American College of Radiology
ACR Appropriateness Criteria®
Clinically Suspected Vascular Malformation of the Extremities

Variant 1:   Upper or lower extremity. Suspected vascular malformation presenting with pain or findings of physical deformity including soft-tissue mass, diffuse or focal enlargement, discoloration, or ulceration. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRA extremity area of interest without and with IV contrast	Usually Appropriate	O
MRI extremity area of interest without and with IV contrast	Usually Appropriate	O
CTA extremity area of interest with IV contrast	Usually Appropriate	Varies
US duplex Doppler extremity area of interest	Usually Appropriate	O
MRA extremity area of interest without IV contrast	May Be Appropriate	O
CT extremity area of interest with IV contrast	May Be Appropriate	Varies
MRI extremity area of interest without IV contrast	May Be Appropriate	O
US extremity area of interest with IV contrast	May Be Appropriate	O
CT extremity area of interest without IV contrast	May Be Appropriate	Varies
CT extremity area of interest without and with IV contrast	Usually Not Appropriate	Varies
Radiography extremity area of interest	Usually Not Appropriate	Varies
Arteriography extremity area of interest	Usually Not Appropriate	Varies

Variant 2:   Upper or lower extremity. Vascular murmur (bruit or thrill). Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRA extremity area of interest without and with IV contrast	Usually Appropriate	O
MRI extremity area of interest without and with IV contrast	Usually Appropriate	O
US duplex Doppler extremity area of interest	Usually Appropriate	O
CTA extremity area of interest with IV contrast	Usually Appropriate	Varies
MRI extremity area of interest without IV contrast	May Be Appropriate	O
CT extremity area of interest with IV contrast	May Be Appropriate	Varies
CT extremity area of interest without and with IV contrast	May Be Appropriate (Disagreement)	Varies
MRA extremity area of interest without IV contrast	May Be Appropriate (Disagreement)	O
US extremity area of interest with IV contrast	May Be Appropriate	O
Arteriography extremity area of interest	May Be Appropriate	Varies
CT extremity area of interest without IV contrast	Usually Not Appropriate	Varies
Radiography extremity area of interest	Usually Not Appropriate	Varies

CLINICALLY SUSPECTED VASCULAR MALFORMATION OF THE EXTREMITIES

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Summary of Literature Review

Introduction/Background

Vascular anomalies encompass a broad range of pathologies histologically composed of vascular type cells. These lesions are most commonly classified by the International Society for the Study of Vascular Anomalies according to their underlying histology as either vascular malformations or vascular tumors. Vascular malformations represent focal structural abnormalities of the vascular tree, typically related to developmental errors during vasculogenesis [1], whereas, vascular tumors are caused by neoplastic cellular proliferation of the endothelium [1]. The extremities are the most common site of these vascular lesions outside of the head and neck [1,2].

Vascular malformations more commonly represent isolated spontaneous lesions yet can be part of one of several syndromes such as Parkes Weber syndrome [1,3]. These typically grow commensurate with patient age, often in conjunction with hormonal changes, such as puberty and pregnancy [3-6]. Therefore, vascular malformations that are present at birth may not present clinically until adolescence or adulthood. These lesions can be broadly divided into high- and low-flow lesions. High-flow malformations include arteriovenous malformations and arteriovenous fistulas, both of which demonstrate arterial flow and arteriovenous shunting. The former tend to be congenital lesions, and the latter are typically acquired as sequela of prior trauma or surgery. High-flow lesions comprise approximately 10% of peripheral vascular malformations and may present with pain, skin discoloration, warmth, or mass with palpable thrill or bruit [6]. Compression neuropathy, soft-tissue ulceration, bleeding, arterial steal phenomenon, and high-output cardiac failure may be seen in extreme cases [4,7]. Low-flow lesions include capillary, venous, and lymphatic malformations and are overall more common than high-flow lesions. Of these, capillary malformations are the most common but rarely require imaging for diagnosis because of their characteristic cutaneous manifestations [3,4]. Venous and lymphatic malformations have a reported prevalence of 1% in the general population with 40% involving the extremities [6]. Although symptomatology is variable, these lesions may present with focal or more generalized extremity pain, swelling, or compressible mass with or without associated skin discoloration. Involvement of the deep tissues, including bone, is not uncommon, and physical examination often underestimates their full extent [2,5].

Vascular tumors are subclassified based on propensity for aggressive and malignant behavior; however, the majority are benign [3]. Infantile hemangiomas are among the most common type of vascular neoplasm. These benign lesions present in infancy or early childhood and demonstrate rapid proliferative growth followed by eventual involution, most often not requiring treatment [7]. Less commonly, vascular tumors such as the intramuscular hemangioma, can present in adulthood with swelling, pain, or mass.

