

## WHAT IS IT: HAEMANGIOMA OR VASCULAR MALFORMATION?

Zacharias Zachariou

Congenital vascular anomalies constitute a large group of lesions in children, and they differ in terms of their localization, outer appearance, as well as histology. Additionally, the nomenclature of this group of lesions has been confusing due to the indiscriminate and interchangeable use of the term haemangioma and vascular malformation in diagnosis and management. Whilst hemangiomas are true endothelial cell neoplasms, vascular malformations are localized congenital defects of vascular morphogenesis.

The aim of this article is to overcome obstacles in communication when describing a vascular lesion by adhering to the correct terminology. This leads inevitably to the correct therapeutic guidelines, management and follow-up of these lesions. The therapy guidelines, management and follow-up of hemangiomas and vascular malformations differ and are beyond the scope of this article. *Acta Medica Medianae 2017;56(2):111-114.*

**Key words:** congenital haemangioma classification, haemangioma, infantile hemangioma, vascular anomalies, vascular malformation

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### Introduction

The most common term for any vascular anomaly is haemangioma (Figure 1) which is a descriptive nomenclature that appears to be "good" for almost any kind of a birthmark with vascular origin.



**Figure 1.** Infantile hemangioma (former strawberry hemangioma)

However, it has been applied at the same time to lesions that involute or never involute, often used together with clearly descriptive names as "strawberry", "cherry-hemangioma", etc. As early as the 19th century, Virchow proposed a classification that focused on the microscopic appearances of the

lesions: "haemangioma", "cavernosum", "racemosum", "simplex" and "cystic". The different biologic behavior of each lesion was not taken into consideration making this classification independent of any predictive value in terms of the management and prognosis.

A new classification of congenital vascular anomalies, based on physical findings, clinical behavior, and cellular kinetics, was proposed by J.B. Mulliken and J. Glowacki in 1982 (1). For the first time, two major groups of pediatric vascular lesions – haemangiomas and vascular malformations, were clearly distinguished. In 1996, this classification was adopted by the International Society for the Study of Vascular Anomalies (ISSVA) (2-5).

### Classification and description

All benign vascular tumors are classified histologically:

1. According to the type of fluid they contain:
  - a. Haemangioma - blood-containing lesion
  - b. Lymphangioma - lymph-containing lesion
2. According to the size of the vascular channels:
  - a. Capillary - small diameter vascular channels
  - b. Cavernous - large diameter vascular channels

Further characteristics that contribute to a biological classification are endothelial cell characteristics, physical findings, clinical appearance, radiological features and natural history. This makes the differentiation between vascular lesions with endothelial cell proliferation (i.e. hemangioma) and lesions with structural anomalies (vascular malformations) (1, 2, 4-7).

Haemangiomas are early infancy tumors cha-

racterized by increased cell turnover of endothelium, mast cells, fibroblasts and macrophages. They are the most common benign soft tissue tumor of infancy, occurring in 12% of all infants with higher frequency in girls (ratio 3:1), whites, premature infants (23%), twins and infants born to mothers of higher maternal age. The most frequent localization is the head and neck region (60%), followed by the trunk (25%) and the extremities (15%). Eighty percent of haemangiomas are singular, but 20% are multiple, usually located in various parts of the body. Infants with multiple cutaneous lesions are suspects of having also visceral haemangiomas. The most common visceral sites are liver, lungs and gastrointestinal tract. Other possible visceral locations are: lymph nodes, spleen, thymus, urinary bladder, gallbladder, pancreas, adrenal glands, meninges, brain and spinal cord.

They can be further grouped into infantile hemangiomas (IHs) and congenital hemangiomas (CHs).

IHs are usually not visible at birth and become apparent in the first eight weeks of life as a patch, a blanched spot or localized teleangiectasia, followed by a rapid proliferative phase for the following 6-12 months and grow into a raised rubbery bright-red tumor resembling a strawberry. A gradual involution and a spontaneous regression follow by the age of 5-9 years. Statistically, 30% of haemangiomas resolve by the third year of life, 70% by the seventh year, and the rest till the age of puberty. An involution starts with some white, grey spots spread over the surface of the lesion, from its center toward the periphery. About 40% of involute lesions might reside with skin atrophy and wrinkling. Previously ulcerated areas leave patches of scars.

