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ISSVA classification: Controversy with the benefit and liability

RAUL MATTASSI

The classification of vascular anomalies of the International Society for the Study of Vascular Anomalies (ISSVA) was approved by the general assembly of the society during the 19th meeting in 2014 in Melbourne, Australia, and updated and approved in 2018 during the general assembly of the society in 2018 in Amsterdam, the Netherlands. This classification is the result of a main job of the French pathologist Michel Wassef, who tried to create a structural basis in which all the different types of vascular anomalies could be included.[1]

The classification has a multilevel structure with a main first step (an overview), which includes all the types of defects and many subgroups in which all the known anomalies are included. One of the main advantages of this classification is the possibility to add different new forms without changing the main base, as this topic is quickly evolving due to active research in the field (Figure 1.1).

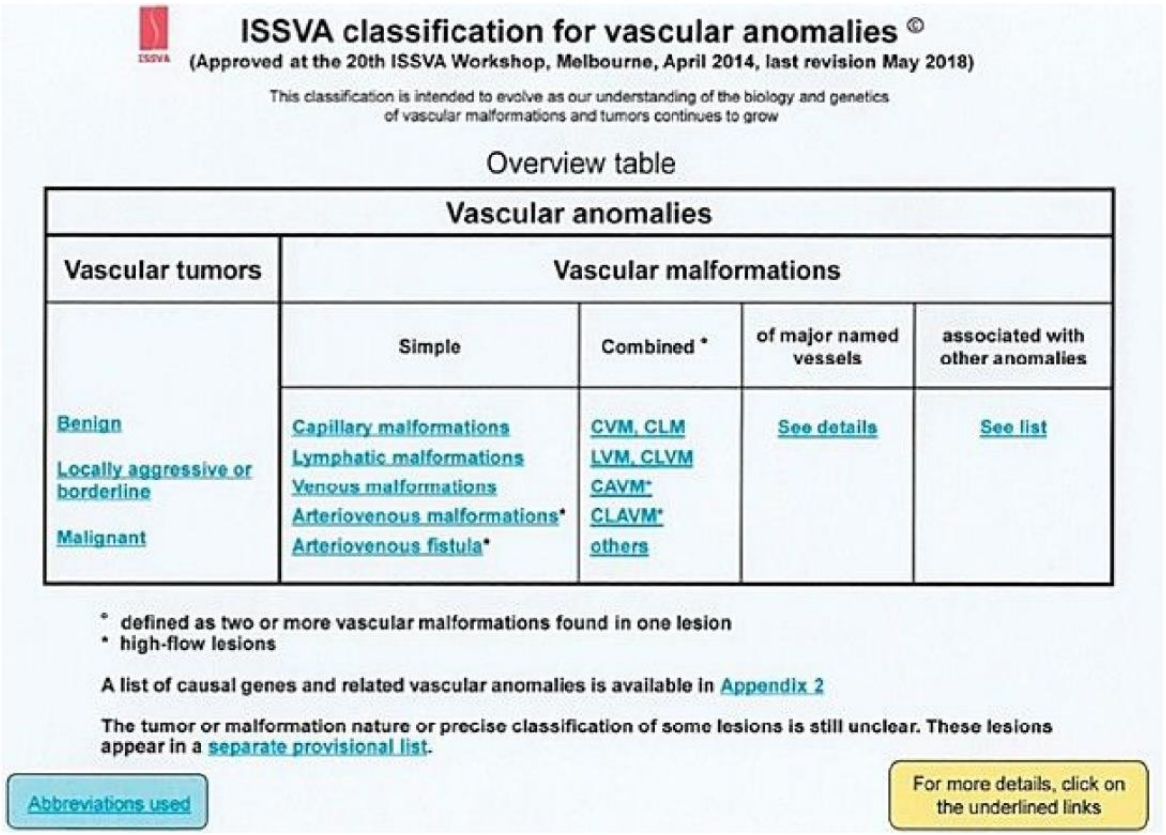

Figure 1.1 International Society for the Study of Vascular Anomalies classification, first overview table, last revision 2018. Available at www.issva.org—by clicking on the underlined links, additional tables open with all subgroups of vascular anomalies.

One of the main updates of the classification, approved in Amsterdam, was the inclusion in the classification of the new genetic data. This rapidly developing field requires a continuous update that is easily possible with such a flexible classification.

To switch over to the new classification from the older one, however, was not quickly achieved by many doctors, as some aspects of the new one were not immediately understood. Old, eponym-/syndrome-based classifications (Klippel–Trenaunay/Parkes Weber syndromes and others) and hemodynamic-/flow-based classifications (fast flow/low flow) remain to cause significant confusion and failed to make a clear distinction between types of malformations (e.g., arteriovenous [AV] malformation and AV fistula).

