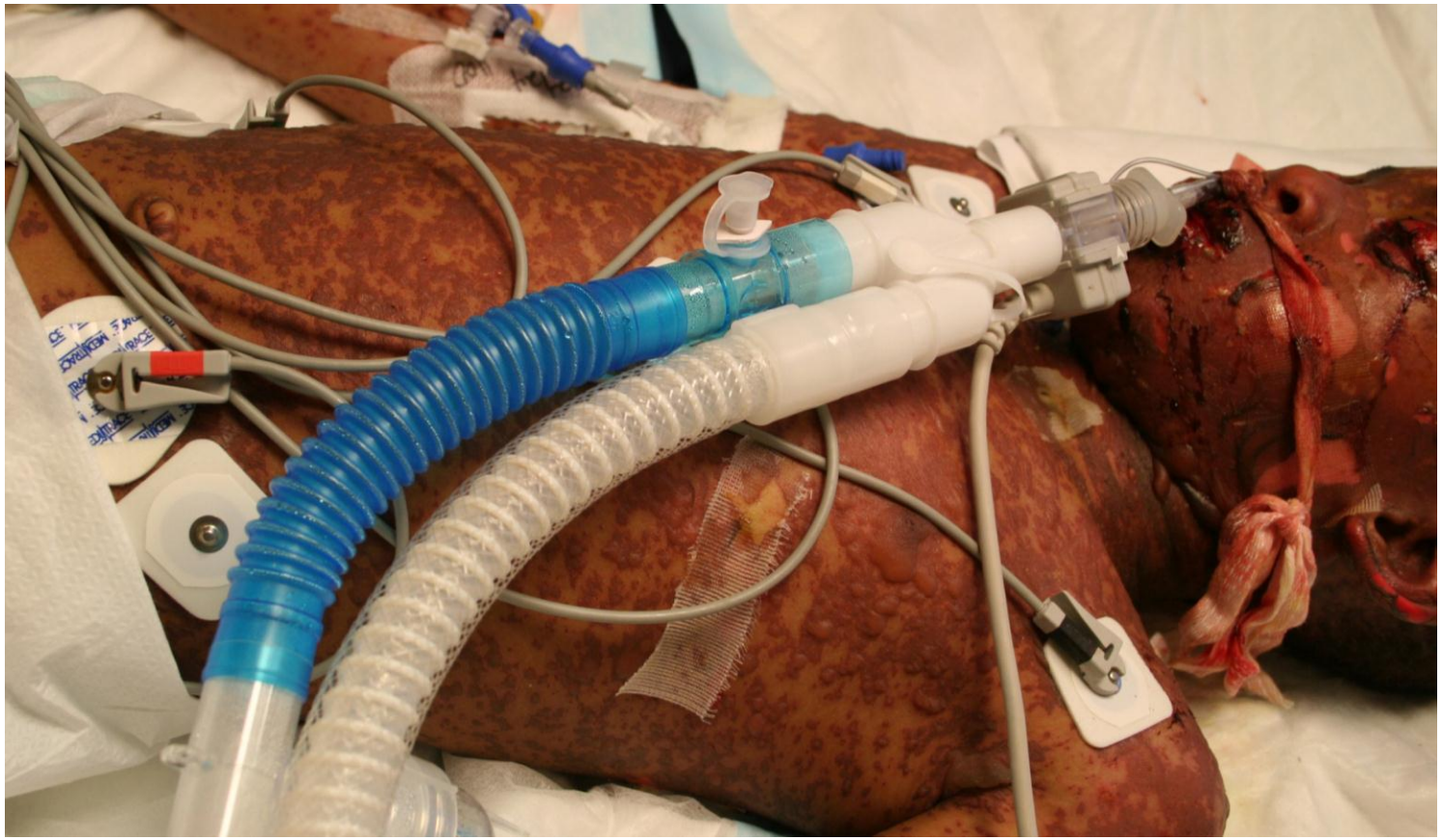

Dermatologic Emergencies

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Alarming Morphological patterns

- Urticaria / Angioedema
- Purpura / Ecchymoses
- Bullae / Sloughing
- Necrosis / Gangrene
- Exfoliative Erythroderma Syndrome
- Generalized/ widespread rashes in the acutely ill febrile patient

Dermatologic Emergencies

Urticaria/angioedema/anaphylaxis

Purpura

Bullous disease

Steven's Johnson syndrome (SJS) / Toxic epidermal necrolysis (TEN)

Erythroderma

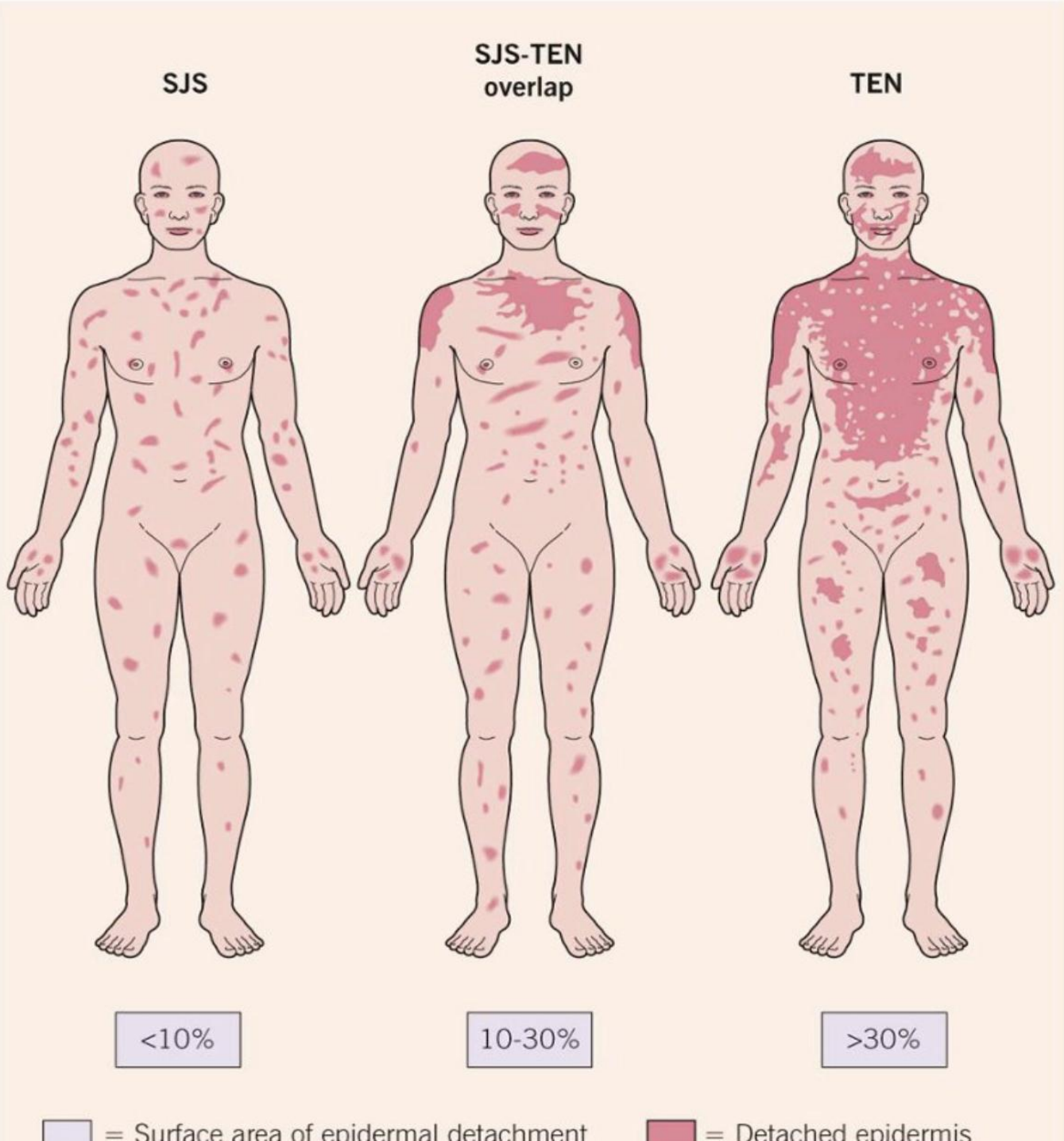
Steven's Johnson syndrome (SJS)
Toxic epidermal necrolysis (TEN)

Steven's Johnson syndrome (SJS)
Toxic epidermal necrolysis (TEN)

- Rare, acute, life-threatening mucocutaneous disease.
- Nearly always drug-related.
- Keratinocyte death → separation of skin at the dermal-epidermal junction.
- Characteristic symptoms: High fever, skin pain, anxiety and asthenia.
- It is crucial to diagnose it early so the causal drug can be discontinued.

Spectrum of disease based on surface area involved

SJS and TEN are variants of an identical pathologic disease and differ only in the percentage of body surface involved.



Steven's Johnson syndrome (SJS)
Toxic epidermal necrolysis (TEN)

- Mortality:

- 5% for patients with SJS.
- 25%-50% for patients with TEN

- Cause:

- Medications (95%)
- Infections, Immunizations (rare)

Steven's Johnson syndrome (SJS)
Toxic epidermal necrolysis (TEN)

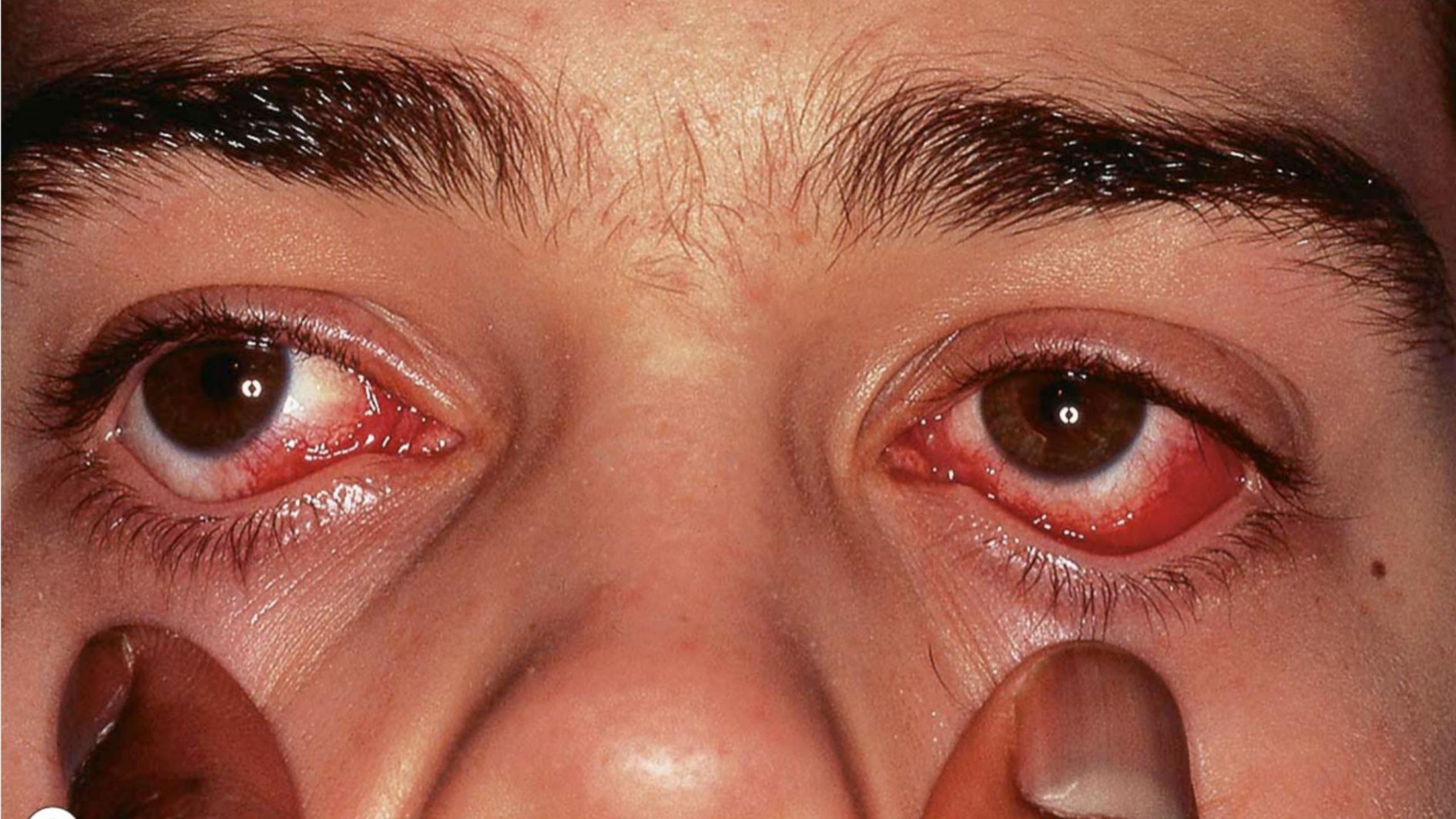
Medications:

