Vascular lesions, malformations, anomalies, tumors, and hemangiomas: what they are, what to do, when to worry, when to refer

What is a Vascular Anomaly?

Scott A. Norton, MD, MPH

Propranolol for uncomplicated infantile hemangiomas

A. Yasmine Kirkorian, MD

Congenital vascular syndromes: diagnostic role of a multidisciplinary clinic Kalyani Marathe, MD, MPH

Complex vascular anomalies: therapeutic role of a multidisciplinary clinic Nancy Bauman, MD (Otolaryngology)

Lasers to treat vascular lesions

Iris Rubin, MD



What is a Vascular Anomaly?

Scott A. Norton, MD, MPH

Chief of Dermatology, CNHS

What exactly is a Vascular Anomaly?

What's the difference between a vascular malformation and a vascular tumor?

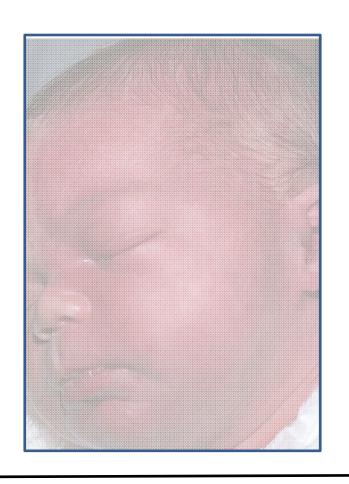
And why does this matter so much?

No financial conflicts of interest.

What is a Vascular Anomaly?

Malformation vs Tumor

(Port-wine stain vs Hemangioma)

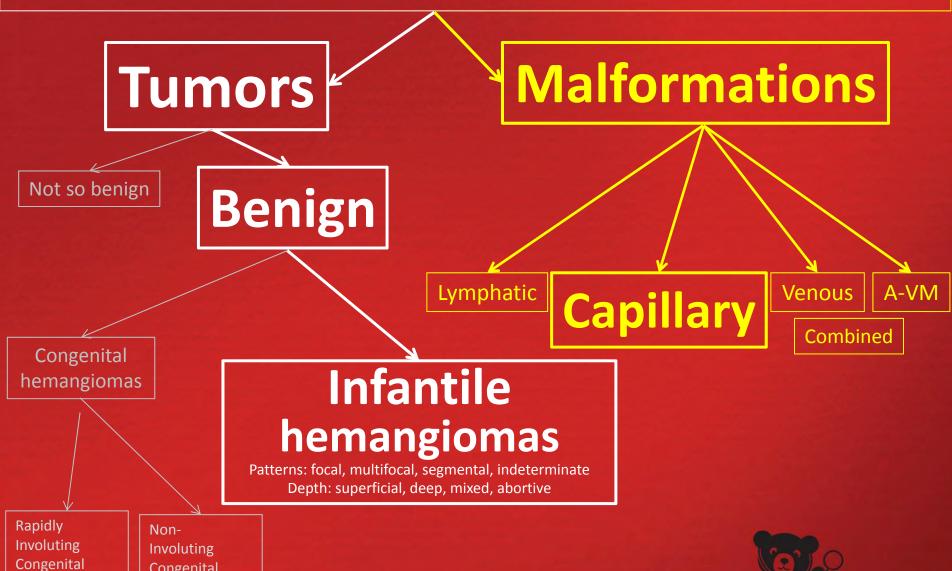




Syndromic vs Non-syndromic Treatable vs Not treatable

Differences: Tumors (hemangiomas) vs malformations

Why does it matter so much?



Hemangiomas

Congenital Hemangiomas



International Society for the Study of Vascular Anomalies (ISSVA) Classification

	Vascular a	Vascular anomalies				
Vascular tumors	Vascular malformations					
	Simple	Combined °	of major named vessels	associated with other anomalies		
Benign Locally aggressive or borderline Malignant	Capillary malformations Lymphatic malformations Venous malformations Arteriovenous malformations* Arteriovenous fistula*	CVM, CLM LVM, CLVM CAVM* CLAVM* others	See details	See list		

Tumors

Not so benign

Benign

Congenital hemangiomas

Rapidly Involuting Congenital Hemangiomas

Non-Involuting Congenital Hemangiomas

Infantile hemangiomas

Patterns: focal, multifocal, segmental, indeterminate

Depth: superficial, deep, mixed, abortive



International Society for the Study of Vascular Anomalies (ISSVA) Classification

Benign vascular tumors
Infantile hemangioma / Hemangioma of infancy
Congenital hemangioma
Rapidly involuting (RICH) *
Non-involuting (NICH)
Pyogenic granuloma (aka lobular capillary hemangioma)
Others
Locally aggressive or Malignant vascular tumors
Kaposiform hemangioendothelioma
Angiosarcoma
Epithelioid hemangioendothelioma
Others

Infantile Hemangiomas (ISSVA Classification)

Pattern

- focal
- multifocal
- segmental
- indeterminate

Different types

- superficial
- deep
- mixed (superficial + deep)
- reticular / abortive / minimal growth
- others

As	ssociation with other lesions
PHACE association / syndrome	Posterior fossa malformations, Hemangioma, Arterial anomalies, Cardiovascular anomalies, Eye anomalies, sternal clefting and/or supraumbilical raphe
LUMBAR (SACRAL, PELVIS) association / syndrome	Lower body hemangioma, Urogenital anomalies, Ulceration, Myelopathy, Bony deformities, Anorectal malformations, Arterial anomalies, and Renal anomalies

