Blisters and scales and marks, oh my!

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Leslie Potter Lawley M.D.
Assistant Professor of Dermatology and Pediatrics
Emory University School of Medicine
leslie.lawley@emory.edu

Leslie P. Lawley, M.D.
Personal/Professional Financial Relationships with Industry

<table>
<thead>
<tr>
<th>External Industry Relationships *</th>
<th>Company Name</th>
<th>Role</th>
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<td>Board of Directors or officer</td>
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<td>Royalties from Emory or from external entity</td>
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<td>Content Author</td>
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<td>Industry funds to Emory for my research</td>
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No conflicts related to this talk. I will discuss off-label use of medications.
Learning Objectives

• Compare and contrast blistering disorders in the neonate
  • Review ddx and diagnostic pearls
  • Focus on Epidermolysis Bullosa and wound care
• Recognize newborn presentation of collodion membrane and implement management strategies
• Identify and differentiate birthmarks (including the subtypes of infantile hemangiomas) that warrant further work up for systemic involvement

abstract

• Skin changes may indicate a severe systemic genetic disorder or a benign and common eruption. The neonatology provider needs to be able to recognize different skin lesions and distinguish when further work up is indicated by certain skin presentations. Basic skin care for neonates with blistering or collodion membrane will be reviewed.
Blisters

Conditions where pustules and/or vesicles predominate

- **Common**: superficial staphylococcal infection, erythema toxicum neonatorum, neonatal pustular melanosis, miliaria, benign cephalic pustulosis, neonatal candidiasis
- **Uncommon**: congenital candidiasis, herpes simplex, scabies, acropustulosis, incontinentia pigmenti
- **Rare**: Listeria, H. influenzae, group A streptoccus, pseudomonas, varicella, cytomegalovirus, aspergillus, eosinophilic pustular folliculitis, erosive pustular dermatosis of the scalp, Langerhans cell histiocytosis, hyperimmunoglobulin E syndrome, pustular eruption in Down syndrome, pustular psoriasis, neonatal Behcets’
Conditions where bullae may predominate

- **Common**: bullous impetigo, sucking blisters
- **Uncommon**: staphylococcal scalded skin syndrome, epidermolysis bullosa
- **Rare**: group B streptococcal infection, pseudomonas, congenital syphilis, varicella, bullous mastocytosis, maternal auto-immune blistering disease (pemphigus, pemphigoid), linear IgA, bullous pemphigoid, toxic epidermolysis necrosis, epidermolytic hyperkeratosis, acrodermatitis enteropathica, membranous aplasia cutis congenita

Common, uncommon, and rare but important to diagnose: INFECTIONS

- Rupture vesicle, pustule, or bullae and culture the base
- Gram stain, KOH, or DFA/ELVIS may give immediate results
<table>
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<tr>
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<th>Congenital candidiasis</th>
<th>Neonatal candidiasis</th>
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<tbody>
<tr>
<td><strong>Frequency</strong></td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td><strong>Acquisition</strong></td>
<td>In utero</td>
<td>Antepartum/post partum</td>
</tr>
<tr>
<td><strong>Cord</strong></td>
<td>May show whitish plaques</td>
<td>Not seen</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>&lt;6 days of life</td>
<td>&gt;6 days after birth</td>
</tr>
<tr>
<td><strong>Sites</strong></td>
<td>Back, skin folds, palms, soles</td>
<td>Oral, diaper area involved</td>
</tr>
<tr>
<td><strong>Morphology</strong></td>
<td>Generalized, erythematous macules, papules/pustules on an erythematous base</td>
<td>Deep beefy red color with moist appearance, scalloped outline, satellite pustules</td>
</tr>
</tbody>
</table>

Adapted from Indian Dermatology Online Journal

**Typical baby “rashes”**

- Erythema Toxicum Neonatorum
- Benign cephalic pustulosis (“baby acne”)
- Transient Neonatal Pustular Melanosis
- Miliaria
Erythema toxicum neonatorum

