American Academy of Pediatrics infantile hemangioma clinical practice guideline: What every dermatologist should know

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DISCLOSURE OF RELATIONSHIPS WITH INDUSTRY

DISCLOSURES

Venthera/Biobridge: Consultant – Consulting Fees
Pfizer Chair Data Safety monitoring board – Fees
Clinical Practice Guidelines (CPG): Relevance for Dermatologists

- AAD Guidelines of Care for IH was Oct 1997
- AAP recommended that US Agency for Healthcare Research and Quality study IH systematically
  - Report issued in 2016
- CPG used this report and updates through Jan 2017 as basis for multi-disciplinary consensus-guided document


ARS question #1
Which clinical finding *not* defined as high-risk in the AAP CPG

1. Segmental lumbosacral IH
2. Periocular IH
3. Presence of 5 or more cutaneous IH
4. Any facial IH >1 cm if infant is 3 months of age or young
5. Breast hemangioma involving nipple or central breast/female infant
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Key Take-aways

- Defines 14 Key-Action Statements in 5 domains
- Most IH small innocuous but significant minority are problematic
- Gives a risk stratification schema
- Emphasizes critical need for early referral for high-risk IH “window of opportunity”
- Defines what is a “hemangioma specialist”
What is “right answer” for PCP?

1. Reassure this is benign and will go away
2. Recheck again at next well-baby check-up
3. Urgent referral to dermatologist
4. Start topical timolol
5. Start oral propranolol
- Retrospective study 184 untreated patients
- Hemangiomas with a **steep or abrupt border** of superficial component had more severe sequelae than those with a smooth border (OR, 3.4; 95% CI, 1.8-6.6; P < .001)
- Less sequelae from very superficial OR deep hemangiomas (OR, 1.6; 95% CI, 0.6-3.8)

Baselga et al. JAMA Dermatol 2016; 152;1239-43.
Main messages for pediatricians/PCPs:
Natural History and Risk Stratification

- IH growth characteristics different than once taught
- IHs involute BUT process may be incomplete
  - Can leave permanent skin changes that may be life altering
  - Especially true for IHs that are thick.
- Emphasis on dual risks: medical AND risk of scarring/disfigurement
Key Messages Re: Timing

- Most rapid IH growth occurs between 1 and 3 months of age.

- Window of opportunity to treat problematic IHs
  - Ideal is by 1 month of age for IH that are potentially high risk

- For highest and high risk: specialist consultation is urgent (e.g. 1-2 weeks)
Defining “highest” and “high” risk

- Life-threatening complications
- Risk underlying abnormalities
- Functional impairment
- Potential causing permanent disfigurement
- Ulceration
Risk Stratification and Need for Consultation

- Highest risk: Timing ≤1 week
- High risk: Timing ≤2 weeks
- Intermediate risk: May or may not need consultation/referral
- Low risk: Typically will not need consultation/referral

Highest risk

- Large (>5 cm) or segmental facial or scalp
  - Risk PHACE, higher risk disfigurment

- Large or segmental lumbosacral or perineal
  - Risk LUMBAR syndrome, higher risk ulceration

- Multifocal IHs (≥5) and abdominal ultrasonography reveals liver hemangiomas:

- Periocular IH causing eyelid asymmetry, lid closure or ptosis, proptosis
High Risk: Most common reason for consultation/referral

- Large segmental IH on trunk or extremities
- Any facial IH ≥ 1 cm if <3 mos of age (or 2 cm if older)
- Scalp ≥2 cm growth phase
- Nasal tip, perioral even smaller
- Breast involvement female infants
- Ulceration any site
Re: Treatment Take-Aways of CPG

- Oral propranolol for problematic IHs requiring systemic therapy
  - Information re: dosing and anticipatory guidance
- Topical timolol to treat some thin and/or superficial IHs
- Surgery and laser most useful for the treatment of residual skin changes after involution
  - May be used earlier for selected IHs
Key Action Statements re: Imaging

- No routine imaging except:
  - diagnosis of IH is uncertain
  - ≥5 cutaneous IHs
  - Risk associated anatomic abnormalities (ie PHACE, LUMBAR

- Ultrasound
  - initial imaging modality when the diagnosis of IH is uncertain
  - looking for liver hemangiomas

- MRI if concern re: associated structural anomalies
Defining “hemangioma specialist”

- Management of IHs is not limited to 1 medical or surgical specialty.
- A hemangioma specialist may have expertise in dermatology, hematology-oncology, pediatrics, facial plastic and reconstructive surgery, ophthalmology, otolaryngology, pediatric surgery, and/or plastic surgery, and his or her practice is often focused primarily or exclusively on the pediatric age group.
More on “hemangioma specialist”

