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

**Variant 1:** Suspected arrhythmogenic cardiomyopathy.

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
</tr>
</thead>
<tbody>
<tr>
<td>MRI heart function and morphology without and with IV contrast</td>
<td>9</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>US echocardiography transthoracic resting</td>
<td>8</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>MRI heart function and morphology without IV contrast</td>
<td>7</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>CT heart function and morphology with IV contrast</td>
<td>6</td>
<td>This procedure is an alternative to MRI if the patient has a pacemaker or other contraindication.</td>
<td>☢☢☢☢</td>
</tr>
<tr>
<td>Arteriography coronary with ventriculography</td>
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<td>☢☢☢</td>
</tr>
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<td>CT chest without IV contrast</td>
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<td>X-ray chest</td>
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<td>FDG-PET/CT heart</td>
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<td>CT coronary calcium</td>
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<tr>
<td>US echocardiography transesophageal</td>
<td>1</td>
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<td>O</td>
</tr>
<tr>
<td>Arteriography pulmonary</td>
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**Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level*
**Clinical Condition:** Nonischemic Myocardial Disease with Clinical Manifestations (Ischemic Cardiomyopathy Already Excluded)

**Variant 2:** Suspected myocardial infiltrative disease.

<table>
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<th>Comments</th>
<th>RRL*</th>
</tr>
</thead>
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<td>9</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>US echocardiography transthoracic resting</td>
<td>8</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>X-ray chest</td>
<td>7</td>
<td></td>
<td>☢</td>
</tr>
<tr>
<td>FDG-PET/CT heart</td>
<td>7</td>
<td>This procedure is as good as MRI for initial diagnosis of sarcoidosis and better than MRI for follow-up.</td>
<td>☢ ☢ ☢ ☢</td>
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<tr>
<td>MRI heart function and morphology without IV contrast</td>
<td>6</td>
<td></td>
<td>O</td>
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<td>5</td>
<td>Perform this procedure for associated valve disease.</td>
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<td>CT heart function and morphology with IV contrast</td>
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<td>This procedure is an alternative to MRI if the patient has a pacemaker or other contraindication to MRI.</td>
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<td>CT coronary calcium</td>
<td>2</td>
<td></td>
<td>☢ ☢ ☢</td>
</tr>
<tr>
<td>Te-99m V/Q scan lung</td>
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<td>☢ ☢ ☢</td>
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<td>Arteriography coronary with ventriculography</td>
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<td>☢ ☢ ☢</td>
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</tr>
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<td>MRI chest without and with IV contrast</td>
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<tr>
<td>MRI chest without IV contrast</td>
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</tr>
<tr>
<td>Arteriography pulmonary</td>
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<td>☢ ☢ ☢ ☢</td>
</tr>
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</table>

**Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level*
**Clinical Condition:** Nonischemic Myocardial Disease with Clinical Manifestations (Ischemic Cardiomyopathy Already Excluded)

**Variant 3:** Suspected hypertrophic cardiomyopathy.

<table>
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<th>Comments</th>
<th>RRL*</th>
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<td>O</td>
</tr>
<tr>
<td>US echocardiography transthoracic resting</td>
<td>9</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>Arteriography coronary with ventriculography</td>
<td>8</td>
<td>Perform this procedure prior to alcohol septal ablation or to assess left ventricular pressure and gradients.</td>
<td>☢☢☢</td>
</tr>
</tbody>
</table>
Clinical Condition: Nonischemic Myocardial Disease with Clinical Manifestations (Ischemic Cardiomyopathy Already Excluded)

Variant 4: Suspected acute/subacute myocardial disease.

<table>
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<tr>
<th>Radiologic Procedure</th>
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<th>Comments</th>
<th>RRL*</th>
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<td>MRI heart function and morphology without and with IV contrast</td>
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<td></td>
<td>O</td>
</tr>
<tr>
<td>US echocardiography transthoracic resting</td>
<td>8</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>X-ray chest</td>
<td>7</td>
<td>Perform this procedure to assess systemic inflammation.</td>
<td>☢</td>
</tr>
<tr>
<td>MRI heart function and morphology without IV contrast</td>
<td>6</td>
<td>Perform this procedure if contrast cannot be given or if right disease is suspected.</td>
<td>O</td>
</tr>
<tr>
<td>US echocardiography transesophageal</td>
<td>4</td>
<td>Perform this procedure primarily to assess left atrial thrombus.</td>
<td>O</td>
</tr>
<tr>
<td>Arteriography coronary with ventriculography</td>
<td>3</td>
<td></td>
<td>☢☢☢☢</td>
</tr>
<tr>
<td>CT heart function and morphology with IV contrast</td>
<td>2</td>
<td></td>
<td>☢☢☢☢</td>
</tr>
<tr>
<td>CTA chest with IV contrast</td>
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<td></td>
<td>☢☢☢</td>
</tr>
<tr>
<td>MRI chest without and with IV contrast</td>
<td>2</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>MRI chest without IV contrast</td>
<td>2</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>FDG-PET/CT heart</td>
<td>2</td>
<td>This procedure may be considered in select cases to look for inflammatory causes of myocarditis-like disorders.</td>
<td>☢☢☢☢</td>
</tr>
<tr>
<td>CT chest without IV contrast</td>
<td>1</td>
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<tr>
<td>CT coronary calcium</td>
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<tr>
<td>Te-99m V/Q scan lung</td>
<td>1</td>
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<tr>
<td>Arteriography pulmonary</td>
<td>1</td>
<td></td>
<td>☢☢☢</td>
</tr>
</tbody>
</table>

