Classification and ultrasound findings of vascular anomalies in pediatric age: the essential

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Received: 14 September 2018 / Accepted: 24 November 2018 / Published online: 28 November 2018
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Abstract

Proper nomenclature is a major obstacle in understanding and managing vascular anomalies. Often the same term is used for totally different types of lesions or, conversely, the same lesion may be labeled with different terms. Although in recent times there has been a greater understanding of the problems concerning vascular anomalies, episodes of improper use of terminology still remain. The aim of this article, starting from the most recent classification of vascular anomalies, is to provide a clinical and instrumental approach to identifying these lesions and to converge towards a clear and unambiguous terminology that must become univocal among the various operators to avoid diagnostic misunderstandings and therapeutic errors.

Keywords Vascular anomalies · Hemangioma · Ultrasound · Doppler · Children

Sommario

L'esatta terminologia è stata il maggior ostacolo alla comprensione e alla gestione delle anomalie vascolari. Spesso lo stesso termine è stato usato per lesioni totalmente diverse o, viceversa, la medesima lesione è stata etichettata con termini diversi. Nonostante in tempi recenti vi sia stata una maggiore comprensione delle problematiche riguardanti le anomalie vascolari, ancora permangono episodi di uso improprio della terminologia. Scopo di questo articolo, partendo dalla più recente classificazione delle anomalie vascolari, è fornire un approccio clinico-strumentale a queste lesioni e convergere verso una terminologia chiara ed univoca che deve diventare comune fra i vari operatori per evitare fraintendimenti diagnostici e errori terapeutici.

Classification and nomenclature of vascular anomalies

Vascular anomalies may be present at birth or arise shortly thereafter. Since they are often very apparent, they have always made a great impression among the observers who have tried to classify them. Obviously, attempts at classification have been conditioned by the historical context and the methodological trends in vogue at the time of observation. This has led to a continuous revision and redefinition of terms and caused increasing confusion, sometimes with fatal results.

From the initial *descriptive classification*—based above all on the external appearance of those lesions which dazzled the first observers—some very suggestive terms emerged, some of which have extended their influence to the present day. For example, a French dermatologist during the early nineteenth century used the generic term “angioses” (i.e.,

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affection of the vascular system) to describe a variety of acquired and congenital lesions, as well as the term “haematomus tuberosus” (“tumor of the blood similar to a potato”) [1, 2].

Next came *anatomopathological classification*, under which Virchow, considered the father of the cellular pathology, in 1863 generally classified all vascular anomalies as “angiomas”, labeling them according to vascular architecture (e.g., “angioma simplex”, “angioma racemosum”, and “angioma cavernosus”) [1, 3].

Beginning in the early 1900s, *embryological classification* became widespread. One of the leading figures of this system was the Italian Edmondo Malan. In his monumental monograph, Malan divides angiomas into “venous,” “truncular,” “arteriovenous”, and “capillary” [1, 4]. Although conceptually interesting, classification on an embryological basis does not appear clinically useful, as it fails to differentiate between involutive lesions and non-involutive ones.

Finally, on the basis of clinical-histological studies, in 1982 Mulliken and Glowacki proposed a *biological classification* that distinguishes lesions based on the behavior of endothelial cells, with lesions broadly classified as *hemangiomas* (lesions in which endothelial hyperproliferation is present) and *malformations* (lesions presenting normal turnover of endothelial cells) [5].

This fundamental distinction remains the basis of modern classification today. In fact, it was adopted and appropriately modified by the International Society for the Study of Vascular Anomalies (ISSVA) in 1996 to provide the medical community with a universal scientific language that would prevent both misdiagnosis and inaccurate management. Following adoption, the classification was reviewed in 2014 and updated in 2018 [6]. Under this classification, the vascular anomalies are divided into two main groups: *vascular tumors*, characterized by a clonal proliferation of endothelial cells, and *vascular malformations*, determined by errors in different developmental stages of embryogenesis but with a normal endothelial cell turnover. Vascular tumors are divided into three groups in relation to their neoplastic aggressiveness. Vascular malformations are divided into four groups in relation to the vascular structures involved.

Imaging techniques

The first level examination of pediatric vascular anomalies is ultrasound (US). The predominantly superficial location allows a morphological evaluation of the lesions and a dynamic examination of the vascularization. The subsequent integration with magnetic resonance is reserved for selected cases and is always a next step compared to the ultrasound examination.

However, the instrumental investigation cannot be separated from a first clinical evaluation. Some elements must be known by the radiologist or sonographer who performs the examination to better interpret the findings of the investigation:

- When was the lesion first noticed?
- Was it already present at birth or did it develop later?
- What was its growth rate?

The role of ultrasound imaging is fundamental in diagnostic orientation and to address the subsequent therapeutic phase. The main elements that the ultrasound examination must look for are:

- the anatomical localization of the lesion by determining the involved layers (e.g., skin, subcutis, muscular layer).
- degree of compressibility of the lesion (a hard, non-compressible lesion must always be suspicious of possible different nature).
- distribution and density of vascularization of the lesion, including recognition of the type of vessels (arterial or venous) involved.
- assessment whether the clinically detectable lesion is single or consisting of multifocal elements.

