American College of Radiology
ACR Appropriateness Criteria®

Soft Tissue Vascular Anomalies: Vascular Malformations and Infantile Vascular Tumors (Non-CNS)-Child

Variant: 1 Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging.

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

Variant: 2 Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
US abdomen	Usually Appropriate	O
US duplex Doppler abdomen	Usually Appropriate	O
US abdomen with IV contrast	May Be Appropriate (Disagreement)	O
Radiography abdomen	Usually Not Appropriate	☢☢
Arteriography abdomen	Usually Not Appropriate	☢☢☢☢☢
CTA and CTV abdomen with IV contrast	Usually Not Appropriate	☢☢☢☢☢
MRA and MRV abdomen without and with IV contrast	Usually Not Appropriate	O
MRI abdomen without and with IV contrast	Usually Not Appropriate	O
MRI abdomen without IV contrast	Usually Not Appropriate	O
CT abdomen with IV contrast	Usually Not Appropriate	☢☢☢☢
CT abdomen without IV contrast	Usually Not Appropriate	☢☢☢☢
CT abdomen without and with IV contrast	Usually Not Appropriate	☢☢☢☢☢

Variant: 3 Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
US area of interest	Usually Appropriate	O
US duplex Doppler area of interest	Usually Appropriate	O
MRA and MRV area of interest without and with IV contrast	Usually Appropriate	O
MRI area of interest without and with IV contrast	Usually Appropriate	O
US area of interest with IV contrast	May Be Appropriate (Disagreement)	O

MRA area of interest without IV contrast	May Be Appropriate (Disagreement)	O
MRI area of interest without IV contrast	May Be Appropriate	O
Arteriography area of interest	Usually Not Appropriate	Varies
Radiography area of interest	Usually Not Appropriate	Varies
CT area of interest with IV contrast	Usually Not Appropriate	Varies
CT area of interest without and with IV contrast	Usually Not Appropriate	Varies
CT area of interest without IV contrast	Usually Not Appropriate	Varies
CTA and CTV area of interest with IV contrast	Usually Not Appropriate	Varies

Variant: 4 Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.

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

Variant: 5 Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.

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

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Summary of Literature Review

Introduction/Background

Soft tissue vascular anomalies (VAs) may be diagnosed prenatally or at any time during life [1-3]. Anomalies of the soft tissues may be located in the extremities, face, scalp, neck, airway, thoracoabdominal wall, mediastinum, lungs, and abdomen (mesentery, retroperitoneum, and viscera). They may be associated with syndromes and may signal the presence of internal vascular lesions. Soft tissue VAs are subdivided into broad categories of vascular malformations (VMs) and vascular tumors (VTs). VTs are true neoplasms with increased mitotic activity and endothelial cell turnover; VMs are composed of abnormal or defective formation of vascular tissue [4] (see Appendix 1).

The prevalence of VMs varies by type: venous malformations (70%), lymphatic malformations (12%), arteriovenous malformations (AVMs) (8%), combined malformation syndromes (6%), and capillary malformations (4%) [5]. They may be divided into simple (further divided into low-flow or fast-flow VMs) or combined. Lowflow, simple VMs often contain 1 type of low-flow vessel: capillary, lymphatic, or venous vessel. AVMs or arteriovenous fistulas (AVFs) are fast-flow, simple VMs. Combined VMs are composed of more than 2 types of simple VM components and may be named for a major vessel [4,6]. Complex anomalies may be associated with overgrowth syndromes, which are often composed of infiltrative venous and lymphatic tissues through thickened subcutaneous fat affecting the trunk and/or limbs [7-10] (see Appendix 2).

VTs are divided into masses that behave in a benign, locally aggressive, borderline, or malignant manner. The most common benign VT is infantile hemangioma, which presents in the newborn period, whereas other VTs, malignant and other aggressive vascular lesions, may be diagnosed at any age. Most benign lesions are observed or treated in a noninvasive manner. Locally aggressive and borderline VTs present shortly after birth and may present with thrombocytopenia and/or a consumptive coagulopathy, which can complicate treatment. Malignant VTs are rapidly growing masses found in children of all ages, are often more aggressive than similar tumors in adults, and may be difficult to accurately diagnose due to poorly differentiated cell type [4,6].

This document pertains mainly to lesions that are easily accessible to imaging unless specified otherwise and does not include a discussion of central nervous syndrome (CNS) VA. For a discussion of VMs of the extremities and lungs in adults, please see the ACR Appropriateness Criteria® topics on “Clinically Suspected Vascular Malformation of the Extremities” [11] and “Clinically Suspected Pulmonary Arteriovenous Malformation (PAVM)” [12] for further guidance.

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: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging.

Infantile hemangiomas are the most common benign neoplasm of infancy, with a prevalence of 4% to 5% [13]. Infantile hemangiomas, distinct from VMs, are a true neoplasm rather than an abnormality of embryonic development of vascular tissue (see Appendix 1). They become clinically evident within the first few weeks of life and progress through phases of latency, growth, and plateau, predictably by the first year of life; in rare cases, growth continues through 24 months of age. With either complete or partial involution, lesion regression is completed by 4 years of age in 90% of cases but may continue through to 8 years of age [13,14]. Risk factors for having infantile hemangiomas include prematurity, White race (3%-10%), and female sex (female:male ratio range 1.4:1 to 3:1) [15].

Most infantile hemangiomas are diagnosed clinically, and imaging is useful for superficial lesions with atypical features or deep lesions, which are difficult to assess physically. Central or segmental location in the face and/or ears, breast, and midline lumbosacral region; lesions ≥4 cm; a presence of ≥5 hemangiomas; age of the patient; and growth rate can indicate the need for imaging [16,17]. In a recent study of 185 untreated infantile hemangiomas, photographs of lesions were studied to evaluate physical characteristics to distinguish the lesions associated with combined superficial and deep components. Therefore, it is important to understand both the superficial clinically evident findings of the surface and margins of the lesion and its extension into deep soft tissues to make recommendations for imaging and course of treatment when indicated.