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level
NONISCHEMIC MYOCARDIAL DISEASE WITH CLINICAL MANIFESTATIONS
(ISCHEMIC CARDIOMYOPATHY ALREADY EXCLUDED)

Expert Panel on Cardiac Imaging: Leena Mammen, MD1; Pamela K. Woodard, MD2; Suhny Abbara, MD3; Sharmila Dorbala, MD4; Cylen Javidan-Nejad, MD5; Paul R. Julsrud, MD6; Jacobo Kirsch, MD7; Christopher M. Kramer, MD8; Rajesh Krishnamurthy, MD9; Archana T. Laroia, MD10; Amar B. Shah, MD11; Jens Vogel-Clausen, MD12; Richard D. White, MD13

Summary of Literature Review

Introduction/Background
Symptomatic myocardial disease, which is not the result of flow-limiting coronary artery disease or prior myocardial infarction, is commonly referred to as nonischemic myocardial disease or cardiomyopathy (CM). Clinical manifestations of nonischemic myocardial disease include arrhythmia, palpitations, heart failure, shortness of breath, dyspnea, lower extremity edema, ascites, syncope, and chest discomfort. At times, the symptoms are so generalized and/or nonspecific that it may be difficult to identify myocardial disease as the etiology of the illness. Early and accurate detection and characterization of myocardial disease therefore becomes critical for appropriate patient treatment and management and to potentially avoid disease progression.

Nonischemic CM is more common in women and younger individuals. In general, the prognosis of nonischemic myocardial disease or CM is better than in ischemic myocardial disease or CM [1], although specific therapy such as revascularization may improve symptoms in the latter. Cardiac dysfunction can further be classified as systolic or diastolic or a combination of both. Most commonly the left ventricle (LV) is affected, but the right ventricle (RV) can also be affected in certain disease states. Systolic heart failure is diagnosed when there is reduction in the ventricular ejection fraction. Symptoms of heart failure occur when the degree of myocardial dysfunction exceeds compensatory mechanisms that occur to maintain adequate cardiac output and oxygenation. An LV ejection fraction (LVEF) less than 50% is below normal range, but some patients are asymptomatic with even lower ejection fractions. Nonischemic myocardial disease can also be secondary to other cardiac or systemic disease processes, such as valvular disease or recurrent tachyarrhythmia.

Conversely, heart failure with normal systolic function, also referred to as diastolic heart failure, is an increasingly recognized illness. There is a growing body of literature regarding diastology, which is the study of relaxation of the myocardium. Various diseases produce diastolic dysfunction and result in inadequate relaxation and filling of the ventricle(s). Myocardial fibrosis and infiltrative diseases such as amyloid and sarcoid are examples. Alternatively, there may be pericardial disease causing constriction of the cardiac chambers, resulting in inadequate diastolic filling. Symptoms of diastolic abnormality may be typical for congestive heart failure as well as symptoms of right heart failure, which include ascites, hepatic congestion, and lower extremity edema.

Overview of Imaging Modalities
In most cases of systolic heart failure, ischemic CM should be considered and excluded prior to assessment for nonischemic CM. Echocardiography (echo) is the mainstay of evaluating left ventricular function due to ease of access and widespread use [2]. Valvular function and diastolic function can also be evaluated. Advanced echo techniques such as pulsed tissue Doppler imaging and 3-D imaging have added diagnostic and prognostic value in the evaluation of nonischemic CM [3-5]. Stress physiology assessment via treadmill exercise, rest and stress radionuclide myocardial perfusion imaging (SPECT MPI), or stress echo are also commonly used in the evaluation of CM primarily to exclude ischemic etiologies [6]. Dobutamine and adenosine stress functional cardiac magnetic resonance imaging (MRI) provide high sensitivity and specificity for ischemia. If these studies are inconclusive, coronary angiography with ventriculography may be considered. In cases of low-to-intermediate likelihood for coronary artery disease, coronary computed tomography angiography (CCTA) can be performed for

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direct coronary artery evaluation to differentiate ischemic versus nonischemic CM [7,8]. Since elevated pulmonary arterial pressure is a predictor of death in patients with heart failure, echo and/or right heart catheterization may be considered [9]. Evaluating right heart pressures is also useful in evaluating restrictive cardiomyopathies.