The ultrasound examination, however, has intrinsic limits. For example, it cannot clearly define the limits in the case of very extensive and particularly deep lesions, and it presents difficulties in exploring some areas such as those near bony and air-filled structures.

Despite the numerous information that the ultrasound examination can provide, it is not always possible to arrive at a precise diagnosis with the color Doppler exam alone. The great semeiological overlap does not always allow for a differential diagnosis, not only between different vascular anomalies but also with other benign or malignant lesions of the soft tissues. In these cases, it is mandatory to use the MRI and, if necessary, the biopsy examination.

Vascular tumors

Vascular tumors are divided into three main categories according to their nature: benign, locally aggressive, and malignant.

Malignant tumors are, fortunately, infrequent among pediatric patients. Given the large number of recognized lesions, we will cover in particular those most frequently encountered in clinical practice. Benign tumors include:

- Infantile hemangioma.
- Congenital hemangioma (RICH, NICH, PICH).

Fig. 1 Infantile hemangioma: proliferative phase in a 5-week-old boy. The lesion healed after treatment with propranolol

Although they are identified with the same term (hemangioma), these two lesions have two substantial differences, one from the clinical point of view and the other from the histopathological point of view:

- Infantile hemangioma occurs and develops only after birth, while congenital hemangioma (as the name suggests) is already fully developed at birth.
- From the histopathological point of view, the lesions are characterized by different behavior of the glucose transporter 1 (GLUT 1) protein. Infantile hemangioma is GLUT 1 +, while congenital hemangioma is GLUT 1 – (this behavior is common with vascular malformations) [7, 8].

Infantile hemangioma

Infantile hemangioma is the most common vascular tumor in children up to 1 year old [9]. Diagnosis is generally clinical, and ultrasound examination is often unnecessary. In any case, US may be required for extensive and deep lesions and during pharmacological follow-up [7, 9, 10].

It should be noted that this lesion has a characteristic clinical history. Mass is never present at birth, but there may be flat prodromal signs on the skin that subsequently increase in volume during the proliferative phase of hemangioma [9]. Infantile hemangioma, in fact, characteristically has two phases: a *proliferative phase*, in which it appears in the first days/weeks of life and grows rapidly for several months (Fig. 1), and an *involution phase*, lasting up to age 8–9 years, in which there is a progressive reduction in

Fig. 2 Infantile hemangioma. **a** Sonogram shows hyperechoic pattern, 8-month-old girl; **b** sonogram shows hypoechoic lobulated pattern, 5-month-old girl

volume with an increase in adipose tissue compared to the vascular tissue [7, 11–13].

- The US aspect of hemangioma also depends on the phase in which it is examined. In the *proliferative phase*, the hemangioma is generally a well-defined mass with a non-homogeneous echostructure and may be predominantly hyperechoic, hypoechoic, or with a lobulated appearance (Fig. 2). Of these patterns, it is difficult to say which is the prevailing one, and the results in the literature are rather discordant. In some cases, the hyperechoic aspect prevails [9], while in others the hypoechoic aspect prevails [14]. At the color Doppler (ECD) exam, infantile hemangioma appears richly vascularized with high vascular density and uniform vessel distribution within the lesion [7, 9, 13, 15] (Fig. 3). Regarding vascular density, some authors have determined the vascular density in relation to the number of vessels per square centimeter, classifying as highly vascularized lesions with more than five vessels per square centimeter [16]. However, this quantitative approach is not very significant at present due to the enormous improvement of modern ultrasound devices (Fig. 4) and is not very useful from the practical point of view, presupposing a technical uniformity of examination usually not obtainable. Spectral analysis detects prevalent presence of arterial flows that show low resistance and relatively high velocities [7, 17–19]

Fig. 3 Color Doppler sonogram shows high vascular density, characteristic of infantile hemangioma

Fig. 4 Infantile hemangioma in 4-month-old girl. B-flow technology shows the very high vascular density

(Fig. 5). It is also possible to identify venous vessels that show slow flow and are generally poorly modulated [14].

Normally, infantile hemangioma is not often examined in the *involution phase*. In this stage, the echogenicity of the lesion increases progressively, due to the increase in adipose tissue and at the same time decreases the vascular density with arterial flows which show a progressive increase in the Resistive Index (RI) [7, 9, 20, 21].

Congenital hemangioma

Congenital hemangioma is by definition already present at birth. Introduced for the first time in 1996 [22, 23], congenital hemangioma, although presenting vascular characteristics similar to infantile hemangioma, is differentiated by clinical history and histopathological aspects [12, 24, 25]. These lesions grow rapidly in the uterus, so they are very evident at birth (Fig. 6). Based on their evolution they are distinguished in *rapidly involuting congenital*

Fig. 5 Infantile hemangioma. Spectral analysis shows arterial flow with low resistance and high velocities

Fig. 6 Rapidly involuting congenital hemangioma (RICH) in 2-day-old boy (same case as of Fig. 8). Newborn with congenital vascular tumor in the occipital region. Biopsy confirmed the diagnosis

Fig. 7 Rapidly involuting congenital hemangioma (RICH) in 1-month-old boy. Sonogram shows heterogeneous subcutaneous mass that contains large visible vessel (arrow)

hemangioma (RICH) and in *non-involuting congenital hemangioma* (NICH). The RICH shows regression by about 14 months, unlike infantile hemangioma whose regression is slower and can even occur over several years [22].

