Vesiculobullous diseases

Vesicle: Asuperficial blister(bulging), 5mm or less in diameter, usually filled with clear fluid.

Bullae: Same as vesicle, but of larger size , more than 5mm in diameter.

Pustule: A blister that filled with a purulent exudate.

Erosion: Partial loss of surface epithelium.

Fissure: A narrow, slit- like ulceration or groove.

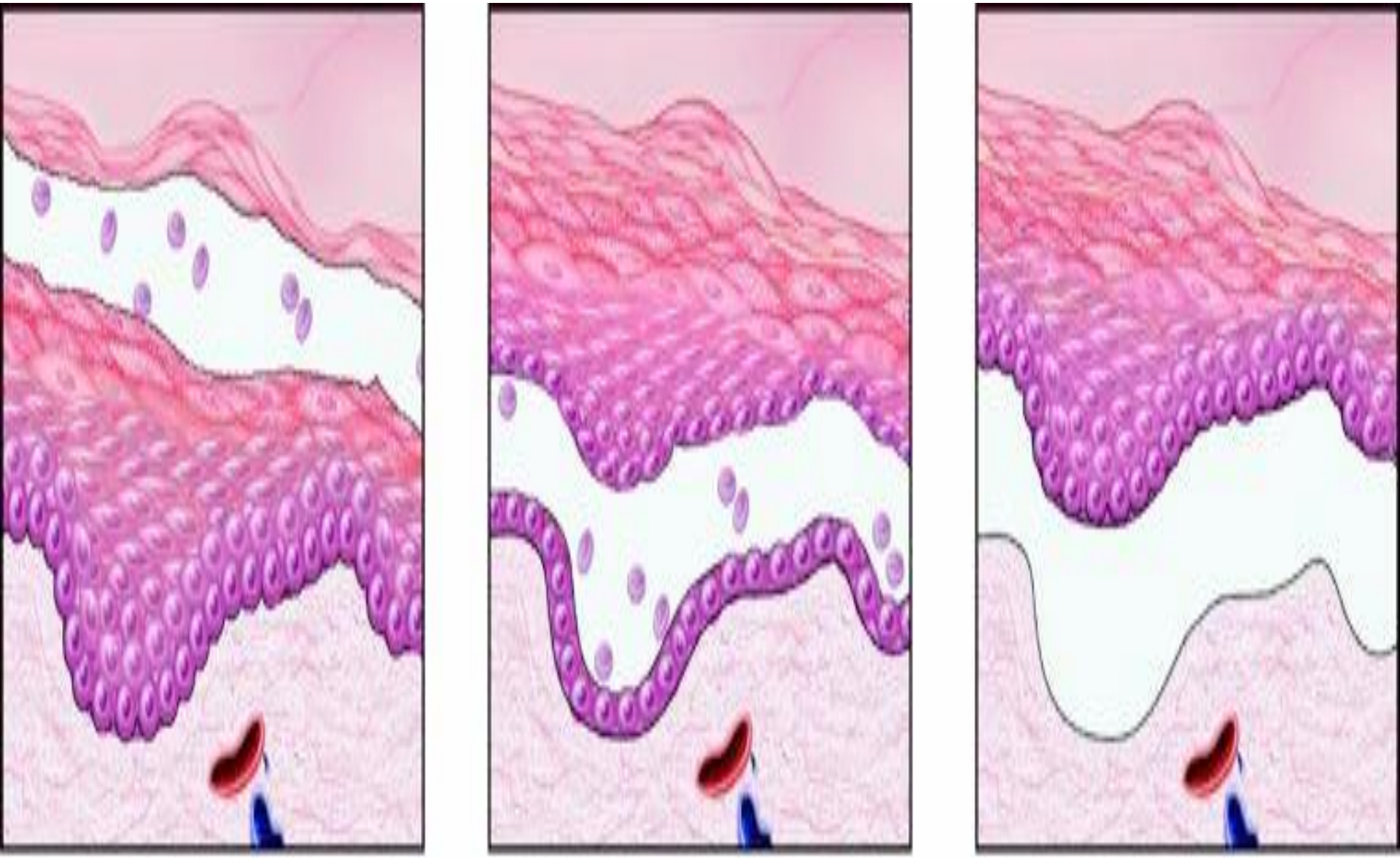
CLASSIFICATION OF VESICULOBULLOUS DISEASES

INTRA EPITHELIAL VESICLES: The lesion is formed within the epithelium

- **Acantholytic vesicles** : This is because of the break down of specialized attachments called the desmosomes
- **Nonacantholytic vesicles**: It is usually in the viral infections because of the death or the rupture of the group of cells.

SUB EPITHELIAL VESICLES: Lesions formed between the epithelium and the lamina propria eg:

- Erthyma multifome
- Phempegoid
- Dermatitis herpetiformis
- Epidermolysis bullosa


A Subcorneal
B Suprabasal
C Subepidermal

PEMPHIGUS VULGARIS

PEMPHGOID

ERYTEMA MULTIFORME

EPIDERMOLYSIS BULLOSA

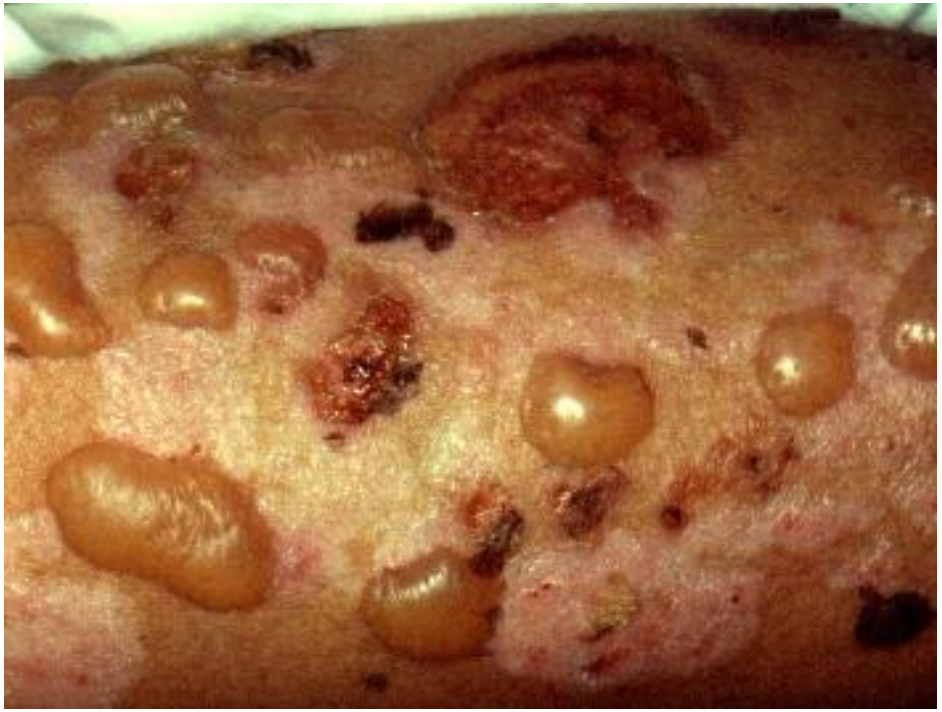
PEMPHIGUS VULGARIS

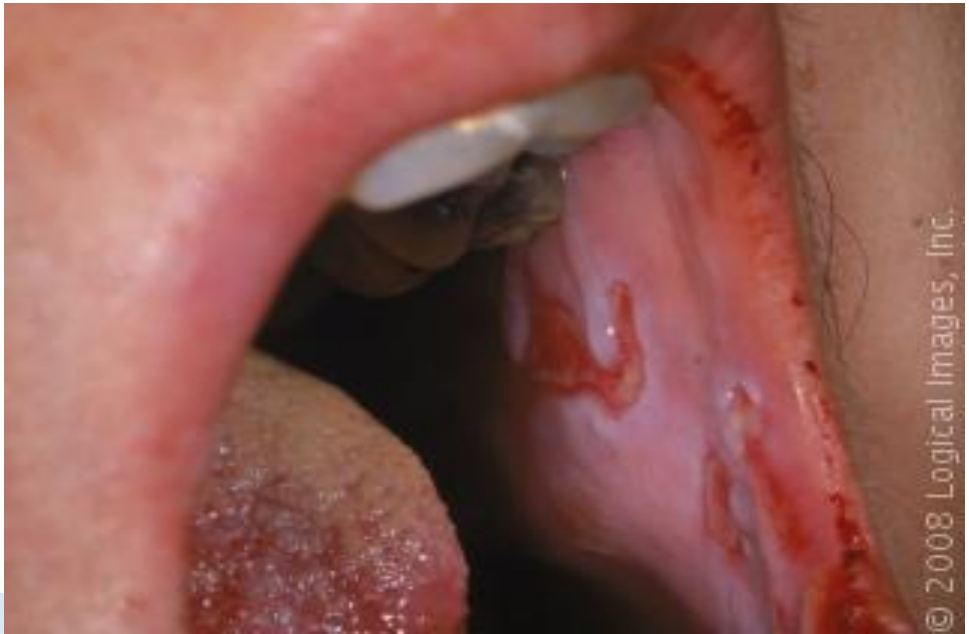
- Autoimmune disease.
- Common in Ashkenazic & Mediterranean jewish .
- Middle aged females.
- Other variants are:
  - Pemphigus Vegitans
  - Pemphigus Foliaceus & Erthematosus
  - Paraneoplastic pemphigus.

PEMPHIGUS VULGARIS

CLINICAL FEATURES:

- Painful ulcers or bulla are formed which are fluid filled.
- They can be formed any where in the oral cavity .
- The bulla is rapidly ruptured leaving a collapsed roof of grayish membrane with a red ulcerated base.The ulcer may look like an apthous ulcer or may be large map shaped.
- Nikolsky sign is positive.

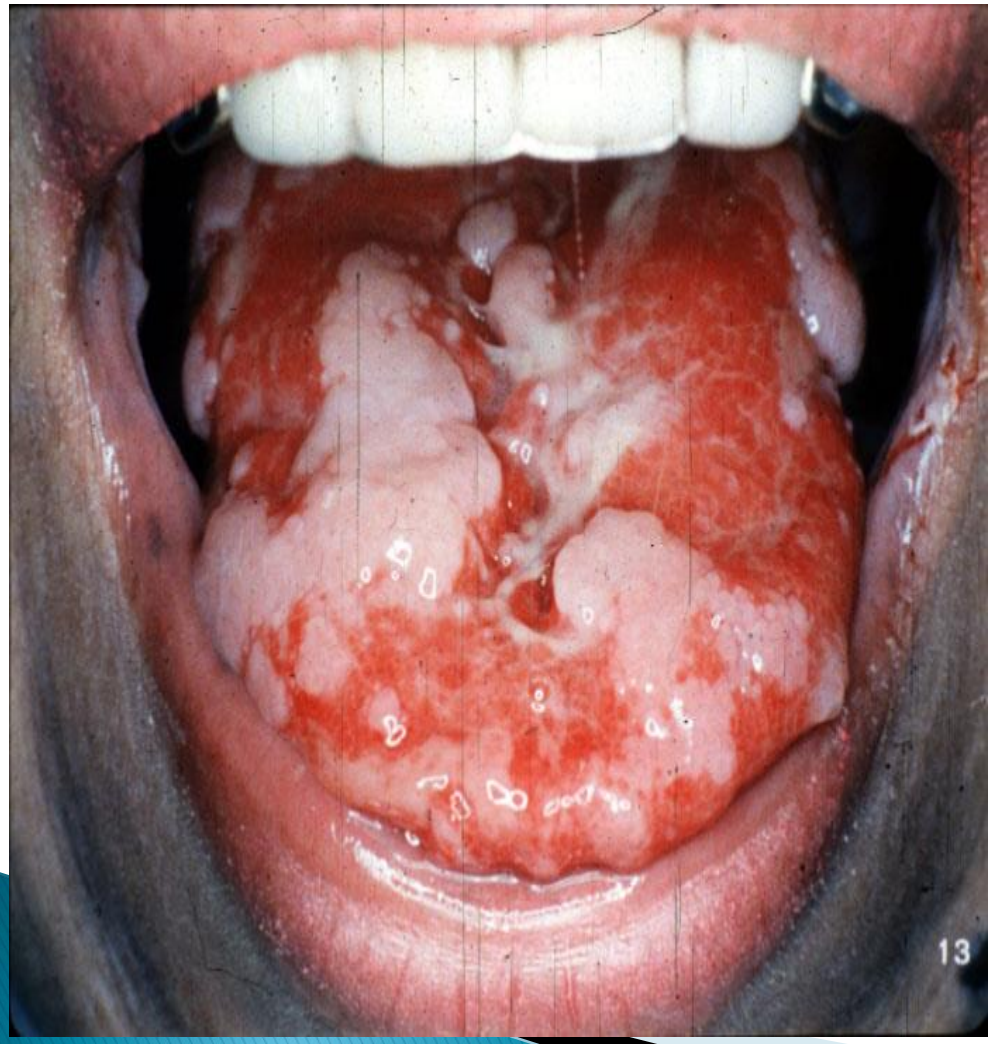


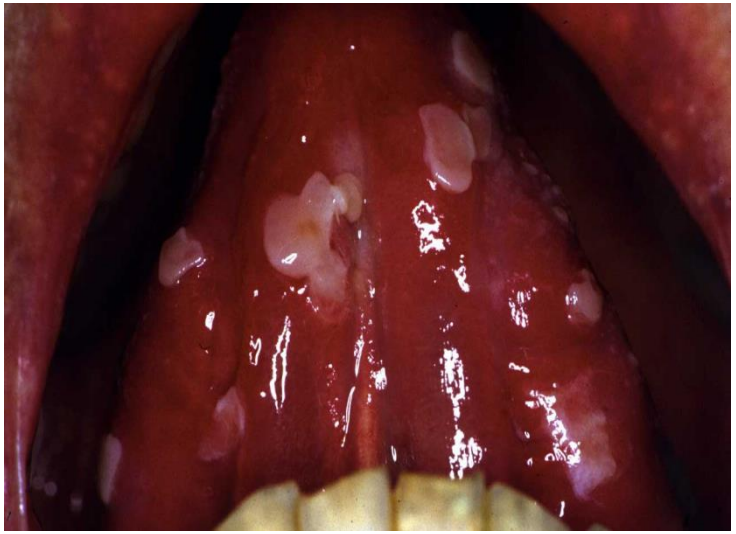


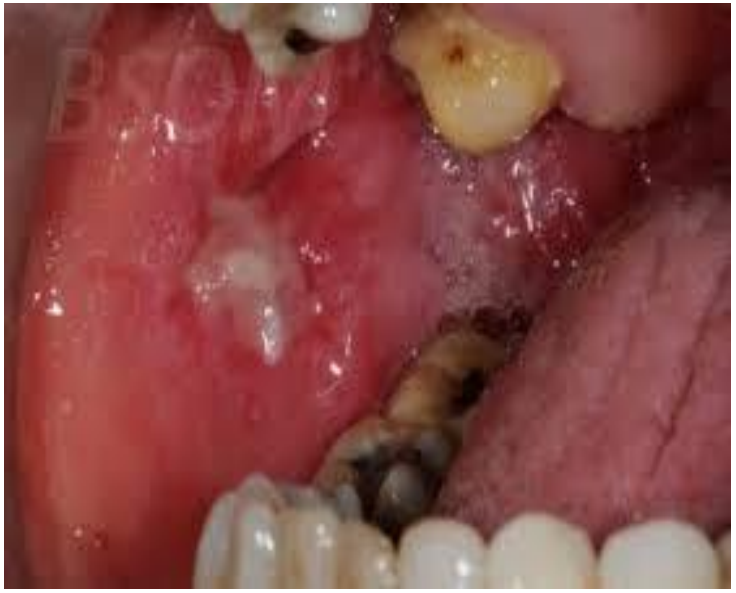
PEMPHIGUS VULGARIS

- Some time the ulcers are joined together to make a confluence this condition is very painful.
- It has a variable course might involve skin, oesophagus, cervix.
- Protein/fluid,electrolyte & weight loss /secondary infections.
- Fatal if untreated.

