Vascular Anomalies: State of the Art Diagnosis and Treatment 2013

Kristina W. Rosbe MD
Director, Pediatric Otolaryngology
Consultant, UCSF Vascular Anomalies & Birthmark Center (BVAC)

Department of Otolaryngology-Head and Neck Surgery
University of California-San Francisco

I have no financial disclosures

Objectives

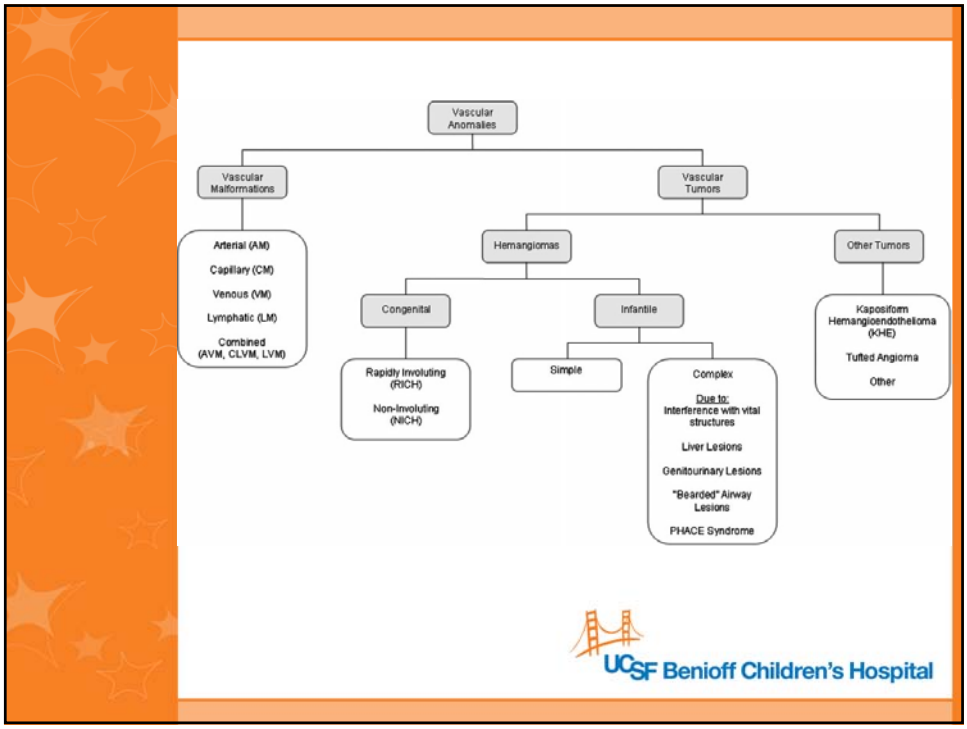
- Be able to distinguish between hemangiomas and vascular malformations
- Become familiar with appropriate diagnostic imaging
- Understand multidisciplinary approach and roles for medical therapy, sclerotherapy, and surgical intervention
- Recognize that increased understanding of the genetics and biology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

