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Letter to the Editor

Terminological precision in vascular anomalies: beyond "lymphangioma" and "haemangioma"

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Dear Editor,

Vascular anomalies, frequently encountered in the paediatric population, represent a heterogeneous group of disorders whose classification has historically been plagued by inconsistent and misleading nomenclature [1]. A significant and persistent issue concerns venous malformations (VMs) and lymphatic malformations (LMs), which are still often incorrectly referred to as "haemangioma" and "lymphangioma", respectively. Despite advances in understanding their distinct pathogenesis and clinical behaviour, these outdated terms remain in common use [2]. The recent 2025 International Society for the Study of Vascular Anomalies (ISSVA) classification update refines this framework, integrating flow characteristics and genetic aetiology, making precise terminology even more crucial [3,4]. This imprecise language misrepresents the fundamental nature of these conditions and has tangible, negative implications for diagnosis, management, and patient perception.

Historical classification often grouped distinct entities under broad, descriptive terms like "haemangioma" and "lymphangioma", prompting the development of a standardised, biologically-based system by the ISSVA [1,3,4]. The critical distinction lies in separating vascular tumours from vascular malformations. The term "haemangioma" should be reserved specifically for benign vascular tumours like infantile haemangioma, characterised by endothelial cell proliferation and a predictable lifecycle [3]. In contrast, "lymphangioma" incorrectly implies a neoplastic process. Embryological and molecular evidence has clarified that LMs are primarily errors in vascular development – malformative in origin, not true neoplasms [2,5].

A key strength of the ISSVA system is its dynamic nature. The 2025 update, for instance, replaced previous categories with a primary division into fast-flow and slow-flow malformations, and developmental anomalies of named vessels, better reflecting clinical behaviour and underlying genetics. According to this system, LMs are categorised as slow-flow malformations. Crucially, the term "lymphangioma" is absent from this classification. Likewise, "haemangioma" is precisely defined under vascular tumours, and its application to non-proliferative venous malformations is explicitly discouraged [3,4].

Why terminology matters?

1. Clinical and therapeutic implications: Mislabelling a LM as a "lymphangioma" or a VM as a "haemangioma" can directly lead to clinical mismanagement. Lymphatic and venous malformations are typically managed with interventions like sclerotherapy or surgery. In stark contrast, true vascular tumours like infantile haemangiomas often respond to pharmacologic agents such as beta-blockers. Using an incorrect diagnostic label may steer clinicians away from effective treatments towards ineffective or even contraindicated ones, potentially compromising patient outcomes. Radiologic and pathologic correlation, including immunohistochemical markers, is essential.

2. Research and data harmonisation: Inconsistent terminology hinders meta-analyses, comparative studies, and the development of robust, evidence-based clinical guidelines. Standardised nomenclature is essential for accurate data aggregation, reliable interpretation of scientific literature, and facilitating collaborative research. The ISSVA framework provides this essential common language.

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A Study design · **B** Data collection · **C** Statistical analysis · **D** Data interpretation · **E** Manuscript preparation · **F** Literature search · **G** Funds collection

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3. Psychosocial Impact: The suffix "-oma" can erroneously suggest a neoplastic tumour, causing unnecessary anxiety and distress for patients and their families. Precision in language to distinguish a static "malformation" from a proliferative "tumour" is crucial for effective counselling, setting realistic expectations, and alleviating undue psychological burden. Accurate terminology is a cornerstone of ethical, patient-centred communication.

The universal adoption of the 2025 ISSVA classification is a practical necessity for modern, precision-based medicine. Clinicians, radiologists, pathologists, researchers, and journal editors should actively discourage outdated terms and champion precise terminology: "lymphatic malformation" and reserving "haemangioma" strictly for its defined vascular tumour counterparts.

A practical, imaging-based algorithm, aligned with the 2025 ISSVA update, is instrumental. The primary questions in radiological evaluation should be: "Is the lesion a proliferative tumour or a malformation?" and if a malformation, "What is the flow dynamics (fast-flow vs. slow-flow)?"

1. Vascular tumour (proliferative): characterised by contrast-enhancing solid tumour stroma.
   - Benign (e.g. infantile haemangioma): often shows flow voids on MRI, high T2 signal, homogeneous contrast enhancement, and high vascularity on Doppler ultrasound. Typically GLUT-1 positive.
   - Borderline/malignant (e.g. kaposiform haemangioendothelioma): imaging characteristics are variable and often aggressive, with invasive features.

2. Vascular malformation (non-proliferative, structural): characterised by dysplastic vascular channels without a solid tumour stroma.
   - Fast-flow malformation (e.g. AVM): identified by a vascular nidus with prominent feeding arteries and early draining veins. MRI often shows prominent flow voids.
   - Slow-flow malformation:
     – Venous malformation (VM): look for phleboliths, a spongiform appearance on US, very high T2 signal on MRI, and progressive, patchy centripetal contrast enhancement.
     – Lymphatic malformation (LM): characterised by cystic spaces, fluid-fluid levels, and contrast enhancement only in the walls or septa.
     – Capillary malformation (CM): typically presents as a cutaneous stain.
     – Combined malformations (e.g. CLVM): display imaging characteristics of more than one simple malformation type.
   - Developmental anomalies of named vessels (e.g. vena cava agenesis): this category includes structural anomalies of major specific blood vessels.

The updated ISSVA classification introduces additional categories to improve diagnostic accuracy. "Potentially unique vascular anomalies (PUVA)" serves as a provisional category for lesions with ambiguous or overlapping characteristics between tumours and malformations, preventing misclassification. In cases of atypical or combined malformations, multidisciplinary collaboration is essential to achieve an accurate diagnosis and ensure optimal patient management. This imaging-centric algorithm reinforces the 2025 ISSVA classification in daily practice.

In conclusion, terminological clarity is the cornerstone of accurate diagnosis, effective interdisciplinary communication, and truly patient-centred care in vascular anomalies. Embracing the updated 2025 ISSVA classification system, supported by practical imaging criteria, reflects our current understanding of disease biology, directly improves clinical outcomes, and ensures we communicate with the precision and empathy our patients deserve.

Disclosures

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4. Conflicts of interest: None.

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