Evidence-Based Medicine in the Treatment of Infantile Hemangiomas

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KEYWORDS

• Vascular anomalies • Infantile hemangioma • Propranolol therapy • Steroid therapy
• Pulsed dye laser therapy

KEY POINTS

- The International Society for the Study of Vascular Anomalies (ISSVA) (www.issva.org) is the presiding organization for the classification of vascular anomalies.
- Oral steroids are now considered a second choice option for the management of infantile hemangiomas (IHs).
- Oral propranolol is becoming the first line of treatment of the management of IHs if observation or laser therapy is not sufficient.
- Surgery has become more accepted as a means of early treatment of hemangiomas and relies on a physician's clinical judgment regarding timing of intervention.
- Multimodality algorithms, specifically addressing individual components of these tumors, result in the best functional and cosmetic outcomes.

HISTORICAL PERSPECTIVE AND CURRENT CLASSIFICATION SCHEME

The broader picture of vascular anomalies must first be elucidated to better understand the disease process of hemangiomas. The history of facial vascular anomalies is one plagued by confusing nomenclature, misdiagnosis, and lack of a unified consensus on the classification of an incredibly broad group of lesions. In 1982, however, the diligent work of Mulliken and Glowacki[1] introduced the first classification system of vascular anomalies. Their study analyzed cellular characteristics of 49 vascular lesions and distinguished vascular tumors from vascular malformations based on histopathology, increased endothelial cell turnover, and differences in clinical history.[1] A decade later, the ISSVA was founded and has since led the way in maintaining international consensus on the classification of vascular anomalies.[2] The classification system has enhanced physician understanding and ability to accurately diagnose and appropriately prescribe therapy in patients afflicted with vascular anomalies.

The current classification scheme for vascular anomalies undergoes frequent updating and redefining by ISSVA (**Table 1**). Currently, IH is classified as a benign vascular tumor. Like all vascular tumors/hemangiomas, IHs are characterized by increased endothelial cell turnover, which differentiates them from vascular malformations.[1] Current theories surrounding the pathophysiology of IHs revolve around its unique expression of glucose transporter protein 1 (GLUT-1), possibly

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Table 1
International Society for the Study of Vascular Anomalies classification for vascular anomalies

Vascular Anomalies				
	Vascular Malformations			
Vascular Tumors	Simple	Combined	Of Major Named Vessels	Associated with Other Anomalies
Benign vascular tumors IH Congenital hemangioma Rapidly involuting congenital hemangioma[a] Noninvoluting congenital hemangioma Partially involuting congenital hemangioma Tufted angioma Spindle-cell hemangioma Epithelioid hemangioma Pyogenic granuloma (also known as lobular C hemangioma) Others Locally aggressive or borderline vascular tumors Kaposiform hemangioendothelioma Retiform hemangioendothelioma Papillary intralymphatic angioendothelioma, Dabska tumor Composite hemangioendothelioma Kaposi sarcoma Others Malignant vascular tumors Angiosarcoma Epithelioid hemangioendothelioma Others	C malformations L malformations V malformations AV Ms[a] AV fistula[a]	Defined as 2 or more vascular malformations identified in 1 lesion. Can be composed of any combination of: C, L, V, AV[a]	Channel-type or truncal malformations	Klippel-Trénaunay syndrome Parkes Weber syndrome Servelle-Martorell syndrome Sturge-Weber syndrome Limb CM + limb hypertrophy Maffucci syndrome Macrocephaly–CM Microcephaly–CM CLOVES syndrome Proteus syndrome Bannayan-Riley-Ruvalcaba syndrome

Abbreviations: AV, arteriovenous; C, capillary; CLOVES, congenital, lipomatous, overgrowth, vascular malformations, epidermal nevi, and spinal/skeletal anomalies and/or scoliosis; CM, capillary malformation; L, lymphatic; M, malformation; V, venous.

[a] High-flow lesions.