Vascular Anomalies:  
State of the Art Diagnosis and Treatment 2013

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Objectives

- Be able to distinguish between hemangiomas and vascular malformations
- Become familiar with appropriate diagnostic imaging
- Understand multidisciplinary approach and roles for medical therapy, sclerotherapy, and surgical intervention
- Recognize that increased understanding of the genetics and biology of these lesions are leading to new potential medical therapies and new clinical trials

Common Misconceptions

- All vascular birthmarks are not “hemangiomas”
- All vascular malformations are not “AVMs”
- 50% of patients referred to the UCSF Vascular Anomalies Center carry an incorrect diagnosis
Diagnosis of Vascular Anomalies

- **More important**
  - age of patient
  - physical examination
  - history (esp. birth/childhood/recent activity)

- **Less important**
  - Imaging
  - Biopsy
Hemangioma

- Period of most rapid growth is between 4-6 weeks
- 80% of growth is done by 3-4 months
- Proliferate over ~6-9 months
  - deep & segmental IH grow longer
- Slowly involute over years

Natural History
Focal vs Segmental

Propranolol for IH

- Risks/Side Effects
  - Bradycardia
  - Hypotension
  - Hypoglycemia
  - Sleep disturbance
  - Hyperkalemia
  - Bronchospasm/exacerbation of asthma
UCSF Propranolol Protocol

Suspension is 4mg/ml
0.15mg/kg TID for 2 days
0.30mg/kg TID for 3 days
0.45 mg/kg TID for 3 days
0.60mg/kg TID is target dose
<6mo should be fed Q 4 h
hold dose for pulse<120

Consensus Statement

Vascular Anomalies: Classification Clinical and Cellular Differences

Vascular Malformation
- Malformed blood vessels
- Present at birth
- Commensurate growth
- F/M: 1/1
- nl endothelial turnover
- nl FGF
- No GLUT 1 staining

Venous Malformation
Venous Malformation: Imaging

discreet/scattered lesion
T1: intermediate signal
T2: high signal
enhancement: homogeneous
  • no flow voids
can involve muscle/bone
  + phleboliths

Venous Malformation: Therapy

• Observation/Education
• Compressive stocking
• ASA/Lovenox (LIC) (check D-dimers and fibrinogen)
• Laser
• Surgery
• Sclerotherapy
• Rapamycin (MTOR)
Lymphatic Malformation

• Classification Systems
  – Microcystic vs. Macro
  – DeSerres
    • Suprahyoid vs. infra
    • Uni vs. Bi
Lymphatic Malformation:
Imaging

- macro- vs. micro-cystic
- multiple cysts
- T1: low (water) signal, unless prior hemorrhage, Rx
- T2: high (water) signal
- fluid-fluid layers typical
Lymphatic Malformation: Therapy

- Observation/Education
- Compressive stocking
- Manual lymphatic drainage
- Surgery
- Sclerotherapy
- Propranolol
- Sildenafil
- Rapamycin

Vascular Lesions: Known Genetic Markers

- Capillary and AVM
  - RASA-1
- Lymphatic Malformations
  - VEGFR3
- Venous Malformations
  - TIE2/TEK
- Hemangiomas of Infancy
  - GLUT1
- Hamartomas and AVM
  - PTEN
Pathways involved in vascular anomalies


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Summary

- Vascular anomalies classifications are useful for prognosis and treatment planning
- History and PE are most important elements for accurate diagnosis
- Education about the natural history of the lesion is important for the patient/family
- Imaging can be helpful to differentiate between LM and VM and determine tissue involved “tip of the iceberg”
- Patients with complex lesions benefit from a multidisciplinary approach in a Vascular Anomalies Center
- New clinical trials are available based on ongoing discoveries and identification of therapeutic targets