On the contrary, the NICH does not show involution and tends to grow with the child [23, 26, 27]. This behavior is very similar to the natural history of vascular malformations.

More recently, a third category has been hypothesized called *partially involutive congenital hemangioma* (PICH) [12], as it is easy to imagine these lesions showing a first involutive phase, similar to the RICH, but at some point the involution stops and these lesions assume the same behavior of the NICH [9, 28].

Congenital hemangioma is much less common than infantile hemangioma. It is generally solitary and occurs with more frequency at the level of the head and limbs near the joints [29].

Sometimes it is not possible, on the basis of clinical history alone and of the instrumental finding, to make a differential diagnosis between infantile hemangioma and congenital hemangioma. If a diagnosis is necessary, it is possible to distinguish the two lesions on the basis of immunohistochemistry with the GLUT 1 research (the infantile hemangioma is positive for GLUT 1, while the congenital hemangioma is negative for GLUT 1). The ultrasound and color Doppler findings can be very similar to what is found in infantile hemangioma; however, it is possible to identify some elements more often present in the congenital form [7, 12, 24, 26, 27, 30, 31].

Congenital hemangioma B-mode ultrasound is more inhomogeneous than infantile hemangioma. It is often possible to identify vascular structures of relatively high diameter that well distinguishable in the lesion (Fig. 7) in comparison to the infantile hemangioma whose vascular structures are of smaller caliber and difficult to distinguish in grayscale. The calcifications are not common but when present they aid in

Fig. 8 Rapidly involuting congenital hemangioma (RICH) in 2-day-old boy. Sonogram shows heterogeneous subcutaneous mass with intralesional calcification (arrow) and large visible vessel (arrowhead)

Fig. 9 Rapidly involuting congenital hemangioma (RICH) in 1-month-old boy. Color Doppler shows high vascular density, and spectral analysis shows the presence of low resistance arterial flow

distinguishing the congenital hemangioma from the infantile hemangioma, where the calcifications are very rare (Fig. 8).

The color Doppler vascular density is very high for congenital hemangioma, similar to that of infantile hemangioma, with arterial flows at high velocities and low resistance (Fig. 9). However, the venous vascular signal is more frequently seen as compared to the infantile form. In some cases, the venous flow tends to be predominant and often corresponds to the larger vessels seen at the B-mode that represent dysplastic veins that cross the lesion transversely. Moreover, it is possible to detect the presence of arteriovenous MicroShunt with turbulent flow on the spectral examination, particularly frequent

Fig. 10 Kaposiform hemangioendothelioma in a 2-day-old girl. The “extended view” sonogram shows a large subcutaneous mass of the abdominal wall (arrows). The mass is heterogeneous with some vessels visible on the gray-scale image. *L* liver, *S* spleen

in the NICH. Finally, the possible presence of thrombi in the larger venous vessels has been described, and this characteristic is never present in infantile hemangioma.

Locally aggressive tumors

Kaposiform Hemangioendothelioma (and Tufted Angioma) Although *Tufted angioma* is categorized as a benign tumor while the *Kaposiform hemangioendothelioma* is considered a locally aggressive tumor [32], the two lesions are treated together because they have many similar aspects.

Both forms may be associated with systemic complications such as the Kasabach–Merritt phenomenon (characterized by thrombocytopenia, microangiopathic hemolytic anemia, and consumption of coagulation factors). This common behavior led to the hypothesis that the two lesions may represent a spectrum of the same neoplasm [13]. The finding of the Kasabach–Merritt phenomenon is indicative of the presence of one of these two tumors [9].

On the US examination these lesions tend to be inhomogeneous with poorly defined margins compared to the surrounding tissues [11, 33–37] (Fig. 10). Tufted angioma is generally more superficial, usually surrounded by subcutaneous fat, with a maximum thickness of 1 cm [9, 33], while the Kaposiform hemangioendothelioma is generally deeper, involving the underlying soft tissues. In addition, there are sometimes calcifications that are not present in the Tufted angioma. Although some authors report that the Kaposiform hemangioendothelioma shows a lower vascular density than infantile hemangioma [33], others argue that the color Doppler shows a high vascular density with the presence of vessels already recognizable at B-mode [9, 38] (Fig. 11).

Fig. 11 Kaposiform hemangioendothelioma in a 2-day-old girl. Color Doppler sonogram confirms the high vascular density of lesion

Malignant tumors

Angiosarcoma The angiosarcoma is an extremely rare tumor. Only 2% of vascular tumors are malignant, and angiosarcoma accounts for 0.5% of all pediatric sarcomas [13]. It is a lesion more frequently localized to the limbs. At the moment there are no pediatric ultrasound reports of soft tissue angiosarcoma [9].

