VESICULOBULLOUS DISORDERS

VESICLE : fluid filled blister < 0.5 cm in size.

BULLA : fluid filled blister > 0.5 cm

CLASSIFICATION

CONGENITAL INHERITED DISORDERS

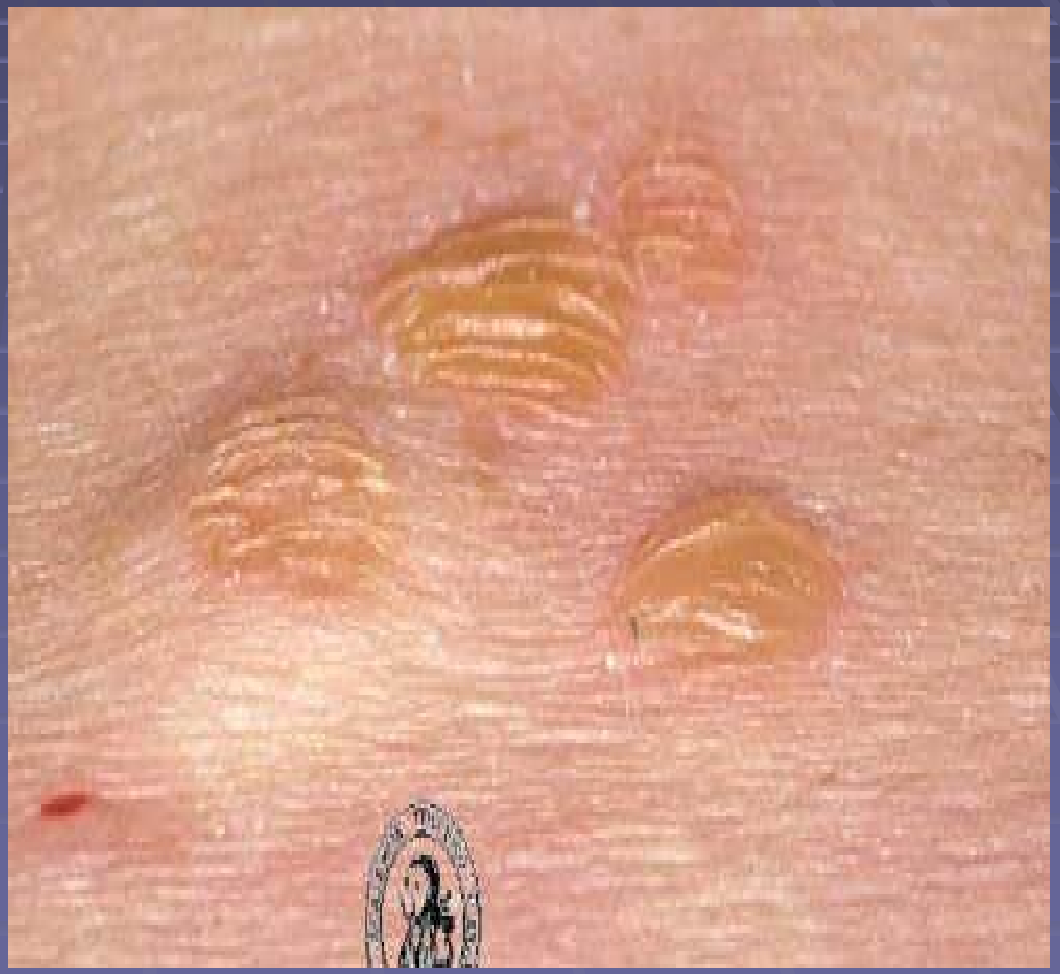
Epidermolysis bullosa
Hailey-2 disease
Relapsing linear acantholytic dermatosis

IMMUNOLOGICAL DISORDERS

Intraepidermal -Pemphigus
Subepidermal - Bullous pemphigoid
Linear IgA disease
EBA (epidermolysis bullosa acquista)
Bullous SLE
Dermatitis herpetiformis

PEMPHIGUS

Pemphix – blister/bubble



Variants of pemphigus

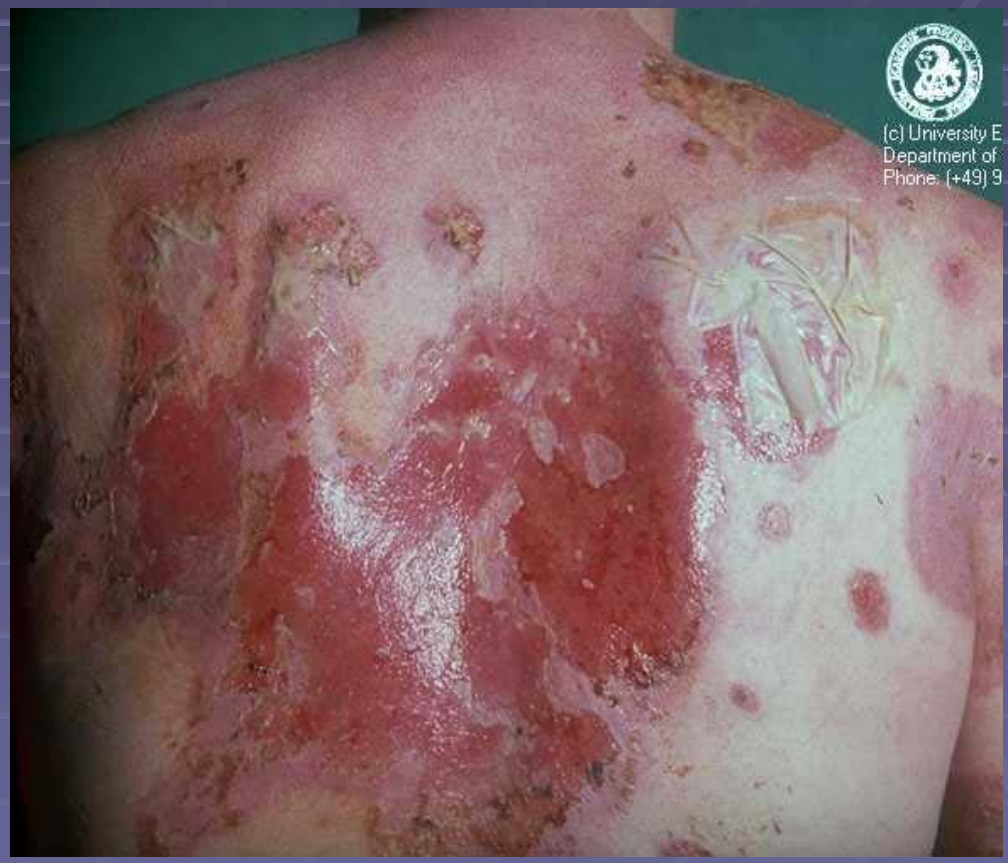
- P.vulgaris
- P.vegetans
- P.foliaceus
- P.erythematosus

Pemphigus vulgaris

Pathogenesis

Autoantibodies- Desmoglein

(present over desmosomes)



Pathology

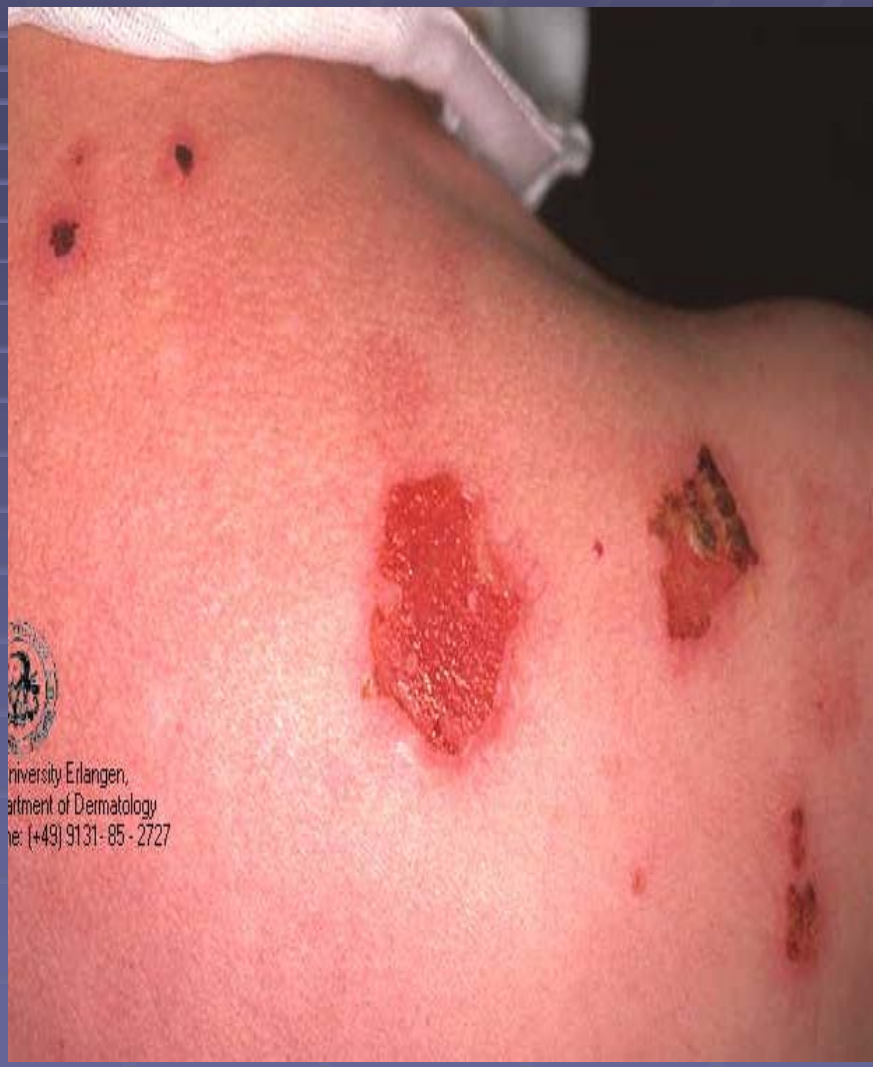
- Suprabasal split

- Row of tombstones:

