What is new in the Management of Hemangiomas

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Vascular Anomalies

- 1/3 children have a vascular birthmark or anomaly
- 1% - Significant & require evaluation & Rx
- 1/1000 - complex & require multimodal Rx
Objectives

- Differentiate hemangiomas from other vascular anomalies and malformations?
- Outline current classification system used by International Society of the Study of Vascular Anomalies (ISSVA)
- Discuss spectrum and patterns of hemangioma presentation
- Outline contemporary pharmacological and surgical treatment of hemangiomas
Not everything is a hemangioma despite appearances!
Not everything is a hemangioma despite appearances!

Kaposiform Hemangioendothelioma

Infantile Hemangioma

Lymphatic Malformation
VASCULAR TUMORS AND MALFORMATIONS

- **Vascular Tumors**
  - Arise by endothelial hyperplasia
  - Small or absent at birth
  - Rapid growth during early infancy
  - Involution during childhood

- **Vascular Malformations**
  - Arise by dysmorphicogenesis
  - Present at birth
  - Growth proportional to child
  - No regression

Mulliken & Glowacki. *Plast Recon Surg* 1982
Classification of Hemangiomas and Vascular Malformations

Vascular Birthmarks

Tumors
- Hemangiomas
- Other Tumors

Malformations
- High Flow
  - Arterial
  - Combined
- Low Flow
  - Capillary
  - Venous
  - Lymphatic
  - Combined

- Accepted by ISSVA (International Society for Study of Vascular Anomalies) in 1996
- Molecular & genetic differences not included
Classification of Hemangiomas and Vascular Tumors

- Hemangiomas
  - Infantile hemangioma
  - Congenital hemangioma
    - RICH
    - NICH
- Other Tumors
  - Hemangioendotheliomas
  - Tufted angiomas
  - Spindle cell hemangiomas
  - Pyogenic granuloma
HEMANGIOMAS

- Most common tumor of infancy
  - Female preponderance
  - More common in premature infants
    - Multiple gestations (10.6%)
    - Placenta previa (4.4%)
    - Pre-eclampsia (11.6%)
  - Chorionic villus sampling
  - Family history uncommon

- More common in caucasians but occurs in all races

- Head and neck > trunk > extremities

- 15 - 20% have multiple lesions
Not all hemangiomas are the same!
Placental Endothelial Markers

- Found in postnatal hemangiomas but not congenital hemangiomas, pyogenic granulomas, vascular malformations, granulation tissue or normal skin:
  - GLUT1: glucose transporter protein
    - Useful marker for infantile hemangioma
  - FcyRII
  - Merosin
  - LeY

North PE, et al, Arch Dermatol 2001;137:559-70
Congenital hemangiomas

- **RICH** - *Rapidly Involuting Congenital Hemangioma*
- **NICH** - *Non-Involuting Congenital Hemangioma* (Pale halo or center)
- These lesions are all GLUT-1 negative
30% visible at birth, seen as red macule: >80% become apparent during 1st few weeks of life

Proliferative phase 6 to 9 months

Involutive phase 1yr to 5 yrs

Most regress completely by age 7 years

20% have residual scaring & fibrofatty tissue
Management Differences

Localized

Multifocal (≥ 6)

Segmental
Periorbital Hemangiomas

- When to worry about visual impact?
  - Amblyopia,
  - Astigmatism
  - Blindness
- Careful exam
- CT/MRI scan
- Propranolol/Steroids
- Patching unaffected eye daily
Bearded distribution – airway involvement

- Airway involvement
- Rebound off steroids
- Proliferates up to 12-18 months
PHACE Syndrome

- Posterior fossa malformations
- Hemangiomas
- Arterial anomalies
- Cardiac anomalies
- Eye abnormalities
- Sternal cleft or supraumbilical raphe syndrome

PHACE: Algorithm

- Ophthalmologic exam
- Cardiology evaluation (ECHO, EKG)
- Neurologic evaluation / Developmental assessment
- MRI/MRA – Head, Neck and Mediastinum
- CBC w/ platelet count – consider other blood work (liver function, renal function, thyroid function)
Lumbosacral Hemangiomas

Associated with:
- Tethered cord
- Lipomas
- Spinal defects
- GU anomalies
- Anorectal malformations

