In recent decades, a great deal has been learned about the histopathology, etiology, and treatment of vascular anomalies, which has caused change in the classification and terminology used to describe these lesions. The International Society for the Study of Vascular Anomalies (ISSVA) classification system, which has been widely embraced by various subspecialists who care for patients with these malformations, provides an approach based on histopathology, clinical course, and treatment. However, use of older nomenclature continues to cause confusion, inaccurate diagnoses, and potential mismanagement. The overarching goal of this article is 2-fold. First, to review the ISSVA classification of vascular anomalies by discussing key pathogenesis, imaging features, and modern treatment of representative lesions from each category; and second, to discuss terminology used to describe vascular malformations with the goal of clarifying dated or confusing terms that remain in use.

**International Society for the Study of Vascular Anomalies Classification**

In 1982, Mulliken and Glowacki\(^1\) proposed a binary classification system for vascular anomalies based on pathologic features. This system, which was adopted by the ISSVA, has since been expanded and is now widely accepted.\(^1\) The importance of the ISSVA system is that it allows a systematic approach to vascular lesions that correlates predictably with clinical history, disease course, and treatment options, making it clinically useful.\(^5\)

The ISSVA classification system divides vascular anomalies into 2 (binary) primary biological categories: (1) vasoproliferative or vascular neoplasms and (2) vascular malformations (Table 1).\(^2\) The major distinction between the 2 categories is whether there is increased endothelial cell turnover, which is ultimately determined by the identification of mitoses seen on histopathology.\(^1,2\) Vasoproliferative neoplasms have increased endothelial cell turnover (ie, they proliferate and undergo mitosis) because they are neoplasms.\(^3\) Vascular malformations do not have increased endothelial cell turnover. Instead, vascular malformations are structural abnormalities of the capillary, venous, lymphatic, and arterial system that grow in proportion to the child.\(^1,3\)

**Imaging Approach to Vascular Anomalies**

Many vascular anomalies can be diagnosed by history and physical examination, making imaging unnecessary. However, when imaging is used, it is important to choose the modality based on the specific lesion and clinical situation, rather than using a one-size-fits-all mentality. Ultrasonography (US) and magnetic resonance imaging (MRI) are the 2 most widely used modalities of choice. US is used for initial screening because of its portability, lack of ionizing radiation, and no requirement of sedation in children. It is relatively simple, noninvasive, and yields good results for evaluating small, superficial and/or suspected, solid visceral lesions. Typically, US is able to determine the basic type of lesion, direct initial management, and plan further imaging evaluation.\(^6,7\) For sonography to be a useful modality, it must include gray-scale, color Doppler, and spectral Doppler tracings to evaluate vascularity and determine types of vessels present.\(^7\) MRI is helpful to further characterize sonographic findings and determine the extent of larger lesions for planning medical, interventional, and/or surgical therapy.

Radiography has limited utility in evaluating vascular anomalies, but when obtained, it may show organomegaly, soft tissue masses, and/or phleboliths.
Phy (CT) is helpful when urgent imaging is required because of its speed and less frequent need for sedation.8

Terminology

In the past, the suffix “oma” was used to describe neoplasms and malformations. However, because malformations are not neoplasms, words such as “lymphangioma” are misnomers, and the terminology is being abandoned. The word “hemangioma” has been used to describe several lesions, which are now known to be distinct pathological entities.9 It is the current consensus that a descriptor is needed for the term “hemangioma” to be meaningful (ie, infantile or congenital).10

Infantile hemangiomas present between 2 weeks to 2 months of life, undergoing a proliferative growth phase until they reach their full size, whereas congenital hemangiomas are distinguished by being fully formed at birth.11,12 Hepatic infantile hemangiomas, which are pathologically the same as those located on the skin and other organs, have unfortunately been referred to as infantile hemangioendothelioma(s), erroneously suggesting they are distinct pathologically.10 Further, histological studies have shown that what was commonly referred to as “hemangioma” of the adult liver, “hemangioma” of the adult vertebra, and orbital “cavernous hemangioma” are actually venous malformations rather than neoplasms.13,14 See Table 2 for comparison of old versus new terms describing vascular anomalies.

