Imaging Pediatric Vascular Lesions

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ABSTRACT

Vascular anomalies are commonly encountered in pediatric and dermatology practices. Most of these lesions are benign and easy to diagnose based on history and clinical exam alone. However, in some cases the diagnosis may not be clear. This may be of particular concern given that vascular anomalies may occasionally be associated with an underlying syndrome, congenital disease, or serious, life-threatening condition. Defining the type of vascular lesion early and correctly is particularly important to determine the optimal approach to management and treatment of each patient. The care of pediatric patients often requires collaboration from a multitude of specialties including pediatrics, dermatology, plastic surgery, radiology, ophthalmology, and neurology. Although early characterization of vascular lesions is important, consensus guidelines regarding the evaluation and imaging of vascular anomalies does not exist to date. Here, the authors provide an overview of pediatric vascular lesions, current classification systems for characterizing these lesions, the various imaging modalities available, and recommendations for appropriate imaging evaluation. (*J Clin Aesthet Dermatol.* 2015;8(12):27–41.)

Vascular anomalies are commonly seen in pediatric patients, with an estimated prevalence of at least 4.5 percent.[1] Although typically benign, some vascular anomalies can be part of a syndrome or may be associated with serious, life-threatening conditions. The care of patients with vascular anomalies often requires a multidisciplinary approach. Dermatologists, radiologists, plastic surgeons, neurologists, otolaryngologists, plastic surgeons, hematologists, and pediatricians may all play an integral role in the management of these patients. Defining the type of vascular lesion early and correctly is particularly important in determining the approach to management of the individual patient. This article will discuss an array of pediatric vascular lesions, review the different imaging modalities available, and provide recommendations for imaging based on lesion type.

BACKGROUND ON VASCULAR ANOMALIES

The term "vascular anomaly" encompasses a wide variety of vascular abnormalities, including both proliferative, tumor-like lesions, which grow rapidly, and developmental anomalies, which tend to be more static in their growth patterns. Vascular anomalies can affect any part of the vasculature from the capillaries to the lymphatic system.[2] Because the term "vascular anomaly" encompasses a variety of different lesions, including hemangiomas, pyogenic granulomas, and arteriovenous malformations (AVM), multiple classification systems have been created. In 1982, Mulliken and Glowacki[3] created a biologic classification system for vascular anomalies based on the cellular and histologic characteristics and clinical course of the lesion. This classification system divided vascular anomalies into malformations (i.e., lesions with stable endothelial turnover) and tumors (i.e., lesions with upregulated endothelial cell growth). These two categories are distinct, with tumors representing a predominantly proliferative process, while malformations are thought to represent an aberration in vessel development. Finn et al[4] demonstrated that, by using history and physical exam along with this classification system alone, 96 percent of childhood vascular anomalies could be correctly classified as either "vascular malformation" or as "vascular tumor". This classification system has since been adopted by the International Society for the Study of Vascular Anomalies (ISSVA) in 1996.[5]

Recently, the ISSVA released a novel, expanded classification system for vascular anomalies.[6] Based on the original classification system created by Mulliken and Glowacki, this new system continues to divide vascular anomalies into tumors and malformations. However, it further delineates subcategories of each. The vascular

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TABLE 1. 2014 ISSVA categorization for vascular anomalies[6]

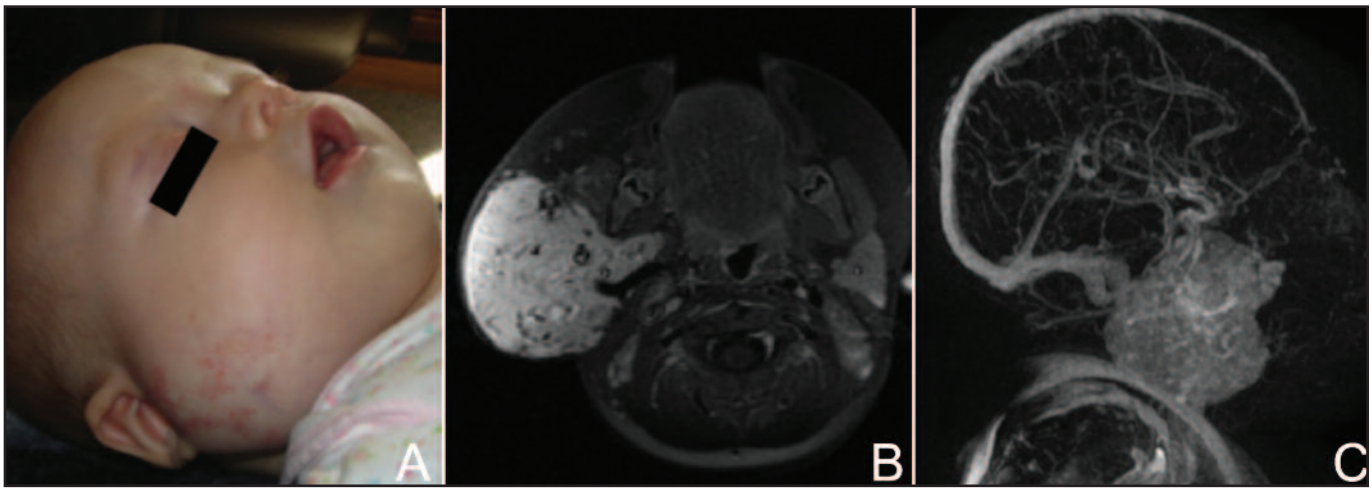
VASCULAR ANOMALIES	TUMORS	**Benign**	Infantile hemangioma, congenital hemangioma, tufted angioma, spindle-cell hemangioma, epithelioid hemangioma, pyogenic granuloma	
		Locally aggressive	Kaposiform hemangioendothelioma, retiform hemangioendothelioma, papillary intralymphatic angioendothelioma (PILA), Dabska tumor, composite hemangioendothelioma, Kaposi Sarcoma	
		Malignant	Angiosarcoma, epithelioid hemangioendothelioma	
	MALFORMATIONS	**Simple**	Capillary	Port-wine stain, telangiectasia, marmorata telangiectatica congenita, nevus simplex
			Lymphatic	Common (cystic), generalized lymphatic anomaly (GLA), channel type, primary lymphedema
			Venous	Common, familial venous malformation cutaneomucosal (VMCM), blue rubber bleb (bean) syndrome, glomulovenous malformation (GVM), Cerebral cavernous malformation (CCM)
			Arteriovenous	
			Arteriovenous fistula	
		Combined	Capillary (C), venous (V), lymphatic (L), arteriovenous (AV) CVM, CLM, CAVM, LVM, CLVM, CLAVM, CVAVM, CLVAVM	
		Associated with other anomalies	Klippel-Trenaunay syndrome, Parkes-Weber syndrome, Servelle-Martorell syndrome, Sturge-Weber syndrome, Maffucci syndrome, CLOVES syndrome, Proteus syndrome, etc.	

tumors are divided into benign, locally aggressive, and malignant groups, and the vascular malformations into simple, combined, and those associated with other anomalies (Table 1). Imaging is often an integral part of determining the proper classification of a vascular lesion, particularly when a vascular malformation is suspected. Therefore, it is particularly important for physicians to understand the proper utilization of imaging modalities currently available.

