BLISTERING CONDITIONS

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OVERVIEW

- CATEGORIES
- HOW TO INVESTIGATE
- FEW CASES OF AUTOIMMUNE BLISTERING CONDITIONS
- EXAMPLES OF OTHER BLISTERING CONDITIONS

Primary blistering conditions

BP, pemphigus, paraneoplastic, EBA, linear IgA, gestational pemphigoid, dermatitis herpetiformis

Drug induced

FDE, TEN/SJS, drug induced BP or pemphigus

Infection

Varicella, erysipelas, hand foot and mouth, HSV, Impetigo, Staph scalded skin syn.

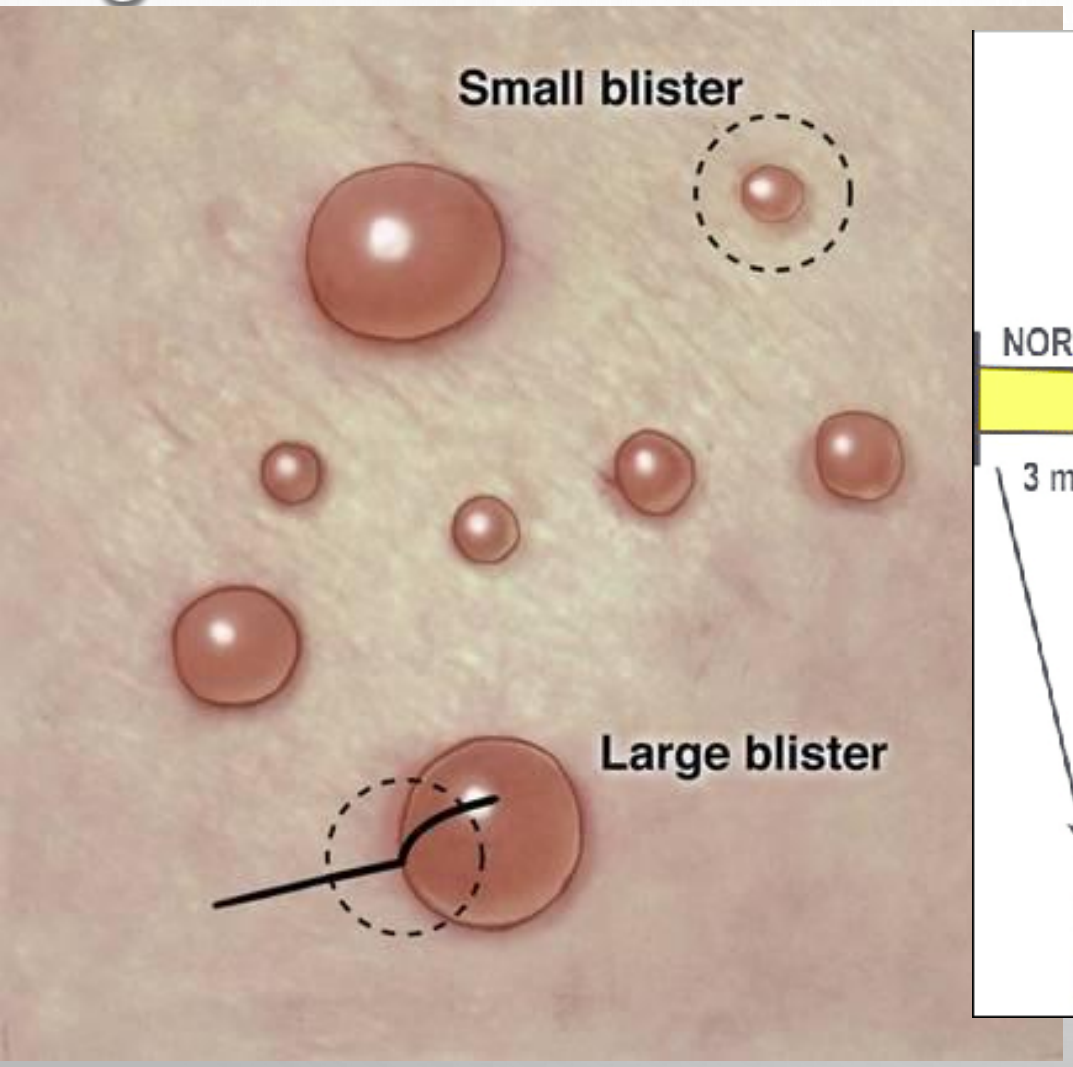
Dermatitis

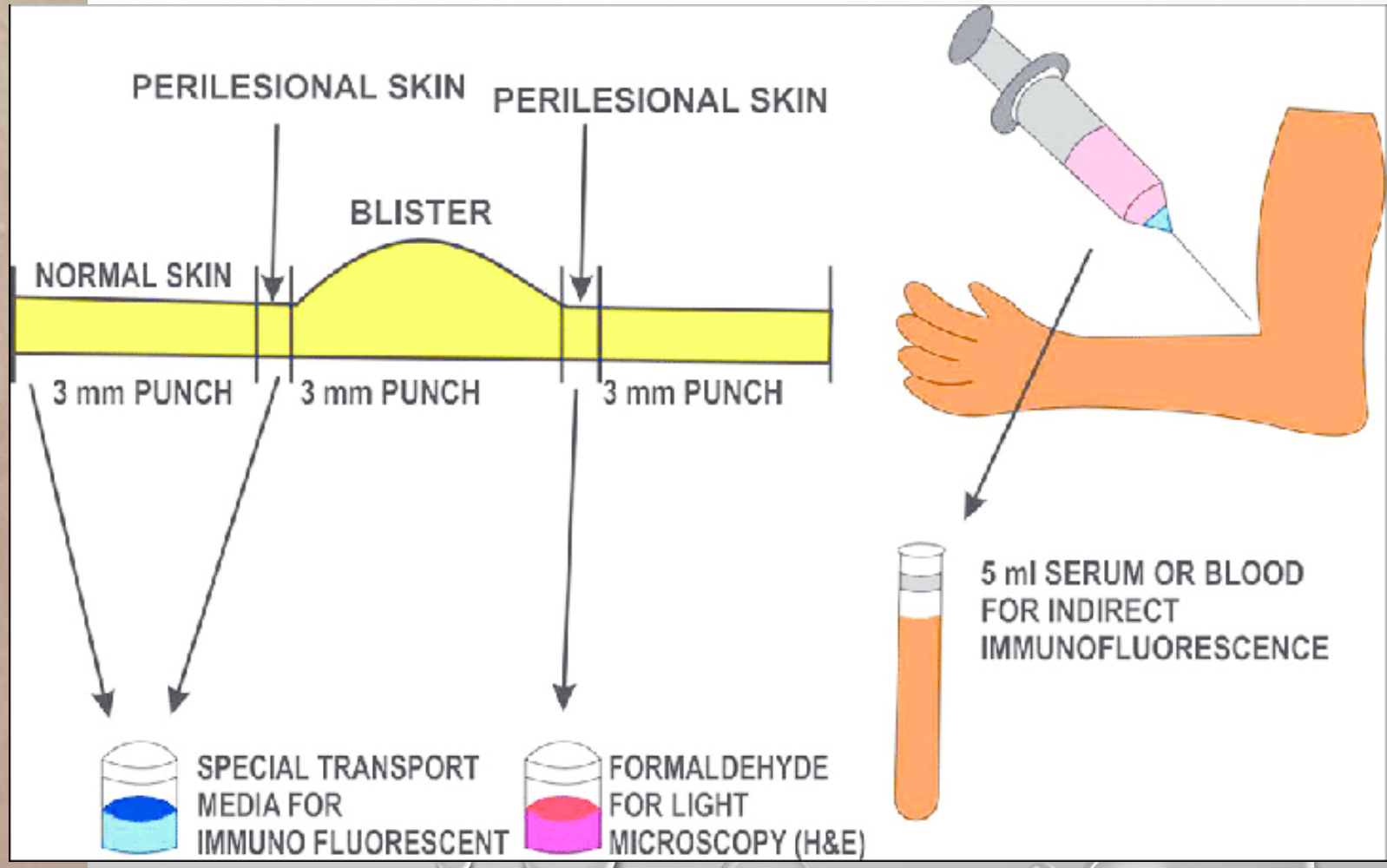
Endogenous eg pompholyx, contact eg plants.

Misc.

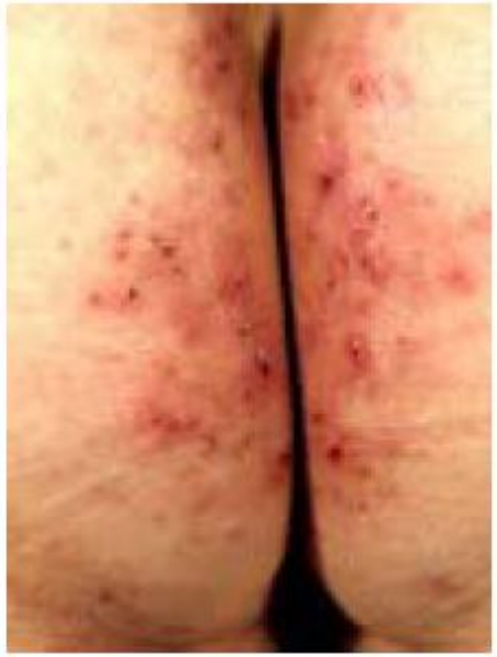
Chilblains, EM, LP, Lupus, PMLE, Insect bites, Porphyria cutanea tarda

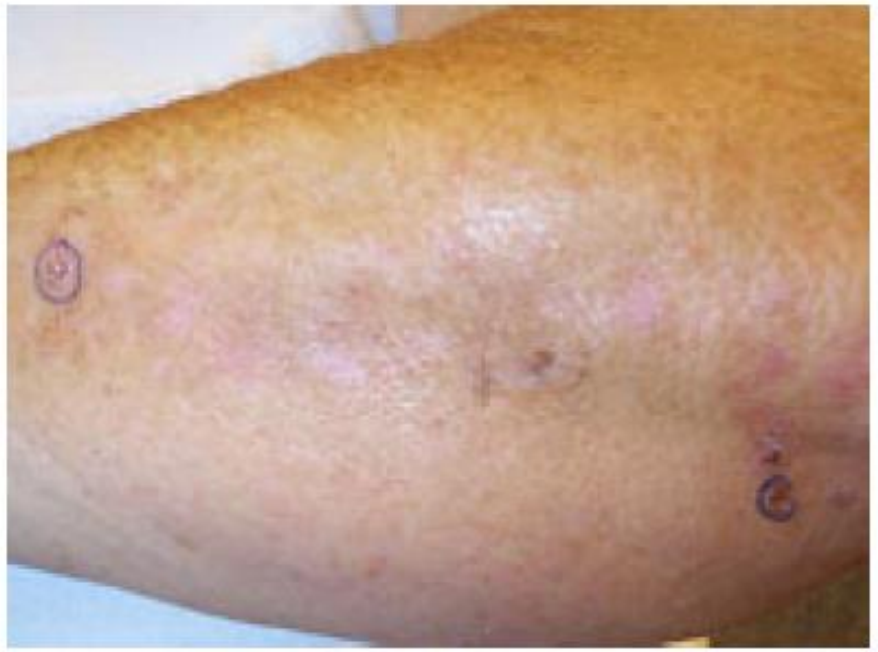
DIAGNOSTIC INVESTIGATIONS





PRIMARY BLISTERING CONDITIONS





A 54-year-old woman presents with a six-month history of intensely itchy papules and vesicles on elbows, knees and buttocks as shown above. She is systemically well other than mild diarrhoea. Blood count is reported as normal. Serum ferritin is 5 µg/L [20-380] and red blood cell folate is 180 nmol/L [600-1600].

Which of the following investigations is most likely to confirm the diagnosis of the rash shown above?

A. IgA endomysial antibodies.

B. Direct immunofluorescence of perilesional skin.

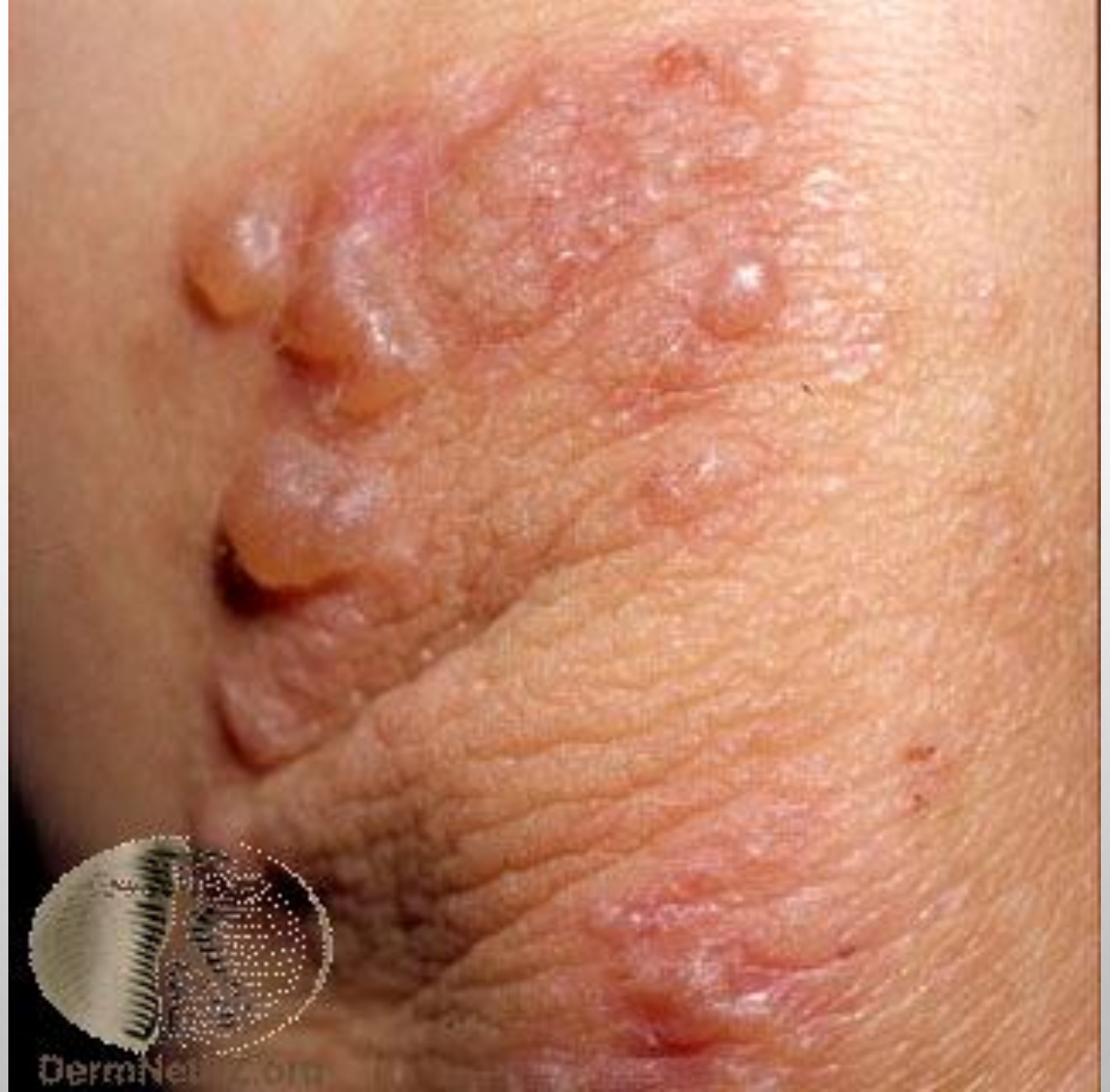
C. Light microscopy of unexcoriated vesicle.