Recent advances in technology allow further reduction of radiation dose from CCTA; newly available dose-reducing techniques include prospective triggering, adaptive statistical iterative reconstruction, and high-pitch spiral acquisition [10-14]. These new lower-dose techniques are appropriate in patients with low heart rates (<65 bpm) who are in sinus rhythm. If function evaluation is desired, the retrospective gating technique must be utilized. If chronic pulmonary embolism is being considered, a chest CTA can be performed [15]. Alternatively, a ventilation perfusion examination can be done if a patient has contraindication to iodinated contrast [16]. The role of positron emission tomography (PET) in the assessment of cardiac sarcoidosis is well established, and its role in cardiomyopathies has been studied to a limited extent for the localization of scar for cardiac resynchronization therapy [17].

Coronary calcium scoring (CCS) is most commonly used for risk stratification in asymptomatic patients. In a large study of 10,377 subjects it has been shown that CCS provides independent incremental information in addition to traditional risk factors in the prediction of all-cause mortality [18].

Cardiac MR (CMR) imaging has emerged as a powerful tool for the diagnosis of ischemic and nonischemic cardiomyopathies [19]. It is now considered the reference standard imaging technique to assess myocardial anatomy, regional and global function, and viability—and often reveals the underlying etiology of heart failure [20,21]. CMR allows a multifaceted approach for the evaluation of new onset heart failure [22]. Advanced MRI techniques with tissue imaging and delayed myocardial enhancement can provide information beyond echo for tissue characterization in CM [23].

In nonischemic CM, delayed myocardial enhancement usually does not occur in a coronary artery distribution and is often mid wall or subepicardial rather than subendocardial or transmural [24]. Localization of pathology can also guide myocardial biopsy to the affected area, increasing its yield [25]. In addition, MRI is increasingly being used for evaluation of genetically positive, phenotypically negative patients for risk stratification [26,27], and the prognostic value of delayed enhancement in CMR has been described [28-30].

**Variant: Suspected Arrhythmogenic Cardiomyopathy**

A patient may suffer from palpitations, and ECG/Holter monitoring may reveal a pattern of arrhythmias, which may indicate a need to exclude underlying structural heart disease. Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is caused by genetic mutations, which result in fibrofatty infiltration of the myocardium, most often the RV. RV dilation and dysfunction are hallmarks of ARVC/D and set the stage for life-threatening arrhythmias [31]. Morphologic and genetic variants of this disease have been investigated [32]. Advances in echo techniques have aided evaluation of the right heart [33]. CT can also provide a significant role in morphologic evaluation of the RV, particularly in patients with implantable cardioverter-defibrillators (ICD) who cannot have MRI examinations [34]. MRI has significant contribution in evaluating the morphology and function of the RV. This is a significant advantage for ARVC/D and also for other diseases such as decompensated RV function from pulmonary hypertension in the setting of chronic pulmonary embolism. Another advantage of MRI is the ability to evaluate RV fibrous tissue, which enhances in a delayed fashion, and RV fat [35].

**Variant: Suspected Myocardial Infiltrative Disease**

Sarcoid and amyloid are the most common infiltrative diseases involving the myocardium [36]. Sarcoid is a granulomatous disease that can affect any organ and has a wide spectrum of clinical manifestations. Cardiac sarcoid may be symptomatic or asymptomatic. The most common clinical presentations are heart block, dilated CM, and ventricular arrhythmias. Because the yield of endomyocardial biopsy for the suspected diagnosis is low from patchy disease, noninvasive imaging is often pursued. Echo and MRI are the most commonly used imaging modalities, although CT is often used in search of mediastinal adenopathy and lung abnormalities [37]. PET using 18-F-fluoro-2-deoxyglucose (FDG-PET) may offer earlier detection of cardiac sarcoidosis and indicate areas of active disease [38]. MRI with late gadolinium enhancement has shown higher sensitivity in the detection of cardiac sarcoidosis than standard clinical evaluations (ECG, thallium scintigraphy, and echo) [39-41]. Early treatment is crucial in improving symptoms and prognosis[42].
Amyloidosis is a systemic condition characterized by the extracellular deposition of amyloid into one or more organ systems. Cardiac deposition leads to an infiltrative/restrictive CM [43]. On imaging, the LV is hypertrophied, the valves and the interatrial septum are often thickened, and the LV exhibits diastolic dysfunction [44]. MRI has an increasing role in the diagnosis of amyloid with characteristic patterns of myocardial delayed enhancement [45].

