Approach to Blistering Diseases in the Pediatric Patient

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Objectives

• To discuss an approach to the evaluation of blisters in pediatric patients
• To compare different patterns of disease presentation in infants versus children
• To understand the role of diagnostic tests in pediatric patients with blistering
Neonates

Concern for Systemic Infection

High

- Bacterial – most common
- Viral – grouped vesicles, maternal risk factors
- Fungal – preterm, low birth weight

Low or Ruled Out

Blisters Predominate

- Erosions Predominate

Isolated Lesions/
Non-progressive

- Sucking Blisters – hand or forearm
- Bullous Impetigo – diaper area
- Aplasia Cutis Congenita (bullous variant) – scalp

Multiple Lesions/
Progressive

- Genetic
  - Epidermolysis Bullosa – congenital absence of skin, traumatic blisters
  - Epidermolytic Ichthyosis – erythroderma
  - Incontinentia Pigmenti
  - Congenital Porphyria – photosensitive, red urine
    Immune Mediated (Maternal Antibodies)
    - Diffuse Cutaneous Mastocytosis – erythroderma, flushing, blisters with rubbing

- Aplasia Cutis Congenita – scalp, extremities
- Hemangioma Precursor – erosion in diaper area

Localized

- Langerhans Histiocytosis – erosions in skin folds and diaper area
- Toxic Epidermal Necrolysis – drug exposure or graft versus host disease/severe combined immunodeficiency
- Congenital Erosive Dermatitis with Reticulate Scarring

Epidermolysis Bullosa

• Genetic disorders of the skin and mucosa
• Manifest with blisters and erosions as a response to minor mechanical trauma
• 4 major types
  – EB Simplex (EBS) – Intraepidermal
  – Junctional EB (JEB) – Intra-lamina lucida
  – Dystrophic EB (DEB) – Sub-lamina densa
  – Mixed (Kindler syndrome)

EB biopsy

- Routine histology most useful to rule out other conditions
- Biopsy an induced blister for IF and EM studies
- Use a lab experienced in EB diagnosis
  - Stanford Dermatopathology (IF and EM)
    - dermatopathology.stanford.edu
  - Beutner Labs (IF only)
    - www.beutnerlabs.com
- Genetic testing is confirmatory and is becoming a first-line test
  - GeneDX: www.genedx.com
EB biopsies are taken from an induced blister

- Choose an area of unaffected skin
- Draw a small circle on the skin
- Anesthetize the area
- Using firm pressure, twist a clean pencil eraser firmly back and forth in the circle OR use a gloved finger or thumb to rub the area
- Erythema should be visible, but a blister may not be apparent.
- Clean the skin, leaving the marked circle
- Biopsy, aiming for approximately one-third of the induced blister and two-thirds normal skin with each biopsy.
Incontinentia Pigmenti

• X-linked recessive; mosaic
• Deletion in *IKBKG*
• Ophthalmology consult crucial in the neonatal period
• Neurologic complications are less common than reported in literature
Diffuse cutaneous mastocytosis

• Diffuse infiltration of the skin by mast cells
• Skin appears thickened, yellowish to tan, with peau d’orange texture
• Risk for systemic disease
  – Flushing
  – Temperature elevation
  – Vomiting, diarrhea, abdominal pain
  – Respiratory distress
Older infants, children, adolescents

Blisters

Infectious
Bullous Impetigo
&
Staphylococcal scalded skin syndrome

• Exfoliative skin diseases caused by epidermolytic toxin-producing *S. Aureus*
• Bullous impetigo = toxin localized to areas of infection
• SSSS = toxin is disseminated
Atypical Hand-Foot-Mouth

• Due to coxsackie virus A6
• Lesions more widespread, facial-acral distribution
• Predilection for areas of eczematous dermatitis (“eczema coxsackium ”) and prior skin injury
Older infants, children, adolescents

Blisters

Immune-mediated
Immune-mediated bullous disease

- Biopsy is needed to make the diagnosis
- Lesional skin for histology
- Peri-lesional skin for IF
Immune-mediated bullous disease: therapy

• Oral corticosteroids for acute control
• IgA disease -> dapsone
• Other adjuncts: antibiotics, topicals
• Steroid sparing immune suppressive agents
• IVIg, Rituximab
Blistering: take home points 1

• After ruling out infections, consider genetic etiologies in neonates with blisters
• Routine histology not useful for EB but is helpful for other conditions
• EB biopsies are taken from induced blisters
Blistering: take home points 2

- In older infants, children and adolescents, consider acute infections and reactive conditions first
- Immune-mediated bullous disorders are uncommon but do occur!
- Diagnosis requires histology and IF studies