Look for:
- Asymmetrical gluteal crease
- Sacral pits
- Masses
- Tufts of hair

Need imaging of the spine!
Hepatic Hemangioma Patterns

- 5 or more should be investigated for hepatic hemangiomas
- Start with a screening abdominal ultrasound
- Echo to rule out cardiac failure

Metry DW, et al, Archives of Dermatology 2004; 140: 591-6
Hepatic Hemangiomas

- **Focal** - May have arterio-venous or porto-venous shunts
- **Multiple skin lesions** common - often asymptomatic
- **Diffuse lesions**
  - High output CHF
  - Abdominal compartment syndrome
  - Profound hypothyroidism
- **Treatment:**
  - Propranolol
  - Corticosteroids
  - Vincristine, Cytoxan
  - Thyroid replacement
- Transplantation
- Only 60% **survive**
Treatment is necessary when hemangiomas...

- Supportive care
- Interfere with vital structures
  - Orbit
  - Airway
- Are life threatening
  - Congestive heart failure
  - Bleeding
  - Airway
- Ulcerated hemangiomas
- Possibility of permanent scarring
- Large facial hemangiomas
- Emotional burden family/child
Oral Systemic Corticosteroids

- Prednisolone 2-3 mg/Kg/day, single AM dose
- Slow tapering starts at 1 month
- Therapy usually lasts 3 to 6 months
- Short term effects: hypertension, Cushing’s features, live vaccine restriction?
- Long term effects: growth, bone mineralization, adrenal suppression?
- Potential rebound of hemangioma when weaning steroids
- Effective in >75% of proliferating hemangiomas
HEMANGIOMAS

Corticosteroid-Induced Regression

Side Effects of Corticosteroids
HEMANGIOMAS

Corticosteroid-Induced Regression

Side Effects of Corticosteroids

Cushingoid

Hypertension

Personality changes

Gastric irritation

Diminished gain of height and weight

Non-systemic fungal infections

(All complications usually resolve with discontinuation of therapy)
Propranolol – paradigm shift

- Propranolol 1-3 mg/kg
- VEGF inhibitor
- No controlled studies but has rapidly become new standard
- Therapy usually lasts 6 months
- Short term effects: hypoglycemia, hypotension, hyperkalemia
- Long term effects? Appears safe
- Potential rebound of hemangioma when weaning propranolol
- Effective in >85% of proliferating hemangiomas
• Vincristine: 0.05 mg/kg weekly for 4-6 wks; then monthly 6-12 months maintenance
  • Experience with Kasabach-Merritt Phenomenon
  • Growing use in steroid resistant hemangiomas
  • Side effects common but manageable

• Interferon-α2a: 1-3 million units/m2
  • Downregulates bFGF, increases cyclin-dependent kinase inhibitors
  • Effective in @ 1/2 of steroid resistant hemangiomas but not on vascular malformations
  • Side effects in infants (25%)
    • spastic diplegia in first year
    • 6 months therapy required

Laser Indications

- Useful for thin lesions, telangectasias
- Appears to be useful for reducing pain and promoting healing of ulcerated hemangiomas
Ulcerated Facial Hemangioma

- Systemic and topical antibiotics
- Extra Thin Duoderm
- Systemic steroids
- Pulse dye laser
Laser Indications

- Is there any convincing evidence that pulsed dye laser hastens resolution or prevents enlargement of hemangiomas?
  - NONE – only anecdotal reports

  - 121 infants uncomplicated hemangioma randomized to observation and PDL at diagnosis (<3 months)
  - Outcome superior in observational group
  - Significantly more scarring in PDL cohort
HEMANGIOMAS: Surgical Resection

Indications

Interfering with function

Ulceration

Mass effect

Cosmetic deformity

Establish Dx
Surgical Excision

• Selective approaches
  • Lenticular
  • Circular excision and purse-string closure
• Curative & cosmetic
Complex Reconstruction

4 years of age

10 years of age
Final Thoughts

Accurate counseling and optimal treatment are highly dependent on:

- Appropriate classification of vascular anomaly
- Understanding the biology & natural history of the lesions
- Diagnostic imaging and histology when appropriate
- Identification of clinical and biological patterns

The medical and self-esteem needs of the child and the family must be considered in planning treatment.
WOFAPS

Thank you!!!