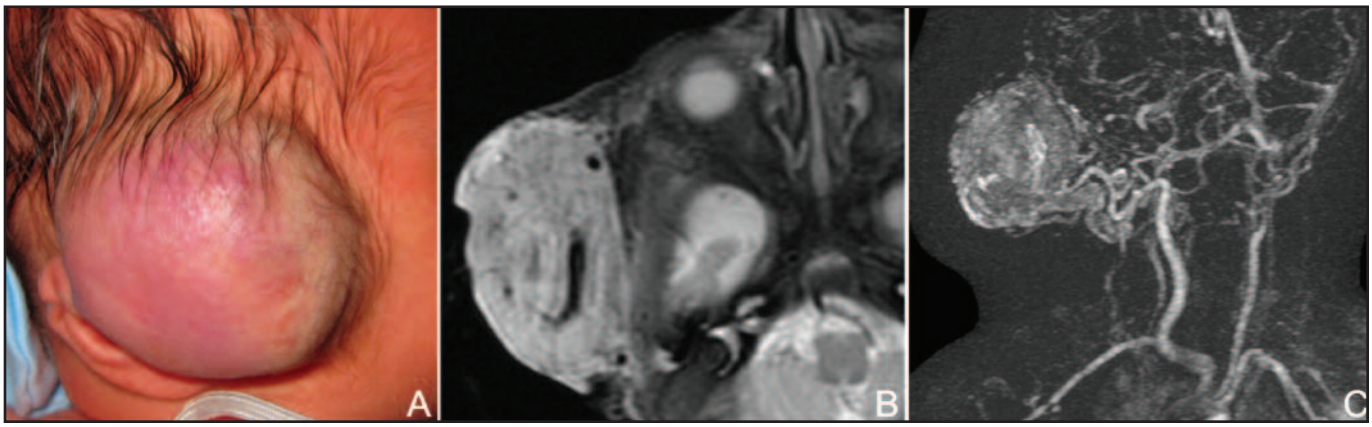
VASCULAR TUMORS (BENIGN, LOCALLY AGGRESSIVE, OR MALIGNANT)

Vascular tumors consist of vascular anomalies with upregulated endothelial cell growth. The most frequently seen vascular tumors are infantile hemangiomas (IH), congenital hemangiomas (CH), pyogenic granulomas (PG), and kaposiform hemangioendotheliomas (KHE).[1]

Infantile hemangiomas. IHs are benign vascular tumors and are considered the most common tumors of infancy, occurring in approximately five percent of infants.[7] IHs are more common in female infants, multiple gestations, and premature infants.[8] They usually appear in the first few days or months after birth and present as a bright red patch if "superficial," a bluish nodule if "deep," or a combination of both if "mixed" (Figure 1A).[8] Occasionally, they may initially present as an area of cutaneous pallor. In general, they present in the first 2 to 4 weeks of life, grow rapidly for 3 to 6 months, and then enter a more quiescent or static phase. They subsequently involute, but the exact onset of this occurrence varies widely. The first signs of involution are lightening and softening of the lesions. Most children will exhibit evidence of involution by 4 to 5 years of life, and if



Figures 1A–1C. (A) Infantile hemangioma on the right cheek, (B) Infantile hemangioma with early, diffuse enhancement with flow voids on gadolinium contrast-enhanced MRI, (C) Mass-like enhancement of an infantile hemangioma on dynamic time-enhanced MRA



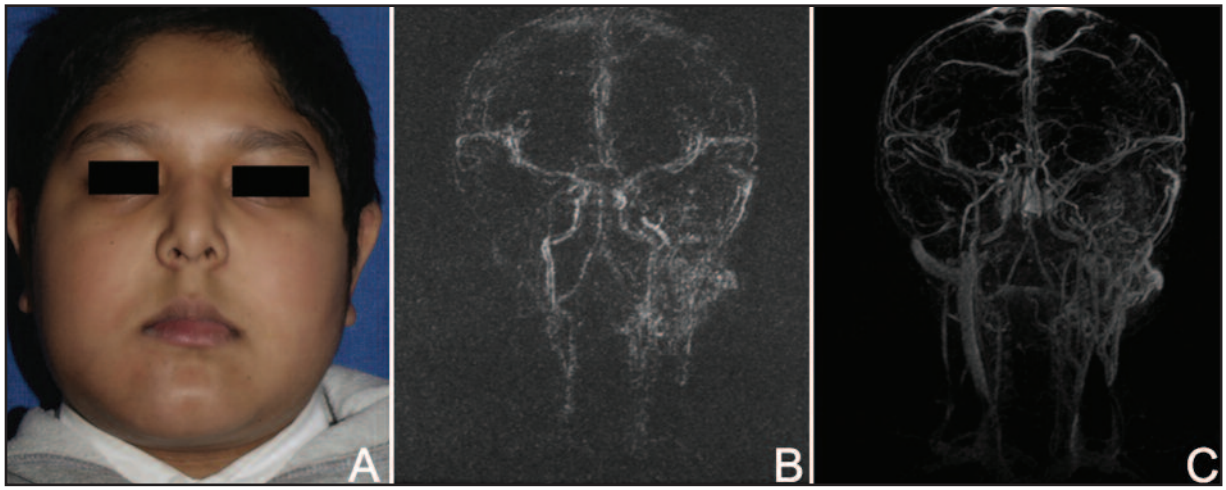
Figures 2A–2C. (A) Congenital hemangioma of the right preauricular area, (B) T2-weighted MRI showing a hyperintense congenital hemangioma, (C) Mass-like enhancement of a congenital hemangioma on dynamic time-enhanced MRA

this does not occur, one should rethink the diagnosis. Though the majority of these lesions may not warrant clinical intervention, some IHs may be associated with functional or cosmetic deformity, ulceration, bleeding, or genetic syndromes.[8]

Congenital hemangiomas. CHs are benign vascular tumors that may present very similarly to IHs (Figure 2A). Unlike IH, however, CHs are much less common and always present at birth (i.e., truly congenital). Two major subtypes of CHs exist: rapidly involuting congenital hemangiomas (RICH) and noninvoluting congenital hemangiomas (NICH).[9] Another, "intermediate," or gradually resolving subtype has recently been identified. RICHs are characterized by rapid involution and complete regression of the hemangioma by approximately two years of age.[9] NICHs, on the other hand, may partially involute; however, complete regression is rare.[9] Complications of congenital hemangiomas may include ulceration, bleeding, thrombocytopenia, and heart failure. Therefore, early identification and, if necessary, treatment of these lesions is the goal.

Pyogenic granulomas. PG, also known as lobular capillary hemangioma, is a benign vascular tumor commonly seen in children and young adults. It is characterized by a small, friable, erythematous papule on the skin or mucous membrane.[10] PGs rarely resolve spontaneously and have a tendency toward ulceration and easy bleeding with trauma.[10] Several treatment options exist for these lesions, including surgical (i.e., shave and destruction) and medical (i.e., topical timolol) intervention.[11–13] When considering the diagnosis of PG, it is particularly important to rule out more serious disorders, such as amelanotic melanoma, spitz nevi, squamous cell carcinoma, basal cell carcinoma, or Kaposi sarcoma.

Kaposiform hemangioendotheliomas and tufted angiomas. KHE and tufted angiomas are two types of locally aggressive vascular tumors that are believed to exist along a spectrum.[14] KHEs and tufted angiomas may be present at birth or develop over time. They typically present as ill-defined subcutaneous erythematous to violaceous firm nodules and may be associated with overlying telangiectasias, hypertrichosis, or hyperhidrosis.[15] Tufted



Figures 3A–3C. (A) Arteriovenous malformation of the right lower cheek, (B–C) Opacification and early venous (i.e., "fast") filling of draining veins in an arteriovenous malformation on dynamic time-enhanced MRA.

angiomas tend to be more superficial and thus may be easier to diagnose clinically than KHEs. It is particularly important to identify and treat these lesions early because they may be associated with the Kasabach-Merritt phenomenon, which often presents as acute enlargement of a pre-existing lesion. This association results from platelet trapping within the tumor, leading to profound thrombocytopenia and consumptive coagulopathy.[15]

Angiosarcoma. Angiosarcoma is a rare malignant vascular tumor that arises from endothelial-type cells in vascular and lymphatic vessel walls.[16] It can arise in the skin, subcutaneous soft tissue, or, more rarely, in bones, but is typically found predominantly in the head and neck.[17] Angiosarcomas are very rare in children, but they are among the most common tumors found following therapeutic radiation. They may also be a complication of breast cancer surgery and long-standing lymphedema in the breast or arm, a phenomenon known as "Stewart-Treves syndrome."[18] Angiosarcomas of the skin or subcutaneous tissue present as violaceous macules or nodules with indistinct borders on clinical exam. They may also be associated with hemorrhage, ulceration, or localized pain and tenderness.[17] Because of the malignant properties and propensity for metastasis of angiosarcomas, early identification and treatment are crucial.