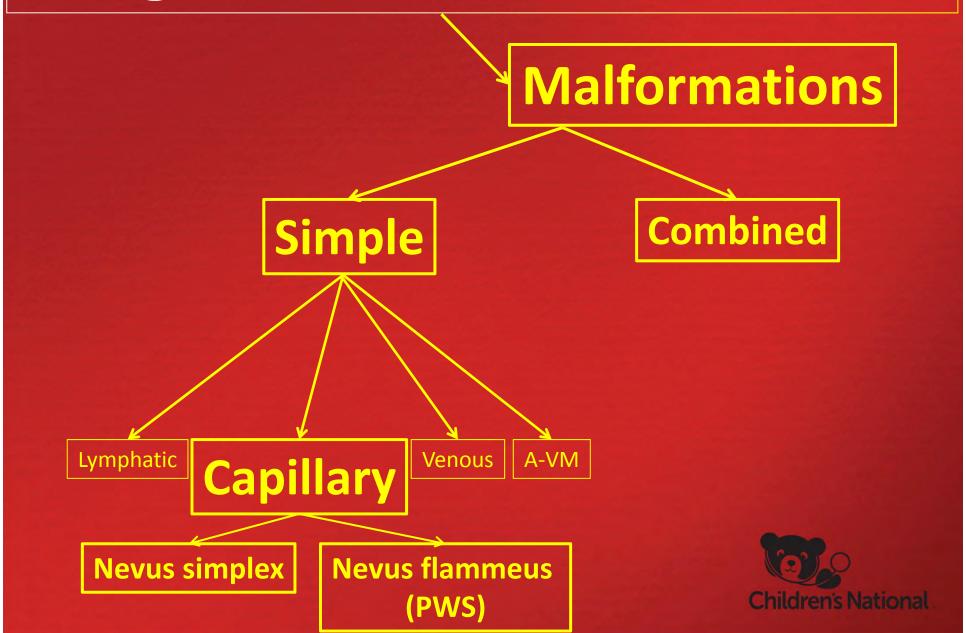
Potential Complications of Infantile Hemangiomas

- Ulcers
- Disfigurement
- Cervicofacial ("beard") hemangiomas can be a marker for subglottic and upper airway IH
- Orbital IH

 visual impairment, amblyopia
- Airway obstruction
- Association with underlying syndromes
 - Typically with segmental IH











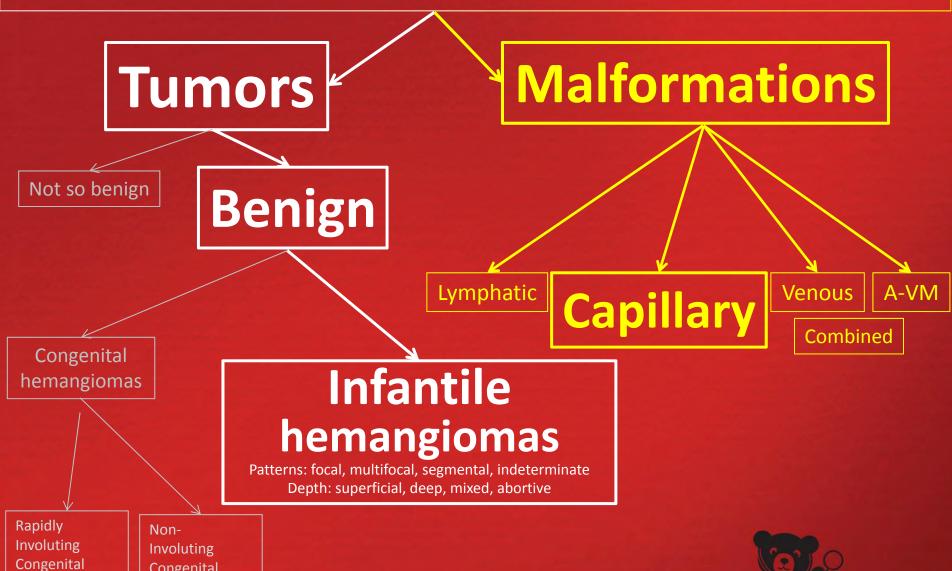


Capillary malformations and genetic correlates

Capillary malformations (CM)	
Cutaneous and/or mucosal CM (aka "port-wine" stain)	GNAQ
CM with bone and/or soft tissue hyperplasia	
CM with CNS and/or ocular anomalies (Sturge-Weber syndrome)	GNAQ
CM of CM-AVM	RASA1
Telangiectasia	
Hereditary hemorrhagic telangiectasia (HHT)	
HHT1	ENG
HHT2	ACVRL1
HHT3	
JPHT (juvenile polyposis hemorrhagic telangiectasia)	SMAD4
Others	
Cutis marmorata telangiectatica congenita (CMTC)	
Nevus simplex / Salmon patch	
Others	

Other vascular malformations & genetic correlates

Vascular malformations associated with other anomalies				
Klippel-Trenaunay syndrome				
Parkes Weber syndrome	RASA1			
Servelle-Martorell syndrome				
Sturge-Weber syndrome	GNAQ			
Limb CM + congenital non-progressive limb overgrowt	h			
Maffucci syndrome				
Macrocephaly - CM (M-CM or MCAP)	PIK3CA			
Microcephaly - CM (MICCAP)	STAMBP			
CLOVES syndrome	PIK3CA			
Proteus syndrome	AKT1			
Bannayan-Riley-Ruvalcaba syndrome	PTEN			



Hemangiomas

Congenital Hemangiomas



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Complex vascular anomalies: Therapeutic role of multidisciplinary VAC TBD

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