Basal cell attached to B.M

-but separate from one another

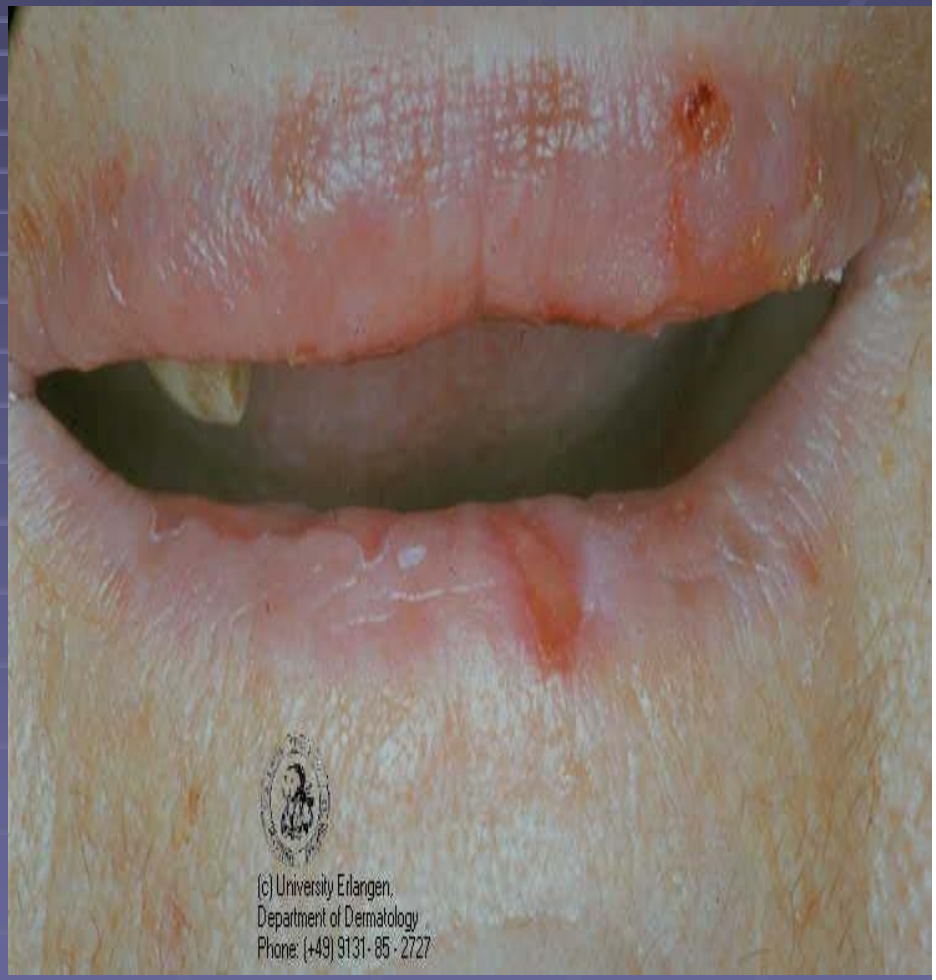
-Acantholytic cells in blister cavity



Clinical features

-Oral lesions in all patients

-Other mucosal sites may be involved

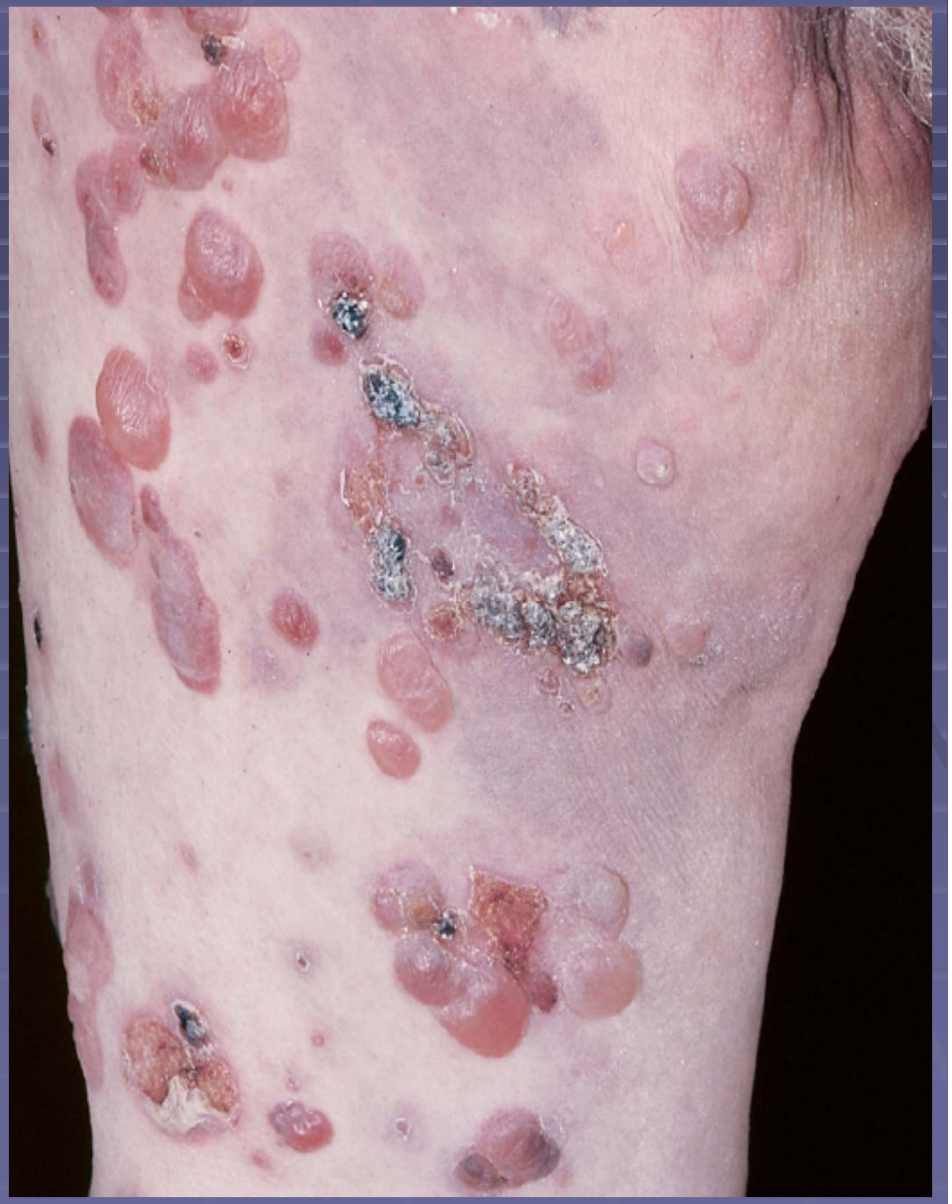


Predilection for

Scalp, face, axillae, groins, pressure points

Lesions

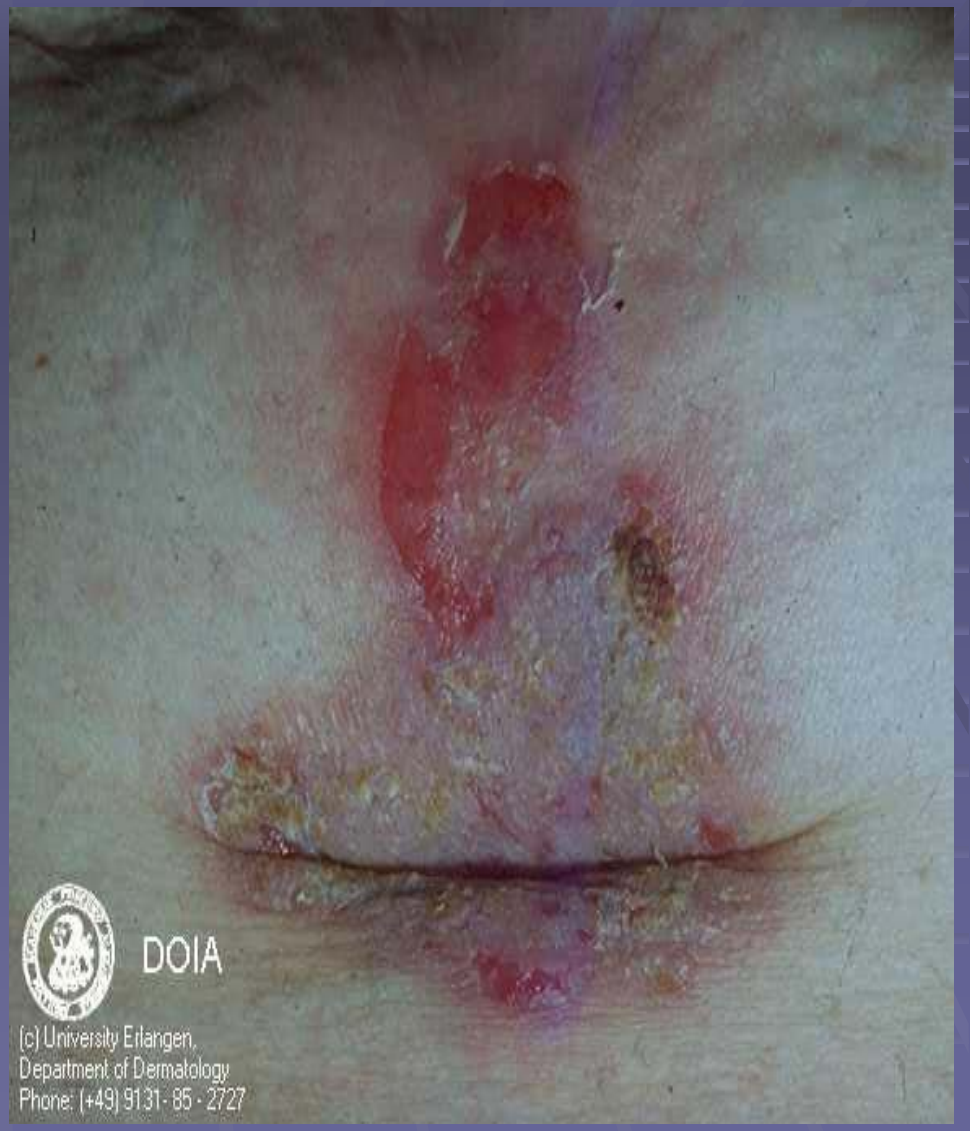
Flaccid blisters filled with clear fluid later fluid becomes turbid/ rupture to form erosions



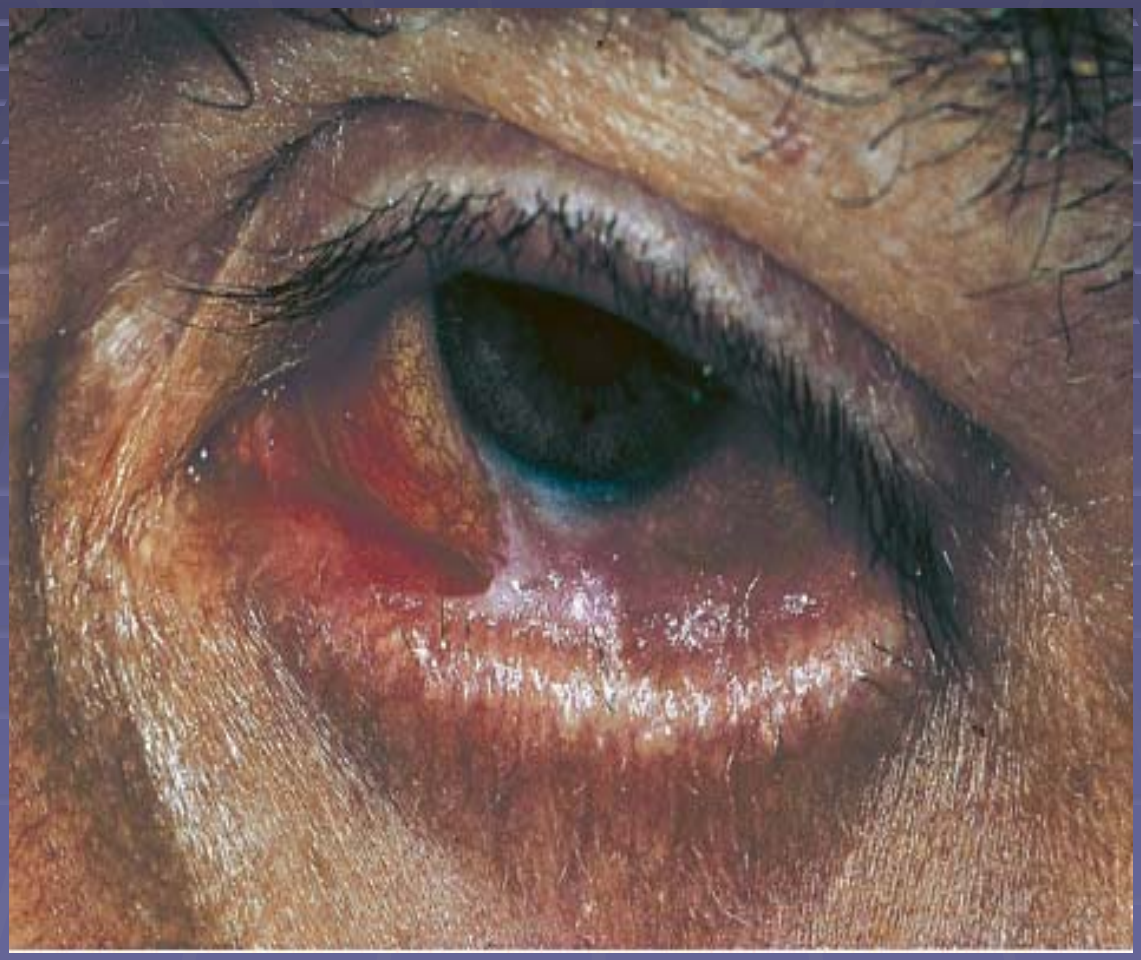
Nikolsky's Sign

Positive(firm sliding pressure separates normal looking epidermis from dermis)

