

MRI Heart Inotropic Stress

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

MRI Heart Vasodilator Stress

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

Echocardiography Transesophageal

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

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 ($\leq 33\%$) [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.

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.

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.

Arteriography Coronary with Ventriculography

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

CT Chest

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

CT Coronary Calcium

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

CT Heart Function and Morphology

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

CTA Coronary

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

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].

MRI Chest

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

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].

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.

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.

Echocardiography Transesophageal

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

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].

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.

- **Variation 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.
- **Variation 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 go to www.acr.org/ac.

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 [146].

Relative Radiation Level Designations		
Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
○	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 (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as “Varies.”

References

1. Bluemke DA. MRI of nonischemic cardiomyopathy. *AJR Am J Roentgenol* 2010;195:935-40.
2. Rajiah P, Raza S, Saboo SS, Ghoshhajra B, Abbara S. Update on the Role of Cardiac Magnetic Resonance in Acquired Nonischemic Cardiomyopathies. *J Thorac Imaging* 2016;31:348-66.
3. Elliott P, Andersson B, Arbustini E, et al. Classification of the cardiomyopathies: a position statement from the European Society Of Cardiology Working Group on Myocardial and Pericardial Diseases. *Eur Heart J* 2008;29:270-6.
4. Arbustini E, Narula N, Tavazzi L, et al. The MOGE(S) classification of cardiomyopathy for clinicians. *J Am Coll Cardiol* 2014;64:304-18.
5. McKenna WJ, Maron BJ, Thiene G. Classification, Epidemiology, and Global Burden of Cardiomyopathies. *Circ Res* 2017;121:722-30.
6. Follath F. Nonischemic heart failure: epidemiology, pathophysiology, and progression of disease. *J Cardiovasc Pharmacol* 1999;33 Suppl 3:S31-5.
7. White RD, Kirsch J, Bolen MA, et al. ACR Appropriateness Criteria® Suspected New-Onset and Known Nonacute Heart Failure. *J Am Coll Radiol* 2018;15:S418-S31.
8. Vogel-Claussen J, Elshafee ASM, Kirsch J, et al. ACR Appropriateness Criteria® Dyspnea-Suspected Cardiac Origin. *J Am Coll Radiol* 2017;14:S127-S37.
9. Ardehali H, Qasim A, Cappola T, et al. Endomyocardial biopsy plays a role in diagnosing patients with unexplained cardiomyopathy. *Am Heart J* 2004;147:919-23.
10. American College of Radiology. ACR–NASCI–SIR–SPR Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA). Available at: <https://www.acr.org/-/media/ACR/Files/Practice-Parameters/body-cta.pdf>. Accessed September 30, 2020.
11. Chan J, Shiino K, Obonyo NG, et al. Left Ventricular Global Strain Analysis by Two-Dimensional Speckle-Tracking Echocardiography: The Learning Curve. *J Am Soc Echocardiogr* 2017;30:1081-90.
12. Nagueh SF, Smiseth OA, Appleton CP, et al. Recommendations for the Evaluation of Left Ventricular Diastolic Function by Echocardiography: An Update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. *J Am Soc Echocardiogr* 2016;29:277-314.
13. Porter TR, Mulvagh SL, Abdelmoneim SS, et al. Clinical Applications of Ultrasonic Enhancing Agents in Echocardiography: 2018 American Society of Echocardiography Guidelines Update. *J Am Soc Echocardiogr* 2018;31:241-74.
14. Harinstein ME, Soman P. Radionuclide Imaging Applications in Cardiomyopathies and Heart Failure. *Curr Cardiol Rep* 2016;18:23.
15. Kalisz K, Rajiah P. Computed tomography of cardiomyopathies. *Cardiovasc Diagn Ther* 2017;7:539-56.