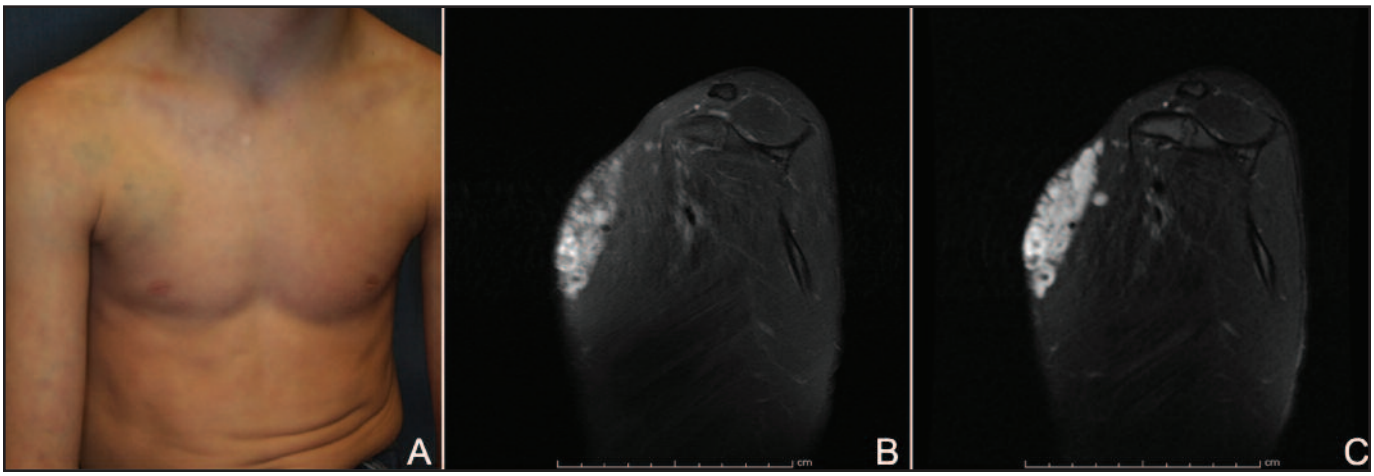
Epithelioid hemangioendothelioma. Epithelioid hemangioendothelioma (EHE) is a rare malignant vascular tumor that arises from endothelial-type cells present in vessel walls.[19] It follows a relatively indolent course with clinical behavior somewhere between that of hemangiomas and angiosarcomas.[20] It is commonly found in the soft tissue, but has also been found in skin, bone, and various organs.[19,21] EHE of the skin or subcutaneous tissue present as erythematous papules, nodules, or plaques, but may also be associated with painful ulceration.[19] Due to the potential for metastases, early identification is crucial.

Vascular tumors are typically easy to identify based on clinical presentation alone. However, in large or complicated lesions, the use of imaging can help better define the true nature of the lesion and facilitate early and effective management.

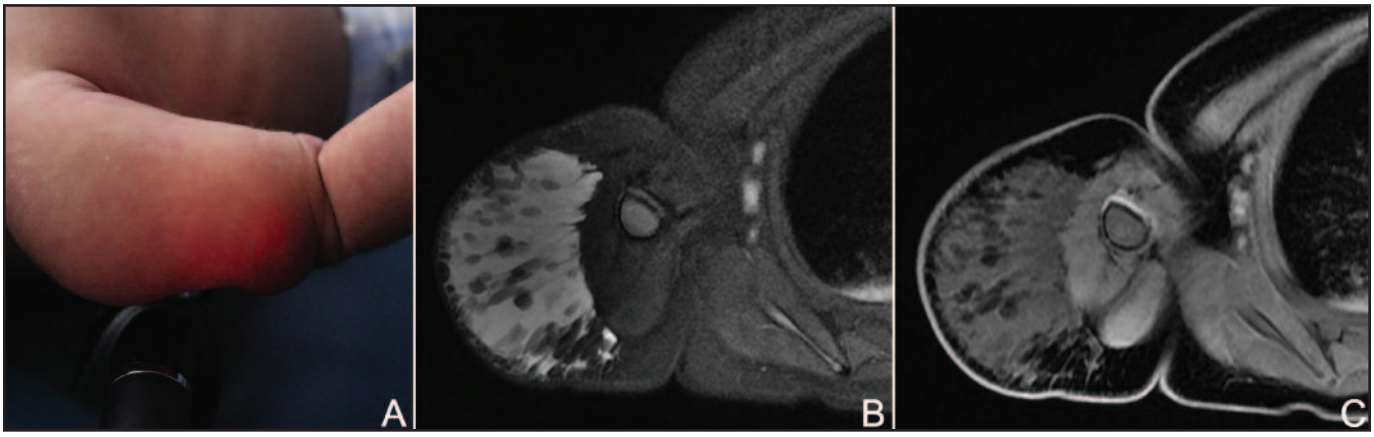
"HIGH FLOW" VERSUS "LOW FLOW" LESIONS

In 1993, Jackson et al[22] developed a radiologic classification for vascular anomalies to help improve categorization and treatment of these lesions. The characteristics of flow are particularly important this schema, useful in determining the specific type of vascular malformation present, and can generally distinguish between arterial, venous, and lymphatic lesions (see discussion below). Under the radiologic classification system, vascular anomalies are divided into "high flow" or "low flow" groups based on their flow dynamics. This classification system is generally capable of providing a sense of whether a lesion is predominantly arterial, venous, or lymphatic in nature. This information is particularly important in devising an appropriate treatment plan for specific lesions, as flow often determines the likelihood of response to a particular therapy.

High flow lesions. High flow lesions contain an arterial component. Examples include hemangiomas, arteriovenous malformations (AVM), and arteriovenous fistulas (AVF) (Figure 3A); these lesions often exhibit "flow voids" on magnetic resonance imaging (MRI), characterized by low signal intensity in vessels containing rapidly flowing blood. Hemangiomas, although composed predominantly of smaller endothelial-like tissue and structures, also contain prominent arteries in addition to veins leading to high flow characteristics on ultrasound (US) and MRI.[23] An AVM is characterized by an aberrant connection between arteries and veins. AVMs are typically present at birth, with estimated prevalence ranging between 5 and 613 per 100,000 persons.[24] AVMs can occur at any body site. When involving the skin or subcutaneous tissue, an AVM can



Figures 4A–4C. (A) Venous malformation of the anterior right chest, (B) Hypointense septated venous malformation on T1-weighted imaging, (C) Hyperintense septated venous malformation on T2-weighted imaging



Figures 5A–5C. (A) Lymphatic malformation of the right upper extremity with transillumination, (B) Multilobulated cystic hypointense lymphatic malformation on T1-weighted imaging, (C) Multilobulated cystic hyperintense lymphatic malformation on T2-weighted imaging

present with localized swelling, discoloration ranging from a slight blush to a deep violaceous color, and in some circumstances a pulsatile mass.[25] Late-stage, untreated AVMs can lead to distal ischemia, pain, ulceration, soft tissue and bony changes, bleeding, and potentially high-output cardiac failure.[25] An AVF is formed through an aberrant connection between an artery and vein. It can arise congenitally, iatrogenically when fistulas are created for dialysis, or traumatically. Clinically, AVF localized in the skin or subcutaneous tissue can lead to swelling, erythematous to violaceous discoloration, and a bruit and palpable thrill. Although typically benign, AVF can cause increased cardiac preload leading to hypotension, fatigue, and eventually heart failure.[26] Therefore, early and aggressive treatment of high flow lesions is particularly important.

