American College of Radiology
ACR Appropriateness Criteria®
Suspected Acute Aortic Syndrome

Variant 1:   Acute chest pain; suspected acute aortic syndrome.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transesophageal	Usually Appropriate	O
Radiography chest	Usually Appropriate	☢
MRA chest abdomen pelvis without and with IV contrast	Usually Appropriate	O
MRA chest without and with IV contrast	Usually Appropriate	O
CT chest with IV contrast	Usually Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Appropriate	☢☢☢
CTA chest with IV contrast	Usually Appropriate	☢☢☢
CTA chest abdomen pelvis with IV contrast	Usually Appropriate	☢☢☢☢☢
US echocardiography transthoracic resting	May Be Appropriate	O
Aortography chest	May Be Appropriate	☢☢☢
MRA chest abdomen pelvis without IV contrast	May Be Appropriate	O
MRA chest without IV contrast	May Be Appropriate	O
MRI chest abdomen pelvis without IV contrast	May Be Appropriate	O
CT chest without IV contrast	May Be Appropriate	☢☢☢
CTA coronary arteries with IV contrast	May Be Appropriate	☢☢☢
MRI chest abdomen pelvis without and with IV contrast	Usually Not Appropriate	O

SUSPECTED ACUTE AORTIC SYNDROME

Expert Panel on Cardiac Imaging: Gregory A. Kicska, MD, PhD[a]; Lynne M. Hurwitz Koweek, MD[b]; Brian B. Ghoshhajra, MD, MBA[c]; Garth M. Beache, MD[d]; Richard K.J. Brown, MD[e]; Andrew M. Davis, MD, MPH[f]; Joe Y. Hsu, MD[g]; Faisal Khosa, MD, MBA[h]; Seth J. Kligerman, MD[i]; Diana Litmanovich, MD[j]; Bruce M. Lo, MD, RDMS, MBA[k]; Christopher D. Maroules, MD[l]; Nandini M. Meyersohn, MD[m]; Saurabh Rajpal, MD[n]; Todd C. Villines, MD[o]; Samuel Wann, MD[p]; Suhny Abbara, MD.[q]

Summary of Literature Review

Introduction/Background

Acute aortic syndrome (AAS) includes the entities of acute aortic dissection (AD), intramural hematoma (IMH), and penetrating atherosclerotic ulcer (PAU). AAS typically presents with sudden onset of severe, tearing, anterior, or interscapular back pain [1]. Symptoms may be dominated by malperfusion syndrome due to obstruction of the lumen of the aorta and/or a side branch when the intimal and medial layers are separated. Risk factors include hypertension, family history, and underlying collagen vascular disorders [2]. Timely diagnosis of AAS is crucial to permit prompt management; early mortality rates are reported to be 1% to 2% per hour after the onset of symptoms for untreated ascending AD [3]. Medical management of acute ascending AD is associated with a mortality rate of nearly 20% by 24 hours after presentation, 30% by 48 hours, 40% to 70% by day 7 [4], and 50% by 1 month [3,5]. The major cause of early death with AAS is aortic rupture [6]. AAS limited to the descending aorta may be managed medically and/or with open surgical or endovascular treatment based on extent of disease, aortic size, malperfusion of end organs, and clinical parameters; medical management is usually undertaken unless the aorta is dilated and/or there is mesenteric or limb ischemia [6]. IMH occurs with approximately 10% of the frequency of AD [7], and can be seen in isolation or in conjunction with AD and PAU.

Imaging findings of AAS include any disruption of the intimal and medial layers, either with an IMH within the media (with or without the presence of a penetrating ulcer) or with an intimal flap. The prevalence of isolated IMHs in patients with suspected AAS is reported to be 21% to 30% [3]. Over time, 28% to 47% of patients may progress to a classic AD, and in other patients, the IMH will resolve with or without aneurysmal enlargement of the aorta [6]. PAUs that disrupt the intima most commonly occur in the mid and distal descending aorta [8].