According to the appearance, IHs can be defined as:

- Focal (localized, raised tumor-like lesion)
- Segmental (flat plaque-like larger lesions that show a geographic segmental distribution)
- Indeterminate (characteristics of both focal and segmental)
  - Superficial (grow just beneath the dermal-epidermal junction)
  - Deep (grow in the lower dermis or subcutaneous tissue)
  - Mixed.

CHs are already present as fully developed lesions at birth and do not exhibit a proliferative phase. They either rapidly involute during the first year of life, called rapidly involuting congenital hemangiomas (RICH), or may never disappear, called non-involuting congenital hemangiomas (NICH). NICH do not show a regression phase and grow proportionately with the growth of the child.

Vascular malformations are not neoplastic lesions; they are pure errors of vascular morphogenesis (8-13). They are almost always sporadic and non-familial.

Despite being congenital, vascular malformations may not be clinically apparent till adolescence or adulthood.

The classified subcategories are based on the predominant type of vessels:

- Capillary venous (CM)
- Lymphatic (LM)
- Arterial (AM)
- Venous (VM)
- Arteriovenous (AVM)
- Combined (see the Table 1.).

Agricultural strategies

**Table 1.** The combined vascular malformations can be specified according to the ISSVA as follows:

|                    |  |        |
|--------------------|--|--------|
| CM + VM            | Capillary-venous                         | CVM    |
| CM + LM            | Capillary-lymphatic                      | CLM    |
| CM + AVM           | Capillary-arteriovenous (Fig.2)          | CAVM   |
| LM + VM            | Lymphatic-venous                         | LVM    |
| CM + LM + VM       | Capillary-lymphatic-venous               | CLVM   |
| CM + LM + AVM      | Capillary-lymphatic-arteriovenous        | CLAVM  |
| CM + VM + AVM      | Capillary-venous-arteriovenous           | CVAVM  |
| CM + LM + VM + AVM | Capillary-lymphatic-venous-arteriovenous | CLVAVM |



**Figure 2.** Capillary-venous-arteriovenous malformation

In addition, all the above named vascular malformations can be classified according to the flow

- High flow lesions
- Low flow lesions.

### Conclusions

For this specific group of lesions, a classification is of utmost importance as the information and the recorded data have to be categorized, so that a proper communication can be achieved, leading to guided treatment plans based on the prognostic knowledge. This classification should be easy to understand and applicable to all involved clinicians.

Consistency in our terminology and classification of vascular lesions will provide profound scientific reports and presentations with an effective communication for the understanding of the pathophysiology, the promotion of research and development of new therapies.

The widely accepted International Society for the Study of Vascular Anomalies (ISSVA) classification differentiates lesions with proliferative endothelium from lesions with structural anomalies and has been very helpful in standardizing the terminologies avoiding inappropriate interchangeable use of the terms hemangioma and vascular malformation.

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## ŠTA JE TO: HEMANGIOMI ILI VASKULARNE ANOMALIJE?

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Urođene vaskularne anomalije čine veliku grupu promena kod dece, koje se razlikuju u pogledu njihove lokalizacije, spoljnog izgleda kao i histološke građe. Pored toga, nomenklatura ove grupe lezija je zbunjujuća zbog neselektivne i neadekvatne upotrebe termina hemangioma i vaskularnih malformacija u dijagnozi i tretmanu. Dok su hemangiomi prave endotelne ćelijske neoplazme, vaskularne malformacije su lokalizovani kongenitalni defekati morfogeneze krvnih sudova.

Cilj ovog članka je da se prevaziđu prepreke u komunikaciji pri opisivanju vaskularnih lezija pridržavajući se ispravne terminologije. Ovo neminovno vodi ispravnoj terminologiji u terapijskim smernicama i praćenju ovih lezija. Smernice za terapiju i praćenje hemangioma i vaskularnih malformacija razlikuju se i izvan su obima ovog članka. *Acta Medica Medianae 2017;56(2):111-114.*

**Ključne reči:** klasifikacija kongenitalnih hemangioma, infantilni hemangiomi, vaskularne anomalije, vaskularne malformacije

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