50% or more of cases begin in the mouth (first to show last to go)







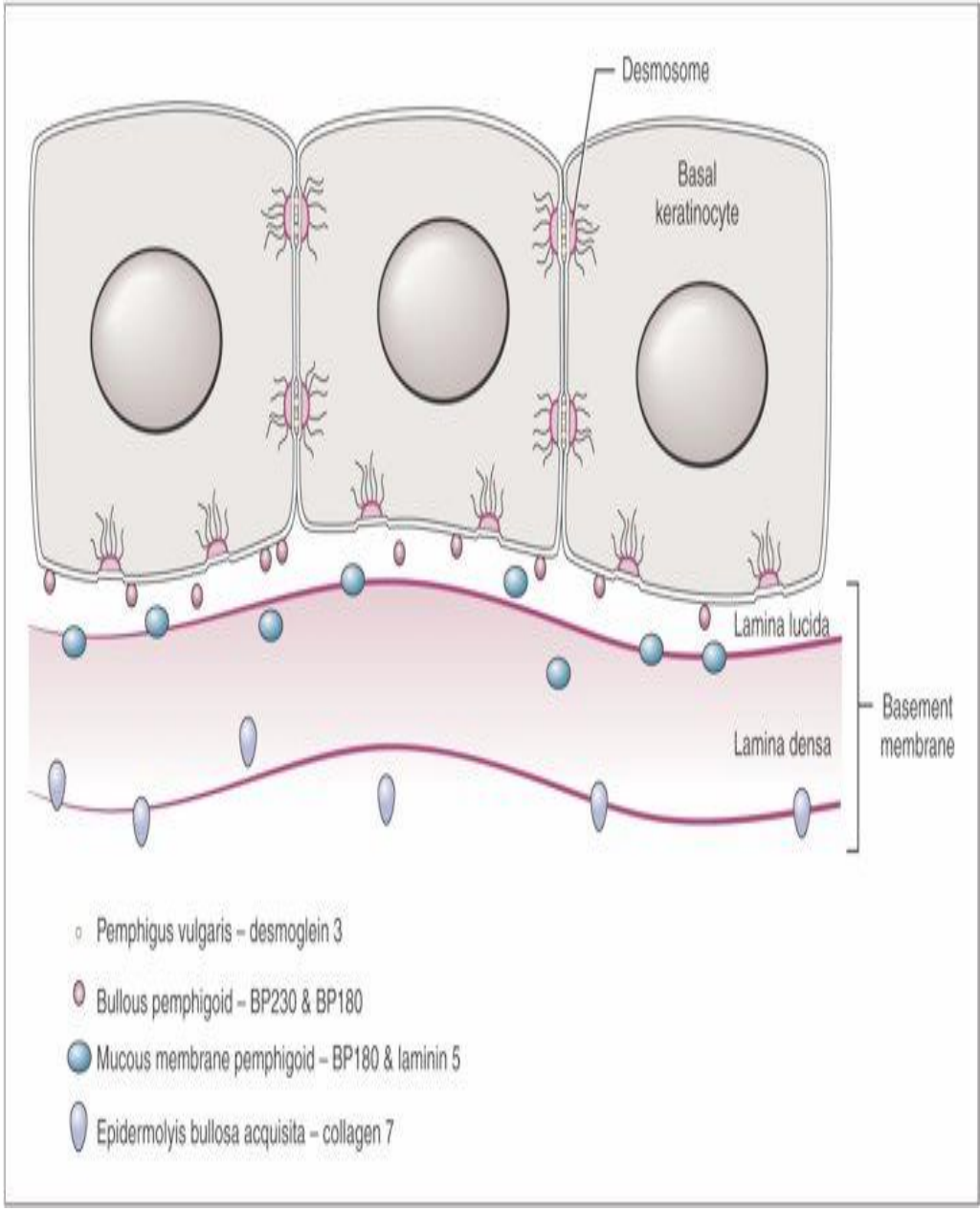
PEMPHIGUS VULGARIS

PATHOGENESIS:

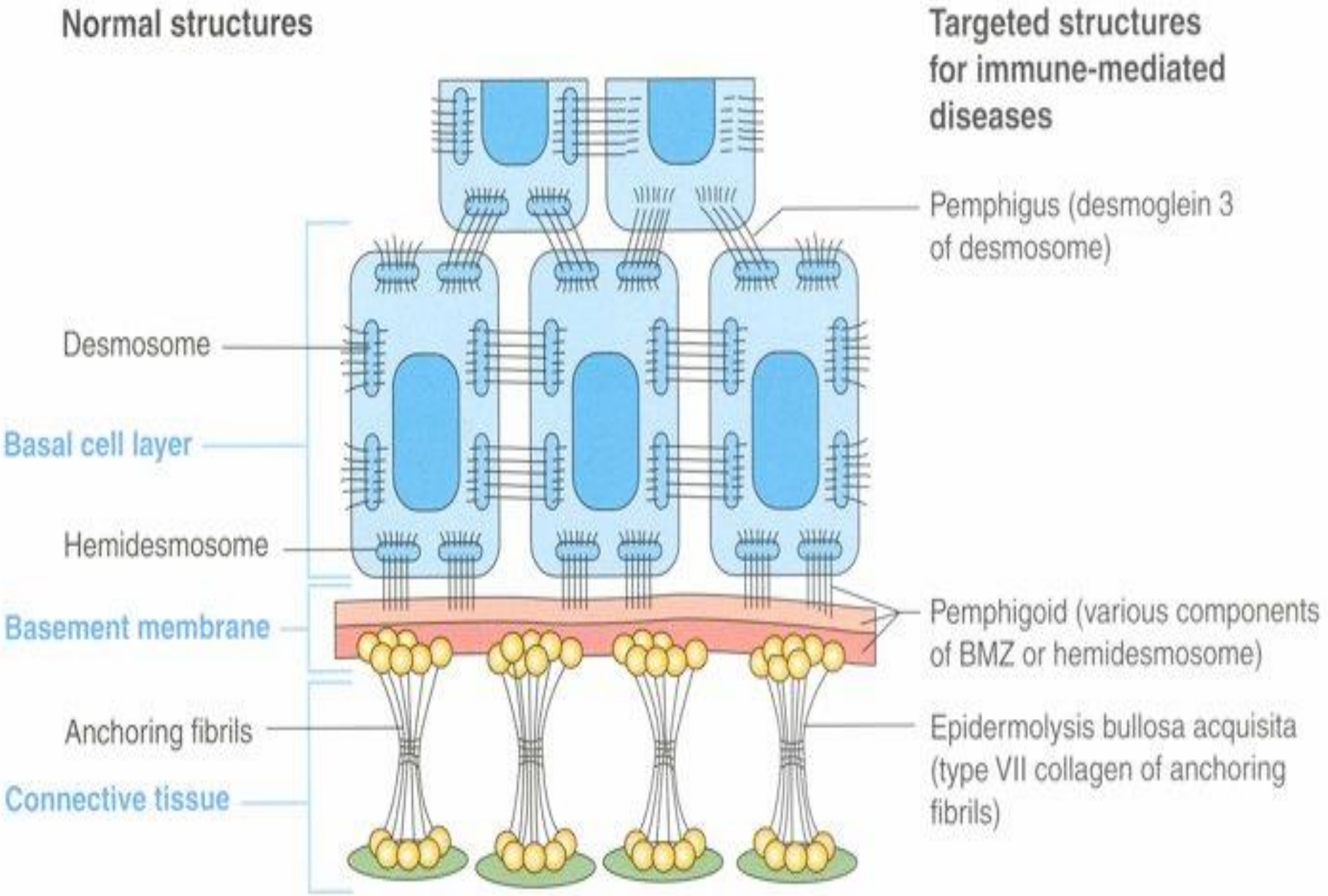
- It is an autoimmune disease
- There are circulating antibodies of type IgG.
- These antibodies are reactive against the desmosomes or the tonofilament complex.
- The destruction or disruption of these tonofilament complex ,resulting in the loss of attachment from cell to cell

PEMPHIGUS VULGARIS

- The epithelial damage is directly proportion to the number of the circulating antibobies.
- The tonofilament or desmosomes are disrupted by a proteolytic enzyme which is released by these antibodies .
- The cell to cell break down also takes place through a complement system but this process is not clearly understood .

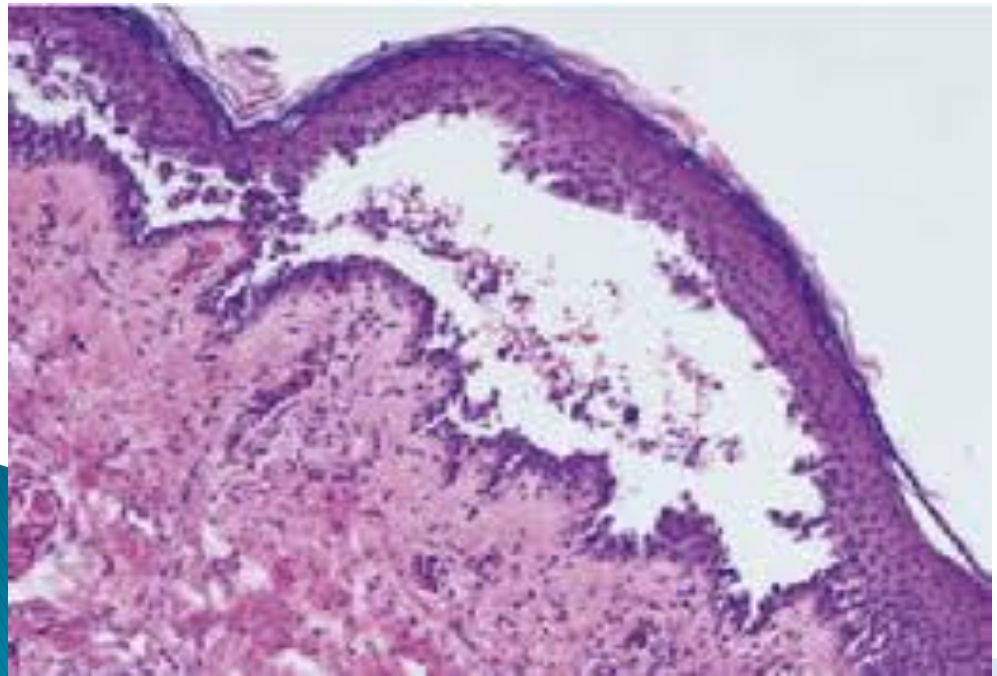


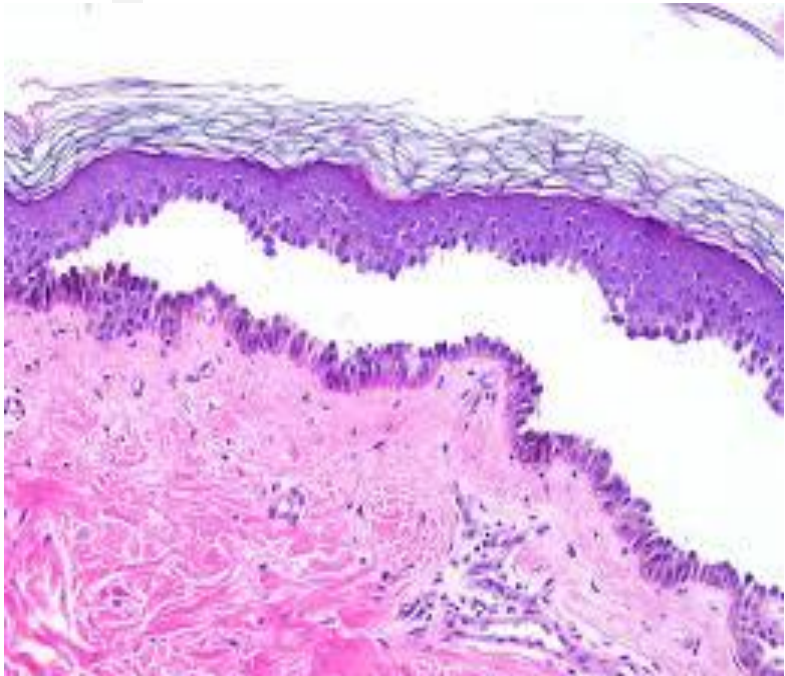
Epithelial attachment apparatus



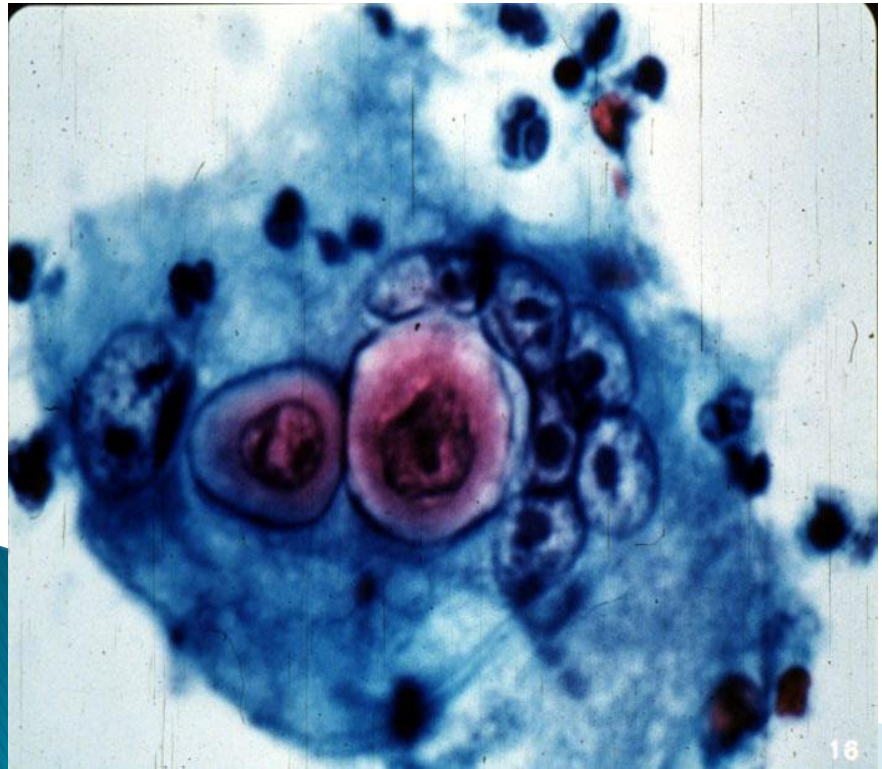
HISTOPATHOLOGY:

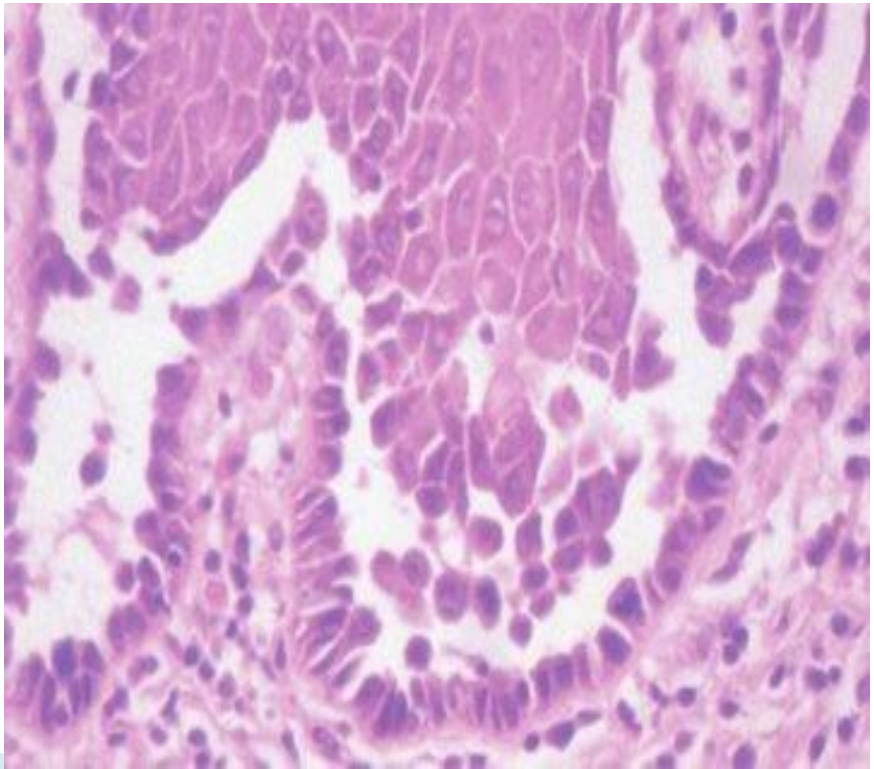
- Intra epithelial vesicles or bulla and cleft like spaces are produced by acantolysis
- These changes are in the stratum spinosum or the prickle cell layer
- The basal cell remain attach to the lamina propria and project into the bulla like tombstones.
- Inflammatory cells are very scanty however eosinophils may be seen.