16. Asferg C, Usinger L, Kristensen TS, Abdulla J. Accuracy of multi-slice computed tomography for measurement of left ventricular ejection fraction compared with cardiac magnetic resonance imaging and two-dimensional transthoracic echocardiography: a systematic review and meta-analysis. *Eur J Radiol* 2012;81:e757-62.
17. Greupner J, Zimmermann E, Grohmann A, et al. Head-to-head comparison of left ventricular function assessment with 64-row computed tomography, biplane left cineventriculography, and both 2- and 3-dimensional transthoracic echocardiography: comparison with magnetic resonance imaging as the reference standard. *J Am Coll Cardiol* 2012;59:1897-907.
18. Lee HJ, Im DJ, Youn JC, et al. Myocardial Extracellular Volume Fraction with Dual-Energy Equilibrium Contrast-enhanced Cardiac CT in Nonischemic Cardiomyopathy: A Prospective Comparison with Cardiac MR Imaging. *Radiology* 2016;280:49-57.
19. Buss SJ, Schulz F, Mereles D, et al. Quantitative analysis of left ventricular strain using cardiac computed tomography. *Eur J Radiol* 2014;83:e123-30.
20. Boogers MJ, van Werkhoven JM, Schuijf JD, et al. Feasibility of diastolic function assessment with cardiac CT: feasibility study in comparison with tissue Doppler imaging. *JACC Cardiovasc Imaging* 2011;4:246-56.
21. Captur G, Manisty C, Moon JC. Cardiac MRI evaluation of myocardial disease. *Heart* 2016;102:1429-35.
22. O'Donnell DH, Abbara S, Chaithiraphan V, et al. Cardiac MR imaging of nonischemic cardiomyopathies: imaging protocols and spectra of appearances. *Radiology* 2012;262:403-22.
23. Messroghli DR, Moon JC, Ferreira VM, et al. Clinical recommendations for cardiovascular magnetic resonance mapping of T1, T2, T2* and extracellular volume: A consensus statement by the Society for Cardiovascular Magnetic Resonance (SCMR) endorsed by the European Association for Cardiovascular Imaging (EACVI). *J Cardiovasc Magn Reson* 2017;19:75.
24. Moon JC, Messroghli DR, Kellman P, et al. Myocardial T1 mapping and extracellular volume quantification: a Society for Cardiovascular Magnetic Resonance (SCMR) and CMR Working Group of the European Society of Cardiology consensus statement. *J Cardiovasc Magn Reson* 2013;15:92.
25. Indik JH, Gimbel JR, Abe H, et al. 2017 HRS expert consensus statement on magnetic resonance imaging and radiation exposure in patients with cardiovascular implantable electronic devices. *Heart Rhythm* 2017;14:e97-e153.
26. Nazarian S, Hansford R, Roguin A, et al. A prospective evaluation of a protocol for magnetic resonance imaging of patients with implanted cardiac devices. *Ann Intern Med* 2011;155:415-24.
27. Russo RJ, Costa HS, Silva PD, et al. Assessing the Risks Associated with MRI in Patients with a Pacemaker or Defibrillator. *N Engl J Med* 2017;376:755-64.
28. Williamson BD, Gohn DC, Ramza BM, et al. Real-World Evaluation of Magnetic Resonance Imaging in Patients With a Magnetic Resonance Imaging Conditional Pacemaker System: Results of 4-Year Prospective Follow-Up in 2,629 Patients. *JACC Clin Electrophysiol* 2017;3:1231-39.
29. Rashid S, Rapacchi S, Vaseghi M, et al. Improved late gadolinium enhancement MR imaging for patients with implanted cardiac devices. *Radiology* 2014;270:269-74.
30. Stevens SM, Tung R, Rashid S, et al. Device artifact reduction for magnetic resonance imaging of patients with implantable cardioverter-defibrillators and ventricular tachycardia: late gadolinium enhancement correlation with electroanatomic mapping. *Heart Rhythm* 2014;11:289-98.
31. Hurrell DG, Nishimura RA, Higano ST, et al. Value of dynamic respiratory changes in left and right ventricular pressures for the diagnosis of constrictive pericarditis. *Circulation* 1996;93:2007-13.
32. Baxi AJ, Restrepo CS, Vargas D, Marmol-Velez A, Ocazonez D, Murillo H. Hypertrophic Cardiomyopathy from A to Z: Genetics, Pathophysiology, Imaging, and Management. *Radiographics* 2016;36:335-54.
33. Marian AJ, Braunwald E. Hypertrophic Cardiomyopathy: Genetics, Pathogenesis, Clinical Manifestations, Diagnosis, and Therapy. *Circ Res* 2017;121:749-70.
34. Patel P, Dhillon A, Popovic ZB, et al. Left Ventricular Outflow Tract Obstruction in Hypertrophic Cardiomyopathy Patients Without Severe Septal Hypertrophy: Implications of Mitral Valve and Papillary Muscle Abnormalities Assessed Using Cardiac Magnetic Resonance and Echocardiography. *Circ Cardiovasc Imaging* 2015;8:e003132.
35. De Cobelli F, Esposito A, Belloni E, et al. Delayed-enhanced cardiac MRI for differentiation of Fabry's disease from symmetric hypertrophic cardiomyopathy. *AJR Am J Roentgenol* 2009;192:W97-102.