The diagnosis of a vascular malformation or neoplasm is frequently made clinically when classic signs and symptoms are present. Imaging is used for confirmation, particularly if the clinical presentation is atypical or vague [3,8] and is generally required for characterization of these lesions. Imaging also plays a critical role in treatment planning, which is often required because of growth, limb deformity, and decreased function, as well as pain. Lesion characteristics such as subtype (high flow versus low flow), depth, and invasion of adjacent structures, as well as inflow and outflow vessels, help in optimal treatment selection [9]. Treatment approach spans the spectrum of conservative to aggressive options, which include compression dressings, sclerotherapy,

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transarterial or transvenous embolization, and surgical resection. Sclerotherapy is often used for low-flow lesions, whereas high-flow lesions are most effectively treated with embolization [8,10].

Special Imaging Considerations

For the purposes of distinguishing between CT and CT angiography (CTA), ACR Appropriateness Criteria topics use the definition in the [ACR–NASCI–SIR–SPR Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA)](#) [11]:

> *"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 recons/reformats. Only in CTA, however, is 3-D rendering a required element. This corresponds to the definitions that the CMS has applied to the Current Procedural Terminology codes.

Discussion of Procedures by Variant

Variant 1: Upper or lower extremity. Suspected vascular malformation presenting with pain or findings of physical deformity including soft-tissue mass, diffuse or focal enlargement, discoloration, or ulceration. Initial imaging.

The body regions covered in this clinical scenario are shoulder, humerus, elbow, forearm, wrist, hand, hip, femur, knee, tibia/fibula, ankle, and foot.

Radiography Extremity

Radiographs are often used as the initial imaging modality in the workup of a patient presenting with nonspecific extremity complaints and may be useful for the exclusion of more common causes of extremity pain and deformity. However, radiographs are of limited utility for the specific purpose of vascular malformation imaging. In the setting of a vascular malformation, radiographs may be normal or show a soft-tissue mass [5,12]. Phleboliths may also be seen and provide a clue to the diagnosis of venous malformations and hemangiomas, which are reported to contain phleboliths in 20% to 67% of cases [3,12]. Lesions located adjacent to bone may be associated with bone changes, including periosteal reaction, remodeling, or signs of destruction, such as cortical scalloping and lucencies. Although such findings may be visible radiographically, they are not specific for the diagnosis of vascular malformations [6,12].

US Duplex Doppler Extremity

Ultrasound (US) with duplex Doppler imaging can be useful for the initial assessment of suspected vascular malformation, particularly if a focal mass or other targetable superficial symptomatology is present. US with Doppler can differentiate high-flow from low-flow malformations, and often provides a specific diagnosis in cases where characteristic vascular malformation features are present [3,5,13,14]. US may also be diagnostic of other lesions within the differential diagnosis for nonspecific extremity complaints [3,15]. Ultimately, this modality is limited in regards to tissue penetration and a small imaging field of view, which may lead to suboptimal evaluation of lesion extent and size, particularly if located deep in the extremity or adjacent to bone [2,8,13]. As a result, cross-sectional imaging, such as MRI, may be needed for more complete evaluation and definitive diagnosis [1,3,13], especially in cases without a targetable focal abnormality.

US Extremity Area of Interest with IV Contrast

There is limited evidence regarding the utility of contrast-enhanced US (CEUS) in evaluating suspected peripheral vascular malformations, but this modality may be considered in select cases. The addition of microbubble contrast may enhance visualization of small arteriovenous shunts and low-flow vessels compared with US with Doppler [16]. CEUS also has potential for quantifying perfusion in vascular malformations, which could be helpful in assessing treatment response [17].

CT Extremity

CT offers the benefit of high spatial resolution and provides comprehensive anatomic detail in the workup of extremity complaints. For the specific purpose of vascular malformation imaging, CT may reveal a soft-tissue mass with or without phleboliths, as well as provide information about size and lesion extent [12]. Bone involvement and acute complications like hemorrhage can also be assessed with CT [3,6]. Intravenous (IV)

contrast administration improves lesion delineation and allows for the assessment of enhancement patterns, which may help narrow the differential diagnosis of a focal finding [3]. However, in general, MRI is the preferred imaging modality when evaluating suspected vascular malformations that are due to its greater soft-tissue contrast and ability to obtain dynamic flow information with MR angiography (MRA) [10].

CTA Extremity
CTA with IV contrast can be used to evaluate a suspected vascular malformation. It is generally of greater utility for a high-flow lesion, such as an arteriovenous malformation, as CTA is capable of delineating the feeding arteries, nidus, and draining veins, which typically characterize these lesions [10]. CTA may also provide some information in regards to lesion extent and invasion into muscular compartments and bones [10]. However, MRI or MRA is the preferred method for suspected vascular malformation imaging that is due to superior soft-tissue contrast and potential for dynamic blood flow imaging [2,10,18].