- Common in term infants
- Rare in premature or <2500gm
- Ave onset 24-48 hrs age (up to 14 days)
  4 main lesions:
  1) Erythematous macules
  2) Wheals
  3) Papules
  4) Pustules
  - Transient lesions (hours to days)
  - Wright’s stain: eosinophils

Benign cephalic pustulosis

- Colonization of the skin with Malassezia furfur
- Onset 2-3 weeks
- Asymptomatic
Transient Neonatal pustular melanosis

• More common in term infants
• Present at birth
3 types of lesions:
1) Pustules (no erythema)
2) Hyperpigmented macules with collarette of scale (ruptured pustules)
3) Hyperpigmented macules
   • Wright’s stain: neutrophils; Gram stain: Neutrophils, no bacteria
   • Pustules resolve in days
   • Macules resolve in weeks-months

Miliaria

• Blockage of sweat ducts at stratum corneum
Scabies

• DDX Vesicles/Pustules on the palms and soles includes:
  Scabies                      Herpes Simplex virus
  Acropustulosis              Varicella Zoster virus
  Bullous pemphigoid          Congenital candidiasis
  Langerhans cell histiocytosis

Incontinentia pigmenti

• 50% with lesions at birth
• 90% with lesions by 2 weeks
• 4 stages in Blaschkoid pattern:
  • Vesicular
  • Verrucous
  • Hyperpigmented
  • Hypopigmented
• Alopecia, nail dystrophy
• Ocular involvement 35%
• CNS in 10-30%
• Dental in 70-95%

• Rule out HSV, impetigo
• Skin biopsy diagnostic
• Peripheral eosinophilia >70%
• Work up:
  • Ophthalmology exam
  • Neurology exam
• Gentle skin care, no specific treatment
Aplasia cutis congenita
(congenital localized absence of skin- CLAS)

• 9 groups described
• Group 6: ACC assoc with EB; includes junctional EB with pyloric atresia
• Group 7: ACC localized to extremities without blistering
• Group 8: ACC due to teratogen
  • Methimazole, VZV, HSV

1 day newborn developing blisters, erosions
Epidermolysis Bullosa

• Widely varied inherited disorders of skin +/- mucosal fragility in response to minor mechanical trauma
Epidermolysis Bullosa

• Incidence 1 in 50,000 births
• 3 main types
  • Simplex (>13 subtypes)
  • Junctional (>7 subtypes)
  • Dystrophic (>11 subtypes)
• Can all look similar in the newborn period
• Varied morbidity and mortality

EB- Diagnosis

• Skin biopsy
  • Induce a blister and biopsy
    • Provides general type with level of split
  • Direct immunofluorescence antibody testing
    • If a protein is completely missing then very informative
    • If partial protein not as helpful
• Genetic testing
  • Most useful for determining prognosis and other risks
  • Simplex- keratin 5,14; plakophilin-1 (ED overlap), plectin (MD or pyloric atresia), integrin α6 or β4 (pyloric atresia), dystrophin (BP ag)
  • Junctional- laminin 5, collagen XVII/BP180, integrin α6 or β4 (pyloric atresia)
  • Dystrophic- collagen VII
Immediate Care in the NICU for EB

- Gentle handling
- Wound care
- NO ADHESIVES
- Monitor for infections
- Ensure adequate nutrition/feeding
- Teach parents how to care for their child
  - Start bonding, getting results on testing/subtype takes time
  - Focus on care of the wounds
  - Focus on feeding and nutrition
  - Genetic counseling

Wound Care in the NICU for EB

- Cover wounds with soothing bandages
  - Makes the neonate more comfortable
  - Allows parents/caregivers to hold their child
  - Help wounds heal
  - Vaseline gauze can be immediately wrapped around affected areas
  - Key is to not allow it to dry and stick to wounds
- Wash hands
  - Before and after dressing changes
  - High risk of infection
Wound Care in the NICU for EB

