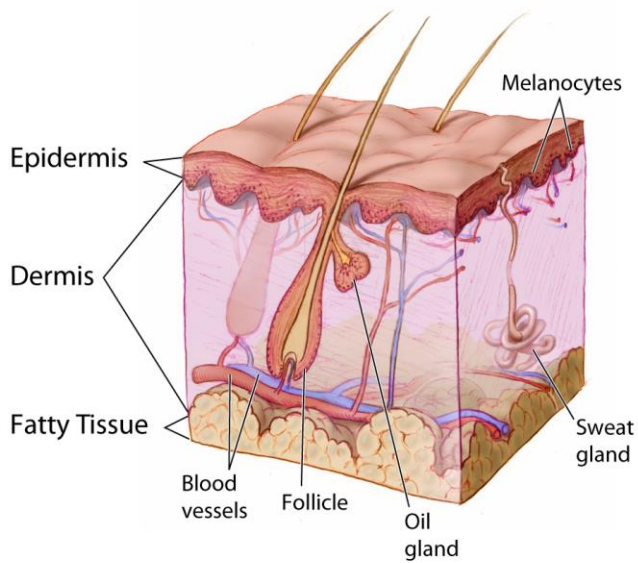


432 Teams

# Dermatology



## Blistering Diseases



Color Code: Original, **Team's note**, **Important**, **Doctor's note**, Not important, **Old teamwork**



Done by: *Muath ALSoliman*

Reviewer: *Bader ALMosned*

Team Leader: *Basil Al Suwaine*

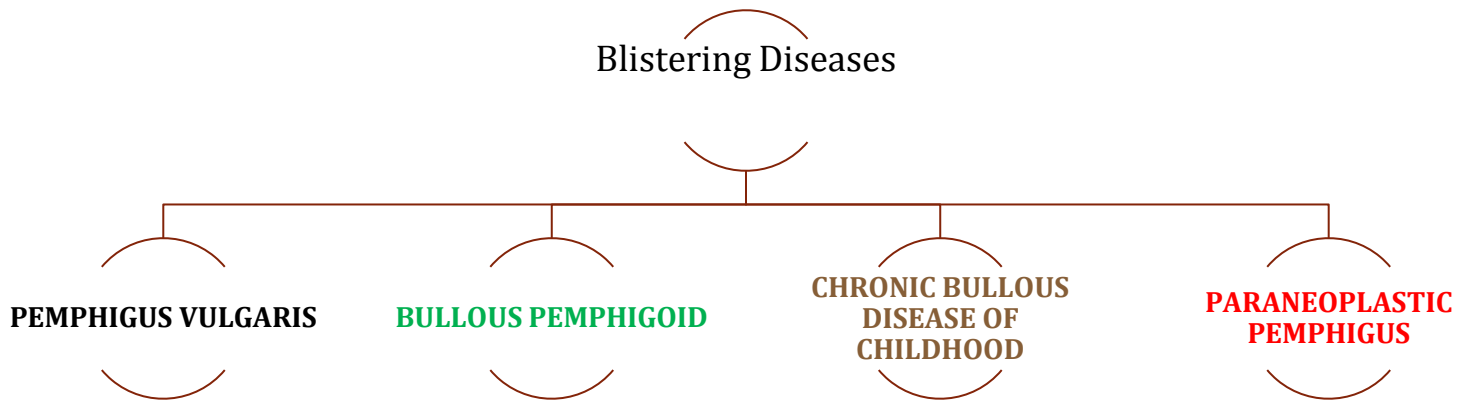
# Objectives

## Objectives

- To know the definition & classification of Blistering diseases
- To recognize the primary presentation of different types of main blistering diseases
- To understand the possible pathogenesis of the main types of blistering diseases
- To have an overview about managements lines of these diseases

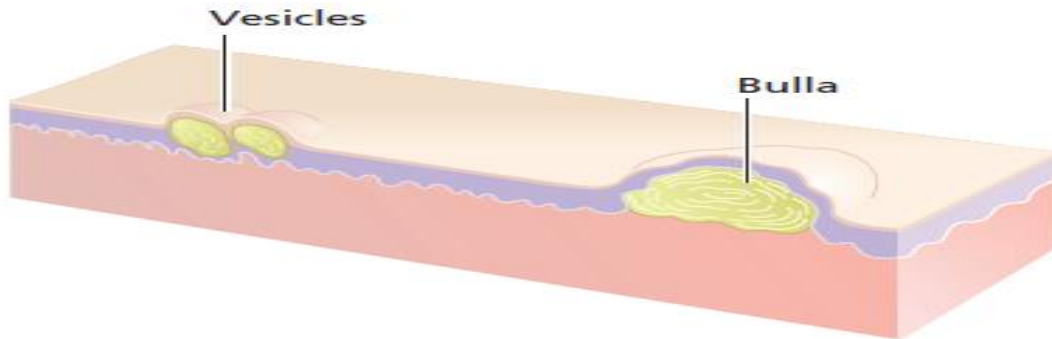
### Note:

- ✓ Doctor mentioned some important note for MCQ exam it will be red and underlined
- ✓ Special thanks for 431team



**Definition:**

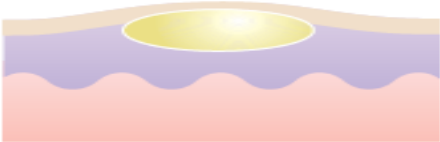
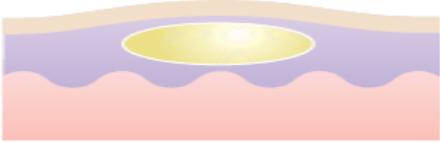

- Vesicles and bullae are raised lesions that contain fluid.
- A vesicle is less than 0.5 cm in diameter.
- A bulla is larger than 0.5 cm in diameter.



**CLASSIFICATION OF VESICULOBULLOUS DISEASES:**

**INTRA EPIDERMAL BLISTERS :** The lesion is formed within the epidermis

**SUB EPIDERMAL BLISTERS :** Lesions formed between the epidermis and the dermis

<b>Location of bullae</b>	<b>Diseases</b>
 <p style="text-align: center;"><b>Subcorneal bulla</b></p>	<p>Bullous impetigo Miliaria crystallina Staphylococcal scalded skin syndrome</p>
 <p style="text-align: center;"><b>Intra-epidermal bulla</b></p>	<p>Acute eczema Viral vesicles Pemphigus Miliaria rubra Incontinentia pigmenti</p>
 <p style="text-align: center;"><b>Subepidermal bulla</b></p>	<p>Bullous pemphigoid Cicatricial pemphigoid Pemphigoid gestationis Dermatitis herpetiformis Linear IgA disease Bullous erythema multiforme Bullous lichen planus Bullous lupus erythematosus Porphyria cutanea tarda Toxic epidermal necrolysis Cold or thermal injury Epidermolysis bullosa</p>

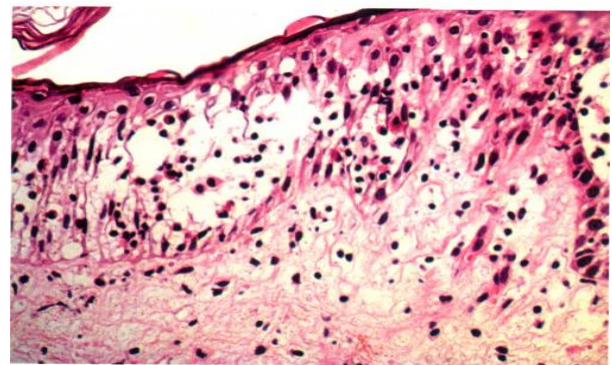
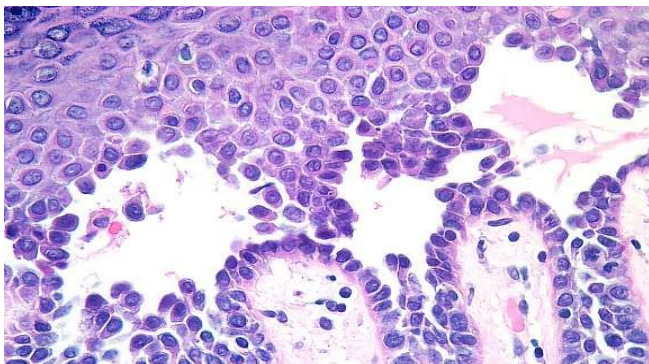
- Accurate pathological diagnosis requires a biopsy of a small newly formed lesion and of perilesional skin for immunopathological studies.

**Diagnostic tests:**

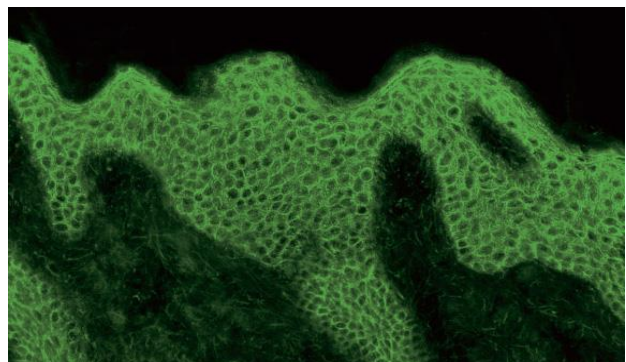
1. **Routine histology**
  - Lesional sample –small bulla or edge of large one.
2. **Direct immunofluorescence**
  - Perilesional sample
3. **Indirect immunofluorescence**
  - Patient’s serum is added to specific substrates that express antigen of interest.
4. **Electron microscopy.**