UCSF Benioff Children's Hospital

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

UCSF Benioff Children's Hospital

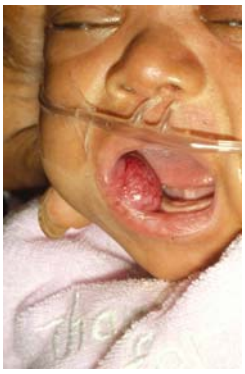


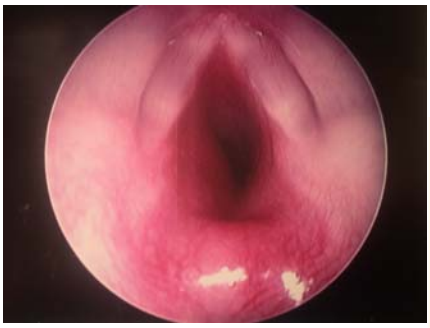
Diagnosis of Vascular Anomalies

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

UCSF Benioff Children's Hospital

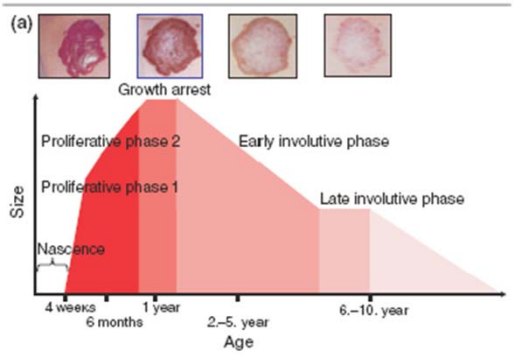
Hemangioma





UCSF Benioff Children's Hospital

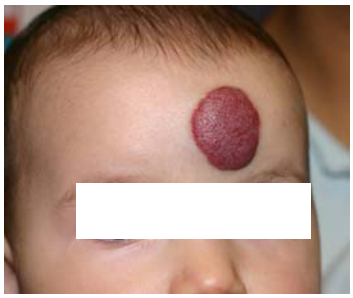
Natural History

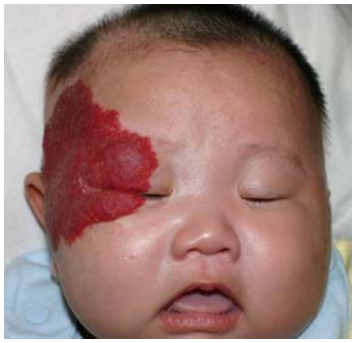


- Period of most rapid growth is between 4-6 weeks
- 80% of growth is done by 3-4 months
- Proliferate over ~6-9 months
  - –deep & segmental IH grow longer
- Slowly involute over years

UCSF Benioff Children's Hospital

Focal vs Segmental





UCSF Benioff Children's Hospital

Propranolol for IH

- **Risks/Side Effects**
  - *Bradycardia*
  - *Hypotension*
  - *Hypoglycemia*
  - *Sleep disturbance*
  - *Hyperkalemia*
  - *Bronchospasm/exacerbation of asthma*

UCSF Benioff Children's Hospital

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

UCSF Benioff Children's Hospital

Consensus Statement

Drolet BA, Frommelt PC, Chamlin SL, et al. Initiation and use of propranolol for infantile hemangioma: report of a consensus conference. *Pediatrics* 2013;131:128-40.

UCSF Benioff Children's Hospital

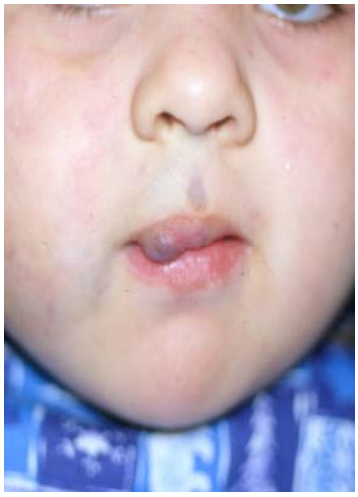
Vascular Anomalies: Classification Clinical and Cellular Differences

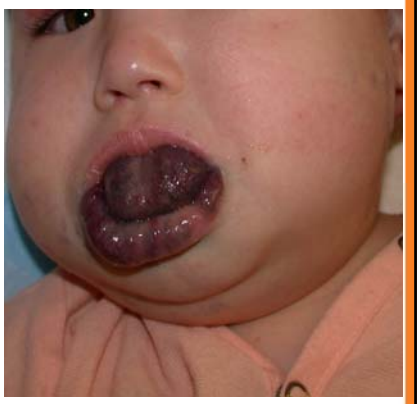
Vascular Malformation

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

UCSF Benioff Children's Hospital

Venous Malformation

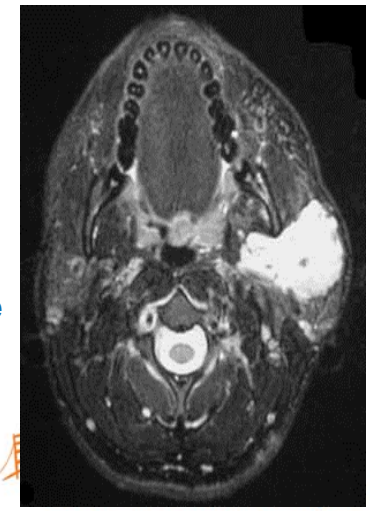




UCSF Benioff Children's Hospital

Venous Malformation: Imaging

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

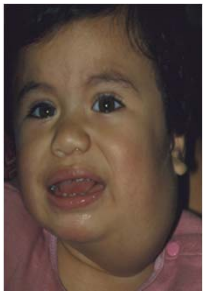
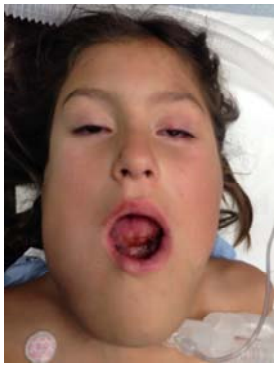


