American College of Radiology
ACR Appropriateness Criteria®

Clinical Condition: **Nontraumatic Aortic Disease**

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9		☢
CT chest with IV contrast	8		☢☢☢
CT chest and abdomen without IV contrast	8		☢☢☢☢
CT chest and abdomen without and with IV contrast	8		☢☢☢☢
CTA chest with IV contrast	8		☢☢☢
CTA chest and abdomen with IV contrast	8		☢☢☢☢
MRA chest without and with IV contrast	8		O
MRA chest and abdomen without and with IV contrast	8		O
US echocardiography transesophageal	7		O
CT chest without IV contrast	7		☢☢☢
CT chest without and with IV contrast	7		☢☢☢
CT chest and abdomen with IV contrast	7		☢☢☢☢
MRA chest without IV contrast	7		O
MRA chest and abdomen without IV contrast	7		O
US echocardiography transthoracic resting	6		O
Aortography chest and abdomen	6		☢☢☢☢
FDG-PET/CT chest and abdomen	5		☢☢☢☢
In-111 WBC scan	5		☢☢☢☢
US intravascular aorta	4		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			***Relative Radiation Level**

NONTRAUMATIC AORTIC DISEASE

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Summary of Literature Review

Introduction/Background

Nontraumatic aortic diseases include congenital, inflammatory, infective, metabolic, neoplastic, postoperative, and degenerative disorders that can affect the lumen or aortic wall or both. 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. Imaging studies are required to assess the anatomy and extent of morphological changes to the aortic lumen and aortic wall and, in some cases, functional changes to the aortic valve, branch vessel involvement, and perfusion of the end organs. Often, aortic disorders involve both the thoracic and abdominal aortic segments, thus requiring imaging of both regions. The clinical symptoms of aortic disease vary widely; 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 mainly to acute aortic syndromes, thoracic aortic aneurysm, atherosclerosis, postoperative aorta, and inflammatory or infective disease of the aorta. Readers are encouraged to review published ACR Appropriateness Criteria® for acute chest pain suspected of coronary syndrome [1], myocardial infarction [1], pulmonary embolism [2], aortic dissection [3], pulsatile abdominal mass [4], and abdominal aortic aneurysm for intervention planning and follow-up [5].

Chest Radiograph

A chest radiograph is a useful tool for the initial examination of suspected aortic disease unless the hemodynamic instability precludes its use. It can help rule out other plausible causes of clinical symptoms, and it provides information about the thoracic aorta as well as the heart, lungs, and ribs, which may be involved in aortic disease. The sensitivity of chest radiograph for detecting thoracic aortic disease varies between 12.4% and 81% [6-9], but sensitivity differs in whether it is read prospectively or retrospectively [10]. Its sensitivity also depends on the clinical question. The reported sensitivity of chest radiograph in acute aortic syndrome (dissection, intramural hematoma, penetrating aortic ulcer, or nondissecting aneurysm) is as high as 64% for detecting aortic disease [11], whereas its sensitivity for detecting saccular arch aneurysms is around 50% [9]. Chest radiographs are usually abnormal (sensitivity of 90%) in the presence of thoracic aortic dissection and a normal aorta, and mediastinum decreases the probability of dissection [12]. For obstructive arch disease and vascular rings, chest radiography is useful in diagnosing or ruling out aortic disease [13,14]. It is also useful in assessing endovascular stent grafts for their integrity (stent fractures, kinking) and displacement. Chest radiography suffers from poor interobserver agreement [8] and lack of sensitivity in assessing the extent of the disease [15], thus requiring more definitive tests for an accurate diagnosis.

Echocardiography

Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) are useful for assessing the heart (for anatomical abnormalities, such as congenital anomalies, intracardiac flow directions, and left ventricular function), pericardium (pericardial effusions and the hemodynamic effects of pericardial effusions), 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); however, TEE is an invasive procedure. For morphological

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assessment and anatomical mapping of the thoracic aorta, TEE is superior to TTE for diagnosing aortic dissection involving the ascending aorta; it has a sensitivity close to 100% (versus 50% for TTE) [16-18], which approaches that of computed tomography (CT) and magnetic resonance imaging (MRI) [19]. However, when TTE is combined with CT angiography (CTA) with contrast, the sensitivity and specificity increase to the level achieved with either TEE or MRI or a combination of TEE and MRI [20]. TEE is highly sensitive (90%) and specific (99%) for detecting aortic intramural hematoma [16]. Penetrating aortic ulcers affecting the ascending aorta can be easily seen with TEE; however, ulcers affecting the arch are poorly identified [21]. In addition, TEE has prognostic value in patients who have a suspected ascending aortic dissection [22]. During endovascular stent graft placement, TEE is superior to angiography in identifying multiple entries of aortic dissection, assessing stent apposition, and detecting endoleaks [23]. Following surgery for ascending aortic dissection, TEE is useful in assessing anastomotic complications, residual or new dissection [24], and aortic valve function; it is superior to contrast-enhanced MRI in evaluating periprosthetic valvular thickening [25]. Echocardiography is of limited value in assessing the supra-aortic vessels and abdominal aorta.