Several studies demonstrate the utility of the current ISSVA terminology.9 One retrospective study of children evaluated in a vascular malformations clinic highlights the confusion surrounding terminology by showing that 69% of patients were initially given a wrong diagnosis, and 53% of parents were given incorrect information regarding lesion progression and treatment.15 Another study found that the term hemangioma was incorrectly used in 71.3% of manuscripts studied, regardless of the author’s discipline.16 The same study noted that patients with mislabeled lesions were more likely to receive improper treatment (20.6%) compared with those classified according to the ISSVA system (0%).16

Vascular or Vasoproliferative Neoplasms

Although a complete discussion of all permutations of vascular tumors included in the ISSVA classification system is beyond the scope of this manuscript, awareness of entities with overlapping terminology will prevent confusion. Infantile hemangiomas, congenital hemangiomas (rapidly involuting congenital hemangiomas [RICH] and noninvoluting congenital hemangiomas [NICH]), tufted angiomas (TA), kaposiform hemangioendotheliomas (KH), spindle cell and epithelioid hemangioendotheliomas, and angiosarcomas are discussed.

Recent Hypotheses and Observations Regarding the Pathogenesis of Hemangiomas

Evidence suggests that vasoproliferative tumors result from vasculogenesis (formation of primitive blood vessels from angioblasts) rather than from angiogenesis (the growth of vessels from preexisting vessels) as previously thought.17-19 Immature stem/progenitor cells are identified within the anomaly that may originate from the placenta.20-23 Studies have identified increased numbers of mast cells, commonly seen in involuting infantile hemangiomas undergoing apoptosis.20,24-26 During the involution phase, mesenchymal stem cells differentiate into adipocytes, whereas capillary lu-

| Table 1 International Society for the Study of Vascular Anomalies Classification System |
|---------------------------------|---------------------------------|
| Vascular (or vasoproliferative) Neoplasms | Vascular Malformations |
| Infantile hemangioma | Slow-flow vascular malformations |
| Congenital hemangiomas | Capillary malformation |
| RICH | Venous malformation |
| NICH | Lymphatic malformation |
| Kaposiform hemangioendothelioma and tufted angiomas (with or without Kasabach–Merritt syndrome) | Fast-flow vascular malformations |
| Spindle cell hemangioendothelioma | Arterial malformation |
| Epithelioid hemangioendotheliomas | Arteriovenous malformation |
| Other rare hemangioendotheliomas (ie, composite, retiform, and others) | Arteriovenous fistula |
| Angiosarcoma | Combined vascular malformations (various combination of the above) |
| Dermatologic acquired vascular tumors (ie, pyogenic granuloma) | |

RICH, rapidly involuting congenital hemangioma; NICH, noninvoluting congenital hemangioma.

| Table 2 Comparison of Previous Terminology and New ISSVA Terminology |
|-------------------------|-------------------------|
| Previous | ISSVA |
| Capillary or cavernous hemangioma of any organ(s) | Infantile hemangioma(s) |
| Infantile hemangioendothelioma(s) of the liver | Hepatic or infantile hemangioma(s) |
| Adult hepatic hemangioma | Venous malformation |
| Adult vertebral hemangioma | Venous malformation |
| Adult orbital cavernous hemangioma | Venous malformation |

mens undergo apoptosis, both of which lead to lesion resolution. Cellular proliferative markers, such as vascular endothelial growth factor (VEGF), are also elevated in proliferating, but not involuting, hemangiomas.

Unique to placental endothelial cells and infantile hemangiomas is the expression of glucose transporter isoform 1 (GLUT1) protein. GLUT1 is not expressed by any other normal tissue or vascular tumor. Placental cells and infantile hemangiomas appear to have the same life cycle, suggesting that hemangiomas may result from embolized placental cells.

Another recent observation is that some hemangiomas occur in a nonrandom, or regional, distribution. This may be from embryologic prominences, which raises the possibility of a neuroectodermal abnormality. One example of this is regional facial hemangiomas, which occur around embryologic fusion lines. The mesenchyme of the fetal head and neck is mostly neuromesenchyme, which favors hemangioblast migration, and may account for the common head and neck location of hemangiomas.

Cases of children with coexistent infantile and congenital (RICH or NICH) hemangiomas, as well as children with initial RICH that at some point have arrested involution, taking on the clinical appearance of a NICH, are reported. This suggests that NICH may be a later stage of RICH or possibly a transformation into RICH.

Somatic mutation with clonal expansion within a single endothelial cell may explain the fact that infantile hemangiomas are GLUT1 positive (GLUT1+) and congenital hemangiomas are not. Finally, extrinsic factors, ie, milieu of the fetus and/or infant, may play an unspecified role in the development of hemangiomas, which is likely multifactorial.