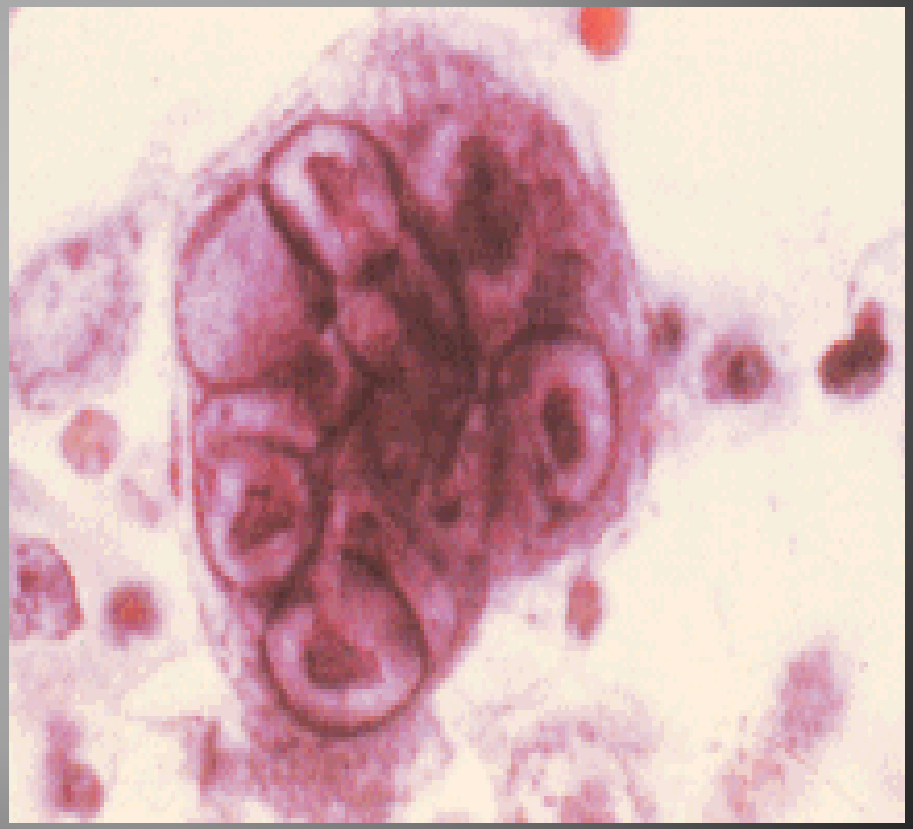
Acantholytic stratum spinosum cells occur singly or are in the forms of clumps lying freely within the blister fluid. These cell loose theire polyhedral morphology rather they are small rounded&contain hyperchromatic nuclei called Tzanck cells.



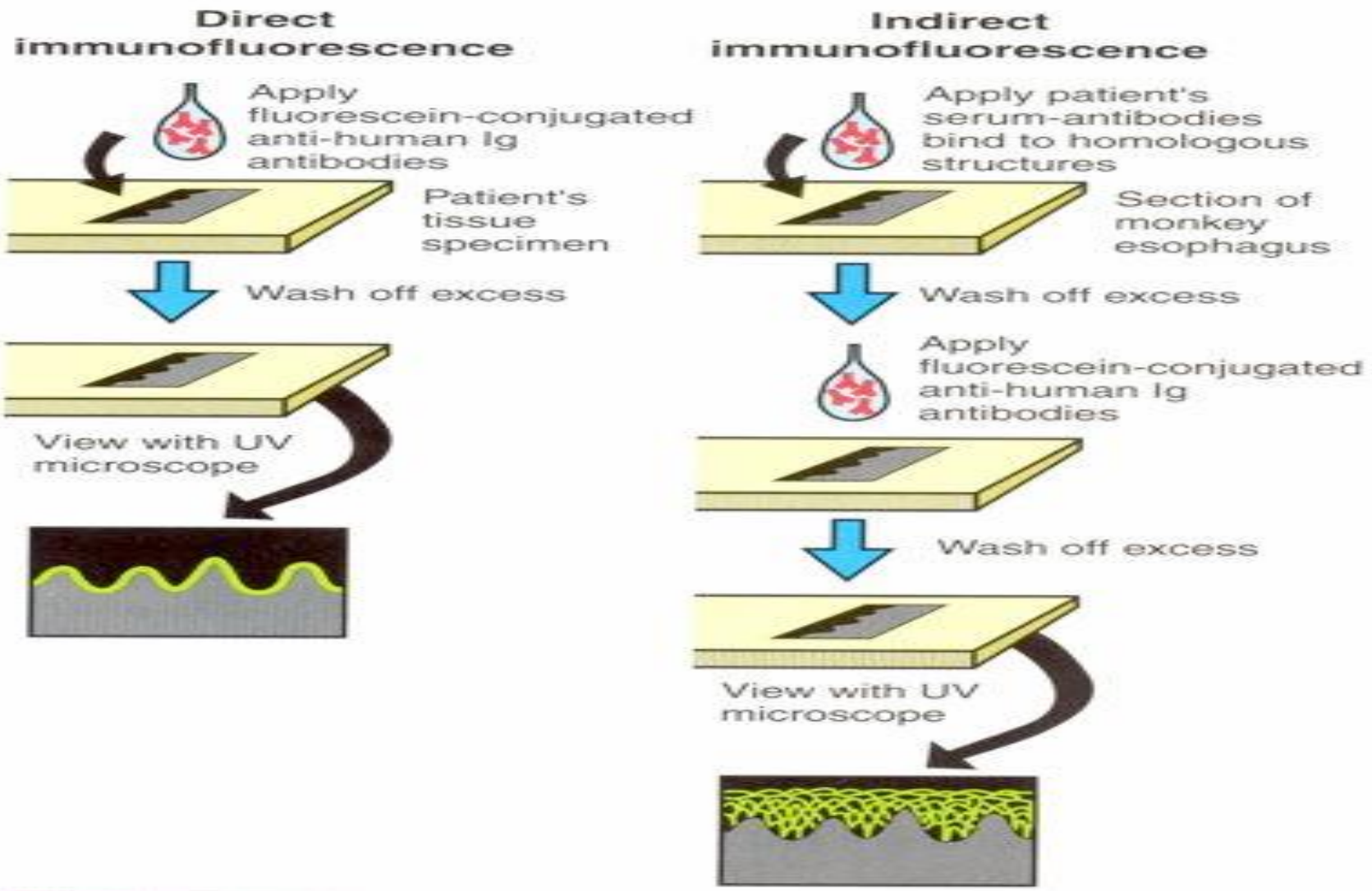


PEMPHIGUS VULGARIS

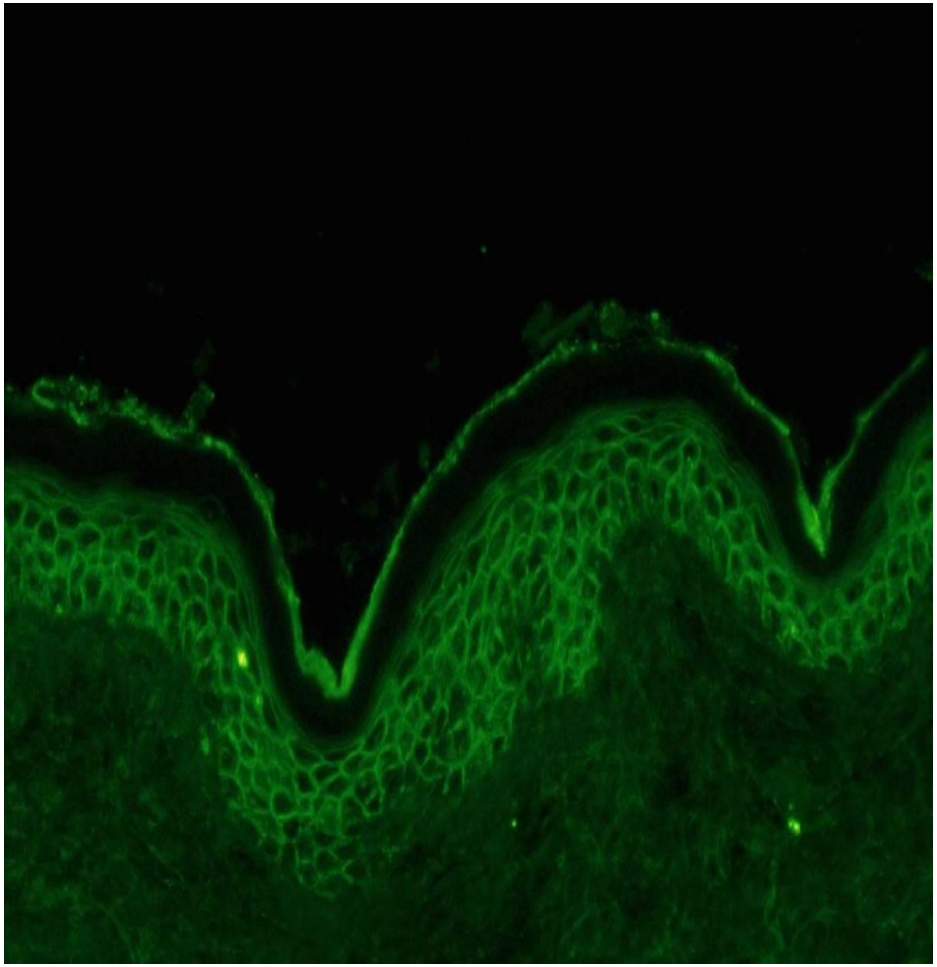
Tzanck cells

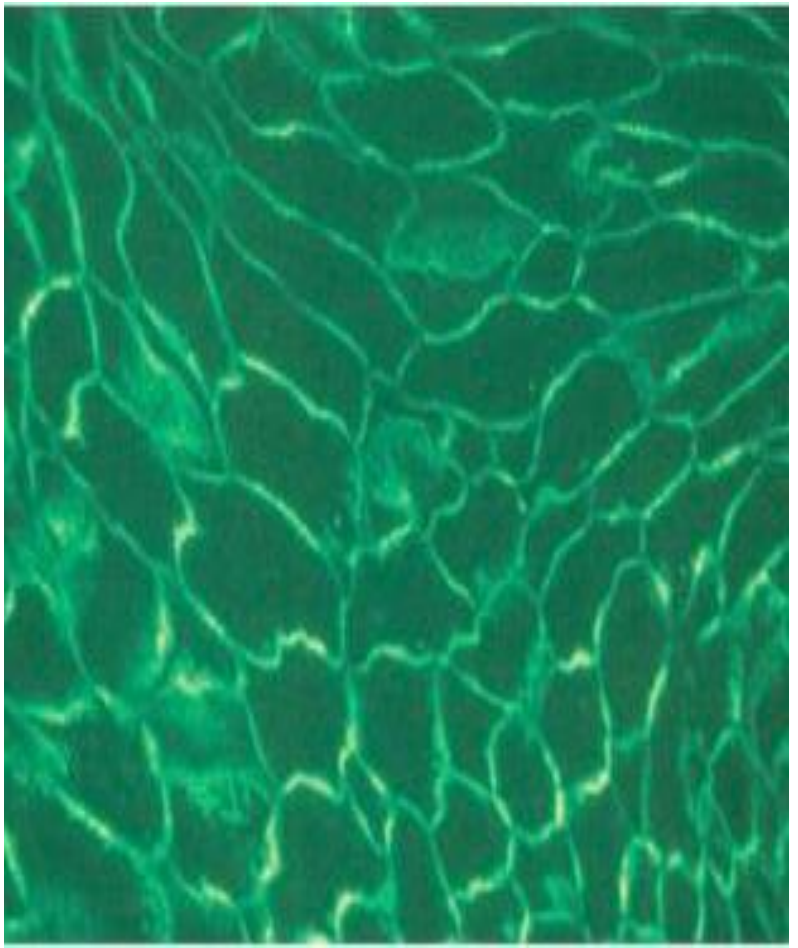


Immunofluorescence techniques



Neville. *Oral and Maxillofacial Pathology, 2nd Edition*. Elsevier, 2002.





Treatment

- Controlled with immunosuppressive(corticosteroids and azathioprine/ cyclophosphamide)
- High mortality when untreated (dehydration, electrolytes imbalance, malnutrition, infection)

PEMPHGOID

- Mucous membrane pemphigoid (cicatricial) CIKA-TRI-CIAL
- Bullous pemphigoid

PEMPHGOID

PATHOLOGY

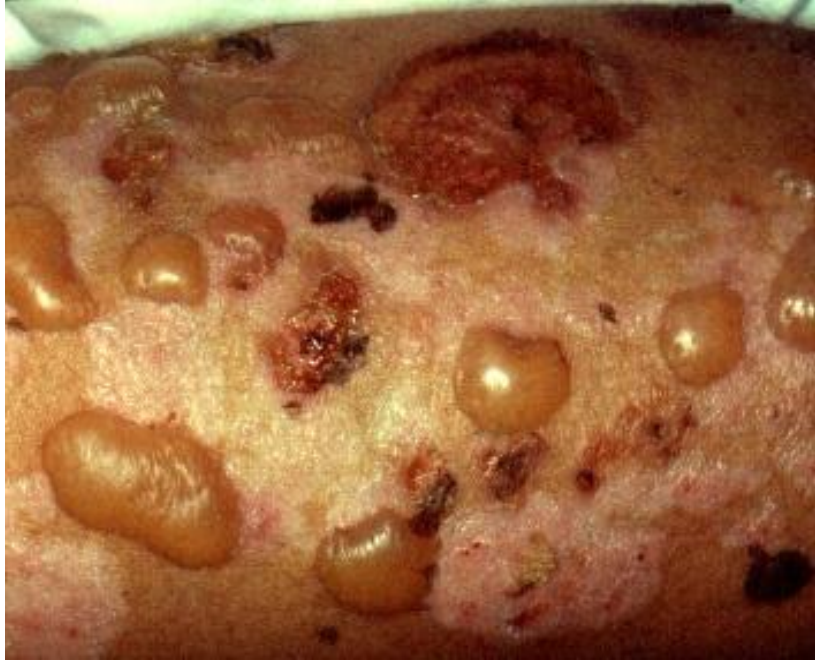
- Autoimmune disease
- Not life threatening
- Elderly females above 60 yrs of age

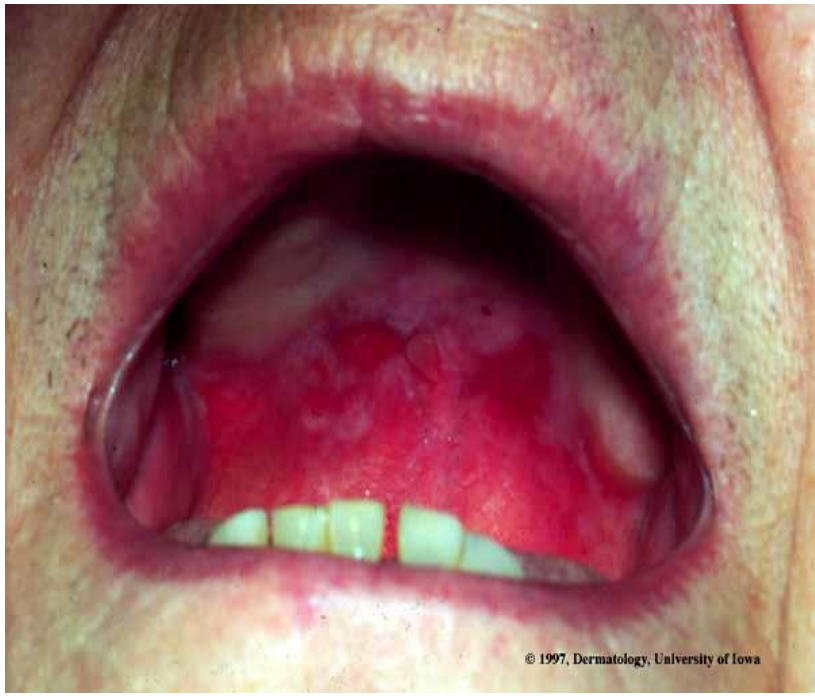
- Loss of attachment and separation of full thickness epithelium from the lamina propria.

