

**American College of Radiology  
ACR Appropriateness Criteria®  
Noncerebral Vasculitis**

**Variant 1: Suspected large-vessel vasculitis (LVV). Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRA chest abdomen pelvis with IV contrast	Usually Appropriate	○
MRA chest abdomen pelvis without and with IV contrast	Usually Appropriate	○
CT chest abdomen pelvis with IV contrast	Usually Appropriate	⊕⊕⊕⊕
CT chest abdomen pelvis without and with IV contrast	Usually Appropriate	⊕⊕⊕⊕
FDG-PET/CT whole body	Usually Appropriate	⊕⊕⊕⊕
CTA chest abdomen pelvis with IV contrast	Usually Appropriate	⊕⊕⊕⊕⊕
Arteriography chest abdomen pelvis	May Be Appropriate	
US duplex Doppler upper extremity	May Be Appropriate	○
MRA chest abdomen pelvis without IV contrast	May Be Appropriate	○
MRA neck with IV contrast	May Be Appropriate (Disagreement)	○
MRA neck without and with IV contrast	May Be Appropriate (Disagreement)	○
MRA neck without IV contrast	May Be Appropriate	○
MRI chest abdomen pelvis without and with IV contrast	May Be Appropriate	○
CTA coronary arteries with IV contrast	May Be Appropriate	⊕⊕⊕
US duplex Doppler aorta abdomen	Usually Not Appropriate	○
US duplex Doppler chest abdomen pelvis	Usually Not Appropriate	○
US duplex Doppler iliofemoral arteries	Usually Not Appropriate	○
US duplex Doppler lower extremity	Usually Not Appropriate	○
MRA coronary arteries without and with IV contrast	Usually Not Appropriate	○
MRA coronary arteries without IV contrast	Usually Not Appropriate	○
MRI chest abdomen pelvis without IV contrast	Usually Not Appropriate	○
MRI heart function and morphology without and with IV contrast	Usually Not Appropriate	○
MRI heart function and morphology without IV contrast	Usually Not Appropriate	○
CT chest abdomen pelvis without IV contrast	Usually Not Appropriate	⊕⊕⊕⊕

**Variant 2:****Suspected medium-vessel vasculitis (MVV). Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
Arteriography chest abdomen pelvis	Usually Appropriate	☼☼☼☼
CTA chest abdomen pelvis with IV contrast	Usually Appropriate	☼☼☼☼☼
MRA chest abdomen pelvis with IV contrast	May Be Appropriate	○
MRA chest abdomen pelvis without and with IV contrast	May Be Appropriate (Disagreement)	○
MRA chest abdomen pelvis without IV contrast	May Be Appropriate (Disagreement)	○
MRI chest abdomen pelvis without and with IV contrast	May Be Appropriate	○
MRI heart function and morphology without and with IV contrast	May Be Appropriate	○
CTA coronary arteries with IV contrast	May Be Appropriate	☼☼☼
CT chest abdomen pelvis with IV contrast	May Be Appropriate	☼☼☼☼
CT chest abdomen pelvis without and with IV contrast	May Be Appropriate	☼☼☼☼
US duplex Doppler aorta abdomen	Usually Not Appropriate	○
US duplex Doppler chest abdomen pelvis	Usually Not Appropriate	○
US duplex Doppler iliofemoral arteries	Usually Not Appropriate	○
US duplex Doppler lower extremity	Usually Not Appropriate	○
US duplex Doppler upper extremity	Usually Not Appropriate	○
MRA coronary arteries without and with IV contrast	Usually Not Appropriate	○
MRA coronary arteries without IV contrast	Usually Not Appropriate	○
MRA neck with IV contrast	Usually Not Appropriate	○
MRA neck without and with IV contrast	Usually Not Appropriate	○
MRA neck without IV contrast	Usually Not Appropriate	○
MRI chest abdomen pelvis without IV contrast	Usually Not Appropriate	○
MRI heart function and morphology without IV contrast	Usually Not Appropriate	○
CT chest abdomen pelvis without IV contrast	Usually Not Appropriate	☼☼☼☼
FDG-PET/CT whole body	Usually Not Appropriate	☼☼☼☼

## NONCEREBRAL VASCULITIS

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### **Summary of Literature Review**

#### **Introduction/Background**

Idiopathic vasculitis is a noninfectious inflammation of the vessels that can lead to serious health consequences. It can be a primary inflammatory process or a secondary process because of an underlying disease. Historically, it has been categorized based on whether inflammation is restricted to blood vessels of particular size as large-vessel vasculitis (LVV), medium-vessel vasculitis (MVV), small-vessel vasculitis, and variable-vessel vasculitis. The large vessels are the aorta and its main branches, the medium vessels are the main visceral arteries and initial branches, and the small vessels are the intraparenchymal vessels and analog veins. There is an overlap between these vasculitis types; for example, LVV predominantly involves large vessels; however, it can also affect medium and small vessels. In variable-vessel vasculitis, there is no predominance of vessel size involvement. Tissue biopsy of the large or medium vessels is often not feasible; therefore, imaging plays a crucial role in diagnosing idiopathic vasculitides. Considering the limitations of spatial resolution across all available modalities for small-vessel vasculitis, this manuscript focused on vasculitis mainly involving the large and medium vessels.