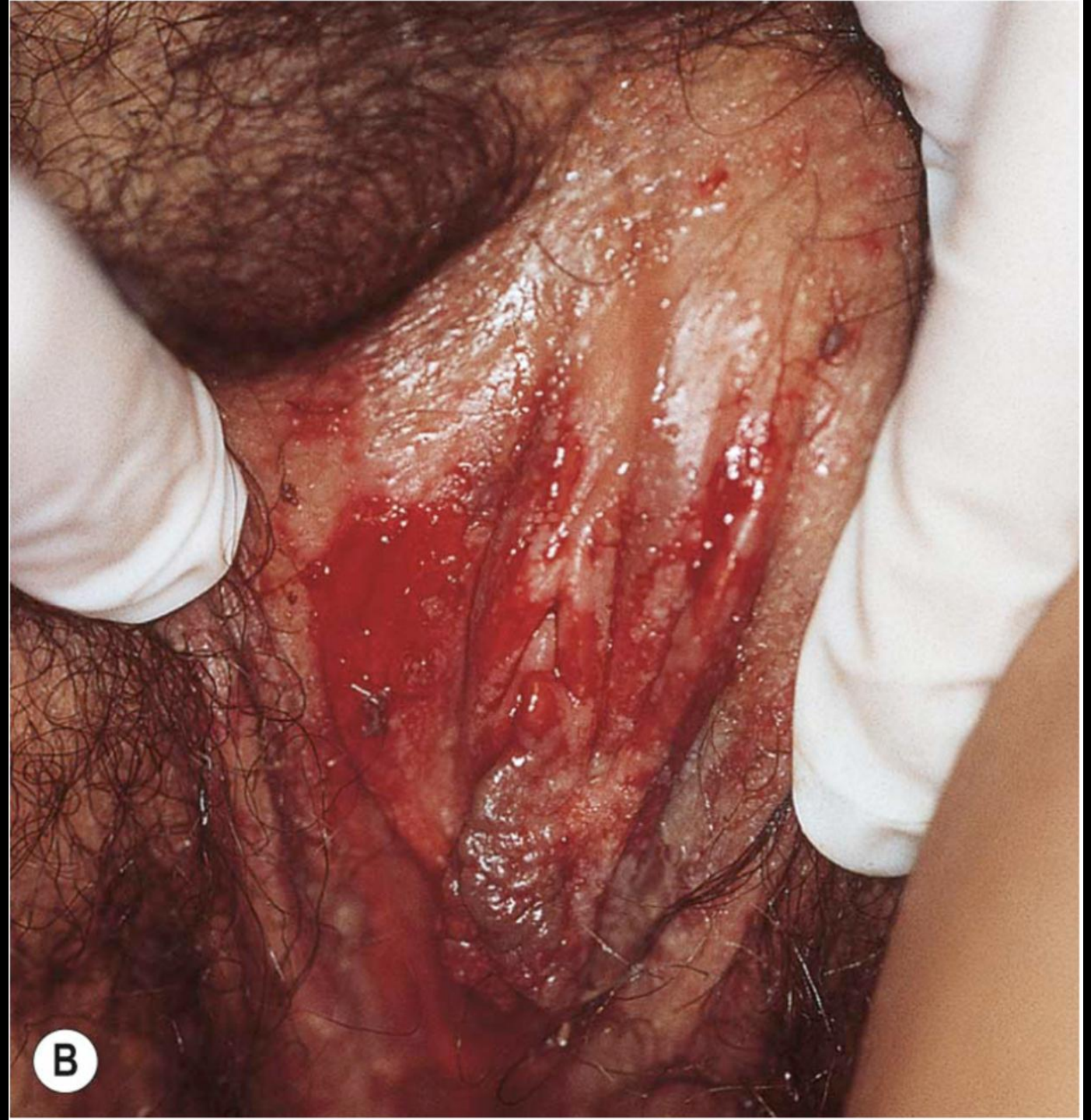
- More than 100 drugs have been identified to date as being associated with SJS/TEN!

- Most common:
- Allopurinol
- Antibiotics (Sulfonamides)
- NSAIDs
- Anti-convulsants.

Clinical features of SJS/TEN

- Initially: Fever, Stinging eyes, and pain upon swallowing.
- These symptoms precede cutaneous manifestations by 1 to 3 days.
- Skin lesions first appear on the trunk, spreading to the neck, face and proximal upper extremities.
- Distal arms and legs are relatively spared (but not the palms/soles).
- Erythema/erosions of the buccal, ocular and genital mucosae are present in more than 90% of patients.
- TEN → epithelium of the respiratory and G.I tract can also occur.
- Skin lesions are usually tender & mucosal erosions are very painful.

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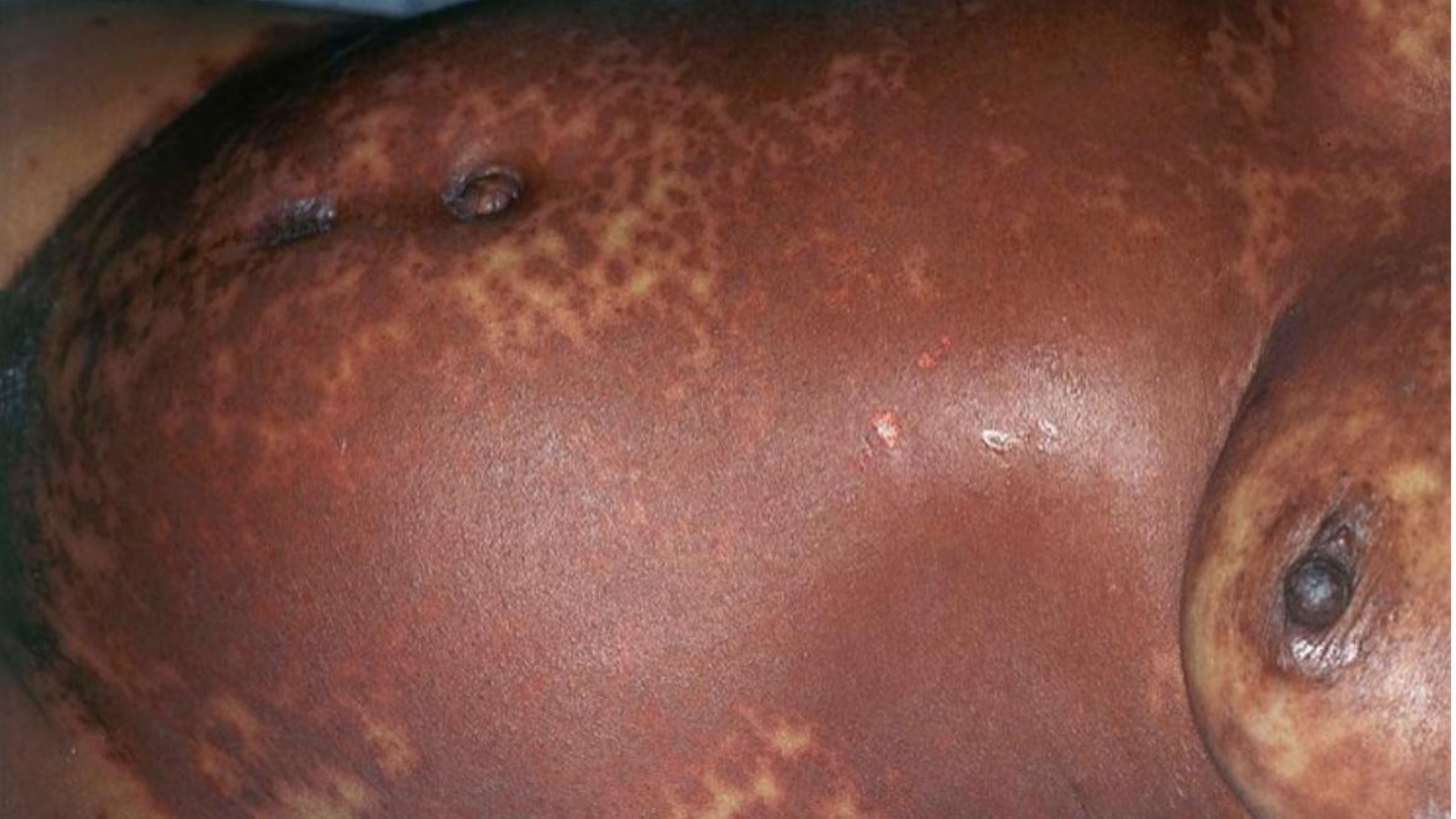
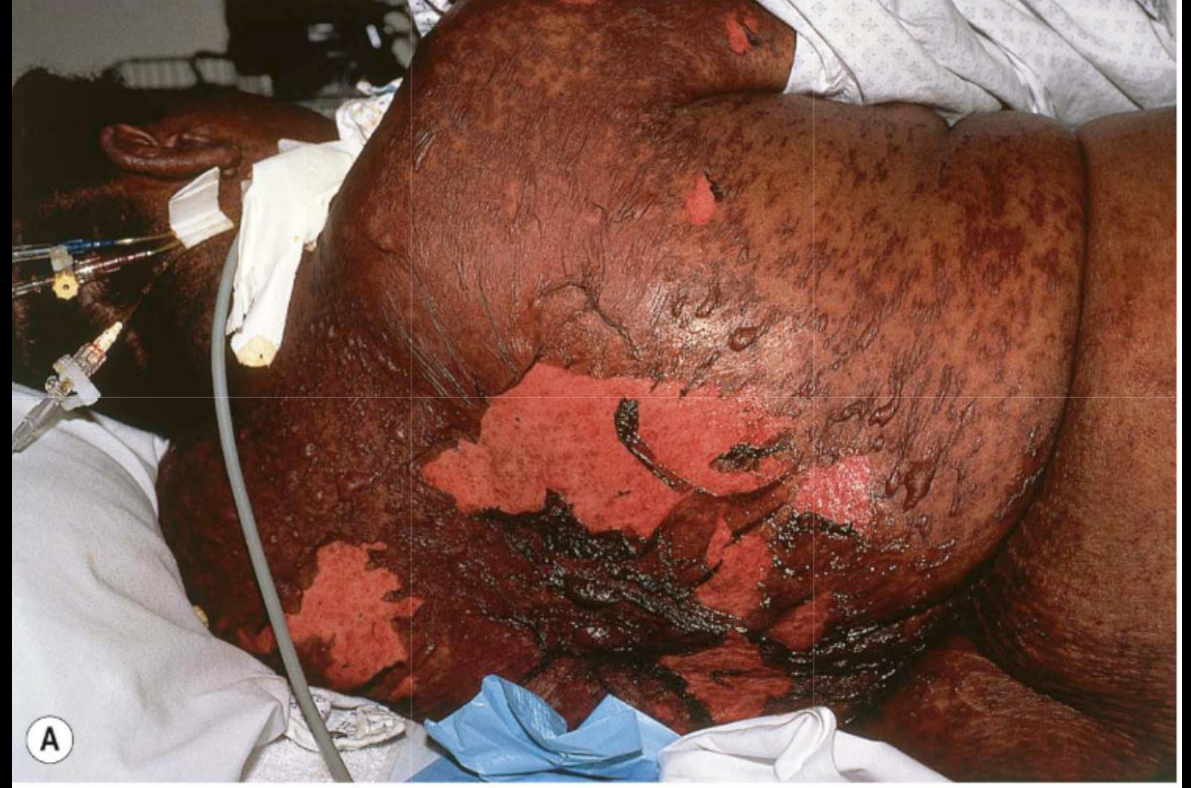
Morphology of skin lesions in SJS/TEN