MRI Extremity
MRI offers superior soft-tissue contrast compared with CT and plays an important role in the workup of suspected vascular malformations and soft-tissue masses. Lesion morphology and internal signal characteristics can be assessed, often allowing for definitive diagnosis [2-4,6,7,9,19]. MRI accurately determines lesion extent and involvement of surrounding structures, both of which are underestimated clinically in up to 76% of cases [2,5,7]. Contrast-enhanced sequences may not be necessary if typical features, such as flow voids, are present; however, the use of IV contrast is preferred for improved specificity and more complete characterization [3,6,8,20]. IV contrast also allows better visualization of the feeding and draining vessels in high-flow lesions, although this is best performed using an MRA protocol [1-3,6,7,21]. Additionally, MRI is useful for the evaluation of other soft-tissue lesions and musculoskeletal pathologies that might be considered in the differential diagnosis for a mass, enlarged extremity, or pain [19,20,22,23].

MRA Extremity
MRA is an excellent imaging option when a vascular malformation is suspected. The typical MRA protocol includes conventional T1 and T2 sequences, which provide anatomic information including lesion size, extent, and internal morphology [6,7]. Dynamic contrast-enhanced MRA, when combined with conventional MRI, has a reported sensitivity of 83% and specificity of 95% for the differentiation of venous and nonvenous malformations [24]. Time-resolved MRA has been shown to be nearly equivalent to arteriography for evaluating dynamic perfusion, allowing for accurate differentiation of feeding arteries and draining veins in high-flow lesions [2,9]. MRA can also be useful in differentiating vascular malformations from other causes of an extremity mass, such as soft-tissue neoplasms, although there may be some overlap in findings [7]. Although noncontrast time-of-flight techniques can be employed, contrast-enhanced MRA is preferred for improved depiction of smaller vessels and dynamic imaging assessment [6,25].

Arteriography Extremity
There is no evidence to support the use of arteriography as the initial imaging evaluation for a suspected vascular malformation because of its invasive nature. MRA is noninvasive and can depict the vascular anatomy of a malformation nearly as well as arteriography [9], making it the preferred initial imaging evaluation. Arteriography does offer the highest resolution imaging of small vessels and superior temporal resolution for assessment of flow dynamics. These advantages may be useful for high-flow lesions when MRA findings are equivocal or when treatment planning requires the highest available vascular detail resolution and/or better estimation of intralesional shunting [5-8].

Variant 2: Upper or lower extremity. Vascular murmur (bruit or thrill). Initial imaging.
The body regions covered in this clinical scenario are shoulder, humerus, elbow, forearm, wrist, hand, hip, femur, knee, tibia/fibula, ankle, and foot.

Radiography Extremity
Radiographs are of limited benefit for the specific purpose of vascular malformation imaging, especially in regard to lesions presenting with a vascular murmur. Radiographs may be normal or show a soft-tissue mass [5,12]. Venous malformations and hemangiomas may contain radiographically visible phleboliths in 20% to 67% of cases, which provide a clue to the diagnosis; however, these lesions typically do not present with a vascular murmur [3,12]. Lesions located adjacent to bone may be associated with bone changes, such as periosteal reaction, remodeling, or signs of destruction, such as cortical scalloping and lucencies. Although such findings may be visible radiographically, they are not specific for the diagnosis of vascular malformations [6,12].

US Duplex Doppler Extremity
US is fast and can often provide initial imaging characterization of vascular malformations [5-7,13]. The presence of a vascular murmur is clinically suggestive of a high-flow malformation, and US with Doppler imaging is generally regarded as a good initial option for the confirmation of a high-flow component. In many cases, US can help differentiate between the various types of vascular malformations and other soft-tissue lesions [5,7,13-15]. However, US has limitations in regard to field of view and tissue penetration that typically limits the ability to completely characterize and delineate the full extent of vascular malformations. Additional cross-sectional imaging, such as MRI, is usually needed for complete characterization [2,8,9].

US Extremity Area of Interest with IV Contrast
Limited evidence is available regarding the utility of CEUS specifically for the evaluation of peripheral vascular malformations, but this may be considered in select cases. Visualization of small arteriovenous shunts may be improved with CEUS compared with US with Doppler [16]. The potential for quantifying perfusion in vascular malformations with CEUS could also be useful in assessing treatment response [17].