LVV includes 2 subtypes: giant-cell arteritis (GCA) and Takayasu arteritis (TAK), of which GCA is more common. GCA is an idiopathic, inflammatory, granulomatous vasculitis involving predominantly the large arteries in older patients (>50 years of age). GCA affects the supra-aortic vessels, especially the extracranial branches of the carotid artery, such as the superficial temporal artery (referred to as cranial-GCA [c-GCA]). Classically, a diagnosis of GCA requires temporal artery ultrasound (US) or temporal artery biopsy. However, a growing body of literature has demonstrated the involvement of the extracranial large arteries, particularly the aorta and its main branches, which is known as large-vessel GCA (LV-GCA). The American College of Rheumatology classification criteria underperforms in classifying patients with LV-GCA. A retrospective study by Muratore et al [1] demonstrated that American College of Rheumatology criteria are inadequate to classify patients with LV-GCA. Therefore, patients with suspected GCA require supplemental imaging studies in addition to temporal artery US or biopsy [2]. In this regard, the trial of tocilizumab for GCA, which is a large randomized controlled trial in GCA, included patients with imaging-confirmed LV-GCA who did not meet the American College of Rheumatology criteria [3]. Interestingly, patients with LV-GCA compared to other patients with GCA, present less frequently with jaw claudication or ischemic symptoms and have a higher incidence of relapse, have greater cumulative glucocorticoid exposure, and are more frequently treated with steroid-sparing agents, such as the interleukin-6-receptor blocker tocilizumab [1]. Polymyalgia rheumatica (PMR) is a disorder in the same disease spectrum as GCA and can be found in association with GCA or as an isolated phenomenon [4].

TAK is also an idiopathic, inflammatory, granulomatous vasculitis mostly reported in young ( $\leq 40$  years of age) women with preferential involvement of the aorta and main branches [5]. It can also involve the pulmonary and coronary arteries [5]. Although 20% of patients have a self-limited disease, most patients demonstrate a relapse. Diagnosing TAK can be challenging and delayed given the clinical heterogeneity of initial presentation, ranging

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from asymptomatic presentations to nonspecific constitutional symptoms and major ischemic events. Although the histopathology of TAK shares similarities with that of GCA, biopsy material from the large arteries is rarely obtained in TAK. The diagnosis of TAK typically requires a combination of physical examination, laboratory findings, and imaging findings.

Variable-vessel vasculitis subtypes such as Cogan or Behcet disease can involve the large vessels, particularly the aorta. Although the literature is limited on imaging features of Cogan or Behcet vasculitis, case reports and series have demonstrated similarities with LVV. Because of the limitation of the peer-reviewed literature, the focus of this manuscript will be on LVV.

MVV predominantly involves medium-sized arteries, although arteries of any size can be involved. Polyarteritis nodosa (PAN) and Kawasaki disease are the 2 types of MVV [6]. PAN affects medium and small visceral vessels (particularly renal arteries), and there is an association with hepatitis B virus. Kawasaki disease is a self-limiting acute necrotizing vasculitis that affects medium and small vessels and is most prevalent in Asian populations. Kawasaki disease commonly affects the coronary arteries in 15% to 20% of patients [7].

## **Special Imaging Considerations**

### **CT and CTA**

CT or CT angiography (CTA) are cross-sectional imaging modalities with an excellent spatial resolution and faster scan time. Although assessment of the vessel wall is possible with contrast-enhanced CT, the proper modality is CTA. CT without intravenous (IV) contrast material is limited for vascular assessment. However, CTA can be acquired as a biphasic study, including noncontrast and arterial phase, or as a triphasic study with the addition of a delayed or venous phase for proper vessel lumen and wall assessment. In addition, electrocardiogram (ECG)-gated CTA exhibits motionless aortic root and ascending aorta [8]. In most cases, CTA of the chest, abdomen, and pelvis ensures coverage of the entire vasculature in the abdomen. In cases of coronary artery involvement, dedicated coronary CTA displays potential vessel involvement.