Vascular malformations

Vascular malformations are the second large class of ISSVA classification [6]. They are a heterogeneous group of congenital lesions present at birth but sometimes not clinically evident [39]. They tend to grow slowly in proportion to the patient, although some conditions—trauma, hormonal changes, internal bleeding, and pregnancy—can lead to sudden and tumultuous growth [1, 39]. The revision of the ISSVA classification of 2018, confirming when already proposed in 2014, divides vascular malformations into four categories: simple, combined, of a major vessel, and associated with other anomalies. These categories are based on the presence of one or more types of vessels combined with each other in the same lesion.

Here we will deal mainly with *simple malformations*, which are in turn divided into further categories according to the vessels involved: capillaries, venous, lymphatic, and arteriovenous.

From a practical point of view, it is useful to distinguish vascular malformations on the basis of Mulliken's proposal [1, 5] according to their behavior to the spectral analysis in *high-flow lesions* and in *slow-flow lesions*. The former always provide for the presence of arteries and often can be difficult to distinguish from vascular tumors such as infantile hemangioma [14].

Capillary malformation

This category includes various forms of lesions, some common and others very rare. In general, being very superficial, clinically they do not present many problems with the exception of esthetic implications. However, their importance lies in the fact that they can often be associated with other diseases, including, for example, Sturge–Weber syndrome, Parkers–Weber syndrome, or to forms of overgrowth [40, 41].

With normal ultrasound probes, the capillary malformation is not visible in a direct way but only as a nonspecific thickening of the skin and subcutaneous tissue compared with the contralateral healthy side. Only with dedicated dermatological probes of 20 MHz is it possible to detect an increase in vascularization [40, 42, 43].

Venous malformation

Venous malformations are the most frequent form of vascular malformations with a prevalence of about 1% [44, 45]. Their presentation may be varied; they may appear as a group of ectatic and dysplastic superficial veins or, more frequently, be deeper and appear as a real mass in soft tissues with a bluish appearance of the superficial skin [1, 39]. These masses are always soft and compressible (Fig. 12). Only in rare cases can they increase in consistency due to the formation of internal clots. Like all vascular malformations, they can rapidly grow and become symptomatic with puberty or pregnancy.

Sonographically, venous malformations appear as well-margined masses with a “spongiform” heterogeneous echostructure (hypoechoic venous spaces separated by hyperechoic septa), which is hypoechoic in comparison to the surrounding tissues. Isoechoic or hyperechogenic echostructure can rarely occur [14, 46]. The mass is always well compressible. Sometimes it is possible to identify anechoic tubular structures that are recognized as vascular channels [47]. Certainly the ultrasound aspect is nonspecific, and it is often not possible to make a differential diagnosis. However, it is a pathognomonic sign that, in addition to the exploration in B-mode, aids in the diagnosis of venous malformation: the presence of a *phlebolith* (i.e., an intralesional calcification). The presence of phleboliths is extremely rare in the other masses of soft tissues in the pediatric age. Unfortunately,

Fig. 12 Venous malformation. Compressible mass in the subcutaneous soft tissue (arrow). Note the dysplastic ectatic superficial veins (arrowheads) and the bluish discoloration of the overlying skin

Fig. 13 Venous malformation in 13-year-old girl. Sonogram shows well-margined masses with a “spongiform” echostructure, which is hypoechoic in comparison to the surrounding tissues. The presence of intralesional phlebolith (arrow)

this sign is not frequent in venous malformations being present, depending on the various series, from 9 to 16% of cases [14, 46] (Fig. 13).

In the color Doppler examination, venous malformations are slow flow lesions. Vascular density is very low. Veins show low-velocity flow with non-modulated spectrum (Fig. 14). Sometimes the flows are so slow that they are not identifiable [46]. In this case, a light compression on the lesion may be useful to reduce the caliber of the vessels and try to increase the velocity of the intravascular flow [34]. The absence of vascularization may also be due to extensive thrombosis of the vessels involved which must therefore be recognized [39]. Venous malformations can often occur near the joints. In this case, it is necessary to identify whether

Fig. 14 Venous malformation in 5-year-old boy. Spectral analysis shows a low-velocity flow with non-modulated spectrum

the lesion has a direct involvement with the joint, since the possibility of hemarthrosis and subsequent damage of the articular cartilages are frequent [48]. Given the extreme variety of clinical and ultrasound presentations of vascular malformations, in case of doubt or, as mentioned, in doubt of joint involvement, it is always advisable to perform an MRI examination.

Lymphatic malformation

The lymphatic malformations are divided into macrocystic, microcystic, and mixed type.

The criterion of distinction between the macrocystic and microcystic form is not univocal. According to some, a 1–2 cm cut-off must be identified to distinguish between the two forms, while according to others, macrocystic lesions should be considered as those that can be reduced by aspiration and sclerotization [8].