A rare, but important, location of infantile hemangiomas is in the airway, because rapid proliferation of the VT can obstruct the airway. Infantile hemangiomas of the airway are most commonly localized in the subglottic airway but may be more diffuse, in a beard-like distribution throughout the soft tissues overlying the mandible and neck [18]. Subglottic infantile hemangiomas may also extend from the neck into the mediastinum. Infantile hemangioma may also be associated with VAs of other organs. PHACE, an acronym for posterior fossa malformations, hemangioma, arterial anomalies, coarctation of the aorta/cardiac defects, and eye abnormalities, is predominantly a neurovascular malformation syndrome [19]. Imaging of the non-CNS lesionsin PHACE syndrome, although not specifically addressed, are included in the recommendations of Variant 1. Imaging recommendations of hepatic hemangiomas found in patients with multiple cutaneous infantile hemangiomas are presented in Variant 2.

In the narrative below, "area of interest" can refer to the following: abdomen, chest, head, neck, pelvis, elbow, face, foot, forearm, hand, hip, knee, lower leg, shoulder, thigh, and upper arm.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging.
A. Arteriography area of interest

There is no relevant literature to support the use of arteriography of the area of interest as the initial imaging modality in infants with infantile hemangiomas.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. B. CT area of interest with IV contrast

When optimal imaging of the airway is required, as with hemangiomas involving the supra- or infraglottic airway or when in a beard-like distribution over the face and neck, CT with intravenous (IV) contrast may be useful. In a study of 11 children with hemangiomas of the upper airway who underwent CT, Koplewitz et al [20] found CT to have an improved definition of the airway lesion, presence, localization, and complete extent of the lesion and a more accurate size assessment compared with bronchoscopy.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. C. CT area of interest without and with IV contrast

There is no relevant literature to support the use of CT area of interest without and with IV contrast as the initial imaging modality in infants with infantile hemangiomas. In the study by Koplewitz et al [20], noncontrast and contrast-enhanced CT scans were performed. It was found that the lesions appeared larger after IV contrast was given and that the complete extent and localization of the lesion could be best defined.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. D. CT area of interest without IV contrast

There is no relevant literature to support the use of CT area of interest without IV contrast as the initial imaging modality in infants with infantile hemangiomas.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. E. CTA and CTV area of interest with IV contrast

There is no relevant literature to support the use of CT angiography (CTA) or CT venography (CTV) area of interest with IV contrast as the initial imaging modality in infants with infantile hemangiomas.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. F. MRA and MRV area of interest without and with IV contrast

Dynamic MR angiography (MRA) and MR venography (MRV) with IV contrast of untreated infantile hemangiomas is capable of showing supplying arterial and draining venous vessels [15].

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. G. MRI area of interest without and with IV contrast

MRI of the area of interest without and with IV contrast in patients with infantile hemangiomas may be useful when clinically determining the complete extent of the lesion is not possible, such as when the infantile hemangiomas of the face and deep facial structures or periorbital and intraorbital extent must be defined, when lumbosacral region infantile hemangiomas are present and underlying tethering or other spinal cord anomaly may be present, and there are beard-type infantile hemangiomas that occupy the pharyngeal region and may affect the oropharyngeal airway. MRI may also be useful in patients with infantile hemangiomas in anatomic locations, when the presence or growth of the lesion may be disfiguring or interfere with sight or hearing, the face, airway, ears, or breast [17].

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. H. MRI area of interest without IV contrast

There is no relevant literature to support the use of MRI of the area of interest without IV contrast as the initial imaging modality for infants with infantile hemangiomas.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. I. Radiography area of interest

There is no relevant literature to support the use of radiography area of interest as the initial imaging modality in infants with infantile hemangiomas.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. J. US area of interest

Ultrasound (US) of the area of interest is useful to distinguish imaging features of infantile hemangiomas from VMs. Paltiel et al [21] studied 49 lesions and Ding et al [14] studied 66 lesions, describing US imaging characteristics of superficial and deep infantile hemangiomas from well-circumscribed mixed echogenicity solid masses with central and peripheral vessels on grayscale US. Both groups showed that US is useful in distinguishing infantile hemangiomas from VMs and for identifying infantile hemangiomas, which may be combined with other VA components [14,21].

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. K. US area of interest with IV contrast

There is no relevant literature to support the use of US area of interest with IV contrast as the initial imaging modality in infants with infantile hemangiomas.

Variant 1: Infant. Clinical signs or symptoms of infantile hemangioma. Initial imaging. L. US duplex Doppler area of interest

US with duplex Doppler is most useful to assess and confirm diagnosis of infantile hemangiomas. Paltiel et al [21] studied 49 lesions and Ding et al [14] studied 66 lesions, showing that a combination of arterial and venous waveforms on US duplex Doppler enables distinguishing infantile hemangiomas from low-flow VMs.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.

Infants and children with multiple cutaneous infantile hemangiomas, defined as ≥5 lesions, may indicate the presence of additional hepatic or other visceral hemangiomas. Infantile hepatic hemangiomas that occur in infants with multiple cutaneous infantile hemangiomas are classified as multifocal or diffuse. The multifocal type is more common and regresses over time, whereas diffuse hepatic hemangiomas lead to hepatomegaly and are associated with poor outcomes including mortality [22].

A multicenter prospective study of 1,656 infants with infantile hemangiomas confirmed that patients with higher numbers of cutaneous infantile hemangiomas have a greater incidence of infantile hepatic hemangioma, 8.3% in patients with 5 to 9 lesions compared with 0.4% in patients with <5 cutaneous lesions. Based on the results of this study, screening liver imaging examination is indicated in patients with 5 cutaneous infantile hemangiomas and for patients up to 9 months of age [23].