• Wound dressing changes: **Set out all supplies**
  • Sterile needles (drain blisters)
  • Antibiotic ointment
  • Ointment such as petrolatum (Vaseline®, Aquaphor®, or Dermaphor®)
  • Contact layer (1st layer to touch skin), ex Mepitel®, Restore®, Vaseline® gauze or Mepilex® Transfer---cut in strips (½ to 1 inch width)
  • Soft, conforming, stretch rolled gauze (1-2 inch) for 2nd layer
  • Tubular dressing for 3rd layer (ex Tubifast® or Surgilast®)

**Contact Layer**

- Restore® bandage
- Mepitel®
- Vaseline® gauze
- Mepilex® transfer
Wound Care in the NICU for EB

Pain control
• Pre-medicating may help
  • Acetaminophen or ibuprofen
  • Oral sucrose
• Oral erosions and ulcers
  • Magic mouthwash
    • 1 part viscous lidocaine, 1 part diphenhydramine, 1 part liquid aluminum hydroxide or magnesium hydroxide
  • Apply with qtip or toothette
• Encourage feeding—may use soft premie or cleft palate nipple
Wound Care for EB: Steps for Dressings

1. **Drain blisters:**
   - Drain if tense or >1cm size (prevents further increase in size)
   - Using sterile needle, puncture side (parallel to skin surface), gently release fluid (do NOT remove roof)

2. **Cleanse Skin:**
   - Mild non-perfumed, non-soap cleanser
   - Rinse erosions and intact blisters with a soapy solution
   - Then rinse with plain water
   - Do NOT rub dry (can pat dry if needed)

3. **Apply ointment:**
   - Gently apply to skin (can apply to contact layer instead)
     - If a tub of ointment use tongue blades to scoop out ointment and prevent contamination
     - Use antibiotic ointment only in areas that look infected

Wound Care for EB: Steps for Dressings con’t

4. **Apply dressings:**
   - First apply non-adhering contact layer (spiral wrap)
   - Wrap conforming rolled gauze over the contact layer to hold it in place
   - Secure with the tubular dressing
   - If needed, tape may be used to secure dressings but ONLY apply tape to the dressing and not directly on the skin
Wound Care in the NICU for EB

Removal of dressings:
- Change dressings daily to every other day
- Change dressings on one limb at a time
  - Reduce risk of natural kicking causing more wounds, bleeding
  - Air on open wounds is painful
- Soak dressings or clothing stuck to wounds with warm water or sterile saline to allow gentle removal

Wound care of EB in a neonate:
- Smaller erosions/vesicles on trunk: Apply petrolatum +/- contact layer
- Note contact layer between diaper and skin
- Diaper: Consider cutting out elastic portion if causing too much friction and blistering.
- Use a thick layer of petrolatum to prevent friction.
- Be creative with using dressing supplies to anchor necessary lines and tubes
- Tubular dressing on the head to create a “hat”
- Contact layer between cheek and tubes
- Can suture (anchoring deep in dermis below level of skin split for blistering)
Wound Care for EB in NICU

• Avoid adhesives!
• Do not debride (allow roof of blister to be a protective dressing)
• Even in less severe forms of EB significant erosions can occur from use of adhesives
• If they must be used (i.e. to anchor an IV or NG; or for surgery) then remove VERY cautiously

Monitor for signs of infection

• Increased drainage from wounds
• Increased pain
• Increased swelling
• Increased warmth
• Malodorous
• Easy bleeding
• Fever
• Progressive worsening of blistering
• Secondary staph, HSV, SSSS
Nutrition in EB babies

- Higher caloric requirement
- Breast feeding may be attempted but monitor weight gain carefully
- Difficulty latching and maintaining strong suck
  - Cleft palate or soft premie nipples
- Allow oral blisters/erosions to rupture and heal on their own
- If not growing well consider gastronomy tube