36. Langer C, Lutz M, Eden M, et al. Hypertrophic cardiomyopathy in cardiac CT: a validation study on the detection of intramyocardial fibrosis in consecutive patients. *Int J Cardiovasc Imaging* 2014;30:659-67.
37. Zhao L, Ma X, Feuchtner GM, Zhang C, Fan Z. Quantification of myocardial delayed enhancement and wall thickness in hypertrophic cardiomyopathy: multidetector computed tomography versus magnetic resonance imaging. *Eur J Radiol* 2014;83:1778-85.
38. Zhao L, Ma X, Delano MC, et al. Assessment of myocardial fibrosis and coronary arteries in hypertrophic cardiomyopathy using combined arterial and delayed enhanced CT: comparison with MR and coronary angiography. *Eur Radiol* 2013;23:1034-43.
39. Maron MS. The role of cardiovascular magnetic resonance in sudden death risk stratification in hypertrophic cardiomyopathy. *Card Electrophysiol Clin* 2015;7:187-93.
40. Maron MS. Clinical utility of cardiovascular magnetic resonance in hypertrophic cardiomyopathy. *J Cardiovasc Magn Reson* 2012;14:13.
41. Puntmann VO, Jahnke C, Gebker R, et al. Usefulness of magnetic resonance imaging to distinguish hypertensive and hypertrophic cardiomyopathy. *Am J Cardiol* 2010;106:1016-22.
42. Puntmann VO, Voigt T, Chen Z, et al. Native T1 mapping in differentiation of normal myocardium from diffuse disease in hypertrophic and dilated cardiomyopathy. *JACC Cardiovasc Imaging* 2013;6:475-84.
43. Kwon DH, Setser RM, Thamilarasan M, et al. Abnormal papillary muscle morphology is independently associated with increased left ventricular outflow tract obstruction in hypertrophic cardiomyopathy. *Heart* 2008;94:1295-301.
44. Pica S, Sado DM, Maestrini V, et al. Reproducibility of native myocardial T1 mapping in the assessment of Fabry disease and its role in early detection of cardiac involvement by cardiovascular magnetic resonance. *J Cardiovasc Magn Reson* 2014;16:99.
45. Vijapurapu R, Nordin S, Baig S, et al. Global longitudinal strain, myocardial storage and hypertrophy in Fabry disease. *Heart* 2019;105:470-76.
46. Dara BS, Rusconi PG, Fishman JE. Danon disease: characteristic late gadolinium enhancement pattern on cardiac magnetic resonance imaging. *Cardiol Young* 2011;21:707-9.
47. Etesami M, Gilkeson RC, Rajiah P. Utility of late gadolinium enhancement in pediatric cardiac MRI. *Pediatr Radiol* 2016;46:1096-113.
48. Hernandez LE. Myocardial stress perfusion magnetic resonance in children with hypertrophic cardiomyopathy. *Cardiol Young* 2018;28:702-08.
49. Ismail TF, Hsu LY, Greve AM, et al. Coronary microvascular ischemia in hypertrophic cardiomyopathy - a pixel-wise quantitative cardiovascular magnetic resonance perfusion study. *J Cardiovasc Magn Reson* 2014;16:49.
50. Nagueh SF, Bierig SM, Budoff MJ, et al. American Society of Echocardiography clinical recommendations for multimodality cardiovascular imaging of patients with hypertrophic cardiomyopathy: Endorsed by the American Society of Nuclear Cardiology, Society for Cardiovascular Magnetic Resonance, and Society of Cardiovascular Computed Tomography. *J Am Soc Echocardiogr* 2011;24:473-98.
51. Elliott PM, Anastakis A, Borger MA, et al. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy: the Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). *Eur Heart J* 2014;35:2733-79.
52. Seward JB, Casclang-Verzosa G. Infiltrative cardiovascular diseases: cardiomyopathies that look alike. *J Am Coll Cardiol* 2010;55:1769-79.
53. Doughan AR, Williams BR. Cardiac sarcoidosis. *Heart* 2006;92:282-8.
54. Ohira H, Tsujino I, Ishimaru S, et al. Myocardial imaging with 18F-fluoro-2-deoxyglucose positron emission tomography and magnetic resonance imaging in sarcoidosis. *Eur J Nucl Med Mol Imaging* 2008;35:933-41.
55. Dubrey SW, Sharma R, Underwood R, Mittal T. Cardiac sarcoidosis: diagnosis and management. *Postgrad Med J* 2015;91:384-94.
56. Salemi VM, Rochitte CE, Shiozaki AA, et al. Late gadolinium enhancement magnetic resonance imaging in the diagnosis and prognosis of endomyocardial fibrosis patients. *Circ Cardiovasc Imaging* 2011;4:304-11.
57. Deux JF, Mihalache CI, Legou F, et al. Noninvasive detection of cardiac amyloidosis using delayed enhanced MDCT: a pilot study. *Eur Radiol* 2015;25:2291-7.

