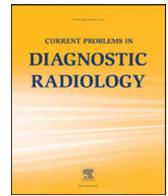




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International Society for the Study of Vascular Anomalies Classification of Soft Tissue Vascular Anomalies: Survey-Based Assessment of Musculoskeletal Radiologists' Use in Clinical Practice

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Aim: There is controversy regarding the diagnosis and classification of vascular anomalies (VA). As torso and extremities are the second most common body part for presentation of VAs, musculoskeletal (MSK) radiologists play a central role in VA classification. The purpose of this study was to evaluate the awareness and clinical use of the International Society for the Study of Vascular Anomalies (ISSVA) classification by MSK radiologists.

Materials and Methods: A Web-based survey was designed and electronically sent to Society of Skeletal Radiology (SSR) members, with 3 questions on demographics and 7 questions on ISSVA classification use and knowledge. The Z-test for binomial proportions is used to assess for statistical significance between subgroups.

Results: The response rate was 12% (130 of 1091), comprised of 64% (83 of 130) academic and 36% (47 of 130) nonacademic MSK radiologists. VAs accounted for only a small (0%–5%) proportion of clinical practice in the majority 92% (119 of 130). Only 17% (22 of 130) of MSK radiologists used the ISSVA classification in practice. Of all respondents, 74% (94 of 127) considered hemangioma a type of vascular malformation (VM). There was no significant difference in the response characteristics between academic and nonacademic radiologists. A greater proportion of MSK radiologists with >5 years' experience provided responses that were discordant with the ISSVA classification compared with less experienced radiologists (0–5 years) in diagnosing hemangiomas in adult patients ($P = 0.02$), and using the presence of phleboliths to diagnose hemangiomas ($P = 0.004$).

Conclusion: Our survey results indicate a lack of familiarity with the ISSVA classification by the MSK radiology community.

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Introduction

Soft tissue vascular anomalies (VA) comprise a spectrum of complex lesions that can involve all regions of the body. Most VAs are often characterized on clinical history, physical examination, and imaging and do not require a tissue diagnosis. After the head and neck, the extremities and trunk are the second most common body parts affected by VAs. Most VA present during childhood. Hence, musculoskeletal (MSK) radiologists and pediatric radiologists play a central role in the diagnosis and classification of soft tissue VA. Currently, the International Society for the Study of Vascular Anomalies (ISSVA) classification system of VAs, which was updated in 2014, is the prevailing classification system routinely used by multidisciplinary teams that diagnose and

manage patients with VAs,^{1–7} particularly among pediatric subspecialties and interventional radiologists.

The ISSVA classification categorizes VAs into 2 major categories: vascular tumors originating from the vascular endothelium with increased cell turnover, and vascular malformations (VMs), which lack the proliferative and mitotic capabilities of neoplastic cells.^{6,8,9} Due to continued advances in histology and genetics, this original classification has evolved with the most recent revision at the 2014 ISSVA workshop in Melbourne, Australia (Table 1).^{6,10} The ISSVA classification enables a systematic approach to VAs used by practitioners in all specialties, correlating clinical history, histopathology, and disease course. Perhaps most importantly, “correct” classification and diagnosis of VA directly informs potential treatment options, making the interpretation of imaging by diagnostic radiologists central to management decisions. Despite this, “incorrect” use of terminology surrounding the ISSVA classification is widespread.^{11–16} Hassanein et al. reported that the term “hemangioma” was used “incorrectly” in 71.3% (228 of 320) of publications per the ISSVA classification scheme, which classifies hemangiomas as proliferative vascular lesions (ie, tumors).¹³ A frequent example of such

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TABLE 1
ISSVA classification of vascular anomalies, modified with permission from 2014 Update¹⁰