### **FDG-PET/CT Whole Body**

Inflammatory cells in the inflamed vessel wall can accumulate fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG); thus, FDG-PET/CT can be used in the diagnosis of vasculitis [9]. Similar to other indications, patient preparation includes 6 hours of fasting before administering the FDG tracer injection and limiting strenuous exercise before the study. Although FDG-PET/CT can be acquired as a standard 60- to 90-minute delay after the injection for vasculitis, a few studies have suggested that a 120- to 180-minute delay could lead to higher diagnostic accuracy [10,11]. In 1 study, the preferred scan time for optimal vessel wall inflammation was 2.5 hours with a target glucose level of <7.0 mmol/L (<126 mg/dL) [12]. In patients with coronary artery involvement/suspected coronary artery vasculitis, myocardial suppression should be achieved via diet, and ECG-gated images should be obtained [13]. Recently, there has been emerging literature describing FDG uptake in the head and neck arteries, particularly in the temporal and maxillary arteries, which could aid the simultaneous diagnosis of c-GCA and LV-GCA [14,15].

### **MRI and MRA**

In LVV, body MRI or MR angiography (MRA), including the chest, abdomen, and pelvis, can be performed to assess vasculitic changes [16]. MRI and MRA sequences are the following: T2-weighted fast spin-echo sequence or inversion recovery sequences, such as short-tau inversion recovery to display edema in the vessel wall; ECG-gated rapid 3-D T1-weighted spoiled gradient-echo pulse sequences can be used for MRA with gadolinium-based agents and maximum-intensity projection reformats; and postcontrast T1-weighted imaging with fat suppression (preferably using the Dixon technique) can be acquired throughout the body. Recently, new sequences have been introduced for LVV diagnosis, such as navigated T1-weighted 3-D black-blood fast turbo spin-echo sequence, which can acquire pre- and postcontrast isotropic 3-D images [17,18]. Last, the ECG-triggered balanced steady-state free-precession sequence provides additional information regarding the aortic root. Gadolinium injection through the veins can create susceptibility artifact along the axillary and subclavian arteries; therefore, maximum-intensity projection images should be examined carefully, and delayed images should be correlated [19].

There is a growing body of literature on high-resolution cranial MRI of the superficial cranial arteries for the diagnosis of c-GCA [20,21]. A 4-point ranking scale is used to classify the affected vessels based on the wall thickness and mural contrast enhancement [20,21]. Although this can be acquired practically during body MRI/MRA studies, discussion of this modality is not in the scope of this manuscript.

## **US and Color Doppler US**

US or color duplex US is a noninvasive imaging modality that has been used in the diagnosis of vasculitis. Particularly, in patients with suspected c-GCA, US/doppler US is recommended as the primary imaging modality [22]. Upper- and lower-extremity US can be added to increase diagnostic yield [23]. However, evaluation of the thoracic and abdominal vasculature is limited with US. Most of the literature on US is focused on its use in the temporal artery and, to a lesser extent, the extremity arteries. There is limited information regarding the use of chest, abdomen, and pelvis US or the diagnosis of LVV.

### **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 (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

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

### **Discussion of Procedures by Variant**

#### **Variant 1: Suspected large-vessel vasculitis (LVV). Initial imaging.**

##### **Arteriography Chest, Abdomen, and Pelvis**

Arteriography or catheter-based arteriography is an invasive imaging modality that was considered the reference standard for LVV diagnosis, particularly for TAK [22]. Although it is an ideal test for assessing luminal narrowing or stenosis with excellent spatial resolution, it fails to demonstrate mural thickening along the vessel. Therefore, assessment of inflammation along the vessel during the early phase of the disease cannot be evaluated. Considering the method's lack of wall assessment, invasiveness, and associated procedural risks, other noninvasive modalities have replaced arteriography, except in specific clinical scenarios (eg, when concomitant measurement of central artery pressure is desirable).

##### **CTA Chest, Abdomen, and Pelvis**

CTA can demonstrate vessel wall abnormalities in LVV, including circumferential mural thickening (>2–3 mm of the vessel without an adjacent zone of atheroma), associated wall enhancement, and perivascular fat stranding [24–26]. Delayed/venous phase image acquisition can emphasize mural enhancement [25,27,28]. In 1 study, significant aortic wall contrast enhancement is defined as an increase >20 HU in attenuation [25]. In 1 meta-analysis, active vasculitis on CTA was defined as vessel wall thickening with enhancement in the outer portion of the vessel wall and a low attenuation ring in the inner aspect of the vessel wall on delayed phase images [29]. Although a low attenuation ring on delayed images and wall enhancement were associated with elevated inflammatory markers, and thus active disease, wall thickening alone was not associated with elevated inflammatory markers [29]. CTA can also depict complications associated with vasculitis, such as luminal narrowing/stenosis or aneurysmal dilation [30,31]. With the multiphasic technique—noncontrast and arterial phases—differentiation of intramural hematoma from vasculitis is possible [27].