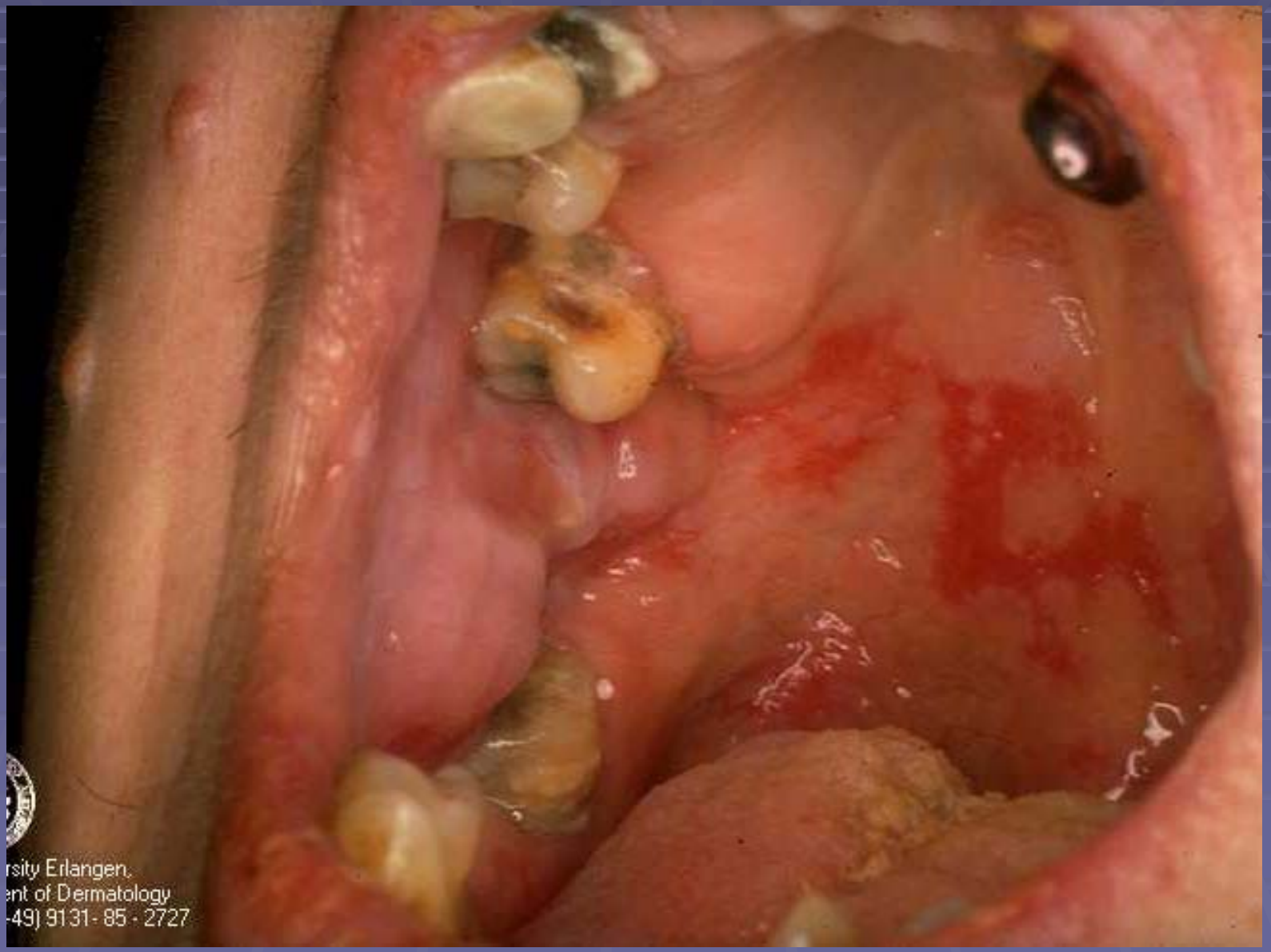
Bulla spread Sign : Positive



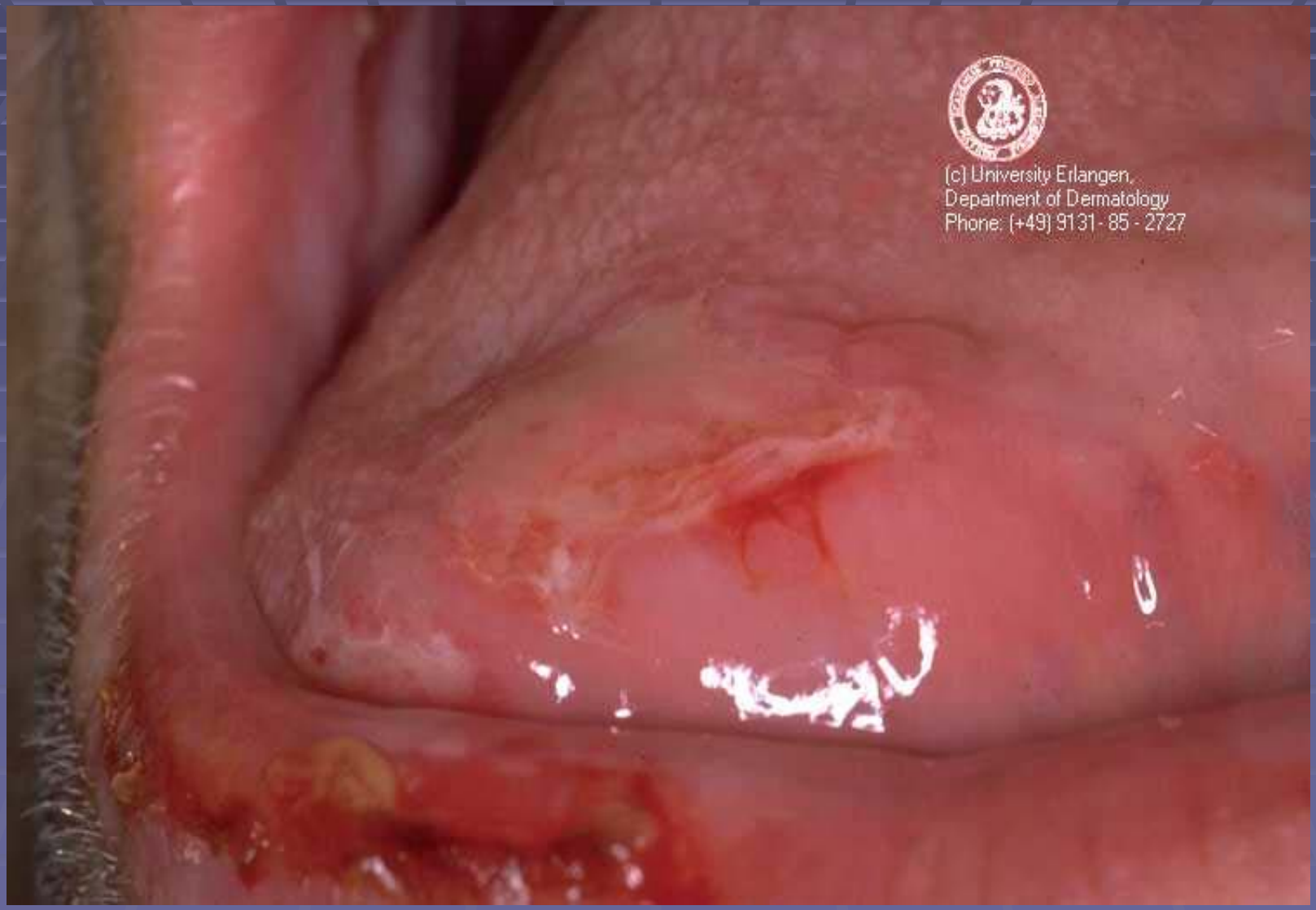
Conjunctival involvement



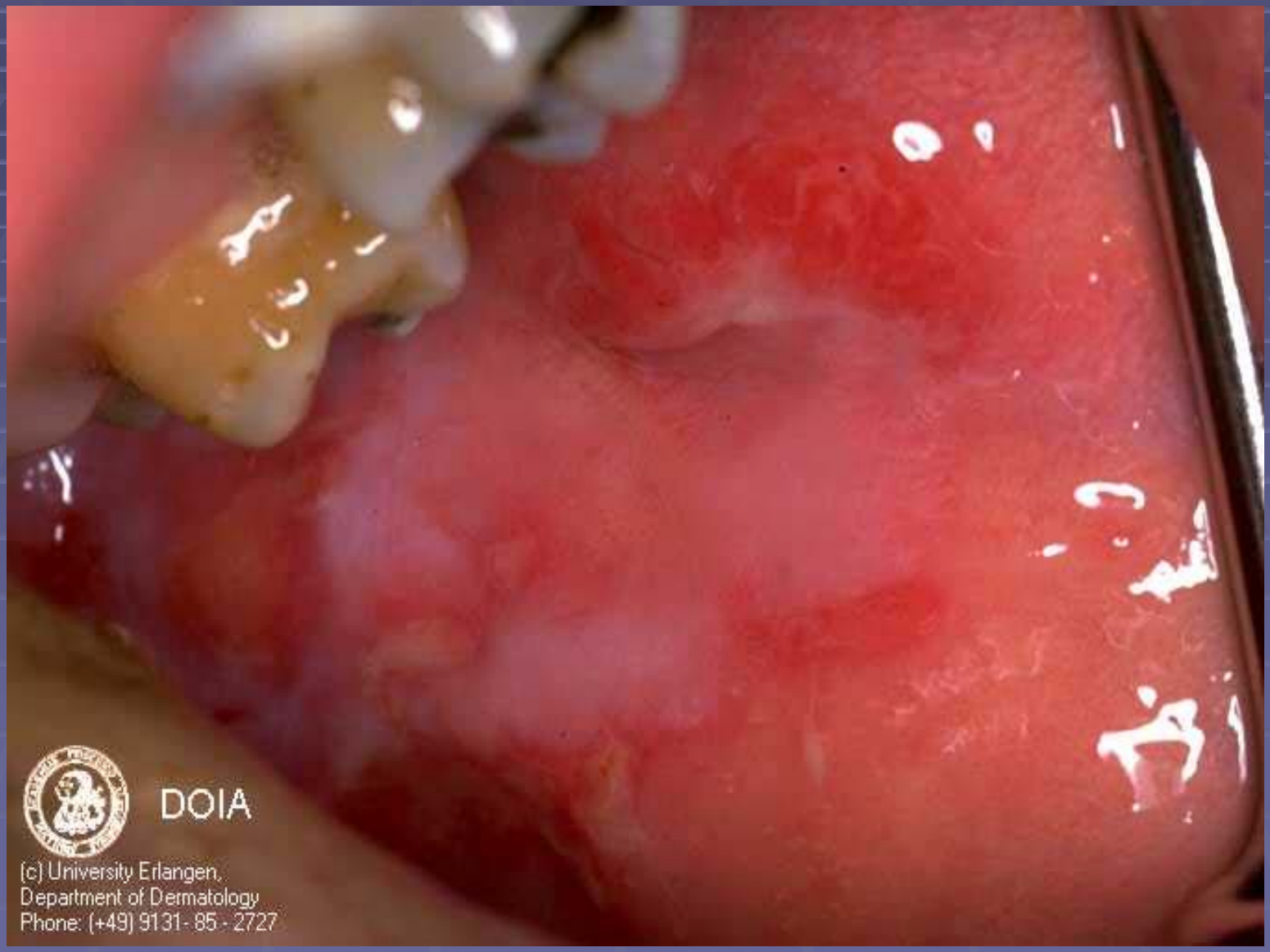
Mucosal involvement



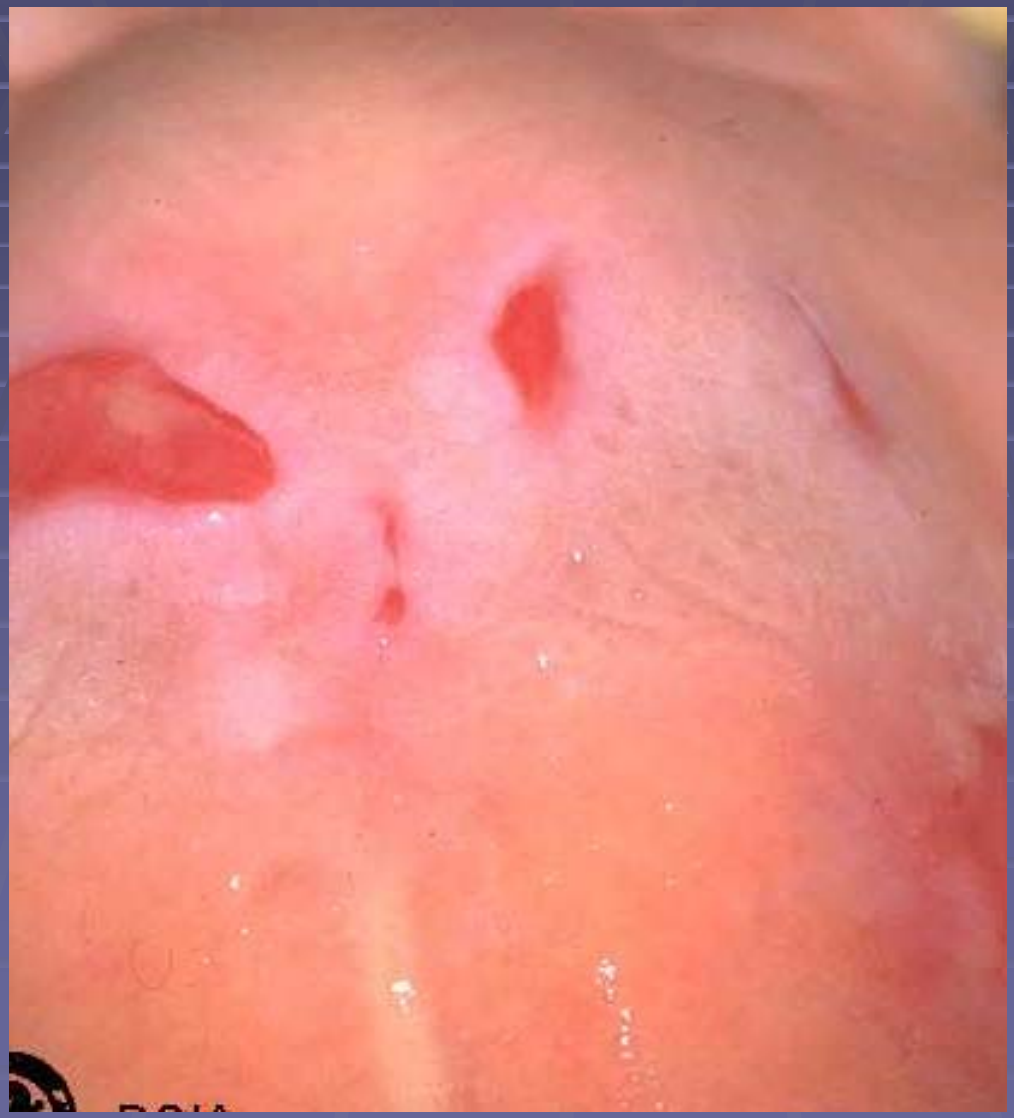
Mucosal involvement



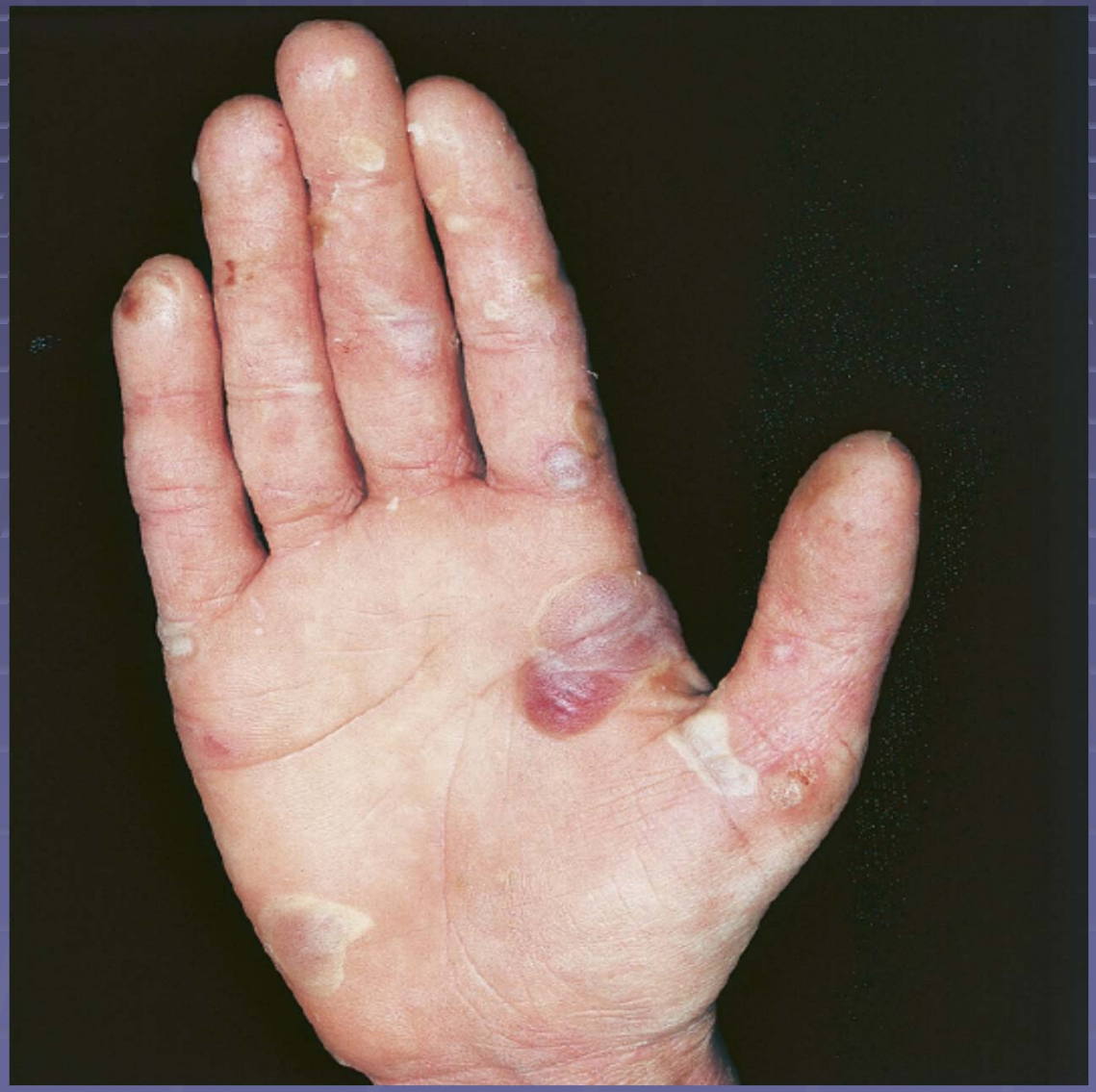
Mucosal involvement



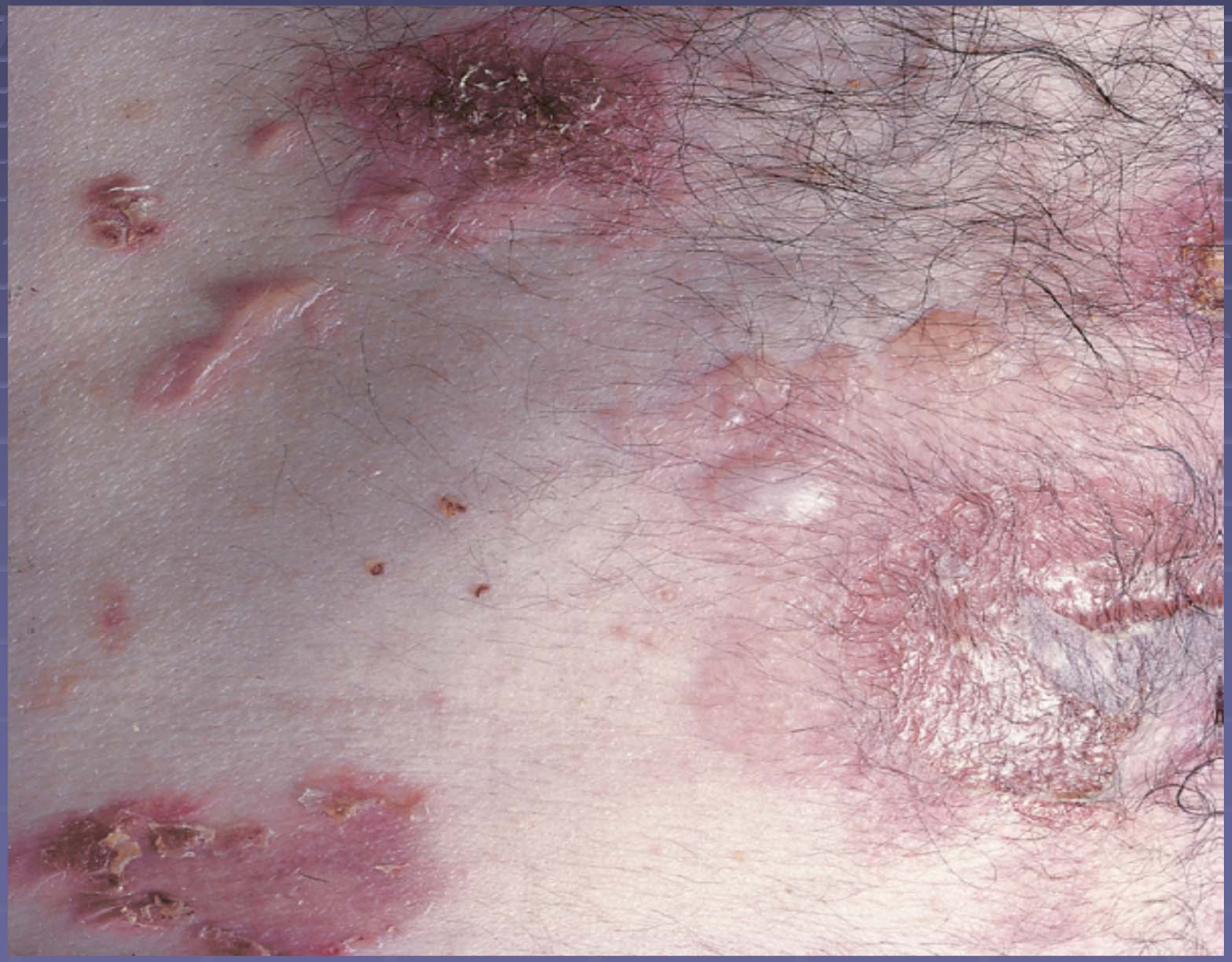
Mucosal involvement



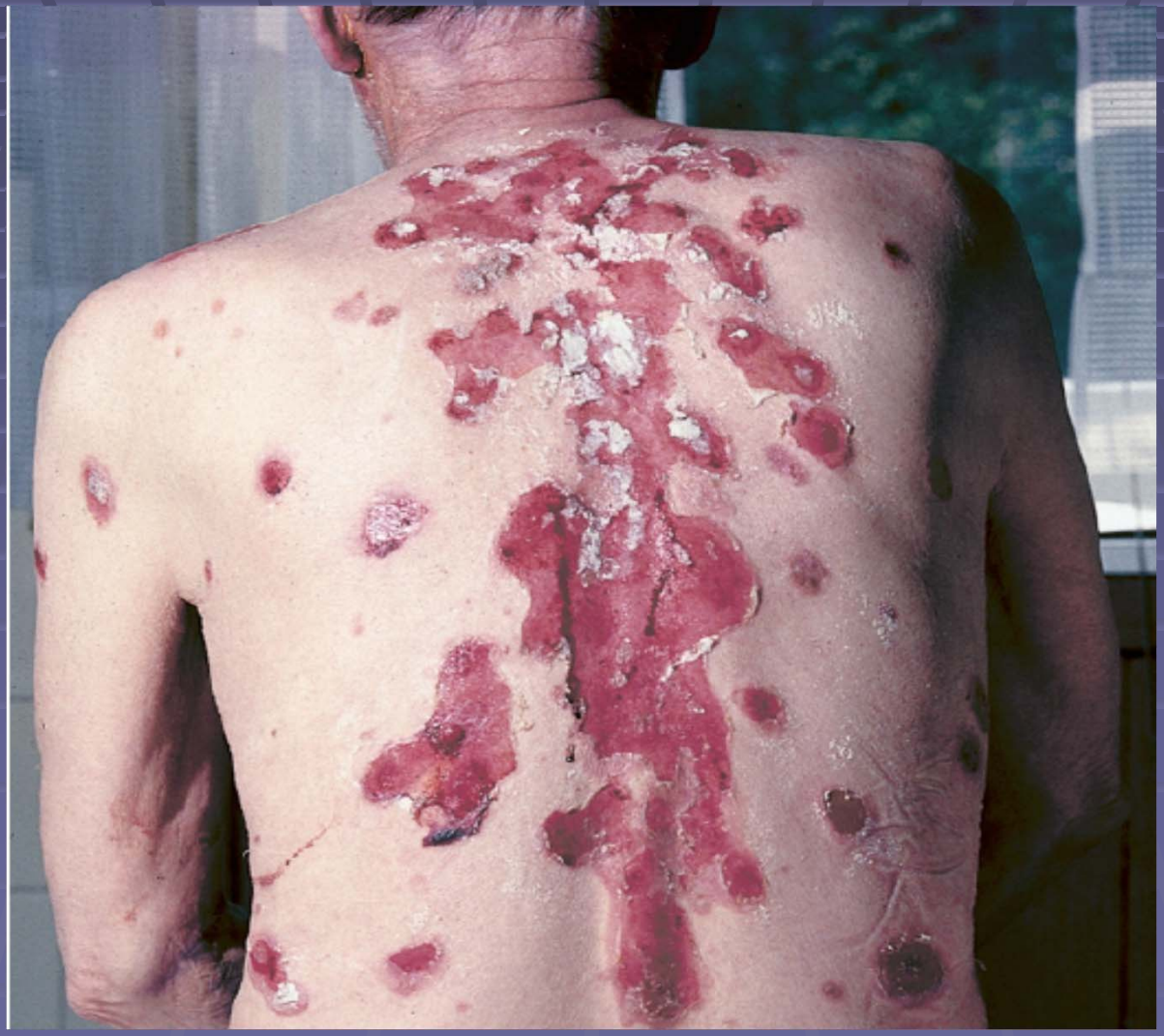
Cutaneous lesions