Low flow lesions. Low flow lesions are present in approximately one percent of the population and encompass all other lesions that do not contain an arterial component including capillary, lymphatic, and venous malformations.[27] Low flow vascular malformations are typically present at birth, but they can arise over time as well.[28] These lesions may be composed of capillaries, lymphatics, veins, or any combination of these components. Therefore, the clinical presentation often varies widely. Venous malformations (VM) (Figure 4A) or lymphatic malformations (LMs) (Figure 5A) are fairly common and, when present in the skin or subcutaneous tissue, appear as bluish soft tissue masses that may increase in size with valsalva maneuver (i.e., crying, defecating, etc.) or when placed in gravity-dependent positions.[28] VMs can present in two different ways: cavitary or dysplastic lesions, with cavitary being the more common of the two. LMs may be macrocystic or microcystic in nature. Microcystic LMs consist of lesions containing multiple cysts less than 2mm in size, whereas macrocystic lesions contain cysts larger than 2mm.[29] Although low flow vascular malformations are generally less dangerous than high flow lesions, the former are often accompanied by pain or discomfort attributed to localized swelling.[30] Identification and treatment of low flow lesions are also important in improving patient quality of life.

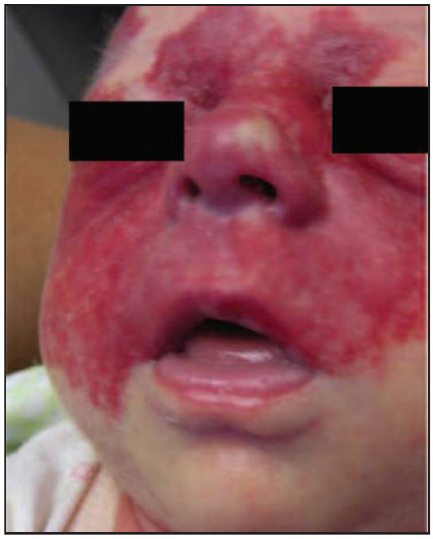


Figure 6. Large facial hemangioma in an infant with PHACES syndrome

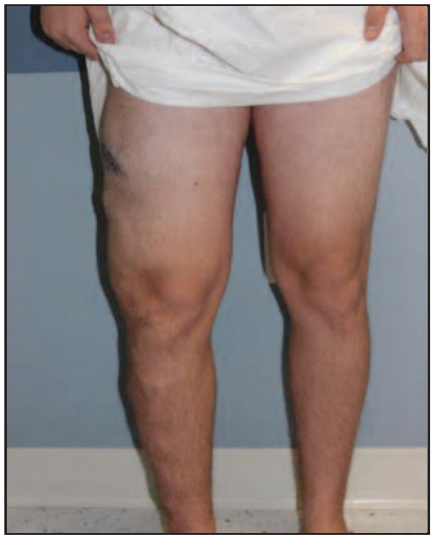


Figure 7. Unilateral vascular malformation with soft tissue and bony hypertrophy leading to leg length discrepancy in a patient with Klippel-Trenaunay syndrome

SYNDROMES ASSOCIATED WITH VASCULAR ANOMALIES

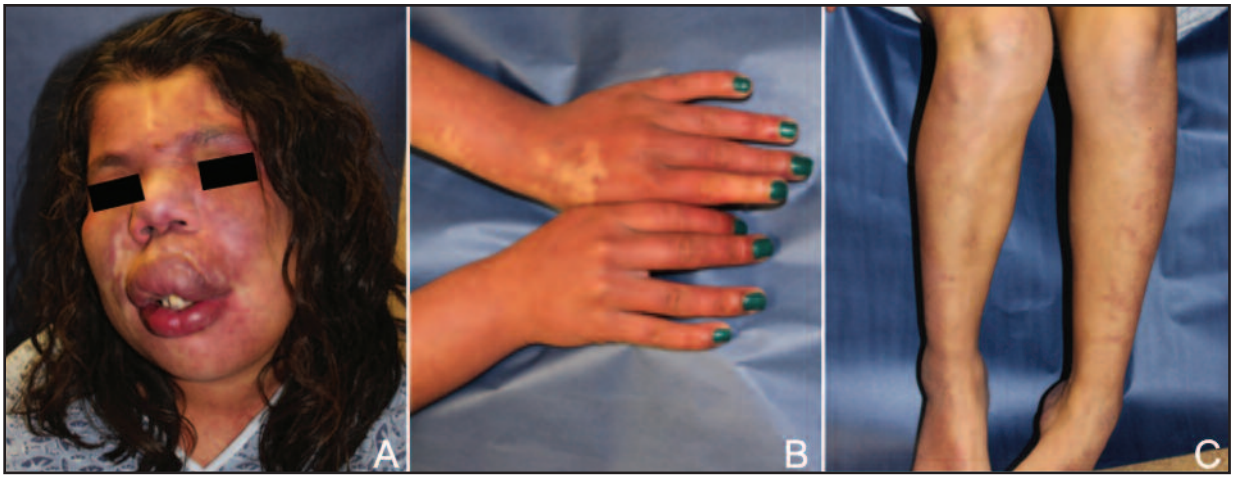
When evaluating a child with a vascular anomaly, it is important to evaluate patients for syndromes associated with specific vascular lesions. Although rare, these conditions may be a potential source of morbidity or mortality for patients, and early identification may lead to better clinical outcomes.

PHACE(S) syndrome is an association of findings characterized by posterior fossa brain malformations, hemangiomas, arterial anomalies, cardiac anomalies, and eye or endocrine abnormalities. Many experts also include "S" to represent sternal clefts and supraumbilical raphes, ventral closure defects, which can also be noted with this disorder. Because of its potential for multiple organ involvement, it is important to diagnose PHACES syndrome early so that infants can undergo careful cardiology, ophthalmology, and neurology exams. It is particularly important to consider this diagnosis in infants with large, segmental facial hemangiomas, as this distribution has been frequently reported in infants with PHACES syndrome (Figure 6).[31,32]

Klippel-Trenaunay syndrome (KTS) is a syndrome that was once thought to be characterized by the triad of 1) port-wine stains; 2) capillary, venous, or lymphatic malformations; and 3) soft tissue or bony hypertrophy that is often associated with limb length discrepancies (Figure 7).[33] Although patients with KTS may typically present with all three manifestations, only two of these three features are necessary for diagnosis. Patients with KTS may frequently present with pain associated with localized swelling from the vascular malformations and significant psychosocial morbidity.[30] Additionally, a higher prevalence of deep vein thromboses and pulmonary emboli has been reported in patients with KTS.[33] Therefore, early identification and monitoring of these patients may help decrease associated comorbidities. In many cases, KTS is used as an all-encompassing term to describe conditions in which port wine stains, vascular malformations, and/or overgrowth are seen, and some experts recommend describing these lesions based on the actual malformations present (e.g., CM vs. VM), rather than using what some view as a "waste-basket" term. Recent evidence suggests that although the majority of those syndromes categorized as KTS arise from PIK3CA mutations, many others may arise from a variety of different mutations.[34] The pathogenesis by which these patients develop vascular lesions and overgrowth may be different, and they may represent unique entities that should perhaps not be grouped together as a single disorder.

Sturge-Weber syndrome (SWS) is a syndrome characterized by capillary malformations arising in multiple tissues (skin, brain, eyes) leading to port wine stains, brain abnormalities such as seizures or mental retardation, and eye abnormalities such as glaucoma (Figures 8A–8C). One potential cause of SWS is thought to be related to a somatic mosaic mutation in GNAQ, a gene involved in regulating intracellular signaling pathways.[35] A mutation in this gene has been found in isolated PWS, as well as affected skin and CNS tissue in patients with SWS. Some have hypothesized that it is the point in fetal development when the mutation occurs that determines extent of involvement, with earlier mutations leading to more significant multi-organ abnormalities. It has been noted that SWS is also more likely to occur if infants have a PWS of the face involving the V1 trigeminal nerve distribution. Recently it has been suggested that forehead involvement may be the most important risk factor, rather than V1 distribution *per se*.[36–38] Because of the significant neurologic and ophthalmologic consequences of SWS, it is important to evaluate any child who has a PWS in a "high risk" area.[36–38]

There are a number of other syndromes associated with vascular anomalies that are best evaluated and managed by a multidisciplinary team. These patients often first present



Figures 8A–8C. Diffuse capillary malformations and deformities in a patient with Sturge-Weber syndrome

to their primary care doctors or to dermatology for skin findings. Therefore, it is particularly important for these "front-line" providers to recognize common presenting signs associated with these syndromes.