**Infantile Hemangioma**

Infantile hemangioma is the most common tumor of infancy and occurs in approximately 4%-10% of infants. It is most frequent in premature Caucasian females with low birth weight, who are products of multiple gestation and have undergone chorionic villous sampling. Maternal risk factors include advanced age, pre-eclampsia, and placental abnormalities. The increased risk of hemangiomas in populations undergoing procedures that may cause trophoblastic

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**Figure 1** Infantile hemangioma in a 4-month-old female patient with a lump on her neck. (A) Photograph of the back of the neck shows a smooth focal lump (arrows). (B) Axial fat-suppressed T1-weighted postcontrast image reveals a well-defined vigorously enhancing mass with internal flow void located in the subcutaneous fat (arrow).

**Figure 2** LUMBAR regional syndrome in a 4-week-old female patient with extensive cutaneous hemangiomas on the lower abdomen and legs, left renal agenesis, and a patent urachus. T1-weighted contrast-enhanced (A) axial and (B) sagittal fat-suppressed magnetic resonance (MR) images demonstrate multiple enhancing well-defined lobular masses consistent with hemangiomas (arrows). Abnormally low conus position (arrowhead) is also shown. L5-Fifth lumbar vertebra.
embolization and placental disruption adds support to the theory of placental origin of hemangiomas.\textsuperscript{21,41} Infantile hemangiomas typically present between 2 weeks and 2 months of life.\textsuperscript{12,40} They may be single or multiple, may involve 1 or many organ systems, and may be focal or regional (segmental). Regional hemangiomas occur along lines of embryological fusion and may be more frequent in Hispanics.\textsuperscript{42} Examples include midline glabella (nasal bridge), low-back, and facial hemangiomas. Infants with regional or with cutaneous hemangiomas should undergo screening US of the head and abdomen to check for additional lesions.\textsuperscript{10,43,44}

Although classically diagnosed by their history and physical examination, biopsied infantile hemangiomas are uniquely GLUT1+ and demonstrate increased endothelial cell turnover. They have a predictable clinical course, including rapid proliferation during the first year of life (proliferative phase), followed by gradual involution over 1-7 years (involuting phase), and then complete regression after 8 years (involuting phase).\textsuperscript{8,11,20,45} Skin is the most common location for infantile hemangiomas, especially of the head and neck (60%), trunk (25%), and extremities (15%).\textsuperscript{12,20} (Fig. 1). Multiple cutaneous hemangiomas are seen in 10%-25% of cases, with \( \geq 1 \) lesion found in 31% of cases.\textsuperscript{46} Associated liver lesions are seen in up to 13% of children with skin lesions, making it the most common extracutaneous site.\textsuperscript{12,47} Liver hemangiomas are subclassified as focal, multiple, or diffuse. Some authors have referred to multiple infantile hemangiomas as hemangiomatosis, and when 3 or more organ systems are involved, disseminated hemangiomas.\textsuperscript{12,40,48} For simplicity, we will use a single term, infantile hemangioma(s).

Although the majority of infantile hemangiomas are self-limited, treatment may be required if complications arise. Ulceration is the most common complication occurring in up to 13% of lesions.\textsuperscript{10} As the size and number (\( > 5 \)) of infantile hemangiomas increase, so does the risk of complications.\textsuperscript{49} Infantile hemangiomas located on the face, those that are regional, and those near vital structures are more likely to have complications requiring treatment.\textsuperscript{49}

In certain locations, infantile hemangiomas may pose important risks or herald possible underlying anomalies. Orbital hemangiomas may cause visual compromise; hemangiomas adjacent to the nose and mouth are at risk for airway obstruction; hemangiomas involving breast bud may impair breast development; and midline lesions on the forehead, nose, and back raise concern for brain and spinal anomalies. The association of hemangiomas with regional/segmental anomalies has been described, the most common of which include posterior fossa anomalies, hemangiomas, arterial anomalies, cardiac anomalies, eye anomalies, sternal anomalies (PHACES) and lower body hemangioma, urogenital anomalies and ulceration, myelopathy, bony deformities, anorectal and arterial malformations, renal anomalies (LUMBAR) associations (Fig. 2) as listed in Table 3.\textsuperscript{50} Liver hemangiomas are associated with high-output congestive heart failure (shunting of blood through the vascular