Venous Malformation: Therapy

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

UCSF Benioff Children's Hospital

Lymphatic Malformation


UCSF Benioff Children's Hospital

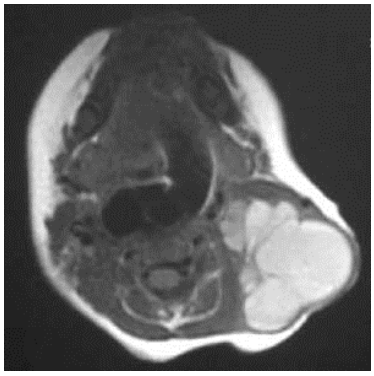
Lymphatic Malformation

- **Classification Systems**
  - *Microcystic vs. Macro*
  - *DeSerres*
    - Suprahyoid vs. infra
    - Uni vs. Bi

UCSF Benioff Children's Hospital

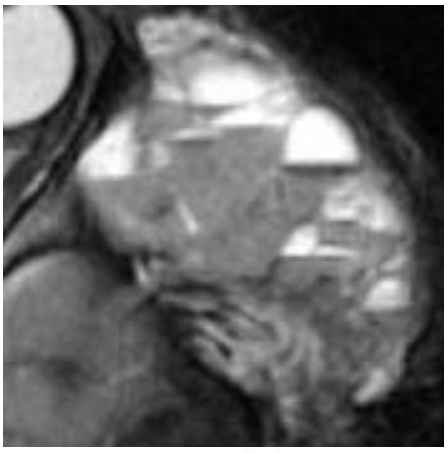
Lymphatic Malformation: Imaging

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



UCSF Benioff Children's Hospital

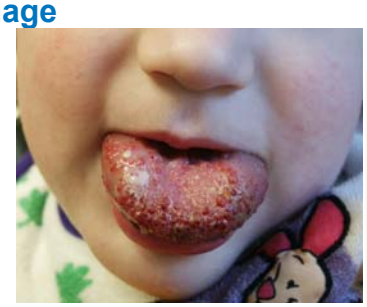
Imaging



UCSF Benioff Children's Hospital

Lymphatic Malformation: Therapy

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



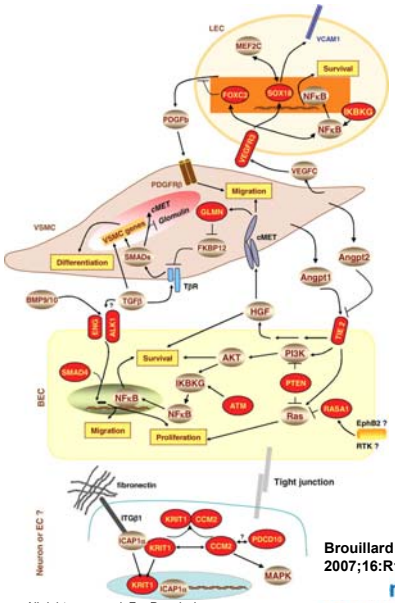
UCSF Benioff Children's Hospital

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*

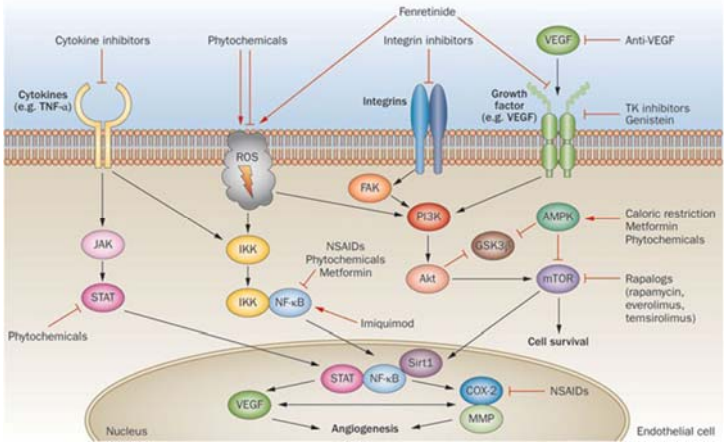
UCSF Benioff Children's Hospital

Pathways involved in vascular anomalies



Brouillard P , and Vikkula M Hum. Mol. Genet. 2007;16:R140-R149

nioff Children's Hospital



UCSF Benioff Children's Hospital

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

UCSF Benioff Children's Hospital

UCSF Birthmark and Vascular Anomalies Center 1991-2013

UCSF Benioff Children's Hospital