VASCULAR ANOMALIES IN CHILDREN

Charleston Pediatric ENT Update
Mar 23, 2024

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MEDICAL UNIVERSITY of SOUTH CAROLINA

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Disclosures

- None

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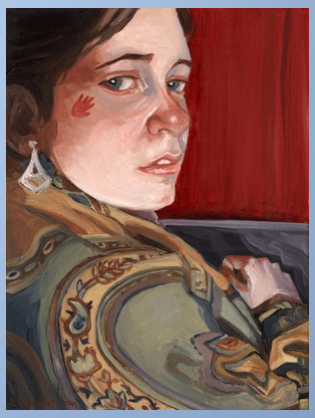
Historic Perspective:

Folklore: "maternal impressions"

- Mother's emotions could be imprinted on her child.
- Pregnant women were warned not to look upon scenes of accidents or slaughtering of animals.
- Mostly negative connotation implying impure thoughts
- Disproved in the 1860's



Nathaniel Hawthorne "The Birthmark"

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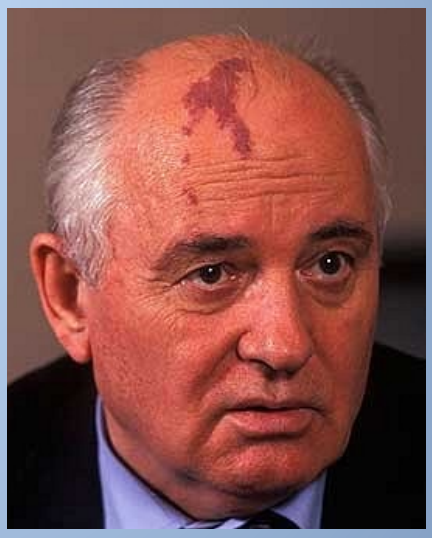
Historic figures

Then …

Harvard President Charles Eliot, mid 1800's

Now…



Soviet Union President Mikhail Gorbachev, late 1990's

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Positive Exposure

https://positiveexposure.org/frame/

- FRAME
- Faces Redefining the Art of Medical Education

- Video…

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Classification

- Multiple confusing terminologies used to describe vascular anomalies:
  - Cavernous hemangiomas
  - Strawberry hemangiomas
  - Portwine stains
  - Cystic hygromas

Until….

- J.B. Mulliken and J. Glowacki

Hemangiomas and Vascular Malformations in Infants and Children: A Classification based on Endothelial Characteristics. *Plastic and Reconstructive Surgery* 1982, 69: 412-422

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Mulliken and Glowacki

- Classification based on clinical and histologic characteristics of the lesions.
- Defined two categories:

Hemangiomas	Malformations
n=26	n=23
Endothelial cell proliferation	Normal endothelial cell cycle
40% present at birth	90% recognized at birth
Rapid postnatal growth and slow involution	Grow commensurately with child

International Society of the Study of Vascular Anomalies (ISSVA)- 1996

Vascular Tumors	Vascular Malformations
<u>Benign</u>	<u>Slow-flow</u>
• Infantile Hemangiomas	• Capillary malformation

• Congenital Hemangiomas
  • NICH
  • RICH
	• Venous malformation
	• Lymphatic malformation

• Hemangioendotheliomas • Kaposiform, Spindle cell, Epithelioid, other rare…	<u>Fast-flow</u> • Arterial malformation
• Dermatologic acquired • Pyogenic granuloma	• Arteriovenous malformation
<u>Malignant</u> • Angiosarcoma, Kaposi's sarcoma, Malignant angioendotheliomas…	• Arteriovenous fistulas Combined vascular malformations

NICH- noninvoluting congenital hemangioma; RICH- rapidly involuting congenital hemangioma

Old Nomenclature to New

HEMANGIOMA	Old Terminology	MALFORMATION
	Capillary	
HEMANGIOMA	Strawberry	
	Capillary-cavernous	
	Portwine	CAPILLARY
	Cavernous	VENOUS
	Venous	
	Cystic hygroma	LYMPHATIC
	Lymphangioma	
	Arteriovenous	ARTERIOVENOUS

Vascular Birthmarks Hemangiomas and Malformations. Mulliken, JB and Young AE WB Saunders Co., Philardelphia, 1988. p32.

Classification

- Classification based on clinical and histologic characteristics of the lesions
- Histology:
  - Vascular Tumors
    - Increased endothelial turnover or PROLIFERATION
  - Vascular Malformations
    - Normal endothelial turnover
    - ABNORMAL vascular MORPHOLOGY

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Malignant • Angiosarcoma, Kaposi's sarcoma, Malignant angioendotheliomas…	• Arterial malformation
	• Arteriovenous malformation
	• Arteriovenous fistulas
	Combined vascular malformations

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ISSVA 2014:

Vascular Anomalies					
Vascular Tumors	Vascular Malformations				
	Simple	Combined	of major named vessels	associated with other anomalies	
Benign Vascular Tumors:	Capillary (C) malformations	Defined as 2 or more vascular malformations identified in 1 lesion. Can be composed of any combination of: C, L, V, AV*	"channel type" or "truncal" malformations	Klippel-Trenaunay synd.	
Infantile hemangioma				Parkes Weber synd.	
Congenital hemangioma:	Lymphatic (L) malformations			Servelle-Martorell synd.	
Rapidly involuting (RICH)*				Sturge-Weber synd.	
Non-involving (NICH)	Venous (V) malformations			Limb CM + limb hypertrophy	
Partially involuting (PICH)				Maffucci synd.	
Tufted angioma	Arteriovenous (AV) malformations*			Macrocephaly - CM	
Spindle-cell hemangioma				Microcephaly-CM	
Epithelioid hemangioma	Arteriovenous fistula*			CLOVES synd.	
Pyogenic granuloma (aka lobular capillary hemangioma)				Proteus synd.	
Others				Bannayan-Riley-Ruvalcaba	
Locally aggressive or borderline vascular tumors:					
Kaposiform hemangioendothelioma					
Retiform hemangioendothelioma					
Papillary intralymphatic angioendothelioma (PILA),Dabska					
Composite hemangioendothelioma					
Kaposi sarcoma					