IMAGING OPTIONS FOR VASCULAR ANOMALIES

The majority of vascular anomalies can be diagnosed by history and clinical presentation alone. However, imaging can be useful to characterize and plan treatment for more complicated lesions. Several imaging options are available for this purpose (Table 2).

Radiographs. Radiographs, or x-rays, have poor soft tissue contrast resolution and confer some degree of ionizing radiation to the patient. As such, they are not commonly used for general imaging of vascular anomalies. Calcified phleboliths, however, may be identified on x-ray and are considered a hallmark of venous malformations. Bony remodeling may also be detected using conventional radiograph.[39] However, radiographs alone are rarely sufficient to provide adequate information to properly characterize a vascular anomaly. There are no absolute contraindications for the use of radiography. However, given the risk of ionizing radiation, it is not used in pregnant women unless there is an emergent indication. Urine beta-HCG testing is usually performed prior to routine radiographs in any female of child-bearing potential.

Computed tomography. Computed tomography (CT) is a technique that uses ionizing radiation to create a multi-dimensional image of the body. CT offers improved spatial resolution and detailed cross-sectional evaluation of the involved anatomy compared to conventional radiography. This technique is often useful in emergent situations as image acquisition time is usually only a few seconds. However, like radiographs, they have a limited role in the imaging work up for vascular anomalies. Furthermore, given the degree of ionizing radiation produced by CT, this technology must be judiciously used in the vulnerable pediatric population.

Vascular anomalies are typically visible on CT, but often appear as nonspecific masses.[40] CT with contrast (non-ionic iodinated contrast) may be more useful for characterizing vascular lesions because the addition of radio-opaque contrast allows mapping of the different arteries and veins within a lesion; however, this advantage is limited by the absence of temporal resolution in that all images are obtained at one point in time. CTs are generally considered safe procedures. However, they are associated with significantly increased exposure to ionizing radiation compared to conventional radiography. Therefore the use of CT is contraindicated in pregnant women. Urine beta-HCG testing is always performed prior to the use of CT unless it needs to be performed emergently, and if imaging is required, US and MRI would be the preferred techniques. Additionally, CT with contrast is relatively contraindicated in patients with a contrast allergy, renal impairment, hyperthyroidism, or a pheochromocytoma due to the risk of anaphylaxis, contrast-induced nephropathy, thyrotoxic crisis, and hypertensive crisis, respectively.[41–44] Relative contraindications in children in particular include syndromes with increased susceptibility to radiation, such as ataxia-telangiectasia, basal cell nevus syndrome, and Nijmegen breakage syndrome.[45]

Ultrasound. In many cases, US may be the initial screening method for patients with suspected vascular anomalies. It is readily available, inexpensive, safe, and imaging can usually be obtained without the need for sedation. US is particularly useful for evaluating superficial vascular anomalies. Basic grey-scale US provides excellent soft tissue contrast and an assessment of the involved anatomy. Doppler US evaluation is useful to determine vascularity and flow dynamics in a particular anomaly and may even be able to determine the type of vessels present.[28,46] Doppler US uses sound waves to measure whether blood is moving toward or away from the probe and the relative speed at which it is moving. The information can then be converted into colors that are overlaid over the

TABLE 2. Comparison of imaging modalities

Imaging Modality	Description	Benefits	Risks
Radiograph	A technique that uses electromagnetic radiation to create an image of the internal structures of the body	• Low cost • Fast • Can detect phleboliths or bony remodeling	• Ionizing radiation • Provides very little useful information
CT Scan	A technique that uses ionizing radiation to create a multi-dimensional image of the body	• Better spatial resolution compared to radiographs • Fast	• Ionizing radiation • Provides little useful information • Cannot be used in children with syndromes that cause increased susceptibility to radiation
Ultrasound	An imaging device that uses sound waves to visualize internal structures of the body. Doppler US uses sound waves to measure the direction and speed at which blood is flowing	• Readily available • Low cost • Safe • Sedation is not needed • Good soft tissue contrast • Can determine vascularity and flow dynamics	• Limited utility for examining deeper anatomy • Inferior to MRI and MRA in evaluating extent of a lesion or muscle/bone involvement
MRI	An imaging technique that uses magnetic fields and radio waves to form images of internal structures in the body	• Superior spatial resolution • Can determine vascularity and flow dynamics • No ionizing radiation • Can run several sequences at the same time	• Expensive • Time consuming • May require sedation in infants and children • Cannot be used in the presence of implanted devices
MRA	A specially tailored MR sequence that uses magnetic fields and radio waves to focus on imaging blood vessels	• Safer than conventional angiography • Good spatial and temporal resolution • Can determine vascularity and flow dynamics	• Expensive • May not be available in all centers • Cannot be used in patients with implanted devices
Dynamic time-resolved MRA	A specific type of contrast enhanced MRA that acquires images every few seconds allowing for improved temporal resolution and evaluation of flow dynamics	• Improved temporal resolution • Can determine vascularity and flow dynamics • Shorter procedure time and decreased contrast material used compared to traditional MRI or angiography	• Expensive • May not be available in all centers • Cannot be used in patients with implanted devices
Angiography	Technique that uses intravascular contrast injected in close proximity to the anomaly and fluoroscopy to visualize blood vessels	• Good visualization of blood vessels	• Invasive

standard US image of blood vessels to determine the flow dynamics, or direction and speed of blood flow. The evaluation of flow dynamics is useful for categorizing vascular anomalies into two groups: high flow and low flow lesions. Ultrasound may have limited utility for the evaluation of deeper vascular lesions and is often suboptimal in evaluating the full extent of a vascular lesion including potential muscle or bone involvement. Despite concerns about the frequent use of 3D and 4D US technology for fetal imaging, US is generally considered the safest imaging modality, and there are no absolute contraindications for its use.[47]

Magnetic resonance imaging. MRI is another commonly used imaging modality for the identification and characterization of vascular lesions. MRI provides superior spatial contrast resolution compared to radiograph or CT allowing detailed soft tissue evaluation.[48] Additionally, like US, MRI can be used to assess the flow dynamics within a vascular lesion. High flow lesions on MRI are often characterized by the presence of flow voids (i.e., areas

emitting no radiofrequency signal).[49] Occasionally, a thrombus or phlebolith can mimic a flow void on MRI in low flow lesions.[49]

Many pediatric providers prefer MRI because it does not expose young patients to ionizing radiation. Due to the lack of ionization radiation, multiple sequences including T1-weighted, T2-weighted, pre- and post-contrast images, and fat saturation can be run in a single exam, each of which is obtained at a different point in time. T1 and T2 are two distinct tissue relaxation constants or characteristics that are used with MRI. In T1-weighted imaging, fat appears bright (hyperintense) and water appears darker (hypointense). Complex fluid containing blood products and proteinaceous debris are typically brighter on T1 and darker on T2, which is the reason fluid-fluid levels are visible. In T2-weighted imaging, both fat and water appear bright. Fat suppression makes fat dark, which emphasizes fluid signal on T2-weighted images and enhancement on post-contrast T1-weighted images.[29] T1-weighted images are typically used for post-contrast imaging. After contrast, vascular tissue becomes brighter indicating enhancement. STIR (short tau inversion recovery) images are a type of T2-weighted image with fat saturation, and are considered a fluid sensitive sequence. Since most pathology, including vascular anomalies, has increased fluid, these sequences are the most helpful in lesion detection. T1-weighted imaging has excellent spatial resolution. It is best for evaluation of anatomical structures and tissue planes within a vascular anomaly, and it may be particularly useful for characterizing the extent and anatomy of lesions if used before and after gadolinium administration.[28]

Together, T2-weighted imaging with fat suppression (or STIR) and post-contrast T1-weighted imaging are considered the most useful sequences for lesion detection and to demonstrate the full extent of vascular anomalies and their relationship with adjacent anatomical structures.[50,51] Contraindications for MRI include the presence of implanted devices, such as cardiac pacemakers, defibrillators, metal parts or wires, pregnancy (it may however be used when a fetus is suspected of having a vascular anomaly), unstable patients, and other concerns, such as claustrophobia or very obese patients.[52,53] Additionally, MRI with contrast agents should not be used in patients with renal failure.[52,53] However, for most of these contraindications, MRI may still be performed if the benefits outweigh the risks.

