American College of Radiology ACR Appropriateness Criteria® Nontraumatic Aortic Disease

Variant: 1 Congenital aortic disease. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	0
Radiography chest	Usually Appropriate	•
MRA chest and abdomen without and with IV contrast	Usually Appropriate	0
MRA chest and abdomen without IV contrast	Usually Appropriate	0
CTA chest and abdomen with IV contrast	Usually Appropriate	∵ • • •
US abdomen	May Be Appropriate	0
US echocardiography transesophageal	May Be Appropriate	0
Aortography chest and abdomen	May Be Appropriate	∵ • • •
CT chest and abdomen with IV contrast	Usually Not Appropriate	∵ • • •
CT chest and abdomen without and with IV contrast	Usually Not Appropriate	∵ ∵ ∵ ∵
CT chest and abdomen without IV contrast	Usually Not Appropriate	∵ ∵ ∵
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	※ ※ ※

<u>Variant: 2</u> Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRA chest and abdomen without and with IV contrast	Usually Appropriate	0
MRA chest and abdomen without IV contrast	Usually Appropriate	0
CTA chest and abdomen with IV contrast	Usually Appropriate	※ ※ ※
FDG-PET/CT skull base to mid-thigh	Usually Appropriate	※ ※ ※
US abdomen	May Be Appropriate	0
Radiography chest	May Be Appropriate	③
CT chest and abdomen with IV contrast	May Be Appropriate	⊗ ⊗ ⊗
CT chest and abdomen without and with IV contrast	May Be Appropriate	⊗ ⊗ ⊗
CT chest and abdomen without IV contrast	May Be Appropriate	⊗ ⊗ ⊗
US echocardiography transesophageal	Usually Not Appropriate	0
US echocardiography transthoracic resting	Usually Not Appropriate	0
Aortography chest and abdomen	Usually Not Appropriate	⊗⊗⊗

Variant: 3 Degenerative or atherosclerotic aortic disease. Initial imaging.

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Procedure	Appropriateness Category	Relative Radiation Level
US abdomen	Usually Appropriate	0
Radiography chest	Usually Appropriate	⊗
MRA chest and abdomen without and with IV contrast	Usually Appropriate	0
MRA chest and abdomen without IV contrast	Usually Appropriate	0
CTA chest and abdomen with IV contrast	Usually Appropriate	⊗ ⊗ ⊗
US echocardiography transthoracic resting	May Be Appropriate	0

CT chest and abdomen with IV contrast	May Be Appropriate	∵ ∵ ∵
CT chest and abdomen without IV contrast	May Be Appropriate	∵ • • •
US echocardiography transesophageal	Usually Not Appropriate	0
Aortography chest and abdomen	Usually Not Appropriate	
CT chest and abdomen without and with IV contrast	Usually Not Appropriate	
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	૽

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Summary of Literature Review

Introduction/Background

Nontraumatic aortic disease can be caused by a wide variety of disorders, including congenital, inflammatory, infectious, metabolic, neoplastic, and degenerative diseases. Such conditions include, but are not limited to, atherosclerosis, aortic dissection, intramural hematoma, penetrating aortic ulcer, aortic aneurysms of various etiologies (degenerative, mycotic, or vasculitis-related), aortic rupture, thrombosis, aortobronchial fistula, congenital disorders, and extrinsic compression from adjacent masses. Diagnostic imaging is essential to assess the anatomy and extent of morphological changes involving the aorta. Nontraumatic aortic diseases may affect the lumen, wall, or perivascular structures. When there is aortic branch vessel involvement, end organ perfusion may be compromised.

Often, aortic disorders involve both the thoracic and abdominal aortic segments, thus requiring imaging of both regions. The clinical symptoms of aortic diseases vary widely. For example, acute aortic syndrome presents acutely with chest pain and elevated blood pressure, whereas atherosclerosis may be asymptomatic and detected incidentally.