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TAKE HOME MESSAGE

- TREATMENT for vascular anomalies depends upon diagnosis.
- "Hemangioma" was incorrectly used in 71.3% (228 of 320 articles). Patients with lesions mislabeled as hemangioma were more likely to receive improper treatment
- History and Physical exam can accurately diagnose **90%** of vascular anomalies

Hassanein, AH., et al. *Evaluation of Terminology for Vascular Anomalies in Current Literature*. PRS Journal. Jan 2011. vol.127: no.1347-351.

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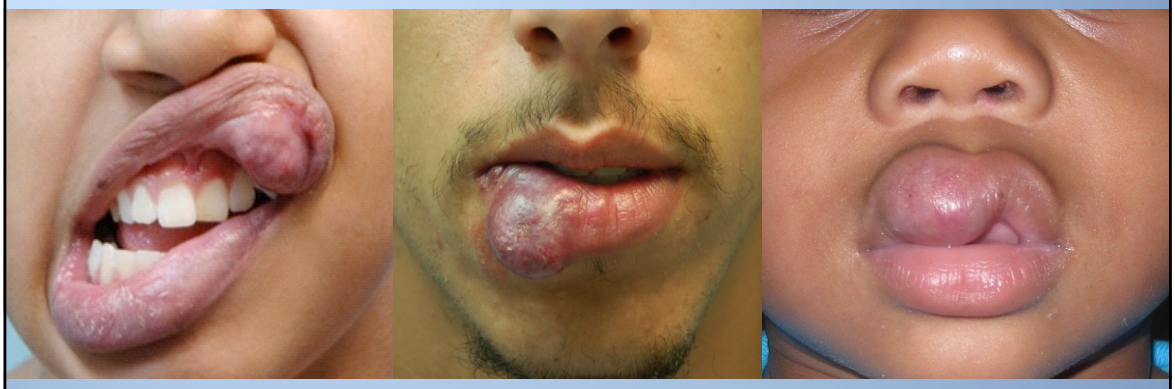
Diagnosis?



Venous Malformation

Arteriovenous Malformation

Hemangioma

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International Society of the Study of Vascular Anomalies (ISSVA)

Vascular Tumors
Benign
• Infantile Hemangiomas

• Congenital Hemangiomas
  • NICH
  • RICH

• Hemangioendotheliomas   • Kaposiform, Spindle cell, Epithelioid, Tufted angioma…
• Dermatologic acquired   • Pyogenic granuloma
Malignant   • Angiosarcoma, Kaposi's sarcoma, Malignant angioendotheliomas…

NICH- noninvoluting congenital hemangioma; RICH- rapidly involuting congenital hemangioma

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Infantile Hemangiomas

- Most common benign vascular tumor of infancy
- Incidence by 1 year of age - **10%**
- Female : Male ratio - **3 : 1**
- Distribution
  - Head and neck- 60%
  - Trunk- 25%
  - Extremities- 15%
- 80% Single, 20% Multiple
- Subglottic hemangiomas become symptomatic at 4- 12 weeks of life and 50% have a cutaneous hemangioma with beard distribution
- Increased frequency with Premature infants <1200g - 23% incidence

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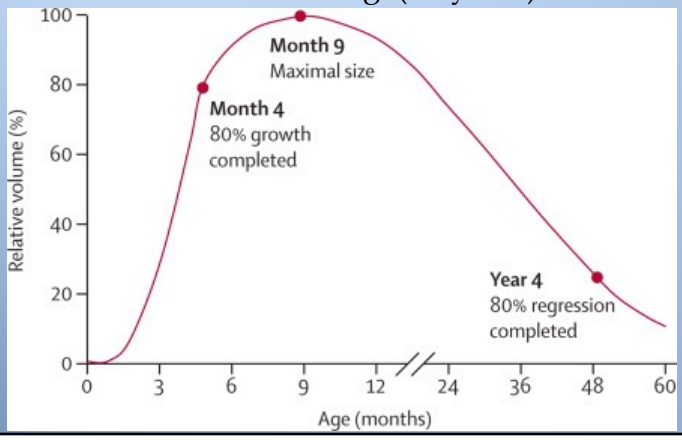
Infantile Hemangiomas

- GLUT-1 Positive
(+ in placental endothelium and infantile hemangiomas)

- 3 Stages of Clinical Presentation:
  - Proliferative Stage (2 weeks - 12 months) May affect vital organs
  - Involuting Stage (1- 5 years)
  - Involuted Stage(>5 years)



Infantile haemangioma.
Christine Léauté-Labrèze, John I Harper, Peter H Hoeger. *The Lancet*. 2017, Available online 13 January 2017
http://dx.doi.org/10.1016/S0140-6736(16)00645-0

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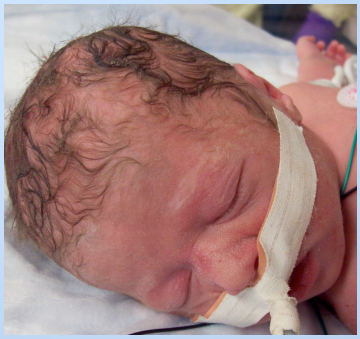
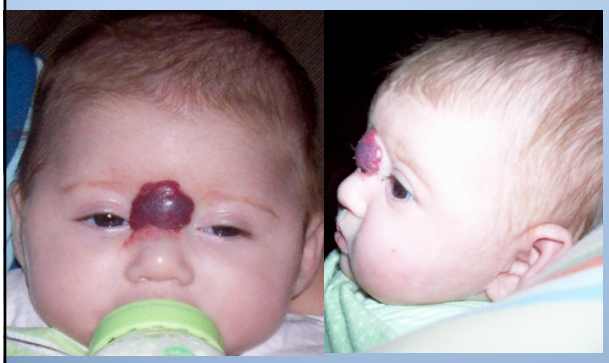
INFANTILE HEMANGIOMA