Other considerations for EB newborns

- Refer parents to DEBRA website (www.debra.org)
  - Invaluable resource for families and providers
  - Nurse educator available to parents and providers
- Encourage parents/family to hold/nurture the child
- Listen for hoarseness that suggests airway involvement
- Minimize placement of tubes if possible
- Parents/caregivers should be involved in wound dressing changes immediately to ensure smooth transition home
**Take home points for care of EB newborn**

- All forms of EB can include large areas of aplasia cutis congenita
- Cannot determine specific form of EB clinically, genetic testing needed
- Gentle wound care
- Monitor nutrition
- No adhesives to skin!
- Involve parents in wound care

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**Scales**
Collodion membrane

Diffuse scale and erythema
Taut skin especially over joints
(note blanching appearance)

Diagnoses to consider in the setting of collodion membrane:

- ARCI- congenital ichthyosiform erythroderma
- ARCI- Lamellar ichthyosis
- ARCI- Self-healing collodion baby
- Neutral lipid storage disease with ichthyosis
- Loricrin keratoderma
- Gaucher disease, type II
- Trichothiodystrophy syndromes
- Sjogren-Larsson syndrome
- Conradi-Hunermann syndrome
- ARCI- Harlequin ichthyosis

65% have ARCI
Immediate care in the NICU of collodion membrane

• Supportive care:
  • Humidified incubator with temperature control
  • Application of bland emollients (for gentle desquamation)
  • Avoid keratolytic agents in infancy
• Excessive transcutaneous fluid and electrolyte loss
  • Monitor electrolytes
  • Monitor fluid intake and output
• Increased metabolic requirements
• Ectropion:
  • Consult ophthalmology
  • Lubricating ointment

Immediate care in the NICU of collodion membrane: possible complications

• Inability to suck properly
• Respiratory difficulties due to restriction of chest expansion
  • Aspiration pneumonia (can be neonatal from squamous material in amniotic fluid)
• Peripheral edema with digital constriction
• Hyponatremic dehydration
• Temperature instability
• Risk for cutaneous and systemic infections
Collodion membrane

- Counsel parents regarding possible underlying diagnoses
- Refer to FIRST
- Emollients!!!
- Hearing tests

Marks
Oh my!

“Bummer of a birthmark, Hal.”
Nevus simplex

• Up to 40% of newborns
  • 81% nape of neck
  • 45% eyelids
  • 33% glabella
• Ectatic capillaries represent persistent fetal circulatory patterns in skin
• Central face, symmetric

Nevus simplex

Does this infant need any work up?

• No scans unless other signs of dysraphism: hair collar sign on scalp or hypertrichosis at sacrum, lipoma
Cutaneous lesions associated with spinal dysraphism

**High index of suspicion**
- Presence of 2 or more cutaneous lesions
- Hypertrichosis
- Dimples (large, >2.5 cm from anal verge, atypical)
- Acrocordons/pseudotails/true tails
- Lipomas
- Hemangiomas
- Aplasia cutis or scar
- Dermoid cyst or sinus

**Low index of suspicion**
- Telangiectasia
- Capillary malformation
- Hyperpigmentation
- Melanocytic nevi
- Small sacral dimples (<2.5 cm from anal verge)
- Teratomas

From Neonatal and Infant Dermatology, Eds Eichenfield and Frieden, 3rd Ed. 2015

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Capillary vascular malformation
“port wine stain”

- Present at birth
- 3/1000 infants
- Face/neck
  - With time darken, more violaceous, thicken, blebs, hyperkeratotic surface
- Limb/trunk
  - Occasionally fade over time
Sturge-Weber Syndrome

- Facial port wine stain
  - Embryonic developmental patterns
  - 8% incidence
  - 24% if hemifacial or bilateral
- Vascular malformations of leptomeninges and the eye
  - Seizures (55-90%)
  - Hemiplegia (30%)
  - Mental retardation (50-65%)
  - Ocular (60%): Glaucoma, nevus of Ota, buphthalmos, blindness

