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ADVANCES IN PEDIATRICS

Vascular Anomalies in Pediatrics

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Keywords

• Hemangioma • Vascular anomaly • Vascular malformation • Venous malformation
• Lymphatic malformation • Arteriovenous malformation
• Kaposiform hemangioendothelioma

Key points

- A standardized classification system allows improvements in diagnostic accuracy.
- Vascular anomalies are divided into two groups: vascular tumors and vascular malformations.
- Multidisciplinary vascular anomaly centers combine medical, surgical, radiologic, and pathologic expertise.

INTRODUCTION

For centuries, vascular birthmarks were referred to by vernacular names derived from the belief that a mother's emotions can affect her child's appearance. The modern era of vascular anomalies started in 1982 when Mulliken and Glowacki divided the field into hemangiomas and malformations [1]. The classification was refined to tumors and malformations in 1996 and updated by the International Society for the Study of Vascular Anomalies in 2014 (Tables 1 and 2) [2]. Vascular anomalies are confusing due to imprecise terminology and similar appearance of distinct lesions [3]. Vascular anomalies affect the endothelium of capillaries, arteries, veins, or lymphatics. The estimated prevalence is 4.5% [3,4]. Vascular tumors are characterized by endothelial hyperplasia. This group consists of hemangiomas and less common pediatric tumors. Vascular malformations occur due to errors in morphogenesis and exhibit normal endothelial turnover. The current classification system provides a clinically useful method for diagnosis, prognosis, and treatment of

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Table 1
Vascular tumors

Benign	Locally aggressive or borderline	Malignant
IH	KHE	Angiosarcoma
CH	—	—
Rapidly involuting		
Noninvoluting		
Partially involuting		
Tufted angioma	—	—

vascular anomalies. An accurate diagnosis is made in 90% of infants and children with history and physical examination. Vascular anomalies are often apparent in infancy as the skin and soft tissues are affected. More and more, vascular anomalies are diagnosed antenatally due to advances in fetal medicine. Pediatricians may be called on to assist with the diagnosis of a vascular anomaly.

VASCULAR TUMORS

Vascular tumors are divided into 3 groups: benign, locally aggressive, and malignant [2]. Malignant vascular tumors is beyond the scope of this text.

Benign vascular tumors

The term, *hemangioma*, has been indiscriminately applied to many different types of vascular lesions. In general, there are 2 types of hemangioma: infantile and congenital.

Infantile hemangioma

Clinical features. Infantile hemangioma (IH) is the most common tumor of infancy and childhood, occurring in approximately 4% of children [5]. The incidence is lower in dark-skinned infants. Increased risk factors for IH include

Table 2
Vascular malformations

Simple	Combined	Associated with other anomalies
CM	CM + VM	Klippel-Trenaunay syndrome
	CM + LM	
	CM + AVM	
LM	LM + VM	CLOVES syndrome
	CM + LM + VM	
VM	—	Parkes Weber syndrome
AVM	CM + LM + VM + AVM	—
	CM + VM + AVM	
Arteriovenous fistula	—	—