Vascular anomalies				
Vascular neoplasms	Vascular malformations			Associated with other anomalies
	Simple	Combined	Named of major vessels	
Benign	Low flow	Low flow	Affect	PHACE* association/syndrome LUMBAR [†] (SACRAL/PELVIS) association/syndrome Klippel-Trenaunay Syndrome Parkes Weber syndrome Servelle-Martorell syndrome Sturges-Weber syndrome Maffucci syndrome CLOVES syndrome Proteus syndrome Bannayan-Riley-Ruvalcaba
<ul style="list-style-type: none"> • Infantile • Hemangioma • Congenital Hemangioma • Rapidly Involuting (RICH) • Noninvoluting (NICH) • Partially involuting (PICH) • Tufted angioma • Spindle cell hemangioma • Epithelioid hemangioma • Pyogenic granuloma 	<ul style="list-style-type: none"> • Capillary (C) • Lymphatic (L) • Venous (V) 	<ul style="list-style-type: none"> • CVM • CLM • LVM • CLVM 	<ul style="list-style-type: none"> • Lymphatics • Veins • Arteries 	
Locally aggressive	High flow	High flow	Anomalies of	Macrocephaly Microcephaly
<ul style="list-style-type: none"> • Kaposiform hemangioendothelioma • Retiform hemangioendothelioma • Papillary intralymphatic angioendothelioma • Kaposi sarcoma 	<ul style="list-style-type: none"> • Arteriovenous (AV) • Arteriovenous fistula (AVF) 	<ul style="list-style-type: none"> • CAVM • CLAVM • CLV • AVM 	<ul style="list-style-type: none"> • Origin • Course • Number • Length • Diameter (aplasia, hypoplasia, stenosis, ectasia / aneurysm) • Valves • Communication (avf) • Persistence (of embryonal vessel) 	
Malignant				
<ul style="list-style-type: none"> • Angiosarcoma • Epithelioid hemangioendothelioma 				

ISSVA Classification of Vascular Anomalies ©2014 International Society for the Study of Vascular Anomalies, Available at: "issva.org/classification."

*PHACE association/syndrome—posterior fossa abnormalities, hemangiomas, arterial intracranial anomalies, cardiac anomalies/coarctation of the aorta, eye anomalies, sternal defects

[†]LUMBAR (SACRAL/PELVIS) association/syndrome—lower body hemangioma, urogenital anomalies and ulceration, myelopathy, bony deformities, anorectal and arterial malformations, renal anomalies.

mis-terminology is the characterization of a routinely encountered hyperintense lesion on both T1 and T2 weighted sequences in an adult vertebra on a spine magnetic resonance imaging as a "hemangioma" even though it has been histologically shown to be a VM and does not exhibit proliferative clinical behavior as a vascular tumor would.^{10,11}

The classification of vascular lesions remains a source of controversy, especially among many MSK radiologists, as highlighted in a recent commentary on benign vascular lesion nomenclature by Kransdorf et al.¹⁶ The authors hypothesized that differences in terminology usage may be dependent on the subspecialty.¹⁶ Although the authors use an ISSVA-based nomenclature in multidisciplinary discussions with treating physicians including interventional radiology, they also use an anatomical-based approach when evaluating focal soft tissue masses in and around joints.¹⁶ The purpose of our study was to assess the awareness and clinical use of the ISSVA classification scheme by MSK radiologists, given that it is an established classification system for physicians caring for patients with VAs.

Materials and Methods

This was an observational study using a Web-based survey. An MSK radiologist (with 14 years of experience who serves as the main consultant for the MSK multidisciplinary tumor team) and a pediatric radiologist (with 14 years of experience who serves as the main VA consultant in a tertiary care children's center) generated a 10-part questionnaire survey (Fig 1). The Society of Skeletal Radiology (SSR) was targeted as the ideal audience as it is comprised of board certified radiologists who devote at least 50% of their time to MSK radiology. In addition, the SSR and thus MSK radiologists were selected due to a

resource based approach as the authors had access to this society. The SSR was contacted with the survey. The survey questions were first reviewed, and then approved by an SSR committee before the distribution of the survey to the active members. The survey was designed for online administration using a commercially available survey sponsor and proprietary web site (<http://www.surveymonkey.com>). The survey was sent out by e-mail to SSR members on November 14, 2014. The results were collected from November 2014 through December 6, 2014.

Three of the ten questions focused on demographics: level of experience, type of practice (academic vs nonacademic) and practice patterns. Six of the ten questions focused on use of the ISSVA classification scheme in clinical practice focusing on the classical clinical and imaging features of the 2 more commonly encountered VA—hemangiomas and venous malformations. One question assessed whether the participant knew about the ISSVA classification system (Fig 1).

Responses were collected electronically without personal identifiable data. Descriptive results were tallied using SurveyMonkey software, and qualitative responses were tabulated. The Z-test for binomial proportions was used to assess for statistical significance. A $P < 0.05$ was considered statistically significant.