Intravascular Ultrasound

Intravascular ultrasound (US) suffers from its invasiveness, limited availability, and dependency on operator skills. Its role in the diagnosis and management of aortic disease is evolving, with reports mainly limited to aortic dissection, intramural hematoma, and penetrating aortic ulcer. It has special value during thoracic aortic endograft placement and is superior to angiography for detecting aortic dissection entry tears, identifying false lumen, involving the branch vessels [26], and assessing stent opposition and endoleak [23]. US is more sensitive than helical CT in detecting penetrating aortic ulcers [27]. It is also useful in detecting small perigraft pseudo-aneurysms that are often not seen on angiography following aortic repair; however, CT and MRI are equally sensitive [28]. Intravascular US is also used to measure the luminal diameter prior to stent graft placement, but its usefulness, when compared with CTA, remains questionable due to the inherent high coefficient of variation in measuring the aortic lumen [29].

Computed Tomography and Computed Tomography Angiography

CT provides information about the aortic lumen, the aortic wall, and surrounding aortic structures. CT without contrast is often sufficient for assessing the presence and extent of aortic aneurysm, ruptured aneurysm, intramural hematoma, and calcified atherosclerosis. It is also useful in assessing the integrity of a stent graft for fractures, kinking, collapse, and migration. CT is also used to detect other pathologies in the lungs, chest wall, and pleura, which can mimic the symptoms of aortic disease [30]. However, CTA with contrast material can be used to accurately delineate aortic lumen [29]; differentiate the thrombus from flowing blood; identify aortic dissection [31], penetrating aortic ulcer [32,33], and intramural blood pools during the evolution of an intramural hematoma [34]; assess branch vessels and end-organ perfusion; assess anatomic suitability for endograft treatment [35]; and evaluate postoperative aorta for anastomotic pseudoaneurysms, graft diameter [36], suture dehiscence [37], residual or new dissection [24,38,39], stent graft integrity, and persistent perfusion of the aneurysm sac following its exclusion by a stent graft, especially with a delayed CTA of 70–120 seconds [5,40,41]. The sensitivity and specificity of CTA is close to 100% for detecting aortic dissection and intramural hematoma [17-19,42,43] and 93% for detecting branch vessel involvement [18]. CTA provides prognostic information in patients with intramural hematoma because ascending aortic involvement, thickness of intramural hematoma, presence of ulcer-like projections, and aortic size predict complications [44,45]. CTA can be used to differentiate between atherosclerotic aneurysms and mycotic aneurysms [46,47]. In cases of a suspected penetrating aortic ulcer, CTA can be used to differentiate a penetrating ulcer from ulcerated plaques [32] and dissection [33] as well as to identify an associated intramural hematoma, pseudoaneurysm, and rupture [48]. Thus, in patients who present with acute aortic syndrome, CTA can provide an accurate diagnosis and help predict the disease progression. However, CT and CTA are limited in evaluating aortic valve function, hemodynamic effects of pericardial effusions, and left ventricular function. Electrocardiograph (ECG)-gated aortic CTA decreases pulsation artifacts of the ascending aorta, allows for a more accurate measurement of the ascending aortic diameter, and potentially increases diagnostic confidence in diagnosing ascending aortic pathology, such as aortic dissection [49]). It may provide information about the aortic root [50], morphology, aortic valve area and function, and aortic wall elasticity [51]. ECG-gated aortic CTA could be obtained at a low radiation dose (similar to that of nongated CTA) [52].

Magnetic Resonance Imaging and Magnetic Resonance Angiography

Noncontrast MRI of the aorta using double-inversion recovery T1-weighted imaging and balanced steady-state free precession (bSSFP), allows for imaging of the aorta, especially when acquisition is ECG-gated [53]. The accuracy of bSSFP is close to 100% for detecting thoracic aortic aneurysm, dissection, intramural hematoma, and penetrating aortic ulcer when measured against the reference standard of magnetic resonance angiography (MRA) with contrast material [54,55]. MRI is superior to CT in differentiating an acute intramural hematoma from atherosclerotic plaque and chronic intraluminal thrombus [56]. MRI can also be used to assess the chronicity of an intramural hematoma and can accurately diagnose aortic dissection. Its sensitivity is superior to TTE but comparable to that achieved with CTA and TEE [19]. It accurately measures aortic or graft diameter [36]; evaluates aortic root [25], periaortic hematoma, and aortic regurgitation [17,36]; and detects aortic thrombus and entry tear of a dissection [17]. It also is used to evaluate the entire thoracic aorta and its branches [57]. The detection of penetrating ulcers can be limited when using noncontrast MRI; however, its use in detecting an associated intramural hematoma and its extent can be well demonstrated [56].