The term “lymphatic malformation” has now replaced the old name of “lymphangioma” (whose suffix ‘oma’ should be applied to lesions with cell proliferation) [1, 39]. Clinically, like the previous ones, the lymphatic malformations are slow-growing lesions that can rapidly increase in size in case of intralesional bleeding. In the case of superficial microcystic forms, it is possible to find the presence of microvesicles present on the skin or on the mucous membranes.

Lymphatic malformations are more frequently superficial, although they can also arise in deeper regions [49]. The most

Fig. 15 Macrocystic lymphatic malformation in 1-month-old boy. Sonogram shows multicystic mass in the thoracic subcutaneous tissue. The cystic spaces are anechoic and separated by septa

Fig. 16 Macrocystic lymphatic malformation in 13-month-old boy. Sonogram shows multicystic mass in the neck subcutaneous tissue. The cystic spaces are hyperechoic from hemorrhage. A fluid–debris level is seen within a cyst (arrow)

frequent sites are the neck, the axillary region, and the mediastinum [1, 7, 34, 39].

With US the *macrocystic lymphatic malformations*, which are the most frequent, appear as lesions containing numerous cystic formations of variable dimensions with liquid content separated by thin hyperechogenic septa (Fig. 15). The lesion is deformable, and compression with the probe alters the shape of the cysts that never collapse completely, unlike the venous malformations [50–52]. At the B-mode, the cystic spaces can be anechoic or have a variable degree of echogenicity in relation to the presence of hemorrhages or infections. In some cases, the formation of fluid–fluid levels is possible (Fig. 16).

The *microcystic lymphatic malformations* often appear as solid hyperechoic formations in which some scattered

Fig. 17 Macrocystic lymphatic malformation in 1-month-old boy. Doppler sonogram shows flow that is confined to the septa. Spectral analysis shows arterial flow with high resistance

cysts may be present. This aspect is due to the presence of numerous cysts that are too small to be displayed with probe resolution [53, 54].

On the color Doppler examination, the malformation is a slow flow lesion. Vascular signals are generally absent. In some cases, it is possible to identify arterial vessels confined in the larger septa. At the spectral examination, the arterial flows show medium–low velocities and, above all, high resistance indices (Fig. 17). This aspect distinguishes them from hemangiomas where arterial resistance is low [14].

Arteriovenous malformation

Arteriovenous malformations consist of an abnormal network of arterial and venous vessels with scarce or absent presence of capillaries.

They are high-flow lesions and are often in differential diagnosis with hemangiomas. The clinical evolution,

Fig. 18 Arteriovenous malformation in 3-month-old boy. Hypoechoic subcutaneous lesion (*) with thin hyperechoic rim (arrows)

from quiescent lesion to hemorrhage and heart failure, is described by the Schobinger classification.¹

On ultrasound examination, the arteriovenous malformations can have a very diversified appearance—from a conglomerate of moderate-sized vessels that determine a lesion that can be detected in soft tissues [55], up to a tangle of vessels that do not determine a real “mass effect” and are therefore poorly identifiable at the B-mode exam and can only be highlighted with the color Doppler exam [14]. One element to consider is the presence of adipose tissue around the arteriovenous malformation due to a fibroadipose proliferation [34] that may result in a thin hyperechoic rim that delimits the lesion (Fig. 18).

At the color Doppler examination, the malformation is a high flow lesion. There are numerous vessels showing “high vascular density” with multidirectional flow. The spectral Doppler analysis shows arterial vessels with high-velocity flow and low resistive index (RI) (Fig. 19). Compared to hemangiomas, the venous vessels have a higher average velocity of flow, with an “arterialized” wave for the transmission of the systolic pulse to the venous bed due to the lack of the capillary network [1, 7, 14, 39] (Fig. 20).

Conclusions

Typically, vascular anomalies may appear as isolated lesions, but both the clinician and the radiologist must know that they are often associated with regional or diffuse diseases

¹ Classification proposed in 1990. The scheme was never published directly by the author but subsequently presented by Mulliken et al. [56–58].

Fig. 19 Arteriovenous malformation in 4-year-old boy. Color Doppler shows arterial flow with low resistance and high velocities

Fig. 20 Spectral venous analysis. **a** Non-pulsatile venous flow in infantile hemangioma; **b** pulsatile venous flow in the arteriovenous malformation

(Table 1). The treatment of these diseases goes beyond the scope of this brief review. For their study, we refer to more specialized texts.

As repeated several times, the overlap of the ultrasound and color Doppler findings, both among the vascular anomalies and neoplastic lesions of different nature, often makes a safe differential diagnosis difficult if not impossible. However, we must remember that ultrasound is often not asked for a diagnosis of nature but an examination that is scrupulous and that gives information as completely and precisely as possible. For these reasons, we consider it useful to propose a report scheme to standardize the examination, so as to reduce as far as possible the dependency on the operator of the ultrasound and to improve the communication between the radiologist and the clinician.

The scheme is shown in Table 2.

