Hamburg Classification in Vascular Malformations: Clinical Applications of the System

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DISCLOSURE OF CONFLICTS OF INTEREST
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Hamburg Classification in Vascular Malformations: Clinical Applications of the System

I do NOT have any relevant financial relationships with any commercial interests.

Congenital vascular malformations (CVMs) represent a group of defective vascular structures as the result of defective development of the vascular system from various stages of embryogenesis.

Therefore, CVMs present at birth as an inborn vascular defect in a variety of the condition involving one or more components of the peripheral circulation systems: arterial, venous, lymphatic and capillary systems.

To add to the confusion, most CVM conditions were initially described using name based eponyms only based on the clinical presentation (e.g. Klippel and Trenaunay syndrome; F.P. Weber syndrome; Servelle and Martorell syndrome; Hippel and Lindau syndrome).

Such old nosology and terminology of CVMs without proper information on etiology, anatomy, and pathophysiology, failed to provide appropriate defining of the characteristics among the CVMs.

This old classification based on clinical presentation alone was unable to provide critical anatomical and pathophysiological information for proper evaluation, diagnosis, and treatment.
Hamburg Classification of Vascular Malformation

- Hence, a new classification of CVMs was mandated to adequately describe the anatomy and pathophysiology of these lesions.
- Malan and Puglionisi proposed a new classification to distinguish the different venous, arterial and other associated malformations for the first time (1964), which became the basis of the subsequent Hamburg Classification and ISSVA Classification.
- They also described the morphological difference between lesions involving the main vessel trunks, often with a direct communication ("truncular" form) and the lesions occurring peripherally as separate defects ("extratruncular" form).

Subsequently, the Hamburg Classification was formulated based on the consensus during a workshop held in Hamburg, Germany, 1988 as a new system to replace the old name based eponyms and to meet the mandate for the contemporary management of the CVMs.

It was further modified to improve its clinical applicability providing additional critical information taking into account the stage when the developmental failure occurred during embryogenesis.

Congenital Vascular Malformation (CVM)

Hamburg Classification*, modified

- Arterial defects
- Venous defects
- Arteriovenous (AV) shunting defects
- Lymphatic defects
- Capillary defects*
- Combined vascular defects

* Based on the consensus on CVM through the international workshop in Hamburg, Germany, 1988.
* Capillary malformation was not included in initial classification.

The origin of this morphological difference between both groups has been explained on the basis of an embryological mechanism as an outcome of the arrest or disturbance in development of the vascular system during various stages of angiogenesis, from the earlier stage where the primitive vascular structures are still in the "reticular network stage" before evolving into mature structures to the later stage of vascular trunk formation.

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- To avoid the confusing term of "angioma" (c.f. hemangioma), Belov et al. re-introduced an old embryologic term: "extratruncular", described by Sabini F.R. (1917), for these 'angiomatous' lesions based on their distinctively different morphology from "truncular defects" during the Hamburg Consensus meeting in 1988.
- This new term 'extratruncular' successfully replaced the often misleading old term "angioma" and stopped the confusion involving the 'angioma versus hemangioma'.

The Hamburg Classification of Vascular Malformation was formulated based on the consensus during a workshop held in Hamburg, Germany, 1988 as a new system to replace the old name based eponyms and to meet the mandate for the contemporary management of the CVMs.

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Hamburg Classification- Embryological subclassification

1. Extratruncular forms
   - Diffuse, infiltrating
   - Limited, localized

2. Truncular forms
   - Obstruction
   - Hypoplasia; Aplasia; Hyperplasia
   - Stenosis; Membrane; Congential spur
   - Dilatation
   - Localized (aneurysm)
   - Diffuse (ectasia)

Developmental arrest at the different stages of embryonal life:
- earlier stage – extratruncular form; latter stage – truncular form.
- 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.

* Capillary malformation was not included in initial classification.
When the embryological defect is originated during the early stage of embryogenesis, the lesion will maintain its unique mesenchymal cell, angioblast characteristics. But, when the defective development occurs during the later stage when the vascular trunk is formed, the lesion would no longer have its potential to grow (e.g. popliteal vein aneurysm, iliac vein stenosis).

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Hence, the worsening of CVM lesions would depend on the type of the endothelial cells present as remnants of the primitive capillary network that maintains its ability to proliferate.

Based on the potential to proliferate and grow when stimulated (e.g. menarche, female hormones, pregnancy, surgery, trauma), the group of the CVMs originating from the earlier stage of embryogenesis was designated to the 'extratruncular' type by S. Belov et. al. in order to distinguish from the 'truncular' type of CVM lesion from the later stage of embryogenesis.

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The growth tendency of extratruncular CVM can also be explained by the most recent genetic theory, based on gene mutations within the tissues. Identification of the causative genes in several defects has permitted more precise diagnosis.

However, the main issue is not the pathology that produces the truncular or extratruncular CVM, but the morphology of the lesion and its clinical behavior, which remain critical for proper clinical management.

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The 'Modified' Hamburg Classification based on embryological characteristics of the CVMs is crucial for clinicians be aware of the potential for growth and proliferation of extratruncular CVM lesions in order to minimize the risk of recurrence and subsequent morbidity.

Extratruncular lesions have a high tendency to progress/worsen or recur after treatment, while truncular lesions do not. Hence, identification and confirmation of this embryological subtype is the first step toward the proper management of every CVM lesion.

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Conclusion

Thank you for your attention!

I dedicate this lecture to late Professor Stefan Belov on his pioneering work to establish new concept on Vascular Malformation.
Truncular Lymphatic Malformation (LM) - Primary Lymphedema

Extratruncular Venous Malformations (VM)

Extratruncular Lymphatic Malformation (LM)

Extratruncular AV Malformation (AVM) in right hip