The guidelines proposed in this document pertain to the abovementioned categories. Readers are encouraged to review other ACR Appropriateness Criteria topics on "Chest Pain-Possible Acute Coronary Syndrome" [1], "Nonischemic Myocardial Disease with Clinical Manifestations (Ischemic Cardiomyopathy Already Excluded)" [2], "Suspected Pulmonary Embolism" [3], "Acute Chest Pain-Suspected Aortic Dissection" [4], "Pulsatile Abdominal Mass, Suspected Abdominal Aortic Aneurysm" [5], "Abdominal Aortic Aneurysm-Interventional Planning and Follow-up" [6] "Lower Extremity Arterial Revascularization-Post-Therapy Imaging" [7], and "Vascular Claudication-Assessment for Revascularization" [8] for further guidance.

Special Imaging Considerations

For the purposes of distinguishing between CT and CT angiography (CTA), ACR Appropriateness Criteria topics use the definition in the <u>ACR–NASCI–SIR–SPR Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA)</u> [9]:

"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.

Initial Imaging Definition

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

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

OR

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

Discussion of Procedures by Variant

Variant 1: Congenital aortic disease. Initial imaging.

Variant 1: Congenital aortic disease. Initial imaging. A. Aortography chest and abdomen

Catheter-based aortography is considered the reference standard for the diagnosis of congenital aortic diseases [10]. Aortography provides information regarding flow and allows hemodynamic measurements to be taken; however, several noninvasive studies can provide similar information. As such, the role of aortography in diagnosing aortic diseases is decreasing as the sensitivity of other noninvasive modalities, such as transthoracic echocardiography (TTE), CTA, and MR angiography (MRA) improves [11-18]. Aortography is now most commonly performed when an intervention is planned.

Variant 1: Congenital aortic disease. Initial imaging. B. CT Chest and Abdomen

There is no relevant literature available to examine the use of CT chest and abdomen with intravenous (IV) contrast alone in the management of congenital aortic disease. There is no relevant literature available to examine the use of CT chest and abdomen without and with IV contrast in the management of congenital aortic disease.

There is no relevant literature available to examine the use of CT chest and abdomen without IV contrast alone in the management of congenital aortic disease. Please see the "CTA chest and abdomen with IV contrast" section below for further discussion.

Variant 1: Congenital aortic disease. Initial imaging.

C. CTA Chest and Abdomen with IV Contrast

As a modality, CTA provides excellent spatial resolution, fast acquisition times, and the ability for 3-D reconstruction [6,16]. Another advantage of CTA is the ability to visualize cardiac structures and coronary arteries, as several congenital aortic processes are associated with cardiac abnormalities [19]. One study found that a prospectively triggered, dual-energy, high-pitch protocol CTA was more accurate than echocardiography in the diagnosis of coarctation [20]. Another series found CTA to be 100% accurate compared with operative findings in evaluating the diameter and length of aortic coarctation [18], whereas others have demonstrated CTA to compare favorably to both operative and catheter-based angiographic findings [11-14]. These findings make CTA a valuable, noninvasive imaging study for aortic characterization that can help to guide future interventions [19]. However, CTA does not provide direct hemodynamic information [10].

Variant 1: Congenital aortic disease. Initial imaging. D. FDG-PET/CT Skull Base to Mid-Thigh

There is no relevant literature for the use of PET using the tracer fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG)-PET/CT imaging in the evaluation of congenital aortic disease.

Variant 1: Congenital aortic disease. Initial imaging. E. MRA Chest and Abdomen

The relevant literature focuses on cardiac MRI, rather than MRA of the chest and abdomen, for evaluating congenital aortic disease. Cardiac MRI is becoming standard practice in evaluating patients with suspected congenital aortic pathology [21]. Even though it has lower spatial resolution than CT, MRA provides important physiologic information, including pressure gradients, extent of collateral flow, contractility of the myocardium, and evaluation of the valves [22]. Physiologic measurements are especially critical in evaluating coarctation where the smallest cross-sectional diameter of the aorta and flow deceleration in the descending aorta measured on velocity-encoded cine MRI are excellent predictors of a hemodynamically significant stenosis [15,17]. Newer 4-D sequences may improve the evaluation of vascular flow and hemodynamics, such as shear stress, pressure gradients, and turbulence [23-25]. Because of cardiac motion, 3-D noncontrast navigator MRA, steady-state 3-D contrast-enhanced MRA, or gated first-pass contrast-enhanced MRA is preferable to evaluate the aortic root [26,27].