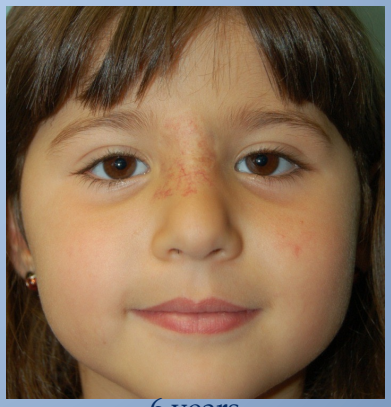


Birth

3 months



6 months



6 years

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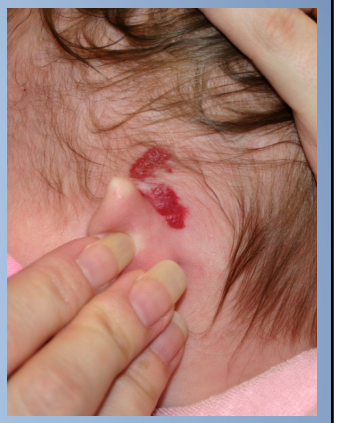
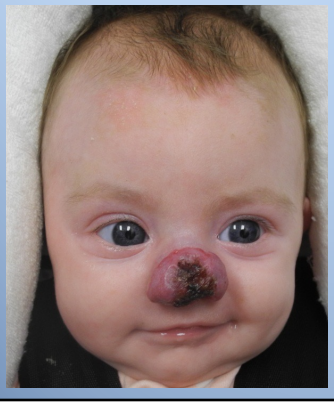
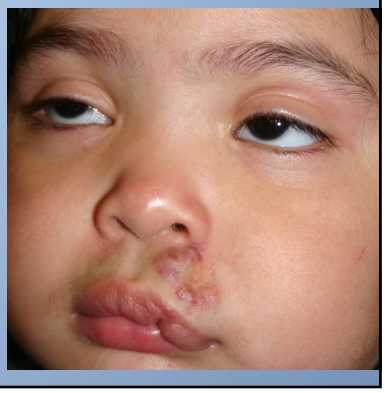
Infantile Hemangiomas

- Superficial
- Deep
- Compound







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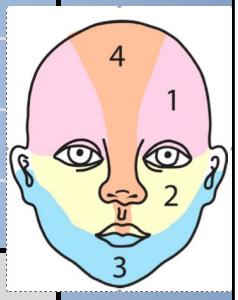
ISSVA 2014:

Infantile Hemangioma	
Pattern	**Types**
focal	superficial
multifocal	mixed/compund
segmental	deep
indeterminate	reticular
Association with other lesions:	
PHACE association:	
Posterior fossa malformations, Hemangioma, Arterial anomalies, Cardiovascular anomalies, Eye anomalies, sternal clefting and/or supraumbilical raphe	
LUMBAR association:	
Lower body hemangioma, Urogenital anomalies, Ulceration, Myelopathy, Bony deformities, Anorectal malformations, Arterial anomalies, and Renal anomalies	



RISK:

⇧ Forehead

⇩ Maxilla

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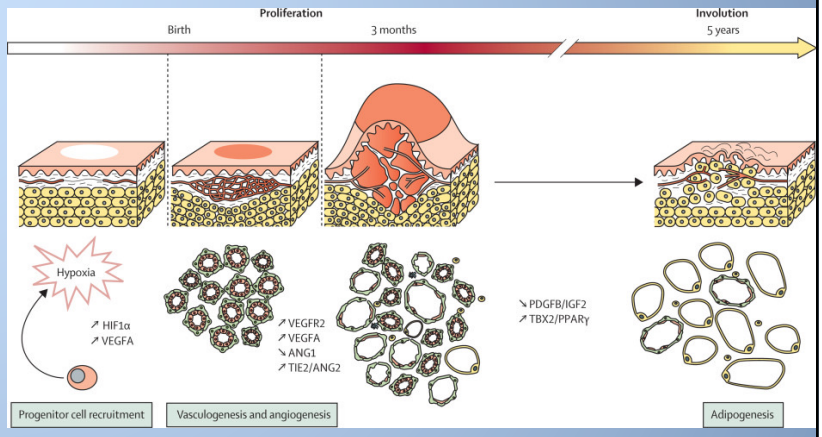
Pathogenesis:

- Hypoxia event
- ↑ HIF 1α
- ↑ VEGF
- ↑ angiogenesis



HIF= hypoxia inducible factor

Infantile haemangioma.
Christine Léauté-Labrèze, John I Harper, Peter H Hoeger. *The Lancet*. 2017, Available online 13 January 2017
http://dx.doi.org/10.1016/S0140-6736(16)00645-0

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Treatment Options

- Observation
- Steroid (oral or local injection)
  - 2-3 mg/kg/day divided
- Propranolol
  - 2-4mg/kg/day divided BID,TID or QID
- Laser (PDL)
- Surgical excision
  - Serial
  - Complete

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Treatment options:

- Propranolol
  - Non-selective Beta-blocker
  - Landmark publication NEJM 2008 Leaute-Labreze et al.
  - Incidental findings of hemangioma resolution in infants on propranolol
  - MARCH 2014:

PROPRANOLOL (HEAMNGEOL™)

IS NOW **FDA** APPROVED FOR TREATMENT OF HEMAGIOMAS

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Hemangeol™

- Brand name(Pierre Fabre) for FDA approved propranolol hydrochloride
- FDA approved in MARCH 2014

- BID Dosing
- 3.4 mg/kg in 2 divided doses daily for 6 months chosen as the optimal treatment regimen in double-blind clinical trial
- Dosing Calculator for determining the appropriate dose for your patients
- Unique graduated syringe with bottle adapter for convenient and safe dosing by parents and caregivers
- Delivers to your home

http://www.hemangeol.com/hcp/

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Initiation and Use of Propranolol for Infantile Hemangioma: Report of a Consensus Conference.

