

## В ПОМОЩ НА ПРАКТИКАТА HELPING PRACTICE

### VASCULAR LESIONS – CONCEPTS AND CLASSIFICATIONS

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### СЪДОВИ ЛЕЗИИ – СХВАЩАНИЯ И КЛАСИФИКАЦИИ

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<p><b>Summary:</b></p> <p><b>Key words:</b></p> <p><b>Address for correspondence:</b></p>	<p>Vascular lesions are heterogeneous group of anomalies with different clinical behavior and histopathological features although many times they look quite similar. The adequate diagnosis is of paramount importance for the appropriate management. From the other hand, historically the descriptive and histologic terms have led to confusion in the communication among medical professionals involved in the treatment and investigation of this group of diseases. The aim of the present article is to make a brief review of the main classifications of vascular anomalies stressing on the most contemporary concepts in the taxonomy of this motley group of lesions.</p> <p>vascular lesions, vascular anomalies, classification, hemangiomas, port wine stains.</p> <p><i>Aylin Shef, MD, Department of Dermatovenereology and Allergology, Military Medical Academy, 3 Sv. G. Sofiyski st. Bg – 1606 Sofia, e-mail: ailin_shef@abv.bg</i></p>
<p><b>Резюме:</b></p> <p><b>Key words:</b></p> <p><b>Адрес за кореспонденция:</b></p>	<p>Съдовите лезии са хетерогенна група аномалии с различна клинична картина и хистопатологични характеристики, въпреки че често изглеждат твърде сходни. Адекватната диагноза е от първостепенно значение за правилното им третиране. От друга страна, погледнато в исторически план, натрупването на множество описателни и хистологични термини води до неяснота в комуникацията между медицинските специалисти, участващи в лечението и проучванията на тази група заболявания. Целта на настоящата статия е да се направи кратък преглед на основните класификации на съдовите аномалии, като се постави акцент върху най-съвременните концепции в таксономията на тази разнородна група лезии.</p> <p>съдови лезии, съдови аномалии, класификация, хемангиоми, порт уайн стейнс</p> <p><i>Д-р Айлин Шеф, Клиника по дерматовенерология и алергология, Военномедицинска академия, бул. „Св. Г. Софийски“ 3, 1606 София, e-mail: ailin_shef@abv.bg</i></p>

## INTRODUCTION

Vascular lesions are a heterogeneous group of anomalies which differ in both their clinical presentation and histopathological features. The adequate diagnosis is of paramount importance because of their distinct differences in morbidity, prognosis, and treatment.

Cutaneous vascular anomalies, comprising a significant part of the vascular lesions, are rare disorders representing errors in vascular development that occur in approximately 0.3% to 0.5% of the population [1, 2]. Historically, the nomenclature of these lesions has been inconsistent. By the use of confusing terminology and classifications, the identification of the vascular anomalies was hampered and has led to confusion. For example, capillary hemangioma, nevus flameus, and port-wine stain (PWS) have all been used in the literature to describe a capillary malformation of the skin [1, 3]. Since management of this multifarious group of lesions depends on the specific vascular malformation, a proper classification and identification is critical. Thus, a brief review of the changing concepts and classifications is of practical and scientific importance for any further studies, commentaries and for the concise communication among the medical professionals which ideally can predict prognosis and guide treatment.

### VASCULAR LESIONS – CHANGING TRENDS

In 1863 Virchow and Wagner in their early published classifications characterized vascular lesions according to the vessel's pathologic appearance [4]. Vascular growths were divided into angiomas (simplex, cavernosum, and racemosum) and lymphangiomas (simplex, cavernosum, and cystoides). The natural history and the biologic behavior of the vascular lesions were not considered and there was a tendency to identify any vascular anomaly as a hemangioma.

A main step along the difficult path to clarity was the work of Mulliken and Glowacki published in 1982 [5], which divided vascular birthmarks into two major categories: hemangiomas and malformations. The Greek nominative suffix “-oma” means “swelling” or “tumor;” however, in modern usage, it denotes a lesion characterized by hyperplasia. This semantic refinement was the key to a binary classification of vascular anomalies as hemangiomas or malformations. This classification was groundbreaking and has served as a cornerstone for the proper identification, investigation, and management of vascular birthmarks.