Cutaneous lesions



Cutaneous lesions



Pemphigus foliaceous

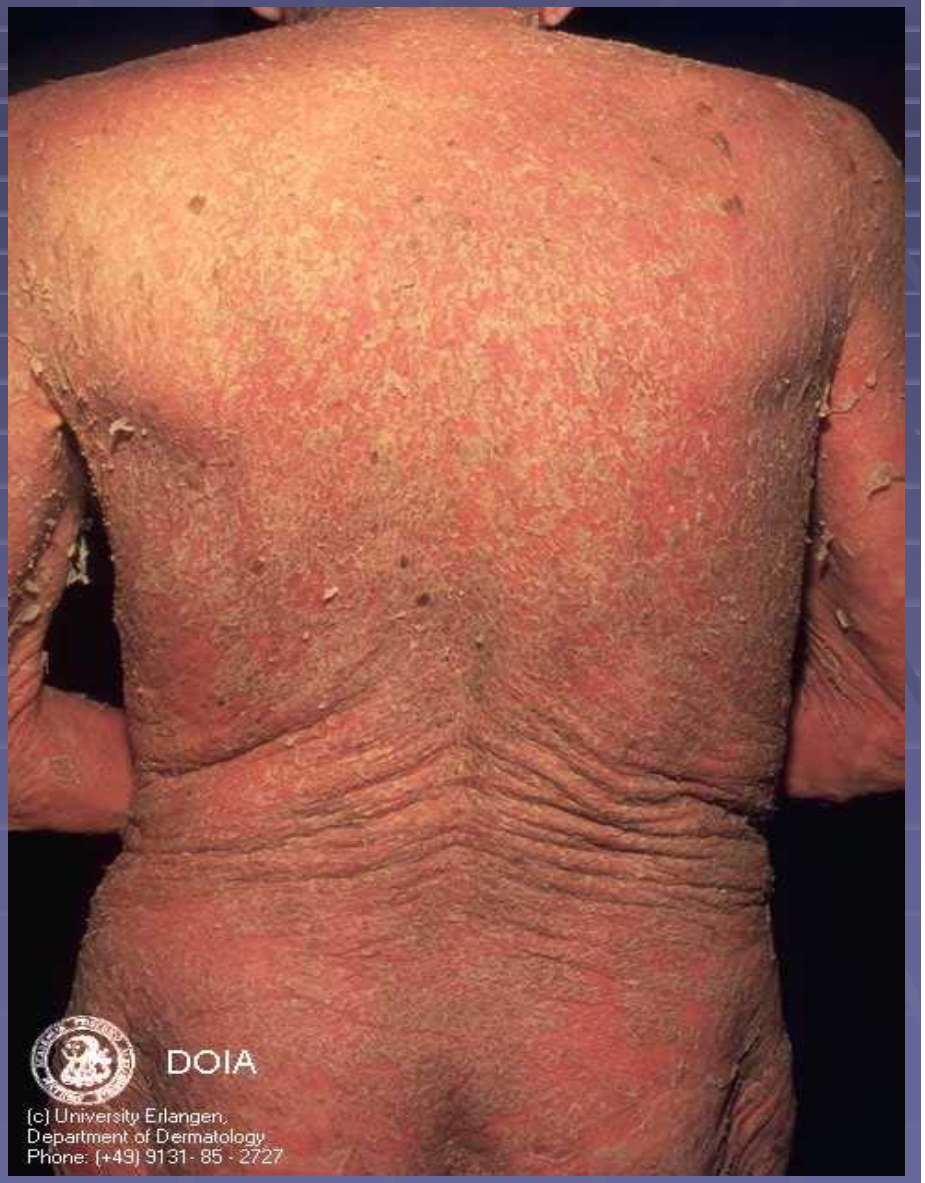
Blistering – **just below corneum**

Blistering may not be obvious

Antigen - Desmoglein – 1

Upper part of body involved

Mucosae - spared



Pemphigus erythematosus

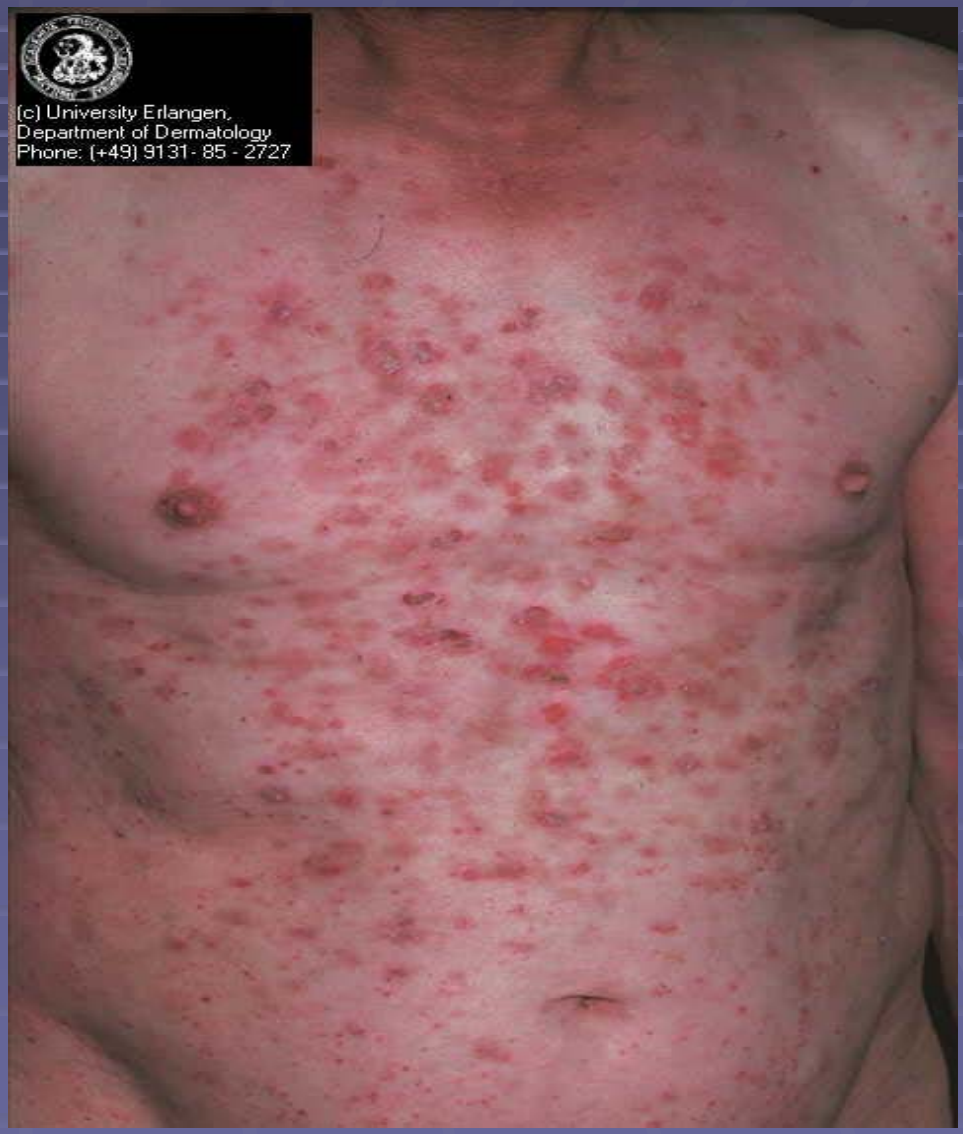
Erythematous, scaly lesions

Nose / cheeks

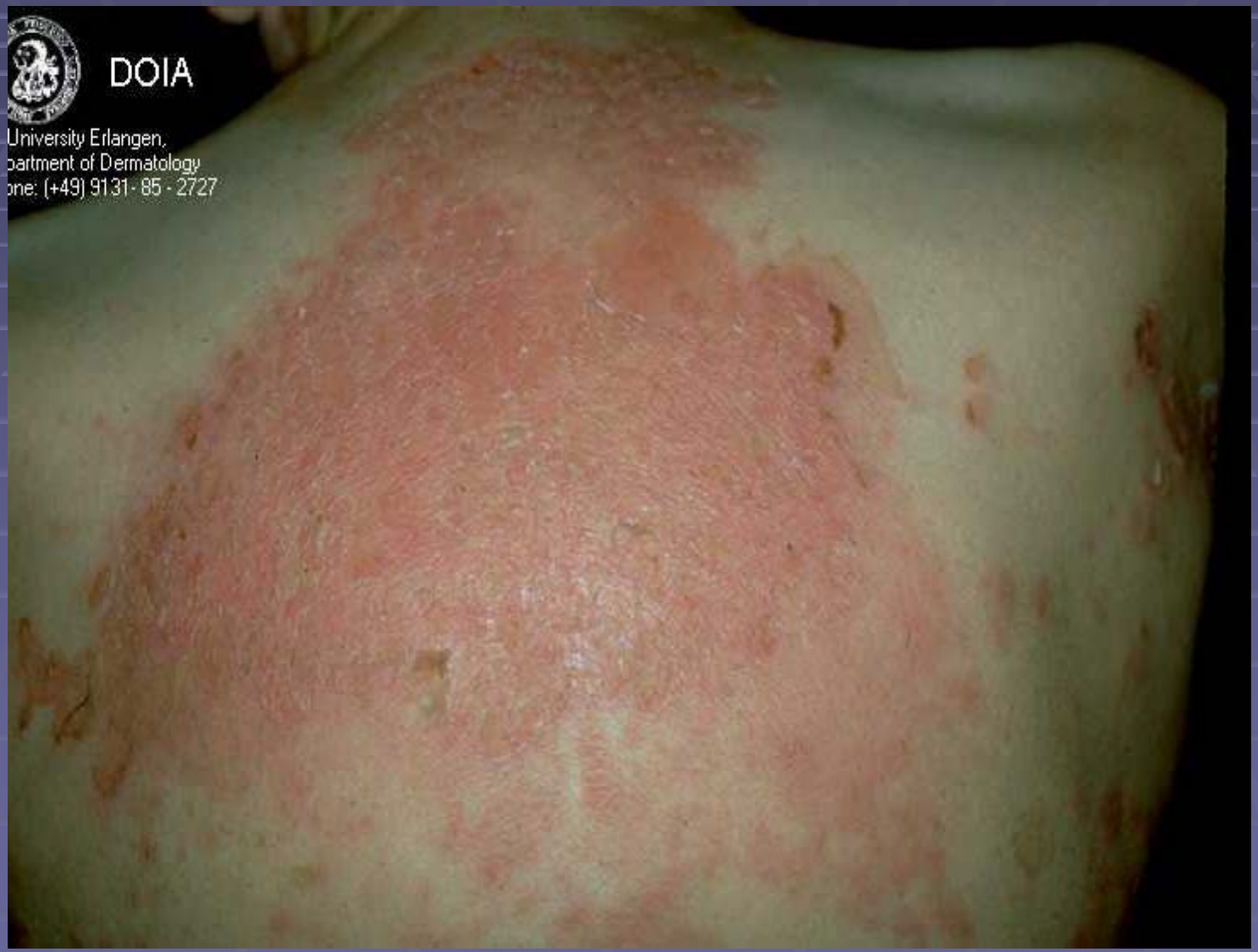
(butterfly distribution)

Lesions over trunk –

similar to P.foliaceous