Results

The response rate was 12% (130 of 1091). Survey respondents answered most questions in the survey, varying from the highest response rate of 100% (130 of 130) on questions 4 and 9 to as low as 97% (126 of 130) for question 6. Of these, 64% (83 of 130) were academic and 36% (47 of 130) were nonacademic MSK radiologists.

1. How long have you been in practice?
 - a. Fellow or resident
 - b. 0-5 years attending, private practice
 - c. 0-5 years attending, academic practice
 - d. 5-10 years attending, private practice
 - e. 5-10 years attending, academic practice
 - f. 10> years attending, private practice
 - g. 10> years attending, academic practice
2. In your practice, are hemangiomas considered to be a type of vascular malformation?
 - a. Yes
 - b. No
3. In your practice, what is the primary imaging method for imaging of vascular anomalies?
 - a. US
 - b. CT
 - c. MRI
 - d. Conventional angiogram
4. What percentage of your practice involves imaging of vascular anomalies?
 - a. none
 - b. 0-5%
 - c. 5-15%
 - d. >15%
5. Of all the vascular anomalies in patients older than age 2, what percentage have you diagnosed with "hemangioma"?
 - a. >50 % of the cases
 - b. 25 to 50 % of the cases
 - c. 0 to 25 % of the cases
 - d. I don't distinguish hemangiomas from other vascular malformations
6. 42 year old male presents with a compressible painful lump in the thigh. MRI demonstrates interspersed fat in the lesion and focal areas of calcification. Which of the following diagnosis would you choose in this patient?
 - a. Hemangioma
 - b. Hemangioendothelioma
 - c. Venous malformation
 - d. Lymphatic malformation
 - e. Arteriovenous malformation
7. What is the most common type of vascular malformation that you encounter in your practice?
 - a. Hemangioma
 - b. Hemangioendothelioma
 - c. Venous malformation
 - d. Lymphatic malformation
 - e. Arteriovenous malformation
8. In your practice, the presence of phleboliths would support your diagnosis toward a...?
 - a. Hemangioma
 - b. Hemangioendothelioma
 - c. Venous malformation
 - d. Lymphatic malformation
 - e. Arteriovenous malformation
9. Do you use the ISSVA (international society for the study of vascular anomalies) classification?
 - a. Yes
 - b. No
 - c. I haven't heard of it.
10. Which of the following statements is true (according to your practice)?
 - a. Cavernous hemangiomas are in fact venous malformations
 - b. Lymphangioma is the proper term to define lymphatic malformations
 - c. Typically, hemangiomas present with calcifications
 - d. Most vascular malformations respond well to propranolol
 - e. None of the above

FIG. 1. Ten-part questionnaire survey submitted to SSR to assess use and awareness of ISSVA classification by MSK radiologists.

Magnetic resonance imaging was the primary modality for imaging of VAs among 95% (122 of 129) of the respondents. Imaging of VAs only accounted for less than 5% of the clinical practices of 92% (119 of 130) of the respondents. Only 17% (22 of 130) of the respondents used the ISSVA classification scheme in practice; 52% (68 of 130) of respondents had not heard of the ISSVA classification and 31% (40 of 130) did not use it.

Among the respondents, 74% (94 of 127) considered a hemangioma to be a VM. In addition, 60% (75 of 126) of respondents stated that they diagnose hemangiomas in adult patients. Similarly, respondents stated that the most common VA in their clinical practice was a hemangioma (Fig 2). A total of 26% (33 of 127) of the respondents recognized that a cavernous hemangioma is in fact a venous malformation (Fig 3), whereas 52% (66 of 127) of the respondents considered cavernous hemangiomas and venous malformations as different entities.

Tables 2 and 3 summarize the results by years of experience (0-5 years vs >5 years in practice), type of practice (academic vs nonacademic) and use of the ISSVA classification scheme in clinical practice. For question 6, diagnosing hemangiomas in adult patients, less experienced MSK radiologists were more consistent with ISSVA classification compared to more experienced counterparts (44% vs 66%, $P = 0.023$). Again, less experienced MSK radiologists were more

FIG. 1 Continued.

consistent with ISSVA in diagnostic value of “phleboliths” in diagnosis of venous malformations compared to those who had been in practice longer (36% vs 64%, $P = 0.0041$). When examining the responses of

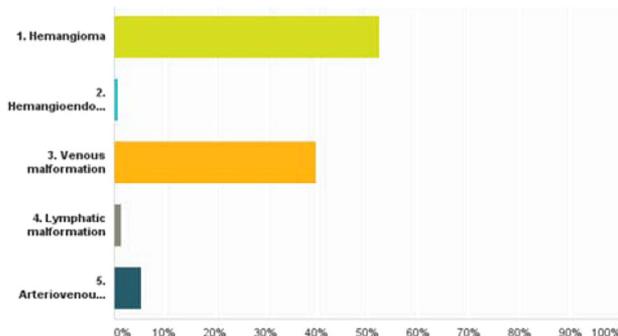


FIG. 2. Responses to question 7 (What is the most common type of vascular malformation that you encounter in your practice?). (Color version of the figure available online.)