Drolet BA, et al. Pediatrics. 2013 Jan: 131(1):128-140.

- PROPRANOLOL
  - Used over 40 yrs in pediatric population for cardiovascular disease
  - Only death related events with IV administered med and over-dosage
  - Oral propranolol has favorable safety profile

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Initiation and Use of Propranolol for Infantile Hemangioma: Report of a Consensus Conference.

Drolet BA, et al. Pediatrics. 2013 Jan: 131(1):128-140.

PROPRANOLOL WORKUP AND INITIATION:

- EKG
  - No consensus for healthy patients
  - 81% patients received in work up
  - Yes
    - 1- Heart rate lower than normal for specific age
    - 2- Family history of cardiac anomaly/arrhythmia or connective tissue disorder
    - 3- Patient has a cardiac abnormality
- Vital monitoring
  - Peak effect at 1-3 hrs after oral dose
  - Obtain baseline Heart rate (HR) and Blood Pressure (BP)
  - Measure HR and BP at 1 and 2 hrs after administration
  - HR easier to assess than BP, (abnormal BP is ~ <64mm Hg for newborn-6mo)
  - Repeat monitoring when increasing dose more than 0.5mg/kg/d
- Glucose check
  - No consensus

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Initiation and Use of Propranolol for Infantile Hemangioma: Report of a Consensus Conference.

Drolet BA, et al. Pediatrics. 2013 Jan: 131(1):128-140.

PROPRANOLOL DOSAGE:

- Available solutions 20mg/5ml
- Target dose: 1-3 mg/kg/day divided TID
- Frequency of Daily-
  - TID with minimum of 6 hrs between doses
  - Surgical literature sometimes reports BID
- Taper Up to goal dose

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Complications Reported with Propranolol Use:

Initiation and Use of Propranolol for Infantile Hemangioma: Report of a Consensus Conference.

Drolet BA, et al. Pediatrics. 2013 Jan: 131(1):128-140.

Complications reported	No. of Patients/ Total No. of Patients in Papers Reporting Complication	Frequency (%) of Complication Among Papers Reporting Said Complication	Overall Frequency (%) of Total of 1175 Patients Reviewed in 85 Papers
Asymptomatic hypotension or hypotension (unspecified)	33/228	14.5	2.8
Symptomatic hypotension	3/46	6.5	0.3
Pulmonary symptoms (bronchoconstriction, bronchiolitis, wheezing, pulmonary obstruction, apneic episode)	16/201	8.0	1.4
Hypoglycemia	10/88	11.4	0.9
Asymptomatic bradycardia or bradycardia (unknown)	11/126	8.7	0.9
Symptomatic bradycardia	1/2	50	0.1
Sleep disturbance (including nightmares)	44/326	13.5	3.7
Somnolence	26/220	11.8	2.2
Cool or mottled extremities	20/225	8.9	1.7
Diarrhea	9/53	17.0	0.8
Gastroesophageal reflux disease or gastrointestinal upset	8/133	6.0	0.7

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Side Effects

- Sleep disturbance
- Pulmonary effects (bronchospasm)

- Atenolol (selective Beta 1)
  - May reduce pulmonary adverse reactions
  - Does not cross BBB so may reduce sleep disturbances

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Contraindications for Propranolol Use:

Initiation and Use of Propranolol for Infantile Hemangioma: Report of a Consensus Conference.
Drolet BA, et al. Pediatrics. 2013 Jan: 131(1):128-140.

Contraindications:
Cardiogenic shock
Sinus bradycardia
Hypotension
Greater than first-degree heart block
Heart failure
Bronchial asthma
Hypersensitivity to propranolol hydrochloride

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Initiation and Use of Propranolol for Infantile Hemangioma: Report of a Consensus Conference.

Drolet BA, et al. Pediatrics. 2013 Jan: 131(1):128-140.

- PHACE Association and PROPRANOLOL
  - Much higher risk involved given cardiac anomalies
  - Cardiology consultation (echo…)
  - MRI/MRA of head/neck/chest
  - Consider neurology consultation if imaging suggestive of high risk for stroke
  - 2 patients reported with stroke with PHACE and propranolol
  - Dosing: TID to decrease fluctuation

PHACES: -Suspect with facial segmental hemangioma
- **P**osterior fossa abnormalities, **H**emangioma, **A**rterial abnormalities, **C**ardiac abnormalities, **E**ye abnormalities, **S**ternal defects

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Propranolol + Timolol gtts (0.5 or 0.1% solution)

3 mo age

20 mo age

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Propranolol versus corticosteroids in the treatment of infantile hemangioma: a systematic review and meta-analysis.

Izadpanah A, Izadpanah A, Kanevsky J, Belzile E, Schwarz K. Plast Reconstr Surg. 2013 Mar;131(3):601-13. PMID: 23142941

- Meta-Analysis
- Objective: Compare the efficacy and complications related to treatment of cutaneous hemangiomas with corticosteroids versus propranolol.
- Methods: Meta-analysis of literature 1965-2012. Out of 1162 articles, 40 studies met final inclusion criteria.
- Results: Meta-analysis data suggested that the pooled response rate for corticosteroids was 71% versus the response rate for propranolol was 97%.