- Understand the time-sensitive nature of IHs during the growth phase and be able to accommodate requests for urgent evaluation;

- Have experience with accurate risk stratification and potential complications associated with IHs
  - Able to provide recommendations for various management options and to discuss R/B/A for specific patients
  - Knowledge of past and emerging medical literature regarding IHs.
Resource for parents and physicians

- Many charts, graphs, supplementary materials
- Goal: Trying to make easier for PCPs to gain confidence in IH management and referral
- Handouts for parents
  - General IH information
  - Information re: propranolol
  - Medication handout
### SUPPLEMENTAL TABLE 22 Risk Level of Les of Varying Types

Using this table, assess the risk posed by the infantile hemangioma you are evaluating. On the reverse side of the page use the flow diagram to determine the action recommended based on risk.

<table>
<thead>
<tr>
<th>Risk Level</th>
<th>Clinical Examples and Reasons for Concern</th>
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| Highest    | - Large (>5 cm) or segmental facial or scalp:  
  - Higher risk of airway hemangiomas (e.g., beard area)  
  - May be associated with PHACE syndrome  
  - High risk of ulceration and/or disfigurement  
  - Large or segmental lumbar or posterior:  
  - May be associated with LUMBAR syndrome  
  - High risk of ulceration and disfigurement  
  - Multifocal IHs (≥5) and abdominal ultrasonography reveals liver hemangiomas:  
  - May be associated with abdominal compartment syndrome, high-output congestive heart failure, and hypothyroidism  
  - Periorbital IH causing eyelid asymmetry, lid closure or ptosis, proptosis, or other findings with potential impact on visual axis:  
  - Risk of strabismus, ptosis, and amblyopia  |
| High       | - Large segmental IH on trunk or extremities:  
  - Risk of scarring and/or disfigurement  
  - Any facial IH ≥2 cm (>1 cm if ≤5 mo of age):  
  - High risk of scarring and/or disfigurement  
  - Nasal tip or lip IH even if <1 cm:  
  - High risk of scarring and/or permanent distortion of anatomic landmarks  
  - Oral:  
  - Risk of ulceration or bleeding, may interfere with feeding  
  - Neck or scalp IH ≥2 cm during growth phase:  
  - Risk of ulceration (neck)  
  - Risk of ulceration, scarring, and/or hair loss (scalp)  
  - Breast:  
  - Risk of permanent changes in breast development (e.g., breast asymmetry) or nipple contour  
  - Ulcerated hemangioma (any site):  
  - Risk of severe pain, scarring and/or disfigurement, and bleeding  |
| Intermediate| - Perimal IH (localized) without ulceration:  
  - Potential for ulceration in this location  
  - Trunk or extremity IH ≥2 cm especially in growth phase or if abrupt transition from normal to affected skin (e.g., ledge effect; Fig 8):  
  - Risk of scarring and/or disfigurement  |
| Low        | - IH ≤2 cm on trunk or extremities in areas easily covered by clothing  
  - IH on trunk or extremities ≥2 cm if gradual transition from normal to affected skin (Fig 13) |

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*See photographic examples at [https://downloads.aap.org/00E2E8C1EUIR_Example_Photos.pdf](https://downloads.aap.org/00E2E8C1EUIR_Example_Photos.pdf)*

*Consultation with a hemangioma specialist may involve a telephone conversation and/or electronic transmission of patient photographs.*
CPG Supplementary Information

- Patient information
- General IH information
- Information re: propranolol
- Medication handout
- Table defining highest, high, intermediate, low risk IH
- Management algorithm

Management algorithm of IH in infants ≤3 months of age

- **Highest**
  - Consult or refer to hemangioma specialist immediately (typically urgent, within 1 week)

- **High**
  - Consult or refer to hemangioma specialist promptly (typically within 2 weeks)

- **Intermediate**
  - Plan to repeat evaluation every 2 to 4 weeks until age 3 months (or older if IH growth continues)

- **Low**
  - Plan to repeat evaluation at next well-child check visit

  - Instruct caregivers to watch for ulceration, bleeding, or other concerns and contact provider if they occur

  - Consult or refer to hemangioma specialist if parental concern or if provider desires additional opinion
Challenges to Implementation of CPG

- Dynamic nature individual IHs
  - Period rapid growth but hard to predict how much, how long
- Wide heterogeneity of IH in size, location, patterns of distribution and depth
- Long-held belief that “IH are benign and will go away”
- Lack of uniform availability of “hemangioma specialists”
Infantile Hemangiomas: 2019

- Highly effective IH therapy available
- AAP CPG tells PCPs who to refer and gives guidance re: timing of referral
- Goal of CPG group is to “move the needle” on more prompt and appropriate referrals
- Implementation challenges remain