- Alteration of rete pegs
- Epithelium forms the roof of the blisters

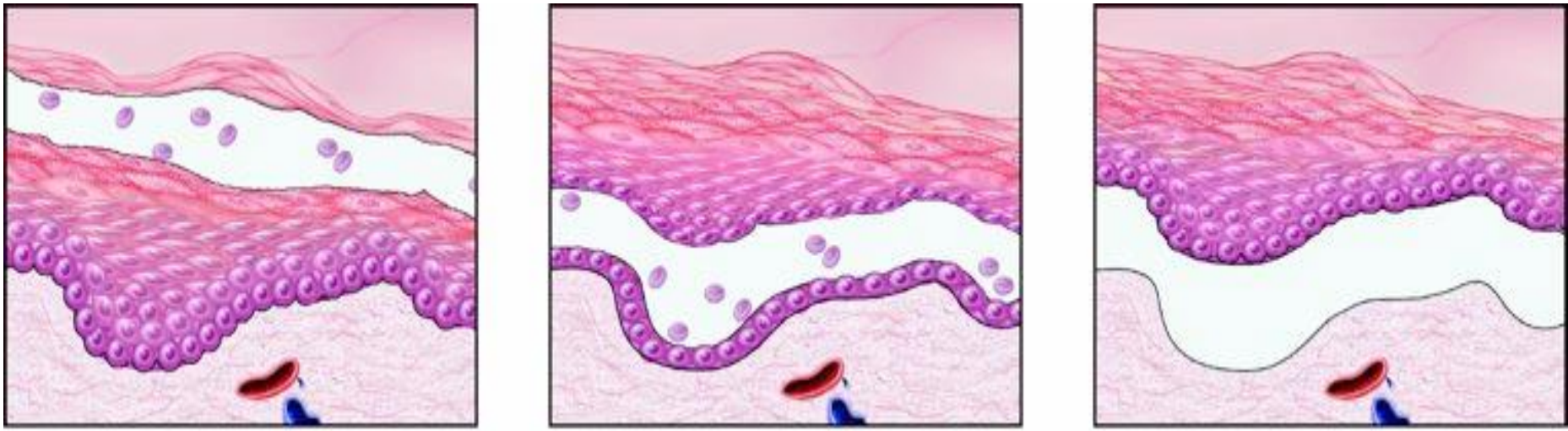
- Auto antibodies to basement membrane protein (laminin 5 & Bp180)

- Inflammatory cells (lymphocytes,neutrophils,eosinophi lare seen in the later stages





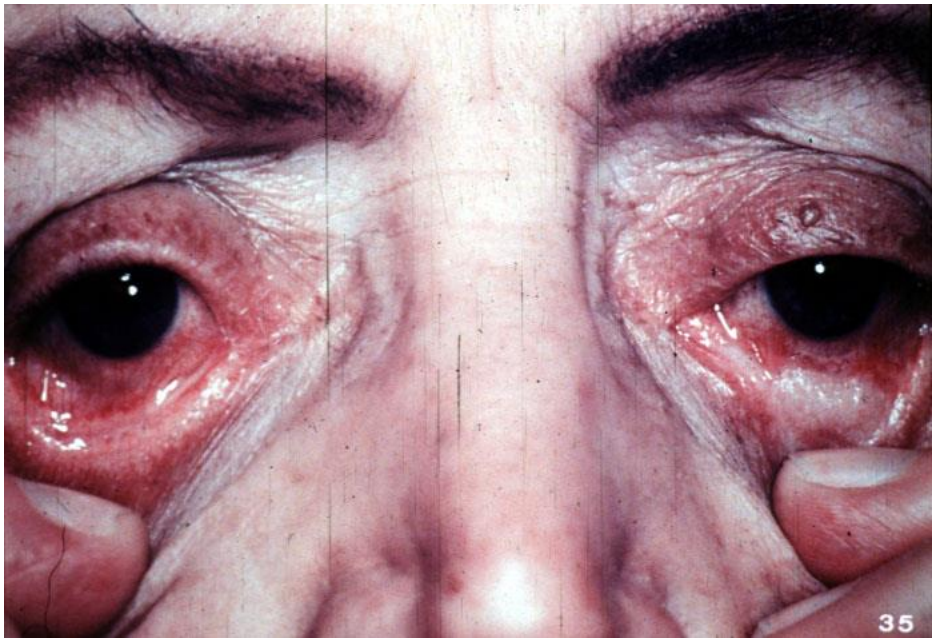
PEMPHGOID

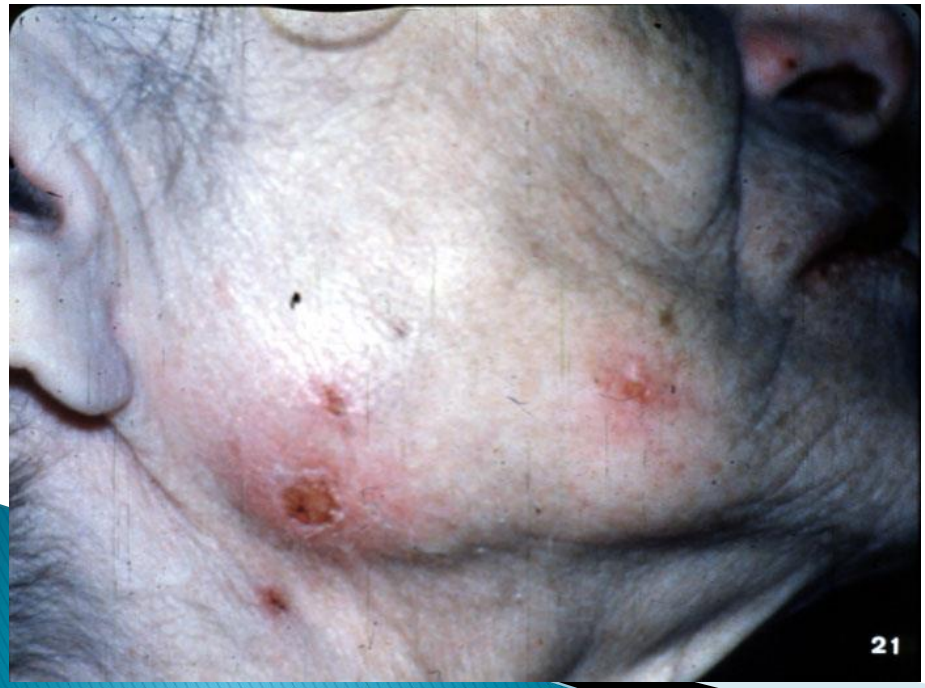


CLINICAL FEATURES (MMP)

- Oral mucosa is the first site- lesions are rarely wide spread
- Subepithelial bullae, ruptured in the later stages.
- Bleeding in the bullae – bleeding blisters
- Slow progress, skin involvement absent or rare
- Involvement of eyes, nose larynx, pharynx and osephaghus

skin rarely affected





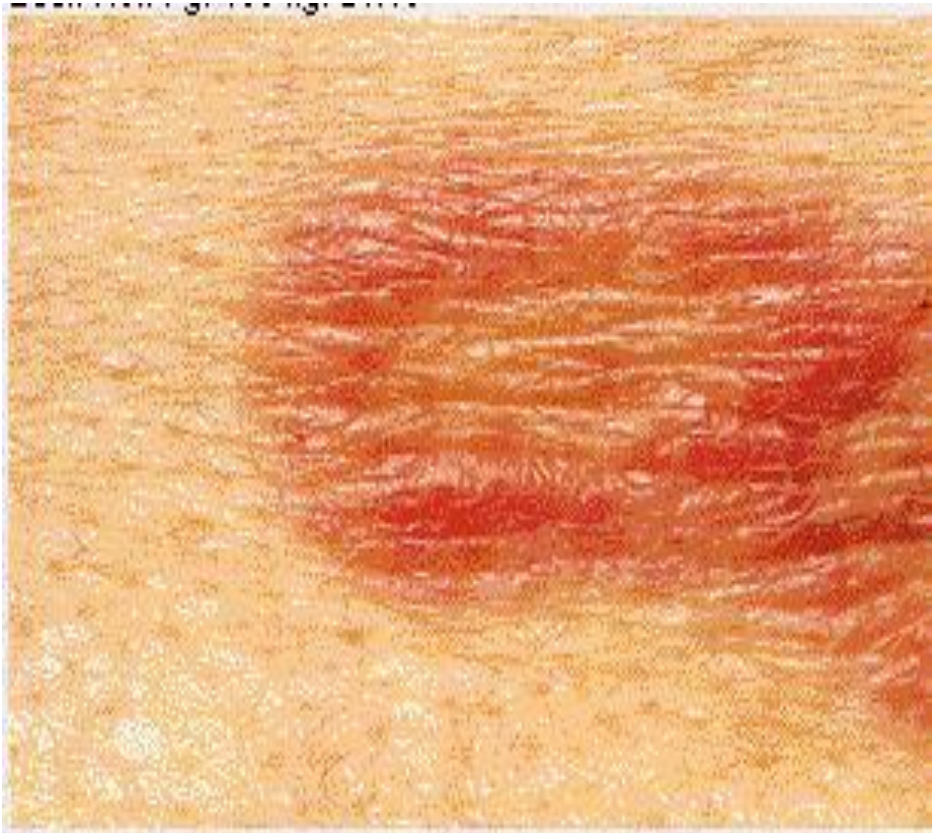
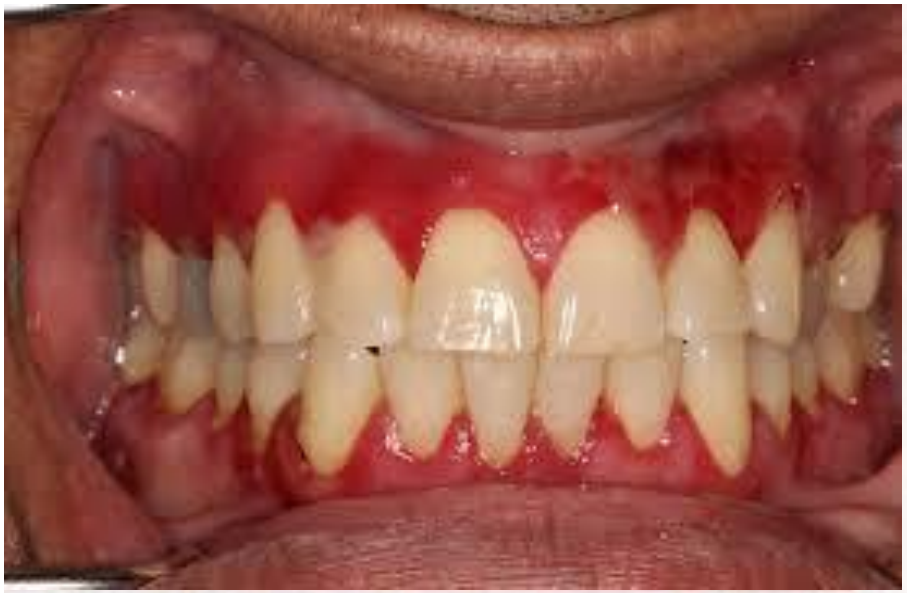


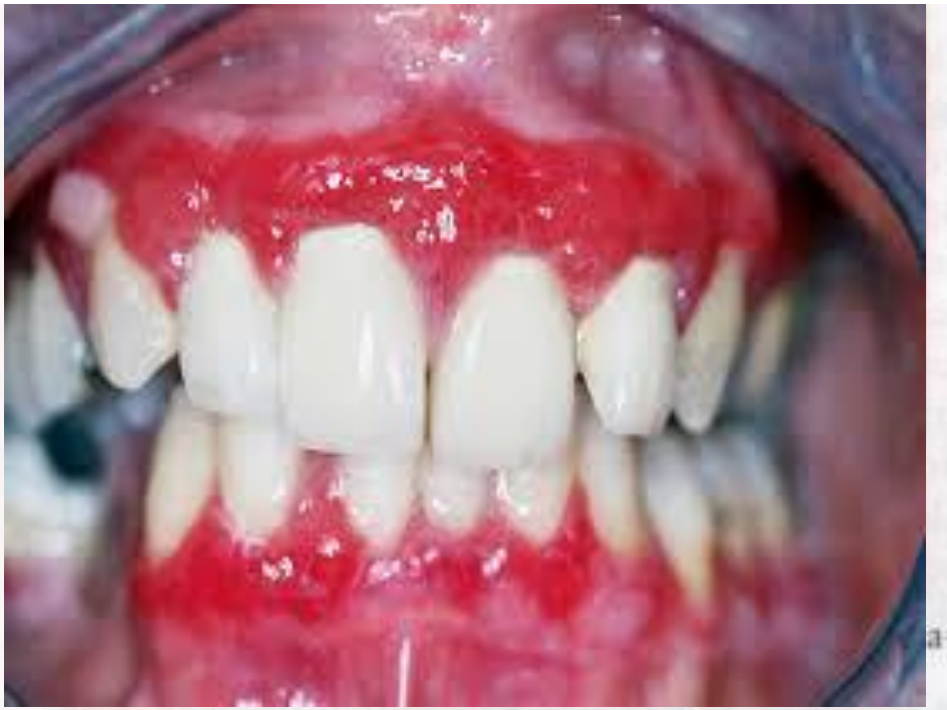
Fig. 21.15 Pemphigoid.

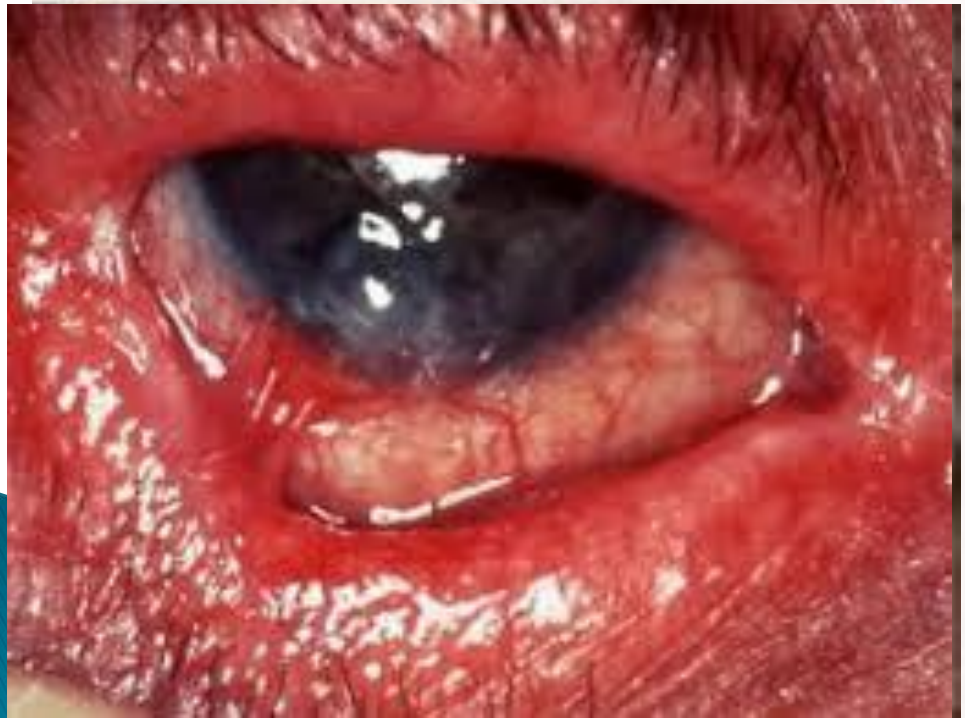
(a) Photo

(a) Clinical features showing incipient blister formation in an area of reddening. The epidermis has already begun to separate in the pale central area.



ure 56.5. Cicatricial pemphigoid: manifesting solely as des-
amative gingivitis.