Table 3 Summary of Regional and Diffuse Associations, Including Vascular Neoplasms

<table>
<thead>
<tr>
<th>PHACES association*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Posterior fossa abnormalities</td>
</tr>
<tr>
<td>Hemangiomas in 5th cranial nerve region</td>
</tr>
<tr>
<td>Arterial intracranial anomalies</td>
</tr>
<tr>
<td>Cardiac anomalies/coarctation of the aorta</td>
</tr>
<tr>
<td>Eye anomalies</td>
</tr>
<tr>
<td>Sternal defects</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>LUMBAR association†</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lower body hemangioma</td>
</tr>
<tr>
<td>Urogenital anomalies and ulceration</td>
</tr>
<tr>
<td>Myelopathy</td>
</tr>
<tr>
<td>Bony deformities</td>
</tr>
<tr>
<td>Anorectal and arterial malformations</td>
</tr>
<tr>
<td>Renal anomalies</td>
</tr>
<tr>
<td>Maffucci syndrome</td>
</tr>
<tr>
<td>Multiple enchondromas and spindle cell hemangioendotheliomas</td>
</tr>
</tbody>
</table>

*Diagnosis of PHACES requires 3 or more of the above, one of which must be hemangioma.
†Other acronyms used to describe the LUMBAR association include “SACRAL” and “PELVIS.” SACRAL, spinal dysraphism, anogenital anomalies, cutaneous anomalies, renal and urogenital anomalies, angiomatous (hemangioma) of the lumbar/sacral region; PELVIS, perineal hemangiomas, external genitalia malformations, lipomeningocele, vesicorectal abnormalities, imperforate anus, skin tag.
mass), hepatomegaly (which can lead to lung compression and respiratory distress), thrombocytopenia, and hypothyroidism (increased type 3 iodothyronine deiodinase activity)47,51-53 (Fig. 3).

If the history, physical examination, and imaging findings are atypical, raising concern for a lesion more aggressive than an infantile hemangioma, tissue biopsy is occasionally required.12 Neuroblastoma, the main differential diagnosis for multiple hepatic hemangiomas, and the most common metastatic infantile hepatic lesion, can overlap in appearance on cross-sectional imaging.54 Fortunately, neuroblastoma is usually distinguished by urine catecholamine screening and abdominal US with identification of a primary retroperitoneal neoplasm.54 Imaging with CT and/or MRI is not able to definitively distinguish hepatic neuroblastoma metastases and infantile hemangiomas. Thus, urine screening and/or abdominal US should precede CT and MRI to avoid unnecessary risk and waste of resources.12,54

Characteristic US and MRI findings of infantile hemangiomas are summarized in Table 4.7 Follow-up sonography is useful to confirm lesion regression.7,47 Smaller lesions are more likely to be homogenous in signal and enhancement, whereas larger lesions tend to be more heterogeneous and more likely to show centripetal enhancement. Enhancement patterns are nonspecific in infantile hemangiomas, and as involution occurs, gradual replacement by fibrofatty tissue causes MRI features to vary accordingly47 (Fig. 4).

Treatment options for hemangiomas vary depending on the type and severity of complications ranging from laser therapy of skin lesions and medical management with antiangiogenic drugs, to embolization, surgery, and, rarely, organ transplant.8,55 Propranolol has recently been used to treat infantile hemangiomas. Other drugs include steroids followed by chemotherapeutic agents, such as vincristine.56,57 In patients with cardiac shunts, aggressive therapy is more likely. Although clinical parameters can predict which patients will require pharmacotherapy, imaging findings predict nonpharmacologic treatment (ie, embolization).12,47