CTA or CT with IV contrast can be combined with FDG-PET as a hybrid model for the early diagnosis of LVV [32–34]. In 1 study, the authors compared CTA with FDG-PET/CT and found that the sensitivity and specificity of CTA for diagnosing GCA were 73% and 78%, respectively, and that the negative predictive value (NPV) and positive predictive value (PPV) were 65% and 85%, respectively [35]. Other studies compared the diagnostic performance of CTA to FDG-PET/CT, and both procedures were similarly able to detect large-vessel involvement in GCA [34,36,37]. In the study by de Boysson et al [36], CTA demonstrated high sensitivity of 95% and specificity of 100% when comparing the diagnosis per patient. However, when the diagnosis was compared per segments of the aorta and branches, FDG-PET/CT was more sensitive for the detection of inflammation, and CTA had lower sensitivity (61%) but still high specificity (98%). Two relatively small studies comparing contrast-enhanced CT and FDG-PET demonstrated comparable results, with a slightly superior diagnostic accuracy of FDG-PET [34,38]. In addition, FDG-PET demonstrated a strong correlation with the inflammatory markers [38]. For TAK diagnosis (not necessarily active disease), sensitivity of 95% and specificity of 100% were demonstrated to be higher with CTA [39,40].

A prospective study showed that almost 49% of patients had persistent wall thickening despite demonstrating a complete clinical response to treatment. Therefore, the role of CTA in disease monitoring and response to treatment of LVV remains unclear [39].

### **CT Chest, Abdomen, and Pelvis**

Although the majority of the literature is based on CTA, there are a few studies focused on contrast-enhanced CT. Recently, 2 studies by Muto et al and Vaidyanathan et al evaluated the role of contrast-enhanced CT and FDG-PET/CT in the diagnosis of LVV [32,34]. Muto et al [32] demonstrated that contrast-enhanced CT shows significantly more thickening of the aortic wall when compared with the control group of patients without vasculitis. In the Vaidyanathan et al [34] study, the authors found that metabolic activity of the aortic wall on FDG-PET/CT has excellent accuracy, with area under the curve [95% confidence interval] for standardized uptake value (SUV)<sub>max</sub> for liver being 0.97 [0.92–1.0], whereas aortic wall thickening on contrast-enhanced CT has good accuracy (0.82 [0.67–0.99]) in the diagnosis of LVV.

### **CTA Coronary Arteries**

Coronary CTA is an ideal imaging modality for detecting coronary involvement in LVV, particularly in patients with TAK [41]. Kang et al [41] reported that 53.2% of patients with TAK had coronary arterial lesions on coronary CTA, regardless of disease activity or symptoms. Of these patients, 28% had coronary ostial stenosis, 36.9% had nonostial coronary arterial stenosis, and 8.1% had coronary artery aneurysm.

### **FDG-PET/CT Whole Body**

Evidence supporting the role of FDG-PET/CT in the diagnosis of LVV has grown substantially in the past decade. A circumferential, segmental intense FDG uptake along the large vessels is considered diagnostic for LVV. The intensity of FDG uptake has been visually graded according to liver FDG uptake—as a qualitative method—with a high reproducibility and interrater agreement (grade 0 = no vascular uptake; grade 1 = vascular uptake < liver uptake; grade 2 = vascular uptake equal to liver uptake; grade 3 = vascular uptake > liver uptake) [33,42–44]. Grade 2 is considered possible LVV, whereas grade 3 is consistent with vasculitis [33]. Considering the segmental involvement, an alternative qualitative assessment was proposed: the Total Vascular Score, in which the visual grading (grade 0–3) of 7 artery segments (thoracic aorta, abdominal aorta, subclavian, axillary, carotid, iliac, and femoral arteries) is incorporated and ranges from 0 to 21 [33,45]. Semiquantitative measurements can be obtained from the vessel via the target-to-background ratio, mostly for research purposes [33,42]. A slightly different validated PET Vascular Activity Score was introduced, where the visual grading system (grade 0–3) added of each 9 arterial systems (ascending, arch, descending, abdominal aorta, right/left carotid arteries, innominate artery, right/left subclavian arteries), which ranges from 0 to 27 [46]. The target-to-background ratio is equal to the average SUV<sub>max</sub> of the artery divided by the average SUV<sub>mean</sub> of a tissue region/background reference tissue [33]. As a background reference tissue, blood pool, liver, or spleen can be used. However, there is contradicting research regarding these background reference tissues; for example, a study by Besson et al [44] demonstrated that the aortic-to-blood pool ratio outperforms the liver and lung ratio. A study by Ahlman et al [47] demonstrated that the use of blood pool to compute a target-to-background ratio might mask the artery wall signal reflective of inflammation, therefore the liver or spleen should be considered as a background reference, but further investigation is required. Although the use of liver was emphasized by another study, FDG uptake in the liver could be highly variable when there is use of steroids or the presence of hepatosteatosis [42]. Overall, qualitative methods have been shown to be more specific than semiquantitative methods but with lower sensitivity [45,48].