58. Treibel TA, Bandula S, Fontana M, et al. Extracellular volume quantification by dynamic equilibrium cardiac computed tomography in cardiac amyloidosis. *J Cardiovasc Comput Tomogr* 2015;9:585-92.
59. Aikawa T, Oyama-Manabe N, Naya M, et al. Delayed contrast-enhanced computed tomography in patients with known or suspected cardiac sarcoidosis: A feasibility study. *Eur Radiol* 2017;27:4054-63.
60. Youssef G, Leung E, Mylonas I, et al. The use of 18F-FDG PET in the diagnosis of cardiac sarcoidosis: a systematic review and metaanalysis including the Ontario experience. *J Nucl Med* 2012;53:241-8.
61. Hanneman K, Kadoch M, Guo HH, et al. Initial Experience With Simultaneous 18F-FDG PET/MRI in the Evaluation of Cardiac Sarcoidosis and Myocarditis. *Clin Nucl Med* 2017;42:e328-e34.
62. Wassmuth R, Abdel-Aty H, Bohl S, Schulz-Menger J. Prognostic impact of T2-weighted CMR imaging for cardiac amyloidosis. *Eur Radiol* 2011;21:1643-50.
63. Zhao L, Tian Z, Fang Q. Diagnostic accuracy of cardiovascular magnetic resonance for patients with suspected cardiac amyloidosis: a systematic review and meta-analysis. *BMC Cardiovasc Disord* 2016;16:129.
64. Dzungu JN, Valencia O, Pinney JH, et al. CMR-based differentiation of AL and ATTR cardiac amyloidosis. *JACC Cardiovasc Imaging* 2014;7:133-42.
65. Karamitsos TD, Piechnik SK, Banyersad SM, et al. Noncontrast T1 mapping for the diagnosis of cardiac amyloidosis. *JACC Cardiovasc Imaging* 2013;6:488-97.
66. Fontana M, Pica S, Reant P, et al. Prognostic Value of Late Gadolinium Enhancement Cardiovascular Magnetic Resonance in Cardiac Amyloidosis. *Circulation* 2015;132:1570-9.
67. Maceira AM, Prasad SK, Hawkins PN, Roughton M, Pennell DJ. Cardiovascular magnetic resonance and prognosis in cardiac amyloidosis. *J Cardiovasc Magn Reson* 2008;10:54.
68. Freeman AM, Curran-Everett D, Weinberger HD, et al. Predictors of cardiac sarcoidosis using commonly available cardiac studies. *Am J Cardiol* 2013;112:280-5.
69. Smedema JP, Snoep G, van Kroonenburgh MP, et al. Evaluation of the accuracy of gadolinium-enhanced cardiovascular magnetic resonance in the diagnosis of cardiac sarcoidosis. *J Am Coll Cardiol* 2005;45:1683-90.
70. Giesbrandt KJ, Bolan CW, Shapiro BP, Edwards WD, Mergo PJ. Diffuse diseases of the myocardium: MRI-pathologic review of cardiomyopathies with dilatation. *AJR Am J Roentgenol* 2013;200:W274-82.