**Table 4 Key Imaging Features of the Most Common Pediatric Vascular Anomalies**

<table>
<thead>
<tr>
<th>Vascular Anomalies</th>
<th>Gray Scale US</th>
<th>Doppler US</th>
<th>MRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infantile hemangioma</td>
<td>Well-defined</td>
<td>Hypervascular</td>
<td>Iso-to-intermediate signal on T1W</td>
</tr>
<tr>
<td></td>
<td>Solid</td>
<td>Arterial and venous waveforms</td>
<td>Bright signal on T2W</td>
</tr>
<tr>
<td></td>
<td>Homogenous</td>
<td>High vessel density (&gt;5 vessels/cm²)</td>
<td>High-intensity flow enhancement</td>
</tr>
<tr>
<td></td>
<td>Variable echogenicity</td>
<td>High Doppler shift (&gt;2 kHz)</td>
<td>on gradient echo</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Internal flow voids</td>
</tr>
<tr>
<td>Vascular malformations</td>
<td></td>
<td></td>
<td>Vigorous enhancement after</td>
</tr>
<tr>
<td>Venous</td>
<td>Solid echogenic mass with phleboliths; often multispacial and compressible</td>
<td>Monophasic (venous) or no flow pattern</td>
<td>contrast administration</td>
</tr>
<tr>
<td>Lymphatic</td>
<td>Variable multicystic, multispacial masses, with or without fluid and/or debris levels</td>
<td>Multispacial, macrocystic mass with no flow except in septa</td>
<td>T1W heterogenous intermediate signal, no flow voids, T2 FSE fat-saturated or short T1 inversion recovery high signal intensity, T1W SE post gadolinium: enhancement</td>
</tr>
<tr>
<td>Venolymphatic</td>
<td></td>
<td></td>
<td>T1W low-intermediate signal intensity; T2W high signal intensity; T1W post gadolinium: no enhancement, except within septa</td>
</tr>
<tr>
<td>Arteriovenous malformations and fistulas</td>
<td>Cluster of vessels with no intervening well-defined mass</td>
<td>Arterial and venous signals from vessels in the lesions with arterialization of venous structures</td>
<td>T1W and T2W sequences show serpiginous signal voids without a focal mass</td>
</tr>
</tbody>
</table>

FSE, fast spin echo; MRI, magnetic resonance imaging; SE, spin echo; T1W, T1-weighted; T2W, T2-weighted; US, ultrasonography.

**Congenital Hemangiomas (RICH and NICH)**

Congenital hemangiomas differ from infantile hemangiomas in that they are fully developed at birth and test negative for the immunohistochemical marker GLUT1.12,58 Two main types of congenital hemangiomas have been described: (1) NICH, which present at birth and demonstrate proportional growth without regression and (2) RICH, which present at birth and regress completely within 2 years.11 Congenital hemangiomas are much less common than infantile hemangiomas and have no gender predisposition.11,59 Because these
tumors undergo their proliferative phase in utero, they may be seen on prenatal imaging. Histologically, variable-sized lobular capillaries with prominent endothelial cells are seen. They have involuting centers characterized by fibrous tissue, large abnormal draining channel, and loss of lobulations.

Congenital hemangiomas are usually solitary and present on the head and limbs near a joint, unlike infantile hemangiomas, which may occur anywhere in the body. Dermatologists may distinguish these lesions by physical examination, where they will observe a pink-to-violaceous color with multiple tiny or coarse telangiectasias and a surrounding pale halo. Involution starts just after birth with RICH, beginning centrally and progressing to atrophy. Most regress within 1-2 years, if not within the first months of life. Interestingly, RICH leave behind a region of thin atrophied skin with little subcutaneous fat, which is in contrast to infantile hemangiomas, which are gradually replaced by fibrofatty tissue. NICHT may partially involute and soften, but complete resolution is not seen.

Imaging features of congenital and infantile hemangiomas overlap. Thus, the radiologist must be aware of the physical examination and clinical history (age at presentation, lesion course) to suggest a possible congenital hemangioma. A study detailing the imaging tendencies of congenital vs infantile hemangiomas, including heterogeneity (72% of NICH, 62.5% of RICH vs 42.3% of infantile hemangiomas), visible vessels (72% NICH, 62.5% RICH vs 15.4% infantile hemangiomas), and calcifications (17% NICH, 37.5% RICH vs none in infantile hemangiomas), has been performed. Although conventional angiography is no longer indicated in the evaluation of hemangiomas, earlier studies of RICH have demonstrated larger flow voids on MRI and arterial aneurysms on angiography (Fig. 6). In addition, angiography of RICH demonstrates a well-circumscribed mass with intense persistent tissue staining in a lobular pattern with enlarged surrounding systemic artery branches. If occasionally needed, magnetic resonance angiography of hemangiomas is sufficient.

Treatment for RICH is similar to infantile hemangiomas. Rarely, antiangiogenic drugs, embolization, and/or surgery may be necessary. Surgery is the treatment of choice for NICH because of lack of regression.