- First: erythematous, dusky red or purpuric macules of irregular size and shape, they have a tendency to coalesce.
- +ve Nikolsky sign.
- Some lesions have a dusky center (Target-like appearance).
- Later: Full-thickness necrosis can develop (can be very rapid).
- The necrotic epidermis detaches from the dermis, fluid fills the space, giving rise to blisters (flaccid blisters).
- The blisters can be extended sideways by slight pressure of the thumb (Asboe-Hansen sign).
- The skin resembles wet cigarette paper.

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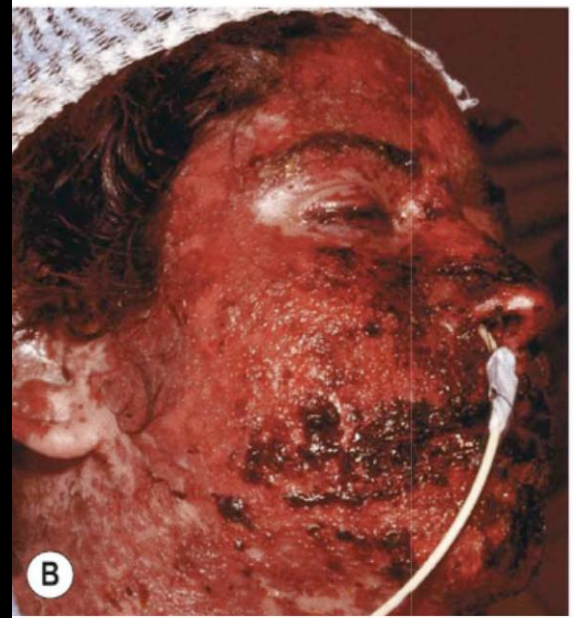
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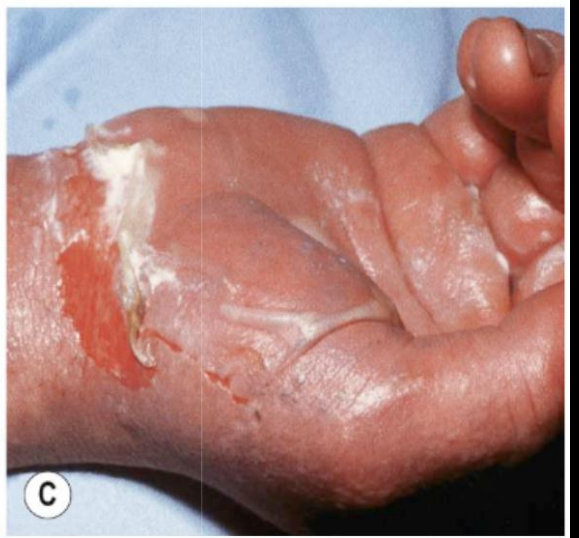
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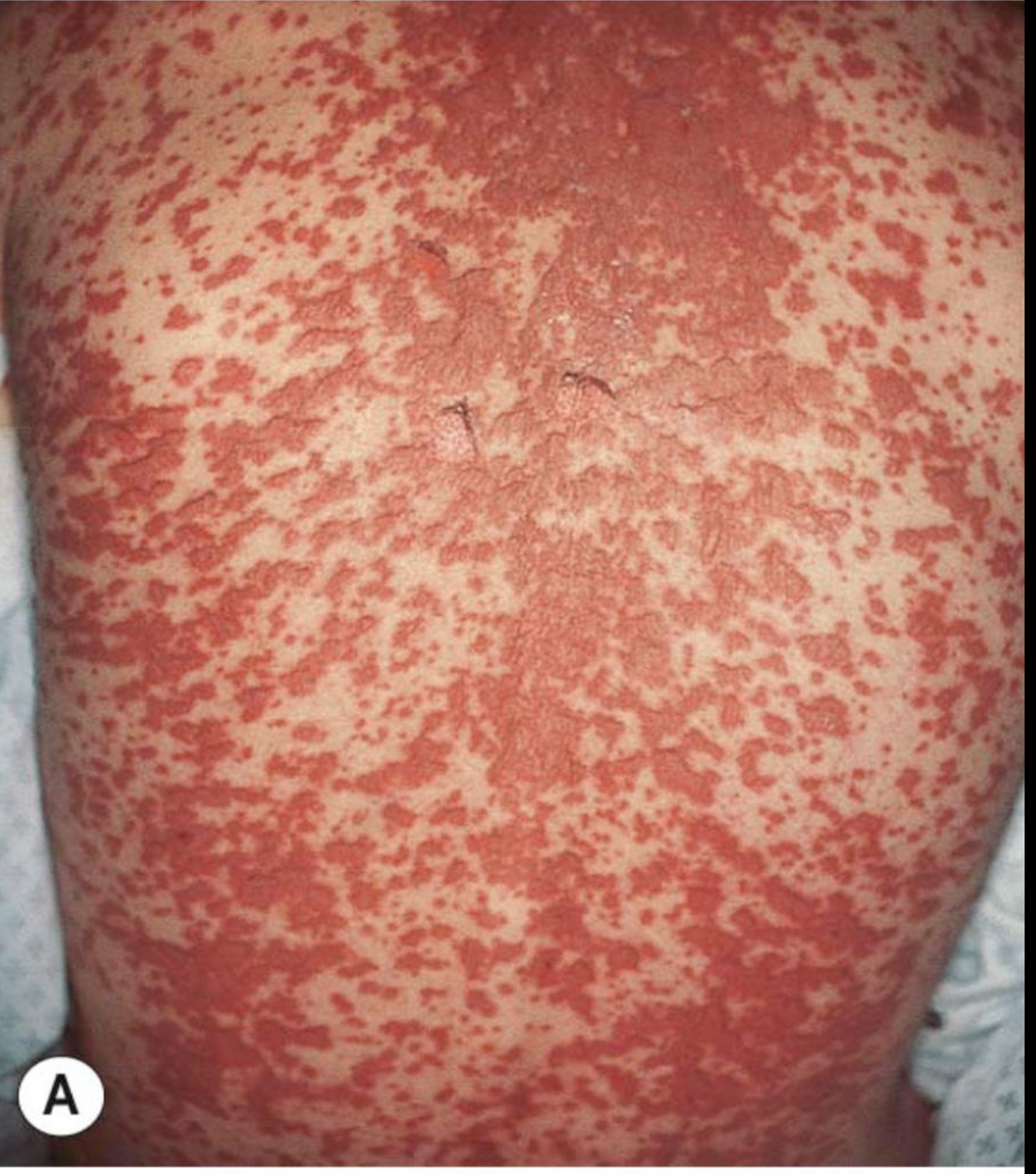
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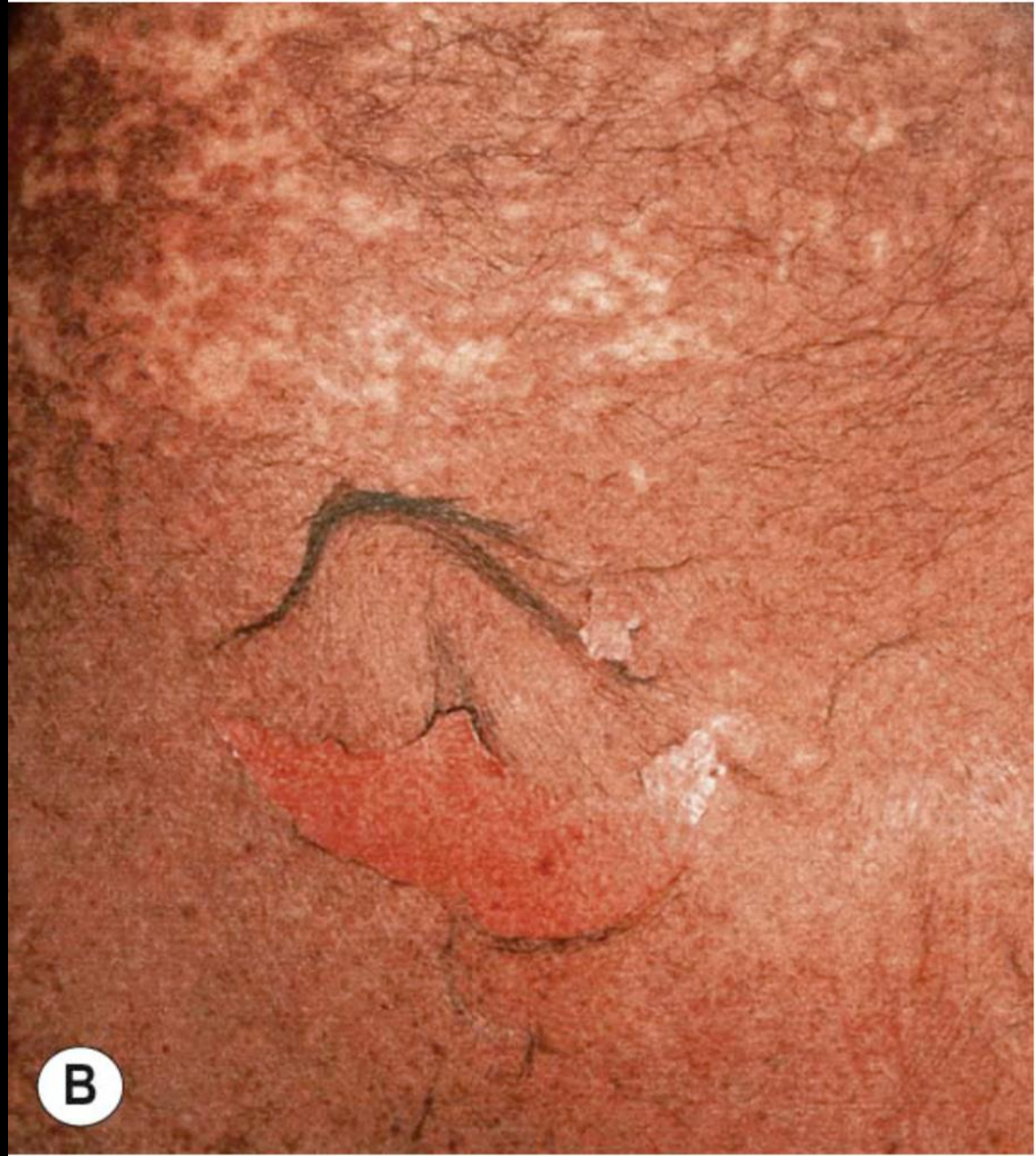
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SCORTEN
A prognostic scoring system for patients with TEN