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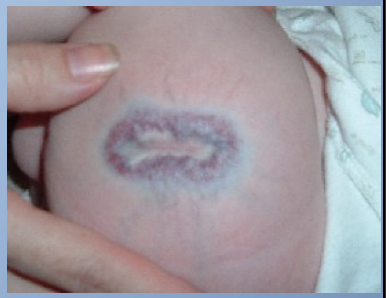
Congenital Hemangiomas

- NICH- noninvoluting congenital hemangioma
- RICH- rapidly involuting congenital hemangioma

- Present at birth
- GLUT-1 negative
- RICH may entirely involute by 1-2 years
- Halo of pallor
- May develop A-V shunting
- Equal sex distribution



http://www.birthmarks.us/case13.htm

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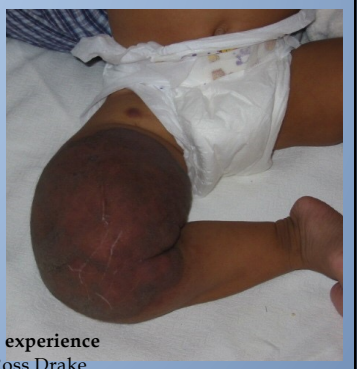
Vascular tumors

Kaposiform Hemangioendotheliomas:

- May be present at birth
- Proliferate into childhood with some regression mid-childhood
- Associated with thrombocytopenia
  - Kasabach- Merritt phenomenon (KMP)
  - <10,000 plt, platelet trapping in the tumor
- Mortality rate can be high (KMP) 14-24%
- CD61 positive
- GLUT-1 and LeY antigen negative
- More common in body than head region
- Appearance- brawny, red-purple, warm tense and edematous



Vincristine for the treatment of Kasabach-Merritt syndrome: recent New Zealand case experience
Kimberley Thomson, Ralph Pinnock, Lochie Teague, Rachel Johnson, Noel Manikkam, Ross Drake
Journal of the New Zealand Medical Association, 16-February-2007, Vol 120 No 1249

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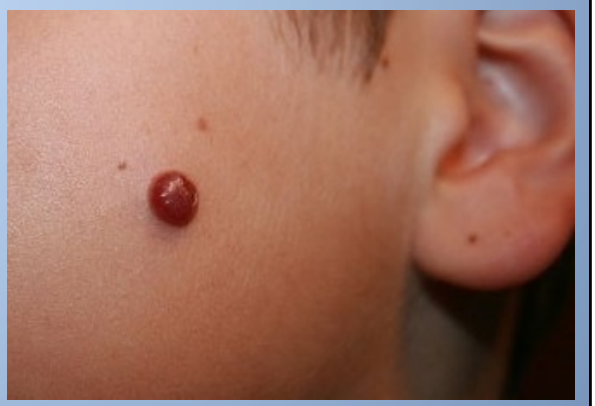
Vascular tumors

Dermatologic acquired- Pyogenic granuloma:

- Common in pediatric population
- Usually on the face
- Small 6.5mm
- Pedunculated
- Can bleed frequently



http://doctorv.ca/cosmetic-services/lump-and-bump-removal/pyogenic-granuloma-detailed-information/

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International Society of the Study of Vascular Anomalies (ISSVA)

Vascular Malformations
Slow-flow
• Capillary malformation
• Venous malformation
• Lymphatic malformation
Fast-flow
• Arterial malformation
• Arteriovenous malformation
• Arteriovenous fistulas
Combined vascular malformations

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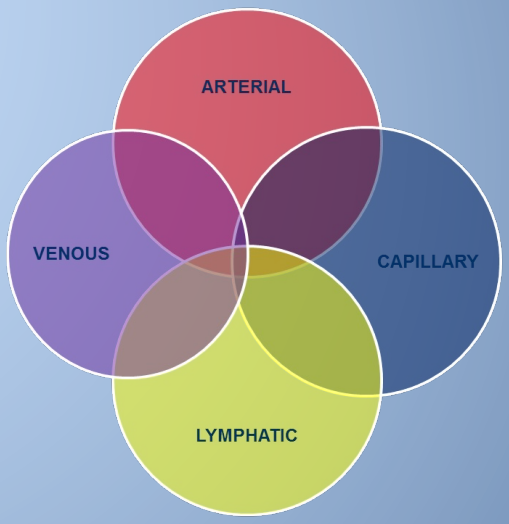
Vascular Malformations

- Normal endothelial turnover
- ABNORMAL vascular MORPHOLOGY
- Present at birth 90%
- Grows commensurately with child



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Capillary Malformation

- Old terminology- Portwine stain
- Equal sex distribution
- Birth prevalence- 0.3%
- Flat, red, sharply demarcated
- Skin becomes more nodular and irregular with age
- If involves forehead distribution distribution- must be evaluated for Sturge Weber syndrome (10-15%)
- TREATMENT- Pulsed Dye LASER
  - Cochrane Review 2011

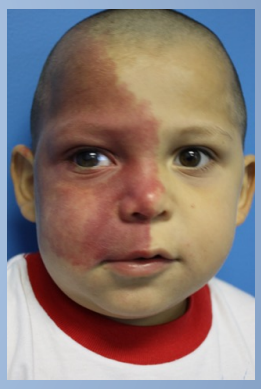

Image provided by Marcelo Hochman

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Man Turns Birthmarks On Arm Into Imaginary World Map

http://geekologie.com

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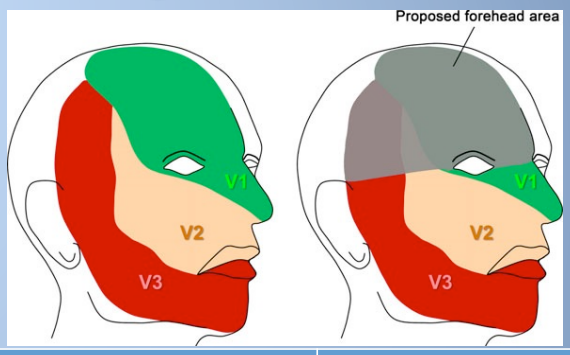
Sturge Weber Syndrome (encephalofacial angiomatosis)

