Cardiac MR Imaging of Nonischemic Cardiomyopathies: Imaging Protocols and Spectra of Appearances

Recent technologic advances in cardiac magnetic resonance (MR) imaging have resulted in images with high spatial and temporal resolution and excellent myocardial tissue characterization. Cardiac MR is a valuable imaging technique for detection and assessment of the morphology and functional characteristics of the nonischemic cardiomyopathy. It has gained acceptance as a standalone imaging modality that can provide further information beyond the capabilities of traditional modalities such as echocardiography and angiography. Black-blood fast spin-echo MR images allow morphologic assessment of the heart with high spatial resolution, while T2-weighted MR images can depict acute myocardial edema. Contrast material–enhanced images can depict and be used to quantify myocardial edema, infiltration, and fibrosis. This review presents recommended cardiac MR protocols for and the spectrum of imaging appearances of the nonischemic cardiomyopathies.

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Recent technical advances have allowed cardiac magnetic resonance (MR) imaging to enter mainstream cardiology imaging practice for cardiomyopathy assessment (1). The development of increasing magnetic field strengths and surface coil channels, rapid k-space sampling, postprocessing techniques, and sophisticated sequences for myocardial characterization have made cardiac MR a powerful tool in the work-up of many complex cardiomyopathies (2). Consensus statements from several international cardiology associations include cardiac MR as a primary imaging technique (1,3,4). In this review, we will outline the cardiac MR protocols that allow optimal characterization of nonischemic cardiomyopathy and illustrate the spectrum of their appearances on cardiac MR imaging studies.

The prevalence of cardiomyopathy in the United States is approximately one in 5439 people, or 0.02% of the population (Table 1). Prevalence varies between the most common type, dilated cardiomyopathy, to the least common, restrictive cardiomyopathy. It is estimated that cardiomyopathy causes approximately 25,000 deaths each year in the United States and follows coronary arterial heart disease as the commonest cause of sudden death (5). Thoracic echocardiography remains the mainstay imaging modality for cardiomyopathy. It allows accurate assessment of chamber dynamics, valvar motion, and real-time Doppler interrogation of intracardiac blood flow and its practical aspects, such as widespread availability and portability, make it a valuable imaging method for cardiomyopathy evaluation (6). Nevertheless, the accuracy of transthoracic echocardiography can be reduced by factors that cause suboptimal acoustic windows, such as chest wall or rib deformities, obesity, and obstructive lung disease. Transesophageal echocardiography resolves this by imaging through the esophagus, with often-improved visualization of the posterior aspect of the heart, which is sometimes not possible via the transthoracic window. Disadvantages are associated with its invasive nature: Intubation of the esophagus is required, and there is an attendant small but documented risk of esophageal perforation, bleeding, and aspiration (7). The patient may also require care by an anesthesiologist.

Cardiac MR is a noninvasive test with excellent spatial and myocardial tissue resolution (2). A combination of sequences allows the detection, localization, and quantification of many pathologic myocardial processes. It has become the reference standard test for accurate quantification of chamber size and function (8). In this article, we will describe the cardiac MR protocols that allow optimum imaging and provide the spectrum of appearances of nonischemic cardiomyopathies.

### Classification of Cardiomyopathy

An increased understanding of cardiomyopathy prompted a revision of the 1995 World Health Organization/International Society and Federation of Cardiology Task Force on the Definition and Classification of Cardiomyopathy with the inclusion of new subgroups of myocardial diseases (9). This contemporary consensus report revised the definition of cardiomyopathy to include “mechanical or electrical dysfunction that usually exhibit inappropriate ventricular hypertrophy and dilatation due to a variety of causes that frequently are genetic.” Under the classification system, the division of cardiomyopathy is split into primary and secondary causes (Tables 2, 3). Primary cardiomyopathies are those disease processes that are uniquely intrinsic to the myocardium. They may be genetic, acquired, or mixed and represent a minority of cardiomyopathies. Secondary cardiomyopathies have multigorgan involvement and are responsible for the majority of cases. There are other classification systems such as the classification by Elliott et al (11) published by the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases, which places an emphasis on phenotypic classification.

### Basic Cardiac MR Protocols for Cardiomyopathy Assessment

The cardiac MR imaging protocol used in cases of cardiomyopathy should be tailored specifically to the suspected type of cardiomyopathy. In this regard, it is important that the radiologist be present to review the images, so that subsequent additional sequences can be determined. There is a tremendous degree of diversity in cardiac MR sequences, but all follow a basic generic protocol (Table 4) (11). Specific additional sequences may be added, depending on the cardiomyopathy. Table 4 outlines the general basic set of cardiac MR imaging sequences for cardiomyopathy evaluation (see also Fig E1 [online]).

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**Content codes:** CA, MR

**Abbreviations:**
- ARVD = arrhythmogenic RV dysplasia
- HCM = hypertrophic cardiomyopathy
- LV = left ventricle
- RV = right ventricle
- SSFP = steady-state free precession

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**Potential conflicts of interest are list at the end of this article.**
Scout images (axial, coronal, sagittal) are useful, particularly sagittal scout images, which represent an underutilized and valuable opportunity to ensure that the heart is positioned at the center of the surface coils, which improves signal to noise. A stack of axial sections of the thorax (most commonly, half-Fourier rapid acquisition with relaxation enhancement or SSFP sequences) is acquired next, to depict major non-cardiac pathologic processes and to provide planning information for the subsequent sequences (Fig E1a [online]). Typically, a set of cine SSFP sections are acquired next in the vertical long axis (so named because this imaging plane depicts the atrium and ventricle; Fig E1b [online], Movie 1a [online]) and horizontal long axis (so named because it depicts all four cardiac chambers; Fig E1c [online], Movie 1b [online]) planes.

These and all subsequent sequences are acquired by using cardiac gating. This is an essential component to overcome image blurring secondary to cardiac contraction. The optimal method is to use electrocardiographic gating, in which image acquisition is triggered from the QRS complex of a three-lead electrocardiogram. For cine SSFP sections, portions of k-space data that compose the image are acquired at the same time point of the QRS complex over several heartbeats, which eliminates image blurring. These sequences usually require a breath hold of 8–12 seconds and, typically, acquisition of two parallel sections (because one may be of suboptimal image quality or miss the ideal image plane). These are followed by a stack of short-axis sections from the annulus to the apex (Fig E1d [online], Movie 1c [online]). These are used to allow quantification of chamber volumes and myocardial function. It is prudent to plan to acquire the first section of the stack on the atrial side of the annulus, because the annulus of the heart contracts toward the apex during systole. Once acquired, the stack of short-axis sections can be interrogated with specific software programs to enable accurate measurements of end-diastolic and systolic volumes and record functional cardiac parameters, such as left and right ventricular end-systolic and diastolic volumes, cardiac mass, stroke volume and ejection fraction. A qualitative analysis of global and regional ventricular function may be provided using a five-point scale (score of 1 for normal; 2 for mild hypokinesia, 3 for severe hypokinesia, 4 for akinesia, 5 for dyskinesia) (12). Although chamber volumes are more meaningful, we also consider it useful to supply an end-diastolic diameter of the LV in our reports, which we measure at the midventricular level. This should measure a mean of 50.2 mm (upper bound of 95% confidence interval, 58.5 mm) for men and 45.6 mm (upper bound, 51.1 mm) for women (13).