In an analysis of 121 children with hepatic hemangioma in the Liver Hemangioma Registry, Kulungowski et al [24] found that 88 children had multiple cutaneous infantile hemangiomas lesions, 68 (77%) of which were multifocal and 20 (23%) of which were diffuse type infantile hepatic

hemangioma.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
A. Arteriography abdomen

There is no relevant literature to support the use of arteriography of the abdomen as the initial imaging modality when screening for hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
B. CT abdomen with IV contrast

There is no relevant literature to support the use of CT of the abdomen with IV contrast as the initial imaging modality when screening for hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
C. CT abdomen without and with IV contrast

There is no relevant literature to support the use of CT of the abdomen without and with IV contrast as the initial imaging modality when screening for hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
D. CT abdomen without IV contrast

There is no relevant literature to support the use of CT of the abdomen without IV contrast as the initial imaging modality when screening for hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
E. CTA and CTV abdomen with IV contrast

There is no relevant literature to support the use of CTA and CTV of the abdomen with IV contrast as the initial imaging modality when screening for hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
F. MRA and MRV abdomen without and with IV contrast

There is no relevant literature to support the use of MRA and MRV of the abdomen without and with IV contrast as the initial imaging modality when screening for hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
G. MRI abdomen without and with IV contrast

There is no relevant literature to support the use of MRI of the abdomen without and with IV contrast as the initial imaging modality when screening for hepatic hemangiomas in infants with

multiple cutaneous infantile hemangiomas. A recent guidance document of the American Society of Pediatric Hematology Oncology Vascular Anomalies Special Interest Group recommends contrast-enhanced MRI of the liver including dynamic sequences if the diagnosis is unclear following Doppler US [25].

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
H. MRI abdomen without IV contrast

There is no relevant literature to support the use of MRI of the abdomen and pelvis without IV contrast as the initial imaging modality when screening for hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
I. Radiography abdomen

There is no relevant literature to support the use of radiography of the abdomen as the initial imaging modality when screening for hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas.

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
J. US abdomen

US of the abdomen is useful as a screening examination for the presence of multifocal or diffuse infantile hepatic hemangioma in patients with ≥5 cutaneous infantile hemangiomas and for infants <9 months of age [23]. US may reveal the presence or absence of infantile hepatic hemangioma and provide guidance for treatment planning or indication for further imaging [13]. Multifocal infantile hepatic hemangiomas are >1 defined spherical discrete hypoechoic lesions and may appear hypoechoic or hyperechoic or have a mixed echogenicity appearance on US; diffuse infantile hepatic hemangiomas show hypoechoic nodules throughout the liver along with hepatomegaly [26].

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
K. US abdomen with IV contrast

US of the abdomen with IV contrast, specifically for evaluation of the liver, for the presence of hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas may be used to increase the sensitivity and diagnostic confidence, particularly for focal lesions that are seen in congenital hepatic hemangiomas rather than the diffuse and multifocal hepatic hemangiomas seen in infantile hemangiomas [26,27]. The addition of IV contrast for US of the abdomen has been shown by El-Ali et al [27] to differentiate infantile hepatic hemangioma in infants from congenital hemangioma of the liver in 5 infants based on the pattern of early and late arterial phase and delayed washout of IV contrast ($P = .0016$). The IV contrast enhancement pattern on US was similar to the manner in which the lesions are known to enhance with IV contrast on CT and MRI examinations [28].

Variant 2: Infant. Multiple cutaneous infantile hemangiomas, screening for infantile hepatic hemangiomas. Initial imaging.
L. US duplex Doppler abdomen

A recent guidance document of the American Society of Pediatric Hematology Oncology Vascular

Anomalies Special Interest Group recommends Doppler US of the liver as the preferred initial imaging study. On US examination imaging features such as multifocal and diffuse patterns are more specific to infantile hepatic hemangiomas and may be used to help differentiate them from congenital hemangiomas of the liver [25]. Solitary lesions may be larger in diameter and of heterogeneous echogenicity, with more prominent peripheral vascular components [26].

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.

VAs are a diverse group of lesions including tumors with benign, locally aggressive, or malignant behaviors and malformations, which may involve low-flow (venous or lymphatic) or fast-flow (arterial) blood flow [4] (see Appendix 1). Many VAs are fully formed before birth and do not clinically present in a fashion typical of infantile hemangiomas. Imaging characterization of the lesion and delineation of its extent are warranted. In the narrative below, "area of interest" can refer to the following: abdomen, chest, head, neck, pelvis, elbow, face, foot, forearm, hand, hip, knee, lower leg, shoulder, thigh, and upper arm.

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
A. Arteriography area of interest

There is no relevant literature to support the use of arteriography area of interest as the initial imaging modality for vascular lesion such as tumor or malformation.

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
B. CT area of interest with IV contrast

There is no relevant literature to support the use of CT area of interest with IV contrast as the initial imaging modality for vascular lesion such as tumor or malformation.

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
C. CT area of interest without and with IV contrast

There is no relevant literature to support the use of CT area of interest without and with IV contrast as the initial imaging modality for vascular lesion such as tumor or malformation.

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
D. CT area of interest without IV contrast

There is no relevant literature to support the use of CT area of interest without IV contrast as the initial imaging modality for vascular lesion such as tumor or malformation.

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
E. CTA and CTV area of interest with IV contrast

There is no relevant literature to support the use of CTA and CTV area of interest with IV contrast as the initial imaging modality for vascular lesion such as tumor or malformation.

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
F. MRA and MRV area of interest without and with IV contrast

Using contrast-enhanced MRA and MRV patterns of high or low signal intensity can help distinguish between lowflow and fast-flow VMs. In a study by van Rijswijk et al [29], they describe a 95% specificity and 83% sensitivity in differentiating venous and nonvenous malformations using dynamic contrast-enhanced MRA. Dynamic 4-D MRA with IV contrast may be used to detect the presence of arteriovenous microshunts in VMs, which have been found to be associated with the presence of phleboliths [30].

Subtraction MRA techniques subtract MR signal from noncontrast-enhanced MR images from a contrast-enhanced MRA, resulting in improved visualization of vascular structures as soft tissue signal is removed [31]. There may also be value in the use of noncontrast MRA as an anatomic survey before performing contrast-enhanced MRA to facilitate planning of field of view and contrast bolus timing [32].

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
G. MRA area of interest without IV contrast

Numerous noncontrast-enhanced MRA sequences are also available, relying on the speed of blood flow (flowdependent) and subject to signal loss in vessels with slow blood flow. Flow-independent noncontrast-enhanced MRA sequences produce bright blood or dark blood pool images and enable imaging a larger field of view. Noncontrast MRA images may incorporate longer scan times and therefore produce higher spatial resolution images[33]. Noncontrast MRA does not provide dynamic flow information provided by contrast-enhanced MRA. Furthermore, although time-of-flight cannot be used to characterize soft tissue components, it does show the feeding and draining vessels of a fast-flow VM [34].