- Facial + leptomeningeal capillary malformations
- Eye anomalies (cataracts)
- Bone/ soft tissue overgrowth
- Developmental delay
- Sporadic (somatic mutation GNAQ)
- 1in 20,000-50,000 live births



Trigeminal nerve distribution of PWS	PWS Patients, n (%)
V1 alone	25 (14.6)
V2 alone	21 (12.3)
V3 alone	9 (5.3)
V1 and V2	63 (36.8)
V1 and V3	2 (1.2)
V2 and V3	10 (5.8)
V1, V2 and V3	41 (24.0)

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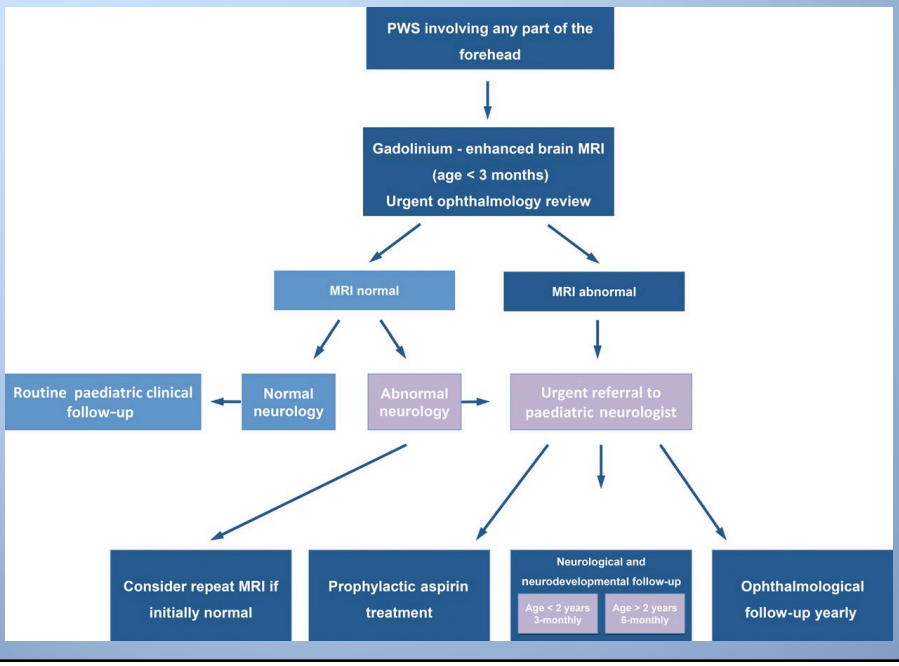
New vascular classification of port-wine stains: Improving prediction of Sturge-Weber risk.

Waelchli, R & E. Aylett, S & Robinson, K & Chong, K.W. & Martinez, A.E. & Kinsler, V.A.. (2014).
British Journal of Dermatology. 171. . 10.1111/bjd.13203.



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Venous Malformation

- Most common type of vascular malformation
- Incidence- 1- 4%
- Present at birth
- Usually unifocal
- Bluish, soft, compressible, non-pulsatile
- Bulge with Valsalva maneuver
- Phlebothrombosis is common

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Treatment Options

- Aspirin
- Sclerotherapy
  - Improved ethanol products
    - decreased toxicity risk
  - Bleomycin
    - contraindicated with pulmonary or renal impairments
  - Doxycycline
- Surgical excision
  - Elevated D-dimers
    - identified in some advanced venous malformations and my increase risk of intraoperative bleeding/coagulopathy
  - Use of low-molecular weight heparin

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Lymphatic Malformations

- Radiologically and Histologically characterized as:
  - Microcystic
  - Macrocystic
  - Combined
- May be present at birth- usually within 2 y.o.
- Skin has bluish hue, soft, non-pulsatile, ballotable
- Can have underlying skeletal overgrowth
- May fluctuate in size from acute illness or intra-lesional hemorrhage, & may become infected.

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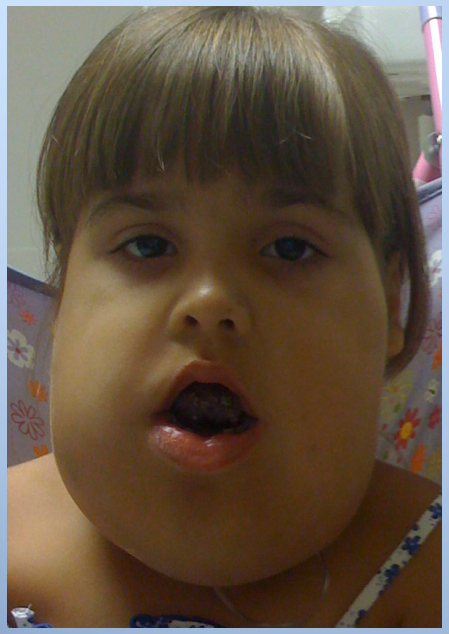
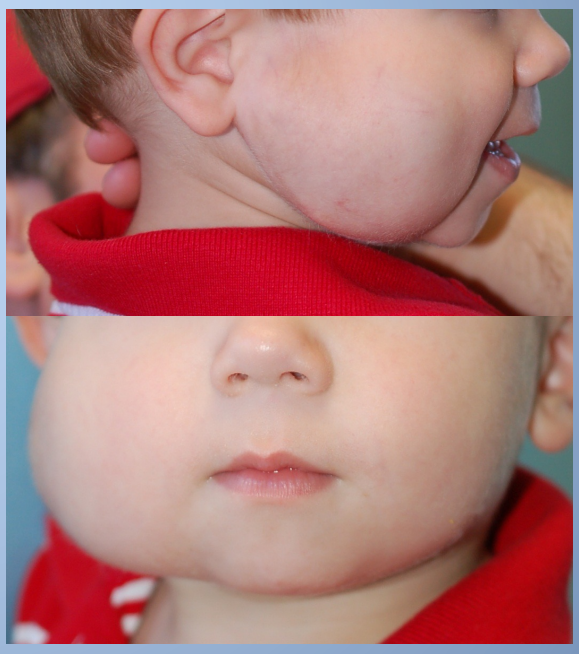
Lymphatic Malformations