The addition of IV contrast for MRA can be beneficial in evaluating congenital aortic diseases. For example, contrast-enhanced MRA has a higher sensitivity, specificity, and accuracy for detecting obstructive aortic anomalies when compared with either TTE or MRA without IV contrast [28]. Contrast-enhanced MRA may also improve visualization of the aorta when compared with fast spin-echo sequences [29].

Variant 1: Congenital aortic disease. Initial imaging. F. Radiography Chest

Chest radiography is a common imaging modality for individuals with suspected congenital aortic processes. Chest radiographs may be helpful in evaluating the contour, size, and location of the thoracic aorta and the great vessels, which, if abnormal, would prompt further investigation [10,21]. In aortic coarctation, a chest radiograph may reveal a characteristic "figure 3" sign or rib notching [30,31]. Chest radiographs can also be useful in excluding other congenital aortic diseases, such as obstructive arch disease and vascular rings [32,33]. However, given the availability of better imaging technologies and radiography's lack of specificity [21,34,35], a more definitive evaluation is typically required for an accurate diagnosis.

Variant 1: Congenital aortic disease. Initial imaging.

G. US Abdomen

Abdominal ultrasound (US) is a common imaging modality for evaluating the aorta. This section will review the relevant literature regarding abdominal US in the management of patients with suspected congenital processes of the aorta. Its role in detecting an abdominal aortic aneurysm is well described in the ACR Appropriateness Criteria topic on "Pulsatile Abdominal Mass, Suspected Abdominal Aortic Aneurysm" [5]. Middle aortic syndrome comprises a small percentage (0.5%–2.0%) of patients with coarctation of the aorta [36]. In this setting, abdominal US is able to detect the narrowing in the aorta [37]. Narrowing in middle aortic syndrome manifests similarly to other vascular stenoses, namely elevated peak systolic velocities, low resistive indexes, prolonged acceleration times, and parvus et tardus waveforms distal to the narrowing [37]. Furthermore, abdominal US may detect heterotaxy syndromes, such as situs invertus, which are variably related to congenital heart disease [38].

Variant 1: Congenital aortic disease. Initial imaging. H. US Echocardiography Transthoracic

TTE is a useful modality during the evaluation of congenital aortic abnormalities, given its association with cardiac abnormalities. For example, the reported incidence of bicuspid aortic valve in the setting of aortic coarctation ranges from 30% to 40% [19]. TTE is often the initial imaging modality when coarctation of the aorta is suspected [16], even though its utility can be reduced in the adult population because of its limited acoustic windows [21]. These limitations can be overcome somewhat through the use of the suprasternal view and Doppler imaging [20]. In addition to anatomic information, TTE can provide valuable physiologic parameters. For instance, Doppler can estimate peak velocities and pressure gradients across a stenosis [39,40]. TTE can also provide information regarding cardiac contractility, direction of flow, and valvular disorders [41]. Despite these advantages, TTE is limited in its ability to evaluate the aortic arch and proximal descending aorta [42,43].

Variant 1: Congenital aortic disease. Initial imaging. I. US Echocardiography Transesophageal

Transesophageal echocardiography (TEE) can provide views of the descending aorta, but physiologic information derived from these views can be inaccurate [21]. TEE is invasive and may not provide additional information than that gained from TTE [10].

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

A. Aortography chest and abdomen

Catheter-based aortography provides high spatial and temporal resolution, but because of its invasive nature and inability to detect changes to the vessel wall, it is considered inferior to cross-sectional imaging modalities. For example, a recent meta-analysis found that CT, MRI, and US were better than catheter-based angiography in detecting vascular lesions resulting from Takayasu arteritis [44]. Aortography is of most benefit when an intervention is planned.

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

B. CT Chest and Abdomen

There is no relevant literature available to examine the use of CT chest and abdomen with IV contrast alone in the management of inflammatory, infectious, neoplastic, or metabolic diseases. Please see the

"CTA chest and abdomen with IV contrast" section below for further discussion.

Overall, the addition of a contrast-enhanced CT scan after an unenhanced CT scan may be of benefit. For suspected vascular infection, one small series found rim enhancement to be the only finding associated with infection that required the administration of IV contrast [45]. Other findings in this series associated with infection did not need IV contrast administration for identification. Vascular neoplasms often do not enhance after the administration of IV contrast [46,47]. There is no relevant literature available to examine the use of CT chest and abdomen without and with IV contrast in the management of either inflammatory or metabolic aortic diseases. Please see the "CTA chest and abdomen with IV contrast" section below for further discussion.