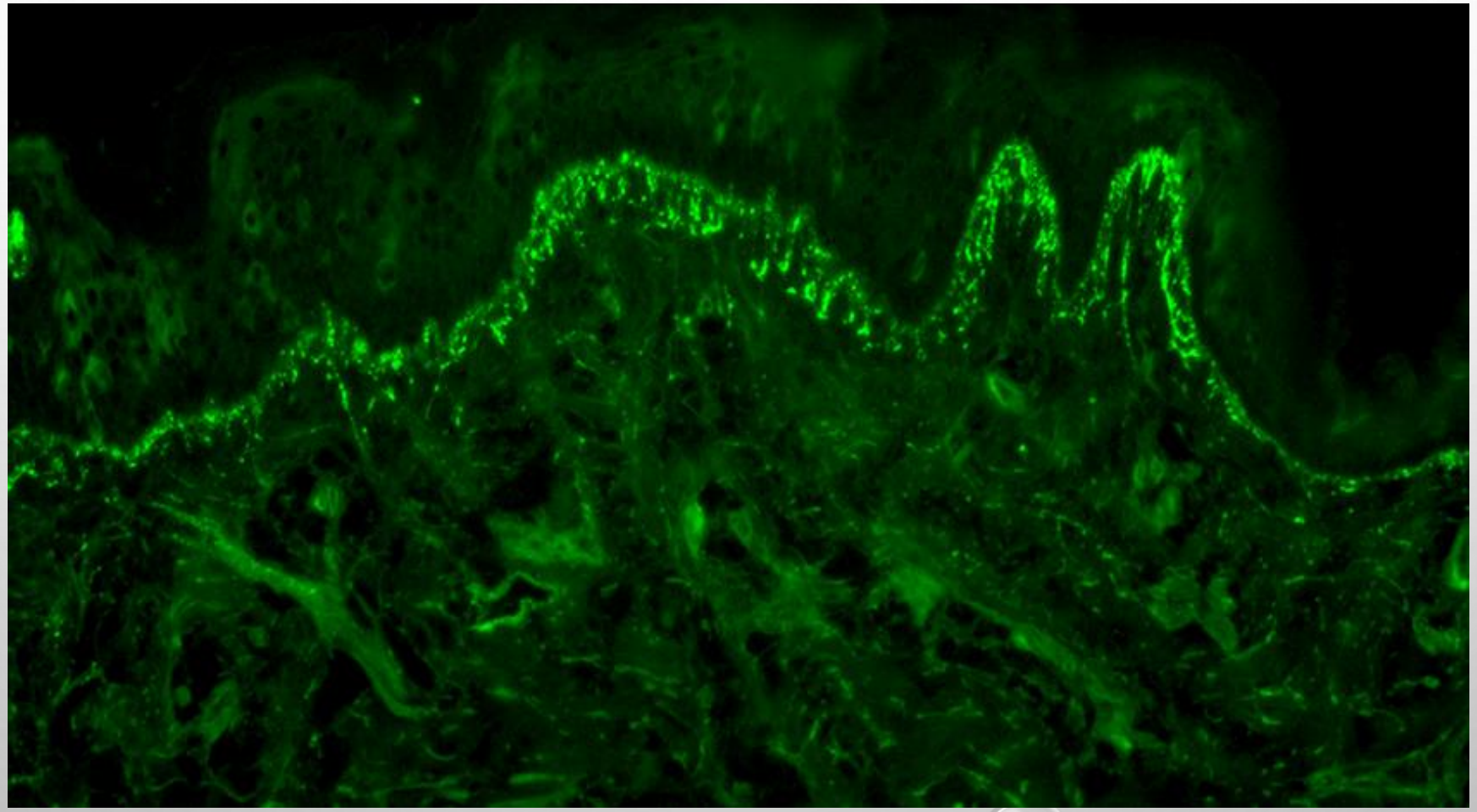
D. Jejunal biopsy.

E. Response to gluten-free diet.

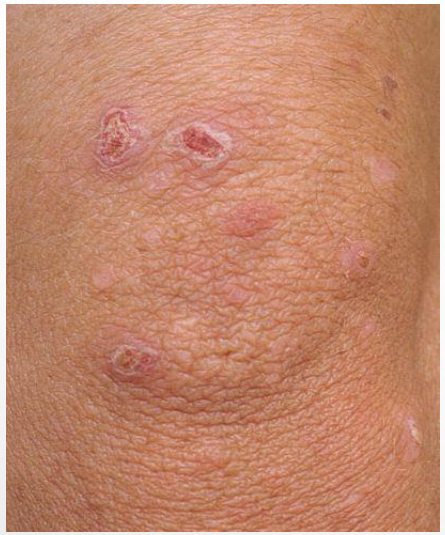
DERMATITIS HERPETIFORMIS

- Immunobullous condition assoc with coeliac disease
- Assoc with HLA DQ2/DQ8
- Gluten → IgA Abs → Attack skin & gut
- Symmetrical, intensely itchy crops of vesicles on elbows & knees
- Ix:
  - Skin biopsy (IgA deposits on DIF)
  - Coeliac serology
  - Small intestine biopsy





How would you treat her?



Most patients obtain excellent control with the following:

Dapsone; initial dose 50mg building up to 300mg daily if necessary, providing haemoglobin level is adequate.

If dapsone results in unacceptable side effects, sulfasalazine 1-2 g/day can be used.

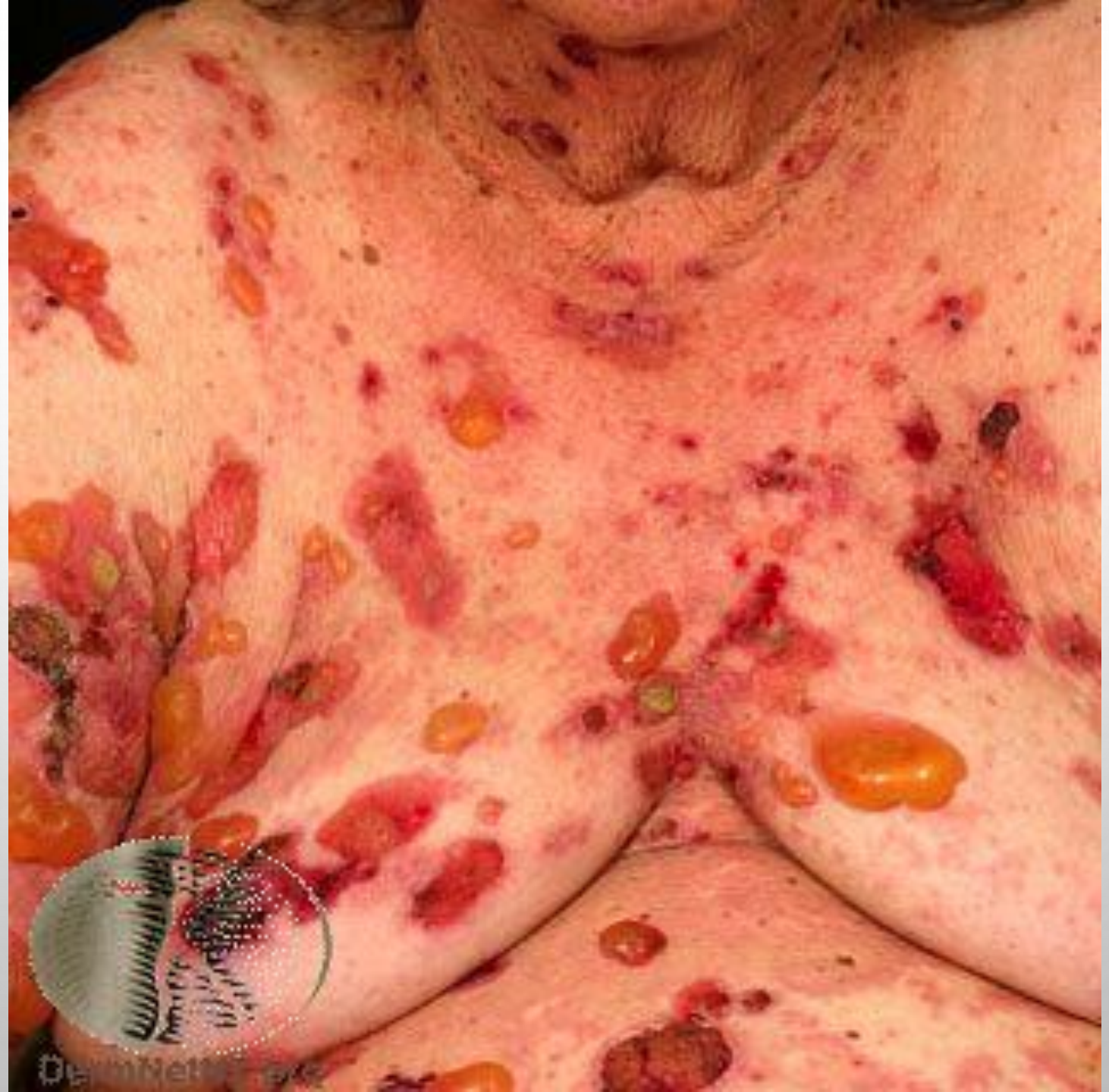
Life long gluten-free diet. Many patients can significantly reduce the requirement for dapsone and are thought to minimise their risk of small bowel lymphoma.

87 YEAR OLD MAN

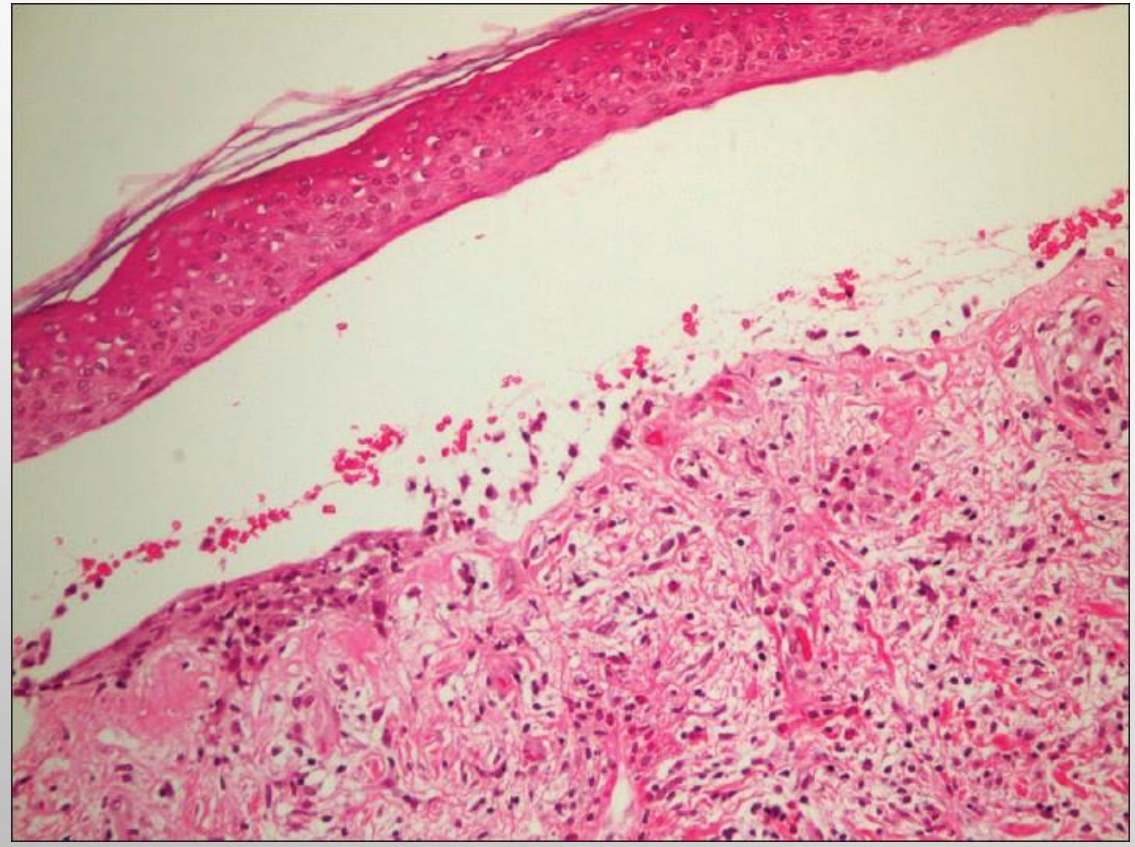
- INTACT BLISTERS ON TORSO AND LIMBS
- SPARES ORAL MUCOSA AND FACE
- DIF SHOWS PERILESIONAL C3 AND IGG AT DERMAL-EPIDERMAL JUNCTION
  - WHICH OF THE FOLLOWING WOULD BE THE MOST APPROPRIATE INITIAL TREATMENT?
    - A. AZATHIOPRINE 2MG/KG/DAY
    - B. TOPICAL HIGH POTENCY CORTICOSTEROIDS
    - C. ORAL PREDNISONE 40MG/DAY
    - D. PLASMAPHERESIS FIVE TIMES OVER TWO WEEKS
    - E. IV CYCLOPHOSPHAMIDE 0.7G/M^2 BODY SURFACE AREA

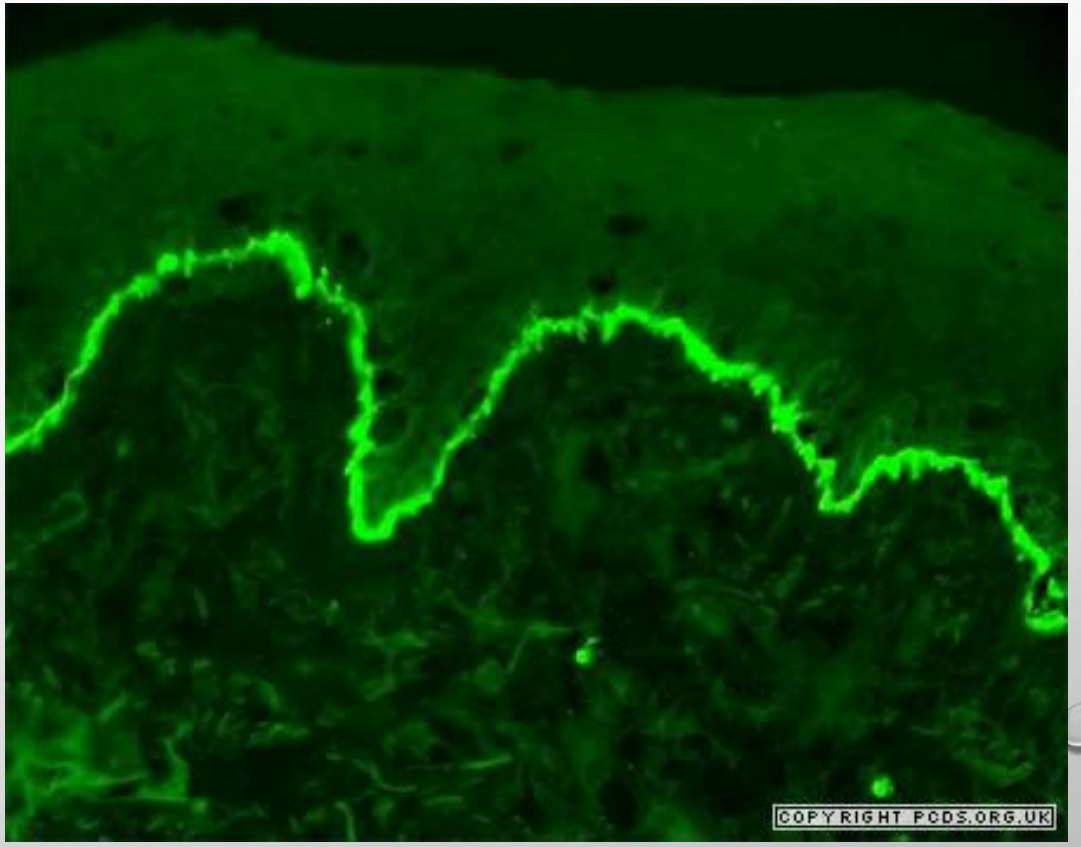
BULLOUS PEMPHIGOID

- Autoimmune blistering disease
- Elderly >60 years
- Itchy plaques → intact blisters
- Spares mucosal membranes
- Pathology
  - Autoantibodies against components of basement membrane: BP180, BP230
  - Sub-epidermal blistering
  - Linear deposition of IgG & C3 along BM