Hemangiomas were differentiated from vascular malformations by their clinical appearance, histo-

pathologic features and biologic behavior. Vascular malformations had an equal sex distribution, whereas hemangiomas were found to be more common in girls. The natural history of hemangiomas involved rapid proliferation for the first several months of life with subsequent spontaneous regression, often leaving fibrous fatty deposition, overlying anetoderma, and telangiectases (Figure 1). Vascular malformations are often recognized at birth and grow proportionately with the child, with many becoming more prominent at puberty. One of the most commonly seen in the daily practice type of vascular malformations are the Port Wine Stains (PWSs) (Figure 2) which have all typical features of the group of vascular malformations (Table 1) [1, 6].

Mulliken's biologic classification has been widely adopted by clinicians to differentiate vascular birthmarks and is the accepted classification of the International Society for the Study of Vascular Anomalies (ISSVA). In 1996, the classification was modified slightly to reflect the importance of other types of vascular tumors that exhibit different clinical and histologic characteristics than the common infantile hemangioma, including kaposiform hemangioendotheliomas, tufted angiomas among others. Consequently, the updated ISSVA/biologic classification divides vascular birthmarks into vascular tumors and vascular malformations. This classification is useful for managing patients and provides a framework for study of these lesions (Table 2) [11].

Beginning in 2013, a group of ISSVA leaders from both the scientific committee and board, with mindful consideration given to the various existing classifications, sought to update and improve the classification of vascular anomalies, both to make it more clinically relevant and flexible and to acknowledge the new knowledge including new genetic and histologic information available since its invention in 1996. This updated consensus classification is intended to be applicable and functional for all medical and surgical specialties and for every organ or tissue. Because the updated classification lists a large number of different diseases, it is presented as a general table containing the main classes of vascular anomalies (Table 3) [15, 16].

Vascular malformations can be further subdivided into groups on the basis of their vascular components and flow characteristics. They may be composed of slow-flow capillary, venous, or lymphatic channels, fast-flow arterial channels, or a combination of each (Table 4) [1]. Each of the four major categories of vascular malformations (capillary, lymphatic, venous and arterial malformations) has a particular histopathologic appearance and all are lined by quiescent endothelium.

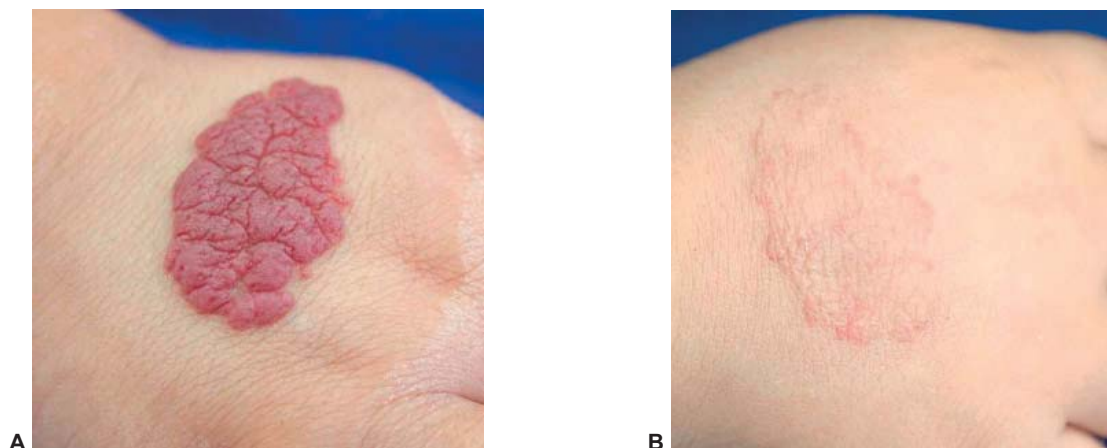


Fig. 1. Infantile hemangioma. (A) Clinical appearance before the start of the treatment; (B) Final result 3 months after the fourth procedure with Long-pulse 1064 nm Nd: YAG laser – post-treatment changes are presented



Fig. 2. Infant with facial capillary malformation, so called “Port-wine stain”