*Tufted Angioma and Kaposiform Hemangioendothelioma*

TA and KH are rare vasoproliferative tumors that present at or shortly after birth. On histology, TA shows vascular tufts of tightly packed capillaries in a cannonball pattern. KH has both vascular and lymphatic components, consisting of irregular infiltrating nodules of compressed vessels. Recent studies have shown differences in the immunostaining patterns of monoclonal antibody D2-40 that may help distinct-
guish TA from KH. Both lesions may occur on the trunk, extremities, head, neck, retroperitoneum, and, rarely, other locations. Expansion into regional nodes and soft tissues is common in KH, but no distant metastases are seen. TA and KH may be associated with Kasabach–Merritt syndrome—thrombocytopenia caused by platelet sequestration. Kasabach–Merritt phenomenon is a generic term referring to thrombocytopenia resulting from a consumptive coagulopathy.

The imaging characteristics of TA and KH are similar to other vasoproliferative neoplasms, although KH tends to be larger, more ill-defined and infiltrative and is more often associated with impressive flow voids because of numerous feeding and draining vessels (Fig. 7). KH are also more likely associated with secondary destructive osseous changes.

Treatment options for TA and KH are similar to those for infantile hemangiomas. The presence of Kasabach–Merritt syndrome portends a poor prognosis with a mortality rate of 30% in KH, and is thus an indication for aggressive treatment. KH demonstrates aggressive local behavior, and a
lack of distant metastases makes wide local excision and supportive treatment the mainstay of therapy.68,72

Hemangioendotheliomas (Spindle Cell, Epithelioid, and Other Variants)

First described in 1986, spindle cell hemangioendothelioma is a rare vascular neoplasm originally thought to be a low-grade angiosarcoma.73,74 Today it is believed to be a benign reactive vasoproliferation that may present at any age throughout the body.73,76 Histologically, lesions are composed of gaping thin-walled vessels containing thrombi, solid areas of spindle cells, and plump endothelial cells.74 Immunohistochemistry is positive for CD31 and factor VIII antigen and negative for CD34.77,78 Interestingly, an association between spindle cell hemangioendothelioma and Maffucci syndrome has been described (refer to Table 3).79

Epithelioid hemangioendothelioma is a rare, slowly progressive, distinct pathological vascular tumor that can occur at any age anywhere in the body. It is most common in the skeleton, a location not primary to vascular neoplasms.1,80,81 Because of the rarity of this lesion, its nonspecific imaging appearance, and its varied locations and ages of presentation, the pathologist is usually the first to diagnose this tumor, and it is not typically part of the radiology differential diagnosis.81,82

Other rare subtypes of hemangioendotheliomas have been described, including composite, retiform, polymorphous, Dabska tumor, and lymphangioendotheliomatosis.76,83 A complete discussion of these subtypes is beyond the scope of this review article.69

Angiosarcoma

By 1999, there were 30 published cases of angiosarcoma. By 2006, 8 cases had been reported in association with multiple infantile hemangiomas.46 Angiosarcoma has a female preponderance, and the average age of onset is 3.7 years.46 Cross-sectional imaging tends to show an aggressive heterogeneous mass with pooling of contrast and multiple synchronous or metastatic lesions in the liver (Fig. 8).

Vascular Malformations

Vascular malformations are congenital morphogenic anomalies of various vessels that can present at any age.84 They are subdivided into 2 categories: (1) slow- or low-flow and (2) fast- or high-flow malformations. Low-flow malformations contain combinations of capillary, venous, and lymphatic components.

Figure 9 Venolymphatic malformation in a 6-year-old female with leg swelling for 2 months. (A) Photograph of the lower extremity shows an ill-defined purple plaque with nodules. (B) Plain radiograph demonstrates several local phleboliths (arrows). (C) Sonogram of the leg shows mixed echogenic, partially cystic (arrow head), and partially solid mass (arrow). (D) Axial T2-weighted fat-suppressed image reveals a multispatial multicystic mass with fluid levels (arrow head) and phleboliths (arrows).

Figure 10 Arteriovenous malformation in a 16-year-old male patient with a left chest wall mass. Contrast-enhanced coronal reconstructed CT image demonstrates a cluster of enhancing vessels in the chest wall (arrow).
High-flow malformations contain arterial components in combination with other vascular structures. In general, pure malformations in either category are rare. Additionally, because capillary malformations and capillary components of malformations typically involve superficial layers of skin (for example, facial malformations of Sturge–Weber syndrome) that are obvious on physical examination but not well seen with imaging, they will not be discussed further here.