BULLOUS PEMPHIGOID



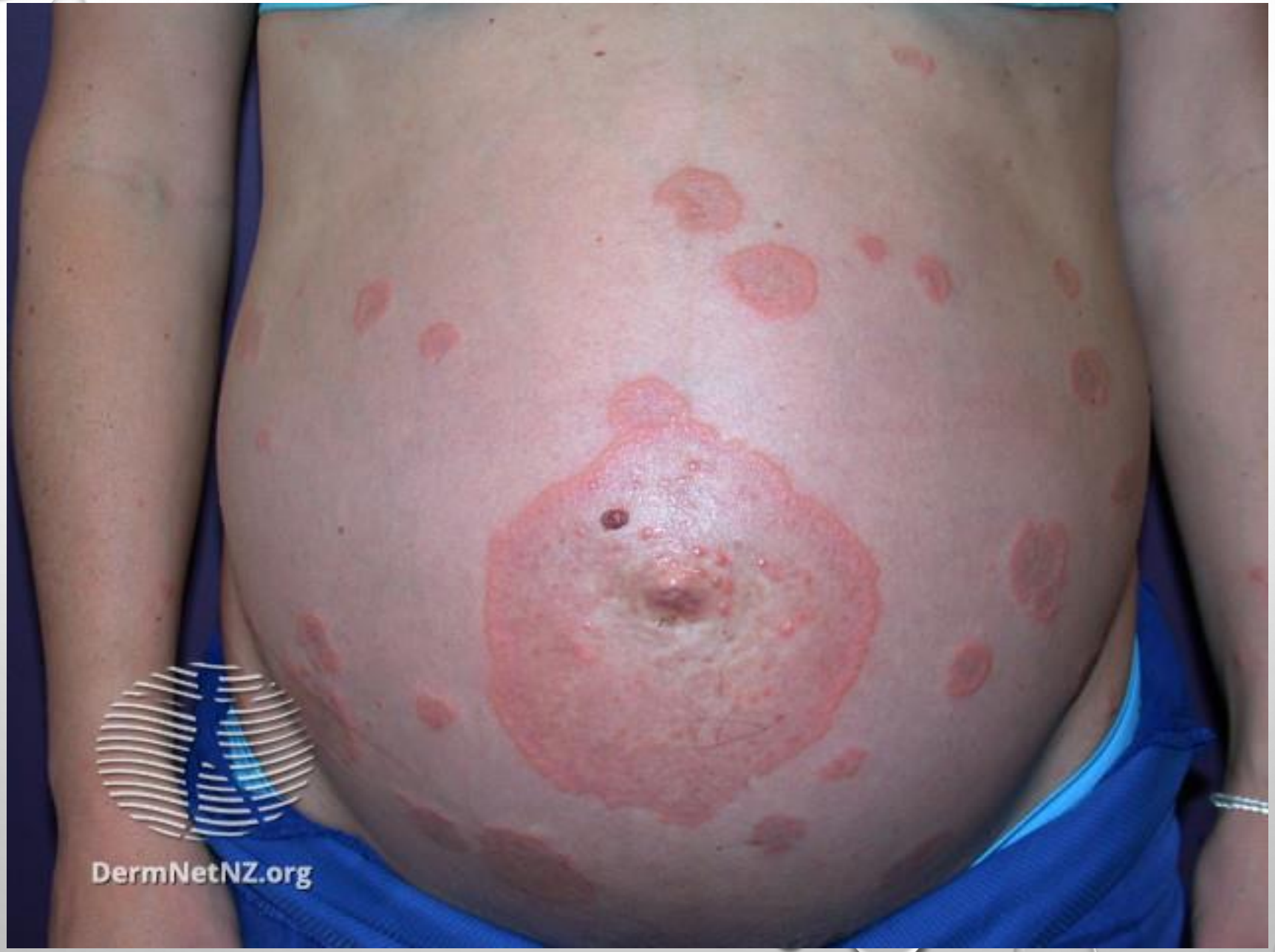


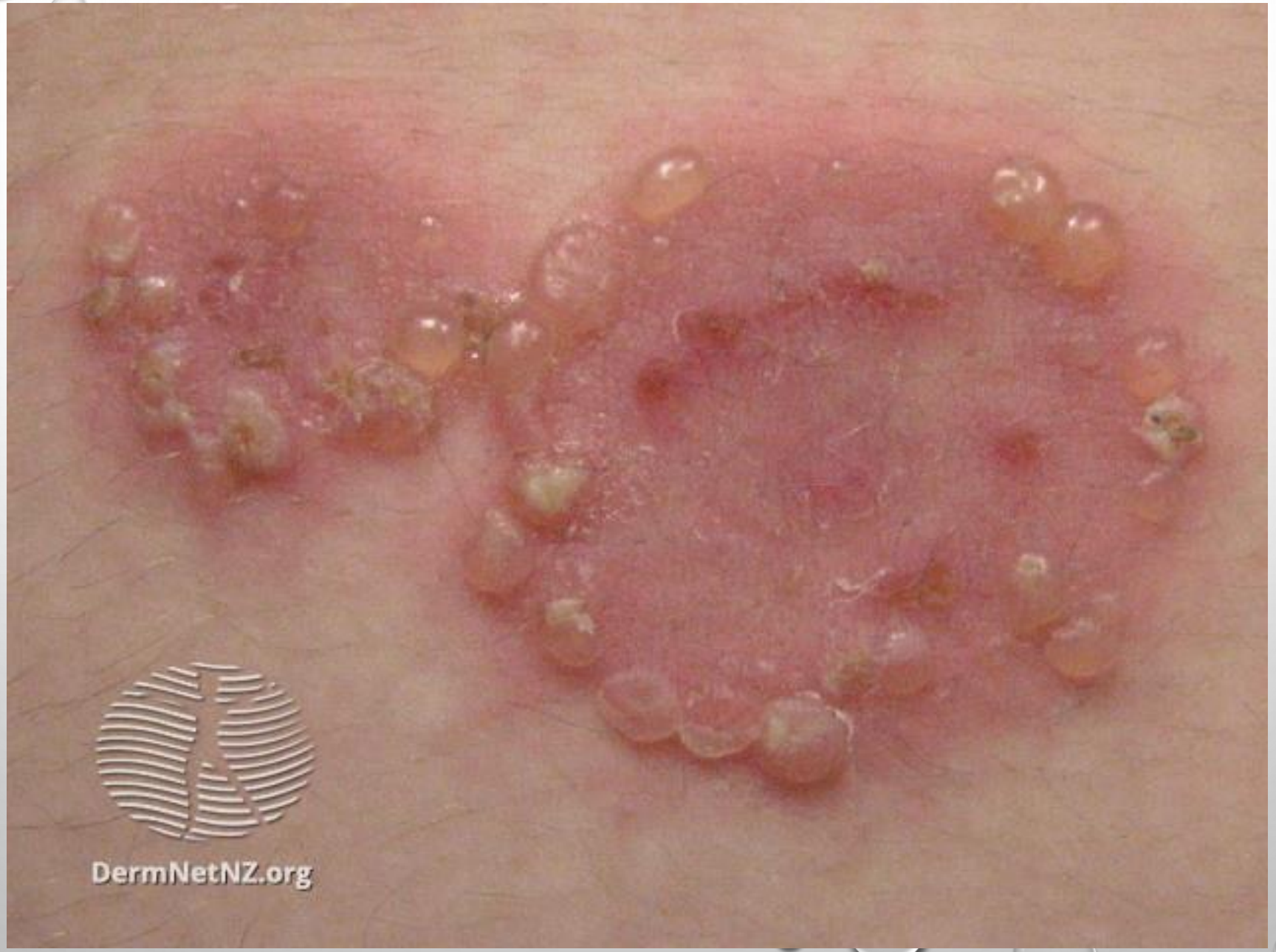
IMMUNOBULLOUS DISEASE

- Treatment
  - Oral corticosteroids ~40mg initially
  - +/- Steroid sparing agent

PEMPHIGOID GESTATIONIS

- Rare pregnancy associated autoimmune skin disease
- Itchy rash developing into blisters
- Most common during 2nd & 3rd trimester
- Symptoms may lessen or spontaneously resolve towards end of pregnancy
- 75-80% of women will experience a flare-up around delivery
- Commencement of menstrual periods, use of OCP or further pregnancies may cause flare-ups

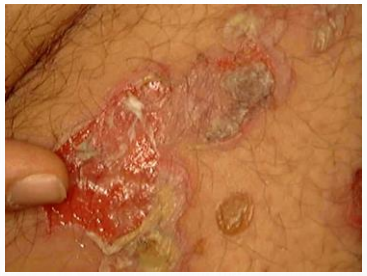

DermNetNZ.org


DermNetNZ.org

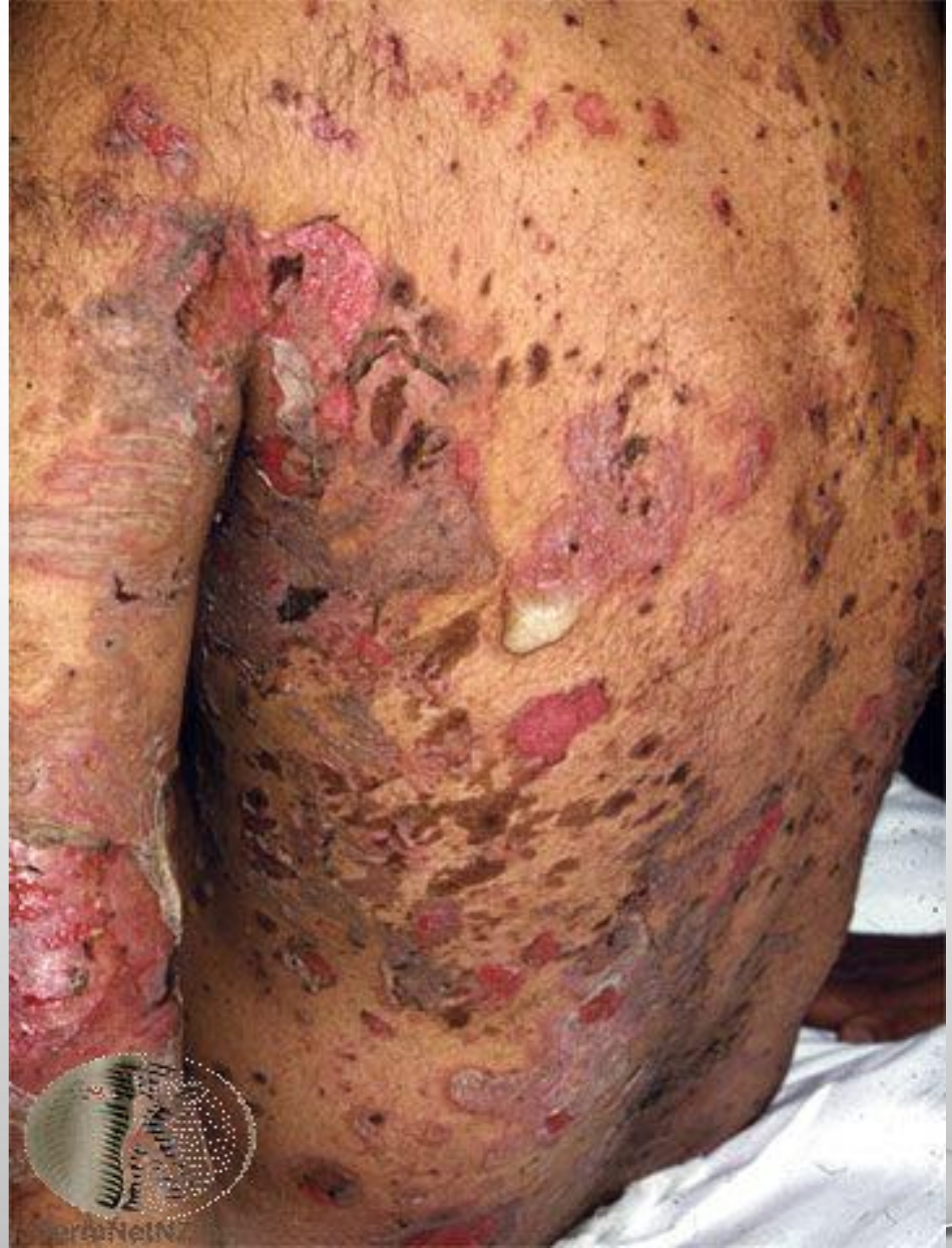
TREATMENT

- Topical corticosteroids in mild disease
- Oral corticosteroids in extensive disease
- Oral antihistamines for itch
- Dapsone may be effective
- Azathioprine or Ciclosporin may be used but safety in pregnancy and breastfeeding must be considered
- Most cases resolves spontaneously within days after delivery

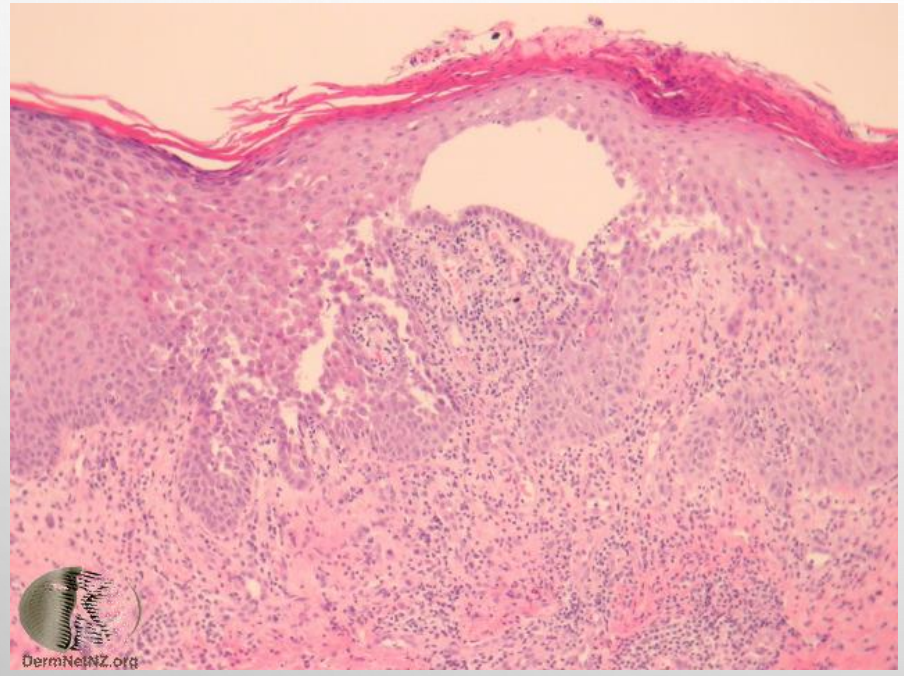
PEMPHIGUS

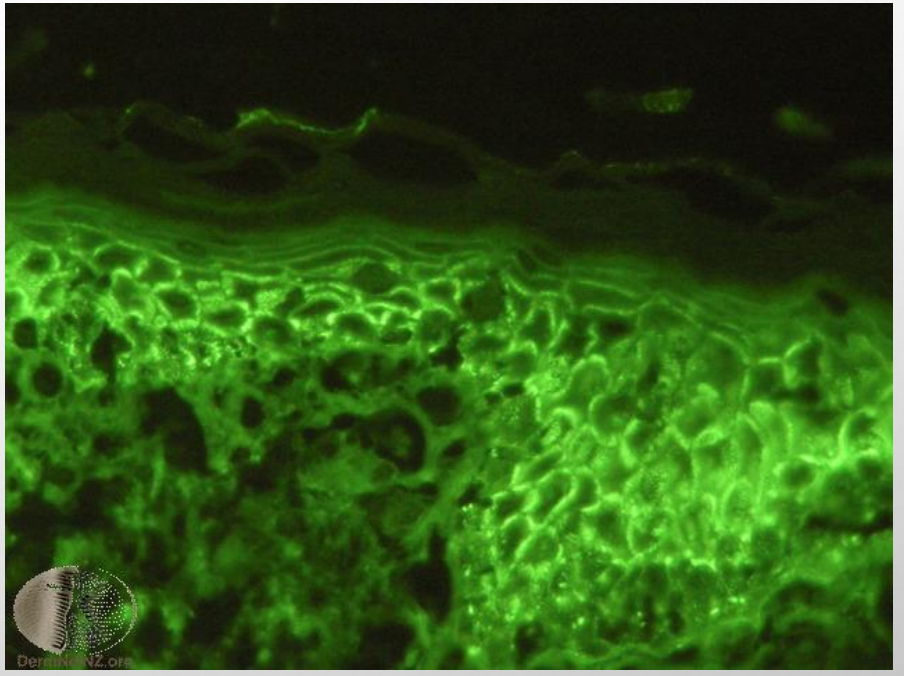


- 2 types
  - Pemphigus vulgaris: mucosal involvement
  - Pemphigus foliaceus: erosions on scalp, trunk
  - Pathology
    - Autoantibodies against intracellular adhesion molecules: Desmoglein 1 & 3
    - Intra-epidermal split → flaccid blisters, erosions
    - DIF: intracellular deposits of IgG, C3