For suspected vascular infection, CT imaging without IV contrast has some value in identifying signs associated with infection, including perivascular stranding, gas, wall thickening, aneurysmal dilatation, and involvement of adjacent bony structures [45,48]. Similarly, periaortic hemorrhage from ruptured aneurysm can be identified on CT without IV contrast [49]. Benign and malignant aortic tumors are exceedingly rare and often difficult to prospectively diagnose on imaging [50]. For suspected primary vascular neoplasms, an irregular soft-tissue density adjacent to the vessel wall may be seen [46,47]. CT imaging without IV contrast has little value in the diagnosis of inflammatory or metabolic aortic diseases.

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

C. CTA Chest and Abdomen with IV Contrast

CTA is routinely used in the diagnosis of inflammatory and metabolic processes of the aorta. For instance, CTA has been shown to be 95% sensitive and 100% specific in the diagnosis of Takayasu arteritis [51], outperforming catheter-based angiography [52]. The use of IV contrast also allows a more accurate assessment of the thickness of the vessel wall, an important marker in these diseases. Wall thickness as measured by CTA was 67% sensitive and 98% specific in identifying patients with clinical evidence of giant cell arteritis (GCA) [53]. Similarly, CTA has been shown to be a valuable imaging tool in the management of patients with Behcet disease [54,55]. CTA has been shown to be highly concordant with FDG-PET/CT (kappa: 0.64–1) in the detection of GCA [56,57]. CTA is considered an essential imaging tool in the diagnosis of vascular infection in which it can demonstrate the extent of vascular involvement, stenoses, aneurysms, wall thickening, and ulcers in addition to perivascular stranding, gas, and involvement of adjacent bony structures [48,58]. Regarding vascular neoplasms, CTA may be useful in evaluating the extent of disease and associated complications but does not add to the initial diagnosis [50].

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

D. FDG-PET/CT Skull Base to Mid-Thigh

FDG-PET/CT may be helpful for the assessment of active vascular inflammation, with sensitivity ranging from 60% to 92% and specificity from 88% to 100% [59,60]. For instance, FDG-PET/CT has been shown to be able to detect aortitis, which is seen in approximately half of patients with GCA [56,61]. Additionally, FDG-PET/CT may be of value in diagnosing GCA in patients who present with only vague clinical symptoms [62] and in patients with only extracranial disease [63]. As noted previously, FDG-PET/CT has been shown to be highly concordant with CTA findings (kappa: 0.64–1) in the detection of GCA [56,57]. In Takayasu arteritis, standardized uptake values are significantly higher in patients with active disease [64]. FDG-PET/CT can also aid in identifying the extent of fibrosis prior to repair in patients with idiopathic aortitis [65]. Because of the often-marked FDG avidity of aortic tumors, FDG-PET/CT can be helpful in

differentiating aortic tumor from bland thrombus, although infectious and inflammatory aortic conditions can similarly demonstrate strong FDG avidity [50]. There is no relevant literature available to examine the use of FDG-PET/CT in the initial diagnosis of vascular infections.

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

E. MRA Chest and Abdomen

Contrast-enhanced MRA techniques have evolved with the introduction of k-space and image-based acceleration techniques, higher field strengths, ultra-fast gradients, and the use of 3-D gradient-echo techniques. Double or triple inversion recovery and balanced steady-state free-precession pulse sequences are acquired before applying contrast-enhanced MRA. Additionally, delayed high-resolution T1-weighted images are acquired to assess aortic wall enhancement, especially in the cases of suspected inflammatory or infectious processes. Contrast-enhanced MRI can be particularly useful in the evaluation of inflammatory conditions as it can detect wall enhancement, which is a sign of active Takayasu arteritis [66]. Moreover, a recent meta-analysis reported that the addition of contrast-enhanced sequences improved the sensitivity of MRA in detecting Takayasu arteritis from 79% to 92% [44]. The same study showed that contrast-enhanced MRA outperformed catheter-based angiography. Similarly, contrast-enhanced MRI allows for improved detection of wall enhancement in both GCA [67-70] and Behcet disease [55,71]. For suspected neoplasms, contrast-enhanced MRI may be able to help differentiate between atheromatous plaque and tumor as well as help to delineate extravascular extension [72]. Regarding infection, the utility of contrast-enhanced MRA is similar to that of CTA in its ability to detect aneurysms, edema, perivascular stranding, gas, and disrupted calcifications [50].