Imaging studies in the evaluation of suspected thoracic AAS should be directed toward confirming its presence; classifying location using the Stanford type A or B (and/or DeBakey types I, II, or III); identifying entry and reentry sites; evaluating the patency of the false lumen; detecting the presence or absence of aortic branch involvement; assessing involvement of the coronary ostia; evaluating aortic valve competency; and determining the presence or absence of extravasated blood in the mediastinal, pleural, or pericardial spaces and sizing of the aorta. Imaging should provide information as to the next step in management, whether medical, open surgical repair, and/or endovascular management. For additional information on aortic imaging, please see the ACR Appropriateness Criteria® topic on "Nontraumatic Aortic Disease" [9].

Classification of Acute Aortic Syndrome

Classification of AAS is based on identifying the location of the most proximal location of the intimal flap and/or IMH. In DeBakey classification type I and type II ADs, the proximal intimal tear is located in the ascending aorta, usually just a few centimeters above the aortic valve. In type I dissection, the intimal flap extends for a variable distance beyond the aortic arch and usually into the descending aorta, whereas in type II the intimal flap is confined to the ascending aorta. DeBakey type III dissection originates in the descending aorta, usually just beyond the origin

[a]University of Washington, Seattle, Washington. [b]Panel Chair, Duke University Medical Center, Durham, North Carolina. [c]Panel Vice-Chair, Massachusetts General Hospital, Boston, Massachusetts. [d]University of Louisville School of Medicine, Louisville, Kentucky. [e]University of Michigan Health System, Ann Arbor, Michigan. [f]The University of Chicago Medical Center, Chicago, Illinois; American College of Physicians. [g]Kaiser Permanente, Los Angeles, California. [h]Vancouver General Hospital, Vancouver, British Columbia, Canada. [i]University of California San Diego, San Diego, California. [j]Harvard Medical School, Boston, Massachusetts. [k]Sentara Norfolk General/Eastern Virginia Medical School, Norfolk, Virginia; American College of Emergency Physicians. [l]Naval Medical Center Portsmouth, Portsmouth, Virginia. [m]Massachusetts General Hospital, Boston, Massachusetts. [n]Ohio State University, Nationwide Children's Hospital, Columbus, Ohio; Society for Cardiovascular Magnetic Resonance. [o]University of Virginia Health Center, Charlottesville, Virginia; Society of Cardiovascular Computed Tomography. [p]Ascension Healthcare Wisconsin, Milwaukee, Wisconsin; Nuclear cardiology expert. [q]Specialty Chair, UT Southwestern Medical Center, Dallas, Texas.

of the left subclavian artery, and propagates antegrade along the descending aorta. Rarely, the proximal intimal tear occurs in an unusual location, such as the abdominal aorta [6].

Stanford type A dissection refers to any dissection involving the ascending aorta and therefore is equivalent to DeBakey type I and type II. Stanford type B dissection is any AD that does not involve the ascending aorta, including dissections that originate in the aortic arch. Stanford type B dissection is therefore equivalent to DeBakey type III [10]. Both the DeBakey and Stanford classifications are ambiguous in regard to AD starting on the aortic arch. Such lesions may be best described as Stanford type B with arch involvement [6,11].

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) [12]:

> *"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.

Discussion of Procedures by Variant

Variant 1: Acute chest pain; suspected acute aortic syndrome

Aortography Chest
Historically, conventional angiography was considered the reference standard for diagnosing AAS with sensitivity, specificity, positive predictive value, and negative predictive value of 88%, 94%, 96%, and 84%, respectively, reported for identification of an intimal flap [13].

The diagnostic accuracy of digital subtraction angiography approaches 98% in some series for identification of an AD. Angiography permits management of patients who are critically ill and can also assess aortic regurgitation and aortic branch vessel involvement (including the coronary arteries) [14]. High frame rates facilitate identification of the intimal tear and the degree of aortic insufficiency. False-negative arteriograms may occur when the false lumen is not opacified, when there is simultaneous opacification of the true and false lumen, and when the intimal flap is not displayed in profile.