-Yellow : Routine histology  
-Green : Direct immunofluorescence



(Routine histology)

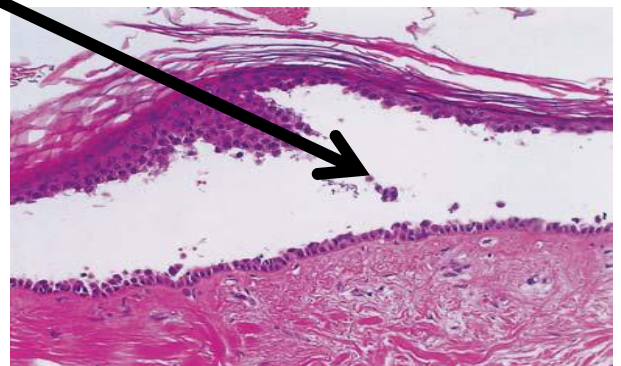
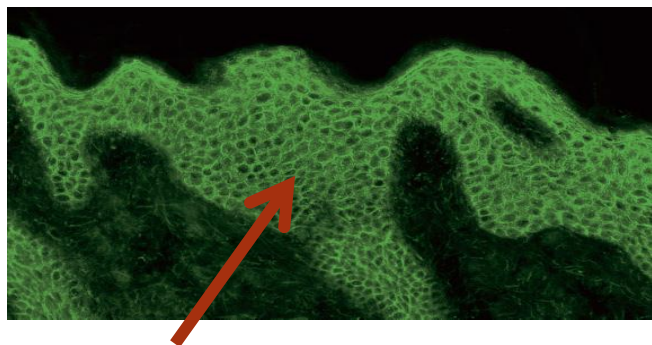
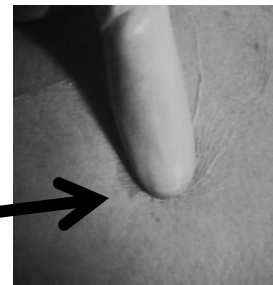


(Sub-epidermal)

(Intra-epidermal)

## Pemphigus Vulgaris > most common

- Pemphigus is a group characterized by blistering of the skin and mucous membranes.
- Age: middle-age 40-60 years.
- Auto-antibodies against
- DESMOSOMES in epidermis and mucosal surface. (IgG , C3)
- Four sub-clinical variants:  
**Pemphigus Vulgaris: is the most common Pemphigus variant, and the form usually responsible for oral lesions,** Others are: Foliacious, vegetens, erythematosus.
- Begins with erosions on mucous membrane then other skin areas.
- Mostly erosions and few flaccid blisters
- Very painful.
- Fatal in all cases if not treated (431 team)
- **+ve Nikolsky's sign:** twisting pressure on normal skin shears skin
- **Secondary infection and disturbance of fluid and electrolyte balance are common complications .**
- **Acantholysis** "Floating cells" is a characteristic feature in routine histology. (Acantholysis is the loss of intercellular connections, such as desmosomes, resulting in loss of cohesion between keratinocytes )



**Immunofluorescence**  
**IgG and C3 " intra-epidermal "**

**Treatment:**

- **High dose systemic steroids** (prednisolone 60-100 mg)
- Immunosuppressive agent such as azathioprine cyclophosphamide , Methotrexate or mycophenolate
- Topical therapy is mainly symptomatic.
- Patient will probably have to remain on systemic steroids for long time.
- Antibiotics
- Biologic Rx: Rituximab or IVIG.
- IF MILD: class III/IV corticosteroid creme / intralesional injection.
- IF SEVER: prednisolone 80 mg & taper in 5 months, immunosuppressive, biological.

<b>Drug-induced PV</b>
------------------------

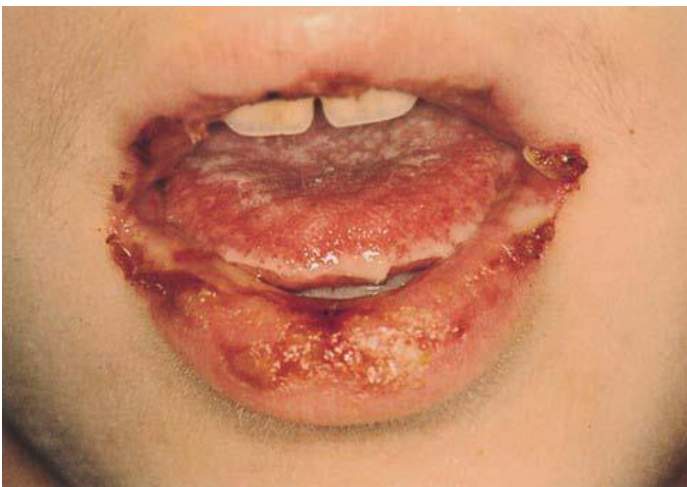
<u><b>Penicillamine , captopril</b></u>
---