71. Greulich S, Deluigi CC, Gloekler S, et al. CMR imaging predicts death and other adverse events in suspected cardiac sarcoidosis. *JACC Cardiovasc Imaging* 2013;6:501-11.
72. Ise T, Hasegawa T, Morita Y, et al. Extensive late gadolinium enhancement on cardiovascular magnetic resonance predicts adverse outcomes and lack of improvement in LV function after steroid therapy in cardiac sarcoidosis. *Heart* 2014;100:1165-72.
73. Puntmann VO, Isted A, Hinojar R, Foote L, Carr-White G, Nagel E. T1 and T2 Mapping in Recognition of Early Cardiac Involvement in Systemic Sarcoidosis. *Radiology* 2017;285:63-72.
74. Anderson LJ, Holden S, Davis B, et al. Cardiovascular T2-star (T2*) magnetic resonance for the early diagnosis of myocardial iron overload. *Eur Heart J* 2001;22:2171-9.
75. Sado DM, Maestrini V, Piechnik SK, et al. Noncontrast myocardial T1 mapping using cardiovascular magnetic resonance for iron overload. *J Magn Reson Imaging* 2015;41:1505-11.
76. Pennell DJ, Udelson JE, Arai AE, et al. Cardiovascular function and treatment in beta-thalassemia major: a consensus statement from the American Heart Association. *Circulation* 2013;128:281-308.
77. Tzelepis GE, Kelekis NL, Plastiras SC, et al. Pattern and distribution of myocardial fibrosis in systemic sclerosis: a delayed enhanced magnetic resonance imaging study. *Arthritis Rheum* 2007;56:3827-36.
78. Ntusi NA, Piechnik SK, Francis JM, et al. Subclinical myocardial inflammation and diffuse fibrosis are common in systemic sclerosis--a clinical study using myocardial T1-mapping and extracellular volume quantification. *J Cardiovasc Magn Reson* 2014;16:21.
79. Syed IS, Martinez MW, Feng DL, Glockner JF. Cardiac magnetic resonance imaging of eosinophilic endomyocardial disease. *Int J Cardiol* 2008;126:e50-2.
80. Nihoyannopoulos P, Dawson D. Restrictive cardiomyopathies. *Eur J Echocardiogr* 2009;10:iii23-33.
81. Phelan D, Collier P, Thavendiranathan P, et al. Relative apical sparing of longitudinal strain using two-dimensional speckle-tracking echocardiography is both sensitive and specific for the diagnosis of cardiac amyloidosis. *Heart* 2012;98:1442-8.
82. Yancy CW, Jessup M, Bozkurt B, et al. 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on practice guidelines. *Circulation* 2013;128:e240-327.