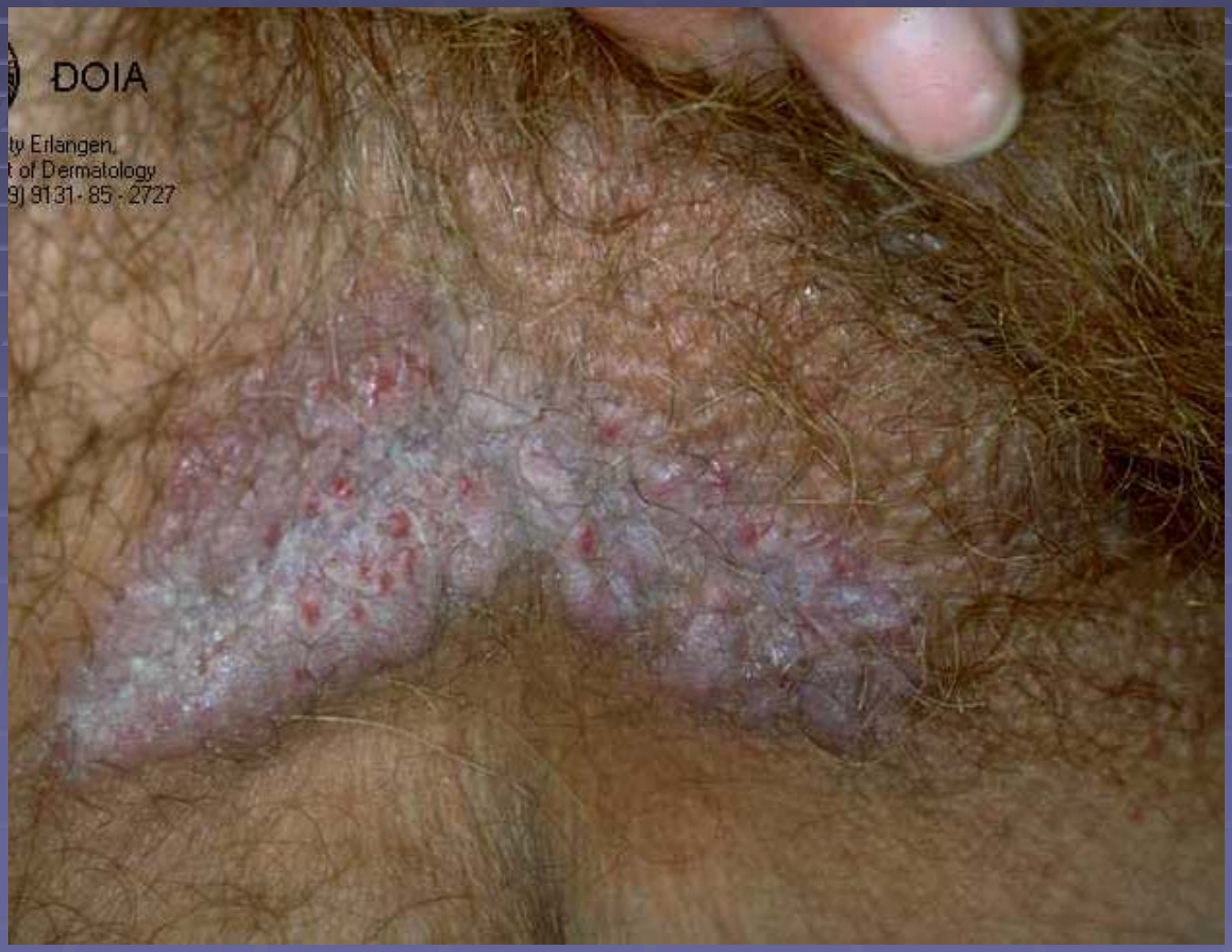


Pemphigus erythematosus

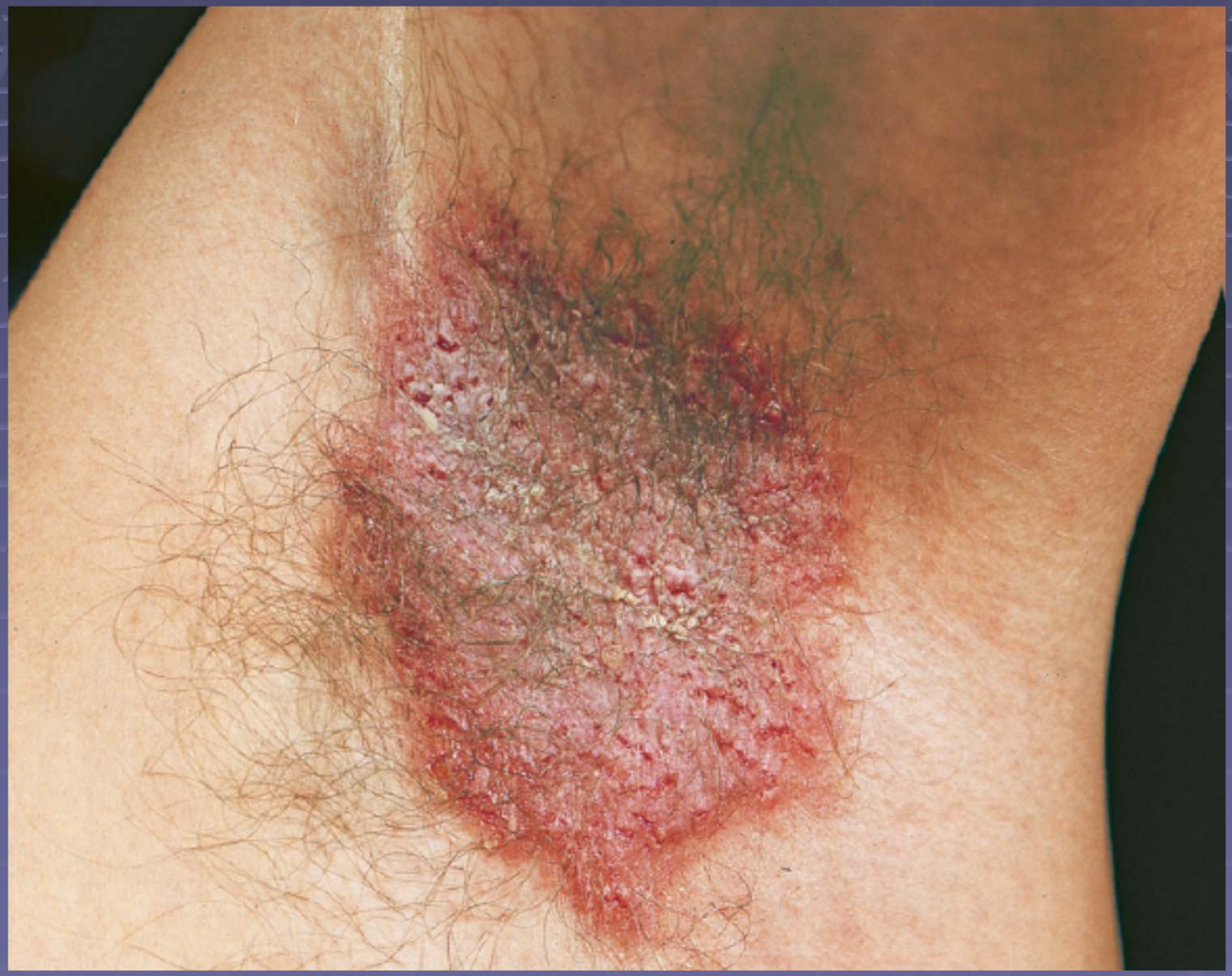


P. vegetans

Vegetating erosions in flexures



P. vegetans



Paraneoplastic pemphigus

Underlying malignancy

Drug Induced pemphigus

Pencillamine, Penicillin, Captopril, Enalapril,

Cephalosporins & Rifampicin

Treatment of pemphigus

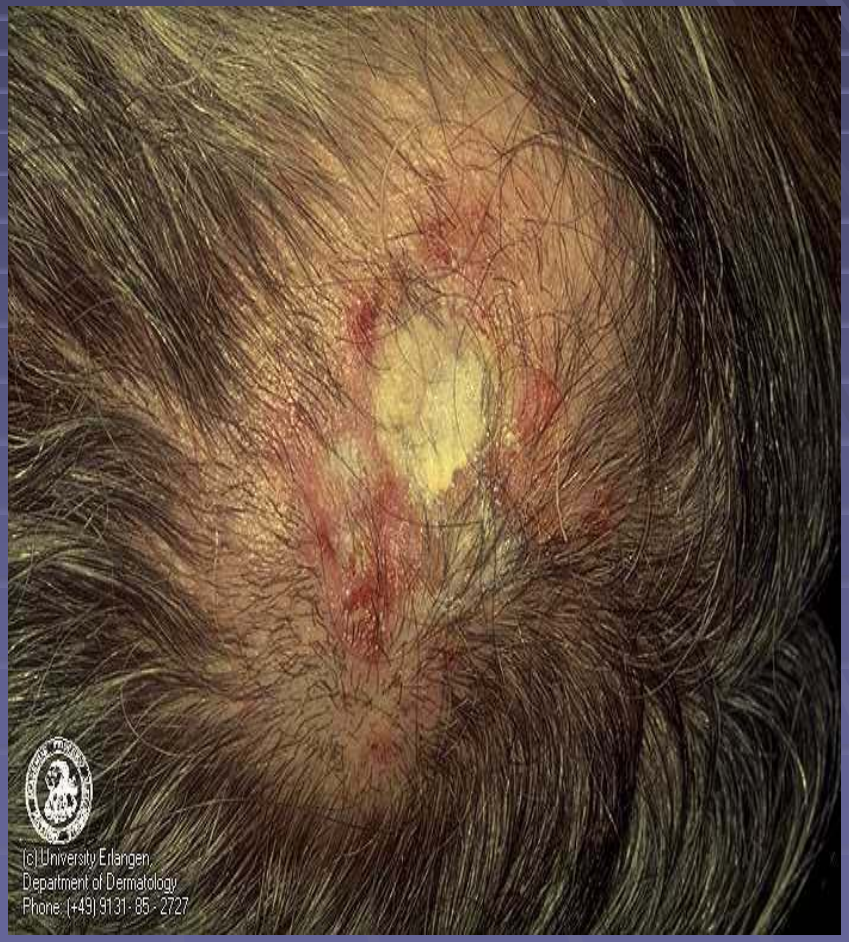
Corticosteroids 1.0 –1.5 mg/kg/day

(Pulse therapy,oral regime)

