ALL CONGENITAL VASCULAR MALFORMATIONS SHOULD BELONG TO ONE OF TWO TYPES: "TRUNCULAR" OR "EXTRATRUNCULAR", AS DIFFERENT AS APPLES AND ORANGES!

Byung-Boong Lee

Center for the Lymphedema and Vascular Malformations, George Washington University, USA

ABSTRACT

Congenital vascular malformation represents a group of anomalous vascular structures caused by defective development through various stages of embryogenesis. In 1988 a workshop for this unique field of vascular pathology was held in Hamburg, Germany, and new classification was evolved on the basis of consensus through this workshop. Congenital vascular malformations are now classified based on their predominant vascular components, namely: arterial malformations, venous malformations, lymphatic malformations, capillary malformations, and arteriovenous malformations. The majority of congenital vascular malformations exist alone as independent forms, but infrequently the malformation can exist as a combined form. Depending on the embryological stage at which the defective development occurs, all congenital vascular malformations present with one of the two distinctive characteristics that originate from the embryological background. Congenital vascular malformations originating from an early stage of embryogenesis are classified as extratruncular lesions, while those with developmental arrest at a later stage of embryogenesis, no longer exhibiting the evolutional potential to grow, are referred to as truncular malformations. In addition to the embryonic characteristics of the malformation that depend on "evolutional potential" originating from the mesenchymal cells, each malformation also has its unique haemodynamic characteristics. Clinically relevant haemodynamic impact is especially found among truncular lesions, such as marginal vein or fistulous arteriovenous malformations.

Key words: congenital vascular malformation, embryogenesis, extratruncular vascular malformation, Hamburg classification, truncular vascular malformation.

EDITORIAL

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ADDRESS FOR CORRESPONDENCE
Byung-Boong Lee
Center for the Lymphedema and Vascular Malformations
George Washington University
2150 Pennsylvania Avenue, NW
20037 Washington DC, United States
e-mail: bblee38@comcast.net

I dedicate my humble editorial review to Dr. Tomasz Drążkiewicz, the founder of the Journal, for his retriment.

Congenital vascular malformation (CVM) represents a group of anomalous vascular structures caused by defective development through various stages of embryogenesis. Therefore, these birth defects can develop anywhere in the (peripheral) vascular system: artery, vein, and/or lymphatics, and present various conditions of vascular defects with different characteristics and behaviours based on the vascular system involved.

Hence, the CVM has remained a symbol of confusion throughout the past era of "angiodysplasia" in the last century, due to such extreme variety of clinical presentation, and often condemned as an enigma of modern medicine based on totally unpredictable behaviour. Nonetheless, subsequently, through a new era of "vascular malformation" for the last four decades, the old concept based on limited knowledge with mostly name-based eponyms and classification has been replaced by a new concept based on advanced knowledge about its aetiology and pathogenesis to clarify its anatomical, pathological, and physiological status.

In 1988 the "historical" workshop for this unique field of vascular malformation was held in Hamburg, Germany for the first time, to open a new era of "vascular malformation", to fulfil the mandate for a contemporary classification to modernise the concept for the advanced management of the CVMs. Subsequently, a new classification was evolved on the basis of the consensus through this workshop led by Professors E. Malan and S. Belov: the ISSVA [International Society for the Study for Vascular Anomaly] Classification, to define the vascular tumour and vascular malformation under the name of "vascular anomaly"; and the Hamburg Classification for

more specific verification of the vascular malformation with its embryological background [1, 2].

The CVM is now classified based on its (pre)dominant vascular components, namely arterial malformation (AM), venous malformation (VM), lymphatic malformation (LM), capillary malformation (CM), and arterio-venous malformation (AVM). The majority of the CVMs exist alone as independent forms, but infrequently the CVM exists as a combined form, often affecting more than one vascular system to make the condition more complicated; as a mixture of various CVMs they are now classified separately as haemo-lymphatic malformation (HLM) [3-6].