MRA without IV contrast has some utility in the diagnosis of inflammatory vascular conditions. For example, one meta-analysis found MRA to be 79% sensitive and 97% specific in the diagnosis of Takayasu arteritis, outperforming catheter-based angiography [44]. MRA without IV contrast is able to identify extracranial involvement in GCA [73]. As discussed above, this modality is unable to provide an assessment of wall enhancement, which is an important marker in many inflammatory conditions. Similar to unenhanced CT, MRA without IV contrast is able to identify aneurysms, edema, perivascular stranding, gas, and disrupted calcifications that may be associated with aortic infections [50]. For suspected neoplasms, an irregular soft-tissue structure may be identified that may or may not enhance after IV contrast administration [46,47].

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

F. Radiography Chest

Chest radiography may be helpful in evaluating the contour, size, and location of the thoracic aorta and the great vessels, which, if abnormal, would prompt further investigation [10,21]. Radiography is not considered an adequate modality to evaluate for inflammatory, infectious, neoplastic, or metabolic aortic diseases.

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

G. US Abdomen

This section will review the relevant literature regarding US in the management of patients with suspected inflammatory, infectious, neoplastic, or metabolic processes of the aorta. Its role in detecting an abdominal aortic aneurysm is well described in the ACR Appropriateness Criteria [®] topic on "<u>Pulsatile Abdominal Mass</u>, <u>Suspected Abdominal Aortic Aneurysm</u>" [5].

Vascular duplex US can identify vascular wall thickening, which is an important marker in patients with generalized vascular inflammation [74], GCA [73,75], Behcet disease [76], and Takayasu arteritis [44]. Additionally, US is better than catheter-based angiography for detecting stenoses, occlusions, and aneurysms from Takayasu arteritis [44]. Nonetheless, it must be remembered that these evaluations primarily focused on nonaortic vessels. In fact, one study compared duplex US to FDG-PET/CT for the detection of extracranial large vessel vasculitis and found that US was only 26% sensitive for detecting aortic involvement. Other authors have also recognized the limitations of US for measuring wall thickness of the aorta [44,75]. For suspected neoplastic processes, US is of little value because it is not able to reliably differentiate between malignant and benign tissue [50]. There is no relevant literature available to examine the use of US in suspected aortic infection.

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

H. US Echocardiography Transthoracic

TTE can view the thoracic aorta (mainly the ascending aorta and, to some extent, the proximal descending aorta and arch) and the aortic valve (for presence and quantification of aortic regurgitation). There is no relevant literature supporting TTE as the initial imaging modality when evaluating infectious, inflammatory, metabolic, or neoplastic processes in the aorta.

Variant 2: Inflammatory or infectious or neoplastic or metabolic nontraumatic aortic disease. Initial imaging.

I. US Echocardiography Transesophageal

TEE may provide views of the descending aorta not appreciated on TTE, but there is no relevant literature supporting TEE as the initial imaging modality when evaluating infectious, inflammatory, metabolic, or neoplastic processes in the aorta.

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging.

This variant includes atherosclerotic disease and degenerative aneurysms involving the thoracic and abdominal aorta. Follow-up imaging following therapy and acute aortic syndromes are discussed in separate ACR Appropriateness Criteria documents.

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging. A. Aortography chest and abdomen

Aortography no longer has a significant role as the initial imaging modality for suspected degenerative or atherosclerotic disease of the aorta, as the availability and accuracy of noninvasive methods continues to increase. Catheter-based angiography remains a critical component of care when an intervention is planned.

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging. B. CT Chest and Abdomen

There is no relevant literature available by which to examine the use of CT chest and abdomen with IV contrast alone in the diagnosis of degenerative or atherosclerotic aortic disease. Please see the "CTA chest and abdomen with IV contrast" section below for further discussion.

There is no relevant literature available by which to examine the use of CT chest and abdomen without and with IV contrast outside of a dedicated CTA in the diagnosis of degenerative or atherosclerotic aortic disease. Please see the "CTA chest and abdomen with IV contrast" section below for further discussion.