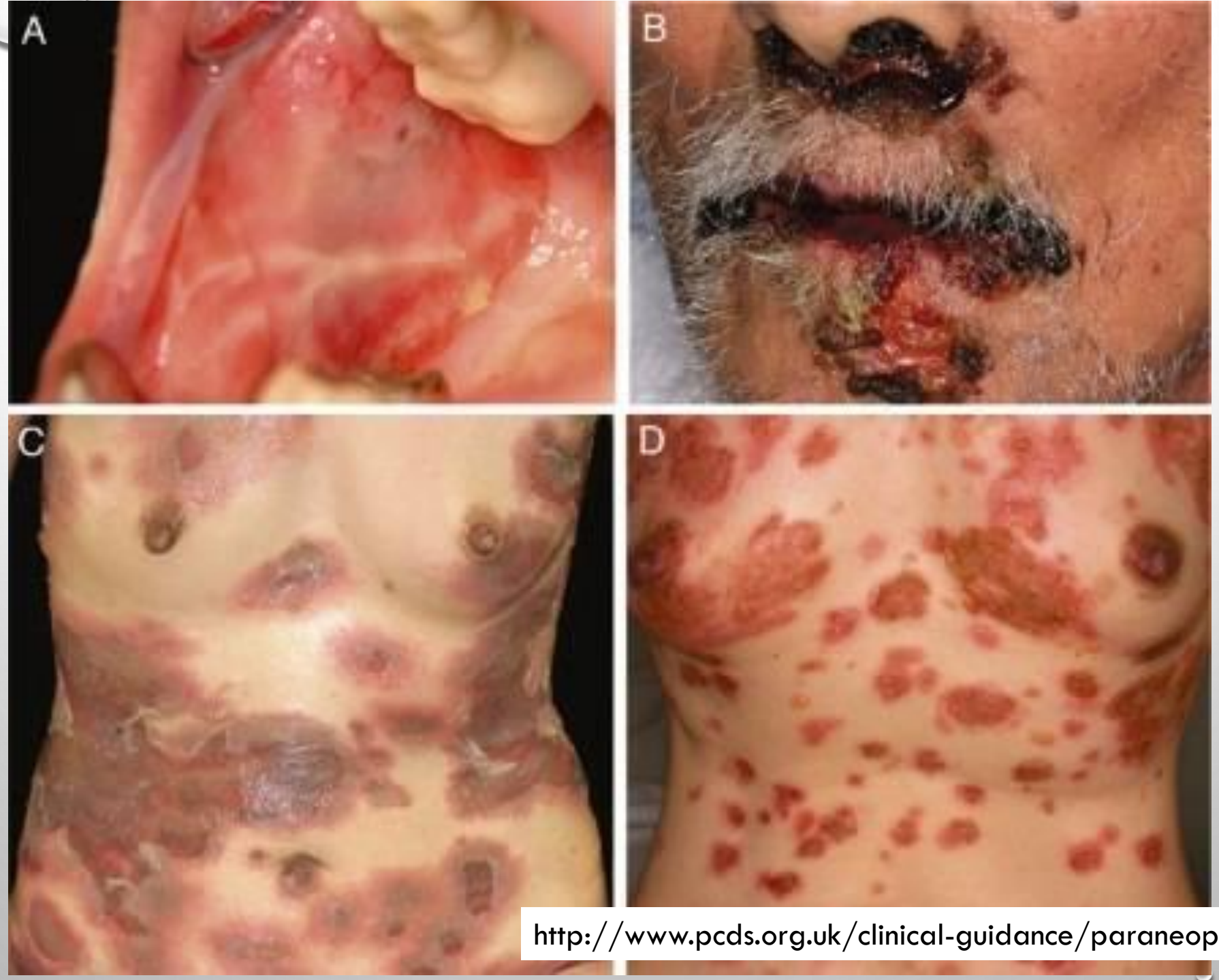
PEMPHIGUS





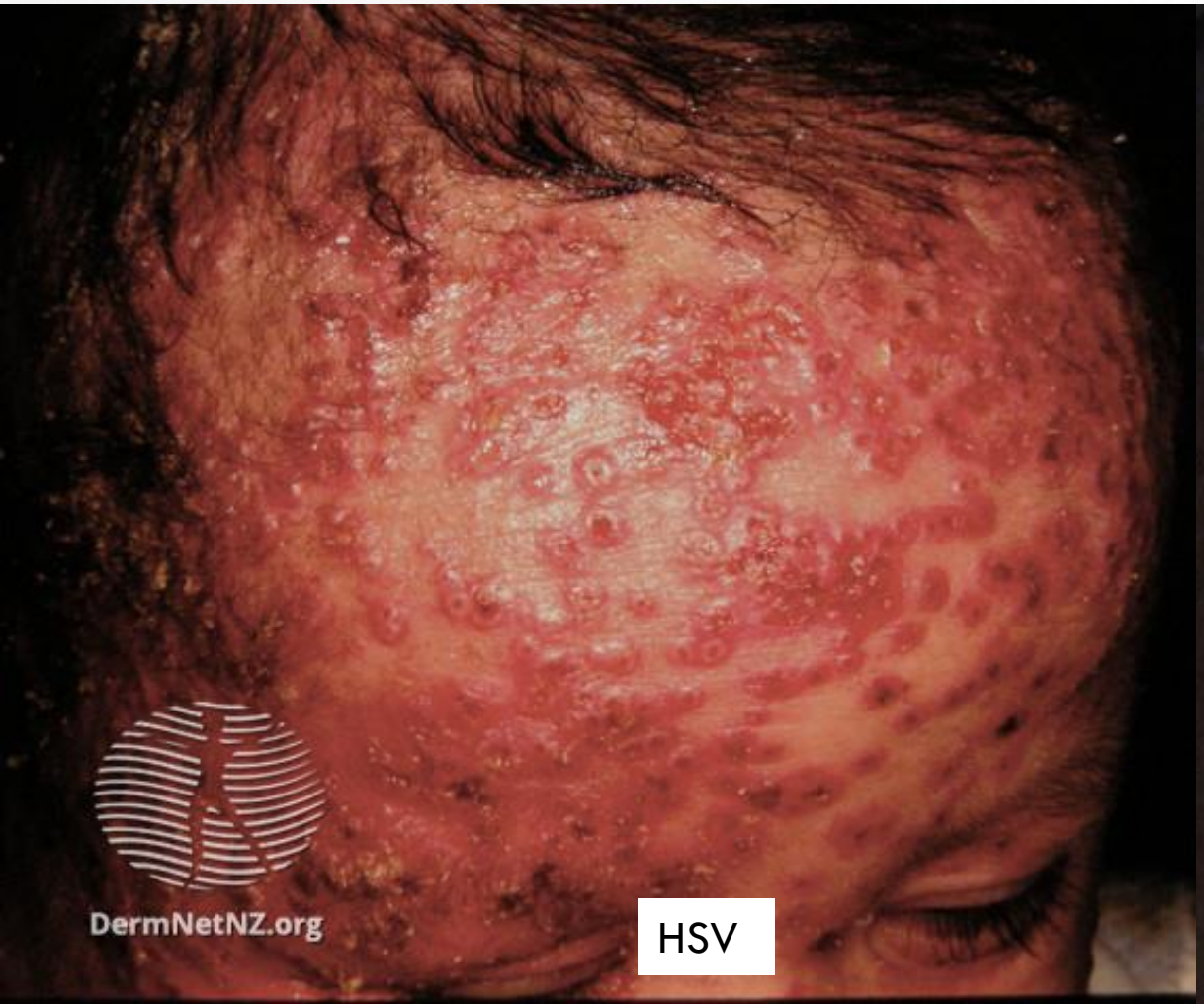
PARANEOPLASTIC PEMPHIGUS

- Rare autoimmune blistering disease affecting skin and/or mucous membranes
- In association with malignancies
- Painful blisters & denuded areas of mouth, lips, oesophagus & skin
- Lease common but most serious form of pemphigus
- Can resolve if cancer cured
- Otherwise largely supportive
- 75-80% die from paraneoplastic pemphigus or underlying cancer