The sensitivity and specificity of the FDG-PET/CT to diagnose LVV is considerably high, ranging from 75% to 90% and from 81% to 98%, respectively, when the American College of Rheumatology criteria are used as a reference standard [42,46,49,50]. As mentioned above, the American College of Rheumatology criteria depend on temporal artery biopsy to detect c-GCA. However, in an extensive study, Luqmani et al [51] demonstrated that temporal artery biopsy has a false-negative rate as high as 61%. Therefore, the specificity of FDG-PET/CT in some studies was likely artificially lowered, considering temporal artery biopsy was falsely negative in extracranial GCA [52].

In their study, Lariviere et al [35] demonstrated that both FDG-PET/CT and CTA have a strong diagnostic yield to diagnose LVV; however, FDG-PET/CT appears to have higher PPV (100%) than CTA (84.6%). Similarly, another study found comparable sensitivity and specificity for FDG-PET/CT and CTA for the diagnosis of LVV [36]. However, per segments of the aorta and branches, FDG-PET/CT was significantly more sensitive than CTA (98% versus 61%) [36,37]. In a small study, the FDG-PET/CT score (visual and quantitative) was noted to be slightly superior to contrast-enhanced CT, with a strong correlation between the inflammatory markers [38]. Similarly,

another study comparing contrast-enhanced CT and FDG-PET revealed that FDG-PET/CT demonstrated excellent accuracy, whereas contrast-enhanced CT mural thickening exhibited good accuracy for the diagnosis of LVV [34].

A large prospective study comparing FDG-PET/CT and MRA demonstrated that FDG-PET/CT provides more information about disease activity, although MRA reveals anatomical changes and vascular damage [53]. Einspieler et al [54] investigated the performance of FDG-PET/MRI in patients with LVV. They assessed the individual value of FDG-PET and MRI, as well as compared FDG-PET/CT with FDG-PET/MRI. The authors concluded that FDG-PET/MRI and FDG-PET/CT produce consistent results and are highly comparable modalities. Similar to earlier studies, FDG-PET detected more abnormal vascular segments than MRA.

In addition, disease activity assessment by PET was associated with the clinical assessment. It has been shown that FDG-PET/CT also has prognostic value in patients with LVV [55,56]. In a multicenter study of 130 patients, inflammation in the aorta on FDG-PET/CT studies was reported to be a risk factor for aortic complications, particularly aneurysmal dilation or dissection [55]. FDG-PET/CT also may predict the long-term clinical outcome of patients with LVV. Two studies demonstrated that increased intensity of FDG uptake and extensive involvement of the vessels appeared to predict a less favorable response to treatment and more likely relapse during the follow-up, although the number of patients was small [46,57].

Most of the aforementioned studies were conducted on GCA, and the sensitivity and specificity for TAK have been reported to be slightly lower. In their meta-analysis of 546 patients, Soussan et al [49] reported a pooled sensitivity and specificity of 90% and 98%, respectively, for GCA diagnosis; however, for TAK, these values were 87% and 73%, respectively. This could be because of the vague inflammatory phase of TAK and the fact that the patients presented during the chronic and “pulseless phase,” therefore imaging may have missed the inflammatory phase. A study conducted in only 17 patients with TAK to assess disease activity demonstrated a sensitivity and specificity of 83% and 90% and a PPV and NPV of 77% and 93%, respectively [58]. FDG-PET/CT was demonstrated to be a reliable imaging modality for the detection of vascular wall inflammation in patients with TAK and was correlated with increased inflammatory markers (particularly C-reactive protein), but otherwise clinically silent disease [59-61]. It has been reported that aortic segments demonstrate the highest SUV<sub>max</sub> values in both GCA and TAK groups, but SUV<sub>max</sub> values were significantly higher in the GCA group [62]. Per the European League Against Rheumatism (EULAR) Recommendations Working Group, FDG-PET/CT is recommended for use as an alternative imaging modality, particularly to MRI/MRA, in patients with TAK [22]. However, a study by Incerti et al [63], in which the authors used both FDG-PET/CT and MRI in 30 patients with TAK, demonstrated that FDG-PET/CT provides information regarding the local inflammation independent of systemic inflammation, although arterial grafts were a potential confounder. Typically, FDG uptake at the site of the grafts in patients with vasculitis does not reflect clinically relevant information regarding disease activity [64].