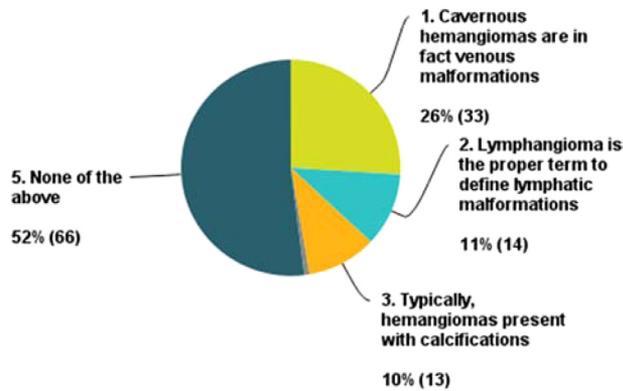


FIG. 3. Response to question 10 (Which of the following statements is true according to your practice?) is demonstrated graphically with approximately 26% of the survey respondents equating cavernous hemangiomas with VMs. (Color version of the figure available online.)

academic radiologists ($n = 67$) alone, 58% (38 of 67) were senior radiologists with greater than 10 years of attending experience. Majority or 76% (50 of 67) of academic radiologists considered hemangioma to be a VM, a distribution similar to the larger group including nonacademic and academic SSR members. When assessing most respondents who used the ISSVA classification scheme ($n = 22$), 59% (13 of 22) considered a hemangioma a VM and 18% (4 of 22) did not distinguish between hemangioma and other VMs. Finally, 54% (50 of 93) of senior or more experienced MSK radiologists and 50% (18 of 36) of less experienced MSK radiologists reported that “I have not heard of the ISSVA classification scheme.”

Discussion

Soft tissue VA are complex lesions with varied imaging appearances and overlapping clinical presentations. Most soft tissue VAs are characterized and often definitively diagnosed on clinical evaluation in conjunction with imaging, without necessitating a biopsy.¹⁴ Hence, correct imaging diagnosis and classification are crucial in determining of proper treatment of VAs. In addition, the use of widely accepted terminology to classify VAs becomes essential for communication between multiple disciplines involved in the care of these patients including diagnostic and interventional radiology, pathology, dermatology, plastic surgery, and other medical or surgical subspecialties.

The ISSVA classification system is an established classification system among many subspecialties, particularly interventional and pediatric radiologists.¹⁻⁷ It derives its foundation from the original histologically based classification proposed by Mulliken and Glowacki⁸ and later revised and adopted in 1996.^{9,10} More recently in 2014, the classification system has been expanded at the ISSVA workshop in Melbourne to incorporate recent advances in knowledge, including newly named anomalies, an association with coagulopathies and identified genes.^{6,10} Knowledge of the ISSVA classification scheme can serve as an important communication tool when discussing VAs in a multidisciplinary setting. Like many other classification schemes within medicine, it serves 2 purposes: first, to organize a larger complex field into clinically meaningful subcategories to guide diagnosis and management and, second, to provide a common terminology that unifies the vocabulary of the diverse group of specialists that care for these patients. Our survey results show limited awareness, knowledge and use of ISSVA classification among MSK radiologists who responded to the survey through SSR (12% [130 respondents out of 1091 known members at the time of the survey]).

One of the first steps in the ISSVA classification of soft tissue vascular anomaly is to distinguish between a vascular neoplasm and a VM. Vascular neoplasms are considered true neoplasms with mitotic potential and cellular hyperplasia whereas VMs not only lack these features but histologically represent errors in vascular “morphogenesis.”¹⁶ In addition, the ISSVA classification incorporates the clinical features of age of onset and the clinical course of the lesion when classifying the lesion appropriately. For example, hemangiomas, the most common vascular neoplasm, are classified as infantile or congenital, and subclassified based on whether involution occurs. Venous malformations, the most common VM, lymphatic malformations, and arteriovenous malformations are congenital and typically do not spontaneously regress, and often require treatment. Such key distinctive clinical features aid in therapeutic management decisions.