Sturge-Weber Syndrome

- Facial PWS + Vascular malformations of leptomeninges and the eye
- Post-zygotic activating mutation in GNAQ
  - Increases proliferation and inhibits apoptosis through RAS paths
- Somatic mosaicism
- Forehead involvement higher risk for associated SWS

Facial capillary vascular malformation and Sturge-Webber Syndrome Risk

- Embryonic patterns to the PWS (not V123—abandon trigeminal terminology)
- Forehead involvement may suggest increased risk
  - Hemi-facial also high risk
- Refer to ophthalmology immediately
- Consider early MRI with gadolinium, weigh risks/benefits
  - Could refer to neurology to assess and assist in decision for imaging
  - Feed and wrap neonates (can avoid sedation)
  - **MRI with and without contrast**: post contrast T2 FLAIR sequence very sensitive for subtle early findings
Capillary malformation vs infantile hemangioma?

- In the first few weeks of life capillary malformations (port wine stains) and early infantile hemangiomas can look very much alike
- Key: monitor for signs of proliferation that indicate an infantile hemangioma (bright red papules, thickening)

Infantile Hemangioma

- Incidence 1-4.5%
- Risk factors: low birth weight, placental abnormalities, female, preterm, multiple gestation, maternal hx miscarriage, fam hx
- Appear birth-6wks
- Precursor in up to 50%
- Superficial, deep, mixed
- Focal vs segmental

IH-Natural History

1. Proliferative stage
   • Rapid growth 0-6mo (especially 5-7 weeks of age)
   • IH reach 80% of final size by 3 mo age
   • 80% of IH reach final size by 5 mo age
   • Slow growth 6-12 mo; rarely in 2nd yr of life

2. Plateau stage
   • stable

3. Involution stage
   • 5-10 years
   • Residual fibro-fatty tissue, texture changes, occasional telangiectasias


Subtypes of Infantile Hemangiomas
May indicate a need for further evaluation

• **Segmental hemangiomas**
  • PHACE(S)
  • LUMBAR/PELVIS/SACRAL
  • Other risks
    • Airway involvement
    • GI involvement

• **Multiple hemangiomas**
  • Multifocal IH with or without extracutaneous disease
Syndromes with segmental IH

• PHACE
  • Posterior fossa malformations
  • Hemangioma
  • Arterial anomalies
  • Cardiac anomalies and aortic coarctation
  • Eye abnormalities
  • Sternal clefting and supraumbilical abdominal raphe


Diagnostic Criteria for PHACE

Diagnosis of PHACE syndrome
• Facial Hemangioma >5cm diameter PLUS 1 major criteria or 2 minor criteria
• IH neck/upper trunk or trunk/prox UE + 2 major criteria

Possible PHACE syndrome
• Facial IH >5cm diameter + 1 minor criteria
• IH of neck or upper torso + 1 major or 2 minor criteria
• No IH + 2 major criteria

<table>
<thead>
<tr>
<th>Organ system</th>
<th>Major Criteria</th>
<th>Minor Criteria</th>
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<tbody>
<tr>
<td>Cerebrovascular</td>
<td>Anomaly of major cerebral arteries: Dysplasia of the large cerebral arteries, arterial stenosis or occlusion with or without moyamoya collaterals, absence or moderate to severe hypoplasia of the large cerebral arteries, aberrant origin or course of the large cerebral arteries, persistent carotid-vertebrobasilar anastomosis</td>
<td>Aneurysm of any of the cerebral arteries</td>
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<td>Structural Brain</td>
<td>Posterior fossa anomaly: Dandy-Walker complex or other hypoplasia/dysplasia of the mid and/or hind brain</td>
<td>Malformation of cortical development Midline brain anomalies</td>
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<tr>
<td>Cardiovascular</td>
<td>Aortic arch anomaly: Coarctation of aorta, Dysplasia Aneurysm Aberrant origin of SC w/ or w/o vascular ring</td>
<td>Ventricular septal defect Right aortic arch/double aortic arch Systemic venous anomalies</td>
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<tr>
<td>Ocular</td>
<td>Posterior segment abnormality: Persistent hyperplastic primary vitreous, Persistent fetal vasculature, retinal vascular anomalies, morning glory disc, optic nerve hypoplasia, peripapillary staphyloma</td>
<td>Anterior segment abnormality: Sclerocornea, cataract, coloboma, microphthalmia</td>
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<tr>
<td>Ventral or midline</td>
<td>Anomaly of the midline chest and abdomen: Sternal cleft or pit, supraumbilical raphe, sternal defects</td>
<td>Midline sternal papule/hamartoma Ectopic thyroid hypopituitarism</td>
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</tbody>
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• **Facial Segmental Hemangioma:**
  • 31% have PHACE, majority >1 extracutaneous finding
  • 91% cerebral arterial anomalies, 67% cardiac anomalies
  • S1 with or without other segments most common