83. Walker CM, Saldana DA, Gladish GW, et al. Cardiac complications of oncologic therapy. *Radiographics* 2013;33:1801-15.
84. Arora NP, Mohamad T, Mahajan N, et al. Cardiac magnetic resonance imaging in peripartum cardiomyopathy. *Am J Med Sci* 2014;347:112-7.
85. Sliwa K, Hilfiker-Kleiner D, Petrie MC, et al. Current state of knowledge on aetiology, diagnosis, management, and therapy of peripartum cardiomyopathy: a position statement from the Heart Failure Association of the European Society of Cardiology Working Group on peripartum cardiomyopathy. *Eur J Heart Fail* 2010;12:767-78.
86. Grothoff M, Pachowsky M, Hoffmann J, et al. Value of cardiovascular MR in diagnosing left ventricular non-compaction cardiomyopathy and in discriminating between other cardiomyopathies. *Eur Radiol* 2012;22:2699-709.
87. Abbas A, Sonnex E, Pereira RS, Coulden RA. Cardiac magnetic resonance assessment of takotsubo cardiomyopathy. *Clin Radiol* 2016;71:e110-9.
88. Kurisu S, Inoue I, Kawagoe T, et al. Prevalence of incidental coronary artery disease in tako-tsubo cardiomyopathy. *Coron Artery Dis* 2009;20:214-8.
89. Ghadri JR, Wittstein IS, Prasad A, et al. International Expert Consensus Document on Takotsubo Syndrome (Part I): Clinical Characteristics, Diagnostic Criteria, and Pathophysiology. *Eur Heart J* 2018;39:2032-46.
90. Redfors B, Ramunddal T, Shao Y, Omerovic E. Takotsubo triggered by acute myocardial infarction: a common but overlooked syndrome? *J Geriatr Cardiol* 2014;11:171-3.
91. Andreini D, Pontone G, Pepi M, et al. Diagnostic accuracy of multidetector computed tomography coronary angiography in patients with dilated cardiomyopathy. *J Am Coll Cardiol* 2007;49:2044-50.
92. Sidhu MS, Uthamalingam S, Ahmed W, et al. Defining left ventricular noncompaction using cardiac computed tomography. *J Thorac Imaging* 2014;29:60-6.
93. Hussain J, Ghandforoush A, Virk Z, Cherukuri M. Viability assessment by multidetector computed tomography in Takotsubo cardiomyopathy. *J Thorac Imaging* 2011;26:W7-8.
94. Otalvaro L, Zambrano JP, Fishman JE. Takotsubo cardiomyopathy: utility of cardiac computed tomography angiography for acute diagnosis. *J Thorac Imaging* 2011;26:W83-5.
95. Ghadri JR, Wittstein IS, Prasad A, et al. International Expert Consensus Document on Takotsubo Syndrome (Part II): Diagnostic Workup, Outcome, and Management. *Eur Heart J* 2018;39:2047-62.
96. Tung R, Bauer B, Schelbert H, et al. Incidence of abnormal positron emission tomography in patients with unexplained cardiomyopathy and ventricular arrhythmias: The potential role of occult inflammation in arrhythmogenesis. *Heart Rhythm* 2015;12:2488-98.
97. McCrohon JA, Moon JC, Prasad SK, et al. Differentiation of heart failure related to dilated cardiomyopathy and coronary artery disease using gadolinium-enhanced cardiovascular magnetic resonance. *Circulation* 2003;108:54-9.
98. Gulati A, Jabbour A, Ismail TF, et al. Association of fibrosis with mortality and sudden cardiac death in patients with nonischemic dilated cardiomyopathy. *JAMA* 2013;309:896-908.
99. Puntmann VO, Carr-White G, Jabbour A, et al. T1-Mapping and Outcome in Nonischemic Cardiomyopathy: All-Cause Mortality and Heart Failure. *JACC Cardiovasc Imaging* 2016;9:40-50.
100. Lopez-Fernandez T, Thavendiranathan P. Emerging Cardiac Imaging Modalities for the Early Detection of Cardiotoxicity Due to Anticancer Therapies. *Rev Esp Cardiol (Engl Ed)* 2017;70:487-95.
101. Thavendiranathan P, Wintersperger BJ, Flamm SD, Marwick TH. Cardiac MRI in the assessment of cardiac injury and toxicity from cancer chemotherapy: a systematic review. *Circ Cardiovasc Imaging* 2013;6:1080-91.
102. Ylanen K, Poutanen T, Savikurki-Heikkila P, Rinta-Kiikka I, Eerola A, Vettenranta K. Cardiac magnetic resonance imaging in the evaluation of the late effects of anthracyclines among long-term survivors of childhood cancer. *J Am Coll Cardiol* 2013;61:1539-47.
103. Renz DM, Rottgen R, Habedank D, et al. New insights into peripartum cardiomyopathy using cardiac magnetic resonance imaging. *Rofo* 2011;183:834-41.
104. Verhaert D, Richards K, Rafael-Fortney JA, Raman SV. Cardiac involvement in patients with muscular dystrophies: magnetic resonance imaging phenotype and genotypic considerations. *Circ Cardiovasc Imaging* 2011;4:67-76.

