July/ 2022

Department of Health Research
Ministry of Health and Family Welfare, Government of India

INDIAN COUNCIL OF MEDICAL RESEARCH
Serving the nation since 1911

Standard Treatment Workflow (STW)

IMMUNOBULLOUS DERMATOSES

ICD-10-L13.8

WHEN TO SUSPECT?

- Appearance of fluid-filled, itchy or painful blisters (either flaccid or tense) on skin, over a normal or erythematous base
- Appearance of raw, erythematous erosions ± crusting on skin
- Appearance of erosions/ blisters inside oral cavity, eyes, nose and genitals

AUTOIMMUNE BLISTERING DISEASES

- Pemphigus vulgaris/ variants
- Pemphigus foliaceus/ variants
- Bullous pemphigoid
- Pemphigoid gestationis
- Mucous membrane pemphigoid
- Linear IgA bullous diseases/ chronic bullous disease of childhood
- Dermatitis herpetiformis
- Epidermolysis bullosa acquisita
- Bullous systemic lupus erythematosus

ADDITIONAL INFORMATION

- Age at onset and duration of blistering
- History of any recent drug intake
- History of prior varicella/ chicken pox
- History of similar illness in family
- History of itching, pain, burning
- Predominant sites affected
- Associated photosensitivity

EXAMINATION

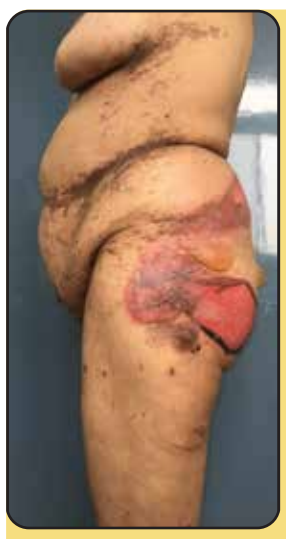
- Are the blisters flaccid or tense?
- Are the erosions crusted?
- Do the blisters contain clear or hemorrhagic fluid?
- Are the blisters umbilicated?
- Is the base of the blisters erythematous/ urticarial?
- Are the blisters healing with or without scarring?
- Are they healing leaving behind hyper/hypopigmentation?
- What is the color of the crust?
- Are mucosae involved?

DIAGNOSIS OF AUTOIMMUNE BULLOUS DISEASES

- **Likely pemphigus group of autoimmune bullous diseases**
  - Flaccid blisters/ erosions ± crusting on skin ± mucosae
  - Usually seen in adults; can rarely affect children
- **Likely sub-epidermal autoimmune bullous diseases**
  - Tense, small to large blisters, containing clear or hemorrhagic fluid, on an itchy erythematous base, commonly healing with hypopigmentation ± scarring
  - Seen in children, adults and elderly (most common is bullous pemphigoid)

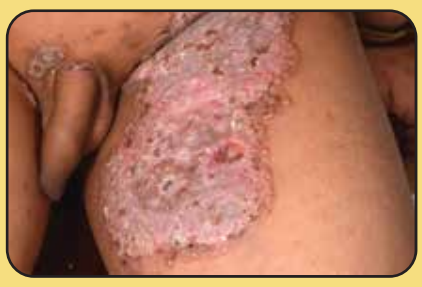
- Get a Tzanck smear
- Get a biopsy for histopathology from margin of a lesion
- Get a peri-lesional biopsy for direct immunofluorescence, if facility is available

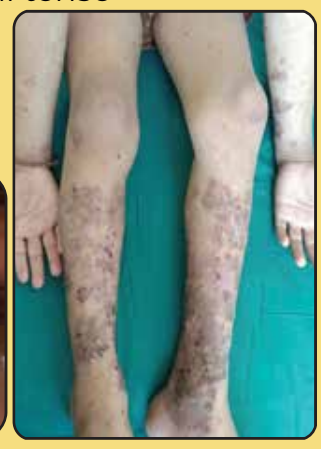
PEMPHIGUS



BULLOUS PEMPHIGOID

- Child < 5 years
- Erosions with peripheral tense blisters
- Urticarial base
- Face/ peri-genital involvement