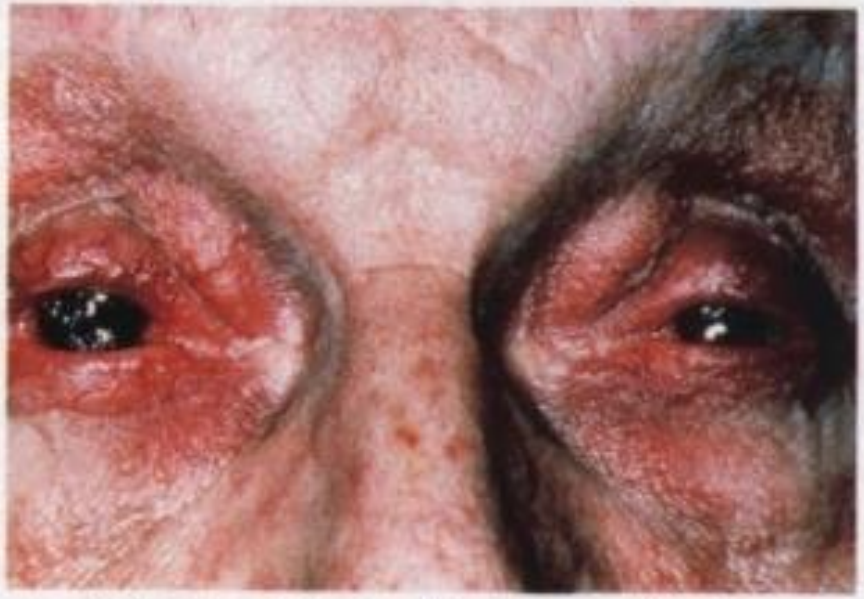
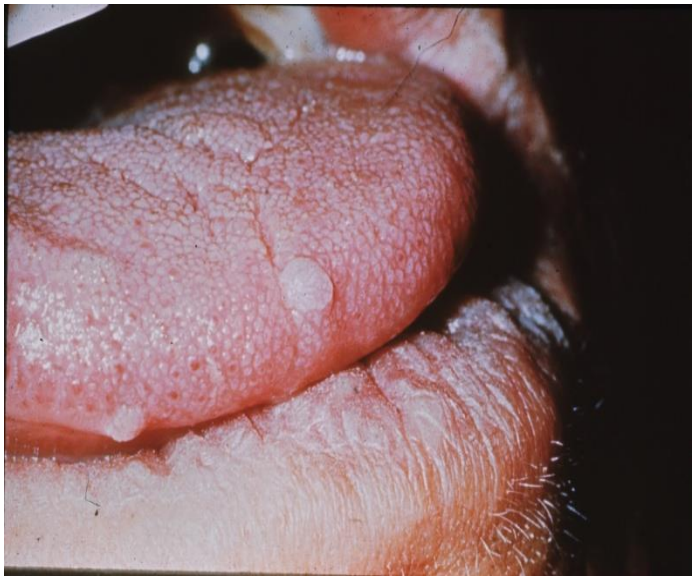
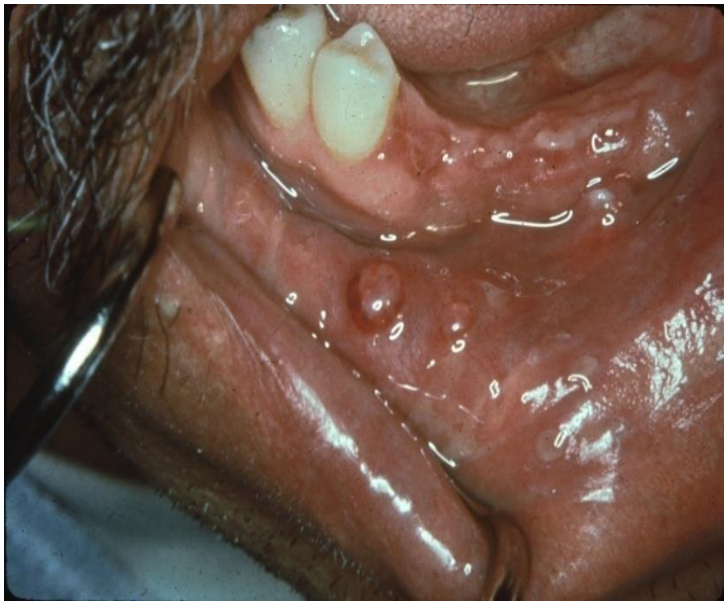
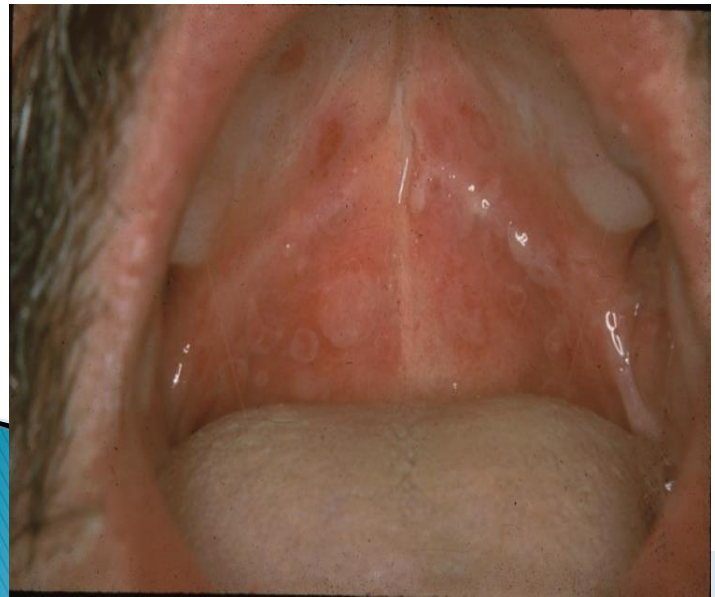


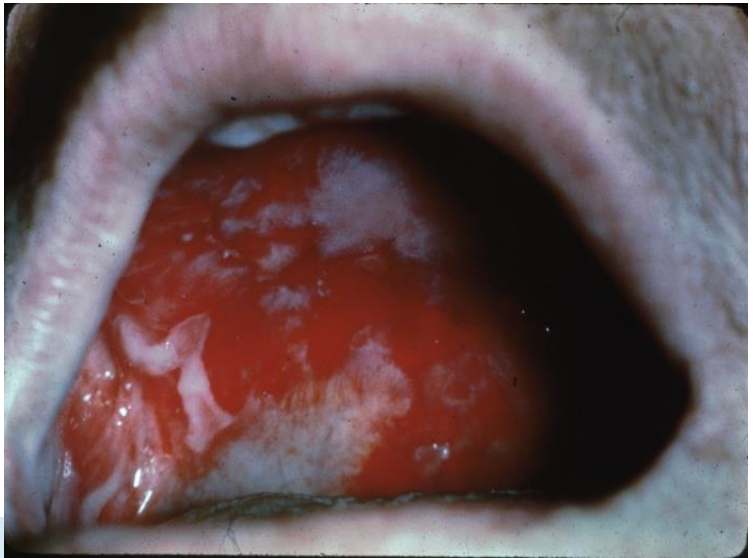
Figure 56.8. Cicatricial pemphigoid: corneal and conjuncti-
val scarring.

Oral lesions of cicatricial pemphigoid

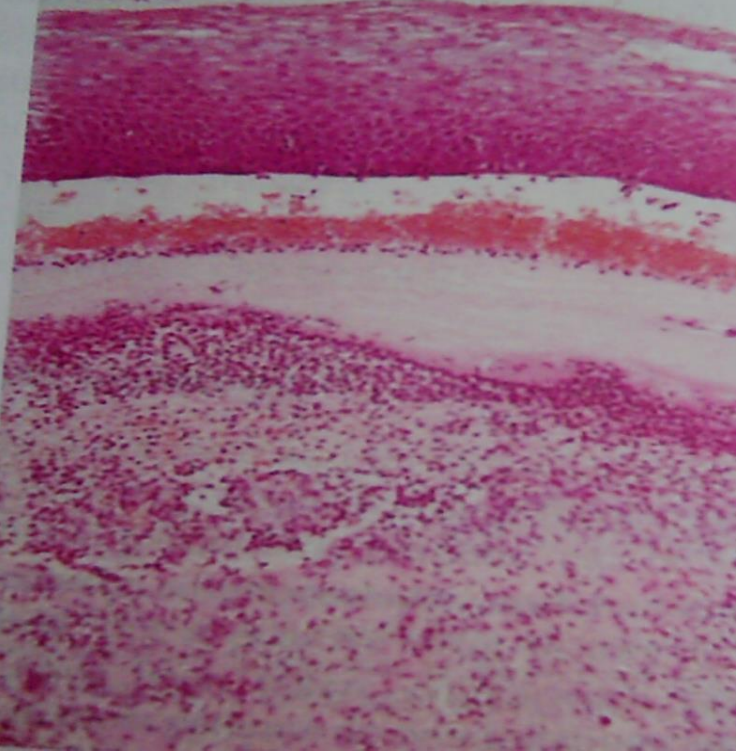








Autoantibodies cause subepithelial blisters



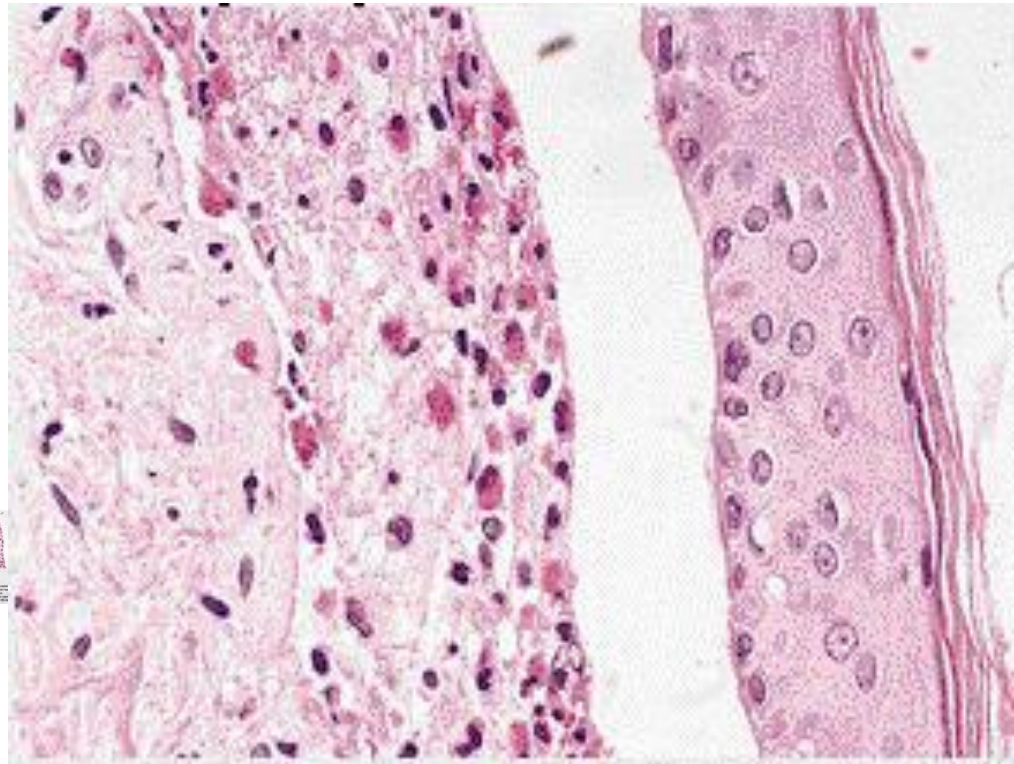
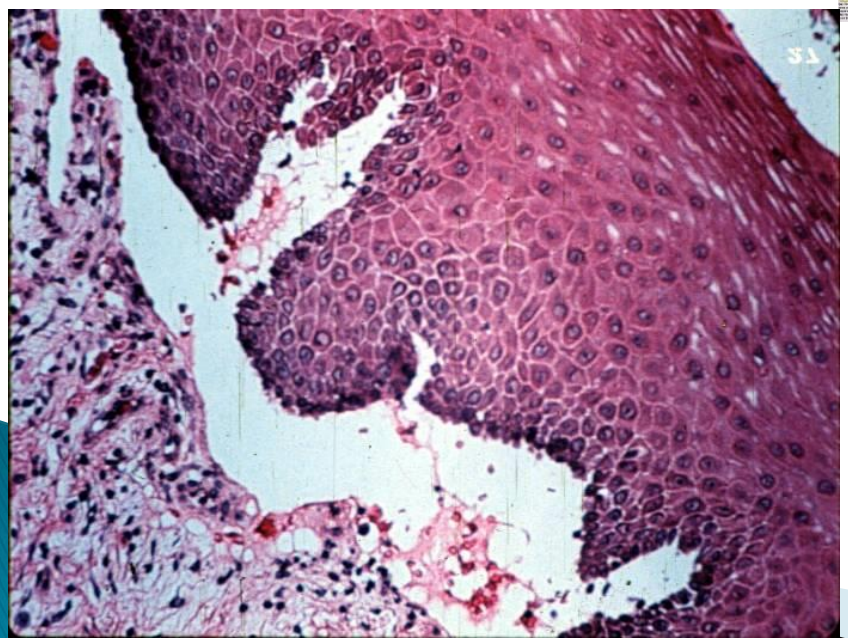


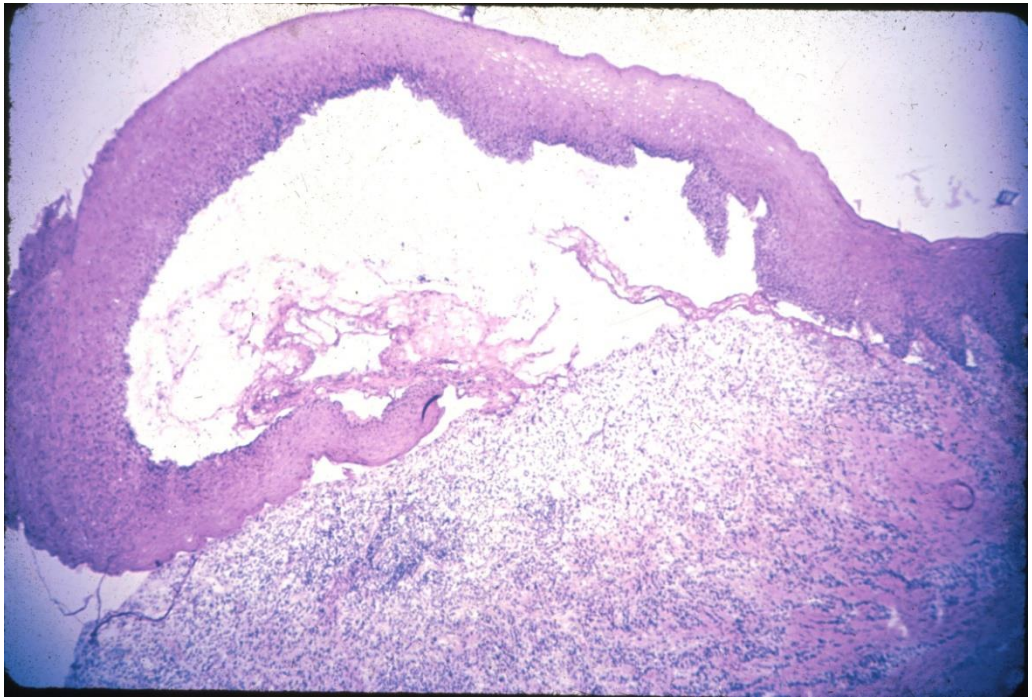
Fig. 21.15 Pemphigoid.

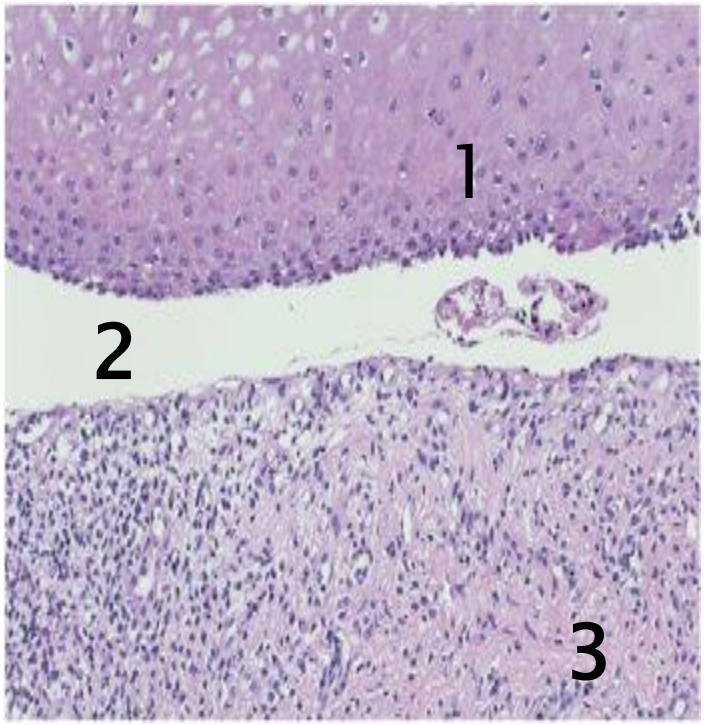
(b) Histology



(b) Histology showing complete separation of the epidermis from the dermis, and a heavy infiltrate of eosinophils.