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
H. MRI area of interest without and with IV contrast

MRI area of interest without and with IV contrast as the initial imaging modality for vascular lesion such as tumor or malformation may be helpful as the initial imaging examination and may be performed contemporaneously with US to investigate a soft tissue mass or skin discoloration.

MRI findings typically show a lobulated and often infiltrative soft tissue mass with T1 hypointense and T2 hyperintense signal, variable vascular flow voids, variable patterns of enhancement, and possibly phleboliths, depending upon the type of lesion. The addition of MRA may be beneficial to making a definitive diagnosis [31].

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
I. MRI area of interest without IV contrast

MRI area of interest without IV contrast as the initial imaging modality for vascular lesion such as tumor or malformation may be helpful as the initial imaging examination and may be performed contemporaneously with US to investigate a soft tissue mass or skin discoloration.

MRI without IV contrast typically shows a lobulated and often infiltrative soft tissue mass with T1 hypointense and T2 hyperintense signal, variable vascular flow voids, and possibly phleboliths, depending upon the type of lesion. The lack of contrast will limit the ability to characterize the type of vessel and characterization of flow through the lesion (high- versus low-flow) and, therefore, the

type of VM being evaluated. The addition of IV contrast and possibly MRA may be beneficial to making a more definitive, accurate characterization of the lesion and eventually the diagnosis [31].

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
J. Radiography area of interest

There is no relevant literature to support the use of radiography area of interest as the initial imaging modality for vascular lesions such as tumor or malformation [35]. Radiographs may reveal calcifications within a soft tissue mass, indicating the diagnosis of VM, but is not typically the initial imaging study when a VA is suspected. Phleboliths occur at the site of microshunts in VMs.

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
K. US area of interest

US may be used to distinguish characteristic features of VTs and to differentiate between low-flow and fast-flow VMs. Aside from infantile hemangioma, VTs reveal well-defined solid tissue components with variable echotexture [21].

Venous VMs can be partially characterized with grayscale US, and certain features such as multiple anechoic spaces, echogenic phleboliths, and expanded soft tissue spaces that are compressible (muscle, subcutaneous fat, dermis layers) can be diagnostic. Lymphatic VMs are also able to be at least partially characterized by grayscale US when multiple anechoic spaces with cysts, which may contain fluid-fluid levels in the event there has been prior infection or hemorrhage into the lesion, that are noncompressible are visualized. Venolymphatic VMs show a combination of the above features. Fast-flow VM-AVMs and AVFs show a cluster of vessels without an associated solid tissue mass [36,37] in contradistinction to hemangiomas.

Congenital hepatic hemangiomas are focal lesions of the liver associated with multiple cutaneous infantile hemangiomas.

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
L. US area of interest with IV contrast

US of the abdomen and pelvis with IV contrast, specifically for evaluation of the liver, for the presence of hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas may be used to increase sensitivity and diagnostic confidence, particularly for focal lesions, which are seen in congenital hepatic hemangiomas rather than the diffuse and multifocal hepatic hemangiomas seen in infantile hemangiomas [26,27]. The addition of IV contrast for US of the abdomen and pelvis has been shown by El-Ali et al [27] to differentiate infantile hepatic hemangioma in infants from congenital hemangioma of the liver in 5 infants based on the pattern of early and late arterial phase and delayed washout of IV contrast ($P = .0016$). The IV contrast enhancement pattern on US was similar to the manner in which the lesions are known to enhance with IV contrast on CT and MRI examinations [28].

Variant 3: Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.
M. US duplex Doppler area of interest

US with duplex Doppler may be used to distinguish characteristic features of VTs, verify arterial

waveforms in fast-flow VMs (arterialization of draining veins), and differentiate between low-flow and fast-flow VMs. Aside from infantile hemangioma, VTs typically demonstrate both arterial and venous waveforms. Venous malformations show multiple anechoic spaces, but the flow in venous VMs may be so slow that it is difficult to perceive on Doppler. Lymphatic VMs are also composed of multiple anechoic spaces with cysts, which may contain fluid-fluid levels in the event there has been prior infection or hemorrhage but will not have Doppler signal. Venolymphatic malformations show a combination of these features. Fast-flow VM-AVMs and AVFs show a cluster of vessels without an associated solid tissue mass [36] with fast-flow on Doppler US.

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.

If initial US imaging raises suspicion for the diagnosis of VM, further imaging is often helpful to visualize the entire extent of the lesion, assessfor the presence of multiple lesions, and evaluate possible involvement of adjacent tissues and organs (including sensitive anatomic regions such as deep facial structures, the airway, orbits, and the spine).

In the narrative below, "area of interest" can refer to the following: abdomen, chest, head, neck, pelvis, elbow, face, foot, forearm, hand, hip, knee, lower leg, shoulder, thigh, and upper arm.

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
A. Arteriography area of interest

Digital subtraction angiography provides an excellent definition of AVM anatomy, in particular the AVM nidus and the number and definition of feeding arteries and fistulas in fast-flow (arterial) lesions, with greater sensitivity than MRA [38]. Before an invasive procedure such as digital subtraction angiography is performed, it is best to have a narrowed differential diagnosis (usually requires prior MRI/MRA to suggest the diagnosis of a fast-flow VM). Diagnostic angiography may confirm the suspicion of a fast-flow VM if this remains in question following MRI/MRA but is typically reserved for symptomatic patients when simultaneous treatment is a leading consideration.

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
B. CT area of interest with IV contrast

Contrast-enhanced CT may provide further anatomic definition of a VM after US in other anatomies, aiding in visualizing phleboliths, thrombus, osseous changes such as erosion or findings related to overgrowth syndromes, and soft tissue involvement, especially in defining the deep or infiltrative extent of a VA [39-41].

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
C. CT area of interest without and with IV contrast

There is insufficient literature reviewing the efficacy of CT area of interest without and with IV contrast in this clinical scenario.

CT of the chest without and with IV contrast has been used for diagnosis in patients with pulmonary arteriovenous malformations (PAVMs), which are seen in children with hereditary hemorrhagic telangiectasia (HHT), as a followup examination for the depiction of precise anatomy

of simple and some complex lesions, after initial screening, which may be performed using transthoracic contrast echocardiography, which confirms the presence of a suspected shunt [42]. Both phases of the CT examination may provide important information in the diagnosis of PAVM [42].