Table 1 Summary of association between vascular anomalies with other lesion

Vascular tumors: association with other lesions	
PHACE association/syndrome	Posterior fossa malformations, hemangioma, arterial anomalies, cardiovascular anomalies, eye anomalies, sternal clefting and/or supraumbilical raphe
LUMBAR (SACRAL, PELVIS) association/syndrome	Lower body hemangioma, urogenital anomalies, ulceration, myelopathy, bony deformities, anorectal malformations, arterial anomalies, and renal anomalies
Vascular malformations: association with other lesions	
Klippel–Trenaunay syndrome: CM + VM ± LM + limb overgrowth	
Parkes Weber syndrome: CM + AVF + limb overgrowth	
Saervelle–Martorell syndrome: limb VM + bone undergrowth	
Sturge–Weber syndrome: facial + leptomeningeal CM + eye anomalies ± bone and/or soft tissue overgrowth	
Limb CM + congenital non-progressive limb overgrowth	
Maffucci syndrome: VM ± spindle-cell hemangioma + enchondroma	
Macrocephaly-CM (M-CM/MCAP)	
Microcephaly-CM (MICCAP)	
CLOVES syndrome: LM + VM + CM ± AVM + lipomatous overgrowth	
Proteus syndrome: CM, VM and/or LM + asymmetrical somatic overgrowth	
Bannayan–Riley–Ruvalcaba sd: AVM + VM + macrocephaly, lipomatous overgrowth	
CLAPO syndrome: lower lip CM + face and neck LM + asymmetry and partial/generalized overgrowth	

CM capillary malformation, VM venous malformation, LM lymphatic malformation, AVM arteriovenous malformation, M-CM macrocephaly-capillary malformation, MCAP megalencephaly-capillary malformation-polymicrogyria, MICCAP microcephaly-capillary malformation, CLOVES congenital lipomatous overgrowth, vascular malformations, epidermal nevi, skeletal/scoliosis and spinal abnormalities, CLAPO lower lip CM + face and neck LM + asymmetry and partial/generalized overgrowth

Table from Ref. [6]

Table 2 Proposal for report scheme of the eco-color Doppler exam

B-mode
Locate the lesion (subcutaneous tissue, beyond the fascia, in the muscle, etc.)
Describe the lesion (hard/soft, margins, echostructure: homogeneous/inhomogeneous)
Measure the lesion (three diameters!)
Describe the tissues surrounding the lesion (thickened/non-thickened)
Color Doppler
Non-vascularized lesion
Vascularized mass
Vascular lesion
Spectral analysis:
“High” or “low” flow lesion
Describe Arterial Resistivity Index (RI)
Describe venous spectrum: “flat” or “arterialized”

Indication of any supplementary investigations and/or follow-up diagnostic hypothesis (if possible)

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, and its later amendments.

Human and animal rights This article does not contain any studies with human or animal subjects performed by any of the authors.

Informed consent Additional informed consent was obtained from all the patients for which identifying information is not included in this article.