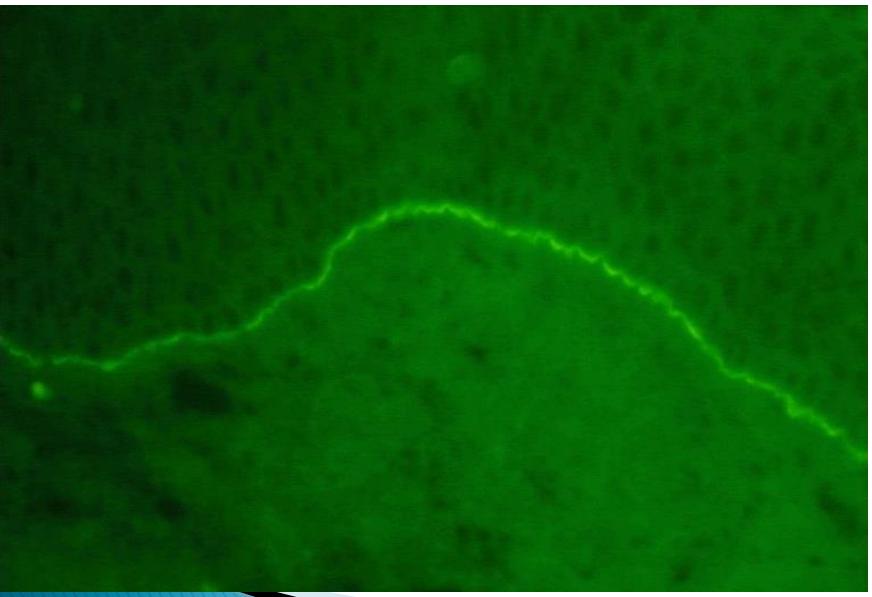
Bullous or cicatricial pemphigoid -- subepithelial blister

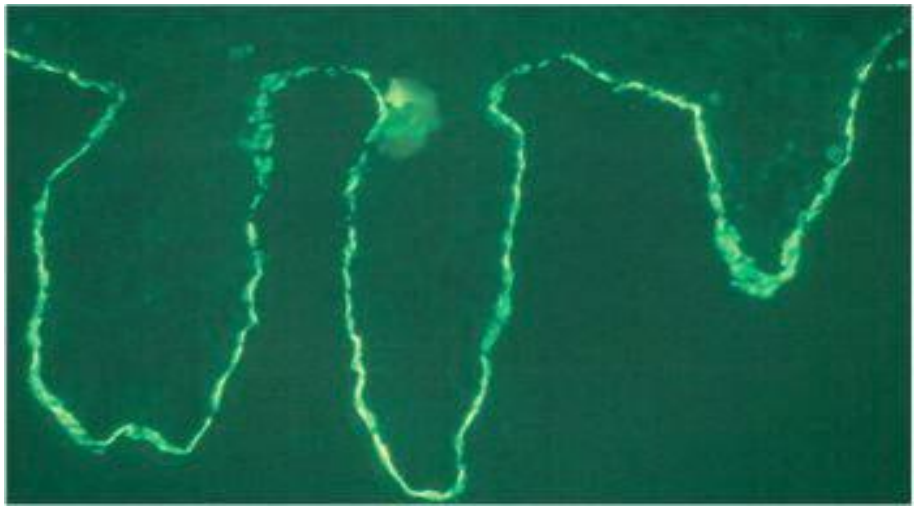




Cicatricial (mucous membrane) pemphigoid; basement membrane immunofluorescent

staining.





Courtesy Dr. Troy E. Daniels.

Regezi. *Oral Pathology: Clinical Pathologic Correlations, 4th Edition*. Elsevier, 2002.

PEMPHGOID

MANAGEMENT

- Confirm diagnosis
- Topical corticosteroids
- Ocular involvement –systemic steroids.

ERYTEMA MULTIFORME

- Mucocutaneous disease
- Males adolosents , young adults are affected more

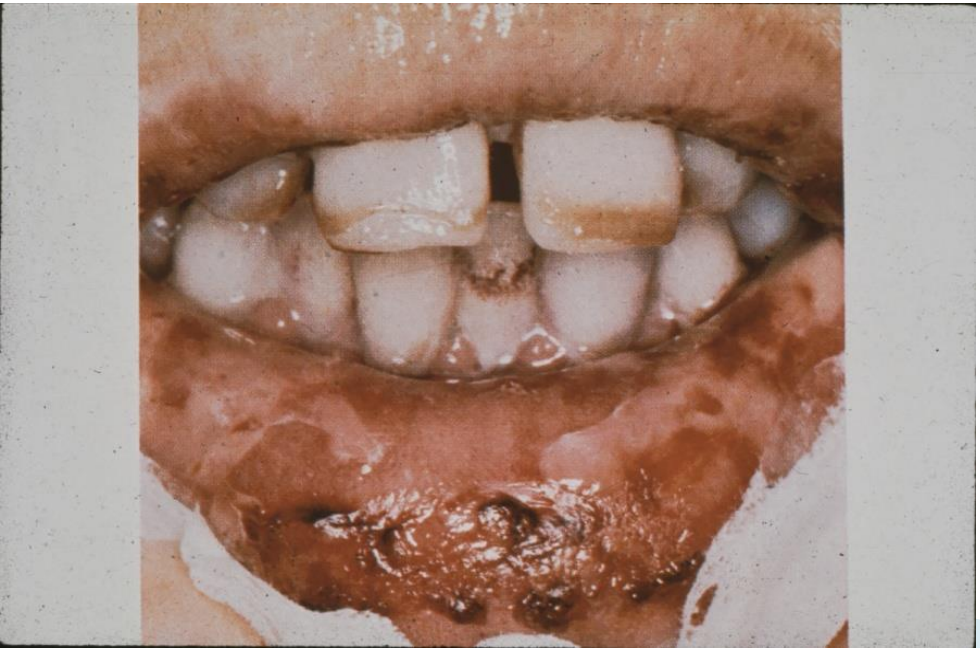
ERYTEMA MULTIFORME

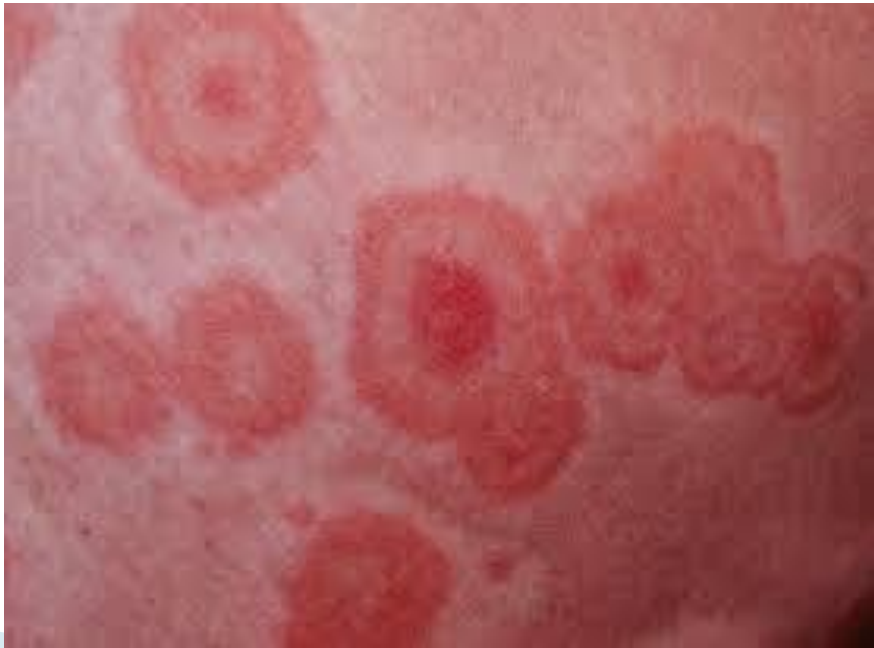
AETIOLOGY /PATHOLOGY

- Unclear aetiology and pathogenesis
- Infections like HSV can trigger this disease
- Drugs like Sulphonamides ,barbiturates
- Suggested cause is also given as to a type III hypersensitivity reaction

Signs during the disease:

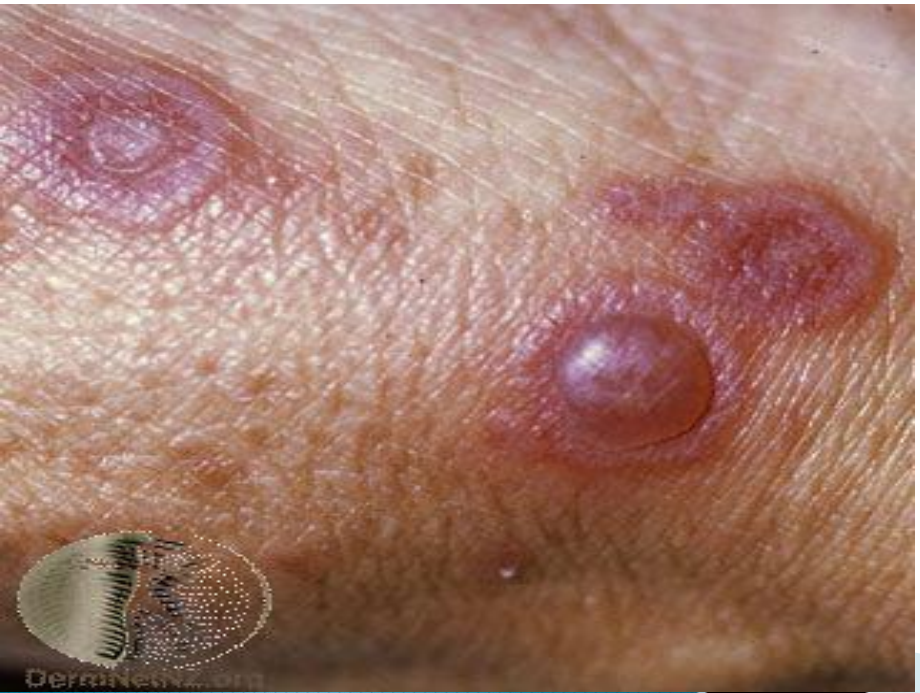
- Red macules – 1cm or more in diameter with cyanotic center
- Lips grossly swollen ,split crusted bleeding
- Widespread fibrin covered erosions and erythema in the mouth.
- Mild fever
- Conjunctivitis may be associated
- Attacks recur at the intervals of several months
- Usually self limiting.

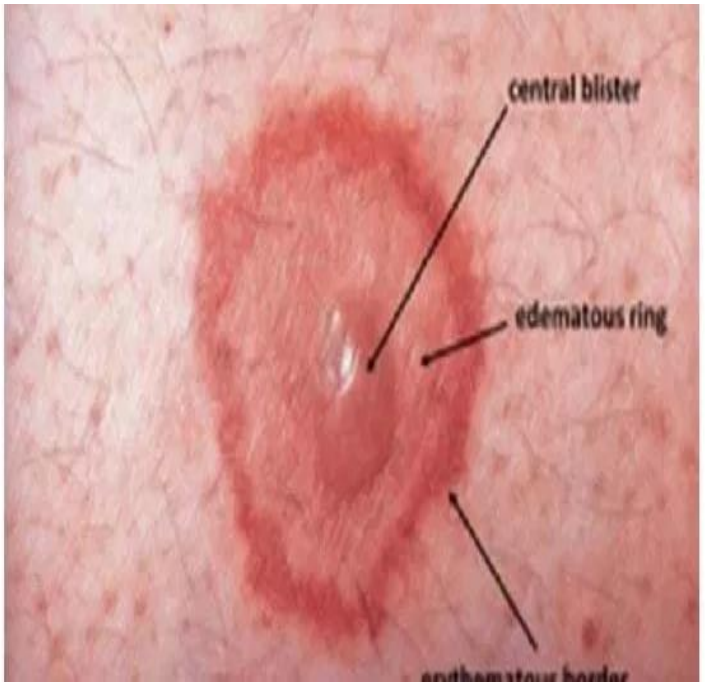




Typical target skin lesion

Central area of vesiculation, surrounded by a concentric erythematous rings resembling target or bull's eye (iris lesion)

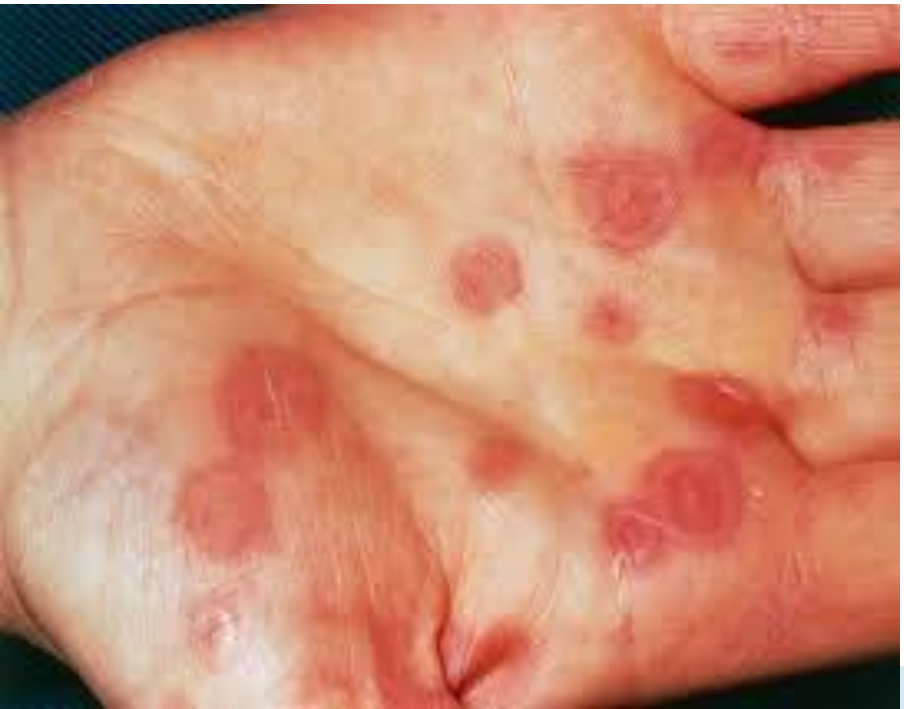


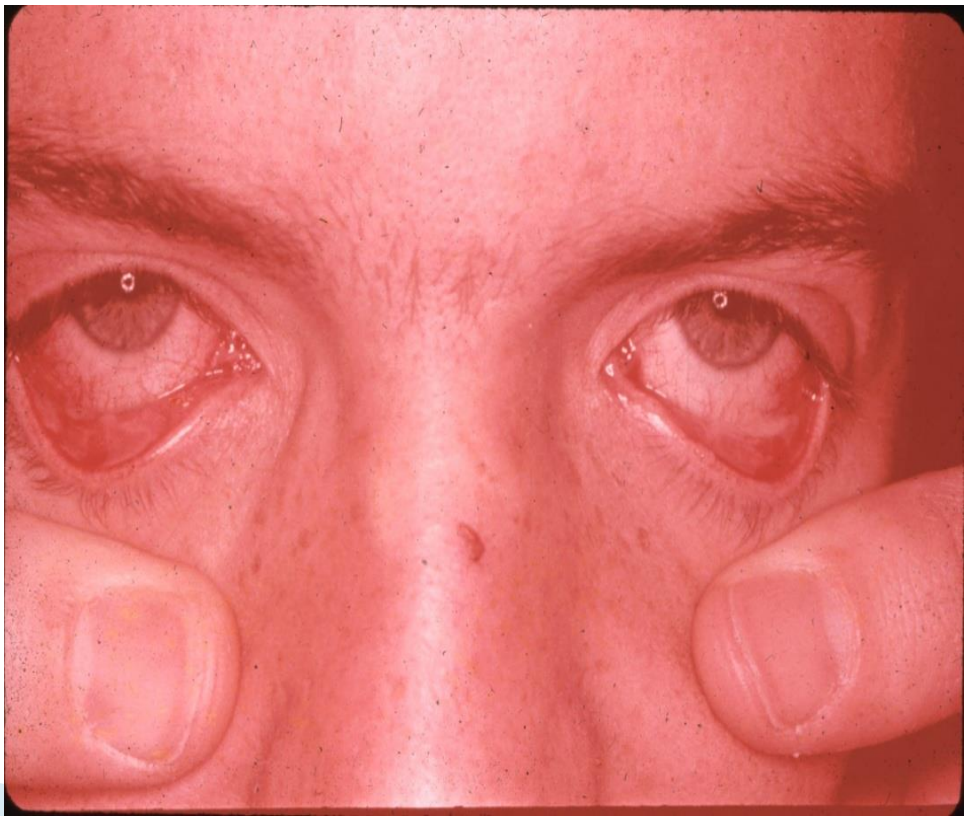


Erythema multiforme with conjunctivitis – in

Stevens Johnson syndrome:

A severe form of EM ,triggered by drugs , involvement of occular-genital-oral& skin lesions





EPIDERMOLYSIS BULLOSA

Definition:

- A large group of clinically similar desquamating disease processes of the skin and mucosa that have in common the separation of the epithelium from the underlying connective tissue and the formation of large blisters at the site of minor truma, that frequently result in extensive and often immobilizing scar formation.