In the case of PAVM feeding and draining, vascular components of the lesion are well defined as they course through the lung. Draining veins are typically 1 to 2 mm larger in diameter than the feeding arteries and can be differentiated on this basis. The intervening nidus is readily identifiable as a nodule [42]. In a retrospective study of 40 patients with 62 PAVMs, Gamondés et al [43] found that the diameter of the draining vein of 2.5 mm or larger was associated with PAVM reperfusion of the lung after embolotherapy with a sensitivity of 98.4% and specificity of 87.7%.

Contrast-enhanced CT may provide further anatomic definition of a VM after US in other anatomies, aiding in visualizing phleboliths, thrombus, osseous changes such as erosion or findings related to overgrowth syndromes, and soft tissue involvement, especially in defining the deep or infiltrative extent of a VA [39-41].

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
D. CT area of interest without IV contrast

There is no relevant literature reviewing the efficacy of noncontrast enhanced CT evaluation of VM. CT without IV contrast is limited in diagnosis of soft tissue and solid organ VMs and VTs because all structures and tissues show homogenous and similar attenuation, which obscures findings or does not allow characterization of the extent of abnormalities.

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
E. CTA and CTV area of interest with IV contrast

CTA and CTV of the upper extremity wasshown by Henzler et al [44] to be the superior imaging modality compared with MRI and US because it provides images with high spatial resolution for excellent delineation of the anatomy and extent of VTs and VMs, providing a vascular map of the lesion for use in treatment planning.

CTA of the chest, using a modified pulmonary CTA and CTV protocol, is used to optimally show the feeding artery, nidus, and draining vein components of the PAVM [42].

CTA and CTV may provide further anatomic definition of a VM after US in other anatomies, aiding in visualizing phleboliths, thrombus, osseous changes such as erosion or findings related to overgrowth syndromes, and soft tissue involvement, especially in defining the deep or infiltrative extent of a VA [39-41,45].

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
F. MRA and MRV area of interest without and with IV contrast

MRA of the area of interest without and with IV contrast may be used to define venous and arterial anatomy, as well as better differentiate the tissue types involved in the VM. MRA and MRV of the area of interest without IV contrast, using flow-dependent or flow-independent techniques, can be used as well. For example, Relaxation Enhanced Angiography without Contrast and Triggering, a

flow-independent T2-weighted noncontrast-enhanced MRA sequence, has been shown to be effective for defining the anatomy of major feeding arteries and draining veins and correlation with contrast-enhanced MRA and MRV [32,46].

Contrast-enhanced 3-D and 4-D dynamic MRA acquisitions are helpful in distinguishing whether flow through the lesion is slow or fast, arterial and venous anatomy, the location of a nidus in an AVM, and the site of vascular fistula in an AVF. Dynamic MRA combined with contrast-enhanced MRI has been shown to have excellent sensitivity (83%) and specificity (95%) in differentiating low-flow from fast-flow VMs [29,47].

Dynamic 4-D MRA with IV contrast may be used to detect presence of arteriovenous microshunts in VMs, which have been found to be associated with presence of phleboliths [30].

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
G. MRI area of interest without and with IV contrast

MRI of the area of interest without and with IV contrast may be used to define the deep and superficial extent of VM using T1-weighted sequences, and T2-weighted images reveal vascular flow voids as well as fluid filled spaces. Areas of signal loss or flow voids are important to document and can help drive the diagnosis (phleboliths versus fast-flow vessels). A well-defined soft tissue mass is not typically identified in AVM. IV contrast shows intense enhancement of involved soft tissues, cyst walls, and/or vascular structures [31].

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
H. MRI area of interest without IV contrast

There is no relevant literature to support the use of MRI area of interest without IV contrast as the next imaging modality for vascular lesion such as tumor or malformation.

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
I. Radiography area of interest

There is no relevant literature to support the use of radiography area of interest as the next imaging modality for VMs [35].

Variant 4: Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.
J. US area of interest with IV contrast

There is no relevant literature to support the use of US area of interest with IV contrast as the initial imaging modality for patients with vascular lesions such as tumor or malformation [48].

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.

VMs are a diverse group of lesions, which are difficult to treat, requiring multiple episodes of interventional embolization/sclerotherapy and or surgical intervention over years of treatment. Before and throughout the treatment course, interval imaging may help for monitoring regression of the lesion and planning approach to the next treatment session.

In the narrative below, “area of interest” can refer to the following: abdomen, chest, head, neck,

pelvis, elbow, face, foot, forearm, hand, hip, knee, lower leg, shoulder, thigh, and upper arm.

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
A. Arteriography area of interest

There is no relevant literature to support the use of arteriography as the initial diagnostic imaging modality for patients with an established diagnosis of a low-flow VM.

In patients with a fast-flow VM, angiography is useful to characterize new or persistent signs or symptoms (recurrent bleeding events, ischemic changes to normal tissues, etc) when simultaneous treatment is planned [49].

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
B. CT area of interest with IV contrast

Although contrast-enhanced CT may provide anatomic definition of a VM if there are persistent anatomical questions after MRI/MRA, there is no relevant literature to support the use of CT area of interest with IV contrast as the initial imaging modality for patients with an established diagnosis of VM presenting with new or persistent signs and symptoms [39].

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
C. CT area of interest without and with IV contrast

There is no relevant literature to support the use of CT area of interest without and with IV contrast as the initial imaging modality for patients with an established diagnosis of VM presenting with new or persistent signs and symptoms.

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
D. CT area of interest without IV contrast

There is no relevant literature to support the use of CT area of interest without IV contrast a as the initial imaging modality for patients with an established diagnosis of VM presenting with new or persistent signs and symptoms.

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
E. CTA and CTV area of interest with IV contrast

There is no relevant literature to support the use of CTA and CTV area of interest with IV contrast as the initial imaging modality for patients with an established diagnosis of VM presenting with new or persistent signs and symptoms.

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
F. MRA and MRV area of interest without and with IV contrast

MRA and MRV of the area of interest without and with IV contrast may be used to update venous and arterial anatomy as well as better differentiate the tissue types involved in the VM. As embolization and sclerotherapy treatments change the vascular channels, MRA can provide updated information.