Disadvantages of angiography include the fact that it is invasive, requires iodinated contrast material, and that it has a limited ability to assess the surrounding structures (eg, presence of mediastinal hemorrhage), which is readily detected by other cross-sectional imaging modalities [1,3].

CT Chest Without IV Contrast
CT chest without intravenous (IV) contrast cannot assess the lumen of that aorta, but AAS can be inferred by identifying displaced aortic calcifications or IMHs, findings that are shown to be 94% and 71% sensitive for diagnosing AAS when using contrast-enhanced CTA as the reference standard [15]. Another study found that abnormal aortic wall contour and paraaortic hematoma were also predictive of AAS. These imaging findings were incorporated into a decision-rule that predicted the presence of AAS with a mean sensitivity of 93% [16]. However, few patients in the derivation data set had PAU as a diagnosis, and the number of patients with AAS in the validation data set was not reported.

CT without IV contrast can identify complications from AAS, such as mediastinal, pericardial, and/or pleural hematoma.

CT Chest With IV Contrast
CT chest with IV contrast can identify the presence of an AAS and the presence of complications such as mediastinal, pericardial, and/or pleural hematoma. There is no relevant literature that compares the diagnostic accuracy of CT chest with IV contrast (delayed-phase enhancement) versus CTA chest (arterial-phase contrast).

CT Chest Without and With IV Contrast

CT chest without and with IV contrast combines the diagnostic capabilities of a CT chest without and a CT chest with IV contrast. In one study of 36 patients, the interrater agreement on diagnosing IMH using CTA with IV contrast alone was $\kappa = 0.65$, a value that increased to 0.92 if the imager was also given a CT without IV contrast at the same time [17].

Dual-energy multidetector CT enables generation of CT chest images without and with IV contrast enhancement by producing virtual unenhanced imaging studies. However, the use of these virtual unenhanced images in place of true unenhanced images remains controversial [18-20].

CTA Coronary Arteries

There is no relevant literature supporting the use of CTA of the coronary arteries for the diagnosis of AAS. CTA of coronary arteries may be used in patients with known AD for which coronary arterial disease classification is needed prior to open surgical repair for an AD involving the ascending aorta.

This document includes the triple-rule-out (TRO) protocol under the topic of CTA of coronary arteries as the TRO includes simultaneous assessment of the pulmonary arteries, aorta, and coronary arteries.

CTA Chest With IV Contrast

CTA can demonstrate the presence of aortic intimal flaps, branch vessel involvement, and entry and reentry sites, as well as identification of penetrating ulcers. The addition of a noncontrast acquisition as part of the CTA examination can be used to confirm the presence of an IMH. CTA chest can also allow for simultaneous assessment of adjacent mediastinal, pericardial, and pleural spaces. CTA chest was the most common initial diagnostic test performed in patients enrolled in the International Registry of Acute Aortic Dissection, and is being used with increasing frequency [21]. CTA was used to diagnose AD with 82% sensitivity and 100% specificity [22]. Numerous prior studies evaluating the accuracy of CT use in diagnosing AD have demonstrated sensitivities of 90% to 100% but lower specificities ranging from 87% to 100% [23-26]. However, these studies evaluated conventional CT, which has largely been supplanted by faster multidetector CT. One multidetector CT study enrolling 57 patients has reported sensitivities and specificities of 100% [22]. CT findings may also be used to predict patient outcome. For example, in one retrospective study of 83 patients with type B dissections, the morphologic features of a dissection were associated with an increase in late adverse events [27].

Isolated IMH can also be diagnosed as hyperdense aortic wall thickening when using CTA chest with IV contrast. In one study of AAS that included 124 patients with IMH, all patients with IMH had aortic wall attenuation >45 HU [28]. In another study of 36 patients, IMH was diagnosed with a 68.4% sensitivity and 96.3% specificity [17]. The interrater agreement on diagnosing was $\kappa = 0.65$, a value that increased to 0.92 if the imager was also given a CT without IV contrast at the same time [17].