- Age >40 years
- HR >120 bpm
- Cancer or hematologic malignancy
- BSA involved on day 1 above 10%
- Serum urea level > 10 mmol/l
- Serum bicarbonate level <20 mmol/l
- Serum glucose level >14 mmol/l

Mortality rate

0-1 → 3.2%

2 → 12.1%

3 → 35.8%

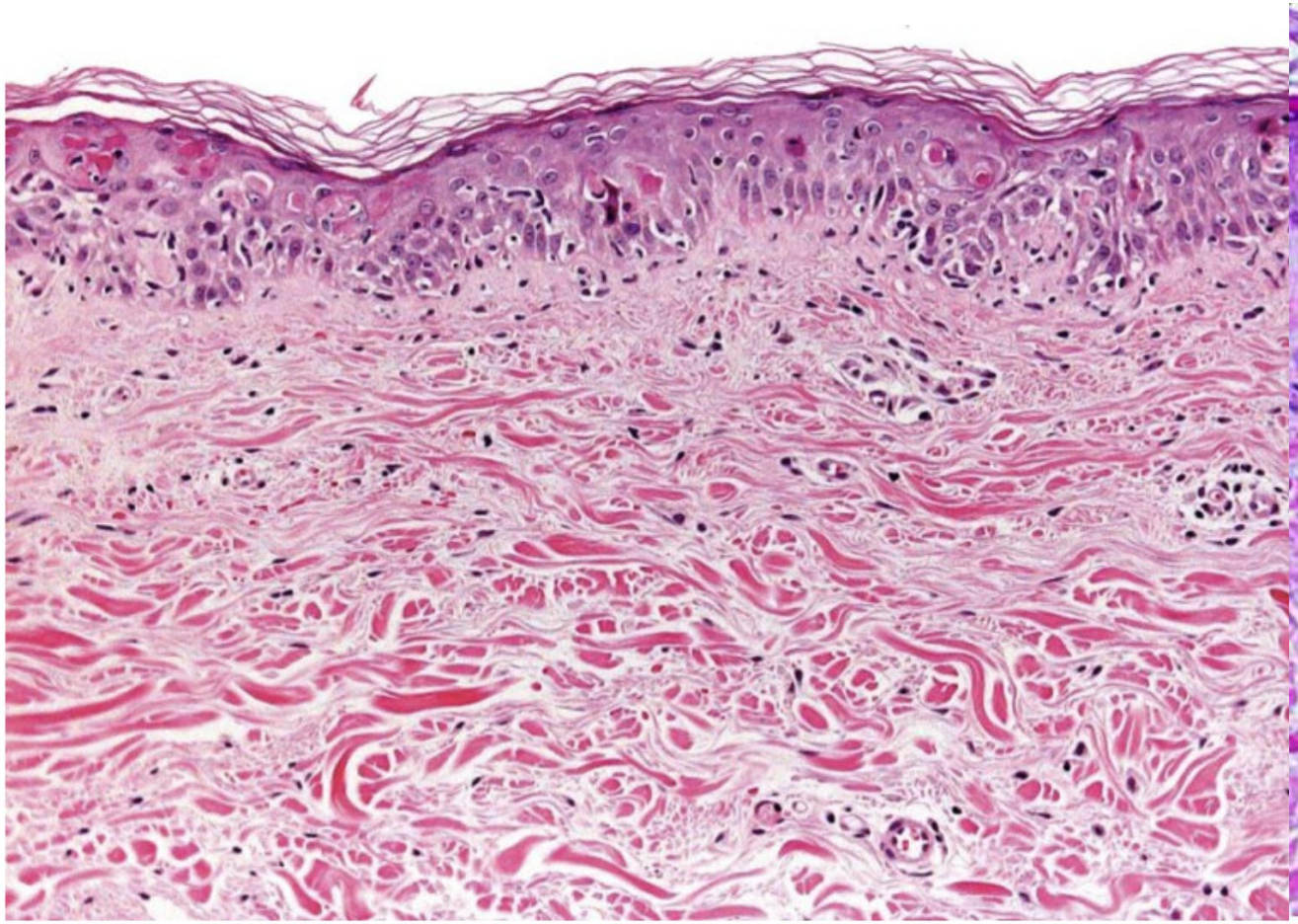
4 → 58.3%

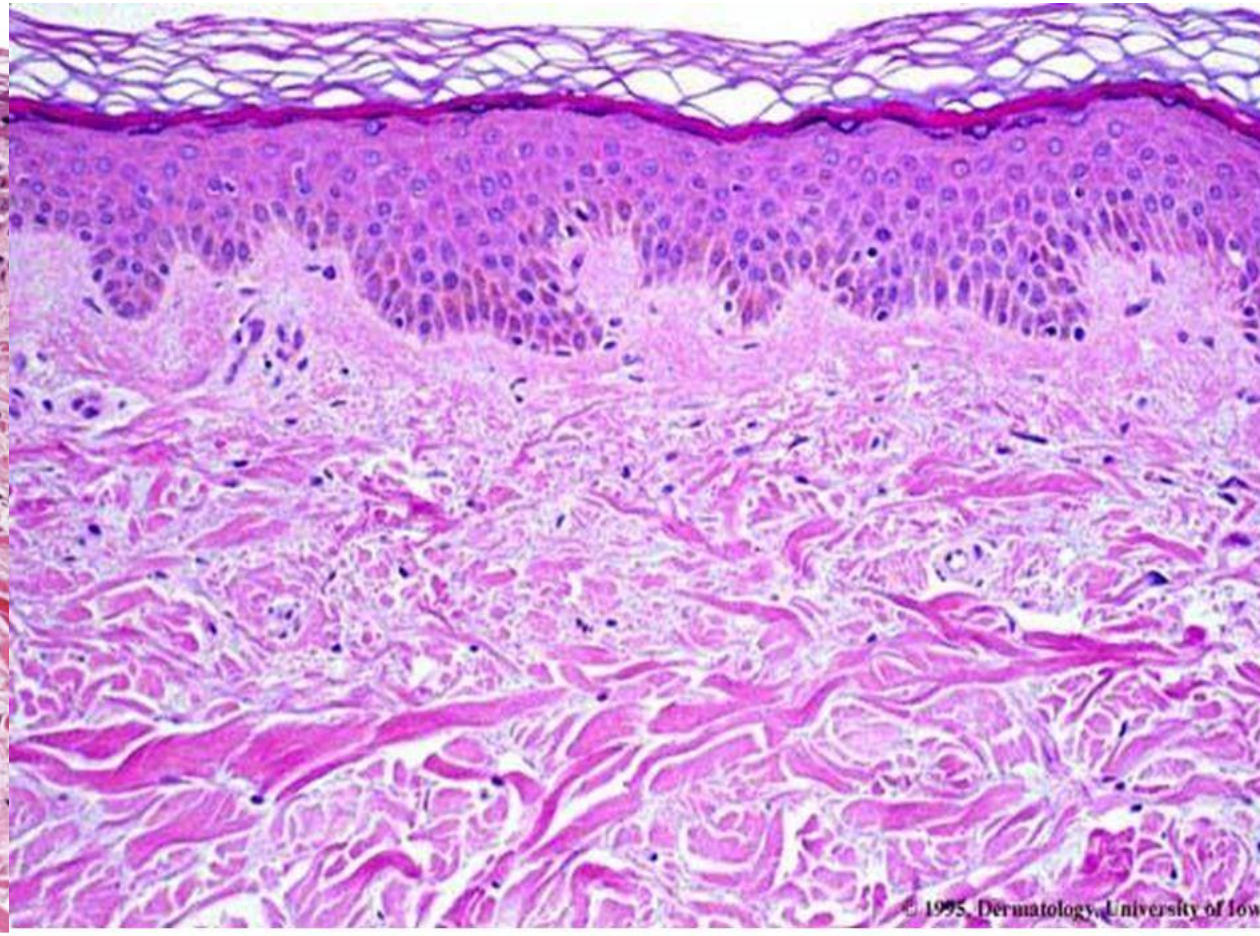
5 or more → 90%

Toxic epidermal necrolysis (TEN)

- Death occurs in 1/3 of pts with TEN (mainly due to infections).
- Best managed in the ICU/Burn unit.
- Eliminating the culprit medication is the most important first step.
- SJS/TEN usually occurs 7-21 after the initiation of the drug (first exposure) and within 2 days in the case of re-exposure to a drug that previously caused SJS or TEN.