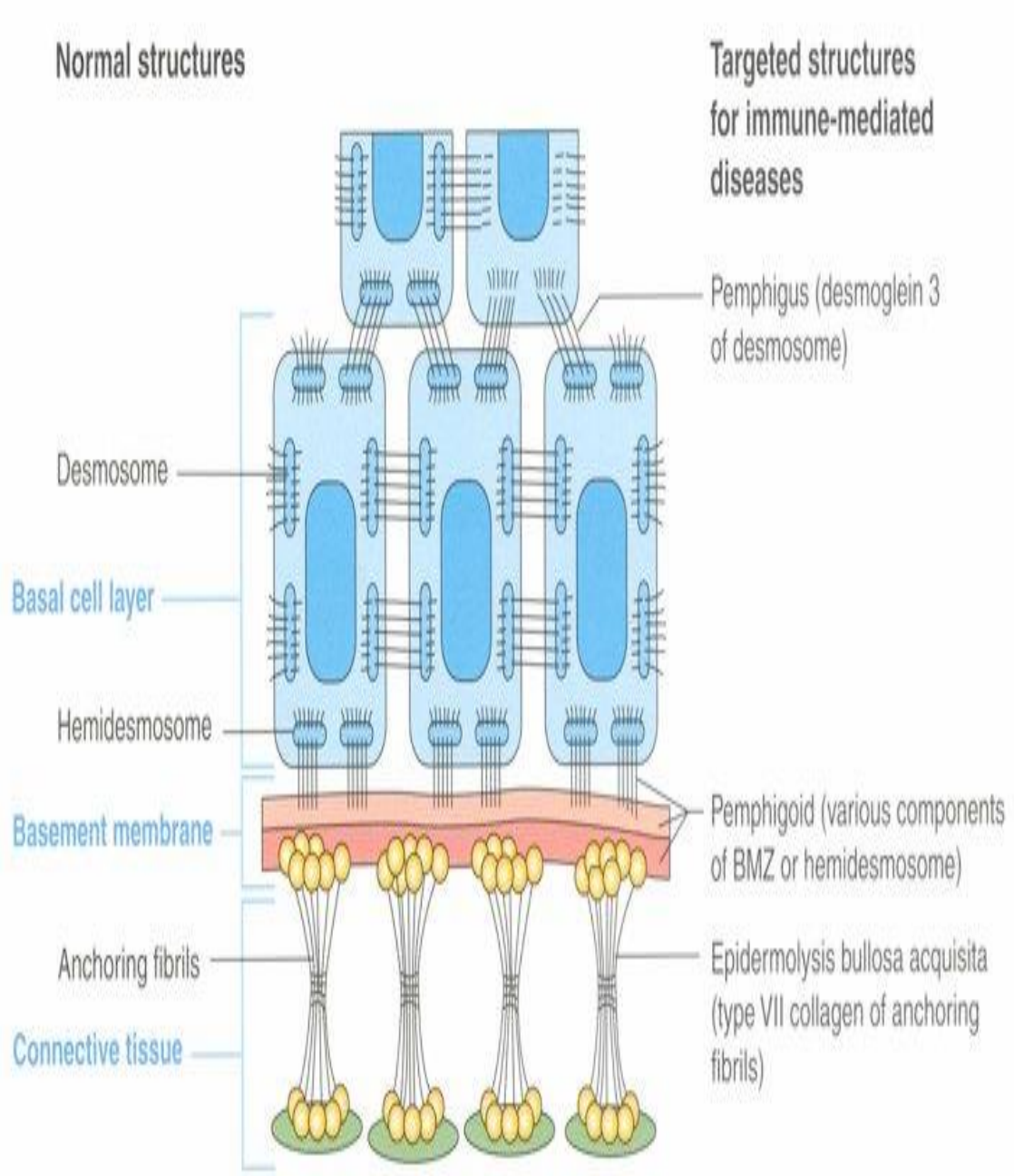
EPIDERMOLYSIS BULLOSA

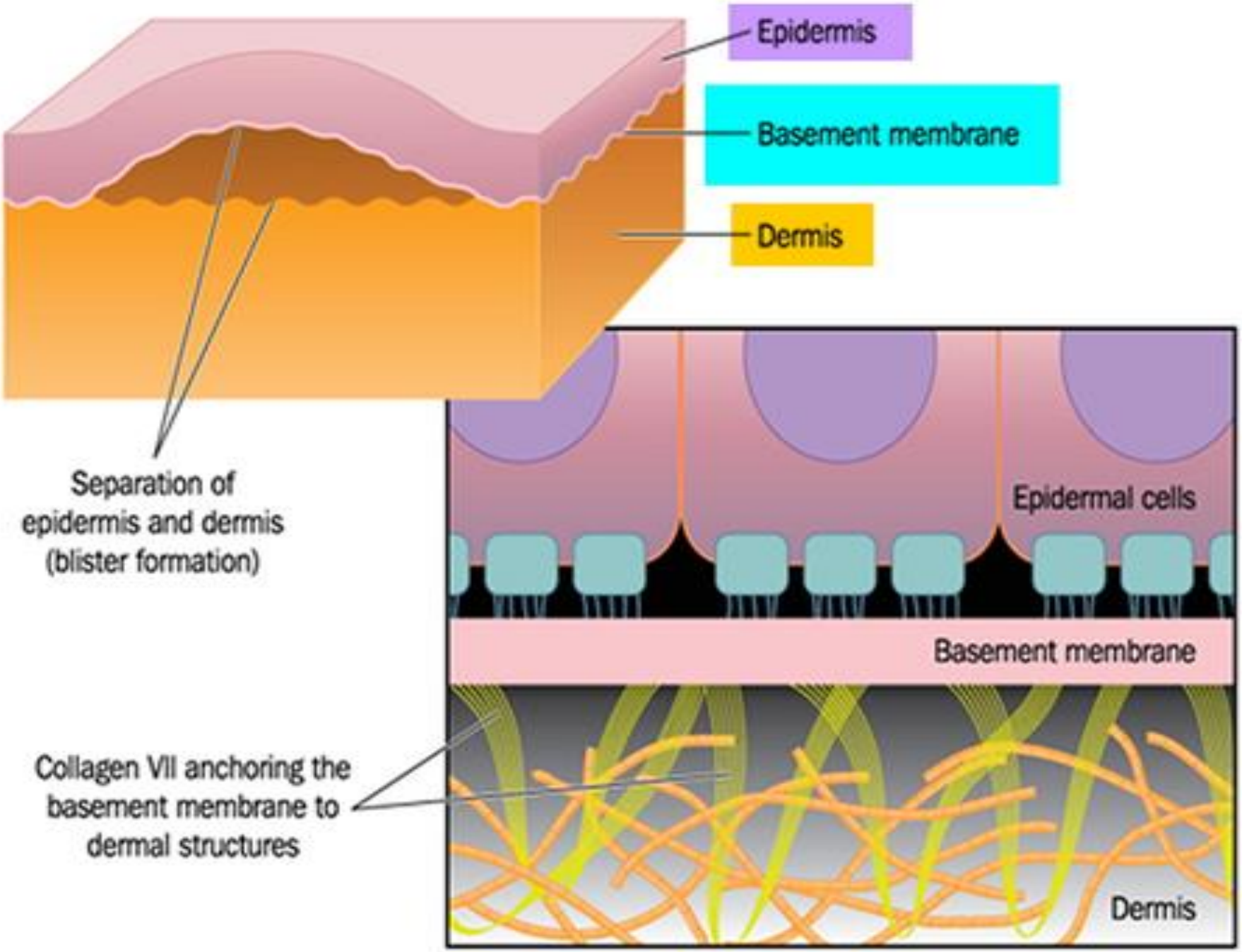
HEREDITARY TYPES:
(simplex, junctional, dystrophic)

- Congenital absence of components:- either in the basal cell layer, hemidesmosome, or anchoring C.T filaments depending on the type of disease.

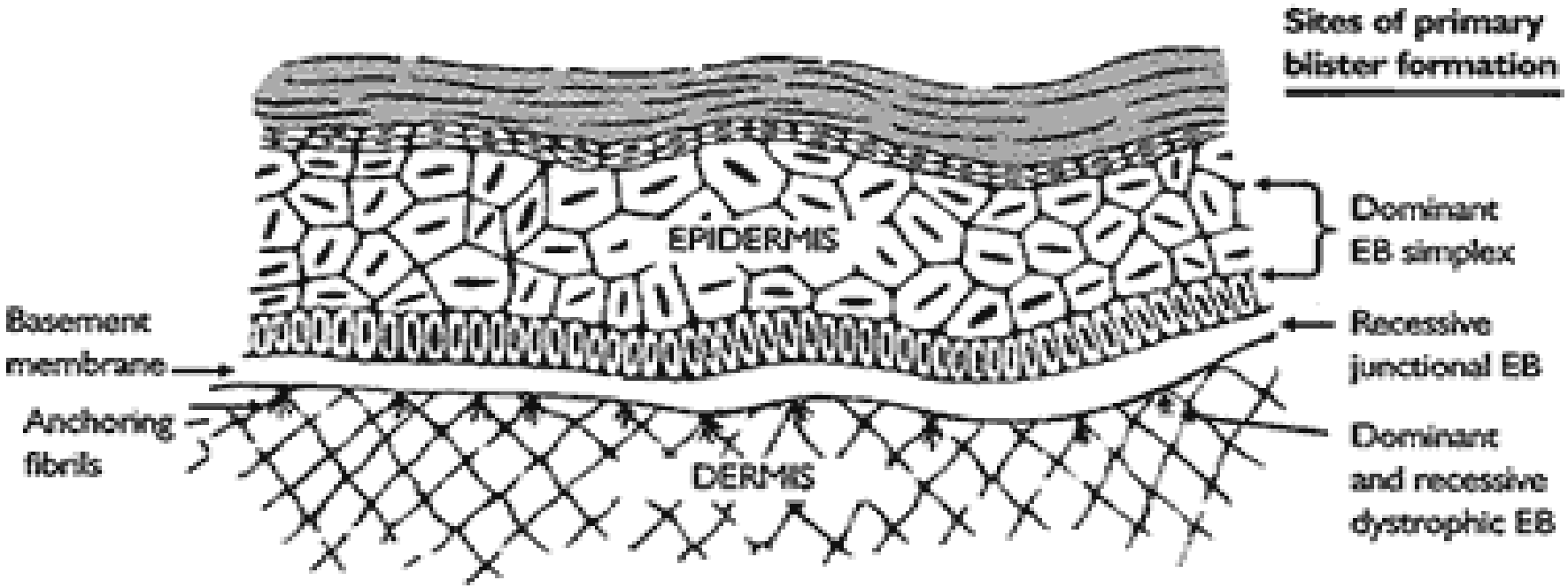
ACQUIRED TYPES:

- Autoantibodies (IgG; sometimes IgA) to type VII collagen of the anchoring fibrils.



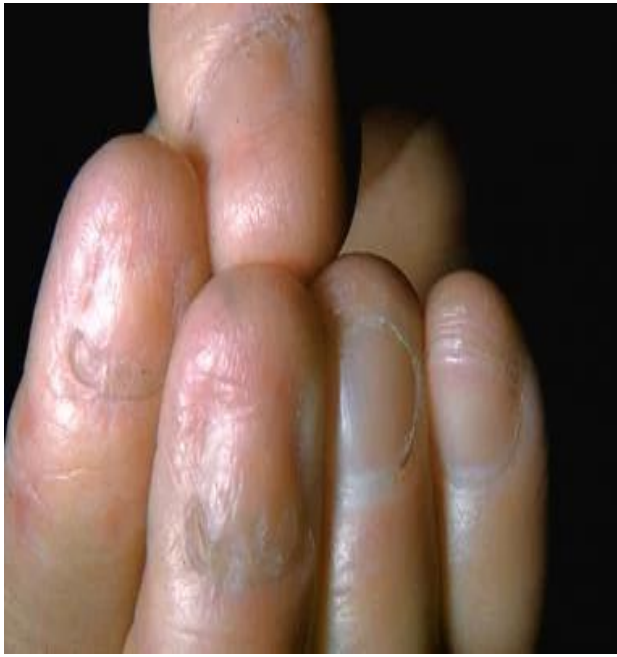

Epidermis
Basement membrane
Dermis
Separation of epidermis and dermis (blister formation)
Epidermal cells
Basement membrane
Collagen VII anchoring the basement membrane to dermal structures
Dermis

EPIDERMOLYSIS BULLOSA

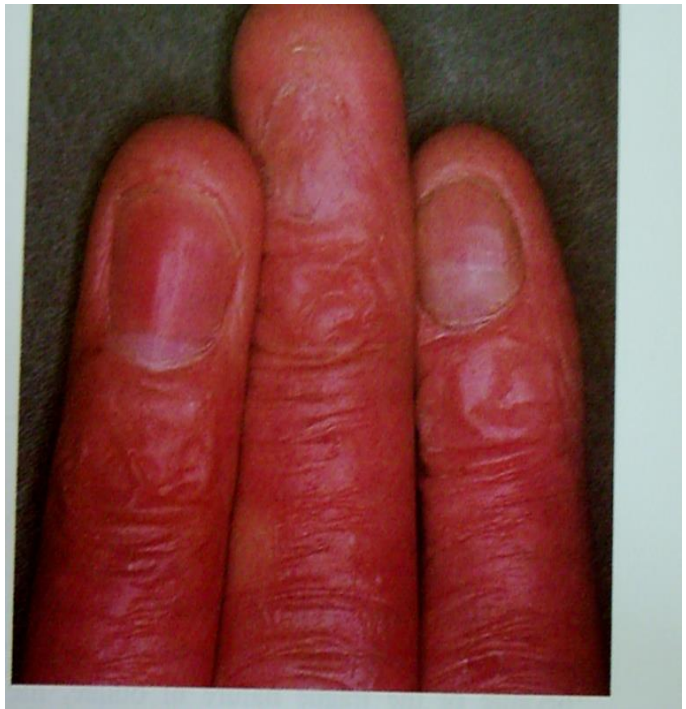


Source: Adapted from Dystrophic Epidermolysis Bullosa Research Association of America (DebRA).

clinical



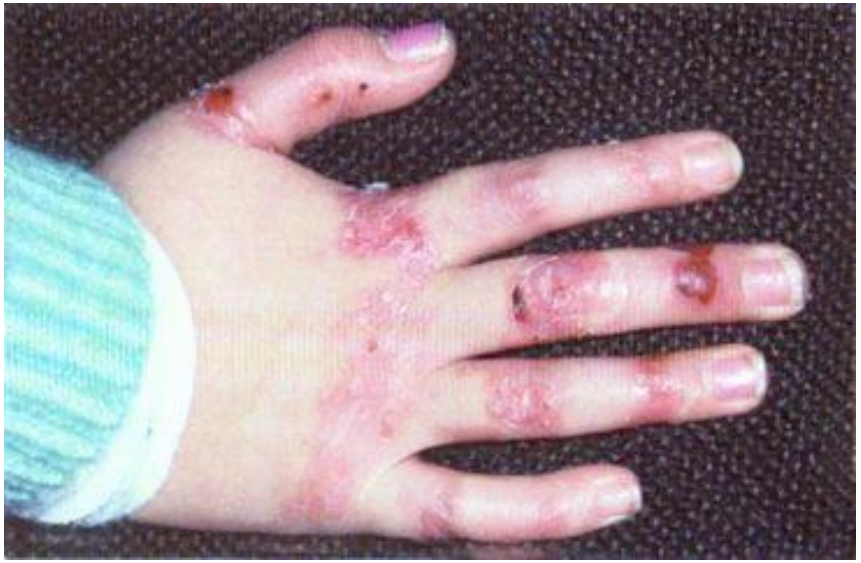
- Onset at infancy & early childhood in the hereditary type
- During adulthood in the aquired type
- Severity depend on type (scarring, and atrophy)
- Teeth are hypoplastic and constricted oral orifice
  resulting from scar contracture
- Nails may be dystrophic in some forms of this disease