Haggstrom AN, Garzon MC, Baselga E, et al Pediatrics 2010; 126:e418
Evaluation of facial segmental infantile hemangioma

- PE (head circ, ventral developmental defects, signs of symptomatic liver hemangioma)
- MRI/A head and neck; upper chest
- Neurologic exam (to neurology if abn exam or MRI/A)
- Echocardiogram
- Ophthalmology exam
- ENT exam (if beard distribution or breathing difficulty)
- Thyroid function tests; CBC, stool guaiac

Long Term Effects of PHACE:

- Speech delay, swallowing difficulties
- Dental enamel hypoplasia
- Hearing loss
- Developmental delays
- GI bleed
- Headaches, migraines, stroke
Facial IH and airway hemangiomas

- Risk of airway IH 27-40% with “beard distribution” (S3)
- Of 108 pts: 17 pts with both facial IH and airway IH
  - All 17 with S3 involved
  - 14/17 symptomatic
  - 8/17 definite PHACE
- Single center retrospective study:
  - 52% PHACE pts had airway IH
  - 2 pts w/o S3 involvement
  - 12% facial IH w/o PHACE have airway IH
- ENT evaluation


Syndromes with segmental IH of the pelvis or lower extremities

LUMBAR syndrome

- Regional association of the distribution of the hemangioma and the associated anomalies
  - T12-L5 area strongly correlated with myelopathies (tethered cord or lipomyelocle/lipo-myelomeningiocele)
  - Anterior iliac crest to perineum associated with urogenital, anorectal, and renal anomalies
  - Vessel anomalies of large arteries found ipsilateral to the IH and may affect limb development
- Minimal growth hemangiomas more common
  - May need biopsy to distinguish from CMTC or PWS
- High incidence of ulceration
- Female predominance (not as high as PHACE)


Evaluation of segmental infantile hemangioma of the lower body (LUMBAR)

- Thorough physical exam of the abdomen, pelvis, and lower extremities with emphasis on genital and paraspinal regions
- If < 3 mo age: ultrasound with color doppler of spine, abdomen, pelvis
- If > 3 mo age: magnetic resonance imaging determined by distribution of the cutaneous hemangioma
- Radiographic scanogram of lower limbs in early pre-school years and consider repeat at 6 yrs age
- Thyroid function tests, CBC, UA, stool guaiac

Multiple Infantile Hemangiomas

• 30% infants have greater than one IH
• Well established association with hepatic IH
• Prospective study of infants with multiple IH:
  ▪ ≥5 cutaneous IH: 16% with hepatic hemangiomas
  ▪ ≤4 cutaneous IH: no liver hemangiomas
• Risks for multiple IH similar to any cutaneous IH
• 8% with hepatic IH required treatment


Multiple Infantile Hemangiomas

Hepatic hemangiomas reported with several morphologies of cutaneous hemangiomas:

• **Miliary**: 30-100 pinpoint to 5 mm sharply demarcated papules
• **One large (>5cm) plus 1 or more small (<5cm)**
• ≥ 6 small
• Solitary large (>5cm)