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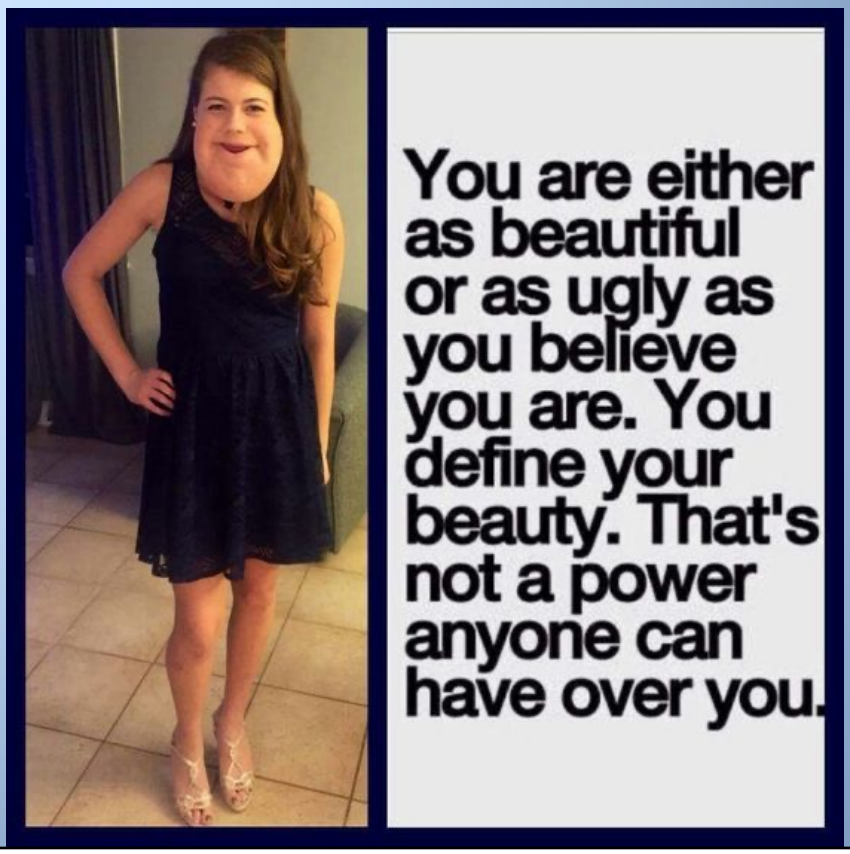

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Treatment Options

- Sclerotherapy (Macrocystic)
  - Doxycycline
  - OK 432 (Picibanil)
- Surgical excision
- CO_2 Laser

- Others:
  - Rapamycin (sirolimus)-
    - Immunosuppressant
    - Phase II trial in Cincinnati Children's Hospital- complete
  - Sildenafil (Viagra)
    - Phase II trial in Stanford- in progress
    - FDA warning with use in children

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Efficacy and Safety of Sirolimus in the Treatment of Complicated Vascular Anomalies

Pediatrics · January 2016

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BACKGROUND AND OBJECTIVES: Complicated vascular anomalies have limited therapeutic options abstract
and cause significant morbidity and mortality. This Phase II trial enrolled patients with complicated vascular anomalies to determine the efficacy and safety of treatment with sirolimus for 12 courses; each course was defined as 28 days.
METHODS: Treatment consisted of a continuous dosing schedule of oral sirolimus starting at 0.8 mg/m2 per dose twice daily, with pharmacokinetic-guided target serum trough levels of 10 to 15 ng/mL. The primary outcomes were responsiveness to sirolimus by the end of course 6 (evaluated according to functional impairment score, quality of life, and radiologic assessment) and the incidence of toxicities and/or infection-related deaths.
RESULTS: Sixty-one patients were enrolled; 57 patients were evaluable for efficacy at the end of course 6, and 53 were evaluable at the end of course 12. No patient had a complete response at the end of course 6 or 12 as anticipated. At the end of course 6, a total of 47 patients had a partial response, 3 patients had stable disease, and 7 patients had progressive disease. Two patients were taken off of study medicine secondary to persistent adverse effects. Grade 3 and higher toxicities attributable to sirolimus included blood/bone marrow toxicity in 27% of patients, gastrointestinal toxicity in 3%, and metabolic/laboratory toxicity in 3%. No toxicity-related deaths occurred.
CONCLUSIONS: Sirolimus was efficacious and well tolerated in these study patients with complicated vascular anomalies. Clinical activity was reported in the majority of the disorders.