Histology





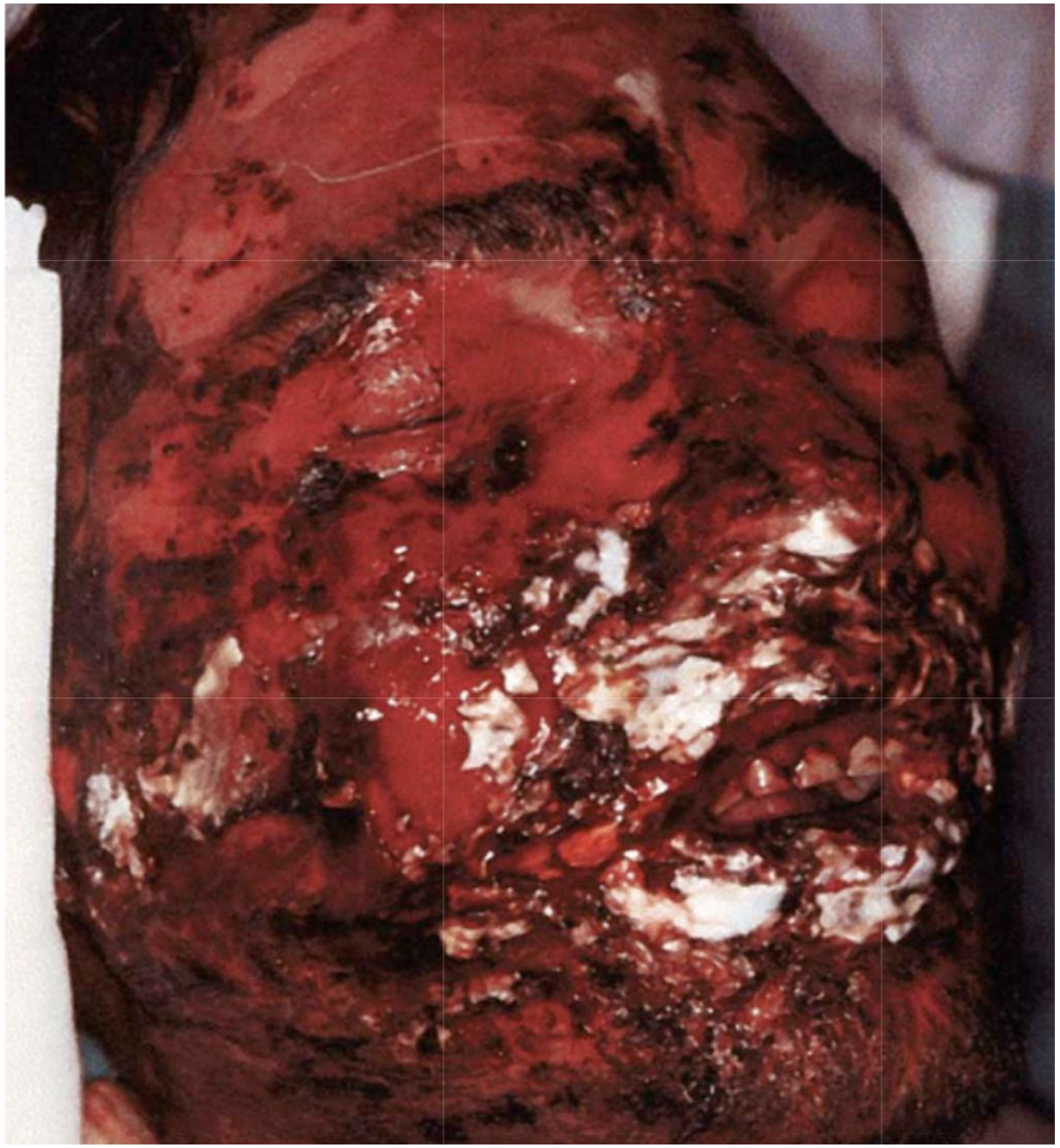
NORMAL SKIN

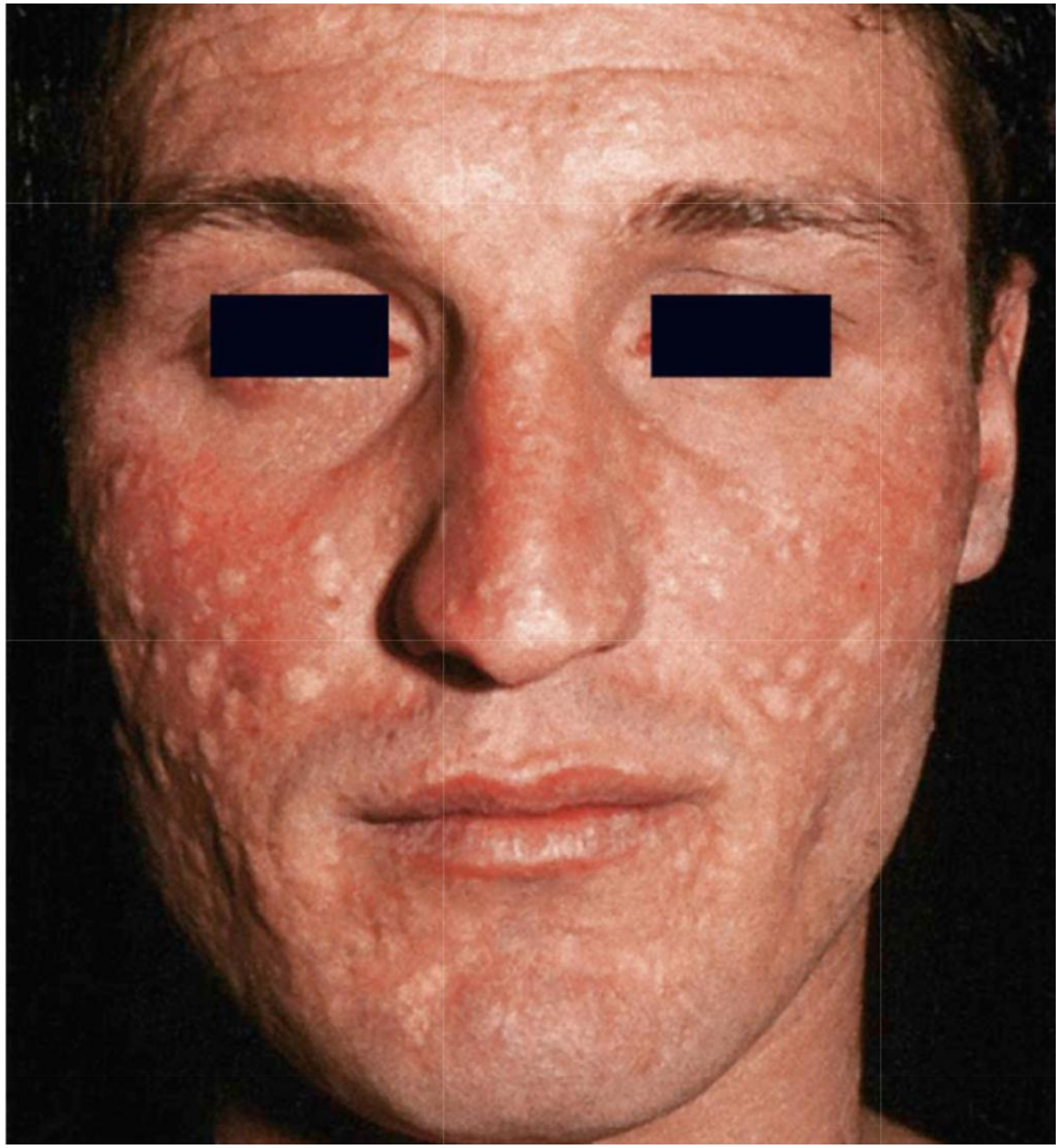
Treatment

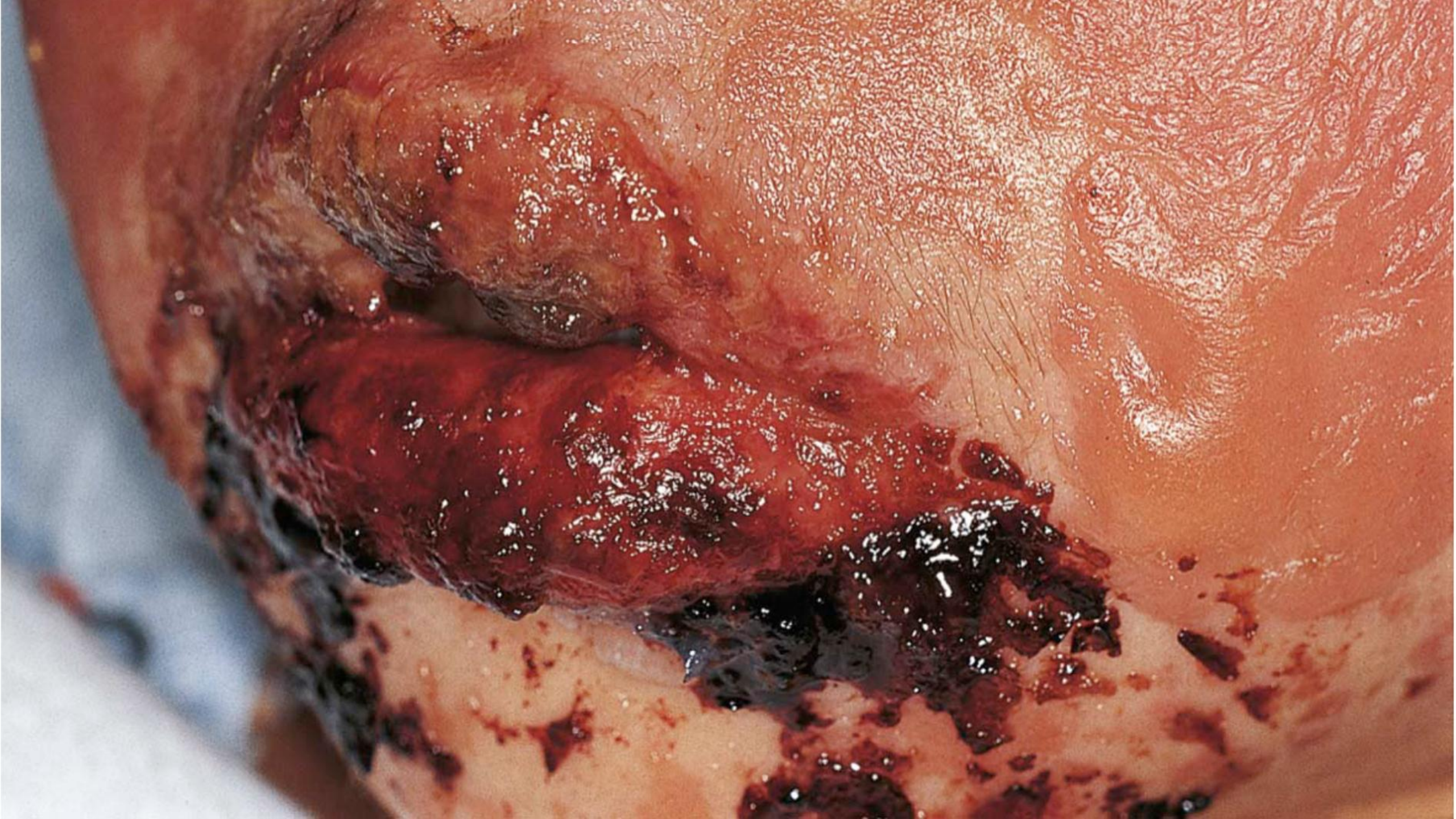
- Supportive care in a burn unit: wound care, hydration, nutritional support..etc
- Regular examination by an ophthalmologist