Therefore, the CVM locates anywhere within the body in a variety of numbers, shapes, extents/degrees, and conditions, either as one single type of (pre)dominant lesion or *mixed* condition with other CVMs. But despite such an extensive variety of lesions, they all have one of two distinctive characteristics originating from the embryological background [7, 8].

Depending on the embryological stage at which the defective development occurs, this embryological event can result in a wide range of clinical presentations of the CVMs with unpredictable clinical course and erratic response to the treatment with a high risk of recurrence and morbidity.

When the defective vascular development occurs in the "earlier" reticular stage of the embryogenesis, the CVM lesion continues to possess unique mesenchymal cell characteristics of evolutional potential to grow. Even after birth, this embryonic tissue remnant continues to grow when the condition is met with sufficient stimulation (e.g. menarche, puberty, pregnancy, female hormone, surgery, injury/trauma).

Such unpredictable behaviour is entirely due to unique *embryological characteristics* as the outcome of developmental arrest in its "early" stage, and "*recurrence*" following ill-planned management became the trademark of this specific group of CVMs. Therefore, this group of CVMs originating from the "early" stage of embryogenesis is classified as "extratruncular" lesion across the board regardless of its type (e.g. venous, lymphatic).

"Extratruncular" lesions exist as a cluster of amorphous vascular tissue infiltrating to the surrounding tissue/structure as an embryonic tissue remnant. But these lesions never have any direct involvement or extension to the named vessel trunks (e.g. femoral, popliteal) and remain as isolated independent vascular lesions (cf. truncular lesion). In other words, any CVM lesion with direct involvement to the named vessels does NOT belong to this extratruncular lesion group.

On the contrary, another group of CVMs, as the outcome of developmental arrest at the "later" stage of embryogenesis, no longer possess this embryological characteristic of evolutional potential to grow because this embryological event occurred long after such unique embryological characteristics were lost following the reticular stage. Hence, they were named/classified separately as "truncular" lesions as opposed to the "extratruncular" lesions to verify this crucial difference: a lack of mesenchymal cell characteristics.

But this defective development occurs while the vessel trunk is formed to make a matured vessel so that all the "truncular" lesions are directly involved with the normally matured vessel trunk to cause defective vessel trunk. Therefore, clinically they present as the condition of obstruction or dilatation of the formed vessel (e.g. vein web, venous aneurysm), or aplasia, hypoplasia, and/or hyperplasia of the vessel development (e.g. agenesis/rudimentary deep vein, absence of femoral artery).

Foetal (truncal) vessels, however, which failed to take a normal involution process to disappear at birth and remained as an embryonic vein (e.g. sciatic vein, marginal vein) after the birth, are also classified as a variant of truncular lesion in view of the outcome of developmental arrest along the later truncal formation stage like other truncular lesions [9, 10].

Nevertheless, all these truncular lesions generate much more serious haemodynamic impact to the involved vascular system (e.g. marginal vein, fistulous AVM, primary lymphedema) in comparison to the extratruncular lesions, although they no longer carry the risk of recurrence and/or continuous growth.

Therefore, precise identification of the CVM lesion either to extratruncular or truncular type is the FIRST step to approach any CVM lesions before consideration of any treatment strategy. All the CVM lesions confirmed as "extratruncular" warrant special precaution on this "recurrence issue", especially for the AVM. The treatment strategy for all extratruncular lesions should be based on the risk of "recurrence". Conversely, all "truncular" lesions should be assessed on their haemodynamic impact with the priority before any consideration for the management in view of potentially serious haemodynamic consequences (e.g. suprahepatic IVC occlusive disease) [7, 8].

CONCLUSIONS

Congenital vascular malformations has embryonic characteristics of "evolutional potential" originating from mesenchymal cells when developmental arrest occurs in the 'earlier' stage of embryonic life: an extratruncular lesion. Congenital vascular malformations also has its unique haemodynamic characteristics affecting each involved vascular system-arterial-venous-lymphatic system, especially among truncular lesion with serious haemodynamic impact. The subsequent management principle and strategy of each malformation lesion should be fundamentally different between extratruncular lesions and truncular lesions.

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