The vast majority of the hemangiomas are “infantile hemangiomas (IH),” the most common tumor of infancy which has a predictable clinical course of proliferation followed by involution. IH located in an anatomically critical area (such as the airway), those that threaten function, ulcerate, are associated with underlying anomalies, are large or disfiguring, require treatment. Propranolol is the primary systemic medical treatment and surgery may be required in cases when involution does not result in full resolution. Congenital hemangiomas, divided into rapidly, partially, and noninvoluting subtypes, are significantly rarer than IH and often require surgical excision after infancy. Unlike hemangiomas, VMs typically do not demonstrate

TABLE 2

Comparison of answers to Questions 2, 6, and 8 as per years of experience, type of practice and use of ISSVA classification scheme

	<i>n</i>	Hemangiomas considered a VM	Diagnosis of 42 year old man with a hemangioma	Presence of phleboliths suggested a hemangioma
Level of experience (y)				
0-5	36	67% (24/36)	44% (16/36)	36% (13/36)
>5 years	91*	76% (69/90)	66% (59/89)	64% (58/91)
<i>P</i> value		0.83	0.023	0.0041
Type of practice				
Academic	67*	76% (50/66)	55% (37/67)	51% (34/67)
Nonacademic	60	72% (43/60)	40% (24/60)	62% (37/60)
<i>P</i> value		0.61	0.73	0.21
Use of ISSVA classification in clinical practice				
Yes	22	59% (13/22)	73% (16/22)	23% (5/22)
No	40	73% (29/40)	63% (25/40)	60% (24/40)
“Has not heard of it”	68*	80% (52/68)	69% (44/64)	63% (42/66)
<i>P</i> value				
Yes vs no		0.26	0.42	0.005
Yes vs “has not heard of it”		0.049	0.73	0.00112

*Refers to total number of radiologists in a particular group. The numerator varies among the different questions due to variable number of respondents answering the questions.

TABLE 3
Answers to question 5 (of all the VAs in patients older than age 2 years, what percentage have you diagnosed with “hemangioma?”) divided by level of experience, type of practice and knowledge of ISSVA classification

	n	>50%	25%-50%	0%-25%	No distinction between hemangioma and VM
Level of experience (y)					
0-5	36	28% (10/36)	11% (4/36)	47% (17/36)	14% (5/36)
>5	91	34% (31/91)	20% (18/91)	29% (26/91)	18% (16/91)
Type of practice					
Academic	67	25% (17/67)	19% (13/67)	37% (25/67)	18% (12/67)
Nonacademic	58	66% (38/58)	2% (1/58)	28% (16/58)	3% (2/58)
Use of ISSVA classification in clinical practice					
Yes	22	5% (1/22)	14% (3/22)	64% (14/22)	18% (4/22)
No	40	40% (16/40)	23% (9/40)	23% (9/40)	15% (6/40)
“Has not heard of it”	66	36% (24/66)	17% (11/66)	30% (20/66)	17% (11/66)

spontaneous involution, therefore treatment is planned based on venous, lymphatic, or arteriovenous composition with either percutaneous embolization or sclerotherapy or surgical resection.

When assessing knowledge or application of the ISSVA classification scheme among MSK radiologists, the survey generated divergent responses, which in part can be explained by only 17% of the respondents being aware and using ISSVA classification system as well as these rare VA comprising only 0%-5% of their entire MSK practice. Most respondents (74% [94 of 127]) considered a hemangioma a type of VM, which is inherently discordant with the ISSVA classification system. In addition, Table 2 shows that a larger proportion of experienced radiologists (>5 years) provided responses that were discordant with the ISSVA classification when compared with less experienced radiologists (0-5 years). This trend may be attributable to changes in radiology residency training increasing familiarity of newer graduates with the ISSVA classification. However, of the survey

TABLE 4
Modified World Health Organization classification of vascular neoplasms¹⁷