Table 1. Comparison between hemangiomas and port wine stains

<i>Pediatric vascular lesions</i>	<i>Hemangiomas</i>	<i>Port-wine stains</i>
<i>Origin</i>	Absent or small at birth. Grow rapidly in early infancy. Unclear origin: possibly a first-trimester developmental error regarding vasculo-genesis and/or angiogenesis or a result of embolized placental cells. Possible autosomal dominant inheritance [7].	Present at birth. Grow in proportion to child's growth. Unclear origin: possibly a result of vascular channel developmental defects or segmental deficiency of autonomic innervation of postcapillary venules.
<i>Prevalence</i>	Affect 1.1-2.6% of the newborns. Usually develop after birth. Affect 10% of Caucasian children within first year [8, 9]. Three times more common in female than male patients [8]. More common in premature infants. Higher prevalence in infants of mother's postchorionic villus sampling.	Affect 0.3-0.5% of the newborns [10]. Equal prevalence in male and female patients. No significance between premature and full-term infants [8]. Associated with Sturge-Weber and Klippel-Trénaunay syndromes. Very rare late onset in adolescents and adults, usually caused by trauma [11].
<i>Diagnosis</i>	Superficial, deep, or mixed vascular tumors. Histology shows plump endothelial cells [7]. Specific growth and involution phases. Many hemangiomas spontaneously disappear, others grow to disfiguring sizes. Ulcerations possible. GLUT1 positive.	Vascular malformation. Usually well defined [12] red macular stains. Histology shows flattened endothelium [13]. Slow growth throughout lifetime. No regression or ulceration. GLUT1 negative.
<i>Who requires treatment</i>	Treatment is controversial because of unpredictable growth of hemangiomas. Life- or function-threatening lesions, lesions in locations that will permanently scar and ulcerated lesions are high priorities for treatment.	All patients require treatment, because of the expansive nature of PWSs, and their tendency to cause psychologic problems [14].
<i>Treatment types</i>	Systemic and direct corticosteroids, vincristine, recombinant interferon alfa-2a and -2b, imiquimod, surgery, laser treatment, cryotherapy, active nonintervention	Laser treatment, plus surgery for individual nodules and soft-tissue hypertrophy, and orthodontic management for complications [11].

**Table 2. Vascular anomalies: ISSVA/Mulliken classification 1996**

Vascular tumors	Vascular malformations	
	Simple	Combined
Infantile hemangioma	Capillary	AVF, AVM
Congenital hemangioma	Lymphatic	CVM, CLVM
Tufted angioma	Venous	LVM, CAVM
Kaposiform hemangioendothelioma	Arterial	CLAVM
Hemangiopericytoma		
Pyogenic granuloma		
Spindle-cell hemangioendothelioma		

Used abbreviations: AVF, Arteriovenous fistula; AVM, arteriovenous malformation; CAVM, capillary AVM; CLAVM, capillary-lymphatic AVM; CLVM, capillary-lymphatic venous malformation; CVM, capillary venous malformation; LVM, lymphatic venous malformation

**Table 3. 2014 ISSVA Classification of Vascular Anomalies**

Vascular tumors	Vascular malformations			
	Simple	Combined	of major named vessels	Associated with other anomalies
Benign	Capillar	AVF, AVM	CAVF	Klippel-Trenaunay syndrome
Locally aggressive or borderline	Lymphatic	CVM, CLVM	PEV	Parkes-Weber syndrome
	Venous	LVM, CAVM		Sturge-Weber syndrome
Malignant	Arterial	CLAVM		

Used abbreviations: AVF, Arteriovenous fistula; AVM, arteriovenous malformation; CAVM, capillary AVM; CLAVM, capillary-lymphatic AVM; CLVM, capillary-lymphatic venous malformation; CVM, capillary venous malformation; LVM, lymphatic venous malformation

**Table 4. Vascular malformations by flow characteristics and vascular components**

Slow flow	Fast flow
Capillary malformations	AVMs
– Port-wine stains	Combined
– Telangiectases	CAVM
Venous malformations	
Glomuvenous malformations	
Lymphatic malformations	
– Macrocystics	
– Microcystics	
Combined	
– CLM (angiokeratoma)	
– CLVM	
– CMTc ( CVM)	

Used abbreviations: AVM, Arteriovenous malformation; CAVM, capillary AVM; CLM, capillary-lymphatic malformation; CLVM, capillary-lymphatic venous malformation; CMTc, cutis marmorata telangiectatica congenital; CVM, capillary venous malformation

## CONCLUSION

In conclusion, we are convinced that any classification system should be first of all a tool for concise and clear communication, which can predict prognosis and guide treatment. It should be relevant, reproducible, reliable, properly validated and most importantly simple to use and understand. In the field of vascular anomalies, even with the big advancement in understanding of pathogenesis and clinical appearance, confusion still exist among professionals in daily practice.

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