9 Hamburg Classification: Vascular Malformation

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Vascular malformations occur in such an enormous variety of forms and types that they have been a symbol of confusion among various vascular disorders through decades. Unfortunately even until today in many places, the differentiation of hemangiomas and vascular malformations is not precisely known or is not accurately used in daily clinical practice. A fundamental statement was the clear differentiation of vascular tumors and vascular malformations within the topic of vascular anomalies [1]. A concept of rational treatment of these different findings can only be gained on the basis of a classification referring to clear anatomic and pathological features [2].

That is why in 1988 following an initiative of Prof. Dr. St. Belov, a consensus conference was performed in Hamburg, Germany (Fig. 9.1) under his leadership and guidance convening international scientists of different specialties. The only topic was to create a classification of congenital vascular malformations, which should be simple, clearly arranged, comprehensible, and implementable in clinical practice. The sessions unanimously resolved that the vascular tumors have to be discussed absolutely apart from the extensive group of the congenital vascular malformations. Following the proposals of Malan [3], those "vascular malformations were differentiated into a number of anatomo-clinical pictures, each with a precise definition of the vascular abnormality, of its evolution and of the therapeutic possibilities." In addition, Malan introduced the concept of the "predominant type of the involved vessel" because he noticed that in vascular malformations, very rarely only one type of vessel alone is affected, but in most cases polyangiopathies have to be dealt with.

In order to define the vascular malformations which were formed extratruncular out of the primitive vascular network during the reticular stage of its embryonal development, the term extratruncular form was installed into the classification. Within this term the limited form is included as well as the infiltrating form which is specific for vascular malformations. In contrast, the vascular malformations which derive from a disturbance in the late phase of the vessel development affect the main vessels and such are called the truncular forms. Malan [3] and Belov [4, 5] are convinced of the idea that the truncular and the extratruncular forms are the result of a defect in the embryonic phase of development of the vessels. The latest results in molecular and genetic research and development demonstrate that this concept may be right [6].

The Hamburg classification [5] was adopted by this working group in 1988 (Table 9.1), and the conclusion in 1993 [4] was published as follows: "(1) the proposed classification of congenital vascular defects based on anatomic and pathological features has proved to be useful in clinical practice. It is valid for vascular defects in all locations (central, visceral, and peripheral),

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AU2 **Fig. 9.1**

includes all types and anatomic forms of vascular malformations, yet is quite simplified. (2) A uniform and universal classification system is necessary for clear communication between the many different specialists dealing with congenital vascular defects. (3) It offers a clear and precise descriptive system to serve as the basis for diagnosis of congenital vascular defects. (4) A unified classification system offers the possibility of uniform analysis and comparative reporting between scientific investigators working in this field around the world" (see Table 9.1).

This conclusion, published by Belov [4], became true, and every specialist dedicated to congenital vascular malformations accepted this Hamburg classification and worked with it efficiently. However, soon the capillary/microvascular form was added, and in 2007 a modified Hamburg classification was proposed and worldwide accepted and recommended [7] (Table 9.2).

Table 9.1 Classification of congenital vascular defects according to their species and anatomic form ("Hamburg Classification 1988")

Anatomical forms		
Species	Truncular	Extratruncular
Predominantly arterial defects	Aplasia or obstruction dilatation	Infiltrating limited
Predominantly venous defects	Aplasia or obstruction dilatation	Infiltrating limited
Predominantly lymphatic defects	Aplasia or obstruction dilatation	Infiltrating limited
Predominantly AV shunting defects	Deep AV fistulae superficial AV fistulae	Infiltrating limited
Combined vascular defects	Arterial and venous, (without AV-shunt) hemolymphatic (with or without AV-shunt)	Infiltrating hemolymphatic limited hemolymphatic

AU3 **Table 9.2**

A. Hamburg classification[a] of Congenital Vascular Malformations (CVMs) – species
Arterial malformation
Venous malformation
Arterio-Venous malformation
Lymphatic malformation
Capillary malformation
Combined vascular malformation
B. Hamburg classification of CVMs[b, c]: forms – embryological subtypes
1. Extratruncular forms
Infiltrating, diffuse
Limited, localized
2. Truncular forms
Obstruction or stenosis
Aplasia; hypoplasia; hyperplasia
Stenosis; membrane; congenital spur
Dilatation
Localized (aneurysm)
Diffuse (ectasia)

ISSVA International Society of the Study for Vascular Anomalies

[a]Original classification was based on the consensus on the CVM through the international workshop held in Hamburg, Germany, 1988, and subsequently modified based on the predominant lesion

[b]Represents the developmental arrest at the different stages of embryonic life: Earlier stage – Extratruncular form; Later stage – Truncular form

[c]Both forms may exist together; may be combined with other various malformations (e.g., capillary, arterial, AV shunting, venous, hemolymphatic and/or lymphatic); and/or may exist with hemangioma)

Further modifications of the Hamburg classification were proposed by the ISSVA (International Society for the Study of Vascular Anomalies) in 1996 and in 2014. While the 1996 modification was elegant in its simplicity, but did not adequately and sufficiently reflect the current understanding of vascular malformations [8], several details were missing. That is why in 2014 another updated and expanded modification was published basing on the original Hamburg classification [9] (Table 9.3). Again capillary (i.e., microvascular), lymphatic, venous and arteriovenous malformations, and arteriovenous fistulas are differentiated. Malformations of the main named vessels are specified again as truncal, and combined extratruncular forms are also considered. In addition, subgroups like "vascular malformations associated with other anomalies" or

t3.1 **Table 9.3** ISSVA classification of 2014

t3.2	Overview table	
t3.3 t3.4	Vascular tumors	Simple vascular malformations(extratruncal)
t3.5	Benigne v.t.	Capillary m.(CM)
t3.6 t3.7	Locally aggressive or borderline v.t.	Lymphatic m. (LM) primary Lymphedema
t3.8	Malignant v.t.	Venous m.(VM)
t3.9 t3.10		Arteriovenous m.(AVM) AV-Fistulas (AVF)
t3.11 t3.12		*Combined vascular malformations(extratruncular)*
t3.13 t3.14 t3.15		*Truncular vascular malformations of maior named vessels*
t3.16		*Lymphatics*
t3.17		*Veins*
t3.18		*Arteries*

104 "provisionally unclassified vascular anomalies"
105 are mentioned. As an appendix the causal genes
106 of vascular anomalies as they are known today
107 are also completely included [10, 11].

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Author Queries

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