References

- Mulliken JB, Burrows PE, Fishman SJ (2013) Mulliken and Young's vascular anomalies—hemangiomas and malformation. Oxford Med Online. <https://doi.org/10.1093/med/9780195145052.001.0001>
- Alibert JL (1817) *Nosologie naturelle ou les maladies du corp humain distribuees par familles*. Caille and Ravier, Paris, pp 349–351
- Virchow R (1863) *Angiome. Die krankhaften Geschwulste*. Hirschwald, Berlin, pp 306–425
- Malan E (1974) Vascular malformation (Angiodysplasias). Carlo Erba, Milan
- Mulliken JB, Glowacki J (1982) Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg* 69:412–422
- International Society for the Study of Vascular Anomalies (2018) ISSVA classification for vascular anomalies—2018. ISSVA
- Kolipara R, Odhav A, Rentas KE et al (2013) Vascular anomalies in pediatric patients: updated classification, imaging, and therapy. *Radiol Clin N Am* 51:659–672
- Wassef M, Blei F, Adams D et al (2015) Vascular Anomalies Classification: recommendations From the International Society for the Study of Vascular Anomalies. *Pediatrics* 136:e203–e214
- Johnson CM, Navarro OM (2017) Clinical and sonographic features of pediatric soft-tissue vascular anomalies part 1: classification, sonographic approach and vascular tumors. *Pediatr Radiol* 47:1–12
- Caprio MG, Di Serafino M, Pontillo G et al (2018) Paediatric neck ultrasonography: a pictorial essay. *J Ultrasound*. <https://doi.org/10.1007/s40477-018-0317-2>
- Kolipara R, Dinneen L, Rentas KE et al (2013) Current classification and terminology of pediatric vascular anomalies. *Am J Roentgenol* 201:1124–1135
- Mulliken JB, Enjolras O (2004) Congenital hemangiomas and infantile hemangioma: missing links. *J Am Acad Dermatol* 50:875–882
- Morrow AC, Gupta A, Patel MN, Adams DM (2016) 2014 Revised Classification of Vascular Lesions from the International Society for the Study of Vascular Anomalies: radiologic–pathologic update. *RadioGraphics* 36:1494–1516
- Paltiel HJ, Burrows PE, Kozakewich HP et al (2000) Soft-tissue vascular anomalies: utility of US for diagnosis. *Radiology* 214:747–754
- Razek AA, Huang BY (2011) Soft tissue tumors of the head and neck: imaging-based review of the WHO classification. *RadioGraphics* 31:1923–1954
- Dubois J, Patriquin HB, Garel L et al (1998) Soft-tissue hemangiomas in infants and children: diagnosis using Doppler sonography. *Am J Roentgenol* 171:247–252. <https://doi.org/10.2214/ajr.171.1.9648798>
- Behr GG, Johnson C (2013) Vascular anomalies: hemangiomas and beyond—Part I, fast-flow lesions. *Am J Roentgenol* 200:414–422. <https://doi.org/10.2214/AJR.11.7852>
- Manaster BJ (2013) Soft-tissue masses: optimal imaging protocol and reporting. *Am J Roentgenol* 201:505–514
- Restrepo R, Francavilla ML, Mas R, Lee EY (2017) Up-to-date practical imaging evaluation of neonatal soft-tissue tumors: what radiologists need to know. *Am J Roentgenol* 2017:1–10
- Wu JS, Hochman MG (2009) Soft-tissue tumors and tumorlike lesions: a systematic imaging approach I. *Radiology* 253:297–316
- Kassarjian A, Dubois J, Paltiel HJ et al (2004) Infantile hepatic hemangiomas: clinical and imaging findings and their correlation with therapy. *Am J Roentgenol* 182:785–795
- Boon LM, Enjolras O, Mulliken JB (1996) Congenital hemangioma: evidence of accelerated involution. *J Pediatr* 128:329–335. [https://doi.org/10.1016/S0022-3476\(96\)70276-7](https://doi.org/10.1016/S0022-3476(96)70276-7)
- Dubois J, Rypens F (2009) Vascular anomalies. *ultrasound clin* 4:471–495. <https://doi.org/10.1016/j.cult.2009.11.002>
- Krol A, MacArthur CJ (2005) Congenital hemangiomas: rapidly involuting and noninvoluting congenital hemangiomas. *Arch Facial Plast Surg* 7:307–311
- Berlioni HP (2009) Classification of infantile hemangiomas and other congenital vascular tumors BT—hemangiomas and vascular malformations. *Hemangiomas Vasc. Malformations*. Springer, Milan, pp 23–34
- Enjolras O, Mulliken JB, Boon LM et al (2001) Noninvoluting congenital hemangioma: a rare cutaneous vascular anomaly. *Plast Reconstr Surg* 107:1647–1654. <https://doi.org/10.1097/00006534-200106000-00002>
- A.E. K-B (2017) Congenital hemangioma: a diagnostic challenge. *Pediatr Dermatol* 34:S203–S204. <https://doi.org/10.1111/pde.13196>
- Nasseri E, Piram M, McCuaig CC et al (2014) Partially involuting congenital hemangiomas: a report of 8 cases and review of the literature. *J Am Acad Dermatol* 70:75–79. <https://doi.org/10.1016/j.jaad.2013.09.018>
- Lowe LH, Marchant TC, Rivard DC, Scherbel AJ (2012) Vascular malformations: classification and terminology the radiologist needs to know. *Semin Roentgenol* 47:106–117
- Gorincour G, Kokta V, Rypens F et al (2005) Imaging characteristics of two subtypes of congenital hemangiomas: rapidly involuting congenital hemangiomas and non-involuting congenital hemangiomas. *Pediatr Radiol* 35:1178–1185. <https://doi.org/10.1007/s00247-005-1557-9>
- Rogers M, Lam A, Fischer G (2002) Sonographic findings in a series of rapidly involuting congenital hemangiomas (RICH). *Pediatr Dermatol* 19:5–11. <https://doi.org/10.1046/j.1525-1470.2002.00011.x>
- International Society for the Study of Vascular Anomalies (2014) ISSVA classification for vascular anomalies—2014. ISSVA. <https://doi.org/10.1542/peds.2014-3673>
- Dubois J, Garel L, David M, Powell J (2002) Vascular soft-tissue tumors in infancy: distinguishing features on doppler sonography. *Am J Roentgenol* 178:1541–1545. <https://doi.org/10.2214/ajr.178.6.1781541>
- Dubois J, Alison M (2010) Vascular anomalies: what a radiologist needs to know. *Pediatr Radiol* 40:895–905. <https://doi.org/10.1007/s00247-010-1621-y>
- Liu Q, Jiang L, Wu D et al (2015) Clinicopathological features of Kaposiform hemangioendothelioma. *Int J Clin Exp Pathol* 8:13711–13718
- Fernández Y, Bernabeu-Wittel M, García-Morillo JS (2009) Kaposiform hemangioendothelioma. *Eur J Intern Med* 20:106–113. <https://doi.org/10.1016/j.ejim.2008.06.008>
- Vanlinooghem O, André J, Bruderer P et al (1996) Tufted angioma, a particular form of angioma. *Dev Neurosci* 19:402–404. <https://doi.org/10.1159/000246161>
- Ferrari A, Casanova M, Bisogno G et al (2002) Malignant vascular tumors in children and adolescents: a report from the Italian and German soft tissue sarcoma cooperative group. *Med Pediatr Oncol* 39:109–114. <https://doi.org/10.1002/mpo.10078>
- Johnson CM, Navarro OM (2017) Clinical and sonographic features of pediatric soft-tissue vascular anomalies part 2: vascular malformations. *Pediatr Radiol* 47:1196–1208. <https://doi.org/10.1007/s00247-017-3906-x>
- Lee MS, Liang MG, Mulliken JB (2013) Diffuse capillary malformation with overgrowth: a clinical subtype of vascular anomalies with hypertrophy. *J Am Acad Dermatol* 69:589–594. <https://doi.org/10.1016/j.jaad.2013.05.030>