Magnetic resonance angiography and dynamic time-resolved magnetic resonance angiography. Magnetic resonance angiography (MRA) is a noninvasive technique that offers a safer and more efficient alternative to conventional angiography. This technique is a specifically tailored MR sequence that focuses on imaging blood vessels. MRA may be obtained with or without gadolinium contrast. MRA is a useful—and often necessary—tool in the initial evaluation of vascular anomalies because it allows for detailed spatial and temporal analysis of the vessels within an anomaly. Standard contrast-enhanced MRA provides very good spatial resolution of a vascular anomaly, taking images of a particular lesion approximately every 15 seconds.[29] This is superior to conventional pre-contrast and post-contrast MRI sequences because it can be used to specifically evaluate blood vessels within a vascular lesion.

Dynamic time-resolved MRA is a technique that is uniquely useful for the imaging workup of vascular lesions. Images are acquired every few seconds allowing for improved temporal resolution.[29] Dynamic time-resolved MRA samples lower spatial frequencies significantly more than higher spatial frequencies and fill in missing data using a specialized algorithm leading to improved temporal resolution.[54,55] The excellent temporal resolution allows assessment of flow dynamics through the arterial, capillary, and venous phases of a lesion and the feeding and draining vessels. This technique therefore permits assessment of flow dynamics and identification of high flow and low flow characteristics of a lesion, which are not possible with conventional MRI or MRA.[54,56] As a result, dynamic time-resolved MRA is the most useful MR sequence for evaluating the flow characteristics within a lesion, distinguishing high flow versus low flow lesions. It also provides other advantages over traditional imaging techniques including decreased procedure time and decreased contrast material used. It is now considered the gold standard for evaluating flow dynamics within a vascular anomaly.[54] Therefore, for the purposes of this discussion, all references to MRA in this paper are made with regard to their appearance on dynamic time-resolved MR angiography. Contraindications for MRA are similar to that of MRI and include the presence of implanted devices, such as cardiac pacemakers, defibrillators, metal parts or wires, pregnancy, unstable patients, and other concerns, such as claustrophobia or very obese patients.[57] Gadolinium should not be used in patients with renal failure or those who are pregnant.[57] However, MRA may still be performed if the benefits outweigh the risks, and with appropriate informed consent from the patient.

Angiography. Catheter angiography is an imaging technique that uses intravascular contrast injected in close proximity to the anomaly and fluoroscopy to visualize blood vessels, making it particularly useful to analyze the vessels in high flow vascular malformations. Angiography is not typically used for initial workup of a vascular anomaly because of its invasive nature. Angiography is typically reserved for confirmation of suspected AVMs or used during imaged guided intervention for these vascular lesions.[58]

APPROPRIATE UTILIZATION OF IMAGING

There are a wide variety of imaging modalities available for evaluating a vascular anomaly. However, imaging can be time consuming, expensive, and potentially harmful to the patient. Therefore, imaging should only be used if it will actually be helpful in the diagnosis or management of a vascular lesion.

As discussed previously, the most commonly utilized modalities for the workup of vascular anomalies include US, MRI, and dynamic time-resolved MRA. MRI, usually in conjunction with MRA, is commonly used when US proves insufficient for diagnosis or when more detailed evaluation is

necessary to plan treatment. However, in clinical cases that suggest complicated vascular anatomy or the likely need for further workup, MRI and MRA may be the more appropriate first-line tools for the initial evaluation of a vascular anomaly.

Benign vascular tumors. ***Infantile hemangiomas.*** Most superficial IHs are easy to diagnose based on clinical appearance alone. However, imaging may sometimes be employed for the evaluation of deeper or atypical lesions. Because IH are dynamic lesions that progress through both proliferative and involution phases, they can have variable presentations on histopathology as well as imaging, depending on their clinical stage.

US with Doppler—On US, IHs are typically well-circumscribed hyper- or hypo-echoic masses with internal vascularity demonstrating low-resistance arterial waveforms.[51,59]

MRI—MRI appearance can be affected by the clinical stage of the IH. During the proliferative phase, IHs are typically well-defined lobulated masses that are hypo- or iso-intense on T1-weighted imaging and uniformly hyperintense on T2-weighted imaging.[23,29] On gadolinium contrast-enhanced MRI, proliferating IHs are characterized by early, diffuse, and uniform enhancement with flow voids (Figure 1B).[23,50] During the involution phase, the decreased arterial blood flow and fat replacement makes the appearance of IHs on MRI more heterogeneous. On T1-weighted imaging, foci of increased signal intensity within the lesion may be found.[23,29]

MRA—High flow tumors typically present as rapid, mass-like enhancement on dynamic time-enhanced MRA, sometimes with early wash out (Figure 1C).

Congenital hemangiomas. CHs, like IHs, can usually be diagnosed based on clinical appearance alone. Imaging may sometimes be used for the evaluation of complicated or atypical lesions. Although CHs comprise two different subtypes, RICH and NICH, they typically present almost identically on imaging.[60]

US with Doppler—Unlike IHs, CHs are typically ill-defined hyper- or hypo-echoic masses on US. Vascular aneurysms, thrombi, and arteriovenous shunting, properties usually not seen in IHs, may be apparent as well.[51,60] However, these findings are generally more prominent on MRI.

MRI—MRI of CHs is very similar to that of IHs, presenting as ill-defined lobulated masses that are hypo- or iso-intense on T1-weighted imaging and hyperintense on T2-weighted imaging (Figure 2B).[23,29] Vascular aneurysms, thrombi, arteriovenous shunting, and large flow voids with evidence of fat stranding may also be visible in MRIs of CHs.[29,40,60]

MRA—High flow tumors typically present as rapid, mass-like enhancement on dynamic time-enhanced MRA, sometimes with early wash out (Figure 2C).

Tufted angioma and kaposiform hemangioendotheliomas. Tufted angiomas and KHEs are considered to be similar vascular anomalies that exist along the same spectrum.[14] Compared to IHs and CHs, tufted angiomas and KHEs are more aggressive, rarely regress without treatment, and may be associated with Kasabach-Merritt phenomenon, making the need for imaging more important in these lesions.[14,23] Because tufted angiomas are generally more superficial in nature, they are often diagnosed clinically and may not require imaging as often as KHE.[23]

US with Doppler—Tufted angiomas and KHEs, like CHs, present as ill-defined masses on US. However, they usually present with variable echogenicity and may show signs of calcifications as well. Vessel density is variable in different lesions and high resistance waveforms may be seen on Doppler examination.[46]

MRI—Like other vascular tumors, tufted angiomas and KHE appear as hypo- or iso-intense ill-defined masses on T1-weighted imaging and hyperintense on T2-weighted imaging.[51] They may also demonstrate involvement of multiple tissue planes, subcutaneous fat stranding, hemosiderin deposits, and adjacent bone destruction or remodeling.[23,29,50] On contrast-enhanced MRI, diffuse, heterogeneous enhancement may be seen, which is arterial phase on MRA.[51]

MRA—Similar to other vascular tumors, such as infantile hemangiomas, these high flow tumors typically present as rapid, mass-like enhancement on dynamic time-enhanced MRA. Therefore, imaging distinction between these and other vascular tumors often relies more heavily on the characteristic seen on conventional MRI sequences, as described above.