Azathioprime 2.5mg/kg/day

Cyclophosphamide 1-3 mg/kg

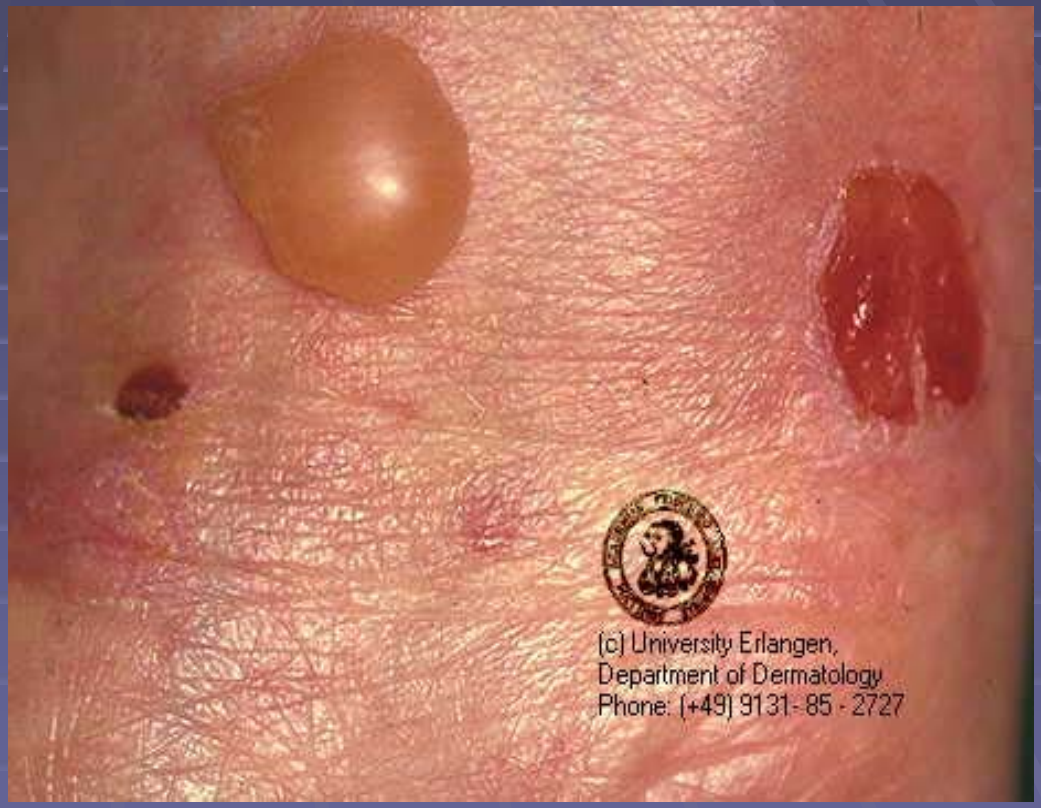
Cyclosporin 5 mg /kg/day



Bullous pemphigoid

-Blistering disease of elderly

Tense , large bullae



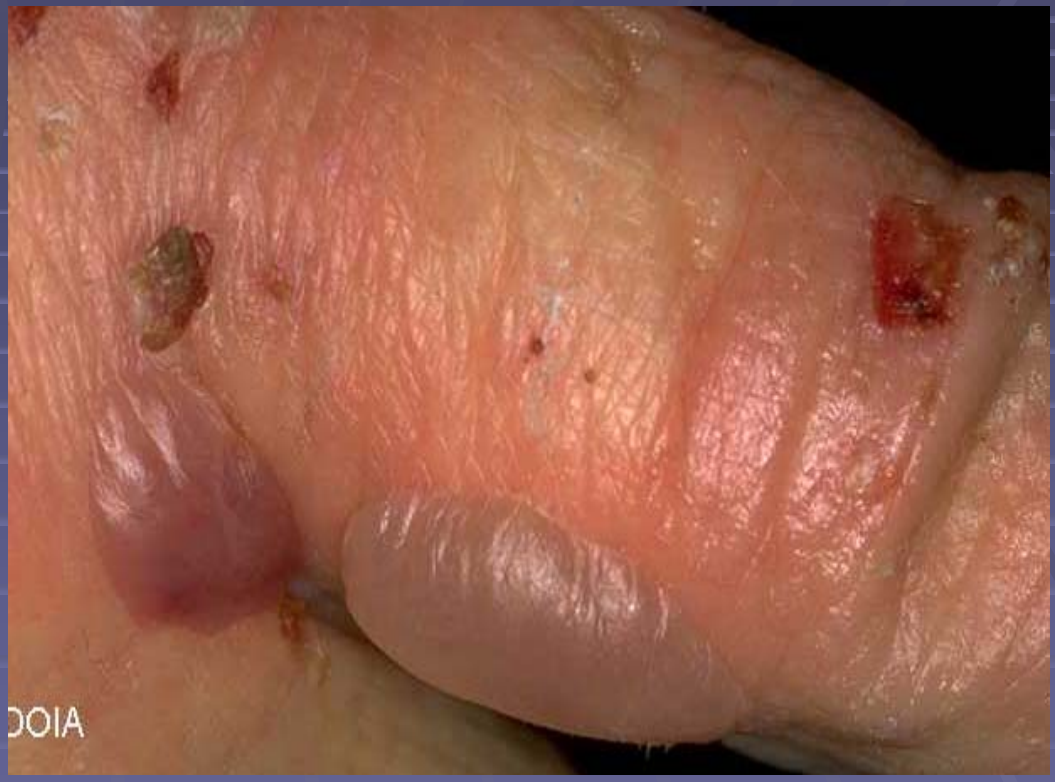
Etiology and Pathogenesis

-Not well defined

Drugs-

(Pencillamine,captopril, antipsychotics, aldosterone antagonists)

-HLADQ_7

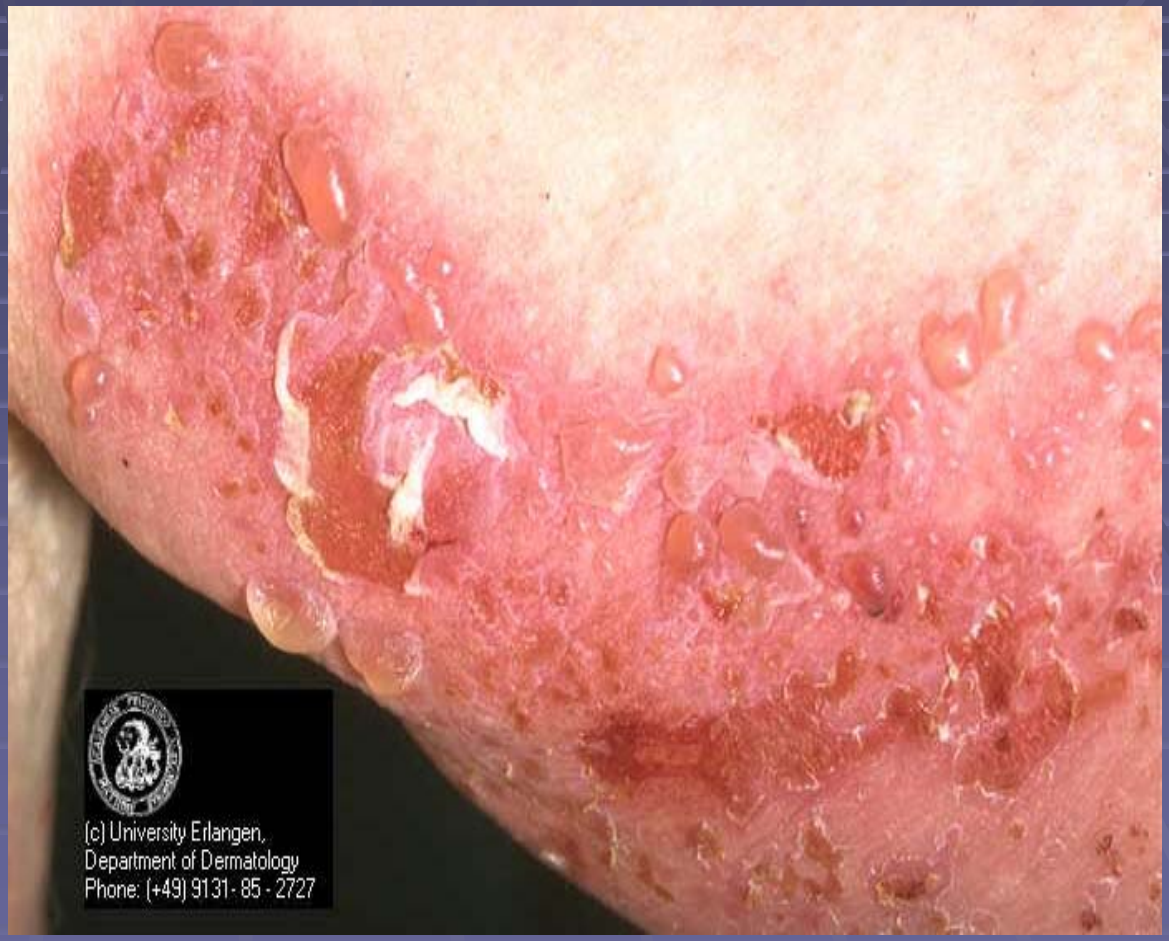


Histopathology

Subepidermal blister

Dermal inflam. infilt.