The most accepted former classification was the Hamburg one (Belov, 1989), which has the advantage of simplicity, only dividing the different types of anomalies in main groups, rendering simple and clear a field that was confusing in the past.[2,3]

The Hamburg classification was an anatomopathological one, with a distinction between defects of the main vessels (truncular forms) and areas of dysplastic vessels in tissues (extra-truncular defects). The distinction is important because extra-truncular defects were considered areas of immature vessels (remnants of primitive vascular tissue) that may recur, while truncular forms may not. The issue is discussed in detail in Chapter 2.

Similar groups (truncular and extra-truncular) exist also in ISSVA classification however in different names. Truncular forms are denominated in ISSVA classification "defects of the main named vessels," an expression a little uncomfortable to use because of its length. Extra-truncular forms are named in ISSVA "simple forms," a term that may be misleading in case of extensive defects.

In the first/original version, ISSVA classification failed to distinguish clearly truncular lymphatic defects, which were included in the "simple" group, but this is now corrected through the latest version.

The general structure of this new updated version, however, is complex, due to the different tables (a total of 19) that cover all types of defects, including some forms that are still not clearly understood. Nevertheless, it can be managed simply by the basic classification on the first overview table (see Figure 1.1), which is roughly similar to the Hamburg classification, with only a new group, the association with other anomalies. To go more in detail, further tables and subclassifications can be consulted (Figure 1.1).

In conclusion, the ISSVA classification is the most modern and complete classification for vascular anomalies. For vascular malformations, it is based on the type of defect, like in the Hamburg classification. The step-by-step structure, from general overview to single groups, allows all known defects to be included and new knowledge to be adapted and expanded. Genetic data inclusion is a further progression.

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2

Hamburg classification: Controversy with the benefit and liability

PETER GLOVICZKI AND DAVID J. DRISCOLL

Classifications are organized knowledge of a subject. As new information accumulates, updates or entirely new, powerful classifications emerge. The best classifications of diseases and disorders are not only all inclusive, but they also guide us in management.

Vascular malformations have been classified in many ways, some simply based on appearance of the lesions or resemblance to fruits, birds, fish, or insects. More complex classifications were arranged according to morphological features, embryological development, endothelial characteristics, and cell biology, hemodynamics, angiographic appearance, or genetics.[1–7] The classification from the International Society for the Study of Vascular Anomalies (ISSVA) was approved at the 20th ISSVA Workshop in Melbourne, Australia, in 2014 and updated in 2018.[8,9] As presented in Chapter 1, the updated ISSVA classification has been gaining traction, particularly because of major recent progress in genetics. It is used frequently now by scientists and physicians worldwide, since this complex classification, based on anatomy, embryology, and genetics, is helpful in management, particularly when deciding on drug therapy based on genetic information.

The Hamburg classification has had an established role in the classification of vascular malformations; it is simple and easier for clinicians for everyday use than the continuously enlarging, updated, and all-inclusive ISSVA classification with over 120 types of vascular anomalies listed now in 13 tables.[9] Using the updated Hamburg classification, physicians can make rapid assessments of the type and extent of the malformation, which is particularly helpful in selecting interventions for management. Clinical practice guidelines of the International Union of Phlebology,[10] the

International Union of Angiology,[11] the Italian Society for Vascular Investigation,[11] the American Venous Forum,[3] and the European Society for Vascular Surgery all recommend at present using the modified Hamburg classification of vascular malformations.[12]

MODIFIED HAMBURG CLASSIFICATION

The Hamburg classification was developed under the leadership of Belov, Loose, and Weber during the seventh Meeting of the International Workshop on Vascular Malformations in Hamburg in 1988.[13] The group working on vascular malformations was formed earlier by Mulliken during their first meeting in Boston, Massachusetts, in 1976. The classification was updated in Seoul, Korea, in 1995, and the currently used modified Hamburg Classification was published in the 2013 Consensus Document of the International Union of Phlebology, by Lee and others.[10] The modified Hamburg classification is primarily an anatomic classification, with an embryological subclassification.