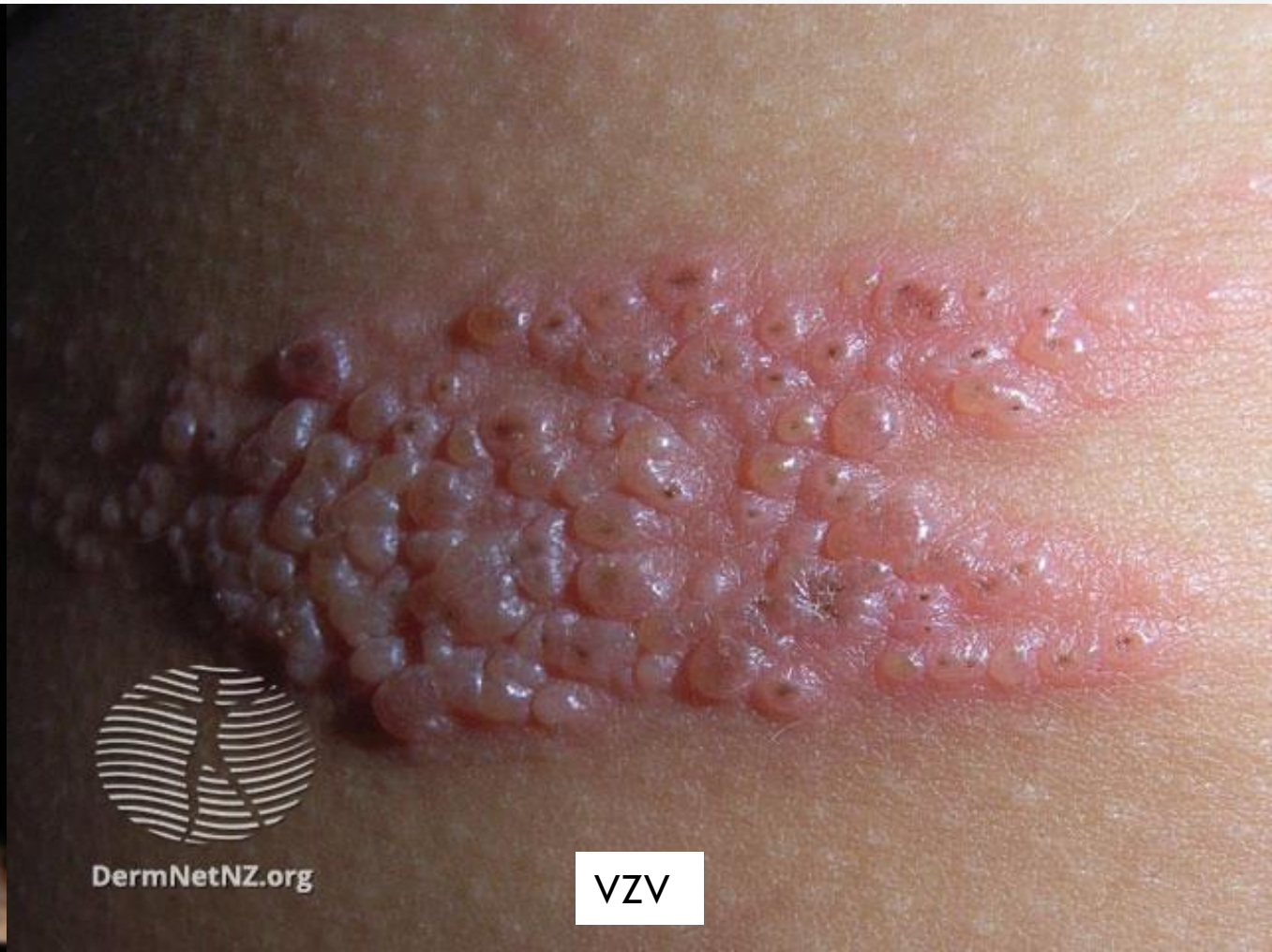


http://www.pcds.org.uk/clinical-guidance/paraneoplastic-pemphigus

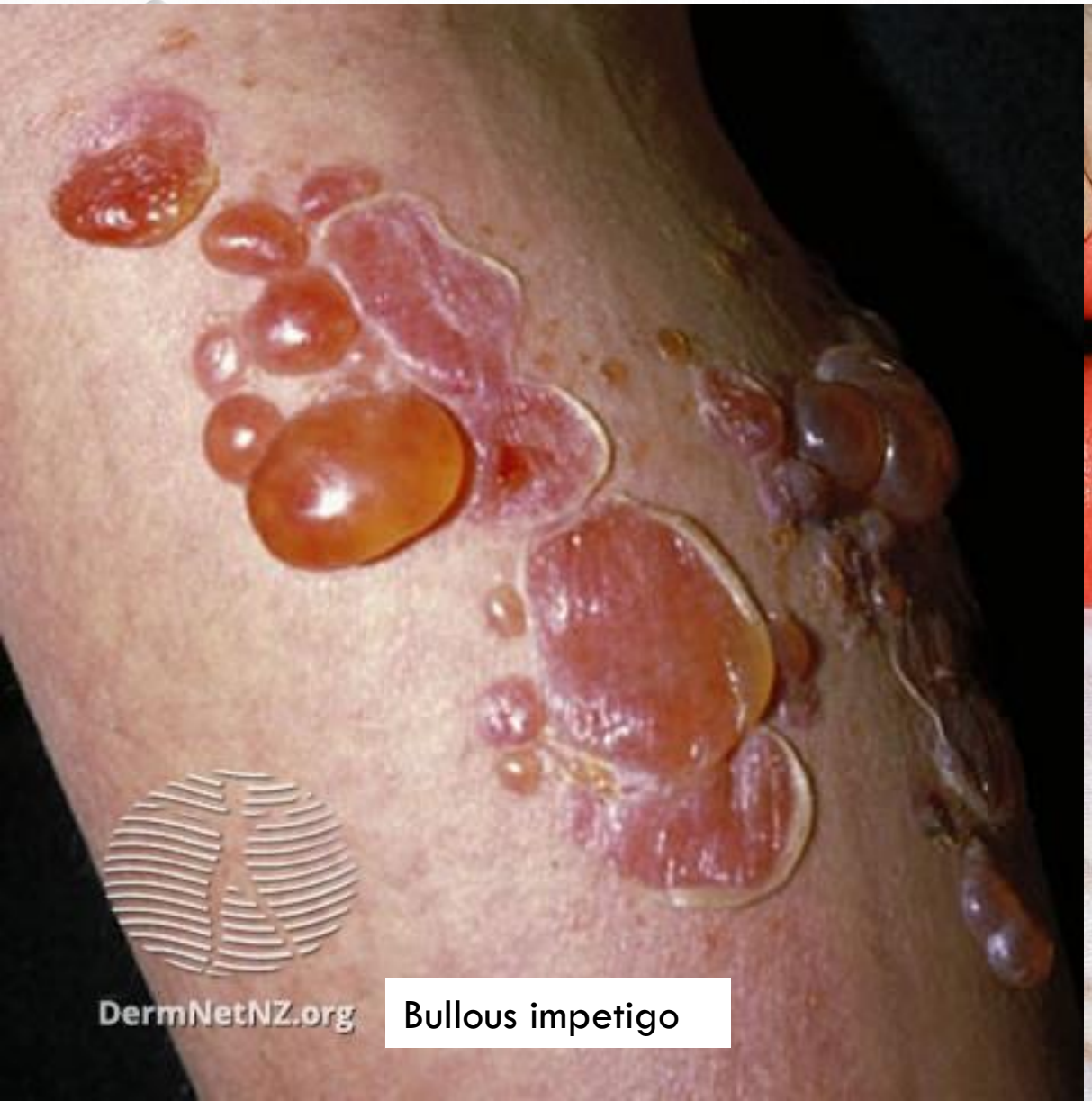
INFECTIOUS



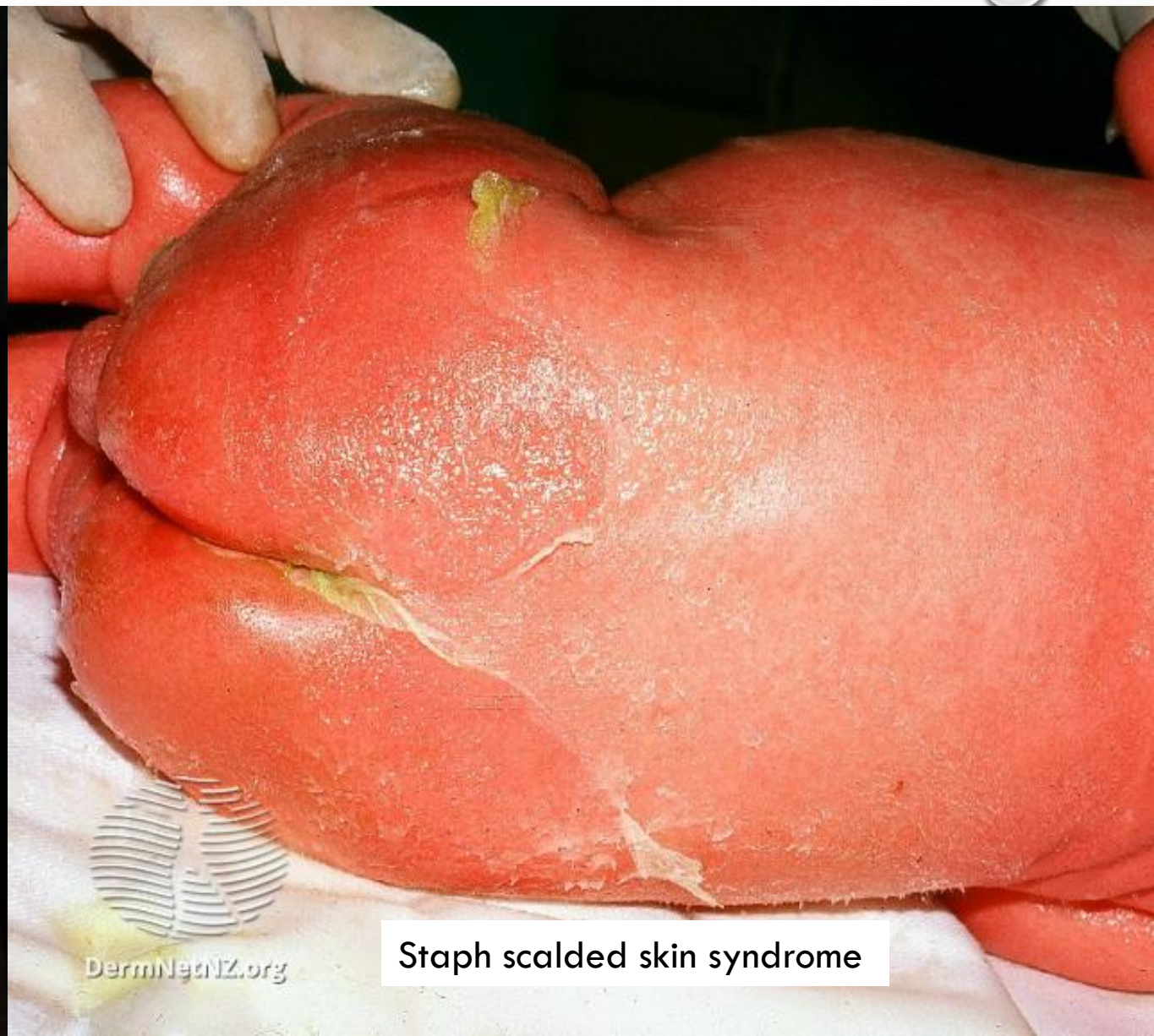
HSV



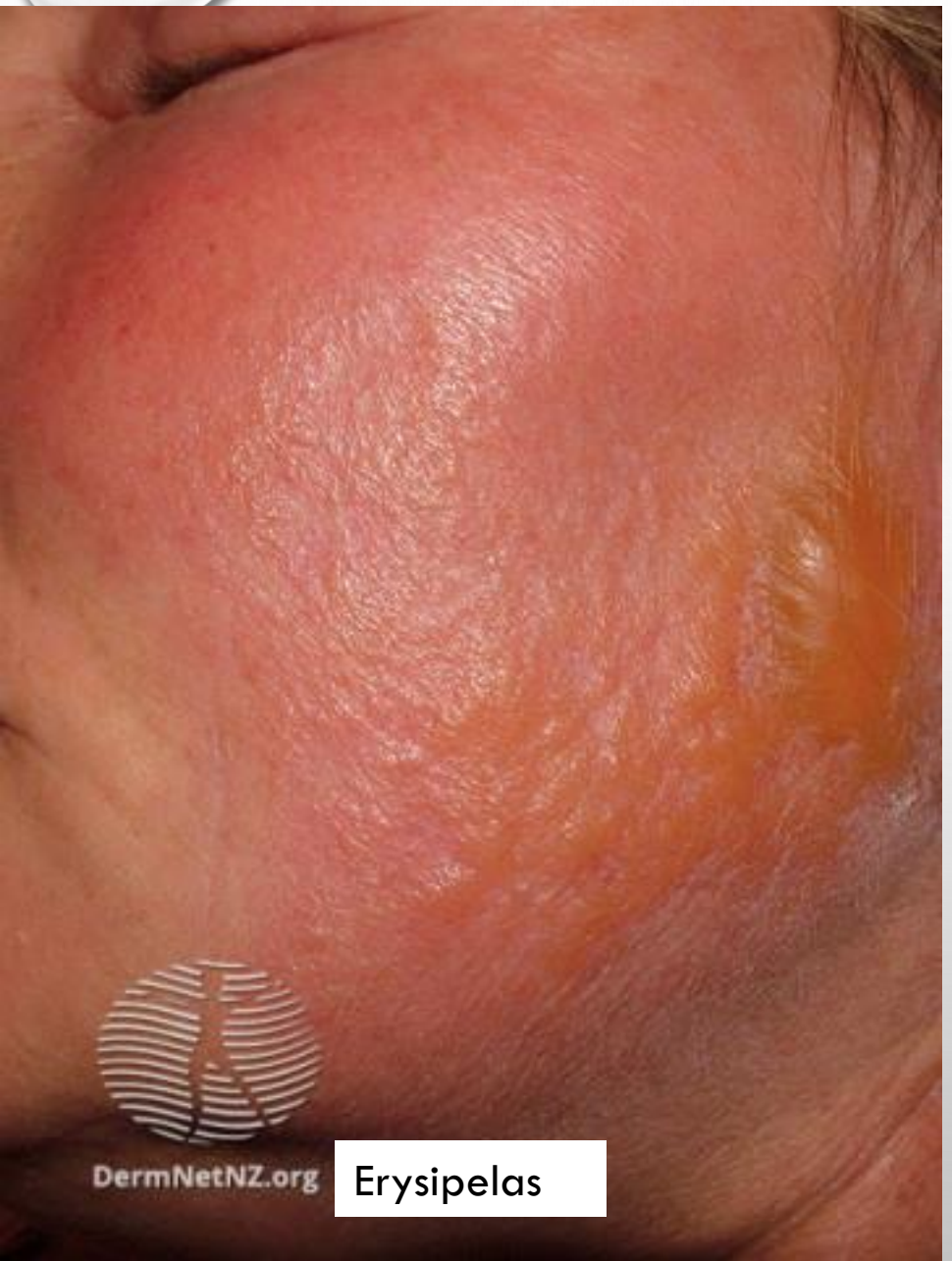
VZV



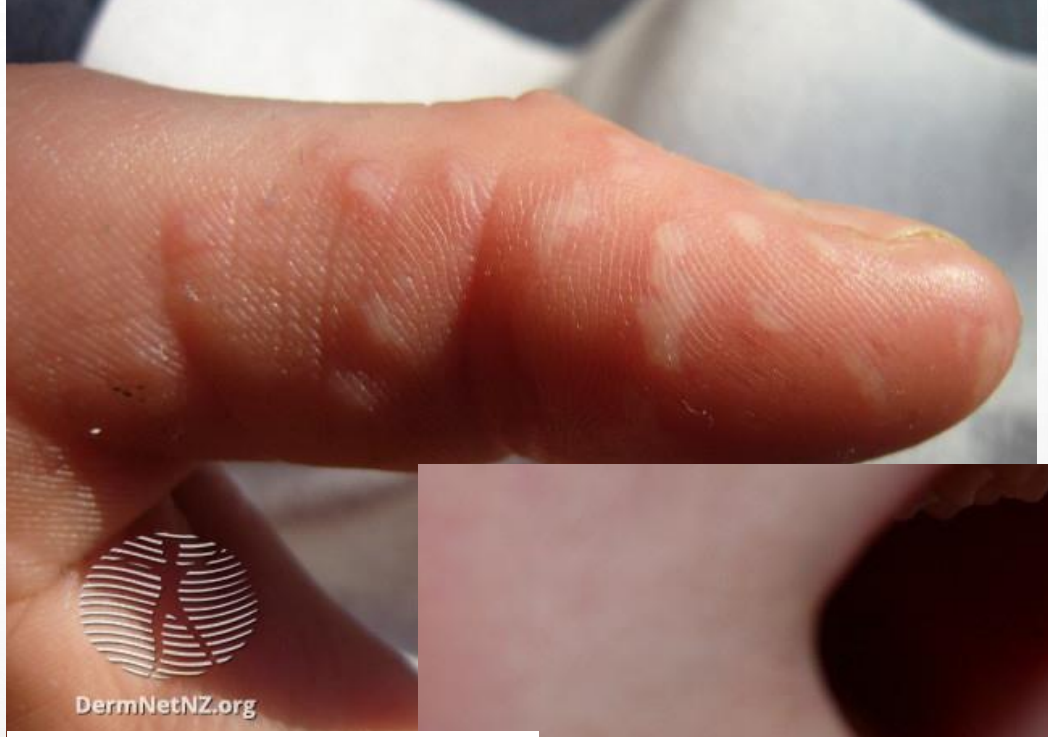
Bullous impetigo

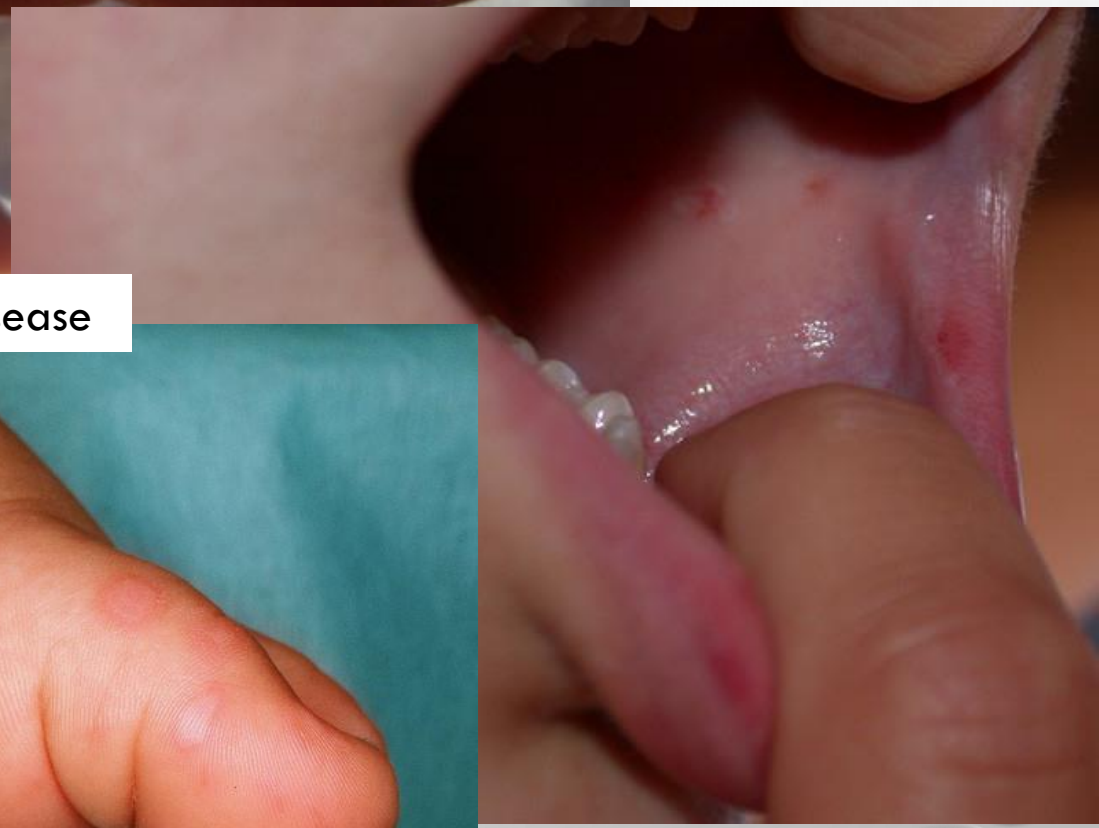


Staph scalded skin syndrome



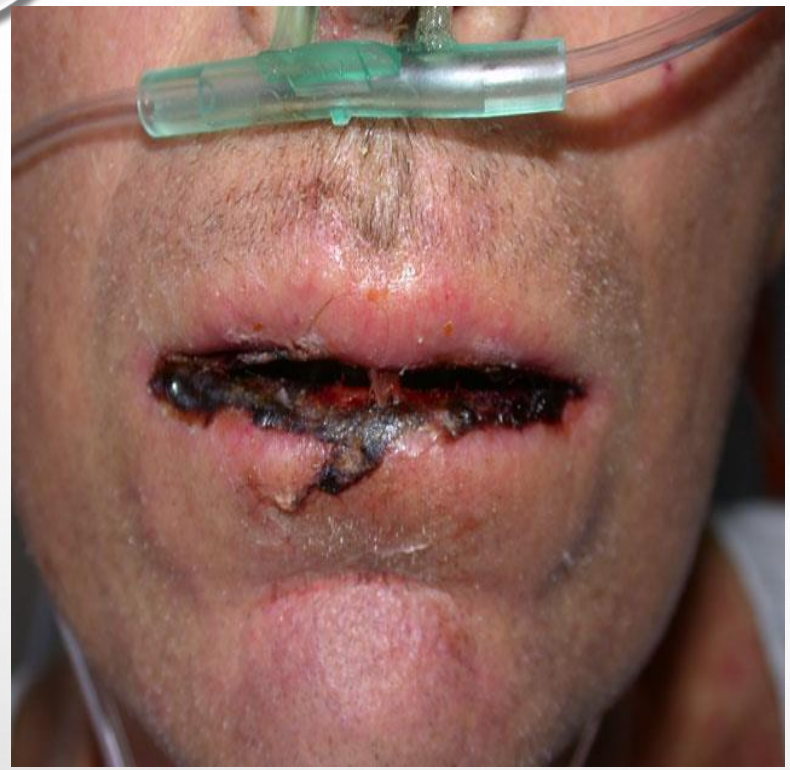
Erysipelas

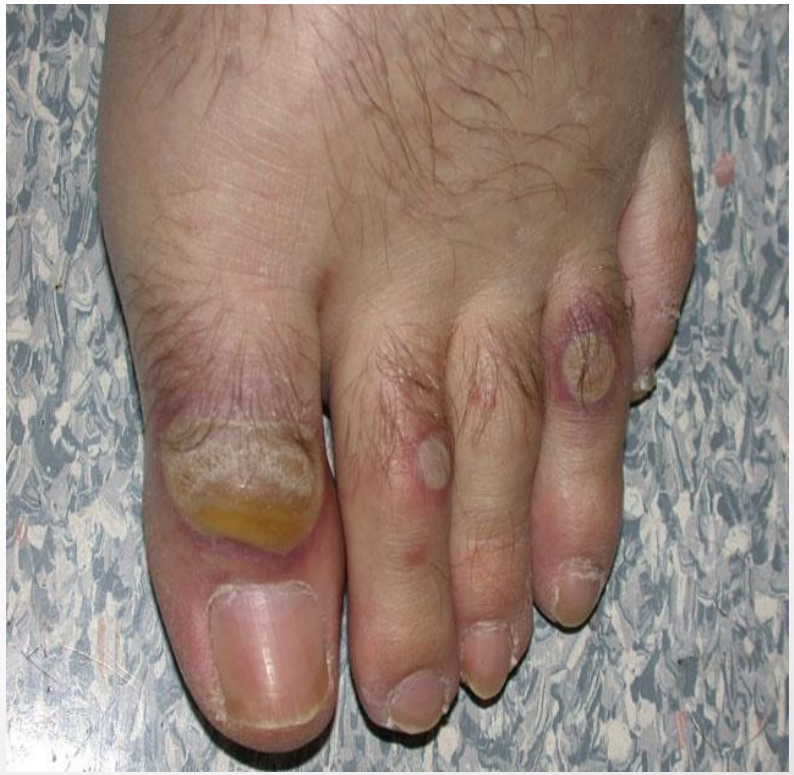




Hand foot mouth disease

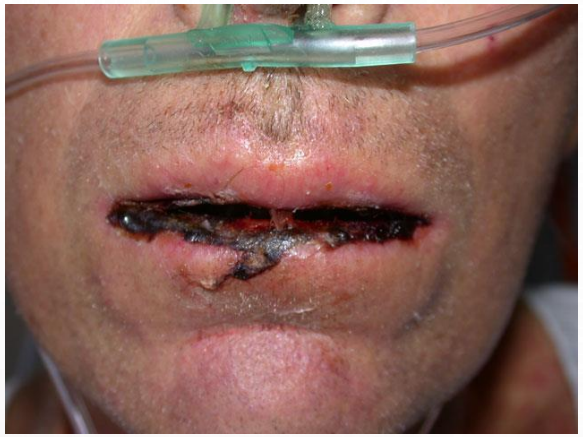
DRUG INDUCED

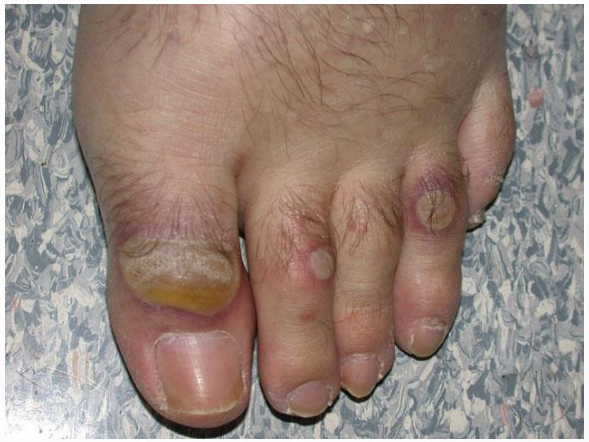




A middle-aged man presents with a fever, prostration, mucocutaneous ulceration and an acute blistering rash mainly affecting his feet.