As mentioned earlier, PMR can be isolated or can occur in tandem with GCA and is often diagnosed based on symptoms. However, it is known that FDG-PET/CT can also demonstrate PMR as FDG uptake in the periarticular and extra-articular structures [65]. Interestingly, a recent study has reported that many patients without cranial symptoms (ie, isolated PMR) often demonstrated LVV on FDG-PET/CT [66]. In addition to PMR, FDG-PET/CT can detect other extravascular findings (such as malignancy or infection) in patients who present with constitutional symptoms similar to vasculitis [13].

In a recent prospective cohort study of patients with suspected LVV, particularly GCA, patients underwent head, neck, and chest FDG-PET/CT. The sensitivity and specificity for the diagnosis of superficial cranial artery vasculitis were reported to be 92% and 91%, respectively, when the temporal artery biopsy was used as a reference test [14]. A similar study was performed by Nielsen et al [15], who demonstrated FDG uptake in the head and neck vessels with a high sensitivity and specificity and concluded that temporal artery biopsy could be omitted. There is also a prognostic value of head and neck FDG-PET/CT, particularly in the vertebral arteries, associated with the risk of ischemic complications [67]. A study by Michailidou et al [68] demonstrated that a pattern of FDG uptake along the head and neck vasculature can help differentiate TAK from GCA involvement without any angiographic abnormalities on MRA images. Patients with TAK present predominantly with carotid artery involvement, whereas more involvement is noted in the posterior cervical circulation (vertebral arteries) in patients with GCA. However, the authors assessed only for arterial damage on MRA, such as aneurysm, stenosis, and occlusion.

The main concern with FDG-PET/CT is that its sensitivity is affected by immunosuppression [42,69,70]. In addition, patients receiving glucocorticoid treatment demonstrate an increase in FDG uptake in the liver, which can lower the diagnostic accuracy when the vessel wall and liver ratio are used [42]. A study showed lower sensitivity

(71%) and specificity (64%) for GCA diagnosis with FDG-PET/CT in patients taking glucocorticoid treatment (for an average of 12 days) [70]. Two studies demonstrated a diagnostic window of the first 3 days after glucocorticoid treatment initiation to perform FDG-PET/CT [71,72]. Last, a problem can be encountered in head and neck FDG-PET/CT in patients with suspected vasculitis, a spill from the physiologic cerebral brain FDG uptake, and vessel juxtaposition to the skin–air interface.

### **MRA Chest, Abdomen, and Pelvis**

Although MRA is considered an ideal imaging modality for vessel wall evaluation with its high tissue resolution ability, the literature on LVV diagnosis is limited.

In a recent study involving 75 patients, when the authors used the temporal artery biopsy as the reference test, the sensitivity and specificity of MRA to diagnose LVV was 79% and 96%, respectively [73]. In the same study, PPV and NPV were 92% and 88%, respectively [73]. However, in a meta-analysis to diagnose TAK only, the pooled sensitivities and specificities were as high as 92% [29]. In addition, another study found that the use of whole-body contrast-enhanced MRA in patients with TAK was useful to diagnose active versus inactive TAK [74]. Similar to CTA, wall thickening and enhancement of the vessel wall on MRA were considered good markers of disease activity [29]. Per EULAR recommendations, MRA was considered as a first imaging modality for patients with TAK, considering the age of this population [22]. Interestingly, a prospective study by Quinn et al [53], demonstrated that MRA identified a greater extent of vascular involvement than PET because MRA can detect both arterial wall abnormalities (wall thickness and edema) and luminal abnormalities (aneurysm, stenosis, and occlusion). However, when investigators assessed disease activity, the interrater agreement was greater for PET scan reads than for MRA reads ( $\kappa = 0.84$  versus  $0.58$ ), indicating that assessment of disease activity by PET is more reliable than MRA. In that study, the authors found that vascular thickness and vascular edema on short-tau inversion recovery sequences were independently associated with the global FDG-PET/CT study interpretation on disease activity [53]. In another study, Einspieler et al [54] demonstrated consistent results between the use of FDG-PET/MRI and FDG-PET/CT. In that study, when FDG-PET and MRI were evaluated independently by an imaging physician, more abnormal vascular segments were detected on FDG-PET [54]. The use of T2-weighted sequences to assess disease activity is controversial in the literature; thus, the EULAR recommendation suggests that edema on T2-weighted images is less sensitive and prone to artifacts [22,75,76].