For all cine acquisitions, parallel imaging techniques can be used to shorten the breath hold. In brief, parallel imaging incorporates the signal and its location from multiple independent cardiac receiver coils to encode multiple MR echoes simultaneously, which speeds up image acquisition (14).

Several additional sequences can be performed at this point, depending on the cardiomyopathy in question. These are described below in more detail for each cardiomyopathy. After these sequences have been performed, late-gadolinium-enhanced images are acquired. The optimum time for late-enhancement image acquisition is usually 10–30 minutes after contrast agent injection (15). For late-enhancement images, optimal contrast differentiation between viable and nonviable myocardium is individually evaluated for each patient by determining the exact time the signal of the normal myocardium is null or black (Fig E1e [online]). The inversion time will vary considerably from patient to patient and must be determined on an individual basis. If the myocardium has a confluent gray appearance, then the inversion time is usually too early (Fig E1f); if it has a mixed appearance, then it is usually too late (Fig E1g). Normal inversion times vary but generally are between 200 and 350 msec, depending on the manufacturer of the MR unit, the cardiac output, the time of image acquisition after contrast agent injection, and the dose of contrast agent administered (15). In contrast-enhanced imaging, inversion time is adjusted as the examination progresses, to allow for contrast agent washout from the myocardium (nulling time will change as this is happening). Increasing the time by 10–20 msec every one to three sections is usually sufficient. The correct inversion time is very important for accurate image interpretation and to prevent polarity artifacts. A useful alternative is acquisition of phase-sensitive images by using an inversion-recovery spoiled-gradient-echo or SSFP sequence (Fig E1h) (16). This minimizes polarity artifacts over a wider range of inversion times than the traditional magnitude sequence, thus resulting in more consistent image quality. However, in our experience it is not as sensitive for detection of small areas of enhancement.

The myocardium is evaluated for the presence, location, and extent of enhancement. A useful method of reference for localization uses the American Heart Association 17-segment model, in which the ventricle is divided into three levels: six basal segments, six middle segments, and four apical segments, with the apex being the last segment (17). A useful method of reference for extent of myocardial enhancement uses a five-point scale for each segment: score of 0 for no late enhancement, 1 = 1%–25% of the affected segment enhanced, 2 for 26%–50% of the affected segment enhanced, 3 for 51%–75% of the affected segment enhanced, and 4 for 76%–100% of the affected segment enhanced (18). Global late-enhancement extent as a percentage of LV myocardium may be calculated by summing the segments with late enhancement (each weighted by the midpoint of enhancement for the segmental score). The contrast-to-noise ratio is also often provided for the late-enhancement sequence. This can be calculated (parallel imaging should be not be used for this calculation) as $SI_{LE} - SI_{RM}/SD_{N}$, where $SI_{LE}$ is mean signal intensity of late-enhancing segments, $SI_{RM}$ is mean signal intensity of remote myocardium, and $SD_{N}$ is the standard deviation of noise. Finally, it is important to emphasize the overall distribution of the
The use of contrast agents in cardiac MR imaging is an important aspect of the utility of cardiac MR for characterizing nonischemic cardiomyopathies. We use a commercially available gadolinium-based contrast agent (gadopentetate dimeglumine) in our protocol. The contrast agent is injected as a bolus (0.2 mmol per kilogram of body weight) via an arm vein, preferably in the antecubital fossa. It may be administered by means of hand injection or infusion pump. This should be followed by a 20–40-mL saline bolus chaser. Gadolinium-based contrast agents are not recommended in patients with renal failure and a glomerular filtration rate of less than 30 mL/kg/min because of the risk of nephrogenic systemic fibrosis.

Table 2

Primary Cardiomyopathies Predominantly Involving the Heart

<table>
<thead>
<tr>
<th>Disease†</th>
<th>Mixed</th>
<th>Acquired</th>
</tr>
</thead>
<tbody>
<tr>
<td>HCM</td>
<td>Dilated cardiomyopathy</td>
<td>Inflammatory (myocarditis)</td>
</tr>
<tr>
<td>ARVD</td>
<td>Restrictive cardiomyopathy</td>
<td>Stress provoked (takotsubo)</td>
</tr>
<tr>
<td>LVNC</td>
<td>Peripartum</td>
<td></td>
</tr>
<tr>
<td>DCM</td>
<td>Amyloidosis</td>
<td></td>
</tr>
<tr>
<td>RCM</td>
<td>Cardiac sarcoidosis</td>
<td></td>
</tr>
<tr>
<td>SCD</td>
<td>Siderotic</td>
<td></td>
</tr>
<tr>
<td>CSC</td>
<td>Scleroderma</td>
<td></td>
</tr>
</tbody>
</table>

† HCM= hypertrophic cardiomyopathy, LV = left ventricle, RV = right ventricle.

* Numbers in parentheses are reference numbers.
three-chamber view (Movie 3[online]), which depicts the left atrium and LV, the mitral and aortic valves, and the proximal ascending aorta (Table 4) by using planimetry and phase-velocity encoding sequences of the LV outflow tract (25). Myocardial tagging can be used to depict detailed myocardial abnormalities in HCM, such as a reduction in posterior rotation, reduced radial displacement and reduced three-dimensional myocardial shortening, although this sequence application has not entered into mainstream imaging practice as yet.

HCM should be distinguished from LV hypertrophy caused by increased afterload, such as in cases of hypertension, aortic stenosis, or athlete’s heart in which the LV wall thickness can measure up to 15 mm. Petersen et al (26) found that that the most useful parameter to enable distinction between HCM and physiologic causes such as athlete’s heart was the ratio of end-diastolic wall thickness to end-diastolic volume. Use of a cutoff value of 0.15 mm/mL/m² yields positive and negative predictive values of 95% and 94%, respectively.