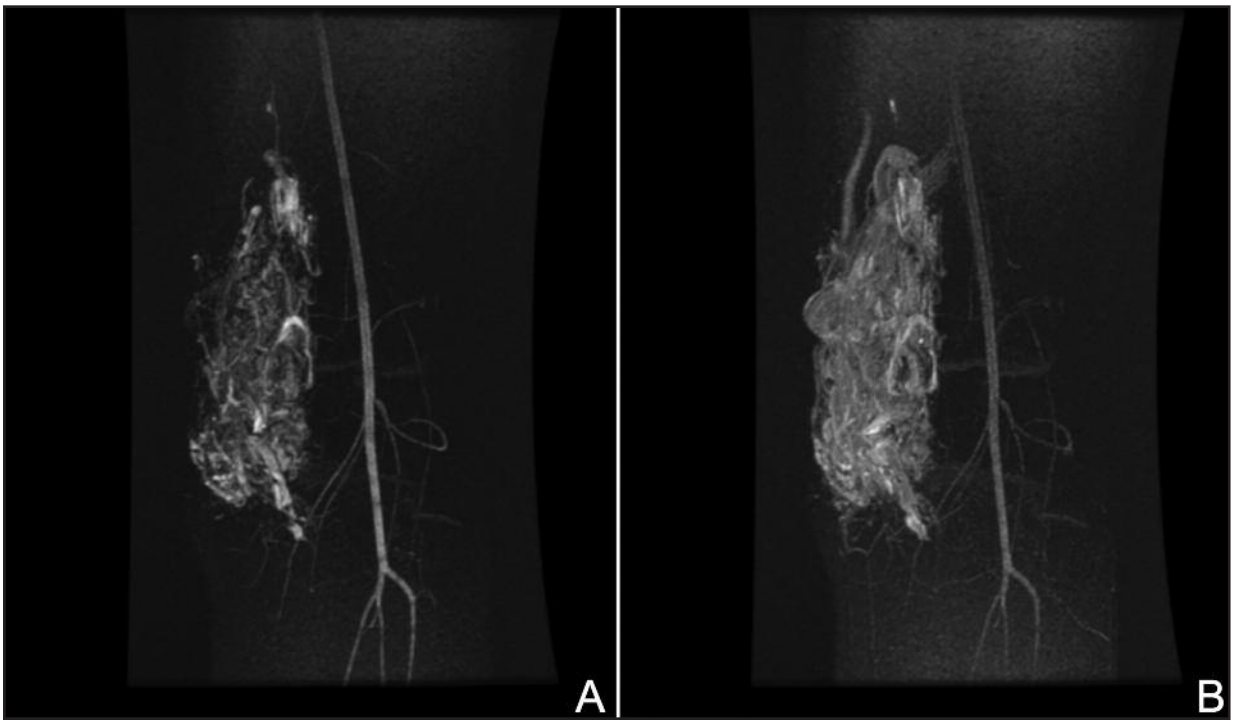
Malignant vascular tumors. ***Angiosarcoma.*** Early diagnosis of angiosarcomas is critical due to their malignant potential. Angiosarcomas have similar imaging features to tufted angiomas and KHEs.[16] Therefore, it is important to be able to recognize clinical and radiographic features that are unique to angiosarcomas, and histopathologic evaluation is invariably required.

US with Doppler—Angiosarcomas appear as irregular hypo-, iso-, or hyperechoic lesions with hyper-vascularization.[20]

MRI—On MRI, angiosarcomas are masses that are isointense on T1-weighted imaging and hyperintense on T2-weighted imaging, with diffuse, robust enhancement on gadolinium administration.[16,61] Because these lesions are prone to hemorrhage and ulceration, evidence of bleeding may be visible as areas of hyperintense signal intensity within the lesion on T1-weighted imaging. However, many of these findings may also be present in benign vascular conditions, making discrimination difficult. A common and helpful feature found in angiosarcomas on MRI is the presence of high flow serpentine vessels within the soft tissue mass.[16]

MRA—Similar to other vascular tumors, such as infantile hemangiomas, these high flow tumors typically present as rapid, mass-like enhancement on dynamic time-enhanced MRA. Therefore, imaging distinction between these and other vascular tumors often relies more heavily on the characteristic seen on conventional MRI sequences, as described above.

Vascular malformations. ***Arteriovenous malformation.*** Arteriovenous malformations (AVMs) are high flow vascular malformations that can often be associated with acute enlargement, overgrowth, hemorrhage, ulceration,



Figures 9A–9B. Slow, progressive enhancement in a venous malformation on dynamic time-enhanced MRA

and high-output heart failure.[23] For this reason, imaging is crucial to determine size, extent, and relationship to adjacent structures and plan treatment.

US with Doppler—On US, AVMs are ill-defined lesions consisting of multiple feeding arteries and draining veins with pulsatile flow. Doppler imaging typically shows high velocity flow with arterialization of the draining veins.[23]

MRI—AVMs are characterized by enlarged high flow feeding arteries and draining veins surrounding areas of large flow voids without the appearance of a well-defined mass.[29] Signal voids can be observed on both T1- and T2-weighted imaging. Areas of hyperintense signaling on T1-imaging correspond to areas of hemorrhage or thrombosis.[29] On gadolinium contrast-enhanced MRI, further evaluation of the feeding arteries and draining veins may be performed.

MRA—On MRA, MRA is extremely useful in the evaluation and treatment planning for AVMs. There is rapid enhancement of a tangle of vessels, often with early enhancement of the dominant draining vein. The dynamic opacification of the nidus and early venous filling of the draining veins in AVMs can often be well-visualized with this imaging modality (Figure 3B–3C).[29]

Venous malformation. Venous malformations (VMs) are low flow lesions that are predominantly found in the head, neck, and extremities.[23,39] They are the most commonly encountered vascular malformation.[39] Imaging of VMs is often performed to determine size, extent, and relationship to adjacent structures and to plan treatment VMs can present in two different ways: cavitary or dysplastic lesions.

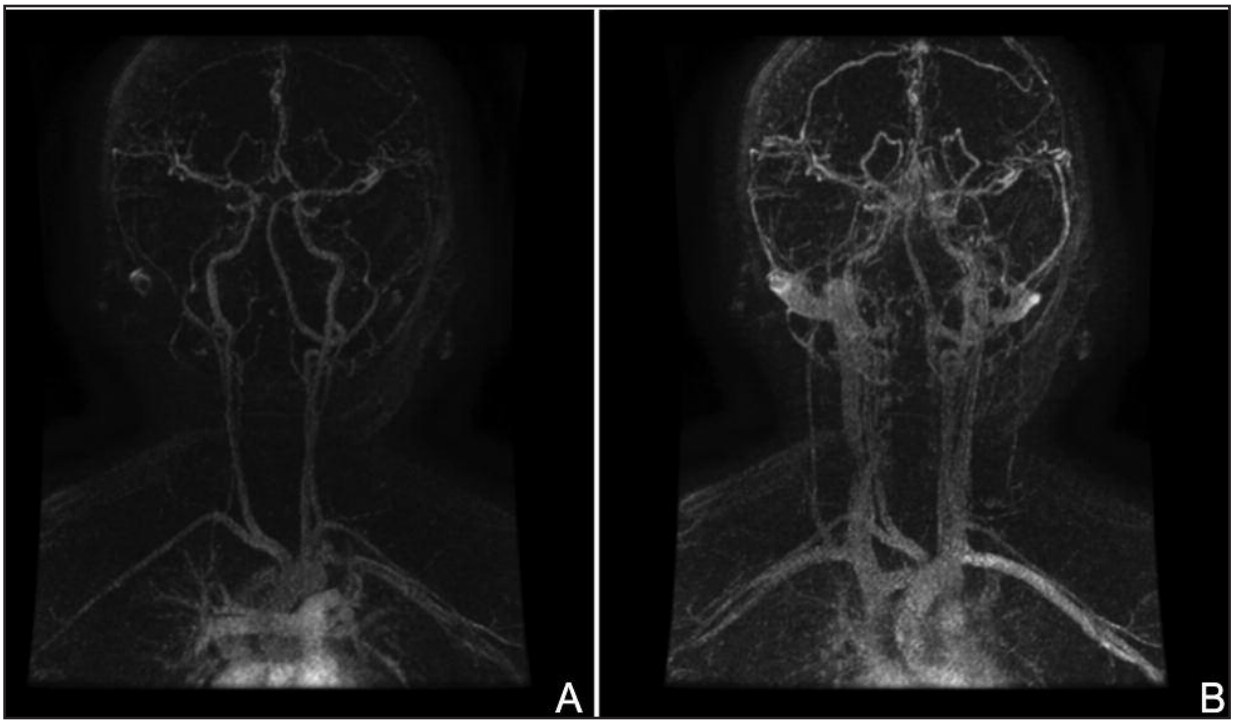
US with Doppler—On US, cavitary VMs appear as hypoechoic, heterogeneous lesions and dysplastic VMs usually demonstrate multiple anechoic, tortuous venous channels.[63] Both types of VMs, however, may have infiltration of the subcutaneous fat, muscles, or fascia.[51,63] On Doppler imaging, monophasic low-velocity flow is common. However, increased flow velocity and the presence of phleboliths may be observed with valsalva maneuvers or manual compression.[63]