41. Peterman CM, Vadeboncoeur S, Mulliken JB et al (2017) Wilms tumor screening in diffuse capillary malformation with overgrowth and macrocephaly–capillary malformation: a retrospective study. *J Am Acad Dermatol* 77:874–878. <https://doi.org/10.1016/j.jaad.2017.06.014>
42. Kim C, Ko CJ, Baker KE, Antaya RJ (2015) Histopathologic and ultrasound characteristics of cutaneous capillary malformations in a patient with capillary malformation–arteriovenous malformation syndrome. *Pediatr Dermatol* 32:128–131. <https://doi.org/10.1111/pde.12188>
43. Alfageme Roldán F, Salguero Fernández I, Zamanta Muñoz Garza F, Roustán Gullón G (2016) Update on the use of ultrasound in vascular anomalies. *Actas Dermosifiliogr* 107:284–293. <https://doi.org/10.1016/j.adengl.2016.02.016>
44. Puig S, Aref H, Chigot V et al (2003) Classification of venous malformations in children and implications for sclerotherapy. *Pediatr Radiol* 33:99–103
45. Legiehn GM, Heran MKS (2008) Venous malformations: classification, development, diagnosis, and interventional radiologic management. *Radiol Clin N Am* 46:545–597. <https://doi.org/10.1016/j.rcl.2008.02.008>
46. Trop I, Dubois J, Guibaud L et al (1999) Soft-tissue venous malformations in pediatric and young adult patients: diagnosis with Doppler US. *Radiology* 212:841–845
47. Behr GG, Johnson CM (2013) Vascular anomalies: hemangiomas and beyond—Part 2, slow-flow lesions. *Am J Roentgenol* 200:423–436. <https://doi.org/10.2214/AJR.11.7853>
48. Dalmonte P, Granata C, Fulcheri E et al (2012) Intra-articular venous malformations of the knee. *J Pediatr Orthop* 32:394–398. <https://doi.org/10.1097/BPO.0b013e31824b29ef>
49. Lin JJ, Fisher J, Caty MG (2000) Newborn intraabdominal cystic lymphatic malformations. *Semin Pediatr Surg* 9:141–145. <https://doi.org/10.1053/spsu.2000.7563>
50. Morrow MS, Oliveira AM (2014) Imaging of lumps and bumps in pediatric patients: an algorithm for appropriate imaging and pictorial review. *Semin Ultrasound CT MRI* 35:415–429. <https://doi.org/10.1053/j.sult.2014.05.005>
51. Wagner JM, Lamprich BK (2014) Ultrasonography of lumps and bumps. *Ultrasound Clin* 9:373–390. <https://doi.org/10.1016/j.cult.2014.02.004>
52. Bansal AG, Rosenberg HK (2017) Sonography of pediatric superficial lumps and bumps: illustrative examples from head to toe. *Pediatr Radiol* 47:1171–1183. <https://doi.org/10.1007/s00247-017-3859-0>
53. Dubois J, Garel L (1999) Imaging and therapeutic approach of hemangiomas and vascular malformations in the pediatric age group. *Pediatr Radiol* 29:879–893. <https://doi.org/10.1007/s002470050718>
54. Elluru RG, Balakrishnan K, Padua HM (2014) Lymphatic malformations: diagnosis and management. *Semin Pediatr Surg* 23:178–185. <https://doi.org/10.1053/j.sempedsurg.2014.07.002>
55. Dunham GM, Ingraham CR, Maki JH, Vaidya SS (2016) Finding the nidus: detection and workup of non-central nervous system arteriovenous malformations. *RadioGraphics* 36:891–903. <https://doi.org/10.1148/rg.2016150177>
56. Mulliken JB (2007) Vascular anomalies. *Grabb Smith's Plast Surg*. [https://doi.org/10.1016/0007-1226\(91\)90114-y](https://doi.org/10.1016/0007-1226(91)90114-y)
57. Mulliken JB (2008) Color atlas of vascular tumors and vascular malformations. *Plast Reconstr Surg* 121:333. <https://doi.org/10.1097/01.prs.0000302375.97376.7f>
58. Enjolras O, Wassef M, et al (2006) Color atlas of vascular tumors and vascular malformations. <http://books.google.com>