When evaluating for HCM, it is important to be aware of several morphologic variants from the classic form, in which the mid or apical ventricular levels may be predominantly affected (Fig 1, Movie 4 [online]). A minority of patients (5%) demonstrate circumferential symmetric hypertrophy.

Late gadolinium enhancement on cardiac MR images in patients with HCM is significantly associated with traditional risk factors for sudden death in HCM (Fig 1) (27). More recently, even in asymptomatic or mildly symptomatic patients with HCM, the presence of late enhancement has been associated with the development of arrhythmias (28). Large HCM cohort studies (29,30) have now shown a strong independent association between late enhancement and surrogate markers of arrhythmia, sudden cardiac death, and implantable cardioverter defibrillator discharge. Finally, with its absence of radiation exposure, serial cardiac MR imaging seems to be an ideal tool for screening relatives of patients with HCM (Fig 1) (31).

Arrhythmogenic right ventricular dysplasia.—Arrhythmogenic right ventricular dysplasia (ARVD) is a rare genetic disorder characterized by progressive loss of myocytes with fibrofatty replacement of RV and, less commonly, LV myocardium. Patients may have ventricular arrhythmias and left bundle branch block at the time of presentation, syncope, or sudden cardiac death. ARVD may be responsible for up to 5% of sudden deaths in young athletes (32) but has a higher prevalence in other countries such as Italy (25% of sudden deaths in young adults) (33). Because of the subtlety of the phenotype, consensus criteria were developed on the basis of structural, functional, and electrocardiographic manifestations. In 1994, the task force of the Working Group on Myocardial and Pericardial Disease of the European Society of Cardiology and the task force of the Scientific Council on Cardiomyopathy of the World Heart Federation proposed diagnostic criteria based on the presence of major and minor criteria that involve structural, histologic, electrocardiographic, genetic, and arrhythmic factors (34). To fulfill the criteria for ARVD, the patient’s condition had to meet two major criteria, one major criterion and two minor criteria, or four minor criteria. These criteria have since been modified in 2010 by the task force (35). In the modified criteria, tissue characterization depicted on cardiac MR images, such as fatty infiltration, have been removed, and more emphasis has been placed on wall motion, volume, and ejection fraction abnormalities (Table 5).

Pathologic descriptions include fibrofatty replacement of myocardial tissue, focal RV wall thinning and/or aneurysm, RV outflow tract enlargement, and RV dilatation (36). RV biopsy has a low diagnostic yield because of the patchy distribution of fibrofatty infiltration and the predominantly epicardial distribution (23). Cardiac MR assessment of ARVD is now based primarily on functional and volume abnormalities of the RV (35). Many centers still provide tissue-characterization imaging of the RV. (a) Morphologic abnormalities include intramyocardial fat deposits (Fig 2a, 2b), focal wall thinning (<2 mm), wall hypertrophy (>6 mm), moderator band hypertrophy, and trabeculation thickening and disarray. (b) Functional abnormalities include focal or global RV wall hypokinesis, focal or global RV dilatation, and focal aneurysms in severe cases (Fig 2c, Movie 5 [online]). The site of involvement of these abnormal-

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**Table 3**

<table>
<thead>
<tr>
<th>Type of Cardiomyopathy</th>
<th>Diseases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infiltrative</td>
<td>Amyloidosis, Gaucher disease, Hurler disease, Hunter disease</td>
</tr>
<tr>
<td>Storage</td>
<td>Hemochromatosis, Fabry disease, glycogen-storage disease (type II, Pompe), Niemann-Pick disease</td>
</tr>
<tr>
<td>Toxicity</td>
<td>Drugs, heavy metals, chemical agents</td>
</tr>
<tr>
<td>Endomyocardial</td>
<td>Endomyocardial fibrosis, hypereosinophilic syndrome (Loeffler endocarditis)</td>
</tr>
<tr>
<td>Inflammatory</td>
<td>Sarcoidosis</td>
</tr>
<tr>
<td>Endocrine</td>
<td>Diabetes mellitus, hyperthyroidism, hypothyroidism, hyperparathyroidism, pheochromocytoma, acromegaly</td>
</tr>
<tr>
<td>Neuromuscular or neurologic</td>
<td>Friedreich ataxia, Duchenne-Becker muscular dystrophy, Emery-Dreifuss muscular dystrophy, myotonic dystrophy, neurofibromatosis, tuberous sclerosis</td>
</tr>
<tr>
<td>Nutritional</td>
<td>Beriberi, pellagra, scurvy, selenium, carnitine</td>
</tr>
<tr>
<td>Autoimmune</td>
<td>Systemic lupus erythematosus, dermatomyositis, rheumatoid arthritis, scleroderma, polyarteritis nodosa</td>
</tr>
<tr>
<td>Electrolyte imbalance</td>
<td>Potassium, phosphate, magnesium deficiencies; anorexia nervosa; laxative abuse</td>
</tr>
<tr>
<td>Cancer therapy</td>
<td>Anthracylines (doxorubicin), cyclophosphamide, radiation</td>
</tr>
</tbody>
</table>
Table 4
Basic Cardiac MR Protocol for Imaging of Cardiomyopathy

<table>
<thead>
<tr>
<th>Sequence</th>
<th>Imaging Plane</th>
<th>Repetition Time (msec)/Echo Time (msec)</th>
<th>Flip Angle (degrees)</th>
<th>No. of Sections</th>
<th>Section Thickness (mm)</th>
<th>Intersection Gap (%)</th>
<th>Other Parameters</th>
</tr>
</thead>
<tbody>
<tr>
<td>Localizers</td>
<td>Coronal, axial, sagittal</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Half-Fourier RARE SSFP</td>
<td>Axial sections of thorax</td>
<td>1000/27</td>
<td>160</td>
<td>Typically 16–20</td>
<td>8–10</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td>Vertical long axis</td>
<td>Perpendicular to mitral annulus and apex of ventricle</td>
<td>3.96/1.12</td>
<td>60</td>
<td>2</td>
<td>6–8</td>
<td>20</td>
<td>Breath hold</td>
</tr>
<tr>
<td>Horizontal long axis</td>
<td>Parallel to mitral annulus and middle of interventricular septum</td>
<td>Similar to vertical-long-axis parameters</td>
<td>Similar to vertical-long-axis parameters</td>
<td>Similar to vertical-long-axis parameters</td>
<td>Similar to vertical-long-axis parameters</td>
<td>Similar to vertical-long-axis parameters</td>
<td></td>
</tr>
<tr>
<td>Short axis</td>
<td>Perpendicular to interventricular septum, through both ventricles (and sometimes atria)</td>
<td>Similar to vertical-long-axis parameters</td>
<td>Similar to vertical-long-axis parameters</td>
<td>Typically 6–12</td>
<td>Similar to vertical-long-axis parameters</td>
<td>Similar to vertical-long-axis parameters</td>
<td></td>
</tr>
<tr>
<td>T2 weighted</td>
<td>Similar to short-axis plane</td>
<td>2 R-R intervals/100</td>
<td></td>
<td>10–12</td>
<td>30–50</td>
<td></td>
<td>Breath hold, black blood, double inversion recovery, fat saturation</td>
</tr>
<tr>
<td>Late gadolinium enhanced</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>T1-weighted scout†</td>
<td></td>
<td>23.49/1/12</td>
<td>50</td>
<td>1 repeated 25–30 times</td>
<td>8</td>
<td></td>
<td>Acquired 10–30 minutes after contrast agent injection</td>
</tr>
<tr>
<td>Formal late-enhancement</td>
<td>2D double-inversion gradient echo</td>
<td>7.0/3.37†</td>
<td>25</td>
<td>Typically 6–12</td>
<td>6–8</td>
<td>0–20</td>
<td>Enhancing areas often confirmed by acquiring vertical-long-axis or 4-chamber oblique image through any abnormal region</td>
</tr>
</tbody>
</table>