MRI—VMs present as hypo- to isointense septated lesions on T1-weighted imaging and hyperintense on T2-weighted imaging (Figures 4B–4C) .[23,29] Rarely, visible fluid-fluid levels or heterogeneous signal intensity on T1-weighted imaging may indicate hemorrhage or thrombosis.[29] Phleboliths are common occurrences in VMs and appear as small foci of signal void on all sequences.[23] On gadolinium contrast-enhanced MRI, VMs demonstrate diffuse or heterogeneous enhancement with small foci of signal voids indicating the presence of phleboliths or thrombi.[23]

MRA—VMs demonstrate slow progressive enhancement throughout the course of the dynamic time-enhanced MRA acquisition compared to similar-appearing high flow lesions (Figures 9A–9B). This is one of the most useful applications of this technology.

Lymphatic malformation. Lymphatic malformations (LMs) are low flow lesions that are commonly confused with other vascular malformations. Imaging of LMs is often performed to confirm the diagnosis; determine size, extent, and relationship to adjacent structures; and plan treatment.

US with Doppler—LM appear as lobulated, septated cystic



Figures 10A–10B. No significant enhancement of a lymphatic malformation of the head on dynamic time-enhanced MRA

masses without internal vascularity on US. They are typically cystic and may appear diffusely hypo- to isoechoic.[62] However, the appearance on US is dependent on whether the LM is macrocystic or microcystic. Cystic masses are usually hypoechoic. However, if the cysts contain protein or hemorrhage, they can appear iso- or hyperechoic. Septations and fluid levels may also be visible.

MRI—MRI may demonstrate LMs infiltrating surrounding tissue or involving multiple tissue planes. LMs appear as multilobulated cystic masses that are hypo- to isointense on T1-weighted imaging and hyperintense on T2-weighted imaging on MRI (Figures 5B–5C).[23,29] However, LMs may also present as hyperintense masses on T1-weighted imaging if they contain protein or hemorrhage. Because of the multilobulated nature of LMs, fluid-fluid levels may also be apparent on imaging.[29] On gadolinium contrast-enhanced MRI, the individual cysts do not demonstrate significant enhancement. However, the septa, which contain vascular elements, may show contrast enhancement.[23,29,62] Macrocystic lesions appear as multilobulated masses with fluid, fluid levels, and enhancement of the septa. Microcystic lesions, on the other hand, can appear as solid masses that show little to no enhancement on imaging.

MRA—LMs demonstrate no enhancement on MRA. However, sometimes when the lesions are large, LMs can lead to distortion of the normal arteries and veins due to localized mass effect (Figures 10A–10B).

FINANCIAL COSTS AND OTHER CONSIDERATIONS

Over the last several decades, there has been a significant rise in the use of imaging tools in medicine. The ongoing improvement of imaging technology has revolutionized the practice of medicine. Diagnostic imaging techniques have reduced the need for invasive procedures and have increased our understanding of several disease processes. However, the combination of increased utilization of imaging along with the use of more expensive technologies has increased the costs of imaging more than two-fold in a 10-year study period.[64] The reported government funded Medicare spending for imaging has more than doubled during the same time period.[64] In 2005, the mean cost for diagnostic radiology, CT, and MRI were $410, $1,565, and $2,048, respectively.[65] These exuberant costs can be a burden on the federal budget, insurance organizations, hospitals, healthcare providers, and patients, alike. These procedures may not always be cost-effective, and physicians must be able to determine when imaging is appropriate.

Another consideration that complicates the use of imaging is the ability to get "permission" to obtain coverage for the diagnostic procedure. Mandatory prior authorization procedures are an attempt on the part of insurance plans to ration use of procedures. Prior authorization is the practice in which health insurance companies can determine whether they will cover a prescribed medication, service, or procedure. Due to the significant rising cost of medical imaging, many insurance companies have implemented prior authorization policies for the use of certain imaging tools to combat overuse and unnecessary costs. Many insurance companies have contracted with Radiology Benefit Managers (RBMs) or companies that determine the need for prior authorization for imaging services using

algorithms based on clinical guidelines and expert opinions.[66] With the introduction of prior authorization policies, the overuse rate of imaging procedures that require prior authorization has decreased, especially in comparison to imaging procedures not requiring prior authorization.[67] However, it is unclear whether prior authorization is beneficial or detrimental to patients and providers overall. It has been reported that primary care physicians and nursing staff spend approximately two hours and 13 hours a week, respectively, on prior authorizations.[68] When time is converted to dollars, it is unclear whether the use of prior authorizations is actually cost effective. Additionally, because of the time-consuming nature, the need for prior authorization may even be detrimental to patients because it may delay or prevent patients from obtaining imaging that is necessary for diagnosis and treatment. Therefore, it is important for physicians to understand the role of prior authorization on the use of imaging for pediatric vascular lesions.

A unique challenge facing the use of imaging in pediatric patients is that imaging procedures often require patients to remain still for a prolonged period of time in an enclosed space. For this reason, frequent use of sedation in the pediatric population is common, especially for more time-consuming or anxiety-inducing imaging procedures, such as CT or MRI. Although generally considered safe, the use of sedation in pediatric imaging studies comes with significant risks as well. Adverse events with sedation in pediatric patients, such as O_2 desaturation, central apnea or airway obstruction, stridor, laryngospasm, and the need for pulmonary or cardiopulmonary resuscitation have been reported with the use of imaging in the literature.[69,70] Additionally, pediatric patients may be more susceptible to prolonged recovery and delayed side effects, such as motor imbalance, GI effects, agitation, and restlessness after sedation for diagnostic imaging studies.[71] The risk of ionizing radiation associated with some imaging procedures in conjunction with the risks associated with the need for sedation exemplifies the particularly vulnerable position of pediatric patients.

Concerns have recently arisen regarding the potential risk of general anesthesia on developing neural tissue. Animal studies have identified neurocognitive deficits that can occur when young animals are exposed to general anesthesia, particularly if it is prolonged or occurs on multiple occasions.[72–75] There has been some suggestion that infants and children may be at risk for similar effects.[76–78] Further investigations are required to better define the true risk for humans, but until this issue is clarified, one must include this theoretical risk in decisions regarding the use of general anesthesia for infants and children. The use of awake "swaddle" techniques in very young infants is a welcome addition to efforts to perform imaging without the need for anesthesia.[79]

CONCLUSION

The care of patients with vascular anomalies can be quite challenging. A clear understanding of the appropriate means of evaluating such children, and the appropriate use of imaging techniques, is crucial. It is particularly important for those involved in front-line care of these patients to recognize vascular anomalies that require imaging and understand what type of imaging is appropriate based on the patient's clinical presentation. The increased use of imaging tools has brought with it increased cost to medical care, as well as some risks, particularly given the need for sedation in most young children. Therefore, risk-benefit of any procedure should always be considered when deciding on appropriate evaluation. Imaging, when used correctly, can be a useful, safe, and cost-effective tool for the characterization of vascular anomalies and for guiding optimal management.

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