

## BULLOUS DISORDERS

### I. Epidermolysis Bullosa--noninflammatory inheritable blistering disorders, skin

fragility

A. E.B. Simplex

B. E.B. Letalis (junctional)

C. E.E. Dystrophica

D. Treatment: trauma avoidance

1. cool environs
2. antibiotics, local wound care
3. steroids
4. Dilantin

### II. Pemphigus

A. Superficial

1. P. foliaceus
2. P. erythematous

B. Deep

1. P. vulgaris
2. P. vegetans

C. flaccid bullae on uninflamed base

D. + Nikolsky sign

E. Pathogenesis: autoantibodies to intercellular cement

F. Histopathology

1. acantholysis
2. intrarepidermal bullae
3. + immunofluorescence intracellularly in epidermis

G. Treatment:

1. Steroids!
2. immunosuppressants

III. Bullous Pemphigoid

- A. Tense bullae on inflamed base
- B. Nikolsky sign
- C. Pathogenesis: autoantibodies to basement membrane zone protein
- D. Histopathology
  1. no acantholysis
  2. intact epidermis overlying bulla with eo's, PMN's
  3. + immunofluorescence at DE junction
- E. Treatment: Steroids, immunosuppressants

IV. Dermatitis Herpetiformis

1. Intensely pruritic papulovesicles over elbows, knees, buttocks
2. gluten enteropathy (asymptomatic)
3. Pathogenesis: IgA antibodies in dermal papillae
4. Histopathology: Necrosis of dermal papillae with PMN's
5. Treatment: Dapsone, gluten free diet, antihistamines

V. Porphyria Cutanea Tarda

A. Blisters on dorsal hands with sun exposure

B. Zygomatic hypertrichosis

C. Marked skin fragility

D. Pathogenesis:

1. defect in uroporphyrinogen decarboxylase

2. increased porphyrins excreted in urine (positive fluorescence)

E. Aggravated by BCP's, EtOH

F. Treatment:

1. phlebotomy

2. antimalarials

3. EtOH avoidance

4. sunscreens