Vascular tumors	
Soft tissue	Bone
Benign	
<ul style="list-style-type: none"> ➤ Hemangiomas of subcutaneous/deep soft tissue <ul style="list-style-type: none"> ○ Capillary ○ Cavernous ○ Arteriovenous ○ Venous ○ Intramuscular ○ Synovial ➤ Epithelioid hemangioma ➤ Angiomatosis lymphangioma 	<ul style="list-style-type: none"> ● Hemangioma Intermediate locally aggressive rarely metastasizing ● Epithelioid hemangioma Malignant ● Epithelioid hemangioendothelioma ● Angiosarcoma
Intermediate (locally aggressive)	
<ul style="list-style-type: none"> ➤ Kaposiform haemangioendothelioma 	
Intermediate (rarely metastasizing)	
<ul style="list-style-type: none"> ➤ Retiform hemangioendothelioma ➤ Papillary intralymphatic angioendothelioma ➤ Composite hemangioendothelioma ➤ Kaposi sarcoma 	
Malignant	
<ul style="list-style-type: none"> ➤ Epithelioid hemangioendothelioma ➤ Angiosarcoma of soft tissue 	

respondents who used the ISSVA classification scheme, a large proportion of responses were still discordant with ISSVA nomenclature. Instead of the ISSVA classification, many MSK radiologists may rely on 2 other similar “anatomic”-based benign vascular lesion classification schemes: the World Health Organization (WHO) classification for soft tissue and bone tumors (Table 4)¹⁷ or the system proposed by Weiss and Enzinger (Table 5).¹⁸ Both these classifications systems are considerably different than the ISSVA system, resulting in confounding use of terminology. Although our survey did not specifically ask what other classification systems were used by those MSK radiologists who did not follow ISSVA classification system, it is possible that WHO or Weiss and Enzinger classification systems were followed by those respondents. The approach of the ISSVA classification is vastly different from the “anatomically” based paradigm of the WHO or Weiss and Enzinger classifications. The WHO classification does not differentiate between tumor, malformation, infection or reactive vascular phenomena (6) and applies the term “hemangioma” to both vascular tumors and VMs, rendering it difficult to interpret the biologic behavior of the mass in question and facilitate communication among different specialists. In the anatomically based approach, the term “hemangioma” is applied to lesions emanating from named vessels, to adhere to the concept that soft tissue tumors are classified on a “histiogenetic basis according to the adult tissue they resemble” and does not incorporate clinical history.¹⁶ Neither the WHO classification system nor the Weiss and Enzinger systems use the term “VM.”

Our observations are similar to other studies that have evaluated the use of ISSVA classification.¹³ In an effort to evaluate ISSVA terminology related to VA, Hassanein et al noted “incorrect” usage of the term “hemangioma” in 71.3% (n = 228) of the 320 articles.⁶ In particular, the diagnostic specialties of pathology and radiology used “incorrect” nomenclature in 69.0% (20 of 29) and 83.8% (31 of 37) of articles, respectively.⁶ In addition, the most common “incorrect” terms used to describe VA per this review were “cavernous hemangioma” and “capillary hemangioma.” Both of these terms are present in the WHO classification of vascular soft tissue lesions as well as the Weiss and Enzinger system, commonly relied on by both pathologists and MSK radiologists⁹ and reflect the general trend regarding classification of VAs among MSK radiologists noted in our survey. There are several major philosophical differences between the anatomically based WHO as well as Enzinger and Weiss classifications of vascular lesions when compared with the ISSVA classification scheme. One of the major advantages and challenges to the ISSVA classification scheme is incorporation of patient history, flow dynamics and disease natural history. The second major distinction is the use of the term “hemangioma.” Unlike the WHO classification, by the ISSVA classification, the term “cavernous hemangioma” should be avoided since these lesions are VMs without neoplastic potential. By the ISSVA classification, “capillary hemangioma” is the same entity as infantile

TABLE 5
Histological classification of vascular tumors as per Weiss and Enzinger

<ul style="list-style-type: none"> • Benign vascular tumors <ul style="list-style-type: none"> ○ Localized Hemangioma <ul style="list-style-type: none"> ▪ Capillary Hemangioma ▪ Cavernous Hemangioma ▪ Venous hemangioma ▪ Arteriovenous hemangioma ▪ Epithelioid hemangioma ▪ Hemangioma of the granulation tissue type ▪ Deep soft tissue hemangioma ○ Angiomatosis • Vascular tumors of intermediate malignancy <ul style="list-style-type: none"> ○ Epithelioid hemangioendothelioma ○ Spindle cell hemangioendothelioma ○ Malignant endovascular papillary hemangioendothelioma • Malignant vascular tumors <ul style="list-style-type: none"> ○ Angiosarcoma ○ Kaposi's sarcoma

Modified from Enzinger and Weiss.¹⁸

hemangioma. In addition, other pervasive terms considered inaccurate by the ISSVA classification (eg, “synovial hemangioma”) are also accepted by the nomenclature of the anatomically based WHO as well as Weiss and Enzinger systems. The term “hemangioma” refers to a neoplasm with mitotic potential in the ISSVA classification while it refers to a soft tissue mass that may closely resemble normal vessels in the two anatomically based schemes.