- Eosinophils,
- neutrophils,
- lymphpcytes



Clinical features

Urticaria like /or eczematous rash

Bullous lesions- after wks to months

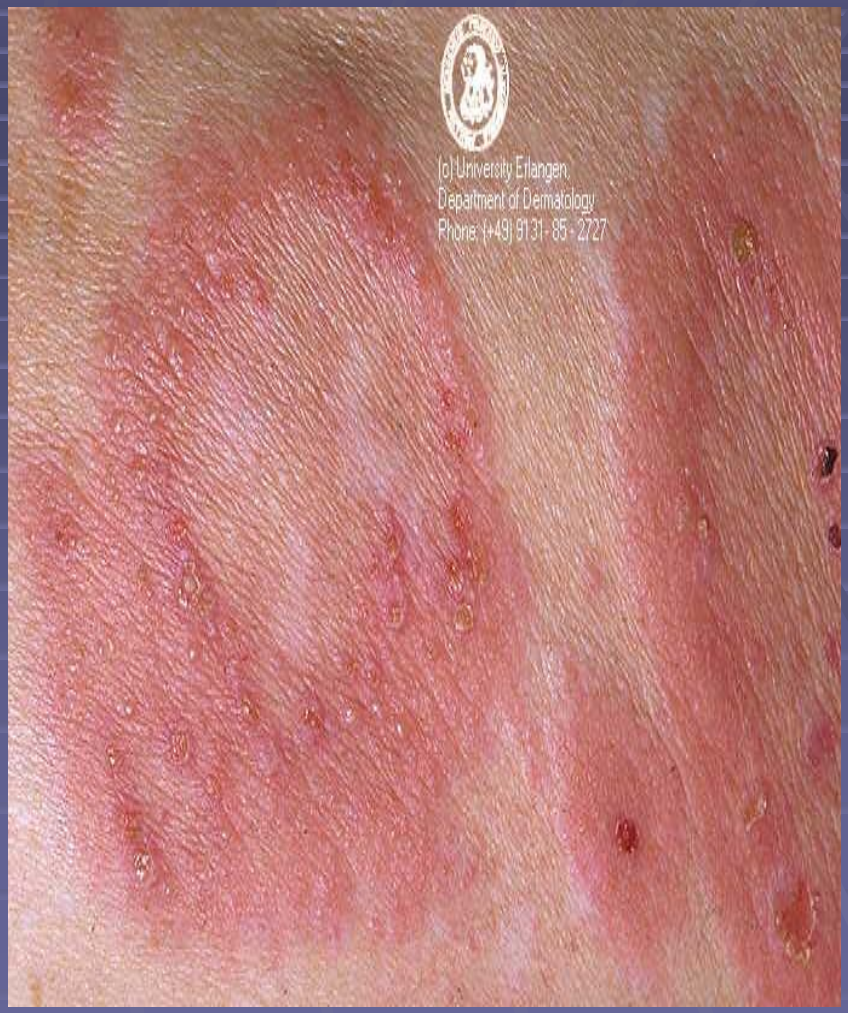
Tense, dome shaped,

upto dia. of 7 cm

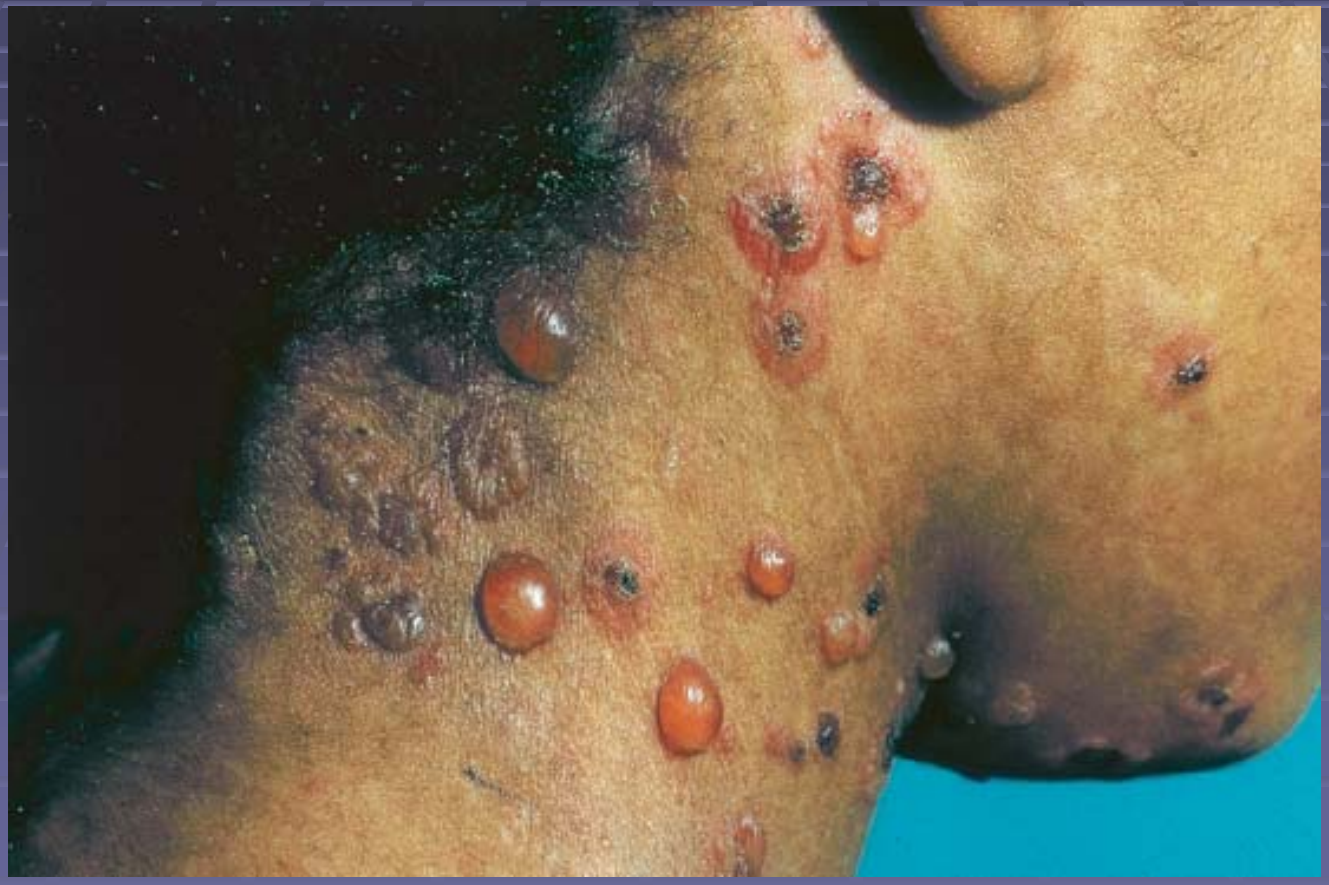
Flexural aspects of limbs/abdomen

Remain intact for several days

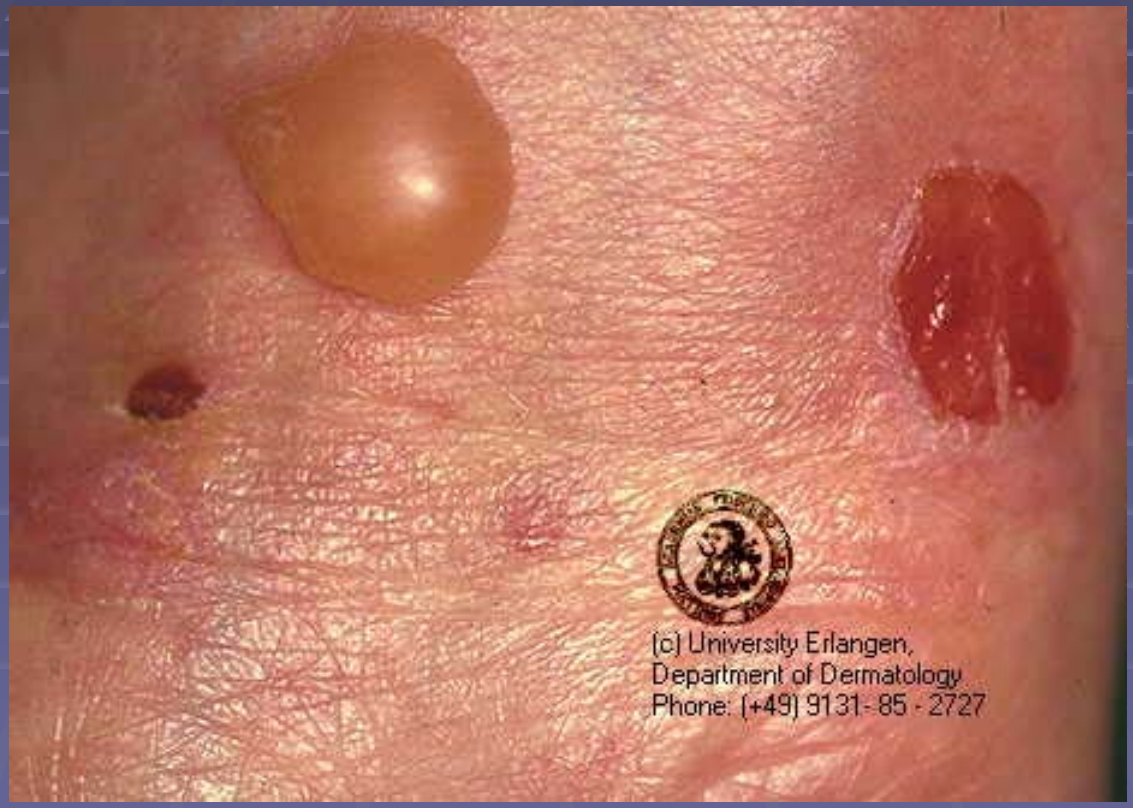
General condition – Good.