Contrast-enhanced 3-D and 4-D dynamic MRA acquisitions are helpful in assessing flow through the lesion before and after treatment, the location of a new nidus in an AVM, and a new vascular fistula in an AVF. Dynamic MRA and MRV combined with contrast-enhanced MRI has been shown to have excellent sensitivity (83%) and specificity (95%) in differentiating low-flow from fast-flow VMs [29,47]. Embolization and sclerotherapy are expected to change the shunting within these low-flow VMs.

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
G. MRI area of interest without and with IV contrast

MRI of the area of interest without and with IV contrast may be used to define the deep and superficial extent of VM with expected treatment changes. IV contrast shows intense enhancement of involved soft tissues, cyst walls, and/or vascular structures [31]. Particular attention may need to be paid to imaging findings that may suggest treatment complications (abscess, tissue necrosis, cellulitis, deep vein thrombosis, etc).

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
H. MRI area of interest without IV contrast

There is no relevant literature to support the use of MRI area of interest without IV contrast as the initial imaging modality for patients with an established diagnosis of VM presenting with new or persistent signs and symptoms.

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
I. Radiography area of interest

Radiographs of the area of interest may reveal calcified phleboliths and embolization material within a previously treated VM; these findings may be helpful in choosing a follow-up imaging study (ie, US for suspected occluded deep vein after sclerotherapy). Radiographs may also better characterize imaging artifacts (embolization coils), which may limit the evaluation of residual VM on MRI/US [35].

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
J. US area of interest

US area of interest may be useful as an initial imaging modality in patients with an established diagnosis of VM when presenting with new or persistent signs or symptoms. However, US may be limited if there is extensive embolization material present. VM occlusions directly due to embolization versus associated soft tissue swelling from treatment changes can be visualized on grayscale US.

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
K. US area of interest with IV contrast

There is no relevant literature to support the use of US area of interest with IV contrast as the initial imaging modality for patients with an established diagnosis of VM presenting with new or persistent signs and symptoms.

It has been shown that the dynamics of blood flow through VM can be determined using contrast-

enhanced US, and that differences in blood flow dynamics as measured using time intensity curve analysis can be shown by comparing pretreatment and post-treatment examinations [50].

Variant 5: Child. Established diagnosis of vascular malformation presenting with new or persistent signs or symptoms. Initial imaging.
L. US duplex Doppler area of interest

US with duplex Doppler may be helpful as an initial imaging modality in patients with an established diagnosis of VM when presenting with new or persistent signs or symptoms. Doppler US can help distinguish changes following

Summary of Highlights

- **Variant 1:** Infantile hemangioma is a benign, true neoplasm, which grows in the first year of life and then regresses in a typical manner. In a child with clinical signs or symptoms of infantile hemangioma, US is usually an appropriate initial imaging examination, particularly for superficial lesions with atypical behavior. MRA and MRV without and with IV contrast, or MRI with and without IV contrast and CT with IV contrast, may be appropriate for determining for lesions of the face, deep facial structures, or periorbital and intraorbital structures, beard-type infantile hemangiomas that occupy the pharyngeal region and may affect the oropharyngeal airway. MRI without and with IV contrast may also be useful in patients with infantile hemangiomas in anatomic locations when the presence or growth of the lesion may be disfiguring or interfere with sight or hearing, the face, airway, ears, or breast, and when in the lumbosacral region. MRA and MRV without and with IV contrast, or MRI with and without IV contrast, are complementary examinations.
- **Variant 2:** In the presence of multiple (≥5) cutaneous infantile hemangioma lesions, screening for infantile hepatic hemangiomas with US abdomen may be appropriate in infants up to 9 months of age. US abdomen with IV contrast may increase the sensitivity to and diagnostic confidence in the detection of congenital hepatic hemangiomas, although there was disagreement among panelists regarding the use of IV contrast for US abdomen.
- **Variant 3:** In a child with clinical signs or symptoms of a VA not suspected to be infantile hemangioma, US, US duplex Doppler, MRA and MRV without and with IV contrast, and MRI without and with IV contrast of the area of interest are usually appropriate. US and US duplex Doppler are complementary examinations and are helpful for differentiation of low-flow and fast-flow VMs and distinction of solid and cystic components. MRA and MRV without and with IV contrast, and MRI without and with IV contrast of the area of interest are complementary examinations, which are useful to visualize the deep and or diffuse extent of lesions and MRA may be beneficial in making a definitive diagnosis. MRI of the area of interest without IV contrast may be appropriate and may be complementary to US of the area of interest. The addition of IV contrast and possibly MRA may be beneficial to making a more definitive, accurate characterization of the lesion and eventually the diagnosis. Panelists disagreed as to whether US with IV contrast or MRA without IV contrast may be appropriate. US with IV contrast is a complementary examination with US and US duplex Doppler of the area of interest. Although there is early evidence that US with IV contrast may increase the sensitivity to and diagnostic confidence in the detection of congenital hepatic hemangiomas, there is not yet evidence of this in the medical literature for other anatomies. MRA without IV contrast may be a complementary examination with US and US duplex Doppler of the area of interest; however,

the addition of IV contrast and possibly MRA may be beneficial to making a more definitive, accurate characterization of the lesion and eventually the diagnosis.

- **Variant 4:** Following US examination, which raises concern for VMs, MRA and MRV without and with IV contrast and MRI without and with IV contrast in the area of interest are usually appropriate and are complementary examinations for defining venous and arterial anatomy, as well as better differentiate the tissue types involved in the VM. Dynamic MRA acquisitions are helpful in distinguishing whether flow through the lesion is slow or fast, arterial and venous anatomy, the location of a nidus in an AVM, and the site of vascular fistula in an AVF. CT with IV contrast and CTA and CTV with IV contrast in the area of interest may be appropriate and are complementary examinations, which may be beneficial in visualizing phleboliths, thrombus, osseous changes such as erosion or findings related to overgrowth syndromes, and soft tissue involvement, especially in defining the deep or infiltrative extent of a VA.
- **Variant 5:** In children with established diagnoses of VMs, with new or persistent symptoms, US duplex Doppler, MRA and MRV without and with IV contrast, and MRI without and with IV contrast in the area of interest are usually appropriate. US duplex Doppler of the area of interest, as it is helpful in distinguishing changes following treatment, as well as new areas of involvement. MRA and MRV without and with IV contrast, and MRI without and with IV contrast in the area of interest are complementary examinations useful for distinguishing the extent of the lesion and for assessing flow through the lesion before and after treatment, location of a new nidus in an AVM, and a new vascular fistula in an AVF. US and US with IV contrast of the area of interest may be appropriate as complementary examinations to US duplex Doppler of the area of interest because it has been shown that the dynamics of blood flow through a VM can be determined using contrast-enhanced US and that differences in blood flow dynamics as measured using time intensity curve analysis can be shown by comparing pretreatment and post-treatment examinations.