What is the name for this syndrome?



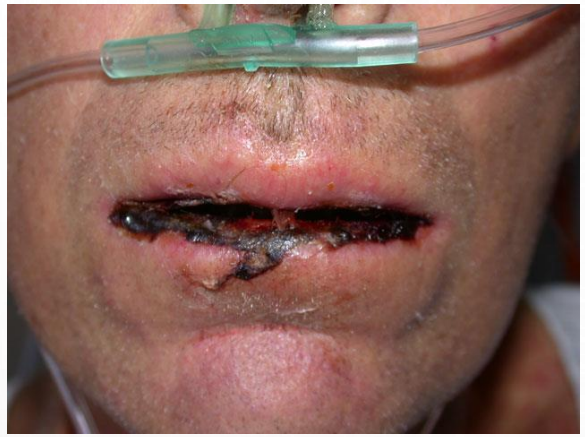


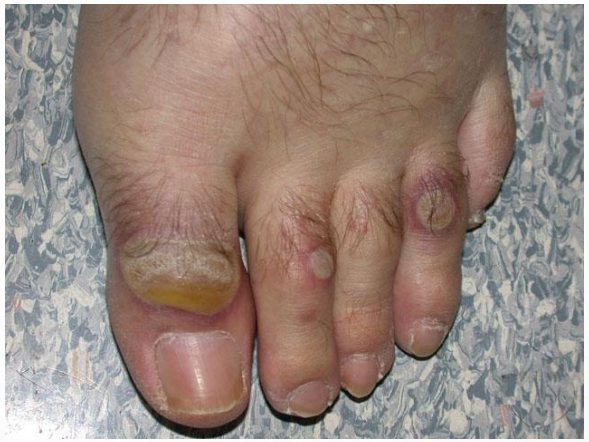
What is the name for this syndrome?

Steven's Johnson syndrome (aka Erythema Multiforme major)

Predominantly a mucosal eruption of erosions and blisters in the oropharynx, on the lips, conjunctivae and genitalia accompanied by fever and prostration

Generally a few target lesions on the distal limbs, hands and feet and these are sometimes bullous



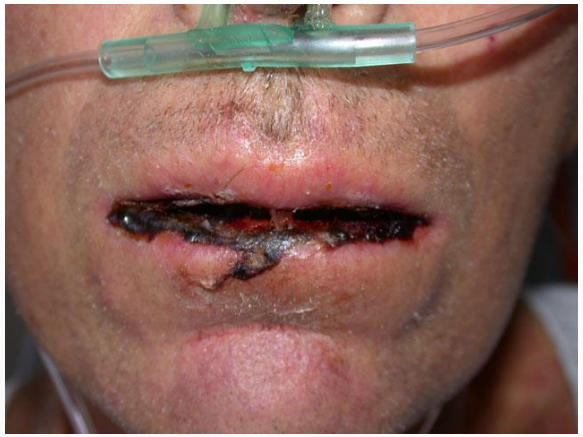


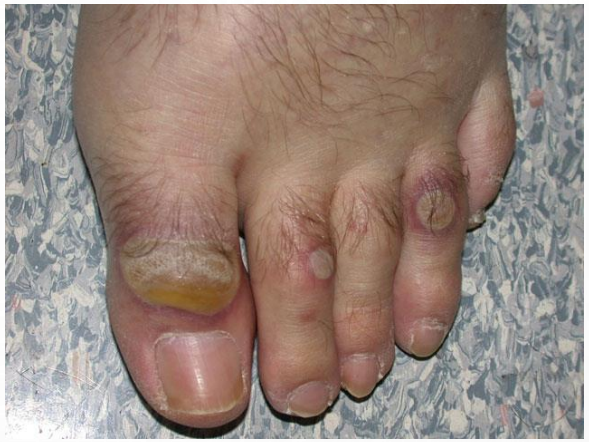
What are the possible causes?

Usually a drug eruption especially to sulphonamides, anticonvulsants, allopurinol and antibiotics prescribed within the previous 3 weeks (rarely up to 8 weeks).

More common in patients with immunodeficiency especially due to HIV.

Contrast this with EM minor, which is usually a reaction to viral infection, especially HSV and Orf. Mycoplasma infection may also provoke EM with mucosal lesions, especially in children.





How would you treat him?

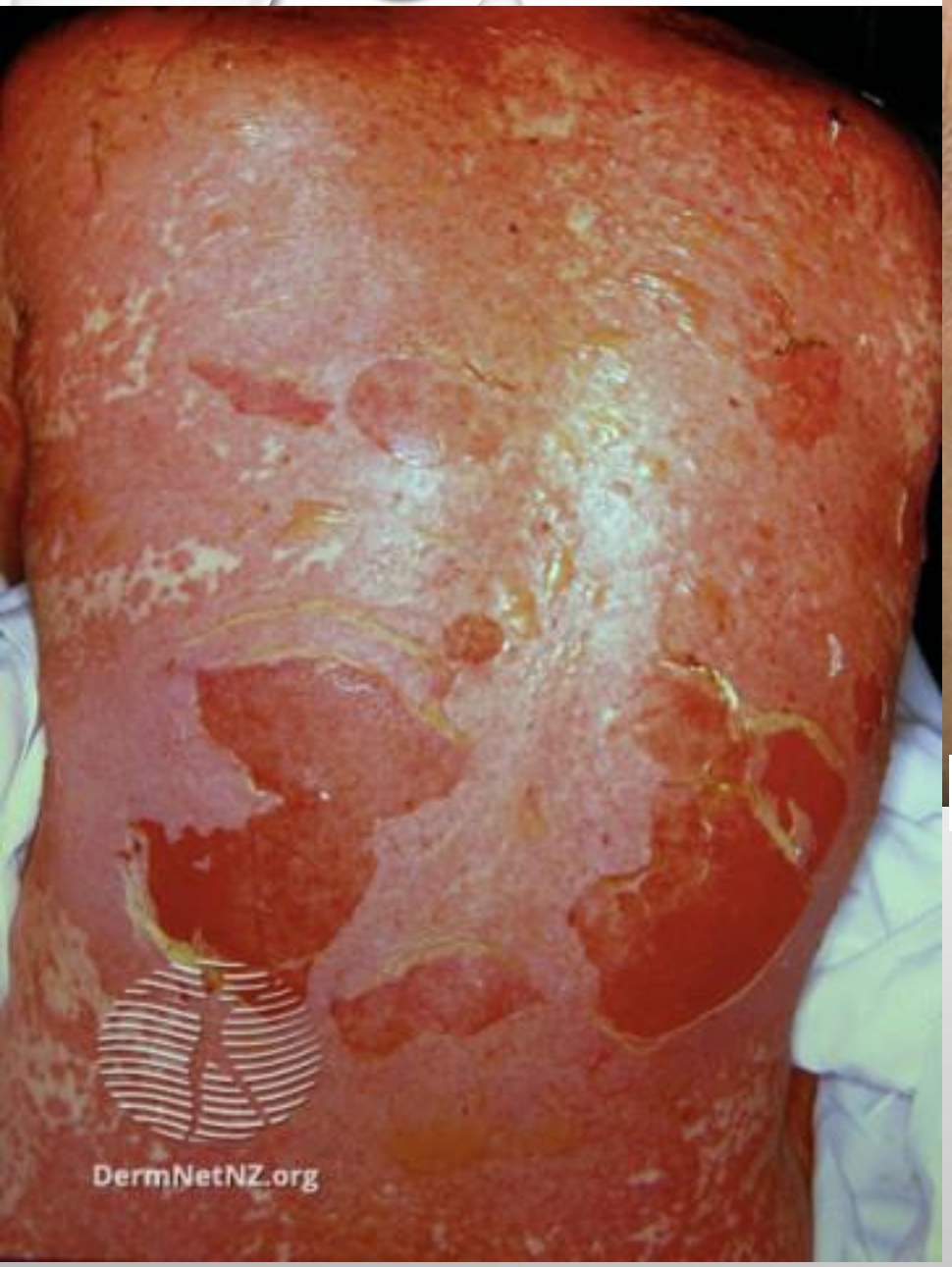
Stop responsible drug

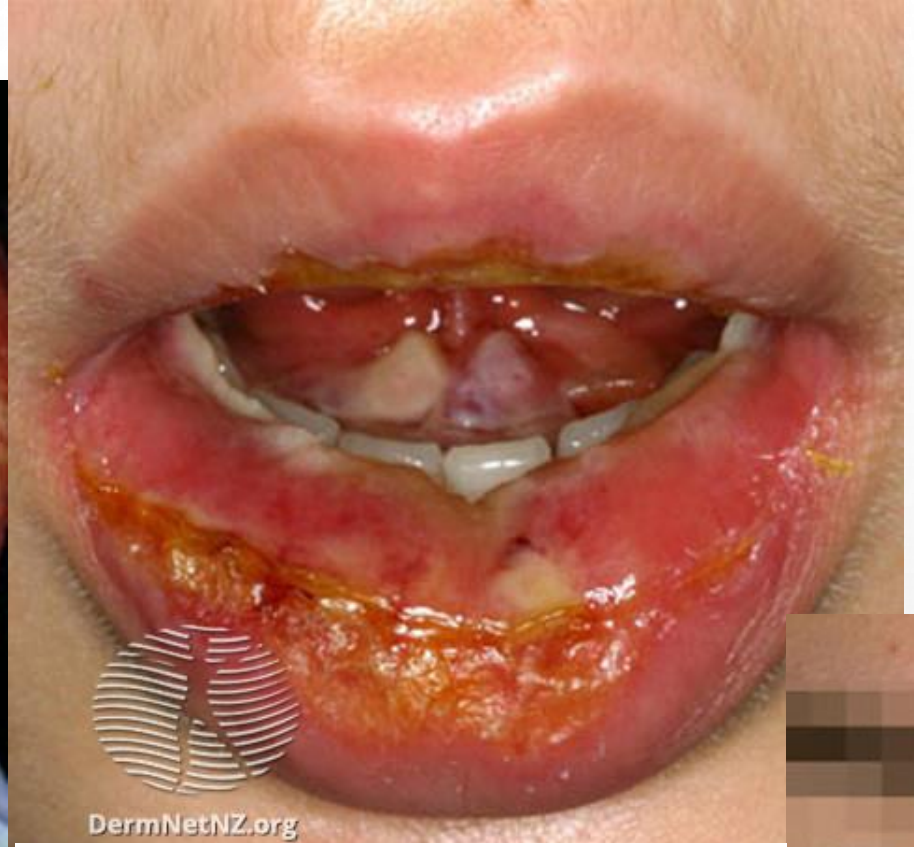
Admission for observation and supportive care (fluid replacement, wound/mouth/eye care, prevent/treat infection)

Oral corticosteroids are avoided in most situations

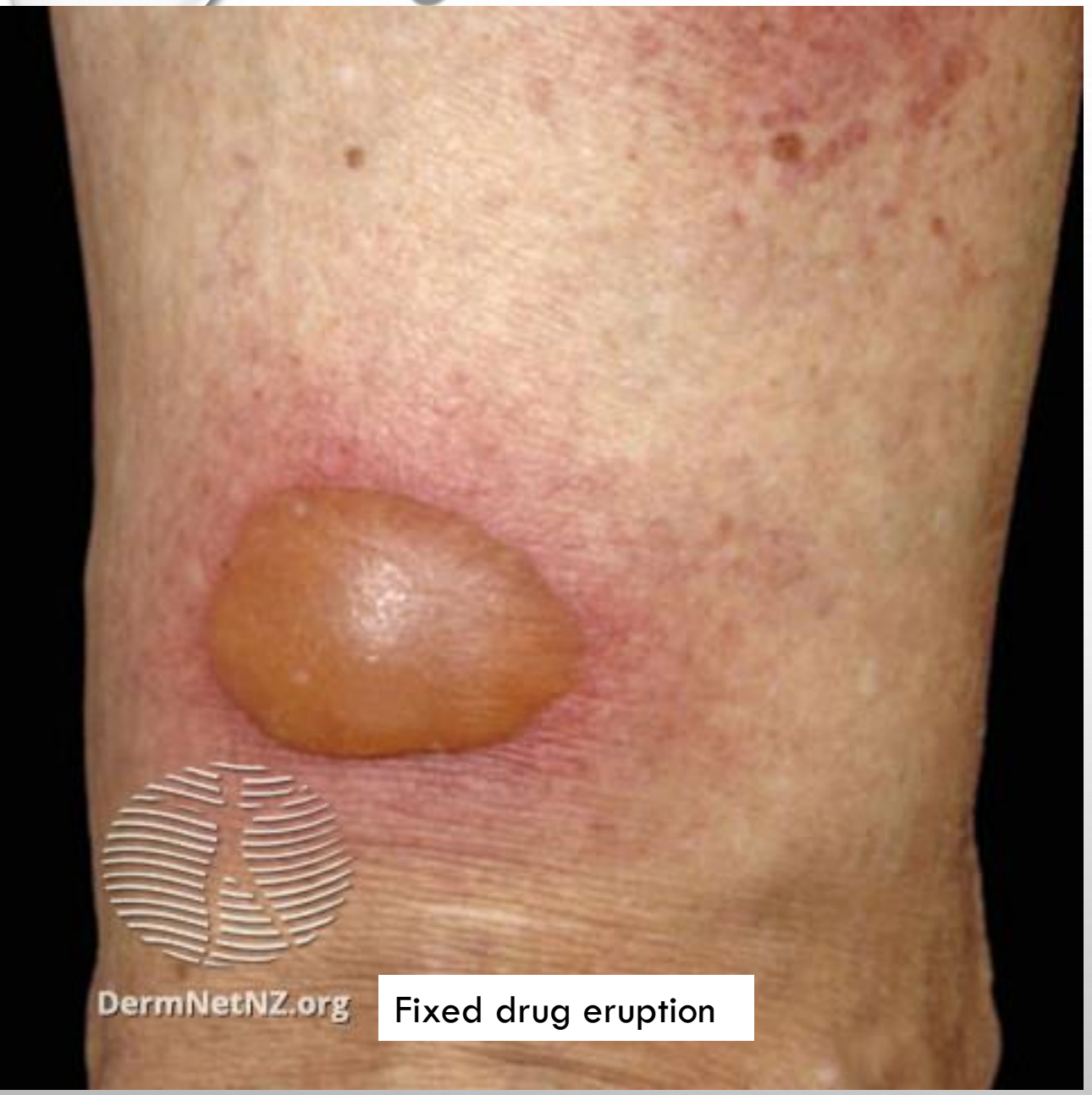
Spontaneous recovery usually occurs within one to three weeks but the occasional case is fatal. Permanent mucosal scarring possible

Toxic epidermal necrolysis (TEN) presents in the same way as EM major but is soon followed by widespread blistering and skin/mucosal shedding. Rx CsA or IVIG. 30% Mortality