CT chest and abdomen without IV contrast can help identify the size and extent of an aortic aneurysm [77]. Unenhanced CT has been shown to be more sensitive (82.6%–88.9%) than US (57.1%–70.4%) in identifying abdominal aortic aneurysms [78]. Both modalities were found to have equally high specificity in this regard (CT: 97.7%–98.4%; US: 99.2%–99.6%) [78]. Several large prospective studies have used CT chest and abdomen without IV contrast to quantify calcified atherosclerotic disease in the aorta [79-84]. However, the clinical utility of this approach as initial imaging is limited because the lack of IV contrast leads to an underestimation of noncalcified atherosclerotic plaque and does not provide an assessment of the aortic lumen [85].

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging. C. CTA Chest and Abdomen with IV Contrast

CTA with IV contrast is an important tool when evaluating the aorta for suspected degenerative and atherosclerotic changes as it provides information about the aortic lumen, the aortic wall, and surrounding aortic structures [86,87]. CT can also be used to detect other pathologies in the lungs, chest wall, and pleura, which can mimic the symptoms of aortic disease [88]. The addition of a venous phase to the CTA appears to increase its ability to identify both benign and malignant incidental pathology in nonvascular structures. [89]. Electrocardiograph (ECG)-gated aortic CTA decreases pulsation artifacts of the ascending aorta, which allows for a more accurate measurement of the ascending aortic diameter and potentially increases diagnostic confidence [90]. CTA provides information about the aortic root [91], aortic valve area and function, aortic wall elasticity, and morphology in general [92,93]. The CTA 3-D data set can be postprocessed and manipulated perpendicular to the flow lumen, allowing for accurate measurements for longitudinal evaluation of aortic growth and lumen diameters as well as planning for endovascular or surgical treatment [87,94,95]. Even though the maximum diameter of the aorta is the most consistent predictor of future rupture, other CTA findings, such as luminal contrast heterogeneity, intraluminal thrombus volume, aortic wall distensibility, and aneurysm geometry, help identify patients at risk for rupture [96-98]. Geometric models from CTA examinations can be used to create computational flow dynamics, which may also be useful in identifying patients with thoracic aortic aneurysms who are at risk for rupture [99].

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging. D. FDG-PET/CT Skull Base to Mid-Thigh

There is no relevant literature for the use of FDG-PET/CT imaging as an initial evaluation of degenerative or atherosclerotic aortic diseases.

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging. E. MRA Chest and Abdomen

Similar to MRA without IV contrast, MRA without and with IV contrast can be used to obtain hemodynamic information about aortic aneurysms from computational flow dynamics or 4-D flow MRI [99]. The addition of IV contrast improves visualization of the aorta and great vessels and decreases overall acquisition time [100-104]. However, there is no definitive evidence that the addition of IV contrast improves the overall accuracy of MRA for degenerative aortic diseases.

MRI without IV contrast using double-inversion recovery T1-weighted imaging and balanced steady-state free-precession MRA allows for imaging of the aorta, especially when acquisition is ECG-gated [105]. The accuracy of balanced steady-state free-precession MRA is close to 100% for detecting thoracic aortic aneurysm, dissection, intramural hematoma, and penetrating aortic ulcer when measured against the reference standard of MRA with IV contrast material [101,106]. It can be used to evaluate the entire

thoracic aorta and its branches [107]. MRA without IV contrast can also be used to create geometric models for computational flow dynamics [99]. Additionally, 4-D flow MRI sequences can be obtained [99]. These sequences are the acquisition of 3-D phase-contrast images in a time-resolved, ECG-gated manner with 3-D velocity encoding. Both computational flow dynamics and 4-D flow MRI can be used to examine hemodynamic information in thoracic aortic aneurysms, such as wall shear stress, flow patterns, and helical flow that may help to identify patients at risk for rupture [99].