* RARE = rapid acquisition with relaxation enhancement, SSFP = steady-state free precession, 2D = two-dimensional.
† Allows determination of optimal inversion time.
‡ Trigger delay set to image in middiastole...
Figure 1: (a, b) HCM in a 52-year-old woman. (a) Short-axis SSFP MR image shows mild asymmetric hypertrophy (16 mm thick) of interventricular septum (solid arrows), as compared with 9-mm-thick lateral free wall (open arrow). Note RV dilatation. (b) Short-axis late-gadolinium-enhanced MR image shows enhancement in hypertrophied segment (arrow), consistent with scar. (c, d) HCM in her 16-year-old son. (c) Short-axis SSFP image shows asymmetric hypertrophy of inferoseptal segment (arrow). (d) Short-axis late-enhancement MR image shows enhancement (arrow) in hypertrophied segment, consistent with scar. (e, f) Apical-variant HCM in a 45-year-old man. (e) Vertical-long-axis SSFP MR image shows asymmetric apical hypertrophy, a variant of HCM that occurs in 5%–10% of cases. (f) Vertical-long-axis late-enhancement image shows scar in hypertrophied segments (arrow).

Cardiac MR Imaging of Nonischemic Cardiomyopathies O’Donnell et al

Important cardiac MR sequences in ARVD.—In our experience, ARVD imaging remains one of the most challenging applications in cardiac MR imaging in terms of sequence performance, time requirements, and image interpretation. Along with the basic imaging protocol outlined above, cardiac MR focuses particular attention on the RV and RV outflow tract (RVOT). Some centers image patients in the prone position to minimize chest wall breathing artifacts. Most centers use a thinner section thickness and intersection gap for this protocol (5–6-mm contiguous sections are typical). Spin-echo or fast spin-echo T1-weighted images (spin-echo studies can be acquired in systole with most MR units, making RV myocardial fat infiltration easier to see, but fast spin-echo studies are less affected by artifacts) can be acquired in an axial oblique and short-axis stack to evaluate for myocardial fat infiltration (fast spin-echo sequences should use double inversion). A double-inversion pulse sequence with a short echo time (30 msec), short echo train length of 28–30, and thin (5-mm) sections is optimum (36). Fat infiltration may be confirmed by repeating the sequence with fat-saturation and demonstrating myocardial signal dropout (Fig 2a, 2b). Note that with the new modified criteria these particular sequences will not be required, but we mention them for completeness (35). Saturation bands above and below the heart also help improve image quality by reducing flow artifacts related to slow-inflowing blood. A small field of view targeted to the RV helps improve the spatial resolution fur-
Table 5

<table>
<thead>
<tr>
<th>Type of Criterion and Modality</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Global or Regional Dysfunction and Structural Abnormalities</strong></td>
<td></td>
</tr>
<tr>
<td>Major</td>
<td>Regional RV akinesia, dyskinesia, or aneurysm AND one of the following (end diastole)</td>
</tr>
<tr>
<td>2D echocardiography</td>
<td>PLAX RVOT ≥ 32 mm (corrected for body size: [PLAX/BSA] ≥ 19 mm/m²)</td>
</tr>
<tr>
<td></td>
<td>PSAX RVOT ≥ 36 mm (corrected for body size: [PSAX/BSA] ≥ 21 mm/m²)</td>
</tr>
<tr>
<td></td>
<td>Fractional area change = 33%</td>
</tr>
<tr>
<td>Cardiac MR imaging</td>
<td>Regional RV akinesia or dyskinesia or dysynchronous RV contraction AND one of the following:</td>
</tr>
<tr>
<td></td>
<td>Ratio of RV end-diastolic volume to BSA ≥ 110 (males) or ≥ 100 mL/m² (females)</td>
</tr>
<tr>
<td></td>
<td>RV ejection fraction ≤ 40%</td>
</tr>
<tr>
<td>RV angiography</td>
<td>Regional RV akinesia, dyskinesia, or aneurysm</td>
</tr>
<tr>
<td>Minor</td>
<td>Regional RV akinesia or dyskinesia AND one of the following</td>
</tr>
<tr>
<td>2D echocardiography</td>
<td>PLAX RVOT ≥ 29 mm to &lt; 32 mm (corrected for body size [PLAX/BSA] ≥ 16 to &lt; 19 mm/m²)</td>
</tr>
<tr>
<td></td>
<td>PSAX RVOT ≥ 32 mm to &lt; 36 mm (corrected for body size [PSAX/BSA] ≥ 18 to &lt; 21 mm/m²)</td>
</tr>
<tr>
<td></td>
<td>Fractional area change ≥ 33%</td>
</tr>
<tr>
<td>Cardiac MR imaging</td>
<td>Regional RV akinesia or dyskinesia or dysynchronous RV contraction AND one of the following:</td>
</tr>
<tr>
<td></td>
<td>Ratio of RV end-diastolic volume to BSA ≥ 100 to &lt; 110 mL/m² (male) or ≥ 90 to &lt; 100 mL/m²</td>
</tr>
<tr>
<td></td>
<td>RV ejection fraction ≥ 40% to 45%</td>
</tr>
</tbody>
</table>

**Myocardial Characterization**

| Major                         | Residual myocytes < 60% at morphometric analysis (<50% if estimated), with fibrous replacement of RV free wall myocardium in one or more samples, with or without fatty replacement of tissue at endomyocardial biopsy |
| Minor                         | Residual myocytes 60%–75% at morphometric analysis (50%–65% if estimated), with fibrous replacement of RV free wall myocardium in one or more samples, with or without fatty replacement of tissue at endomyocardial biopsy |