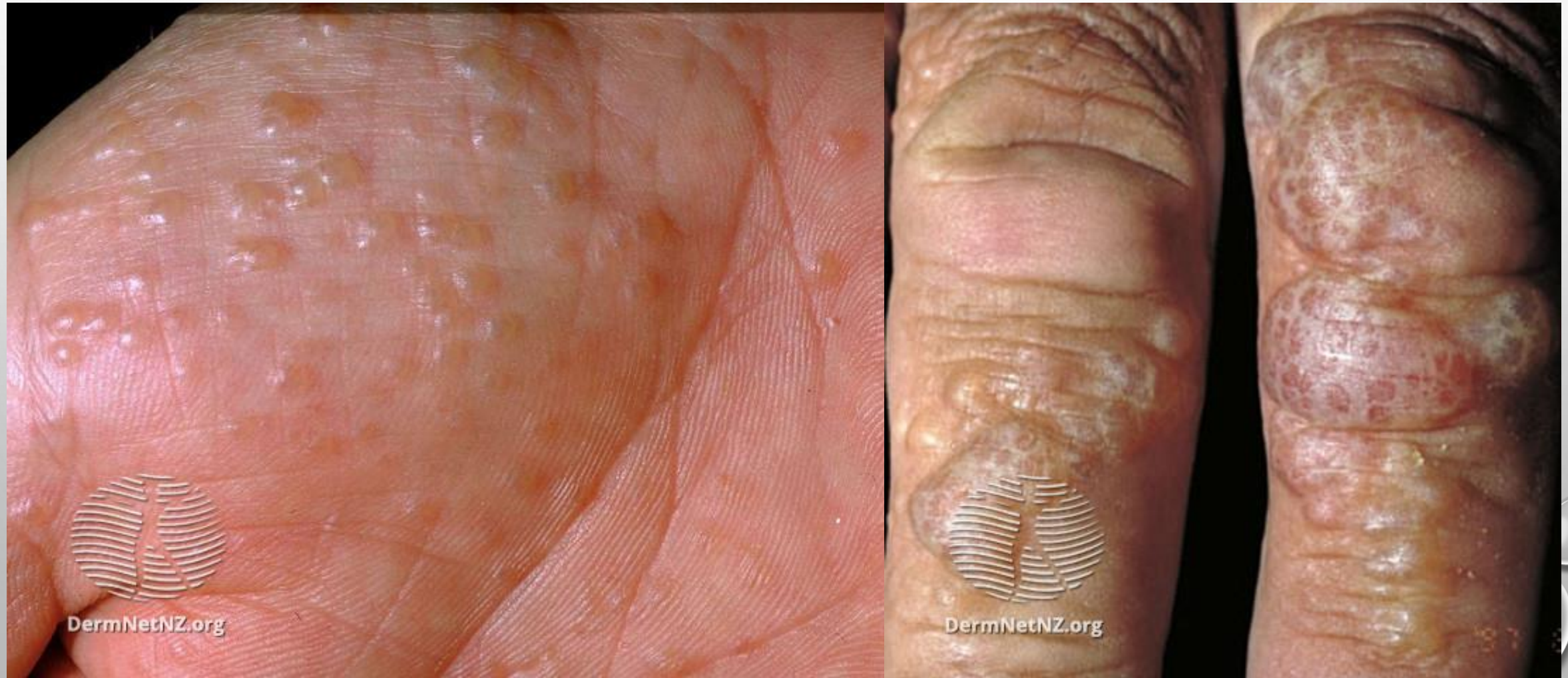
Toxic epidermal necrolysis / TEN



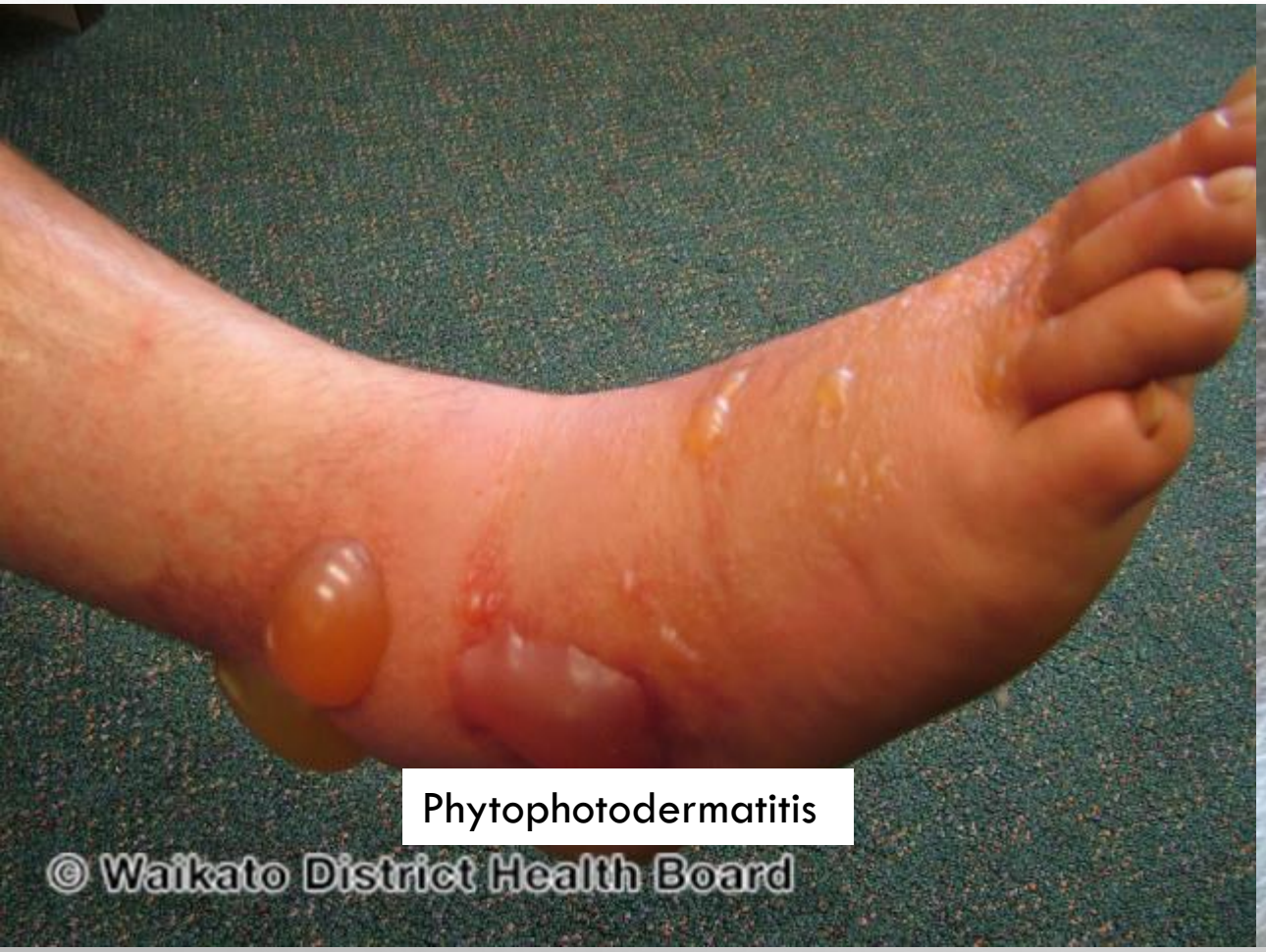
Fixed drug eruption

DERMATITIS

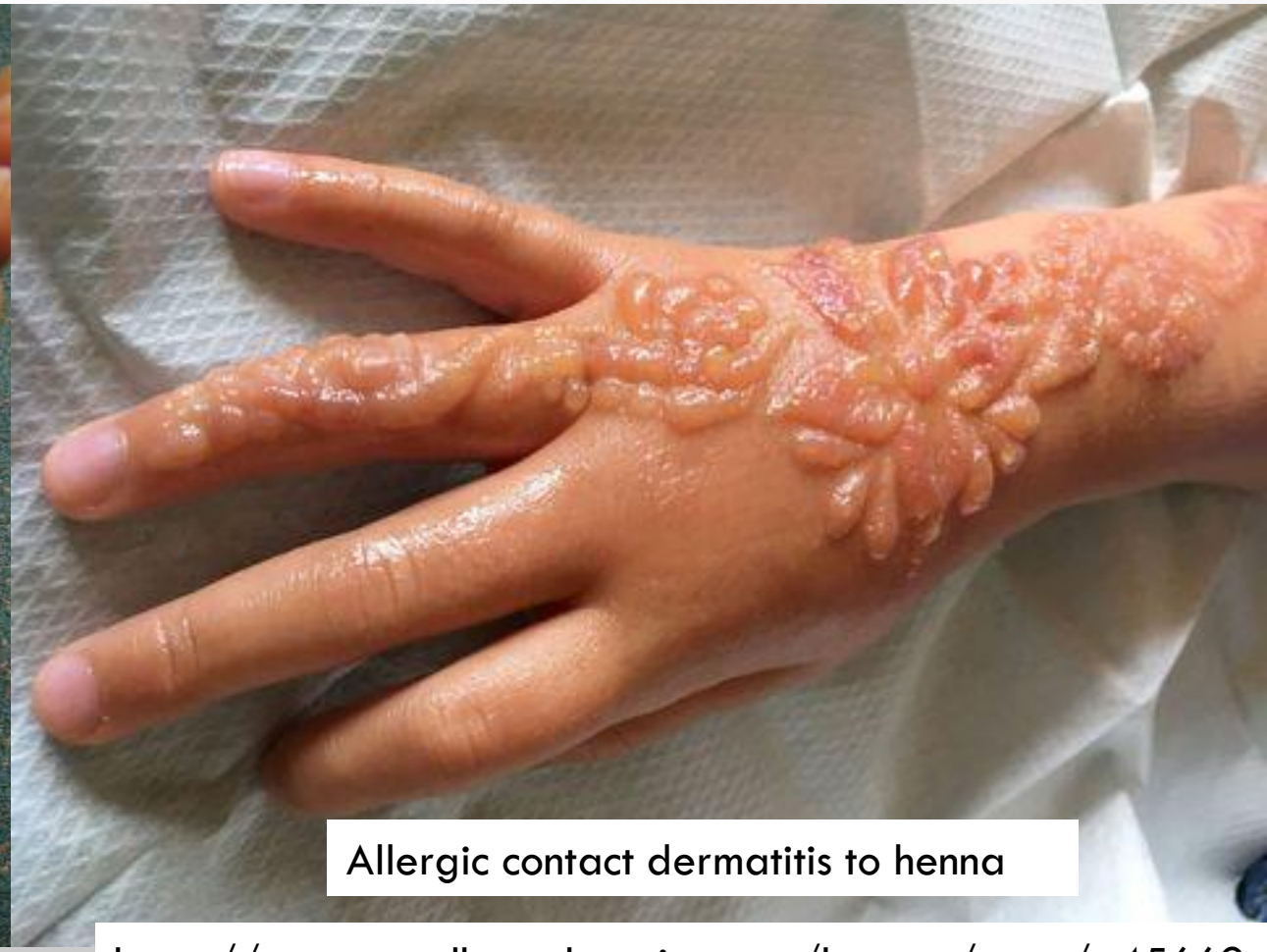
POMPHOLYX / VESICULAR HAND DERMATITIS



ALLERGIC CONTACT DERMATITIS



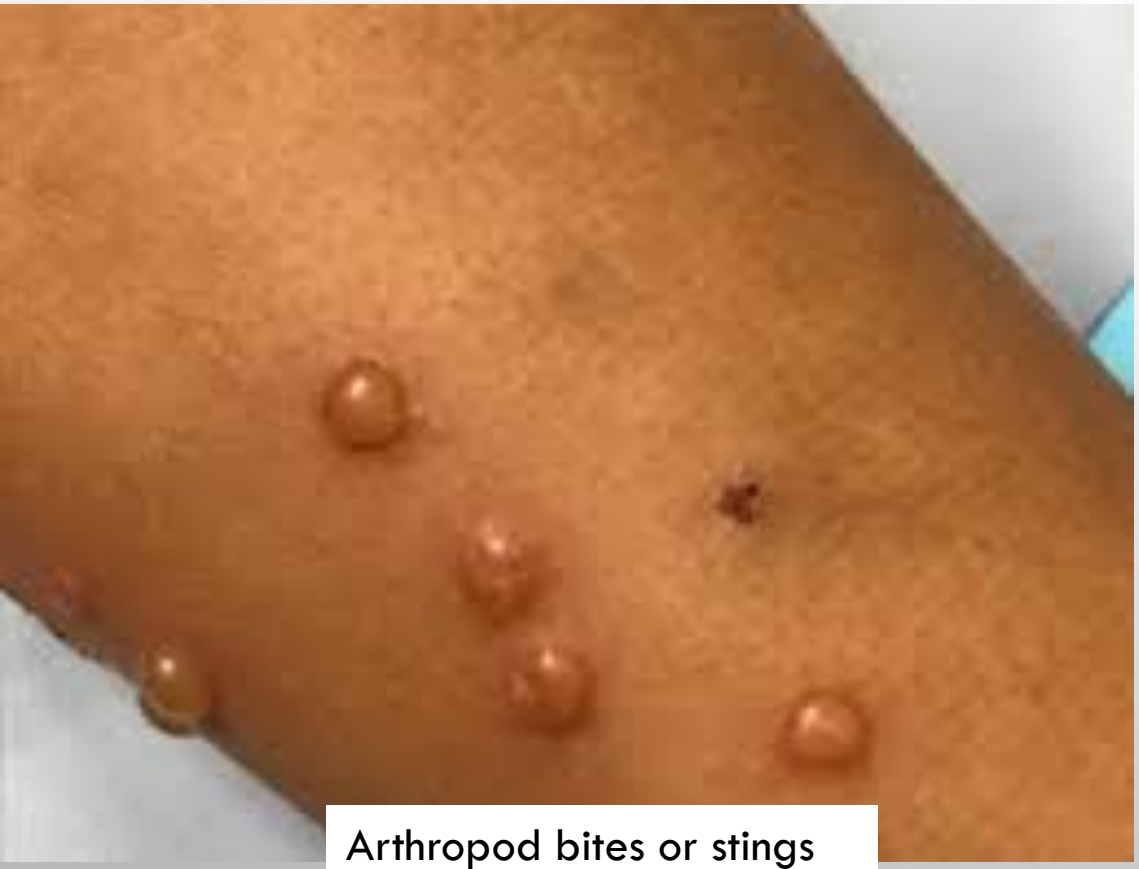
Phytophotodermatitis



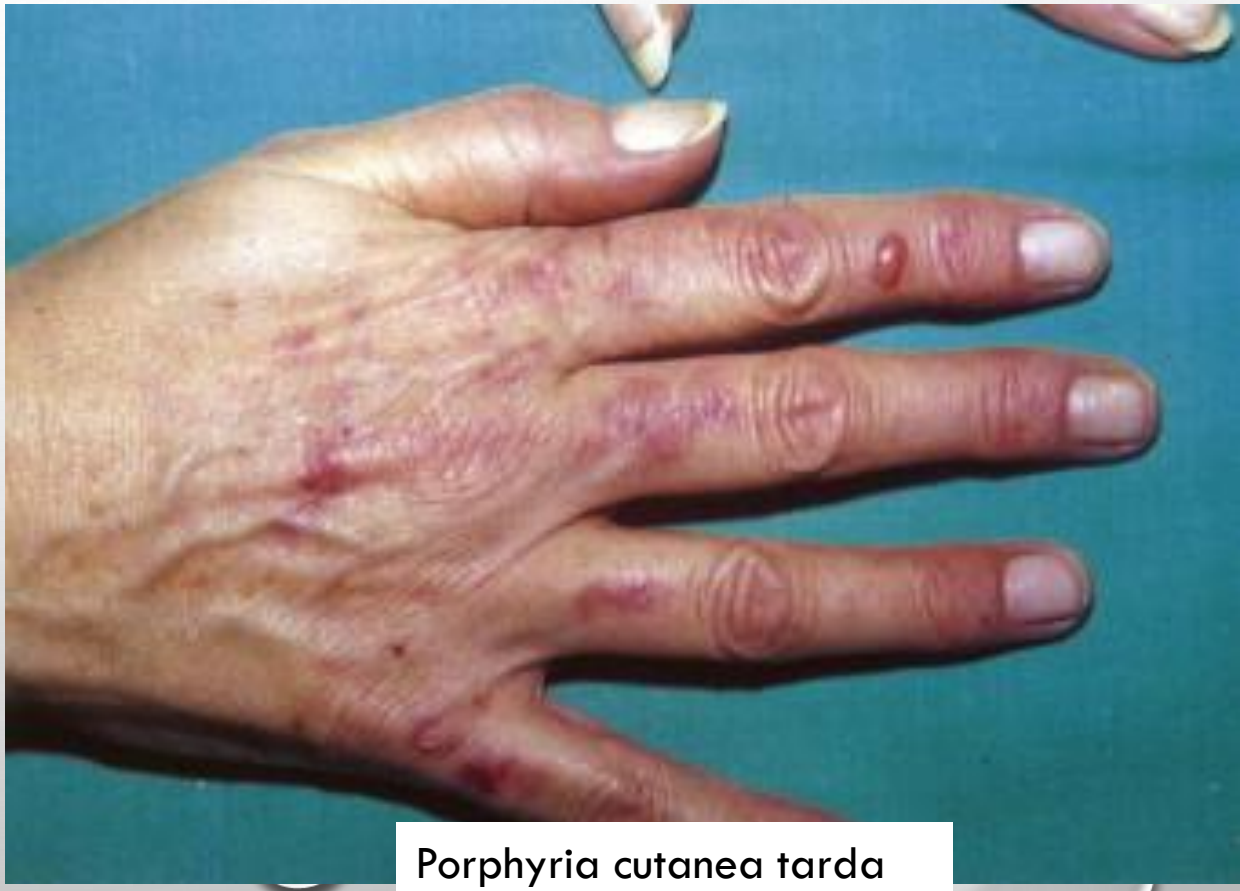
Allergic contact dermatitis to henna

https://www.goodhousekeeping.com/beauty/news/a45669/black-henna-tattoo-scars-little-girl/