Primary anatomic classification

The primary anatomic classification distinguishes the subgroups based on predominant anatomy. Although the original classification did not include all segments of the circulatory system, in the new, updated classification, the anatomical subclasses are all inclusive, and arterial, venous, arteriovenous, capillary, lymphatic, and mixed vascular malformations are distinguished (Table 2.1). Parts 2, 3, 4, and 6 of this book discuss in detail each anatomic class. Over 70% of the vascular malformations are mixed, and these frequently complex abnormalities may include capillary, venous, arteriovenous, or lymphatic elements. Although the Hamburg classification discourages the use of eponyms, many have been widely accepted and used. The list of some of the clinical syndromes with vascular malformations includes Klippel–Trenaunay (Figure 2.1a through c), Parkes Weber, Servelle–Martorell, Sturge–Weber, Rendu–Osler–Weber, von Hippel–Lindau, Kasabach–Merritt, Proteus, and Maffucci syndromes, among others. Syndromic classifications are presented in detail in Chapter 3 and in Part 5 of this book.

Table 2.1 Modified Hamburg classification of congenital vascular malformations

A. Anatomical classification (predominant vascular bed)
- Arterial malformation
- Venous malformation
- Arteriovenous malformation
- Lymphatic malformation
- Capillary malformation
- Combined vascular malformation

B. Embryological subclassification

1. Extra-truncular forms
   - – Infiltrating, diffuse
   - – Limited, localized
2. Truncular forms
   - • Obstruction (occlusion or stenosis)
     - – Agenesis
     - – Aplasia
     - – Hypoplasia
     - – Coarctation
     - – Hyperplasia
     - – Membranous obstruction
     - – Congenital spur
   - • Dilatation
     - – Aneurysm (localized)
     - – Ectasia (diffuse)
   - • Persistence of embryonic vessels
   - • Anomalies of the origin, course, number, and length of major vessels
   - • Anomalies of venous valves
   - • Abnormal arteriovenous communication of truncal vessels

Source: Adapted from Lee BB et al. *Int Angiol*. 2015;34:97–149.

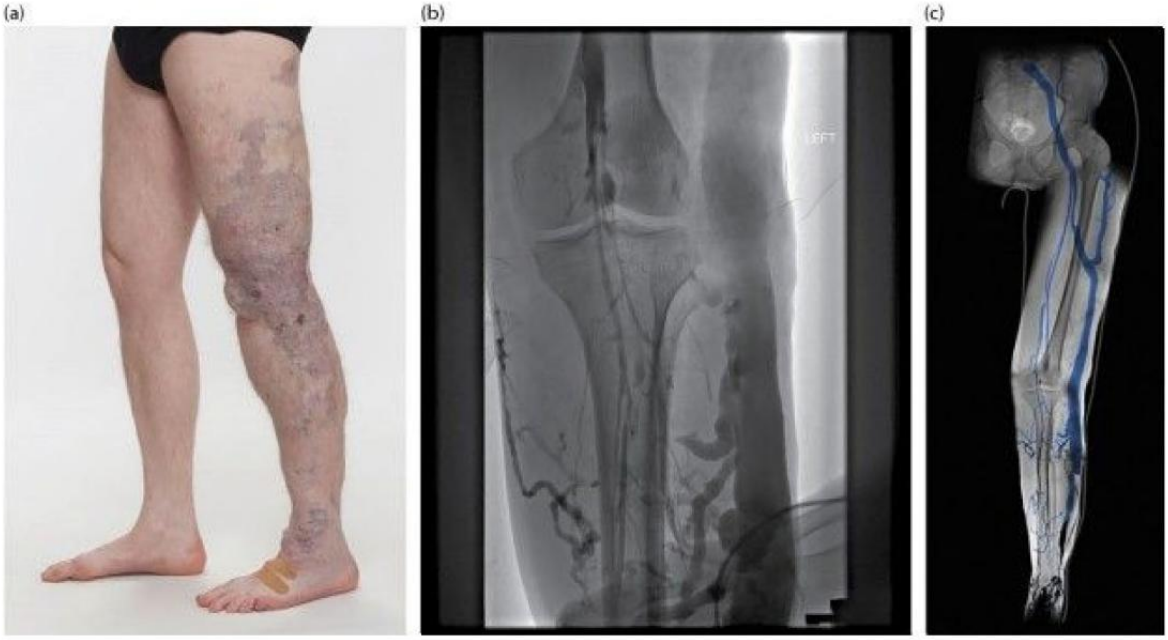

Figure 2.1 (a) Thirty-five-year-old male with advanced symptomatic Klippel–Trenaunay syndrome, presenting with a larger and longer left lower extremity, phlebo-lymphedema, extensive capillary malformation, and atypical lateral varicosity with a persistent lateral embryonic vein. (b) Venogram shows the huge persistent lateral embryonic vein and a patent, although diminutive deep venous system. (c) Venogram of the entire leg and pelvis. (From Rathore A et al. *J Vasc Surg Venous Lymphat Disord*. 2018;6:523–525.)