**Repolarization Abnormality**

| Major                         | Inverted T waves in right precordial leads (V1R, V2R, V3R) or beyond in individuals > 14 y old (in absence of complete right bundle-branch block) ORS ≥ 120 msec |
| Minor                         | Inverted T waves in leads V1 and V2 in individuals > 14 y old (in absence of complete right bundle-branch block) or in V4, V5, or V6 |
|                              | Inverted T waves in leads V1, V2, and V3, and V4 in individuals > 14 y old in presence of complete right bundle-branch block |

**Depolarization or Conduction Abnormality**

| Major                         | Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of T wave) in right precordial leads (V1R, V2R, V3R) |
| Minor                         | Late potentials on signal-averaged ECG in one or more of three parameters in absence of QRS duration ≥ 110 msec on standard ECG |

**Arrhythmias**

| Major                         | Nonsustained or sustained ventricular tachycardia of RV outflow configuration, left bundle-branch block morphology with inferior axis (positive QRS in leads II, III, and aVF and negative in aVL), or of unknown axis |
| Minor                         | > 500 ventricular extra systoles per 24 h (Holter) |

**Family History**

| Major                         | ARVC/D confirmed in first-degree relative who meets current task force criteria |
| Minor                         | ARVC/D confirmed pathologically at autopsy or surgery in first-degree relative |
|                              | Identification of pathogenic mutation categorized as associated or probably associated with ARVC/D in patient being evaluated |

Table 5 (Continues)
Cardiac MR imaging for ARVD carries a high sensitivity but low specificity when compared with traditional task force criteria (40). This high sensitivity may in part be attributed to the rigorous limitations of the task force guidelines, which easily demonstrate the overt forms of the disease. Because the task force criteria have been suggested to be relatively insensitive to less overt forms of the disease (41), modified criteria have been proposed, such that the presence of any minor criterion in a first-degree relative of a patient with proved ARVD is regarded as clinical disease expression (35). When cardiac MR images are assessed by using these modified criteria, they frequently show abnormal findings, which suggests a role in depicting initial manifestations of disease (35, 41). In one study (41), cardiac MR imaging had 96% sensitivity and 78% specificity in a genotyped subset of patients, only 46% of whom had satisfied the task force criteria. In that study, the addition of the cardiac MR results would have enabled diagnosis in a further 30% of proved gene carriers and in 75% of patients who prospectively satisfied the modified criteria only. That said, such an approach would lead to an increased number of false-positive cases in patients who subsequently turn out to be genotype negative. Risk stratification in asymptomatic gene carrier relatives of an index case has not been fully elucidated.

It is important when interpreting cardiac MR images for ARVD to be
aware that fatty infiltration of the RV free wall has been found in healthy subjects, elderly patients, obese individuals, long-term steroid users, and in cases of idiopathic RV outflow tract tachycardia (42). Furthermore, RV myocardial late enhancement has been described in chronic right-sided myocarditis, sarcoidosis, Chagas disease, and Enterobacter infection (37). We emphasize that the interpretation of cardiac MR imaging abnormalities should be made with caution and should not lead to a diagnosis of ARVD on the basis of cardiac MR findings alone. At our center, we recommend a multidisciplinary approach that involves family history, electrocardiogram and Holter findings, imaging, and results from histopathologic and genetic analyses before making a diagnosis of ARVD (35,37).

LV noncompaction.—LV noncompaction is currently classified as a primary genetic cardiomyopathy (9). It is characterized by the presence of an extensive noncompacted myocardial layer lining the cavity of the LV and impaired LV systolic function. Many patients are asymptomatic and have normal LV systolic function, but others develop cardiac failure, thromboembolism, and malignant arrhythmias (43,44). The compaction ratio can be quantitatively analyzed by measuring the thickness (in millimeters) of the noncompacted myocardium, as compared with the compacted myocardium. This can be done on a myocardial segmental basis by using the standard 17-segment American Heart Association model (17). A noncompacted-to-compacted ratio greater than 2.3 on cardiac MR images is considered diagnostic of LV noncompaction (45). The apical antero- and inferolateral segments of the LV are the most commonly affected. RV involvement has also been reported (46).

Important cardiac MR sequences in LV noncompaction.—SSFP sequences can clearly demonstrate noncompaction features, which appear as a loose network of interwoven trabeculae associated with deep myocardial recesses (Fig 3, Movie 6 [online]). This noncompacted trabecular layer may demonstrate perfusion defects after gadolinium chelate injection, suggesting microcirculatory abnormalities (47). Late-gadolinium-enhancement sequences may depict trabecular late enhancement, which appears to improve the correlation between cardiac MR findings and progressive clinical stages of disease (Fig 3) (48). An advantage of cardiac MR imaging is the ability to acquire images in any obliquity. In our experience, radial vertical-long-axis projections ensure that each section acquired passes through the center of the ventricle and the apex, minimizing the potential to overestimate the ratio of compacted to noncompacted myocardium.

Mixed Cardiomyopathies

Dilated cardiomyopathy.—Dilated cardiomyopathy is characterized by dilatation of the cardiac chambers coupled with impaired contraction of the ventricles. The ventricular chambers exhibit increased diastolic and systolic volume and a low ejection fraction (49). There are many causes, but the majority (50%) are idiopathic (50). Idiopathic dilated cardiomyopathy is the most common cause of heart failure in the young, with an estimated prevalence of at least 36.5 per 100 000 persons in the United States. The symptoms and signs at presentation are progressive dyspnea and orthopnea in the majority of patients. Arrhythmias and sudden death may also occur.

Histopathologic features demonstrate interstitial fibrosis and a numeric decrease in myocyte units. Alterations in the genetic expression of proteins that regulate cardiac muscle contraction have been detected (51). Such abnormalities may be patchy, resulting in a low diagnostic yield from myocardial biopsy. Echocardiography remains the principal examination for dilated cardiomyopathy, but cardiac MR imaging provides an optimal assessment of chamber size and systolic dysfunction. Systolic dysfunction is the most important independent predictor of outcome in dilated cardiomyopathy, and evaluation of diastolic filling allows further identification of subgroups with divergent long-term prognoses (52).