**Slow Flow**

Venous, lymphatic, and venolymphatic malformations are the most common types of vascular malformations with an overall prevalence of up to 1% in the general population. At histology, purely venous and purely lymphatic malformations are rare. The venous portions of malformations contain abnormally formed and dilated superficial or deep veins, the walls of which are thin because they lack smooth muscle. The lymphatic portions of these malformations are formed from collections of lymph vessels filled with serous fluid. Research suggests that altered cell signals may be involved in the formation of lymphatic malformations, such as vascular endothelial growth factor C, which causes lymphatic hyperplasia.

The clinical presentation of low-flow vascular malformations varies because of the many combinations of venous and lymphatic components, as well as variation in size and location. Predominantly venous lesions may present at birth, but they are also seen in later years. Predominantly lymphatic malformations are often apparent at birth and nearly all are present by 2 years of age. The natural history of venolymphatic malformations consists of slow steady enlargement. Venolymphatic malformations present with soft, easily compressible, blue masses that may swell in dependent positions or when venous pressures increase (ie, crying or valsala). They may be small and localized or extensive with infiltration throughout an anatomical region. Although venous malformations most often involve the face, limbs, and/or trunk, they may be found within the internal viscera, bones, and skeletal muscle.

Lesions previously described as “intraosseous hemangiomas” are now pathologically known by their lack of GLUT1 to be venous malformations. Imaging of low-flow vascular malformations involves US for general characterization of cystic (mostly lymphatic) versus solid (mostly venous) components. MRI is often performed to determine lesion extent and plan for treatment. Phleboliths, a specific feature of venous components of vascular malformations, may be seen on plain radiographs when calcified and on MRI before or after calcification (Fig. 9). Both US and MRI are able to demonstrate the typical multispacial, multicystic, and/or partially solid nature of slow-flow
venolymphatic malformations, allowing a diagnosis. Imaging findings are summarized in Table 4.

Treatment of predominantly venous malformations is determined by the extent and location of the lesion. Although many are managed expectantly, some require treatment because they are painful, are in a location where they can cause significant morbidity, or are threatening vital organs (ie, airway compromise). Therapeutic options range from observation and compression garments for palliation of pain and swelling, to sclerotherapy of mostly cystic mass and, finally, surgical excision. However, because of their frequent infiltrative and widespread nature, therapy is often challenging and incomplete.

High-Flow Vascular Malformations

Arteriovenous malformations (AVMs) and arteriovenous fistulas (AVFs) are typically congenital and acquired malformations, respectively. They are characterized by a cluster of arterial and venous channels without a significant solid identifiable mass. AVFs lack the nidus that AVMs contain, and usually occur within the brain. Histologically, AVMs and AVFs consist of dysplastic arteries that drain into arterialized veins forming a vascular nidus in AVMs bypassing capillary beds. The prevalence of AVMs is unknown, but estimates range from 5 to 613 per 100,000 persons. AVMs and AVFs may present with pain, ulceration, ischemic changes, bleeding, and congestive heart failure. On physical examination, they may be warm pink patches on the skin with an underlying vascular murmur or thrill. Common locations include intracranial, intramuscular, muscle, and subcutaneous fat. AVMs may be single, multiple, or part of a genetic disorder, such as hereditary hemorrhagic telangiectasia syndrome (Osler–Weber–Rendu) (refer to Table 4). Imaging characteristics are summarized in Table 4. Lesions are often multispacial and hypervascular on color Doppler US. Lesion extent is best determined with MRI, which shows numerous flow voids (because of turbulence) and hyperintense signal on gradient echo as well as angiographic sequences. Intraosseous lesions may benefit from contrast-enhanced CT.

Therapy for high-flow malformations is preceded by angiography for complete mapping of vessels. The first-line therapy for AVMs and AVFs is embolization. Surgery, or a combination of therapies, may be needed in some children.

Regional Associations and Diffuse Syndromes

Vascular malformations can occur as primary lesions or as a part of a regional or diffuse syndrome (Table 5). Treatment of regional and/or diffuse syndromes varies according to the type of anomalies and symptoms (Fig. 12).

Conclusions

Much has been learned about the histopathology malformations in recent decades, causing change in classification and terminology. The ISSVA classification divides vascular anom-
Studies also indicate that these vascular anomalies are not neoplasms and may not be tumors. Radiologists can best use the ISSVA classification system by correlating imaging findings with patient history and physical findings. Consistent use of this system will help patients receive the correct diagnosis and treatment.

References