Contrast-enhanced MRA techniques have evolved with the introduction of k-space manipulation and short acquisition times with 3-D gradient echo techniques. Double inversion recovery and bSSFP pulse sequences are acquired before applying contrast-enhanced MRA. Additionally, delayed high-resolution, T1-weighted images are acquired to assess the aortic wall enhancement, especially in cases of suspected inflammatory aortic aneurysm, aortitis, or mycotic aneurysm. T2-weighted imaging of the aorta is helpful in assessing vessel wall edema in cases of suspected aortitis. Contrast-enhanced MRA is highly accurate in diagnosing aortic dissection [19], localizing entry tears, detecting slow flow in the false lumen, and assessing branch vessel involvement, thus providing all the information required for therapy planning [58]. Combined with noncontrast techniques, contrast-enhanced MRA allows for the accurate detection and localization of penetrating aortic ulcers, intramural hematoma, and ulcerated plaques [59]. Following endovascular repair with nitinol-based thoracic endografts, MRA can be used to detect endoleaks, and its sensitivity surpasses that of CTA [60]. MRA clearly shows thickening and enhancement of the aortic wall and is useful in assessing disease activity in patients who have known vasculitis and suspected inflammatory or mycotic aneurysms [46]. Following surgical repair, MRA can detect pseudoaneurysms at the anastomotic sites, graft diameter, dissection recurrence, and aortic root morphology; it can also be used to evaluate the entire thoracoabdominal aorta and supra-aortic branches [25]. Similarly, contrast-enhanced MRA has a higher sensitivity, specificity, and accuracy for detecting obstructive aortic arch anomalies, when compared with TTE and MRI without contrast [61].

Aortography

The role of aortography in diagnosing aortic disease is decreasing as the availability and higher sensitivities of other noninvasive modalities, such as TTE, CTA, and MRA increases. As expected, aortography remains a part of the interventional procedures for treating aortic disease. Aortography delineates the lumen but provides no direct information about the aortic wall and mural adherent pathology. It has limitations in detecting entry tears, differentiating an atherosclerotic ulcer from a penetrating aortic ulcer, and assessing the extent of mural thrombus [56]. Early reports suggested a similar sensitivity between aortography and contrast-enhanced CT for detecting an aortic dissection, but they noted a higher specificity for aortography [7]. Aortography is useful for diagnosing adult congenital vascular anomalies; however, CTA and MRA provide similar information. Aortography is also useful for assessing postoperative aorta, after surgical and endovascular therapies; however, CT/MRA superseded its use due to their noninvasiveness and higher sensitivity.

Ultrasonography

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 "Pulsatile Abdominal Mass, Suspected Abdominal Aortic Aneurysm" [4]. US tends to underestimate the size of abdominal aortic aneurysm by 4 mm [4] and may not accurately delineate the margins of the aneurysm and involvement of the visceral branches [4].

Fluorodeoxyglucose-Positron Emission Tomography

The role of positron emission tomography with fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG-PET) in evaluating aortic disease is evolving. Recent reports have suggested that a higher FDG uptake in patients who have acute aortic syndrome can predict disease progression and future complications [62]. Similarly, its role in assessing disease activity in patients who have large-vessel vasculitis is being investigated. Its sensitivity appears to be low

(around 60%) in patients with a low C-reactive protein/erythrocyte sedimentation rate [63]. The additional prognostic value of FDG-PET in these patients is not known, but localization of active disease sites is possible with FDG-PET.

Indium-Labeled Leukocyte Scintigraphy

Indium-labeled leukocyte scintigraphy is useful for assessing infected aneurysms, graft infection, and inflammatory aneurysms. Its sensitivity approaches 90% for detecting mycotic aneurysms [47]. For abdominal aortic graft infection, both contrast-enhanced MRI and indium-labeled leukocyte scintigraphy provide similar negative predictive values, but MRI has a higher positive predictive value and may be more valuable modality [64].

Summary

- The literature supports the continued use of chest radiography for the initial evaluation of suspected thoracic aortic disease.
- In patients with acute aortic syndromes, CTA without and with contrast provides the most clinically relevant information. CTA is also useful for planning endovascular therapy and postoperative aorta follow-up.
- MRI and MRA provide similar information to that of CTA and are best suited for use with stable patients.

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.

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".		

Supporting Documents

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

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