### **MRI Chest, Abdomen, and Pelvis**

An extensive literature search revealed that isolated MRI without angiography (MRA) is limited to a study by Kato et al [76]. In this study, the authors performed the late gadolinium enhancement sequence after contrast injection to the patients with TAK to assess disease distribution and activity of vessel wall inflammation. They concluded that the late gadolinium enhancement sequence has utility in detecting the extent; however, disease activity may be difficult to determine using late gadolinium enhancement alone.

### **MRA Coronary Arteries**

There is no relevant literature to support the use of MRA coronary arteries in the diagnosis of LVV.

### **MRA Neck**

There is no isolated or specific literature to support the use of MRA of the neck in the diagnosis of LVV. However, a recent study by Michailidou et al [68] demonstrated that neck MRA can be used along with chest MRA. However, the authors used MRA to assess vascular damage, such as aneurysm, stenosis, or occlusion.

### **MRI Heart Function and Morphology**

There is no relevant literature to support the use of MRA of the coronary arteries in the diagnosis of LVV.

### **US Duplex Doppler Aorta Abdomen**

There is a lack of evidence in the literature for the use of US duplex Doppler abdominal aorta in the diagnosis of LVV. US can be used in the assessment of and screening for abdominal aortic aneurysm. Although the abdominal aortic wall can be visualized with US, again, there is lack of evidence in the literature regarding the use of US for the diagnosis of vasculitis. One study by Loffler et al [77], in which FDG-PET/CT was used a reference test, sensitivity of US for LVV was demonstrated to be 26%.

### **US Duplex Doppler Chest, Abdomen, and Pelvis**

There is a lack of evidence in the literature for the use of US chest, abdomen, and pelvis in the diagnosis of LVV. In particular, thoracic aorta evaluation with US is hampered by the lungs [78]. US can be used in the assessment of



and screening for abdominal aortic aneurysm. Although the abdominal aortic wall can be visualized with the US, again, there is lack of evidence in the literature regarding the use of US for the diagnosis of vasculitis.

### **US Duplex Doppler Upper Extremity**

US is mainly used in the temporal arteries, and there is abundant literature discussing its high sensitivity and specificity in c-GCA diagnosis, when temporal artery biopsy is used as a reference tool [40]. However, it can also be used in the assessment of the upper-extremity vessels [23,79]. In particular, the assessment of the upper-extremity (axillary) arteries with US in patients with LVV can increase the diagnostic yield [2,78,80]. The classic US finding is described as a “halo” sign, which is a concentric hypoechoic rim of the wall around the artery lumen seen on 2 planes [23]. The compression sign is another feature of an inflamed vessel (it refers to the visibility of the vessel or incomplete compression of the vessel wall) following compression with the US transducer [81,82].

In a study by Loffler et al [77], in which FDG-PET/CT was used as a reference test, the sensitivity and specificity of US for the diagnosis of LVV were 80% and 70%, respectively (PPV 80%, NPV 70%). In their study, when authors analyzed separately, sensitivities of US for LVV diagnosis in the axillary and subclavian arteries were 72% and 71%, respectively. In TAK vasculitis, US can demonstrate a wide range of imaging findings, including hypoechoic rim/halo sign and hyperechoic rim around the vessel wall [83]. Although the echogenicity of the vessel wall is not helpful in distinguishing between acute or chronic inflammatory changes, it has been reported that active areas tend to have a thicker vessel wall [23]. A study demonstrated a good correlation between contrast-enhanced US and FDG-PET/CT findings [84]. Regarding the prognostic value of US, a study by Czihal et al [85] showed that concomitant temporal and upper-extremity vessel abnormality on US was associated with the poor treatment response in patients with GCA.

### **US Duplex Doppler Lower Extremity**

There are limited data in the literature for the use of US duplex Doppler lower extremity in the diagnosis of LVV. In a study by Loffler et al [77], in which FDG-PET/CT was used as a reference test, sensitivity of US for LVV diagnosis in the common femoral arteries was demonstrated to be 17%.

### **US Duplex Doppler Iliofemoral Arteries**

There is a lack of evidence in the literature for the use of US duplex Doppler iliofemoral arteries in the diagnosis of LVV.

## **Variant 2: Suspected medium-vessel vasculitis (MVV). Initial imaging.**

### **Arteriography Chest, Abdomen, and Pelvis**

Although digital subtraction angiography has lost its role as a diagnostic method for confirming a clinically suspected diagnosis of LVV, it is still a crucial modality in the diagnosis of MVV. Classic imaging findings in PAN are microaneurysms [86]. Similarly, for Kawasaki disease, catheter angiography is considered to be an ideal imaging modality [87].