**Figure 3**

LV noncompaction in a 46-year-old woman. (a) Vertical-long-axis SSFP MR image shows marked increase in the noncompacted layer of myocardium (arrow), such that the ratio of compacted to noncompacted myocardium at the apical level was greater than 2.3, consistent with LV noncompaction. (b) Vertical-long-axis late-gadolinium-enhanced MR image demonstrates enhancement in the LV trabeculae, an appearance associated with a more advanced stage of disease.
suggested that these may represent coronary emboli–induced ischemic cardiomyopathy cases or ruptured coronary plaques that have subsequently recanalized (53). The presence of midwall fibrosis on cardiac MR images has been shown to have prognostic implications in patients with dilated cardiomyopathy as an independent predictor of the combined endpoint of mortality from all causes and cardiovascular hospitalization and of the development of ventricular tachycardias (54).

**Restrictive cardiomyopathy.**—This condition is characterized by ventricular diastolic dysfunction and resultant biatrial enlargement with relatively normal ventricular size and systolic function (35). The pathophysiologic changes are due to reduced myocardial compliance, which elevates ventricular pressures with small increases in volume causing decreased filling in diastole, resulting in diastolic heart failure with preservation of systolic function (36,57). Symptoms range from increasing dyspnea and exercise intolerance to palpitations, syncope attacks and conduction disturbances.

Cases may be divided into myocardial or endomyocardial (23). The myocardial cases may be subdivided into noninfiltrative, infiltrative and storage disease and the endomyocardial subgroup are made up of a number of underlying causes including endomyocardial fibrosis, drugs, anaphylactides and carcinoid (56). Clinically, restrictive cardiomyopathy is frequently difficult to distinguish from constrictive pericarditis (58). Identifying pericardial thickening and nonbreakage of myocardial tag lines across the pericardium helps distinguish these conditions (59,60). Endomyocardial biopsy may also be helpful in distinguishing restrictive from constrictive cardiomyopathy, particularly if entities such as amyloidosis or hemochromatosis are suspected clinically (23). However, if pericardial thickening or CMR features of restrictive cardiomyopathy are identified and are compatible with the clinical and echocardiographic picture of constrictive or restrictive physiology then biopsy may be obviated.

**Important cardiac MR sequences for restrictive cardiomyopathy.**—Consensus statements have clarified several indexes used to measure diastolic heart failure (61). SSFP images typically show the heart with biatrial enlargement and normal ventricular size (Fig 5, Movie 8 [online]). Several parameters of ventricular relaxation can be calculated from the short-axis SSFP stack, including time to peak filling, early diastolic filling time, and rate of peak filling (62), which are altered in restrictive cardiomyopathy. Studies have also shown the feasibility of using phase-velocity-encoding sequences to measure mitral inflow (the E/A ratio) with good correlations with echocardiographic findings (63). The E/A ratio is a transmitral flow measurement of peak flow velocity in early diastole (E wave) and during atrial contraction (A wave). In diastolic dysfunction, a greater portion of end-diastolic volume results from late filling rather than early filling because of the stiffness of the ventricle, therefore reducing the E/A ratio.

A further useful sequence in restrictive cardiomyopathy is the application of myocardial tags. This technique provides detailed quantitative assessment of myocardial strain or deformation seen in diastolic dysfunction (64).

**Acquired Cardiomyopathies**

**Inflammatory (myocarditis).**—Myocardial inflammation is a nonspecific response to various insults such as viral or bacterial infection, cardiotoxic agents, catecholamines, infarction, or mechanical injury (9). In North America and Europe, viral infection remains the commonest cause. Several viruses have been associated with myocarditis, based on the detection of viral genome within cardiac tissue. The commonest include Coxackie B virus, non-Coxackie enteroviruses, certain strains of adenovirus, parvovirus B19, and Epstein-Barr virus (65). The sequela of severe viral myocarditis is dilated cardiomyopathy, which is thought to occur in up to 10% of cases (66). Myocarditis may manifest with a variety of symptoms, including chest pain, recent onset of heart failure, atrial or ventricular arrhythmias, cardiogenic shock, or even sudden death.

Endomyocardial biopsy results are considered the reference standard in diagnosis (67). The 1987 Dallas criteria require lymphocytic infiltration associated with myocyte injury in the absence of ischemia. Such criteria are very specific but have a low sensitivity, related to sampling error caused by patchy involvement of the myocardium (68), and high interobserver variability in interpretation (69).
Important cardiac MR sequences in myocarditis.—T2-weighted fast spin-echo MR imaging has been found to be useful in cases of acute myocarditis, because it depicts edema (70). The sequence is a breath-hold black-blood T2-weighted triple inversion-recovery sequence (repetition time/echo time msec/inversion time msec, two R-R intervals/65/140); typically, three to four short-axis sections (8–12-mm thick, echo train length of 32, 256 × 256 matrix) are acquired (Fig 6). An edema ratio, which is a quantitative measure of active inflammation, can be calculated based on the ratio of abnormal to normal myocardial signal intensity, with values greater than or equal to 2 being considered abnormal. To calculate the edema ratio from T2-weighted images, a region of interest encompassing the entire LV myocardium and a second region of interest encompassing the entire visible right erector spinae or latissimus dorsi muscle (skeletal muscle), depending on the homogeneity of the muscle signal intensity, are recorded on the same section. The mean myocardial signal intensity (SI myo) is related to the mean skeletal muscle signal intensity (SI skm) by using the equation ER = SI myo/SI skm, where ER is the edema ratio. An ER greater than or equal to 2 is used to determine the presence of active inflammation (70). It should be noted that several problems exist with T2-weighted sequences for cardiac MR imaging: (a) Phased-array coils cause regional myocardial signal variation; (b) slow-flowing blood in the trabeculae results in high signal intensity adjacent to the endocardium, which can make subendocardial abnormalities difficult to interpret; (c) through-plane motion reduces cardiac signal intensity; and (d) qualitative analysis is subjective. Thus, when calculating the edema ratio care must be taken that an efficient coil intensity-correction algorithm is implemented, and quantitative analysis is recommended (71). A useful second sequence for myocarditis makes use of T1-weighted spin-echo or fast spin-echo sequences (R-R interval/21, echo train length of four, 350–400-mm field of view, 512 × 512 matrix) performed with a body coil before and about 15 seconds after intravenous injection of contrast material to obtain five identical axial sections encompassing the myocardium (Fig 6) (72). A saturation band may be positioned across the atria to reduce the signal from slow-flowing atrial blood. A global enhancement ratio of myocardial enhancement to skeletal muscle enhancement can be calculated, with values greater than or equal to 4 being considered abnormal. To calculate the global enhancement ratio, regions of interest that include both the entire LV and normal skeletal muscle are recorded on the same section. The ratio of myocardial signal intensity before and after contrast administration is the global enhancement ratio (72). Note saturation band across the atria to null signal from slow-flowing blood. (d) Horizontal-long-axis late-gadolinium-enhancement MR image shows enhancement that indicates myocardial edema and necrosis in interlateral segment, characteristic of acute myocarditis (straight arrow). Note two smaller foci of enhancement in anterior segment (curved arrow).
myocardium and the skeletal muscle at the same level are mapped on unenhanced T1-weighted images and are copied to the contrast-enhanced images. The average signal intensities of the myocardium and skeletal muscles before and after contrast enhancement are measured according to the following: 