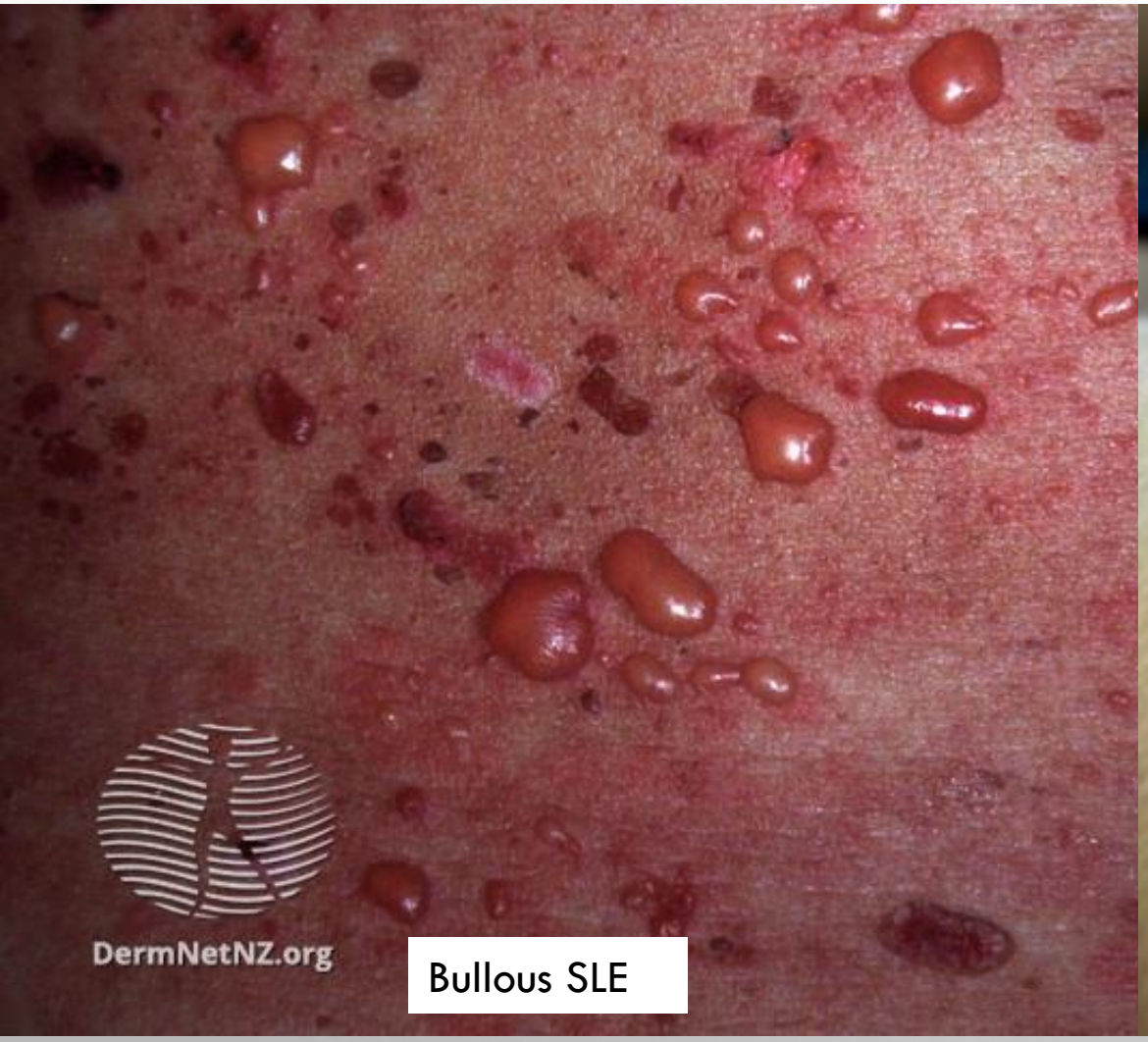
OTHER CAUSES



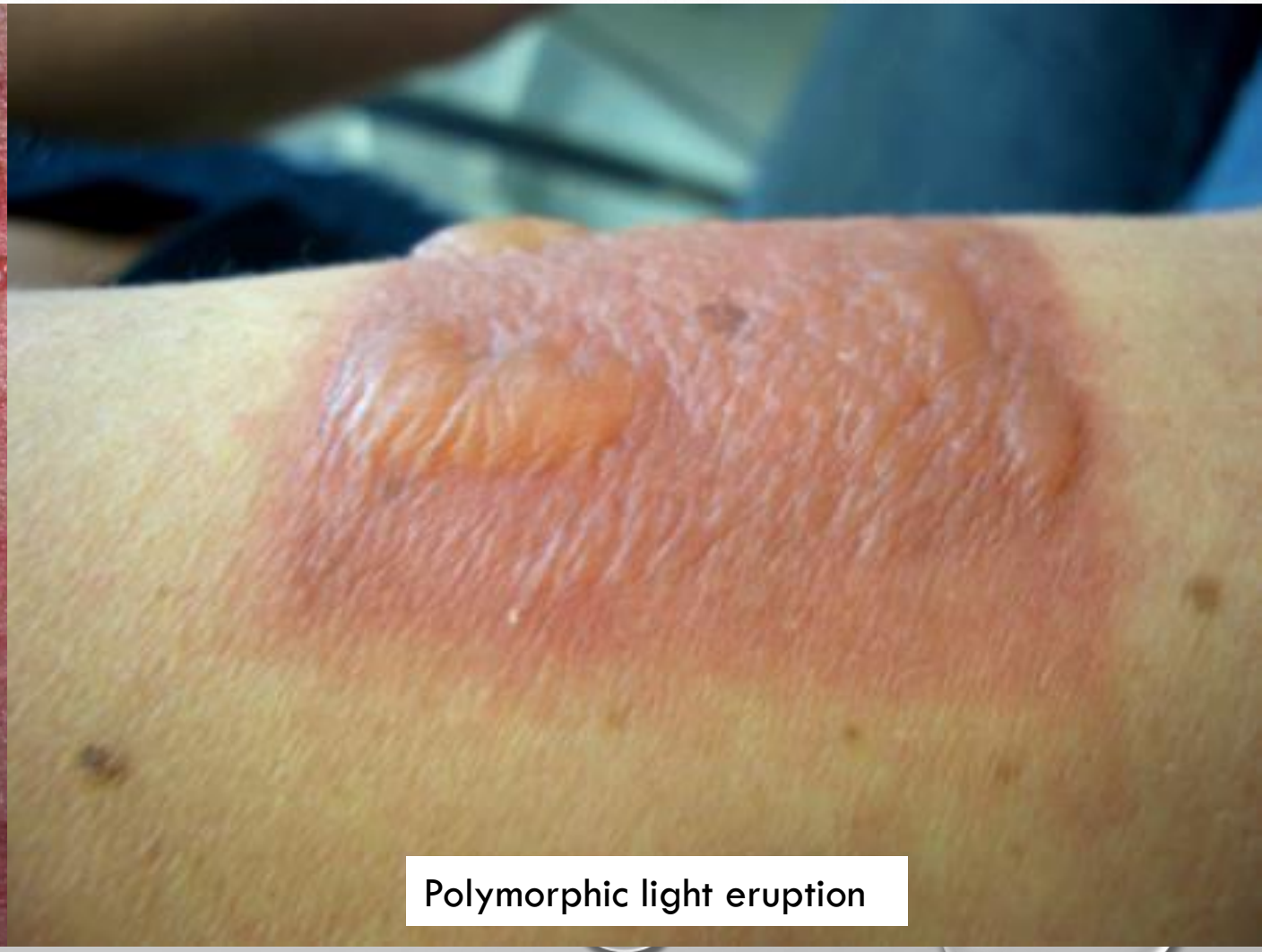
Arthropod bites or stings



Porphyria cutanea tarda

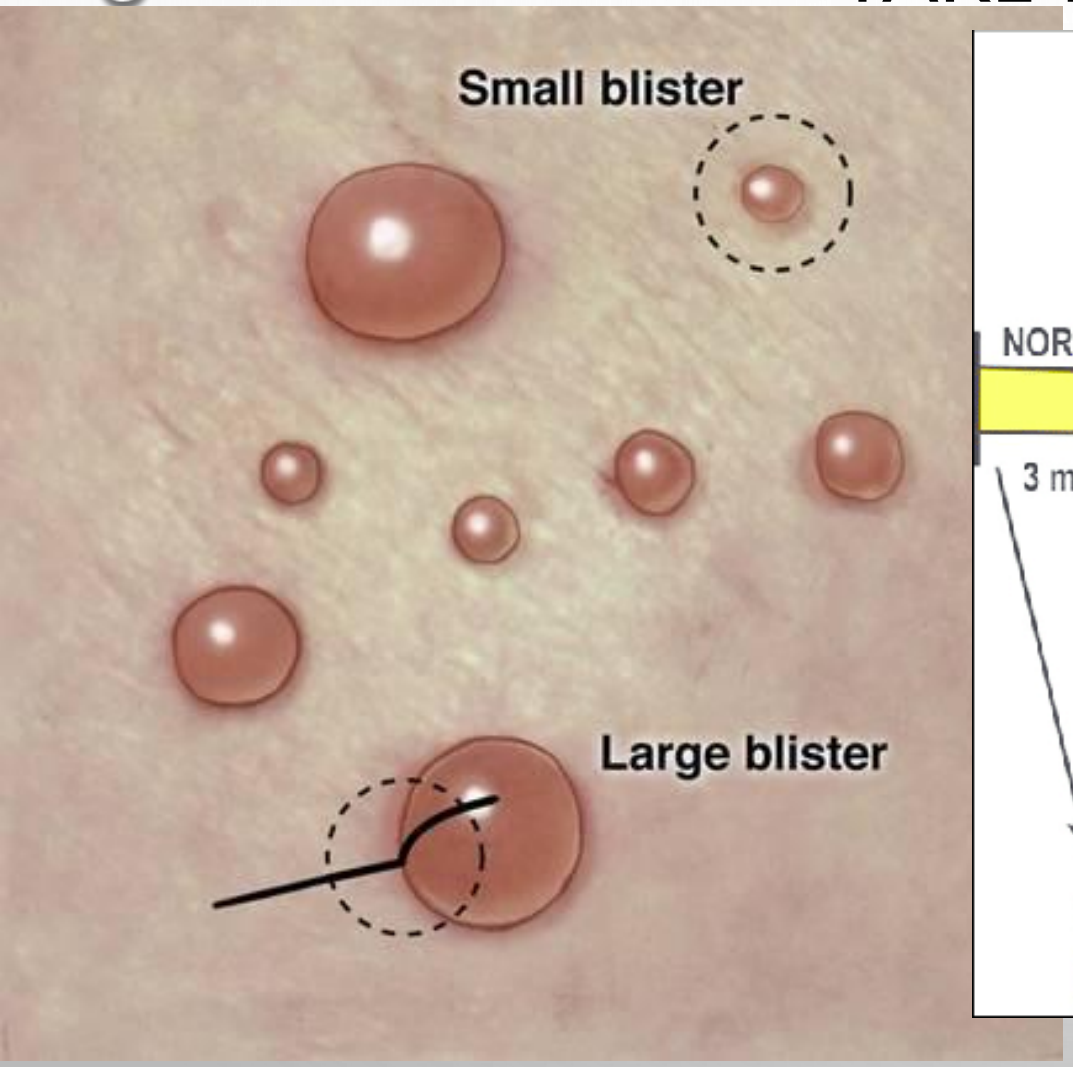


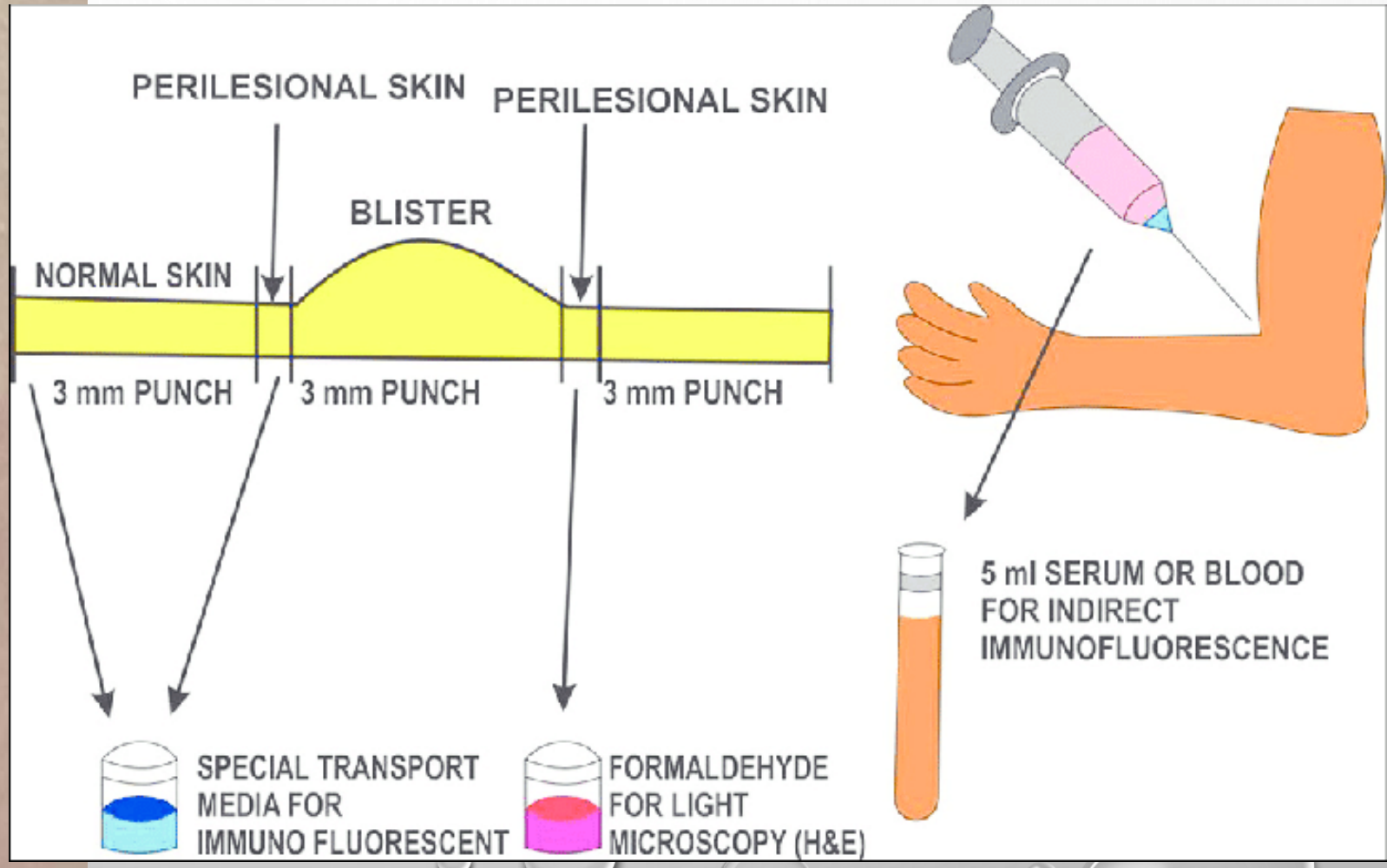
Bullous SLE



Polymorphic light eruption

TAKE HOME MESSAGE





THAT'S ALL FOLKS