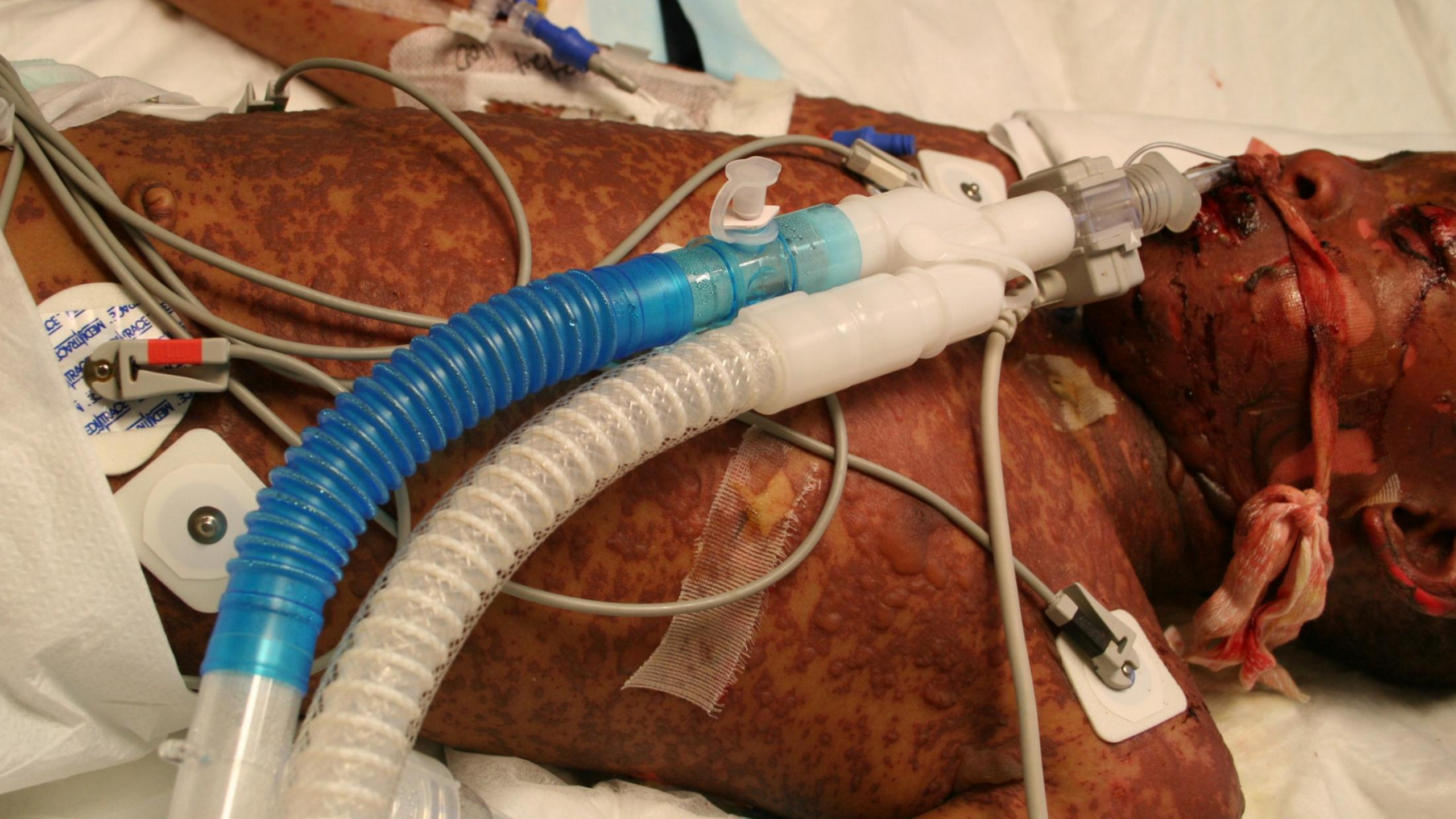
- To date, no specific therapy has shown efficacy in prospective, controlled clinical trials.

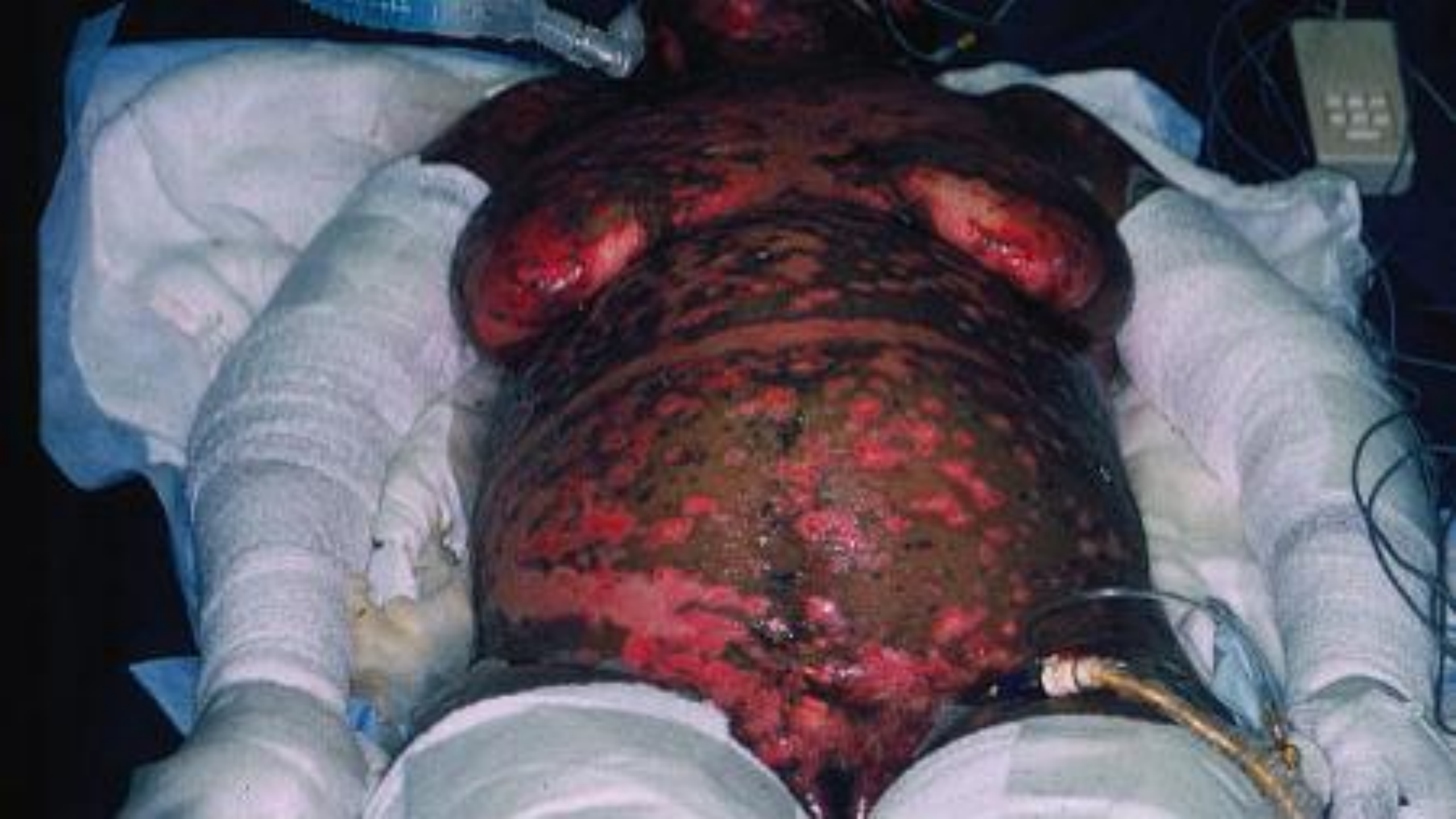
- Cyclosporine
- Cyclophosphamide
- Systemic steroids
- IVIg











Erythroderma

Erythroderma

- Generalized redness and scaling of >90% of the skin surface.
- Considered a serious, at times life-threatening condition.
- It does not represent a disease but rather a clinical presentation of a variety of diseases.
- M > F (avg age is ~50 yrs)

Causes of erythroderma

- Idiopathic
- Atopic Dermatitis
- Psoriasis
- Drug reaction
- Cutaneous T cell lymphoma (CTCL)
- Pityriasis rubra pilaris (PRP)

- Causes in children:
- Ichthyoses
- Immunodeficiences, infections
- Dermatitis, Psoriasis

Causes of Erythroderma

Pre-existing dermatosis (psoriasis, eczema)

50%

Drugs

15%

Lymphoma, leukemia

10%

Undetermined

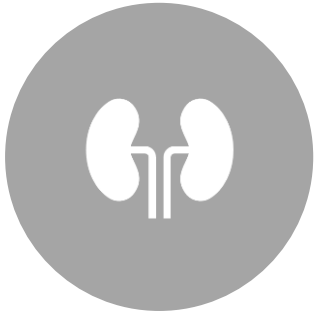
25%

Clinical features of erythroderma

- Erythema precedes exfoliation by 2-6 days.
- Pruritis in 90% of patients.
- Palmoplantar keratoderma.
- Nail changes in 40%.
- Diffuse non-scarring alopecia.

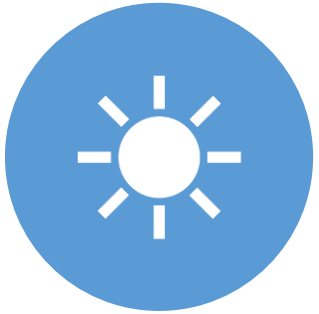
Systemic manifestations

GENERALIZED PERIPHERAL LYMPHADENOPATHY (50%)