### **CTA Chest, Abdomen, and Pelvis**

There is limited literature on the use of CTA to diagnose MVV. Singhal et al [88] reported positive CTA studies in 15 of 27 patients with PAN. The most common CTA finding was aneurysm, followed by stenosis/occlusion, and the renal artery was the most commonly involved artery [89]. Splenic and renal infarcts were the most visceral abnormalities. Therefore, CTA can be utilized in the diagnosis of the MVV.

### **CT Chest, Abdomen, and Pelvis**

There are sparse studies on the use of CT to diagnose MVV. CT of the chest, abdomen, and pelvis is an ideal imaging modality to assess end organ abnormalities in patients with MVV. A study by Singhal et al [88] reported splenic and renal infarcts were the most visceral abnormalities.

### **CTA Coronary Arteries**

Considering that Kawasaki disease targets the coronary arteries, there is a significant role for CTA in the diagnosis and follow-up of these patients [7,90]. However, there is a lack of evidence in the literature for the use of CTA coronary arteries in adult population.

### **FDG-PET/CT Whole Body**

FDG-PET/CT can assess relatively large vessels, so its usefulness in assessing MVV is limited [91].

### **MRA Chest, Abdomen, and Pelvis**

There is no relevant literature to support the use of MRA chest, abdomen, and pelvis in the diagnosis of the MVV. However, similar to CT chest, abdomen, and pelvis, MRA can be utilized in MVV to assess parenchymal changes of the visceral organs when there is a clinical concern.

### **MRI Chest, Abdomen, and Pelvis**

There is no relevant literature to support the use of MRI chest, abdomen, and pelvis in the diagnosis of the MVV. However, similar to CT chest, abdomen, and pelvis, MRI can be utilized in MVV to assess parenchymal changes of the visceral organs when there is a clinical concern.

### **MRA Coronary Arteries**

There is no relevant literature to support the use of MRA coronary arteries in the diagnosis of the LVV. However, there are some case reports describing the usage of coronary MRA in Kawasaki disease.

### **MRA Neck**

There is no relevant literature to support the use of MRA neck in the diagnosis of MVV.

### **MRI Heart Function and Morphology**

There is no relevant literature to support the use of MRI of the heart in the diagnosis of MVV. However, in the right clinical context (eg, Kawasaki disease), cardiac MRI can be utilized to assess complications.

### **US Duplex Doppler Aorta Abdomen**

There is no relevant literature to support the use of US duplex Doppler aorta abdomen in the diagnosis of MVV.

### **US Duplex Doppler Chest, Abdomen, and Pelvis**

There is no relevant literature to support the use of US duplex Doppler chest, abdomen, and pelvis in the diagnosis of MVV.

### **US Duplex Doppler Upper Extremity**

There is no relevant literature to support the use of US duplex Doppler upper extremity in the diagnosis of MVV.

### **US Duplex Doppler Lower Extremity**

There is no relevant literature to support the use of US duplex Doppler lower extremity in the diagnosis of MVV.

### **US Duplex Doppler Iliofemoral Arteries**

There is no relevant literature to support the use of US duplex Doppler iliofemoral arteries in the diagnosis of MVV.

### **Summary of Recommendations**

- **Variation 1:** MRA of the chest, abdomen, and pelvis with IV contrast, or MRA of the chest, abdomen, and pelvis without and with IV contrast, or CT of the chest, abdomen, and pelvis with IV contrast, or CT of the chest, abdomen, and pelvis without and with IV contrast, or CTA of the chest, abdomen, and pelvis with IV contrast, or FDG-PET/CT of the whole body is usually appropriate for initial imaging in a patient with suspected LVV. These procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). The panel did not agree on recommending MRA of the neck with IV contrast or MRA of the neck without and with IV contrast for initial imaging in a patient with suspected LVV as an isolated imaging modality. There is insufficient medical literature to conclude whether or not these patients would benefit from these two procedures in this clinical scenario. Diagnostic imaging in this patient population is controversial but may be appropriate.
- **Variation 2:** Arteriography of the chest, abdomen, and pelvis or CTA of the chest, abdomen, and pelvis with IV contrast is usually appropriate for initial imaging in a patient with suspected MVV. These procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). The panel did not agree on recommending MRA of the chest, abdomen, and pelvis without and with IV contrast or MRA of the chest, abdomen, and pelvis without IV contrast for initial imaging in a patient with suspected MVV. There is insufficient medical literature to conclude whether or not these patients would benefit from these two procedures in this clinical scenario. Diagnostic imaging in this patient population is controversial but may be appropriate.

## 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](http://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 [92].

Relative Radiation Level Designations		
Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
○	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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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.