105. Tandon A, Villa CR, Hor KN, et al. Myocardial fibrosis burden predicts left ventricular ejection fraction and is associated with age and steroid treatment duration in duchenne muscular dystrophy. *J Am Heart Assoc* 2015;4.
106. Hagenbuch SC, Gottliebson WM, Wansapura J, et al. Detection of progressive cardiac dysfunction by serial evaluation of circumferential strain in patients with Duchenne muscular dystrophy. *Am J Cardiol* 2010;105:1451-5.
107. Jacquier A, Thuny F, Jop B, et al. Measurement of trabeculated left ventricular mass using cardiac magnetic resonance imaging in the diagnosis of left ventricular non-compaction. *Eur Heart J* 2010;31:1098-104.
108. Ashrith G, Gupta D, Hanmer J, Weiss RM. Cardiovascular magnetic resonance characterization of left ventricular non-compaction provides independent prognostic information in patients with incident heart failure or suspected cardiomyopathy. *J Cardiovasc Magn Reson* 2014;16:64.
109. Dodd JD, Holmvang G, Hoffmann U, et al. Quantification of left ventricular noncompaction and trabecular delayed hyperenhancement with cardiac MRI: correlation with clinical severity. *AJR Am J Roentgenol* 2007;189:974-80.
110. Thuny F, Jacquier A, Jop B, et al. Assessment of left ventricular non-compaction in adults: side-by-side comparison of cardiac magnetic resonance imaging with echocardiography. *Arch Cardiovasc Dis* 2010;103:150-9.
111. Stacey RB, Andersen MM, St Clair M, Hundley WG, Thohan V. Comparison of systolic and diastolic criteria for isolated LV noncompaction in CMR. *JACC Cardiovasc Imaging* 2013;6:931-40.
112. Kawel N, Nacif M, Arai AE, et al. Trabeculated (noncompacted) and compact myocardium in adults: the multi-ethnic study of atherosclerosis. *Circ Cardiovasc Imaging* 2012;5:357-66.
113. Weir-McCall JR, Yeap PM, Papagiorcopulo C, et al. Left Ventricular Noncompaction: Anatomical Phenotype or Distinct Cardiomyopathy? *J Am Coll Cardiol* 2016;68:2157-65.
114. Eitel I, von Knobelsdorff-Brenkenhoff F, Bernhardt P, et al. Clinical characteristics and cardiovascular magnetic resonance findings in stress (takotsubo) cardiomyopathy. *JAMA* 2011;306:277-86.
115. Ferreira VM, Piechnik SK, Dall'Armellina E, et al. Non-contrast T1-mapping detects acute myocardial edema with high diagnostic accuracy: a comparison to T2-weighted cardiovascular magnetic resonance. *J Cardiovasc Magn Reson* 2012;14:42.
116. Nakamori S, Matsuoka K, Onishi K, et al. Prevalence and signal characteristics of late gadolinium enhancement on contrast-enhanced magnetic resonance imaging in patients with takotsubo cardiomyopathy. *Circ J* 2012;76:914-21.
117. Naruse Y, Sato A, Kasahara K, et al. The clinical impact of late gadolinium enhancement in Takotsubo cardiomyopathy: serial analysis of cardiovascular magnetic resonance images. *J Cardiovasc Magn Reson* 2011;13:67.
118. Scally C, Rudd A, Mezincescu A, et al. Persistent Long-Term Structural, Functional, and Metabolic Changes After Stress-Induced (Takotsubo) Cardiomyopathy. *Circulation* 2018;137:1039-48.
119. Thavendiranathan P, Poulin F, Lim KD, Plana JC, Woo A, Marwick TH. Use of myocardial strain imaging by echocardiography for the early detection of cardiotoxicity in patients during and after cancer chemotherapy: a systematic review. *J Am Coll Cardiol* 2014;63:2751-68.
120. Bauersachs J, Arrigo M, Hilfiker-Kleiner D, et al. Current management of patients with severe acute peripartum cardiomyopathy: practical guidance from the Heart Failure Association of the European Society of Cardiology Study Group on peripartum cardiomyopathy. *Eur J Heart Fail* 2016;18:1096-105.
121. Zhang X, Yuan L, Qiu L, et al. Incremental value of contrast echocardiography in the diagnosis of left ventricular noncompaction. *Front Med* 2016;10:499-506.
122. Kalapos A, Domsik P, Forster T, Nemes A. Left ventricular strain reduction is not confined to the noncompacted segments in noncompaction cardiomyopathy-insights from the three-dimensional speckle tracking echocardiographic MAGYAR-path study. *Echocardiography* 2014;31:638-43.
123. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the Task Force Criteria. *Eur Heart J* 2010;31:806-14.
124. te Riele AS, Tandri H, Bluemke DA. Arrhythmogenic right ventricular cardiomyopathy (ARVC): cardiovascular magnetic resonance update. *J Cardiovasc Magn Reson* 2014;16:50.
125. Huizar JF, Ellenbogen KA, Tan AY, Kaszala K. Arrhythmia-Induced Cardiomyopathy: JACC State-of-the-Art Review. *J Am Coll Cardiol* 2019;73:2328-44.