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 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.

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 (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."

References

1. Crivelli L, Millischer AE, Sonigo P, et al. Contribution of magnetic resonance imaging to the prenatal diagnosis of common congenital vascular anomalies. Pediatr Radiol 2021.
2. Francavilla ML, White CL, Oliveri B, Lee EY, Restrepo R. Intraabdominal Lymphatic Malformations: Pearls and Pitfalls of Diagnosis and Differential Diagnoses in Pediatric Patients. [Review]. AJR Am J Roentgenol. 208(3):637-649, 2017 Mar.

3. Oliver ER, Coleman BG, DeBari SE, et al. Fetal Lymphatic Malformations: More Variable Than We Think?. J Ultrasound Med. 36(5):1051-1058, 2017 May.

4. International Society for The Study of Vascular Anomalies: ISSVA classification for vascular anomalies©. Available at: https://www.issva.org/UserFiles/file/ISSVA-Classification-2018.pdf.

5. Sadick M, Muller-Wille R, Wildgruber M, Wohlgemuth WA. Vascular Anomalies (Part I): Classification and Diagnostics of Vascular Anomalies. [Review]. ROFO Fortschr Geb Rontgenstr Nuklearmed. 190(9):825-835, 2018 Sep.

6. Monroe EJ. Brief Description of ISSVA Classification for Radiologists. Tech Vasc Interv Radiol 2019;22:100628.

7. Asilian A, Kamali AS, Riahi NT, Adibi N, Mokhtari F. Proteus Syndrome with Arteriovenous Malformation. Adv Biomed Res 2017;6:27.

8. Tan WH, Baris HN, Burrows PE, et al. The spectrum of vascular anomalies in patients with PTEN mutations: implications for diagnosis and management. J Med Genet 2007;44:594-602.

9. Turnbull MM, Humeniuk V, Stein B, Suthers GK. Arteriovenous malformations in Cowden syndrome. J Med Genet 2005;42:e50.

10. Uller W, Fishman SJ, Alomari AI. Overgrowth syndromes with complex vascular anomalies. Semin Pediatr Surg 2014;23:208-15.

11. Obara P, McCool J, Kalva SP, et al. ACR Appropriateness Criteria® Clinically Suspected Vascular Malformation of the Extremities. J Am Coll Radiol 2019;16:S340-S47.

12. American College of Radiology. ACR Appropriateness Criteria®: Clinically Suspected Pulmonary Arteriovenous Malformation (PAVM). Available at: https://acsearch.acr.org/docs/3094113/Narrative/.

13. Leaute-Labreze C, Harper JI, Hoeger PH. Infantile haemangioma. Lancet 2017;390:85-94.

14. Ding A, Gong X, Li J, Xiong P. Role of ultrasound in diagnosis and differential diagnosis of deep infantile hemangioma and venous malformation. J Vasc Surg Venous Lymphat Disord. 7(5):715-723, 2019 09.

15. Restrepo R, Palani R, Cervantes LF, Duarte AM, Amjad I, Altman NR. Hemangiomas revisited: the useful, the unusual and the new. Part 1: overview and clinical and imaging characteristics. [Review]. Pediatr Radiol. 41(7):895-904, 2011 Jul.

16. Leaute-Labreze C, Baselga Torres E, Weibel L, et al. The Infantile Hemangioma Referral Score: A Validated Tool for Physicians. Pediatrics 2020;145.

17. Restrepo R, Palani R, Cervantes LF, Duarte AM, Amjad I, Altman NR. Hemangiomas revisited: the useful, the unusual and the new. Part 2: endangering hemangiomas and treatment. [Review]. Pediatr Radiol. 41(7):905-15, 2011 Jul.

18. Darrow DH. Management of Infantile Hemangiomas of the Airway. Otolaryngol Clin North Am 2018;51:133-46.

19. Bayer ML, Frommelt PC, Blei F, et al. Congenital cardiac, aortic arch, and vascular bed anomalies in PHACE syndrome (from the International PHACE Syndrome Registry). Am J Cardiol. 112(12):1948-52, 2013 Dec 15.

20. Koplewitz BZ, Springer C, Slasky BS, et al. CT of hemangiomas of the upper airways in children. AJR Am J Roentgenol 2005;184:663-70.

21. Paltiel HJ, Burrows PE, Kozakewich HP, Zurakowski D, Mulliken JB. Soft-tissue vascular anomalies: utility of US for diagnosis. Radiology. 214(3):747-54, 2000 Mar.

22. Dickie B, Dasgupta R, Nair R, et al. Spectrum of hepatic hemangiomas: management and outcome. J Pediatr Surg 2009;44:125-33.

23. Ji Y, Chen S, Yang K, et al. Screening for infantile hepatic hemangioma in patients with cutaneous infantile hemangioma: A multicenter prospective study. J Am Acad Dermatol 2021;84:1378-84.

24. Kulungowski AM, Alomari AI, Chawla A, Christison-Lagay ER, Fishman SJ. Lessons from a liver hemangioma registry: subtype classification. J Pediatr Surg 2012;47:165-70.

25. Iacobas I, Phung TL, Adams DM, et al. Guidance Document for Hepatic Hemangioma (Infantile and Congenital) Evaluation and Monitoring. J Pediatr. 203:294-300.e2, 2018 12.

26. Xu M, Pan FS, Wang W, et al. The value of clinical and ultrasound features for the diagnosis of infantile hepatic hemangioma: Comparison with contrast-enhanced CT/MRI. Clin Imaging. 51:311-317, 2018 Sep - Oct.