CTA with IV contrast also enables detection of focal intimal disruptions or also ulcer-like projections as prognostic indicators in IMH. In one study of 107 patients, aorta-related death or need for invasive surgery was increased when focal intimal disruptions or ulcer-like projections were present (hazard ratio, 24.43) [29].

CTA with IV contrast can be used to diagnose penetrating PAU, but contrast does not enable additional prognostic imaging findings. PAU prognosis has been associated with the presence or absence of aortic rupture, but in one study, aortic diameter, ulcer size, or the presence of IMH did not affect prognosis [30]. Compared with CTA chest, TRO CT protocols can be used to assess for additional, potentially fatal possibilities such as pulmonary embolism and acute coronary syndrome [31,32]. It has been reported that TRO CT can safely eliminate further diagnostic testing in >75% of patients in the appropriate patient population performed specifically for assessing AD [33].

CTA Chest, Abdomen, and Pelvis With IV Contrast

CTA of the chest, abdomen, and pelvis allows for imaging of both the thoracic and abdominal aorta, and it provides assessment of extension of the dissection along the thoracic, abdominal, and pelvic branch vessels with one injection of IV contrast and a single breath-hold acquisition. Postprocessing of the volumetric data set, using multiplanar reformatting and 3-D volume rendering, facilitates evaluation of the location and course of the intimal flap [34], branch vessel, and visceral organ involvement. A meta-analysis of 3 studies examining CTA for the diagnosis of dissection showed a pooled sensitivity of 100% and a specificity of 98% [21]. When CTA is used as a first-line test, the International Registry of Acute Aortic Dissection reported a sensitivity of 93% for diagnosing dissection [35].

MRA Chest Without IV Contrast
There is no relevant literature supporting the use of MR angiography (MRA) chest without IV contrast for the diagnosis of AAS. Several studies report noncontrast MRA techniques and compare them with contrast-enhanced techniques, but studies were neither blinded to the reference standard or nor did they contain control subjects [36,37].

MRA Chest Without and With IV Contrast
MRA of the chest has a reported sensitivity and specificity of 100% for diagnosing AD, has sensitivity of 85% and specificity of 100% for identifying the site of entry, and has 100% sensitivity and specificity for identifying thrombus and the presence of a pericardial effusion [24-26,35,38-40]. Excellent sensitivity (92%–98%) and specificity (100%) have been documented for contrast-enhanced MRA in acute and chronic AD [31,35].

MRA Chest, Abdomen, and Pelvis Without IV Contrast
There is no relevant literature supporting the use of MRA chest, abdomen, and pelvis without IV contrast for the diagnosis of AAS.

Although no literature directly examines the accuracy of diagnosing AAS with MRA chest, abdomen, and pelvis without IV contrast, the accuracy for identification of an AD in the chest is similar to that of an isolated MRA chest without IV contrast with the addition of MRA abdominal and pelvic assessment.

MRA Chest, Abdomen, and Pelvis Without and With IV Contrast
There is no relevant literature supporting the use of MRA chest, abdomen, and pelvis without and with IV contrast for the diagnosis of AAS.

Although no literature directly examines the accuracy of diagnosing AAS with MRA chest, abdomen, and pelvis without IV contrast, the accuracy for identification of an AD in the chest is similar to that of an isolated MRA chest without IV contrast with the addition of MRA abdominal and pelvic assessment.

MRI Chest, Abdomen, and Pelvis Without IV Contrast
There is no relevant literature supporting the use of MRI chest, abdomen, and pelvis without IV contrast for the diagnosis of AAS. In contrast to MRA, MRI does not contain sequences designed to specifically produce images of the aorta, such as gating, 3-D data sets, thin sections, or double-oblique planes. MRI contains sequences that produce images in orthogonal planes, with greater slice thickness.