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Arteriovenous Malformation (AVM)

- High flow lesion
- Intracranial > Extracranial > Extremity > Truncal > Visceral
- Palpable thrill/ bruit
- Progress over time as collateral flow is recruited to the "nidus" low resistance shunt
- If ulcerate, can cause pulsatile bleeding
- Schobinger staging of AVM evolution (1990):

Stage	Features
I	Quiescence: pink-bluish, warmth
II	Expansion: pulsatile, bruit
III	Destruction: Pain, ulceration, bleeding
IV	Decompensation: cardiac failure

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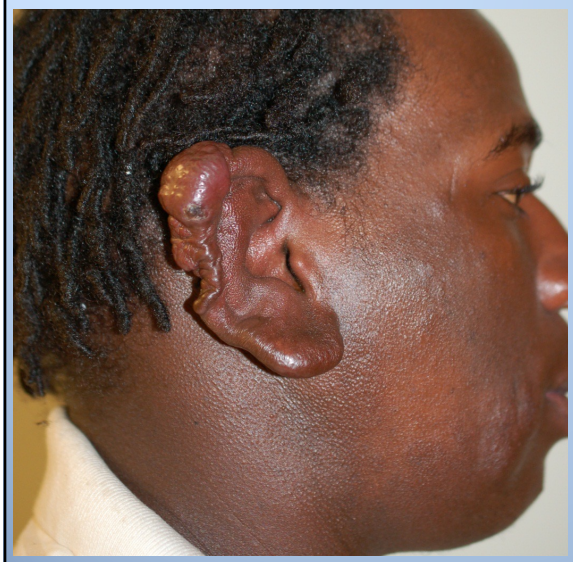
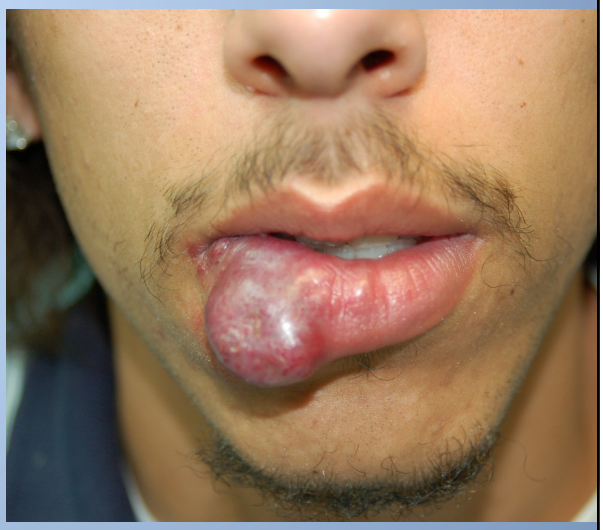
Arteriovenous Malformation





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Treatment Options

- Embolization
  - Onyx (ethylene-vinyl alcohol polymer)
- Surgical excision
- Cure is difficult…

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Genetics

- Increasing recognition of genetic links to vascular anomalies
- Infantile hemangioma- Familial clustering, 5q
- PHACE (posterior fossa abnormalities, hemangiomas, arterial abnormalities, cardiac abnormalities, eye colobomas)- possibly 7q33
- Capillary malformations- sporadic or inherited in AD pattern
- Venous Malformation- sporadic or inherited in AD pattern, TIE2/TEK
- Lymphatic malformations- sporadic, AR, AD. Multiple genes, VEGFR3 associated.

TAKE HOME MESSAGE

- TREATMENT for vascular anomalies depends upon diagnosis.
- "Hemangioma" was incorrectly used in 71.3% (228 of 320 articles). Patients with lesions mislabeled as hemangioma were more likely to receive improper treatment
- History and Physical exam can accurately diagnose **90%** of vascular anomalies

Hassanein, AH., et al. *Evaluation of Terminology for Vascular Anomalies in Current Literature*. PRS Journal. Jan 2011. vol.127: no.1347-351.

International Society of the Study of Vascular Anomalies (ISSVA)

Vascular Tumors	Vascular Malformations
Benign	Slow-flow
• Infantile Hemangiomas	• Capillary malformation

• Congenital Hemangiomas
  • NICH
  • RICH
	• Venous malformation

• Hemangioendotheliomas • Kaposiform, Spindle cell, Epithelioid, other rare…	• Lymphatic malformation
• Dermatologic acquired • Pyogenic granuloma	Fast-flow
Malignant • Angiosarcoma, Kaposi's sarcoma, Malignant angioendotheliomas…	• Arterial malformation
	• Arteriovenous malformation
	• Arteriovenous fistulas
	Combined vascular malformations

NICH- noninvoluting congenital hemangioma; RICH- rapidly involuting congenital hemangioma

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ISSVA 2014:

Vascular Anomalies				
Vascular Tumors	Vascular Malformations			
	Simple	Combined	of major named vessels	associated with other anomalies
Benign Vascular Tumors:	Capillary (C) malformations	Defined as 2 or more vascular malformations identified in 1 lesion. Can be composed of any combination of: C, L, V, AV*	"channel type" or "truncal" malformations	Klippel-Trenaunay synd.
Infantile hemangioma				Parkes Weber synd.
Congenital hemangioma:	Lymphatic (L) malformations			Servelle-Martorell synd.
Rapidly involuting (RICH)*				Sturge-Weber synd.
Non-involving (NICH)	Venous (V) malformations			Limb CM + limb hypertrophy
Partially involuting (PICH)				Maffucci synd.
Tufted angioma	Arteriovenous (AV) malformations*			Macrocephaly - CM
Spindle-cell hemangioma				Microcephaly-CM
Epithelioid hemangioma	Arteriovenous fistula*			CLOVES synd.
Pyogenic granuloma (aka lobular capillary hemangioma)				Proteus synd.
Others				Bannayan-Riley-Ruvalcaba
Locally aggressive or borderline vascular tumors:				
Kaposiform hemangioendothelioma				
Retiform hemangioendothelioma				
Papillary intralymphatic angioendothelioma (PILA),Dabska				
Composite hemangioendothelioma				
Kaposi sarcoma				

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TAKE HOME MESSAGE

- TREATMENT for vascular anomalies depends upon diagnosis.
- Classification system: ISSVA
  - www.issva.org

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- NOVA- National Organization of Vascular Anomalies

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THANK YOU!

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- >30 syndromes with vas half
- 28 genes linked

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