ISSVA classification is founded on the original work of Mulliken and Glowacki,^{8,9} which analyzed cellular characteristics and clinical course of 49 cutaneous vascular lesions in children. In addition to histology, hemangiomas ($n = 26$) were categorized based on clinical history of rapid growth during early infancy vs rapid growth followed by regression.⁸ The remainder of the VMs ($n = 23$) were present at birth and grew proportionally with the child, and were comprised of arterial, capillary, venous, or lymphatic elements.⁸ This initial work has since evolved based on advances in histology and genetics and is accepted across multiple disciplines.^{1-7,10} Despite recent efforts within the radiology literature to familiarize the community with the ISSVA nomenclature,¹⁸⁻²⁸ some radiologists debate its value.¹⁶ Furthermore, like many classification schemes, ISSVA classification can be considered fallible or confusing; for example, Kaposi sarcoma is considered a locally aggressive or borderline neoplasm while epithelioid hemangioendothelioma is considered malignant. The ISSVA scheme provides a common language and an adaptable schema structured to incorporate new scientific advances in our understanding of VAs; moreover, it is used among multiple disciplines that treat VA; as such, we are in favor of more widespread adoption in the radiology community.

Our study has limitations. The survey response rate was 12%. Although the total number of responses is more than adequate to derive statistically meaningful conclusions in the subanalyses presented here, the relatively small fraction of SSR members who responded leaves open the issue of self-selection bias. It is possible that radiologists with a particular interest in VMs may have been more likely to complete the survey. However, the expected effect of this bias would be to overstate radiologists' knowledge and awareness of VMs. Thus, the near-universal lack of familiarity with the

ISSVA system amongst MSK radiologists may thus be even more pervasive than described here. The data presented here reflect survey responses by radiologists rather than an audit of radiology reports from a true practice setting. Although we assessed the type of practice, we did not specifically ask whether the MSK radiologists practiced in a hospital or institution with multidisciplinary approach to VAs. We did not directly ask whether or not they followed other classification systems such as WHO or Weiss and Enzinger if they did not follow ISSVA.

Based on our survey results, MSK radiologists have a relatively low level of awareness of the ISSVA classification system, and very few appear to have incorporated it into the structure of their daily practice. We believe that this represents a missed opportunity, because while practice styles vary among different hospitals or institutions, adherence to a common classification system and nomenclature has the potential to decrease ambiguity in a multidisciplinary setting and improve patient care for complex diseases such as VAs. Management of VA requires a multidisciplinary approach but a shared or common language used by the various subspecialties involved in the diagnosis and treatment of these challenging lesions. Concurrent use of less commonly used classification schemes or different classification scheme by each subspecialty may contribute to confusion. Further research should focus on best practices for broadening adoption of the ISSVA system (eg, structured reporting) as well as demonstrating if and how such adoption impacts variability in the management and outcomes of patients with VAs. Alternatively, various organizations and subspecialties dealing with vascular neoplasms and anomalies can consider generating consensus statements regarding ISSVA use.