126. Hasdemir C, Yuksel A, Camli D, et al. Late gadolinium enhancement CMR in patients with tachycardia-induced cardiomyopathy caused by idiopathic ventricular arrhythmias. *Pacing Clin Electrophysiol* 2012;35:465-70.
127. Nakajima T, Kimura F, Kajimoto K, Kasanuki H, Hagiwara N. Utility of ECG-gated MDCT to differentiate patients with ARVC/D from patients with ventricular tachyarrhythmias. *J Cardiovasc Comput Tomogr* 2013;7:223-33.
128. Bourfiss M, Vigneault DM, Aliyari Ghasebeh M, et al. Feature tracking CMR reveals abnormal strain in preclinical arrhythmogenic right ventricular dysplasia/ cardiomyopathy: a multisoftware feasibility and clinical implementation study. *J Cardiovasc Magn Reson* 2017;19:66.
129. Vigneault DM, te Riele AS, James CA, et al. Right ventricular strain by MR quantitatively identifies regional dysfunction in patients with arrhythmogenic right ventricular cardiomyopathy. *J Magn Reson Imaging* 2016;43:1132-9.
130. Saberniak J, Leren IS, Haland TF, et al. Comparison of patients with early-phase arrhythmogenic right ventricular cardiomyopathy and right ventricular outflow tract ventricular tachycardia. *Eur Heart J Cardiovasc Imaging* 2017;18:62-69.
131. Piers SR, Tao Q, van Huls van Taxis CF, Schaliij MJ, van der Geest RJ, Zeppenfeld K. Contrast-enhanced MRI-derived scar patterns and associated ventricular tachycardias in nonischemic cardiomyopathy: implications for the ablation strategy. *Circ Arrhythm Electrophysiol* 2013;6:875-83.
132. Ferreira VM, Piechnik SK, Dall'Armellina E, et al. T(1) mapping for the diagnosis of acute myocarditis using CMR: comparison to T2-weighted and late gadolinium enhanced imaging. *JACC Cardiovasc Imaging* 2013;6:1048-58.
133. Prasad M, Hermann J, Gabriel SE, et al. Cardiorheumatology: cardiac involvement in systemic rheumatic disease. *Nat Rev Cardiol* 2015;12:168-76.
134. Chagas disease in Latin America: an epidemiological update based on 2010 estimates. *Wkly Epidemiol Rec* 2015;90:33-43.
135. Torreao JA, Ianni BM, Mady C, et al. Myocardial tissue characterization in Chagas' heart disease by cardiovascular magnetic resonance. *J Cardiovasc Magn Reson* 2015;17:97.
136. Dambrin G, Laissy JP, Serfaty JM, Caussin C, Lancelin B, Paul JF. Diagnostic value of ECG-gated multidetector computed tomography in the early phase of suspected acute myocarditis. A preliminary comparative study with cardiac MRI. *Eur Radiol* 2007;17:331-8.
137. Patriki D, Gresser E, Manka R, Emmert MY, Luscher TF, Heidecker B. Approximation of the Incidence of Myocarditis by Systematic Screening With Cardiac Magnetic Resonance Imaging. *JACC Heart Fail* 2018;6:573-79.
138. Chu GC, Flewitt JA, Mikami Y, Vermes E, Friedrich MG. Assessment of acute myocarditis by cardiovascular MR: diagnostic performance of shortened protocols. *Int J Cardiovasc Imaging* 2013;29:1077-83.
139. Ferreira VM, Schulz-Menger J, Holmvang G, et al. Cardiovascular Magnetic Resonance in Nonischemic Myocardial Inflammation: Expert Recommendations. *J Am Coll Cardiol* 2018;72:3158-76.
140. Mahrholdt H, Wagner A, Deluigi CC, et al. Presentation, patterns of myocardial damage, and clinical course of viral myocarditis. *Circulation* 2006;114:1581-90.
141. Berg J, Kottwitz J, Baltensperger N, et al. Cardiac Magnetic Resonance Imaging in Myocarditis Reveals Persistent Disease Activity Despite Normalization of Cardiac Enzymes and Inflammatory Parameters at 3-Month Follow-Up. *Circ Heart Fail* 2017;10.
142. Schumm J, Greulich S, Wagner A, et al. Cardiovascular magnetic resonance risk stratification in patients with clinically suspected myocarditis. *J Cardiovasc Magn Reson* 2014;16:14.
143. Regueiro A, Garcia-Alvarez A, Sitges M, et al. Myocardial involvement in Chagas disease: insights from cardiac magnetic resonance. *Int J Cardiol* 2013;165:107-12.
144. Acquatella H, Asch FM, Barbosa MM, et al. Recommendations for Multimodality Cardiac Imaging in Patients with Chagas Disease: A Report from the American Society of Echocardiography in Collaboration With the InterAmerican Association of Echocardiography (ECOSIAC) and the Cardiovascular Imaging Department of the Brazilian Society of Cardiology (DIC-SBC). *J Am Soc Echocardiogr* 2018;31:3-25.
145. Gomes VA, Alves GF, Hadlich M, et al. Analysis of Regional Left Ventricular Strain in Patients with Chagas Disease and Normal Left Ventricular Systolic Function. *J Am Soc Echocardiogr* 2016;29:679-88.

146. American College of Radiology. ACR Appropriateness Criteria® Radiation Dose Assessment Introduction. Available at: <https://www.acr.org/-/media/ACR/Files/Appropriateness-Criteria/RadiationDoseAssessmentIntro.pdf>. Accessed September 30, 2020.

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.