*higher risk of hepatic Hemangioma with this morphology

Canty at al South Med Journal 2014;107:159-64.
Hepatic hemangiomas

1. **Focal- 15% with cutaneous IH**
   - Congenital, solitary, large, regress within months (RICH- like)
   - May be associated with AV shunting and heart failure

2. **Multifocal- 77% with cutaneous IH**
   - Most common and usually associated with multiple cutaneous IH, often asymptomatic but can have AV shunting and high output cardiac failure

3. **Diffuse- 53% with cutaneous IH**
   - Poor clinical outcomes; massive hepatomegaly, abdominal compartment syndrome, severe hypothyroidism

Evaluation of the patient with ≥ 5 cutaneous infantile hemangiomas

- Abdominal exam
- Vitals
- **Abdominal ultrasound**
  - if <4 wks age, repeat at 1 mo later
  - If u/s positive assess with CT or MRI

- If diffuse involvement of liver with hemangiomas: need baseline size, evaluate cardiac function (echo/ECG) and thyroid function

- If focal or multifocal hepatic IH: follow with serial u/s frequently until no longer growing then q3 mo; cardiac evaluation if signs of AV shunting

- Labs: CBC, stool guaiac, thyroid screen


Multifocal lymphangioendotheliomatosis with thrombocytopenia (MLT)

- Vascular papules at birth and new ones develop
- GI bleeding severe
- Thin walled vascular channels lined by prominent hob-nailed endothelial cells, LYVE-1+ and Ki-67+
- Severe thrombocytopenia
- Coagulopathy
- Poor response to most medications
- Common in GI, lung, bone
- Rare liver involvement


Kaposiform hemangioendothelioma (KHE)

- Rare neoplasm
  - In skin, chest wall or retroperitoneum
- Often associated with Kasabach-merritt (KM)
- Typically tumor present at birth or soon after, 75% occur in early infancy
Kasabach-Merritt phenomenon

- Assoc with kaposiform hemangioendothelioma and tufted angioma
- Commonly appears in first few months
- Tenderness, rapid growth and bruising of soft tissue tumor with widespread purpura
- Different from coagulopathy seen with vascular malformations

- Consumptive coagulopathy
  - Low platelet
  - Low fibrinogen
  - Elevated D-dimers
  - Elevated PT and PTT
  - Hemolytic anemia
- Mortality 10-30%

Kasabach-Merritt phenomenon

- Multidisciplinary approach:
  - Corticosteroids
  - Vincristine
  - Interferon-alpha
  - Surgery
  - Arterial embolization
  - Sirolimus
  - Xray irradiation
  - Fibrinogen or FFP, avoid platelet transfusion
Epidermal Nevus Syndrome

- Extensive epidermal nevus or nevus sebaceous in lines of Blaschko
- Multiple forms of ENS described
- Associated abnormalities of CNS, MSK system, or eyes
- Refer to ophthalmology
- Follow neurodevelopment (refer to neurology if abnormal)

Giant congenital melanocytic nevus and satellite nevi

- Present at birth as dark brown to black plaques +/- verrucous or cobblestone surface and terminal hairs
- Color variations
- May develop superimposed papules or nodules with time (also in smaller CMN)
- Satellite nevi may be present at birth and/or continue to develop in infancy
- Risks: Melanoma (5%) and symptomatic neurocutaneous melanosis (4%)
- Regular skin checks with dermatology and standard monitoring of neurodevelopment
Blisters and Scales and Marks, oh my!

- Broad differential for blisters in the newborn
- Rule out infection
- Gentle wound dressings for epidermolysis bullosa
- Involve the caregivers of EB babies early
- Collodion membrane: monitor electrolytes, use emollients, humidified chamber
- Facial port wine stain has risk of Sturge-Weber syndrome, early MRI recommended
- Facial segmental hemangioma risk of PHACE syndrome and needs work up
- Lower body/sacral hemangiomas work up for LUMBAR syndrome
- Multiple hemangiomas rule out liver involvement