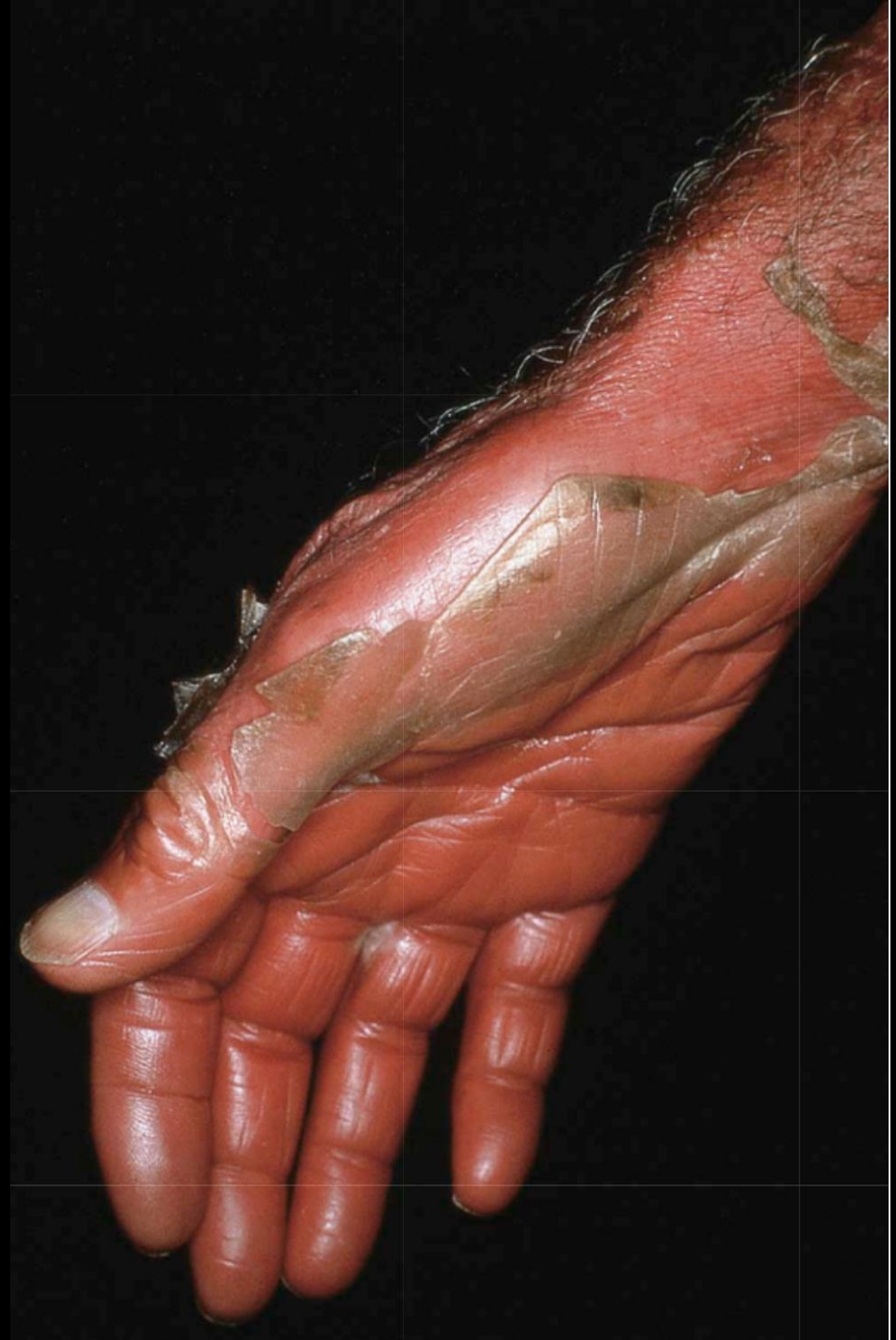


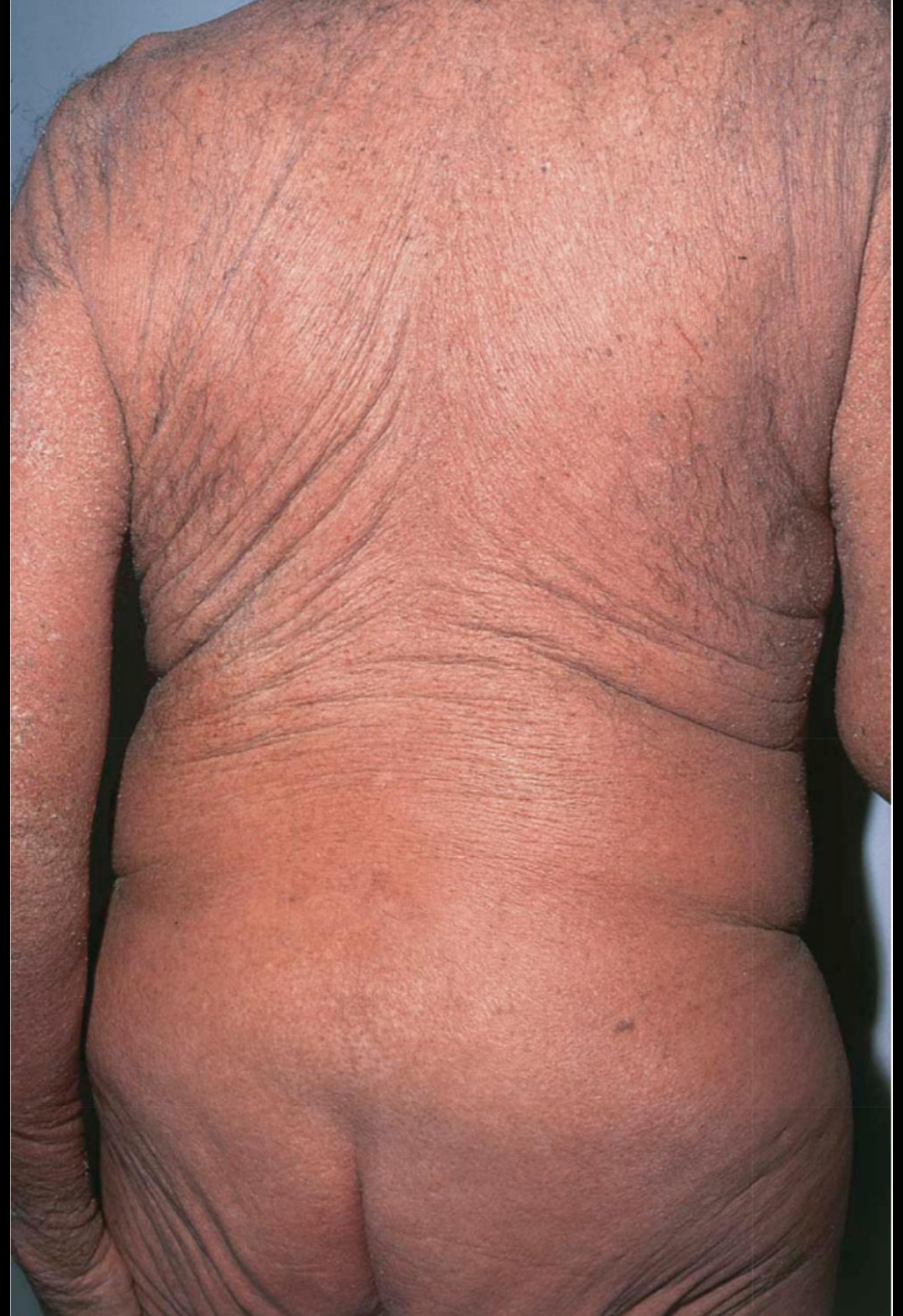
PEDAL OR PRETIBIAL EDEMA IN ~50% OF PATIENTS

TACHYCARDIA, RISK OF HIGH OUTPUT CARDIAC FAILURE (ESP. IN THE ELDERLY)



THERMOREGULATORY DISTURBANCES (HYPER-HYPO THERMIA)





Manifestations based on causative disease

1) Psoriasis:

- Nail changes (Oil-drop, onycholysis, nail pits)

2) Atopic dermatitis:

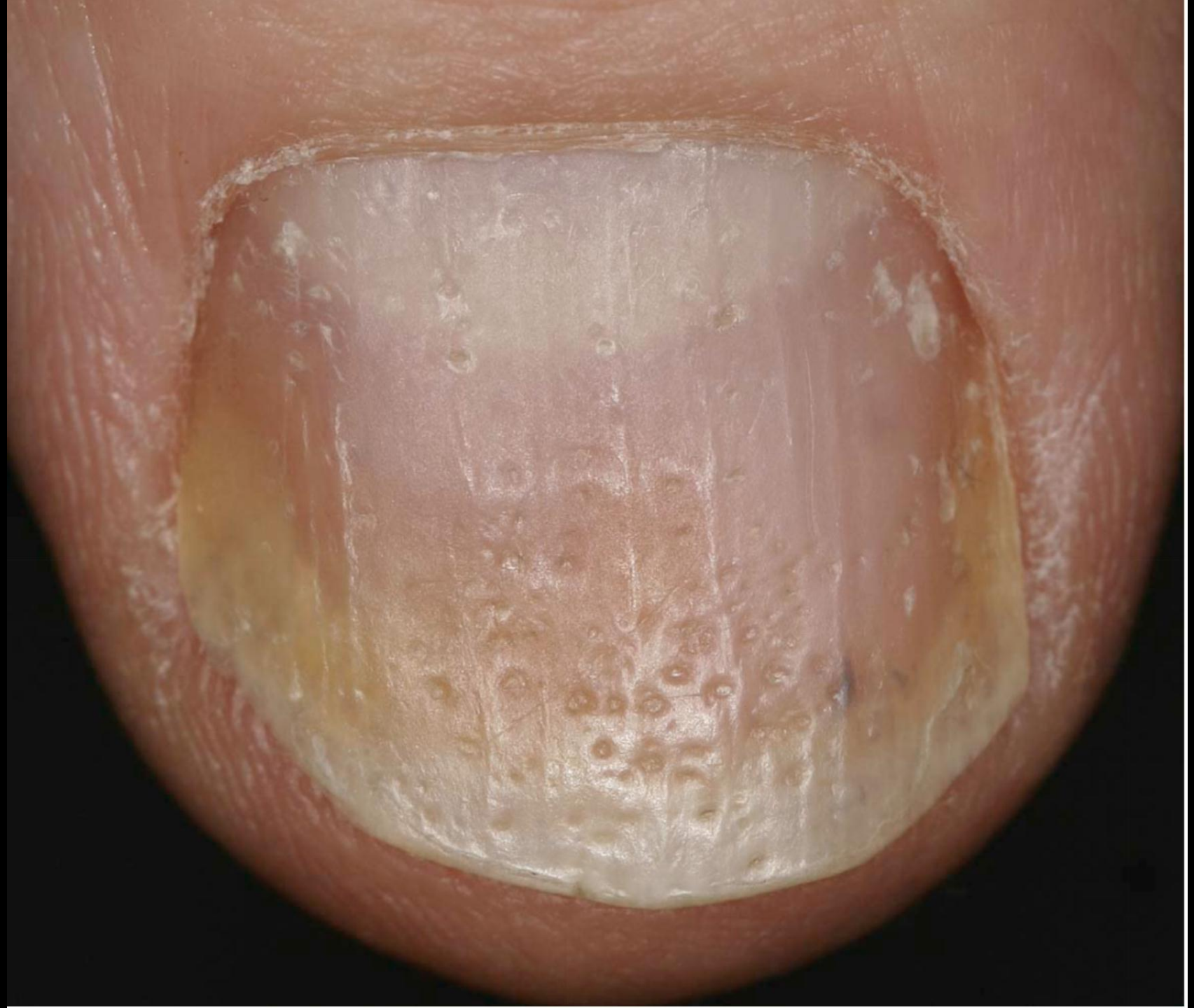
- Pruritis is intense
- Lichenification

3) Drug reactions:

- Morbiliform or scarlatiniform exanthem

4) Idiopathic erythroderma:

- Elderly men
- Lymphadenopathy and extensive palmoplantar keratoderma.
- Peripheral edema



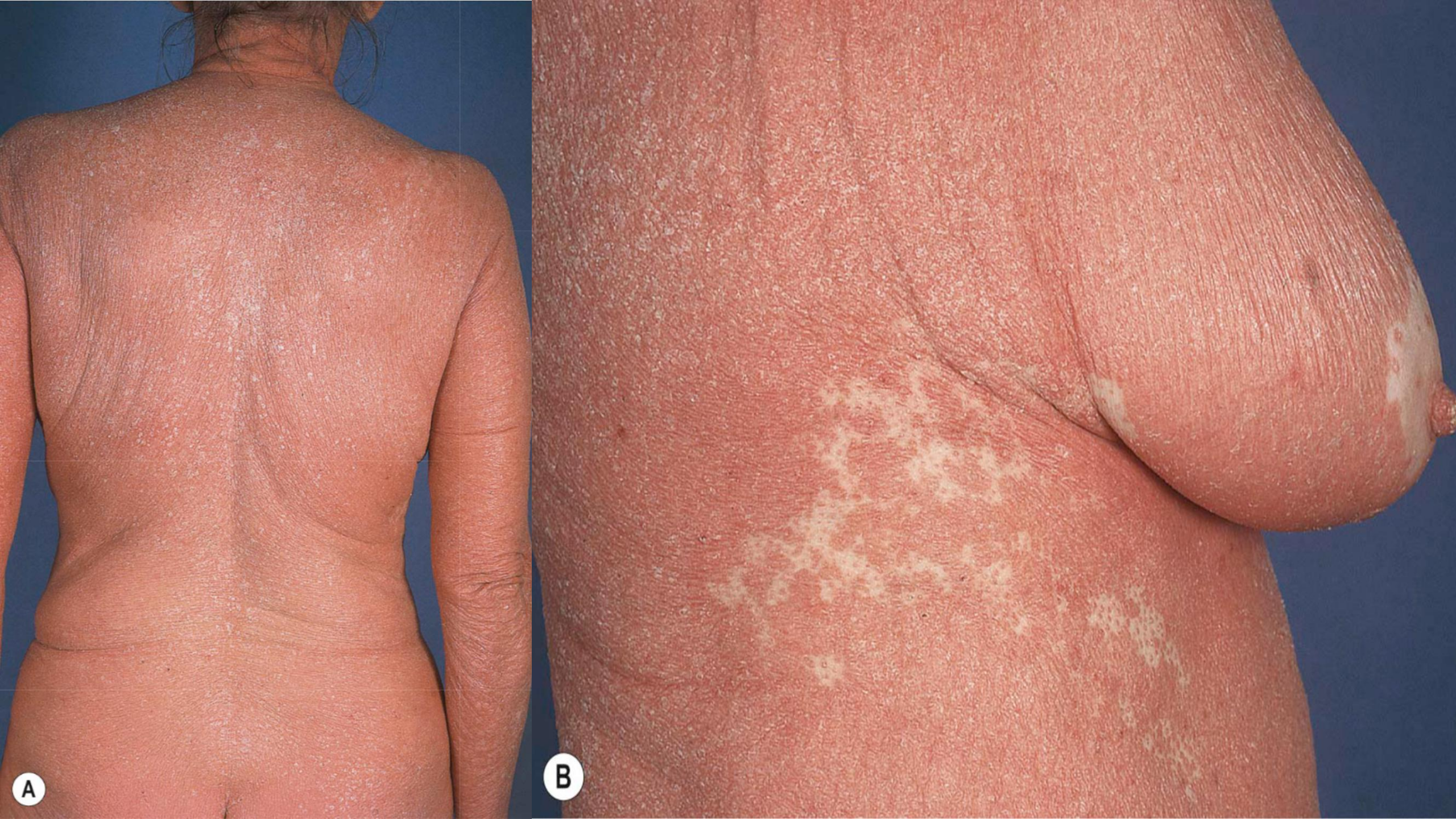
Manifestations based on causative disease

5) CTCL:

- Sezary syndrome: Erythroderma, Malignant T lymphocytes and generalized lymphadenopathy.
- Painful fissured keratoderma, diffuse alopecia, leonine facies.