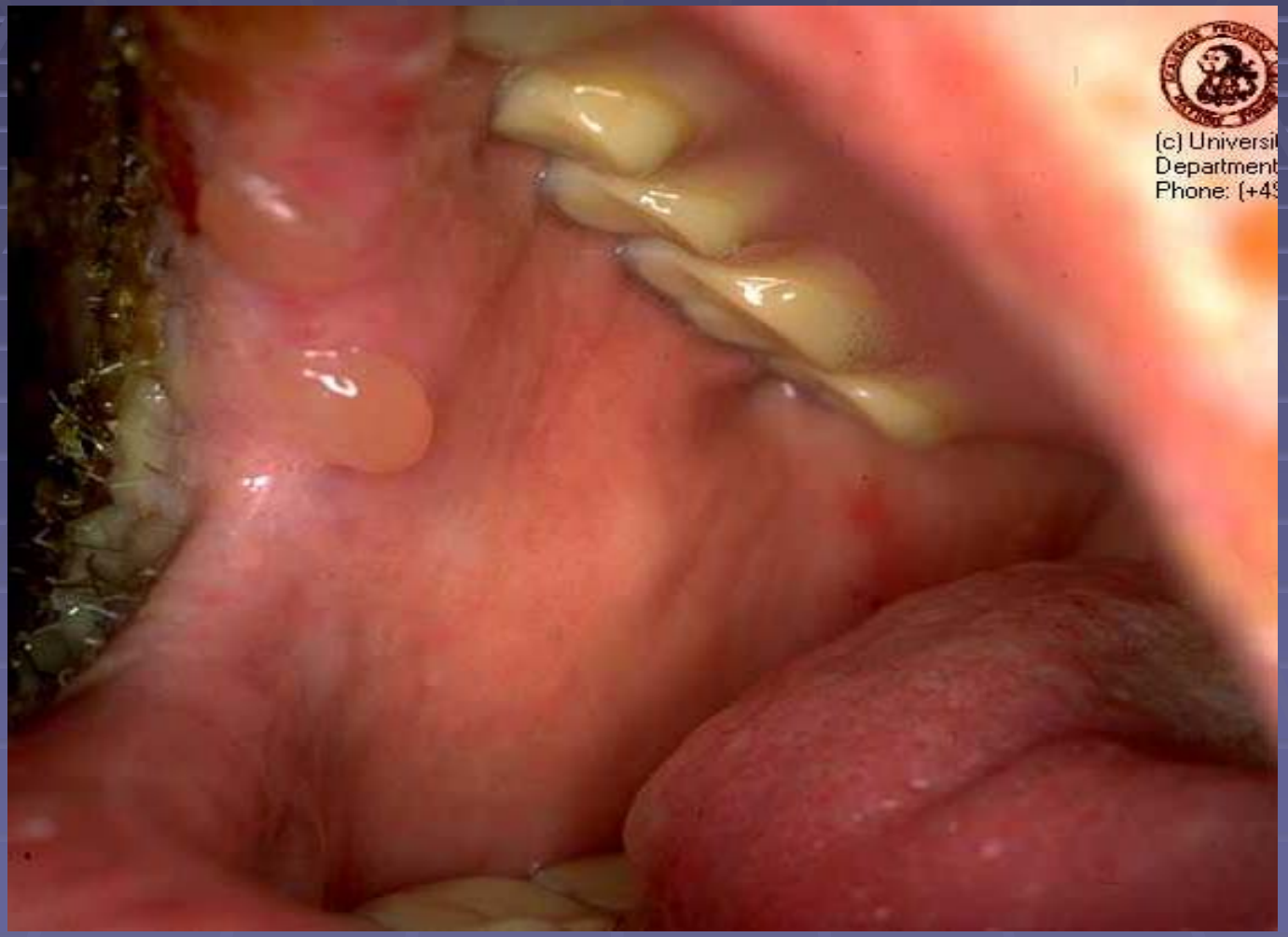
Bullous pemphigoid



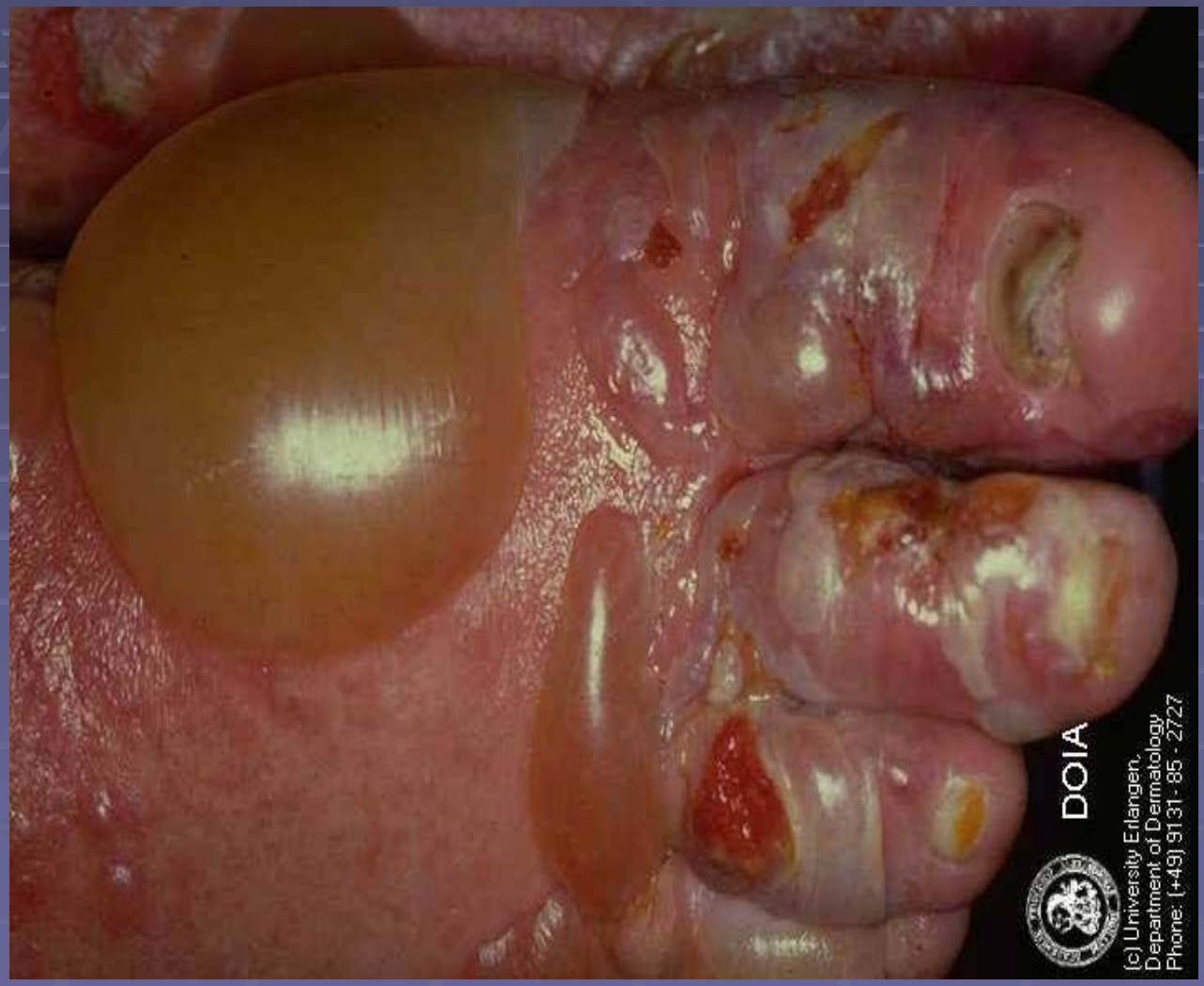
Bullous pemphigoid



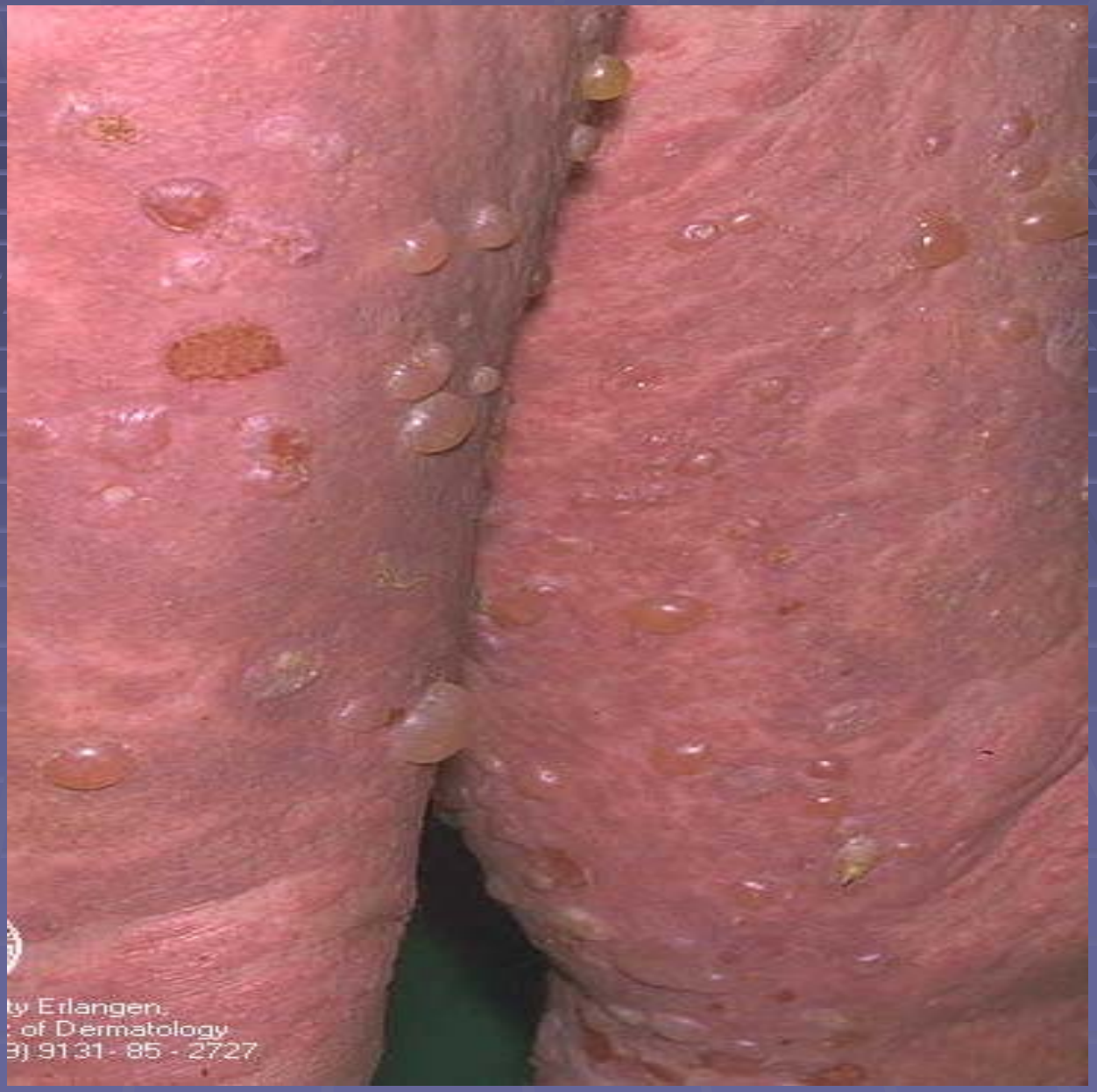
Bullous pemphigoid



Bullous pemphigoid



Bullous pemphigoid



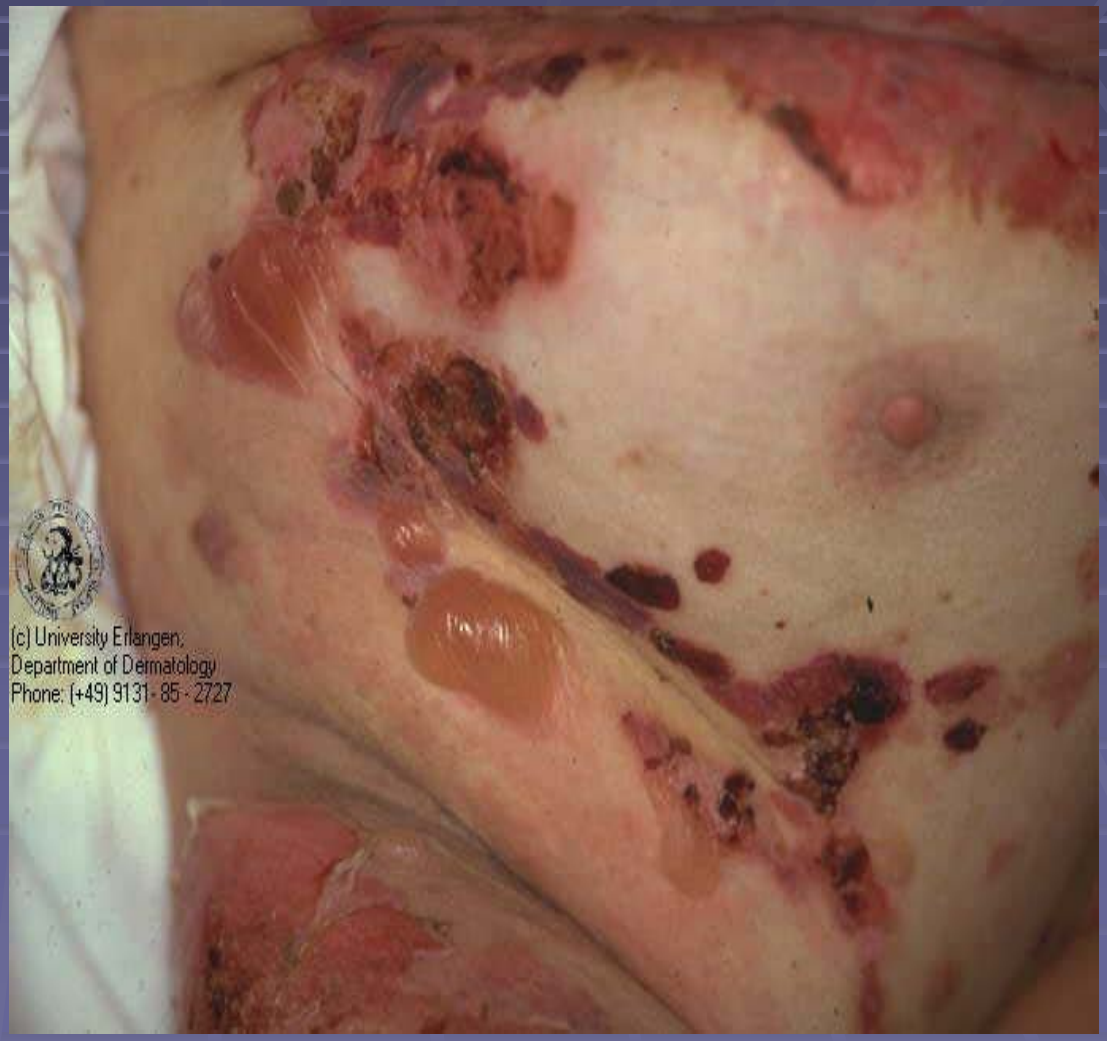
Treatment

Topical steroids

Systemic steroids

–20-40 mg/day

Dapsone



Differences between pemphigus and pemphigoid

Pemphigus	Pemphigoid
Middle age	Elderly
Flaccid blister	Tense
Remains intact for short time	Long
No other lesion preceding bullae	Urticaria/eczema
General condition – deteriorated	Good

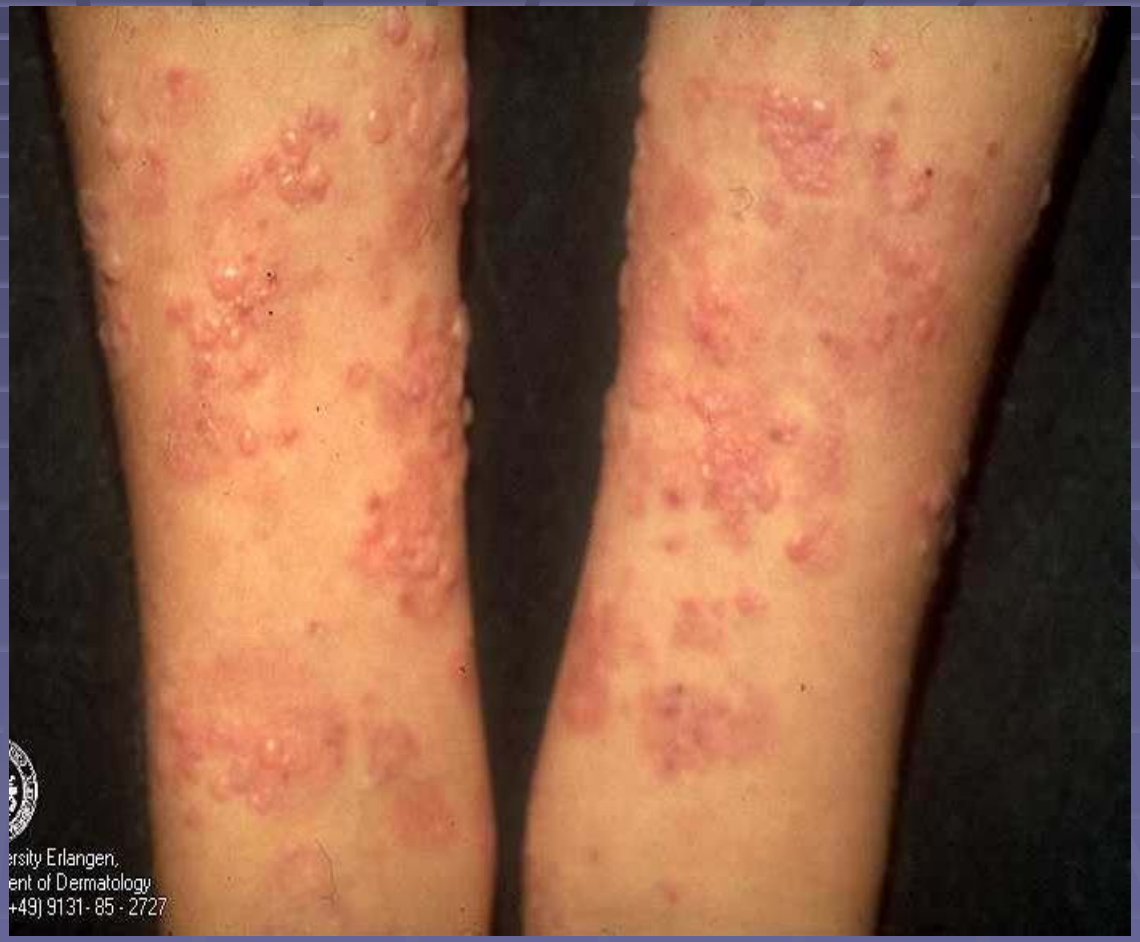
DERMATITIS HERPETIFORMIS

Rare,

Intensely pruritic chronic,

Recurrent

Papulovesicular disorder



Clinical features

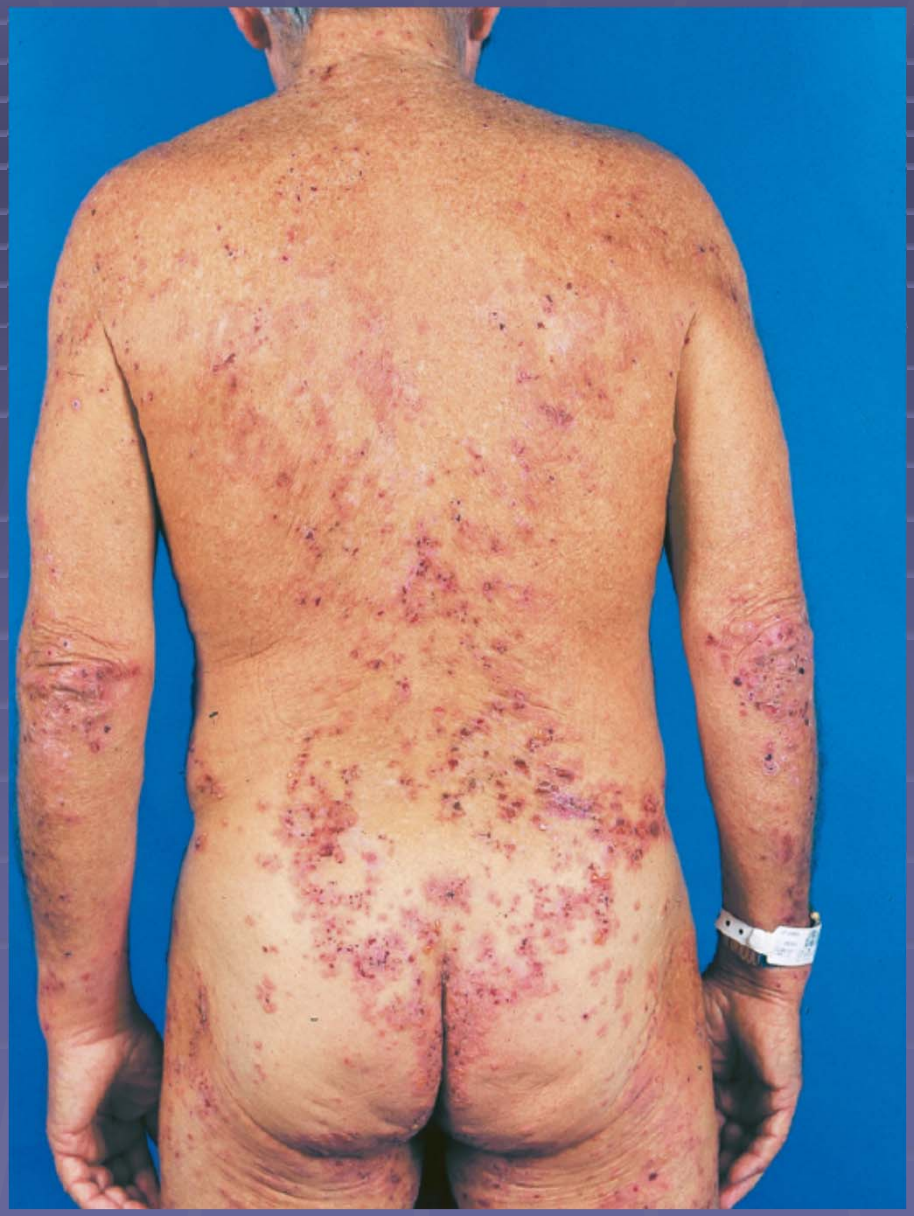
Age - 20-50 yrs

Pruritus-first/predominant symptom

Early lesions-
papules, weals or grouped vesicles

Extensor aspects of body
Gluten sensitive enteropathy

DERMATITIS HERPETIFORMIS



Treatment

Gluten free diet

Dapsone :100-200 mg/day.