CT Extremity
CT is a fast high-resolution modality that may be of some benefit, particularly in the acute setting, as this modality can rapidly provide information regarding lesion extent and assess for acute complications, such as hemorrhage [6]. CT can also be useful for the assessment of osseous involvement and is sensitive for the detection of vascular malformation–associated intralesional phleboliths [3,12]. IV contrast administration typically improves delineation of vascular malformations and allows for the assessment of enhancement patterns that may help narrow the differential diagnosis of a focal finding [3]. However, given the limited soft-tissue contrast and inability to evaluate flow characteristics, CT is generally not a preferred modality for the investigation of a suspected vascular malformation [3,6,12].

CTA Extremity
CTA features comparatively high spatial resolution, which allows for the characterization of a vascular nidus, enlarged feeding arteries, and draining veins that are frequently encountered in high-flow lesions that often present clinically with a vascular murmur [10]. CTA may also be useful for the assessment of other vascular-related pathologies, such as vasculitis and compression syndromes [10]. However, poor soft-tissue contrast and limited temporal resolution are drawbacks to this modality, and MRA is typically preferred over CTA [10,18].

MRI Extremity
The high tissue contrast of MRI makes it a preferred modality to assess the extent and distribution of vascular malformations, which are often underestimated by physical examination alone [2,5]. MRI is also a good option to evaluate for other soft-tissue lesions that may be included within a differential diagnosis of bruit on clinical examination [19,23]. The use of IV contrast improves lesion characterization and optimizes visualization of the surrounding anatomy [3,6,20]. Although MRI is generally considered high yield for evaluating lesion extent and often can distinguish between the various types of vascular malformations and soft-tissues masses, its evaluation of flow dynamics and intralesional vascular anatomy is limited when compared with MRA [3].

MRA Extremity
MRA has emerged as the preferred modality for assessing vascular malformations, particularly in patients with a vascular murmur and suspected high-flow malformation, due to its exceptional ability to delineate inflow and outflow anatomy noninvasively. Time-resolved MRA has been reported to rival conventional angiography for the portrayal of both functional flow dynamics and anatomic detail [9]. Combined conventional and dynamic contrast-enhanced MRA has a reported sensitivity of 83% and specificity of 95% for the differentiation of venous and nonvenous malformations [24]. Furthermore, MRA protocols typically include conventional high soft-tissue contrast T1 and T2 sequences which accurately assess the internal characteristics and extent of vascular malformations. This modality can also assess for and characterize other possible soft-tissue masses which may be included in the clinical differential diagnosis [6,7]. One potential weakness of time-resolved MRA is its underestimation of shunt volumes in vascular malformations which may be better evaluated with arteriography [7].

Arteriography Extremity
Arteriography offers high temporal and spatial resolution images of the vascular anatomy associated with high-flow lesions such as arteriovenous malformations, including the inflow and outflow vessels as well as intralesional shunting [5,7]. Although the presence of a vascular murmur increases the suspicion of a high-flow

lesion, MRA is usually the preferred initial imaging examination due to its noninvasive nature and nearly equivalent accuracy for providing vascular information, as well as depicting lesion morphology and extent [6,9]. Arteriography may be needed when MRA findings are equivocal or higher resolution images of vascular detail are needed for treatment planning [5-7].

Summary of Recommendations

- **Variant 1**: MRA of the upper or lower extremity without and with IV contrast, MRI of the upper or lower extremity without and with IV contrast, CTA of the upper or lower extremity with IV contrast, or US duplex Doppler of the upper or lower extremity are usually appropriate for the initial imaging of a suspected vascular malformation presenting with pain or findings of physical deformity. This includes a soft-tissue mass, diffuse or focal enlargement, discoloration, or ulceration. 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 area of interest for these four procedures covered in this clinical scenario includes the following body regions: shoulder, humerus, elbow, forearm, wrist, hand, hip, femur, knee, tibia/fibula, ankle, and foot.

- **Variant 2**: MRA of the upper or lower extremity without and with IV contrast, MRI of the upper or lower extremity without and with IV contrast, US duplex Doppler of the upper or lower extremity, or CTA of the upper or lower extremity with IV contrast are usually appropriate for the initial imaging of a vascular murmur (bruit or thrill). 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 area of interest for these four procedures covered in this clinical scenario includes the following body regions: shoulder, humerus, elbow, forearm, wrist, hand, hip, femur, knee, tibia/fibula, ankle, and foot. Additionally, the panel did not agree on recommending CT of the upper or lower extremity without and with IV contrast and MRA of the upper or lower extremity without IV contrast for this clinical scenario; both of these examinations may be of some benefit; however, other examinations are considered more appropriate for this purpose.

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 rating round tabulations for each recommendation.

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

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® Radiation Dose Assessment Introduction document [26].

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

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