\[ \text{SI}_{\text{postmyo}} - \text{SI}_{\text{premyo}} / \text{SI}_{\text{preskm}} - \text{SI}_{\text{preskm}} \]

where \( \text{SI}_{\text{postmyo}} \) and \( \text{SI}_{\text{premyo}} \) are myocardial signal intensity after and before contrast enhancement, respectively, and \( \text{SI}_{\text{postskm}} \) and \( \text{SI}_{\text{preskm}} \) are skeletal muscle signal intensity after and before enhancement, respectively.

Finally, late-enhancement sequences may demonstrate late enhancement in patients with acute myocarditis, which is most characteristically depicted in the inferolateral segment of the LV (Fig 6), although changes in the septal segments are also well described. Generally, when evaluating for myocarditis, a combination of sequences is used, with the highest diagnostic accuracy being obtained when two of three are positive (sensitivity, 76%; specificity, 95.5%; overall diagnostic accuracy, 85%) (70,73). Cardiac MR has particular clinical utility in the acute setting because patients with acute myocarditis often have an acute coronary syndrome at presentation, and it may be difficult to differentiate myocarditis from myocardial infarction on the basis of clinical findings.

**Peripartum cardiomyopathy.**—Peripartum cardiomyopathy is a rare cardiomyopathy that occurs in previously healthy women who develop symptoms of heart failure during the peripartum period (74). The cause of peripartum cardiomyopathy is unknown, although it is thought to originate from two broad categories of disease, inflammatory and noninflammatory. Of the inflammatory causes, viral myocarditis is the most common, whereas for noninflammatory causes several factors are hypothesized to play a role, including malnutrition, genetics, hormone function, and increased adrenergic tone (75). The diagnostic criteria are (a) onset of heart failure in the last month of pregnancy or in the first 5 postpartum months, (b) absence of a determinable cause of cardiac failure, and (c) absence of demonstrable heart disease before the last month of pregnancy. Cardiac MR imaging is a useful tool to confirm the presence of cardiomyopathy (76). Risk factors for peripartum cardiomyopathy include advanced maternal age, multiparity, African race, twins, gestational hypertension, and tocolysis (77). Cardiac MR is also a useful tool for follow-up, with only half of patients recovering full function (78).

**Figure 7**

**Peripartum cardiomyopathy.**—Specific cardiac MR imaging appearances have been relatively recently described in patients with peripartum cardiomyopathy (Fig 7, Movie 9 [online]) (78,79). Cine SSFP sequences allow an excellent assessment of LV function (79). Late gadolinium enhancement may demonstrate late enhancement predominately in the midmyocardium, involving the anterior and anterolateral segments (80). These abnormalities may regress over time, corresponding to an improvement in LV function.

Gadolinium-based contrast agents should be used with caution during pregnancy. Although there are no documented teratogenic effects, evidence in the literature is scarce and predominantly relies on animal data and limited case series. Gadopentetate dimeglumine inadvertently administered in three cases at 1, 3, and 5 months gestation and in another 11 women at 16–37 weeks gestation were not associated with subsequent fetal abnormality (81,82). Gadopentetate dimeglumine (0.1 mmol/kg) used to image the placenta was not associated with adverse effects on the fetus. Similarly, no negative outcomes were experienced in a study of 27 women who were administered gadolinium in the first trimester of pregnancy (83).

**Takotsubo cardiomyopathy.**—Takotsubo cardiomyopathy is a relatively recently described cardiomyopathy provoked by emotional stress and is most commonly seen in postmenopausal women (84). The clinical manifestation is similar to that of acute myocardial infarction with acute onset of congestive heart failure and typical electrocardiogram changes in the anterior leads, with absence of significant coronary artery disease at coronary angiography. Takotsubo cardiomyopathy is characterized by an acute transient stunning of the apical myocardium resulting in a transient apical ballooning with bulging out of the LV apex and a hypercontractile LV base. It is likely that there are multiple causative factors, including vasospasm and an abnormal response to catecholamines. Histopathologic analysis has revealed that in the acute phase, vacuoles of different sizes induce cellular hypertrophy along with increased intracellular glycogen (85). Abnormal-
Cardiac Amyloidosis

Amyloidosis is classified into many different forms on the basis of the amyloid precursor protein: immunoglobulin light chain (AL) derived—amyloidosis, reactive (secondary) AA amyloidosis, transthyretin (ATTR)-related hereditary amyloidosis, and β2-microglobulin (Abeta2M)-derived dialysis-related amyloidosis (88). Primary systemic amyloidosis (immunoglobulin light chain) and hereditary transthyretin (TTR) types cause cardiomyopathies that result in restrictive ventricular filling and, usually, a poor prognosis (89). Cardiac accumulation of these various proteins in the insoluble fibrillar amyloid conformation occurs principally in the myocardial interstitium. There often is associated endomyocardial fibrosis leading to diastolic dysfunction.