27. El-Ali AM, McCormick A, Thakrar D, Yilmaz S, Malek MM, Squires JH. Contrast-Enhanced Ultrasound of Congenital and Infantile Hemangiomas: Preliminary Results From a Case Series. AJR Am J Roentgenol. 214(3):658-664, 2020 03.

28. Anupindi SA, Biko DM, Ntoulia A, et al. Contrast-enhanced US Assessment of Focal Liver Lesions in Children. Radiographics 2017;37:1632-47.

29. van Rijswijk CS, van der Linden E, van der Woude HJ, van Baalen JM, Bloem JL. Value of dynamic contrast-enhanced MR imaging in diagnosing and classifying peripheral vascular malformations. AJR Am J Roentgenol. 178(5):1181-7, 2002 May.

30. Hammer S, Uller W, Manger F, Fellner C, Zeman F, Wohlgemuth WA. Time-resolved magnetic resonance angiography (MRA) at 3.0 Tesla for evaluation of hemodynamic characteristics of vascular malformations: description of distinct subgroups. Eur Radiol. 27(1):296-305, 2017 Jan.

31. Flors L, Leiva-Salinas C, Maged IM, et al. MR imaging of soft-tissue vascular malformations: diagnosis, classification, and therapy follow-up. Radiographics 2011;31:1321-40; discussion 40-1.

32. Tan EJ, Zhang S, Tirukonda P, Chong LR. REACT - A novel flow-independent non-gated non-contrast MR angiography technique using magnetization-prepared 3D non-balanced dual-echo dixon method: Preliminary clinical experience. Eur J Radiol Open 2020;7:100238.

33. Fleecs JB, Artz NS, Mitchell GS, Chan SS. Non-contrast magnetic resonance angiography/venography techniques: what are my options?. [Review]. Pediatric Radiology. 52(2):271-284, 2022 Feb.

34. Vilanova JC, Barcelo J, Villalon M. MR and MR angiography characterization of soft tissue vascular malformations. Curr Probl Diagn Radiol 2004;33:161-70.

35. Hammer S, Zeman F, Fellner C, Wohlgemuth WA, Uller W. Venous Malformations: Phleboliths Correlate With the Presence of Arteriovenous Microshunts. AJR Am J Roentgenol. 211(6):1390-1396, 2018 12.

36. Kollipara R, Dinneen L, Rentas KE, et al. Current classification and terminology of pediatric vascular anomalies. [Review]. AJR Am J Roentgenol. 201(5):1124-35, 2013 Nov.

37. van Es J, Kappelhof NA, Douma RA, Meijers JCM, Gerdes VEA, van der Horst CMAM. Venous thrombosis and coagulation parameters in patients with pure venous malformations. Netherlands Journal of Medicine. 75(8):328-334, 2017 Oct.Neth J Med. 75(8):328-334, 2017 Oct.

38. Tan KT, Simons ME, Rajan DK, Terbrugge K. Peripheral high-flow arteriovenous vascular malformations: a single-center experience. J Vasc Interv Radiol 2004;15:1071-80.

39. Johnson JB, Cogswell PM, McKusick MA, Binkovitz LA, Riederer SJ, Young PM. Pretreatment imaging of peripheral vascular malformations. J Vasc Diagn 2014;2014:121-26.

40. Li JL, Liu HJ, Cui YH, et al. Mediastinal hemangiomas: Spectrum of CT and MRI findings - retrospective case series study and systematic review of the literature. Eur J Radiol. 126:108905, 2020 May.

41. Merrow AC, Gupta A, Patel MN, Adams DM. 2014 Revised Classification of Vascular Lesions from the International Society for the Study of Vascular Anomalies: Radiologic-Pathologic Update. Radiographics 2016;36:1494-516.

42. Saboo SS, Chamarthy M, Bhalla S, et al. Pulmonary arteriovenous malformations: diagnosis. Cardiovasc Diagn Ther 2018;8:325-37.

43. Gamondes D, Si-Mohamed S, Cottin V, et al. Vein Diameter on Unenhanced Multidetector CT Predicts Reperfusion of Pulmonary Arteriovenous Malformation after Embolotherapy. Eur Radiol. 26(8):2723-9, 2016 Aug.

44. Henzler T, Vogler N, Lange B, et al. Low dose time-resolved CT-angiography in pediatric patients with venous malformations using 3rd generation dual-source CT: Initial experience. Eur J Radiol Open 2016;3:216-22.

45. Ghouri MA, Gupta N, Bhat AP, et al. CT and MR imaging of the upper extremity vasculature: pearls, pitfalls, and challenges. Cardiovasc Diagn Ther 2019;9:S152-S73.

46. Yoneyama M, Zhang S, Hu HH, et al. Free-breathing non-contrast-enhanced flow-independent MR angiography using magnetization-prepared 3D non-balanced dual-echo Dixon method: A feasibility study at 3 Tesla. Magn Reson Imaging 2019;63:137-46.

47. Lidsky ME, Spritzer CE, Shortell CK. The role of dynamic contrast-enhanced magnetic resonance imaging in the diagnosis and management of patients with vascular malformations. J Vasc Surg. 56(3):757-64.e1, 2012 Sep.

48. Rauch M, Schild HH, Strunk H. Contrast enhanced ultrasound of a hepatic soft tissue angiosarcoma metastasis. Case report. Med Ultrason 2014;16:271-3.

49. Park KB, Do YS, Kim DI, et al. Predictive factors for response of peripheral arteriovenous malformations to embolization therapy: analysis of clinical data and imaging findings. J Vasc Interv Radiol. 23(11):1478-86, 2012 Nov.

50. Wiesinger I, Schreml S, Wohlgemuth WA, Stroszczynski C, Jung EM. Perfusion quantification of vascular malformations using contrast-enhanced ultrasound (CEUS) with time intensity curve analysis before and after treatment: First results. Clin Hemorheol Microcirc. 62(4):283-90, 2015 Sep 25.

51. American College of Radiology. ACR Appropriateness Criteria® Radiation Dose Assessment

Introduction. Available at: https://edge.sitecorecloud.io/americancoldf5f-acrorgf92a-productioncb02-3650/media/ACR/Files/Clinical/Appropriateness-Criteria/ACR-Appropriateness-Criteria-Radiation-Dose-Assessment-Introduction.pdf.

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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.

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