References

- Hyodoh H, Hori M, Akiba H, et al. Peripheral vascular malformations: Imaging, treatment approaches, and therapeutic issues. *RadioGraphics* 2005;25 (suppl 1): S159–71.
- Moukaddam H, Pollak J, Haims AH. MRI characteristics and classification of peripheral vascular malformations and tumors. *Skeletal Radiol* 2009;38:535–47.
- Ernemann U, Kramer U, Miller S, et al. Current concepts in the classification, diagnosis and treatment of vascular anomalies. *Eur J Radiol* 2010;75:2–11.
- Behr GG, Johnson C. Vascular anomalies: Hemangiomas and beyond—Part 1, fast-flow lesions. *Am J Roentgenol* 2013;200:414–22.
- Behr GG, Johnson CM. Vascular anomalies: Hemangiomas and beyond—Part 2, slow-flow lesions. *Am J Roentgenol* 2013;200:423–36.
- Wassef M, Blei F, Adams D, et al. Vascular anomalies classification: recommendations from the international society for the study of vascular anomalies. *Pediatrics* 2015;136 (1). e203–14.
- Donnelly Lane F, Adams Denise M, Bisset George S. Vascular malformations and hemangiomas. *Am J Roentgenol* 2000;174:597–608.
- Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: A classification based on endothelial characteristics. *Plast Reconstr Surg* 1982;69:412–22.
- Mulliken JB, Glowacki J. Classification of pediatric vascular lesions. *Plast Reconstr Surg* 1982;70:120–1.
- ISSVA Classification of Vascular Anomalies 2014 International Society for the Study of Vascular Anomalies. Available at: “issva.org/classification.”
- Hand JL, Frieden IJ. Vascular birthmarks of infancy: Resolving nosologic confusion. *Am J Med Genet* 2002;108:257–64.
- MacFie CC, Jeffery SL. Diagnosis of vascular skin lesions in children: An audit and review. *Pediatr Dermatol* 2008;25:7–12.
- Hassanein AH, Mulliken JB, Fishman SJ, et al. Evaluation of terminology for vascular anomalies in current literature. *Plast Reconstr Surg* 2011;127:347–51.
- Greene AK, Rogers GF, Mulliken JB. Intraosseous hemangiomas are malformations and not tumors. *Plast Reconstr Surg* 2007;119:1949–50.
- Bruder E, Perez-Atayde AR, Jundt G, et al. Vascular lesions of bone in children, adolescents, and young adults. A clinicopathologic reappraisal and application of the ISSVA classification. *Virchows Arch* 2009;454:161–79.
- Kransdorf MJ, Murphey MD, Fanburg-Smith JC. Classification of benign vascular lesions: History, current nomenclature, and suggestions for imagers. *Am J Roentgenol* 2011;197:8–11.
- Jo VY, Fletcher CD. WHO classification of soft tissue tumours: an update based on the 2013 (4th) edition. *Pathology* 2014;46:95–104.
- Enzinger FM, Weiss SW. Benign tumors and tumorlike lesions of blood vessels. In: Enzinger FM, Weiss SW, eds. *Soft tissue tumors*, 3rd edn, St Louis: Mosby; 1995:579–626.
- Eifert S, Villavicencio JL, Kao TC, et al. Prevalence of deep venous anomalies in congenital vascular malformations of venous predominance. *J Vasc Surg* 2000;31:462–71.

20. Pappas DC Jr, Persky MS, Berenstein A. Evaluation and treatment of head and neck venous vascular malformations. *Ear Nose Throat J* 1998;77:918–22.
21. Pascarella L, Bergan JJ, Yamada C, et al. Venous angiomas: Treatment with sclerosant foam. *Ann Vasc Surg* 2005;19:457–64.
22. Rebeiz E, April MM, Bohigian RK, et al. Nd-YAG laser treatment of venous malformations of the head and neck: An update. *Otolaryngol Head Neck Surg* 1991;105:655–61.
23. Papp DF, Khanna J, McCarthy EF, et al. Magnetic resonance imaging of soft-tissue tumors: determinate and indeterminate lesions. *J Bone Joint Surg Am* 2007;89:103–15.
24. Lowe LH, Marchant TC, Rivard DC, et al. Vascular malformations: classification and terminology the radiologist needs to know. *Semin Roentgenol* 2012;47:106–17.
25. Fayad LM, Hazirolan T, Bluemke D, et al. Vascular malformations in the extremities: emphasis on MR imaging features that guide treatment options. *Skeletal Radiol* 2006;35:127–37.
26. Hochman M, Adams DM, Reeves TD. Current knowledge and management of vascular anomalies: I. Hemangiomas. *Arch Facial Plast Surg* 2011;13:145–51.
27. Tekes A, Koshy J, Kalayci TO, et al. Mitchell Vascular Anomalies Flow Chart (SEM-VAFC): a visual pathway combining clinical and imaging findings for classification of soft-tissue vascular anomalies. *Clin Radiol* 2014;69:443–57.
28. Kollipara R, Odhav A, Rentas KE, et al. Vascular anomalies in pediatric patients: updated classification, imaging, and therapy. *Radiol Clin North Am* 2013;51:659–72.