Important cardiac MR sequences in amyloidosis.—A diffuse decrease in signal intensity on T1-weighted fast spin-echo images may be found in cardiac amyloid, although this requires formal measurement in a region of interest (90,91). Ventricular myocardial thickening affects the right and left ventricles, and a useful distinguishing feature from HCM is that it generally results in a diffuse rather than a focal pattern of hypertrophy (Fig 9, Movie 11 [online]). Thickening (>6 mm) of the interatrial septum and posterior right atrial wall is also suggestive of cardiac amyloidosis (63). Marked thickening of the LV wall is associated with a survival time of less than 6 months (64). Aside from the fast spin-echo sequences, late-enhancement sequences have been found to demonstrate diffuse or subendocardial late enhancement (Fig 9) (89,92). It can be challenging to depict these areas on cardiac MR images, because the total amyloid protein load results in rapid washout of gadolinium-based contrast material from the myocardium. Thus, it can be difficult to determine the optimal inversion time to null normal myocardium, because it may be unclear which myocardial areas are normal (93). Typically, a short-axis image will be acquired by using multiple inversion times. The time at which remote myocardium passes through the null point should be used to render normal myocardium as bright regions and areas of amyloid disposition as bright regions. The time to commence imaging for late enhancement is shorter than that for other cardiomyopathies and should begin early (5 minutes after contrast agent injection). Focal enhancement correlates significantly with areas of regional hypokinesis or akinesis (92). When compared with the diagnostic test of choice of endomyocardial biopsy, cardiac MR imaging provides good sensitivity (80%) and high specificity (94%) (93). The authors of one study (94) showed that the abnormal gadolinium kinetics, specifically the 2-minute post-gadolinium intramyocardial inversion time difference between subepicardium and subendocardium at a threshold value of 23 msec, predicted mortality with 85% accuracy.
Cardiac abnormalities are caused by infiltration of sarcoïd granulomas (95). The classic clinical manifestation is heart block; however, other clinical features of sarcoïd heart disease include congestive heart failure, cor pulmonale, supraventricular and ventricular arrhythmias, conduction disturbances, ventricular aneurysms, pericardial effusion, and sudden death. About 7% of patients with sarcoïdosis develop cardiac symptoms, but postmortem studies have revealed cardiac involvement in 20%–30% of patients (96). Accurate diagnosis relies on endomyocardial biopsy findings for noncaseating granulomas or on a positive biopsy result from noncardiac tissue and cardiac abnormalities for which other possible causes have been excluded (95).

**Important cardiac MR sequences in sarcoïdosis.**—T2-weighted fast spin-echo images (usually short axis) may depict sarcoïd lesions as patchy hyperintense areas in myocardium, which may be transmural and characteristically causes LV wall thinning (Fig 10, Movie 12 [online]) (97). Late gadolinium enhancement may also demonstrate sarcoïd lesions (96). Such lesions are characteristically patchy and usually appear in the midwall and subepicardium but may involve any layer of ventricular myocardium and can mimic coronary artery disease (98). Careful attention should be paid to the RV myocardium and RVOT, which can also show enhancement. If a tissue diagnosis is required, cardiac MR imaging may provide optimal guidance for endomyocardial biopsy (99). Improvement in cardiac symptoms and MR findings has been observed after steroid therapy (97,100).

**Siderotic Cardiomyopathy**

Cardiac iron overload is a common problem worldwide affecting patients with β-thalassemia major (101). A mutation in the β-globin gene causes defective erythropoiesis that, in its homozygous form, results in severe anemia. Over 70% of these patients die of heart failure. Measurement of myocardial iron by using biopsy specimens is invasive and difficult to use for monitoring. Myocardial iron content cannot be predicted on the basis of serum ferritin or liver iron levels, and conventional assessments of cardiac function can only demonstrate those with advanced disease. Severe LV systolic dysfunction and heart failure are signs of advanced cardiac siderosis, which usually does not respond as well to chelation therapy as do earlier disease stages. Early diagnosis and treatment of myocardial iron overload is likely to prevent the mortality seen in patients with established ventricular dysfunction. The conventional treatment for severe myocardial disease with heart failure is long-term, continuous, high-dose intravenous deferoxamine. Preliminary evidence suggests that this approach is effective and may result in a reversal of diastolic heart failure.

**Important cardiac MR sequences in iron overload.**—Short-axis SSFP images provide an accurate assessment of global ventricular function that may be affected in iron overload cardiomyopathy.
(Movie 13 [online]). Furthermore, several studies have now proved the utility of specific T2*-weighted sequences for noninvasive quantification of ventricular iron deposition (102). The sequence can be performed by acquiring a single 10-mm-thick short-axis midventricular section of the LV at 8–10 echo times (2.6–16.7 msec at 2.02-msec increments) with standard shimming in a single breath hold and a flip angle of 20°, a matrix of 128 × 128 pixels, a field-of-view of approximately 40 cm, and a sampling bandwidth of 1950 Hz/pixel (Fig 11a). A delay of 0 msec after the R-wave trigger is usually chosen to obtain a high-quality image when blood flow and myocardial wall motion artifacts are minimized. A homogeneous full-thickness region of interest is placed in the LV septum encompassing both epicardial and endocardial regions. The signal intensity of this region is measured for each image and plotted against echo time to form an exponential decay curve (Fig 11b).

To derive mean T2*, an exponential trend line is fitted with an equation in the form $y = K_e^{-TE/T2*}$, where $K_e$ is a constant, TE is echo time, and $y$ is the signal intensity on the image (Fig 11c).

Several studies have shown a clear relationship between reduced myocardial T2* (<20 msec), indicating iron overload, and LV dysfunction. Cardiac MR imaging has been used to evaluate different chelation regimes specifically for their action on the myocardium, and myocardial T2* increase correlates well with the recovery of LV function after intravenous iron chelation (101,102). The sequence is also useful for other causes of myocardial iron overload such as hemochromatosis, sickle cell disease, and in cases where multiple blood transfusions are required (eg, certain myelodysplasias).

**Scleroderma**

Scleroderma is a connective tissue disorder characterized by vascular and fibrotic lesions. It predominates in the skin but can also be found in the lungs, kidneys, and heart (103). Myocardial fibrosis has been documented to be a common finding in pathology and autopsy studies (104).

The exact prevalence is difficult to assess because patients often have occult disease, and autopsy studies are biased toward those with the most severe disease. The development of clinical symptoms and signs is generally a poor prognostic indicator, with a 5-year survival rate of 30%. The reasons for this poorer prognosis include the development of progressive RV and LV failure, ventricular arrhythmias, coronary artery disease, and pericardial disease (105).

**Important cardiac MR sequences in scleroderma.**—Specific cardiac MR appearances have been relatively recently described for late-enhancement sequences (Fig 12, Movie 14 [online]) (106). Late-enhancing myocardial segments may be identified in up to 66% of patients, with a characteristic linear midmyocardial distribution predominantly affecting the basal and midventricular levels. Its presence is associated with development of arrhythmias.

**Unknown Cardiomyopathy: A Practical Approach**

When a patient is referred for cardiac MR imaging, an important initial task is to first gain as much clinical information about the suspected diagnosis Obtaining electrocardiographic and echocardiographic findings is very valuable, as well. Do not acquire or report on cardiac MR images in a clinical vacuum—discuss the case with the referring doctor if possible. A practical imaging approach is to perform (after acquisition of the appropriate localizers) a set of axial sequences through...
Cardiac MR is an excellent imaging technique for assessing the morphologic and functional characteristics of cardiomyopathy. The use of late-enhancement sequences has an important impact on the ability to characterize the myocardium and also aids in improving clinical risk stratification. Finally, the lack of ionizing radiation makes cardiac MR imaging an important tool in screening and serial assessment of progressive myocardial diseases.

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