For aortic atherosclerotic disease, both MRA without IV contrast and MRA without and with IV contrast can be used to collect physiologic information about aortic flow in order to provide an assessment of aortic wall stiffness and to generate 4-D flow MRI [42,99]. One disadvantage of MRI of the chest and abdomen without and with IV is that it may underestimate the thickness of atherosclerotic plaques compared with other modalities [108]. One advantage of MRI of the chest and abdomen is that it can be used to evaluate the composition of atherosclerotic plaques for lipids, fibrosis, calcifications, and intraplaque hemorrhage [95,109]. Because of improved visualization and decreased acquisition times, the use of IV contrast for MRA of the chest and abdomen is preferred, but sufficient information can also be obtained without the use of IV contrast [100-104,110].

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging. F. Radiography Chest

The chest radiograph is a helpful initial imaging evaluation for the diagnosis of degenerative aortic disease, especially if an acute complication is suspected. However, because of the lack of sensitivity in assessing the extent of disease [35], more definitive tests are required. Regarding atherosclerotic aortic disease, some studies have used lateral lumbar radiographs to both quantify and correlate atherosclerotic disease in the aorta to bone mineral density [111], patient diet [112], and more generalized atherosclerotic disease [113]. In clinical practice, however, the greatest utility of radiography would be to prompt additional imaging if an abnormality is identified [114].

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging. G. US Abdomen

The role of US is limited to evaluating the abdominal aorta and its branches and extracranial cerebral vasculature. Its role in detecting an abdominal aortic aneurysm is well described in the ACR Appropriateness Criteria [®] topic on "<u>Pulsatile Abdominal Mass, Suspected Abdominal Aortic Aneurysm</u>" [5].

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging. H. US Echocardiography Transthoracic

TTE is useful in assessing aneurysms involving the aortic root and ascending aorta. TTE is also an excellent tool for evaluating the abdominal aorta. For instance, a recent meta-analysis of studies examining the use of TTE for abdominal aortic aneurysm screening in adults found that TTE was able to visualize the aorta in 86% of patients [115]. However, TTE does not provide a full evaluation of the entirety of the aorta in many patients, which limits its utility as an initial imaging modality [95]. Yet, because the majority of thoracic aortic aneurysms are located within the proximal segments of the aorta, TTE may suffice for screening and serial measurements of aortic root diameters [116]. TTE can provide physiologic information. For example, one study found that patients with ascending aortic aneurysms had reduced elasticity and increased stiffness of the vessel wall [117]. Regarding the detection and characterization of aortic atherosclerotic disease, TTE is generally considered an unreliable imaging technique [42].

Variant 3: Degenerative or atherosclerotic aortic disease. Initial imaging.

I. US Echocardiography Transesophageal

There is no relevant literature for the use of TEE as an initial imaging modality in the evaluation of degenerative or atherosclerotic aortic diseases.

Summary of Highlights

- Variant 1: CTA chest and abdomen with IV contrast, MRA chest and abdomen without and with IV contrast, MRA chest and abdomen without IV contrast, radiography of the chest, or US echocardiography transthoracic resting is usually appropriate for the initial imaging of congenital aortic disease. With the exception of radiography of the chest, these procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). Chest radiography, although appropriate as an initial imaging modality, typically requires confirmation through a more definitive imaging examination because of a lack of specificity.
- **Variant 2:** CTA chest and abdomen with IV contrast, FDG-PET/CT skull base to mid-thigh, MRA chest and abdomen without and with IV contrast, or MRA chest and abdomen without IV contrast is usually appropriate for the initial imaging of inflammatory, infectious, neoplastic, or metabolic nontraumatic aortic disease. These procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- Variant 3: CTA chest and abdomen with IV contrast, MRA chest and abdomen without and with IV contrast, MRA chest and abdomen without IV contrast, radiography of the chest, or US of the abdomen is usually appropriate for the initial imaging of degenerative or atherosclerotic aortic disease. With the exception of radiography of the chest, these procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). Chest radiography, although appropriate as an initial imaging modality, typically requires confirmation by a more definitive imaging examination because of a lack of sensitivity.

Supporting Documents

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

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

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable riskbenefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be

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

Relative Radiation Level Information

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

Relative Radiation Level Designations

Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
0	0 mSv	0 mSv
જ	<0.1 mSv	<0.03 mSv
☆ ❖	0.1-1 mSv	0.03-0.3 mSv
❖ ❖ ❖	1-10 mSv	0.3-3 mSv
	10-30 mSv	3-10 mSv
	30-100 mSv	10-30 mSv

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

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Disclaimer

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.

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