MRI Chest, Abdomen, and Pelvis Without and With IV Contrast
There is no relevant literature supporting the use of MRI chest, abdomen, and pelvis without and with IV contrast for the diagnosis of AAS. In contrast to MRA, MRI does not contain sequences designed to specifically produce images of the aorta, such as gating, 3-D data sets, thin sections, or double-oblique planes. MRI contains sequences that produce images in orthogonal planes, with greater slice thickness.

Radiography Chest
Chest radiographs can identify an enlarged aorta and or mediastinum and can identify complications such as pleural fluid. A substantial portion of patients with AAS may have a normal chest radiograph, and further imaging should be pursued despite a normal chest radiograph in cases of suspected AAS. In most cases when patients with an AAS have an abnormal radiograph, the findings are nonspecific [41]. The most recent report from the International Registry of Acute Aortic Dissection showed that in the last 10 years, the incidence of abnormal radiographic findings has decreased in patients with AD [10]. In that report, radiographs were reported as abnormal in 52% of patients with type A AD, whereas previously it was 61%. Abnormal findings were present in 39% of patients with type B AD, whereas previously it was 56%. Radiographs were reported to be completely normal in 36% to 38% of patients with AD. In another study that measured the diagnostic performance of radiography for all diagnoses of AAS, including IMH, PAU, and ruptured aneurysm, chest radiography had a sensitivity of 70.8% and a specificity of 82.5% [42].

US Echocardiography Transesophageal
Transesophageal echocardiography (TEE) has a reported sensitivity, specificity, and negative predictive value of up to 100% for diagnosis of AD [40,43-45]. Principal limitations of TEE are its blind spots in the distal ascending aorta and proximal transverse arch that are obscured by the air-containing trachea and left main bronchus. Diagnostic problems may also be encountered in the ascending aorta in which reverberation artifacts can result in false-positive diagnosis of AD [46].

The accuracy of TEE may be enhanced with the addition of IV ultrasound (US) contrast. In one study of 66 patients, conventional TEE were compared with contrast-enhanced TEE for the diagnosis of AAS [47]. Of the 22 patients in the study with AD, TEE identified 20 patients and contrast-enhanced TEE identified all 22, suggesting increased accuracy in diagnosis of penetrating aortic ulcers and IMHs with IVUS contrast administration.

US Echocardiography Transthoracic Resting

Transthoracic echocardiography (TTE) has a reported sensitivity of 59% to 85% and a specificity of 93% to 96% [39,40,44,48] for detection of AD. This provides greater utility for ruling in as opposed to ruling out a diagnosis of AAS. It is useful in the diagnosis of dissection involving the ascending aorta and can diagnose the hemodynamic significance of pericardial effusions, the degree of aortic regurgitation, and left ventricular function [25]. TTE is of marginal value in diagnosing distal descending thoracic dissections because of the limited availability of US windows [31]. Although the sensitivity of TTE in detecting descending ADs was previously reported to be lower (31%–80%) than CTA and MRA, recent technical innovations like harmonic imaging and microbubble contrast enhancement have been demonstrated to improve sensitivity of TTE in detecting descending dissections to 84% [49].

Summary of Recommendations

- **Variant 1:** US echocardiography transesophageal, radiography chest, or MRA chest abdomen pelvis without and with IV contrast, MRA chest without and with IV contrast, CT chest with IV contrast, CT chest without and with IV contrast, CTA chest with IV contrast, or CTA chest abdomen pelvis with IV contrast is usually appropriate for the imaging of a patient with acute chest pain and suspected AAS. These procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).

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 [50].

Relative Radiation Level Designations		
Relative Radiation Level*	**Adult Effective Dose Estimate Range**	**Pediatric Effective Dose Estimate Range**
O	0 mSv	0 mSv
☢	<0.1 mSv	<0.03 mSv
☢☢	0.1-1 mSv	0.03-0.3 mSv
☢☢☢	1-10 mSv	0.3-3 mSv
☢☢☢☢	10-30 mSv	3-10 mSv
☢☢☢☢☢	30-100 mSv	10-30 mSv
*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies."		

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