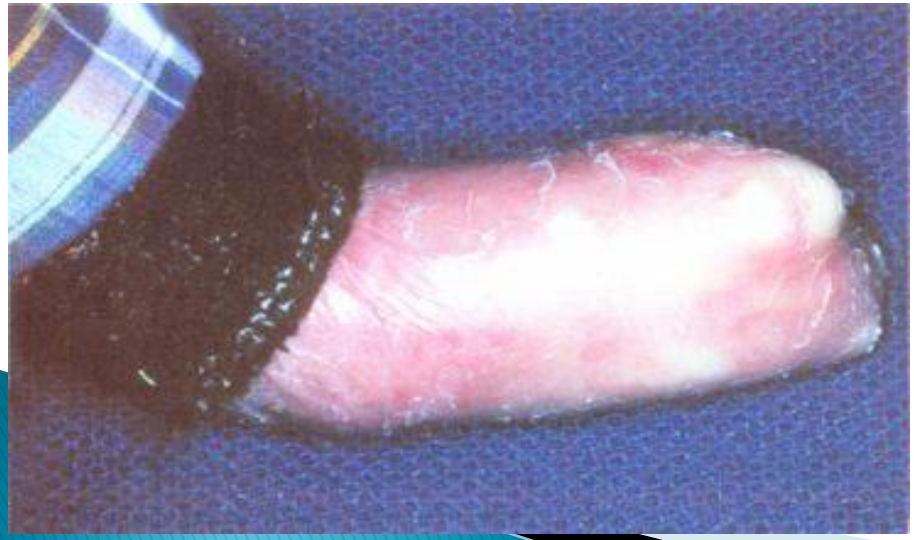


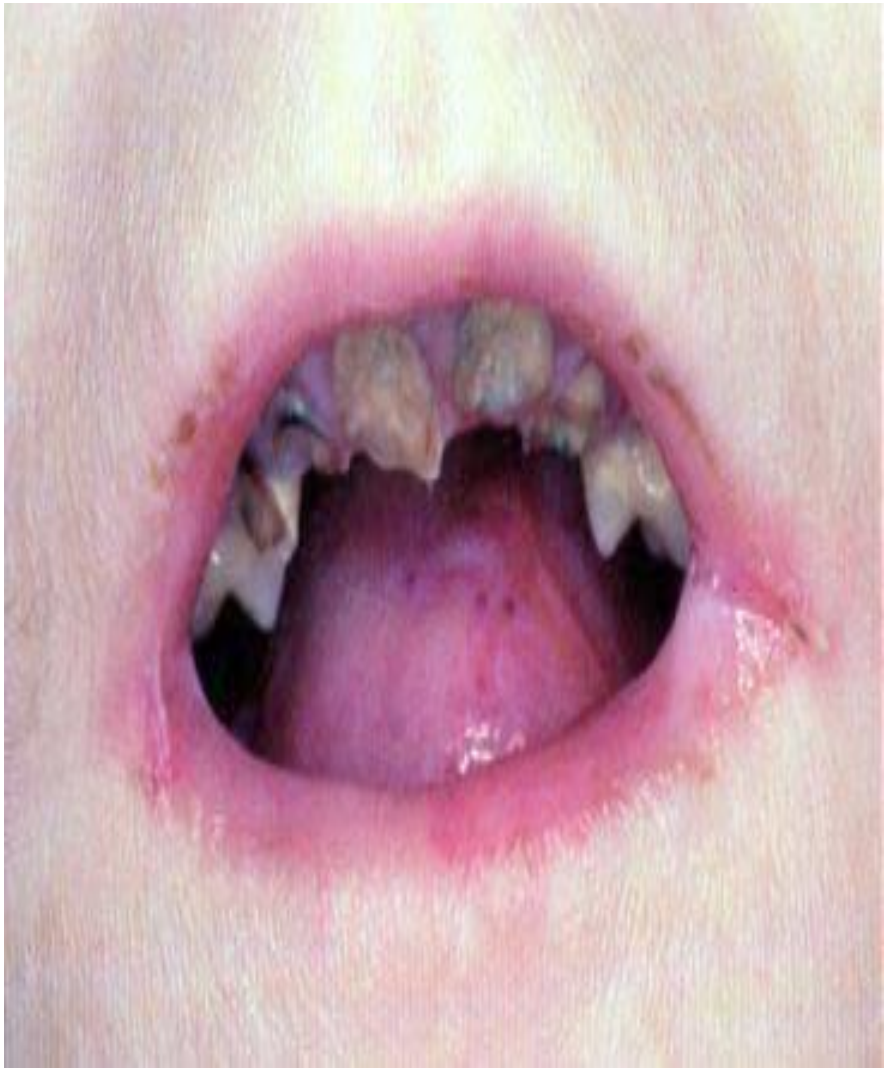
Treatment:

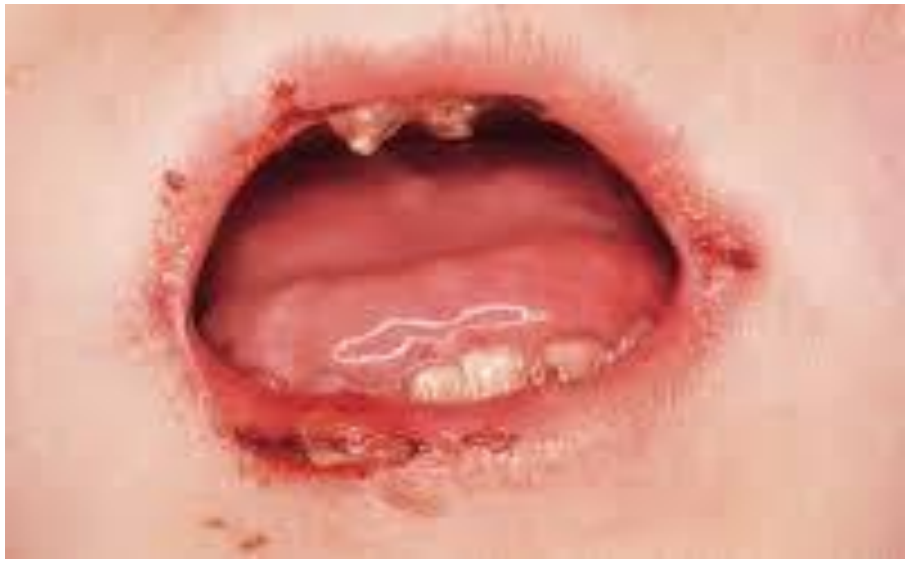
avoidance of trauma, supportive measures & chemotherapeutic agents

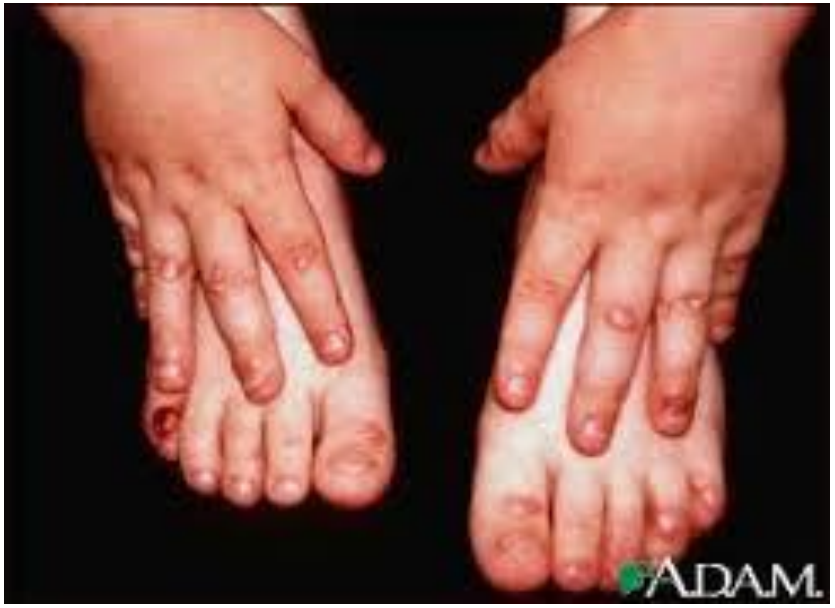
Epidermolysis bullosa

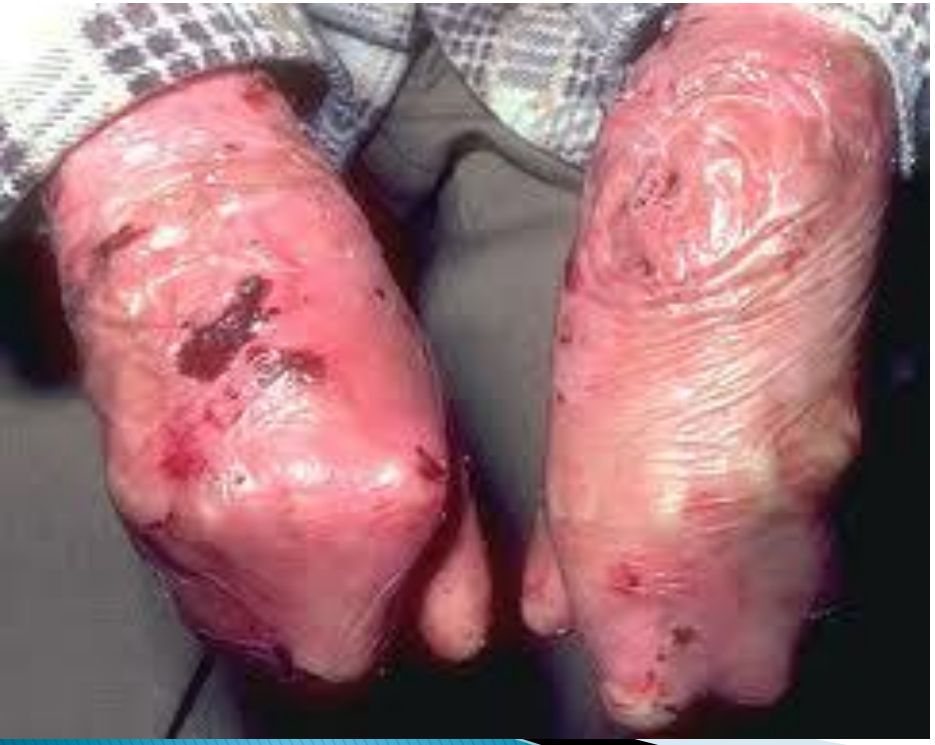


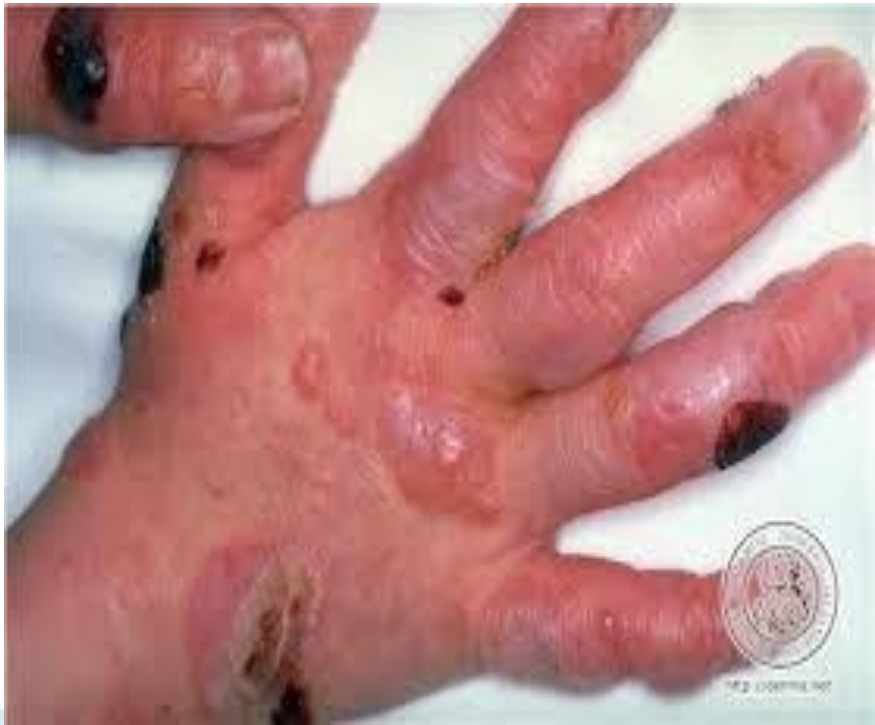




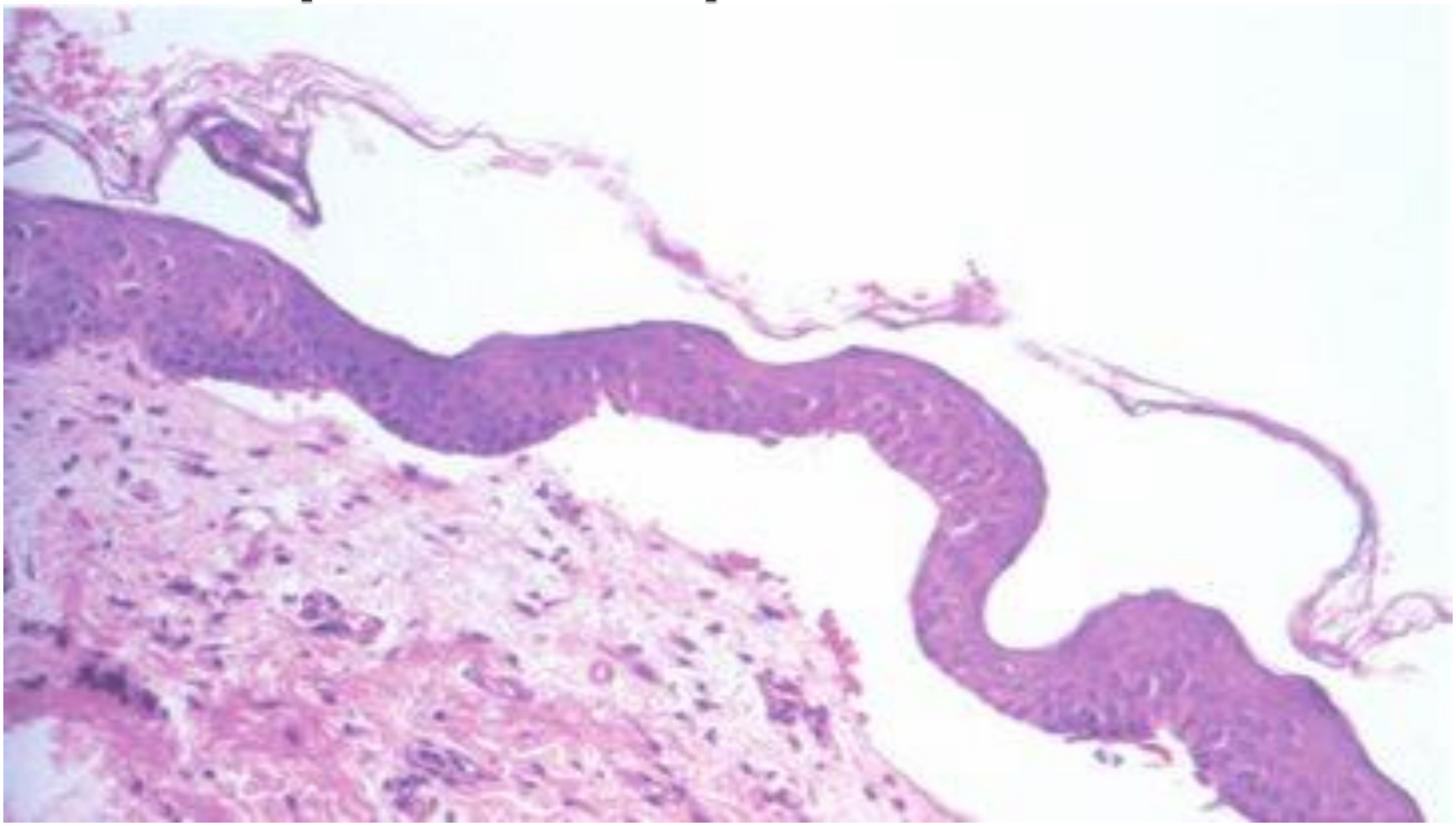



A.D.A.M.





Epidermolysis bullosa



Complete separation of the epithelium from the connective tissue is seen in this photomicrograph of a tissue section obtained from a patient affected by a junctional form of epidermolysis bullosa.

Neville. *Oral and Maxillofacial Pathology, 2nd Edition*. Elsevier, 2002.