CHRONIC BULLOUS DISEASE OF CHILDHOOD

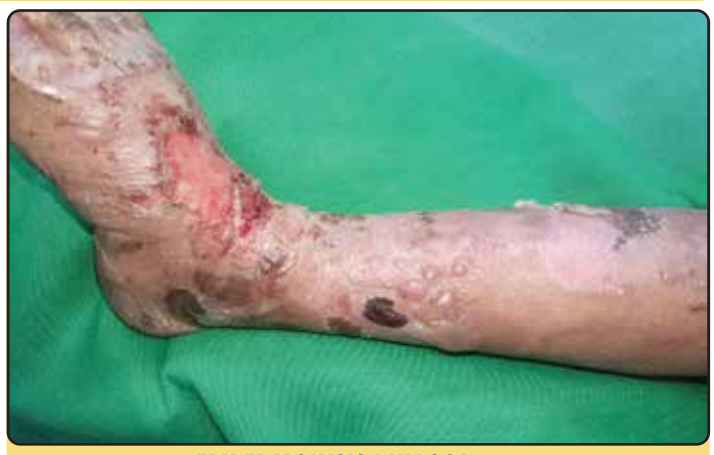
RED FLAG SIGNS

- Fever ± chills and rigors
- Hypotension (indicating hypovolemia due to fluid loss or sepsis)
- Altered sensorium (indicating dyselectrolytemia or sepsis)

DIFFERENTIAL DIAGNOSES

- **Bullous Impetigo, Varicella, Stevens Johnson Syndrome/TEN***
- **Epidermolysis bullosa,** a hereditary blistering disease with onset in neonatal period or infancy and predominantly affecting pressure sites; presence of scarring on limbs, acral areas, trunk and abnormality of the teeth or nails
- Consider **Congenital syphilis** in a neonate- get VDRL for mother and child

*Refer to STW on Bacterial Infections; Varicella and Herpes Zoster and cADR Part B



EPIDERMOLYSIS BULLOSA

GENERAL MEASURES

- Monitor temperature, respiratory rate, pulse rate
- Administer antibiotics if lesions are infected and foul smelling
- Fluid-electrolytes balance
- Get hemogram, basic biochemistry including renal and hepatic function tests, blood sugar
- Get pus culture and if sepsis is suspected, also blood culture
- Supportive management
  - Clean non-adherent dressings
  - Maintain hygiene with normal soap bath
  - Topical antibiotics
  - Aspiration of large blisters with 18G needle if needed
  - Avoid deroofing the blisters as the roof of the blister acts as a natural dressing

- Maintain oral hygiene (if involved)
  - Chlorhexidine mouth wash
  - Brush teeth with pediatric brush with small head and soft bristles
  - Avoiding eroding gingival margin
- Maintain skin hygiene (if involved)
  - Diluted potassium permanganate bath/ potassium permanganate compresses on localized lesions/ thick crusted lesions
  - Emollients/ coconut oil application
  - 2% savlon scalp wash
- Encourage oral intake (fluids and calories); consider other comorbidities
  - Liquid/ semisolid diet for oral erosions

PEMPHIGUS (START TREATMENT ONLY IF FACILITY FOR MONITORING AND MANAGEMENT OF COMPLICATIONS OF TREATMENT IS AVAILABLE)

- **Mucosal/ mucocutaneous with body surface area <5%**
  - Oral Prednisolone (0.5 mg/kg/day), with one or more of the following
    - Azathioprine (2-3 mg/kg/day)
    - Mycophenolate mofetil (35mg/kg/day, start at a lower dose)
    - Cyclophosphamide (1-2 mg/kg/day)
    - Methotrexate (0.3mg/kg/week)
    - Dapsone (100-150 mg/day)
- **Mucocutaneous with body surface area >5%**
  - At primary level-Stabilize patient, initiate general measures and refer to a specialist/ tertiary level
  - To be managed at a tertiary level
    - Dexamethasone- Cyclophosphamide pulse therapy
    - Rituximab

BULLOUS PEMPHIGOID (START TREATMENT ONLY IF FACILITY FOR MONITORING AND MANAGEMENT OF COMPLICATIONS OF TREATMENT IS AVAILABLE)

- **Limited (<10% body surface area)**
- Start treatment and refer to tertiary level
- Topical Clobetasol propionate (upto 30 gm/day)
- Oral Prednisolone (0.5 mg/kg/day) ±
  - Dapsone (100-150 mg/day)
  - Doxycycline (100- 200 mg/day)
  - Niacinamide (500 mg thrice/day)
  - Azathioprine (2-3 mg/kg/day, start at a lower dose)
  - Mycophenolate mofetil (35mg/kg/day, start at a lower dose)
  - Methotrexate (0.3mg/kg/week)
- **Extensive (>10% body surface area)**
- To be managed at a tertiary level
- Oral Prednisolone (0.75- 1 mg/kg/day) ±
  - Dapsone
  - Doxycycline
  - Niacinamide
  - Azathioprine
  - Mycophenolate mofetil
  - Methotrexate

CORRECT DIAGNOSIS; PREVENTION/ TREATMENT OF SEPSIS; AND REGULARITY OF TREATMENT BRINGS BEST RESULTS