---

### Paraneoplastic pemphigus > **Malignant**

---

- The least common and most severe type of pemphigus is paraneoplastic pemphigus (PNP). This disorder is **a complication of cancer**.
- usually lymphoma and Castleman's disease. It may precede the diagnosis of the tumor. Painful sores appear on the mouth, lips, and the esophagus.
- Complete removal and/or cure of the tumor may improve the skin disease.
- Both Intra epidermal and sub epidermal blister.
- Autoantibody ( **IgG , IgA , C3** )



---

## Bullous pemphigoid > good prognosis

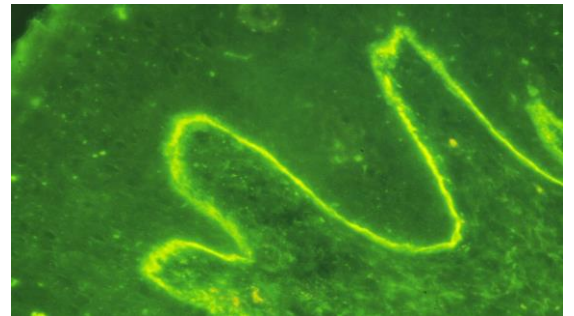
---

- Characterized by **large, tense, intact blisters** on an erythematous base. it can erupt to form erosions.
- Mainly in older age group.
- more than 60 y.
- The prognosis is usually good.
- **Linear band on immunofluorescence.**
- **Antigens identified are in hemidesmosomes.**



### Clinical features :

- Elderly patents.
- Large tense blisters on upper arms and thighs.
- Eczematous base .
- Itch rather than pain.
- Oral lesions are less frequent than pemphigus.



### Pathology :

- Sub epidermal between epidermis and dermis the epidermis forms the roof of the blister.
- Antigens identified are in hemidesmosomes.
- Immunoglobulin and complement are deposited in the lamina lucida of the basement membrane in a linear band.

### Treatment :

- Topical Steroids
- Severe pemphigoid :Systemic steroids , but unlike pemphigus, usually possible to discontinue Rx.
- The addition of either azathioprine enable the oral steroid dose to be reduced more rapidly.
- Milder may also respond very well to potent or moderately potent topical steroids alone.
- Other treatment: Antibiotics like tetracyclines group



---

## CHRONIC BULLOUS Disease OF CHILDHOOD

---

- Chronic blistering dis. which occur in children, usually starts before the age of 5yrs
- Small and large blisters appears predominantly on the lower trunk, genital area, and thighs
- May also affects the scalp and around the mouth
- New blisters form around healing old blisters forming **a CLUSTER OF JEWELS**
- Course: is chronic and spontaneous remission usually occurs after an average of 3-4 yrs
- **IgA autoantibodies** binds to proteins at dermo epidermal junction as linear pattern like the pattern of bullous pemphigoid



### CLINICAL FEATURES :

- Circular clusters of large intact blisters and can erupt to form erosions
- It involves the perioral area, lower trunk, inner thighs and genitalia
- Blistering may spread all over the body

### INVESTIGATION :

- Skin Biopsy will show **subepidermal splits**
- Direct IF reveals **IgA** along the BM of the epidermis in a **linear pattern**

### TREATMENTS :

- **Oral dapsone**  
(may cause hemolysis , check G6PD or methemoglobinemia )
- Sulphonamides and immunosuppressants
- Erythromycine
- Flucloxacillin

## Questions ( 431 team )

1 . Fifty five year old teacher presented with sever oral mucusal ulceration. Moreover, he developed numerous flacid blisters which ruptured easily leaving only erosions. The most likely diagnosis is:

- A. Bullous Pemphigoid
- B. Pemphigus Vulgaris
- C. Pyoderma gangrenosum
- D. Stevens-Johnson syndrome
- E. Toxic Epidermal Necrolysis

2. The mortality in Pemphigus Vulgaris is due to:

- A. Inadequate food intake
- B. Renal failure
- C. Secondary Infection
- D. Squamous cell carcinoma
- E. None of the above

3. keratinocytes detach from their neighbors & float free in the blister a process called:

- A. Acantholysis.
- B. CLUSTER OF JEWELS.
- C. Sub-epidermal splits
- D. Acanthosis

4. Patient diagnosed with blister disease, the IF showed aggregation of IgG along with the basement membrane zone, what's the Most likely DX?

- A. Pumphigus vulgaris.
- B. Bollus Pumphigoid
- C. Dermatitis herpetiformis
- D. Chronic bulls disease of childhood

**Answers:**

1-B

2-C

3-A

4-B