6) PRP:

- Salmon to orange color.
- Follicular keratotic papules on the knees, elbows and dorsal fingers.
- Islands of sparing.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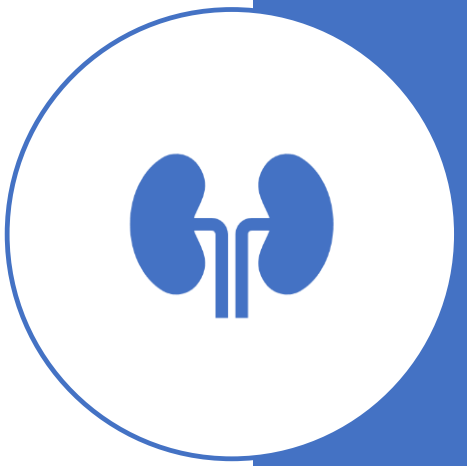
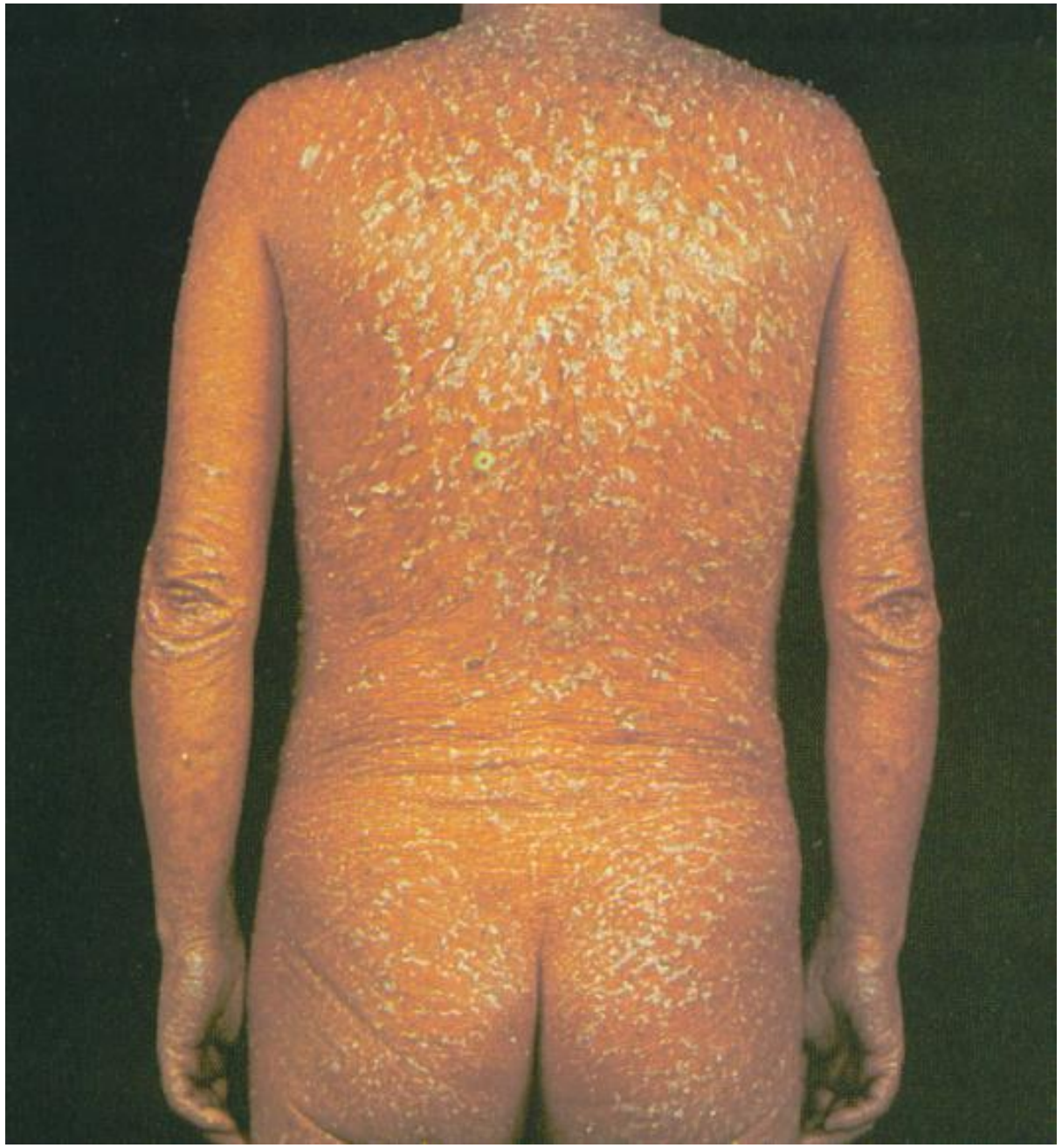
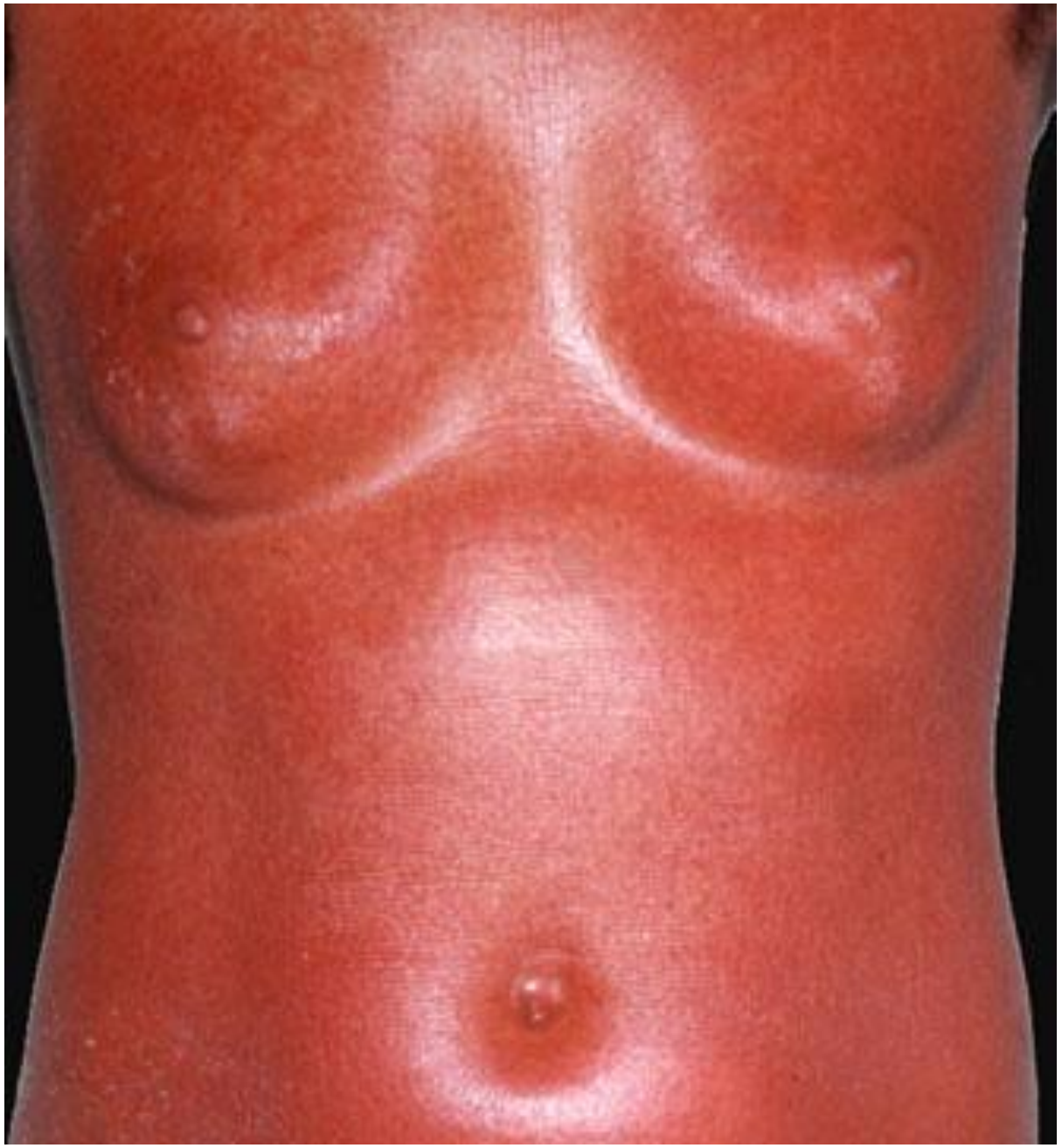
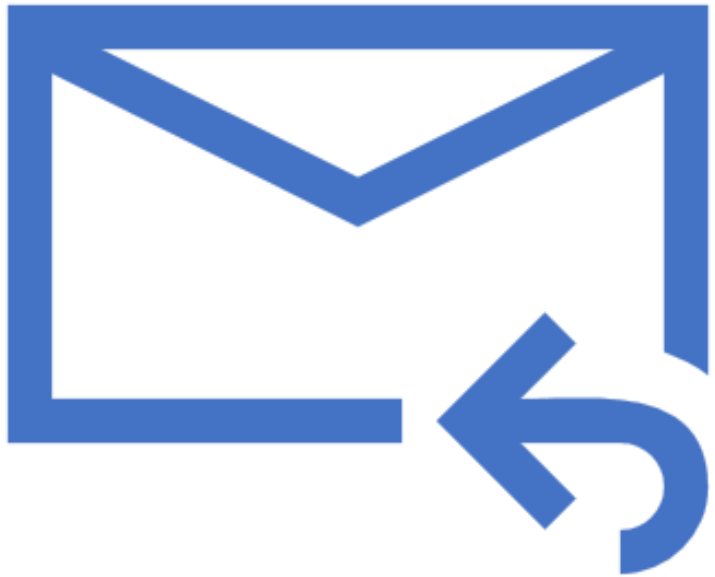
Treatment

- Hospitalization may be required.
- Regardless of cause: Nutritional assessment, correction of fluid and electrolyte imbalance, prevention of hypothermia and tx of secondary infections.
- Idiopathic: Topical and systemic corticosteroids. Anti-histamines.
- Treat the cause of erythroderma.









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