Chagas disease is rare in the United States but may be seen in the southern states and in California, especially in immigrants and visitors from Central and South America. Migratory movements can raise the suspicion of this parasitic infiltrative myocardial disease. CMR has emerged as the noninvasive tool to evaluate the myocardial fibrosis typical of Chagas disease [46].

Iron overload CM is considered an infiltrative cardiomyopathy, which results in diastolic and systolic dysfunction. It may be primary with a genetic basis as in hemochromatosis. It may also be secondary due to various transfusion dependent anemias [47,48]. Advances in cardiac MRI have allowed not only the diagnosis but also the quantification of myocardial iron with T2* evaluations for the purposes of early diagnosis and guiding chelation therapy [49-51].

**Variant: Suspected Hypertrophic Cardiomyopathy**

Hypertrophic CM (HCM) is a genetically based myocardial disease with a wide spectrum of genotypic abnormalities and phenotypic presentations. The distribution of the hypertrophy may be concentric, involving the entire LV, or may asymmetrically affect the LV. Most commonly, the septum or apex is hypertrophied if the distribution is asymmetric. Although echo remains the mainstay modality for diagnosis, diagnostic imaging can be performed with CT and MRI as well [52,53]. LV outflow obstruction and systolic anterior motion of the mitral valve are characteristic features of hypertrophic obstructive cardiomyopathy (HOCM) and can be evaluated by echo and CMR. True short-axis views of the LV by CMR can be helpful in obtaining accurate measurement of LV wall thickness, one of several criteria for ICD placement. MRI may be particularly useful in distinguishing hypertensive CM from symmetric HCM by characteristic enhancement patterns [54]. Fabry disease can be considered when there is symmetric hypertrophy, and MRI may also be helpful in differentiating Fabry disease and HCM [55].

**Variant: Suspected Acute/Subacute Myocardial Disease**

Acute and subacute nonischemic myocardial diseases can cause troponin elevation and mimic ischemic causes of chest pain. Myocarditis is inflammation of the myocardium and may have a viral etiology or be idiopathic. Echo might show global or regional wall motion abnormalities, which may be concerning for ischemia. CMR provides additive information with the ability to image tissue edema and characteristic midwall or subepicardial patterns of delayed enhancement. This can be very useful in differentiating troponin elevation in acute myocardial infarction [56,57]. In addition, associated pericardial thickening or inflammation can be evident.

Stress CM, also known as takotsubo CM, produces transient myocardial dysfunction, which resolves in a period of days to weeks [58]. The wall motion abnormalities are regional, dyskinetic, and in a non-coronary distribution. The most common variant is apical ballooning, with occasional variations of basal and mid-ventricular ballooning. The patient may present with acute chest pain worrisome for myocardial infarction, and imaging is helpful to diagnose this condition in the absence of flow-limiting coronary artery disease [59,60]. Cardiotoxic medications such as chemotherapeutic agents can have either temporary or permanent effect on myocardial function and should also be considered in acute/subacute development of systolic cardiac dysfunction.[61].

**Suspected Familial or Genetically Transmitted Cardiomyopathy**

Familial and/or genetically transmitted cardiomyopathies are increasingly recognized, and several have been described in the text above. Heart failure in complex congenital heart diseases is multifactorial and may have a genetic basis, but this is beyond the scope of this document. The imaging for these cardiomyopathies is quite varied depending on the particular disease and therefore will not be covered in the variant tables. The most commonly recognized myocardial diseases with a genetic basis are HCM, dilated CM, and ARVC/D. Less common are deposition diseases such as Fabry disease and hemochromatosis. Isolated left ventricular noncompaction is a rare but increasingly recognized congenital CM characterized by prominent trabecular, deep intertrabecular recesses, and thickened myocardium with 2 distinct layers (compacted and noncompacted) [62-65].
Summary

- Imaging plays an important role in the diagnosis of nonischemic cardiomyopathies.
- Newer technologies involving CT and MRI have shown promise in improving earlier detection and improved accuracy of diagnosis of several cardiomyopathies.
- Although it is expected that traditional imaging modalities such as echo will continue to play a primary role in nonischemic cardiomyopathies, MRI in particular is expected to play an increasing role in both diagnosis and management.

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, both because of 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 to 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.

<table>
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<th>Pediatric Effective Dose Estimate Range</th>
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<tr>
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</tr>
<tr>
<td>☢☢☢☢☢</td>
<td>30-100 mSv</td>
<td>10-30 mSv</td>
</tr>
</tbody>
</table>

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

Supporting Documents

For additional information on the Appropriateness Criteria methodology and other supporting documents go to www.acr.org/ac.

References


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.