# DERMATOLOGIC EMERGENCIES

JONATHAN L. BINGHAM, MD, FACMS, FAAD
FELLOWSHIP TRAINED MOHS MICROGRAPHIC SURGEON
BOARD CERTIFIED DERMATOLOGIST

# DISCLOSURE

- I HAVE NO CONFLICTS OF INTEREST.
- I WILL BE SPEAKING ON OFF-LABEL USE OF MEDICATIONS.

# HOW TO APPROACH THIS TOPIC

- DERMATOLOGIC CONDITIONS THAT BRING PATIENTS TO THE EMERGENCY DEPARTMENT
- CONDITIONS THAT DERMATOLOGISTS CONSIDER EMERGENCIES
- CONDITIONS WITH DERMATOLOGIC SIGNS/SYMPTOMS THAT ARE EMERGENCIES

# Patients presenting to the ED with a dermatologic complaint have, for the purposes of this lecture, a dermatologic emergency.

1. TRUE
2. FALSE

From: **Skin Conditions That Bring Patients to Emergency Departments**

Arch Dermatol. 2011;147(1):118-120. doi:10.1001/archdermatol.2010.246

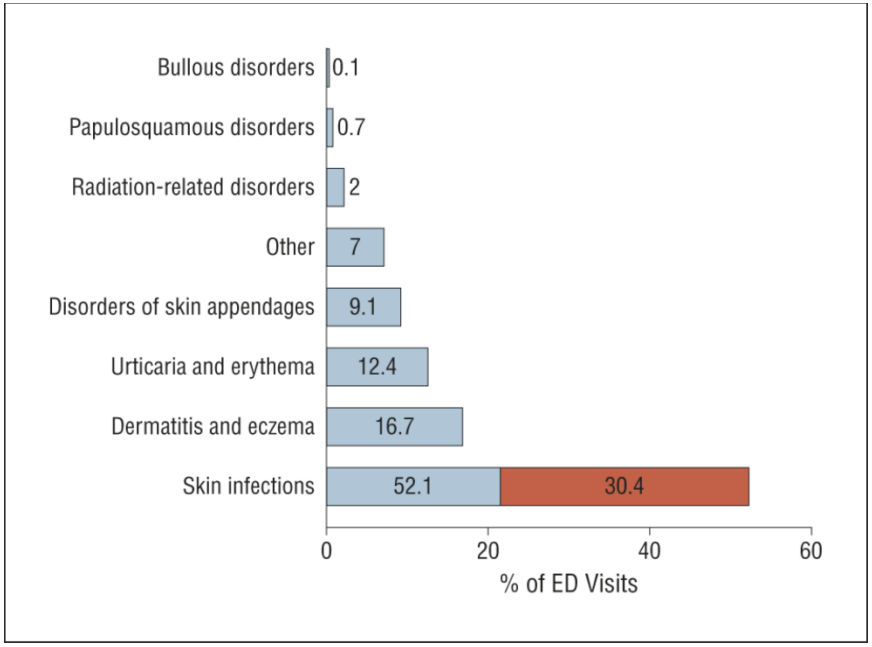



Figure Legend:

Percentage of each diagnostic category among patients seen in the emergency department (ED) for skin conditions. Among patients with skin infections, the fraction of those diagnosed as having cellulitis is in red.

# DERMATOLOGIC EMERGENCIES

- STEVEN JOHNSON SYNDROME/TOXIC EPIDERMAL NECROLYSIS
- KAWASAKI DISEASE
- MENINGIOCOCCEMIA/PURPURA FULMINANS
- ROCKY MOUNTAIN SPOTTED FEVER
- NECROTIZING FASCIITIS
- ENDOCARDITIS
- ERYTHRODERMA
- DRESS SYNDROME

# DERMATOLOGIC EMERGENCIES CONT'D

- DRUG REACTIONS
- INFECTIONS
  - BACTERIAL (NECROTIZING FASCIITIS), VIRAL (HSV) OR FUNGAL (MUCOR)
  - IMMUNOCOMPROMISED/OPPORTUNISTIC
- AUTOIMMUNE EXACERBATIONS
- INFLAMMATORY (ERYTHRODERMA)
- ENVIRONMENTAL (BURNS)
- SIGNS OF ABUSE

# Steven Johnson Syndrome and Toxic Epidermal Necrolysis are primarily differentiated by amount of body surface area (BSA) involved.

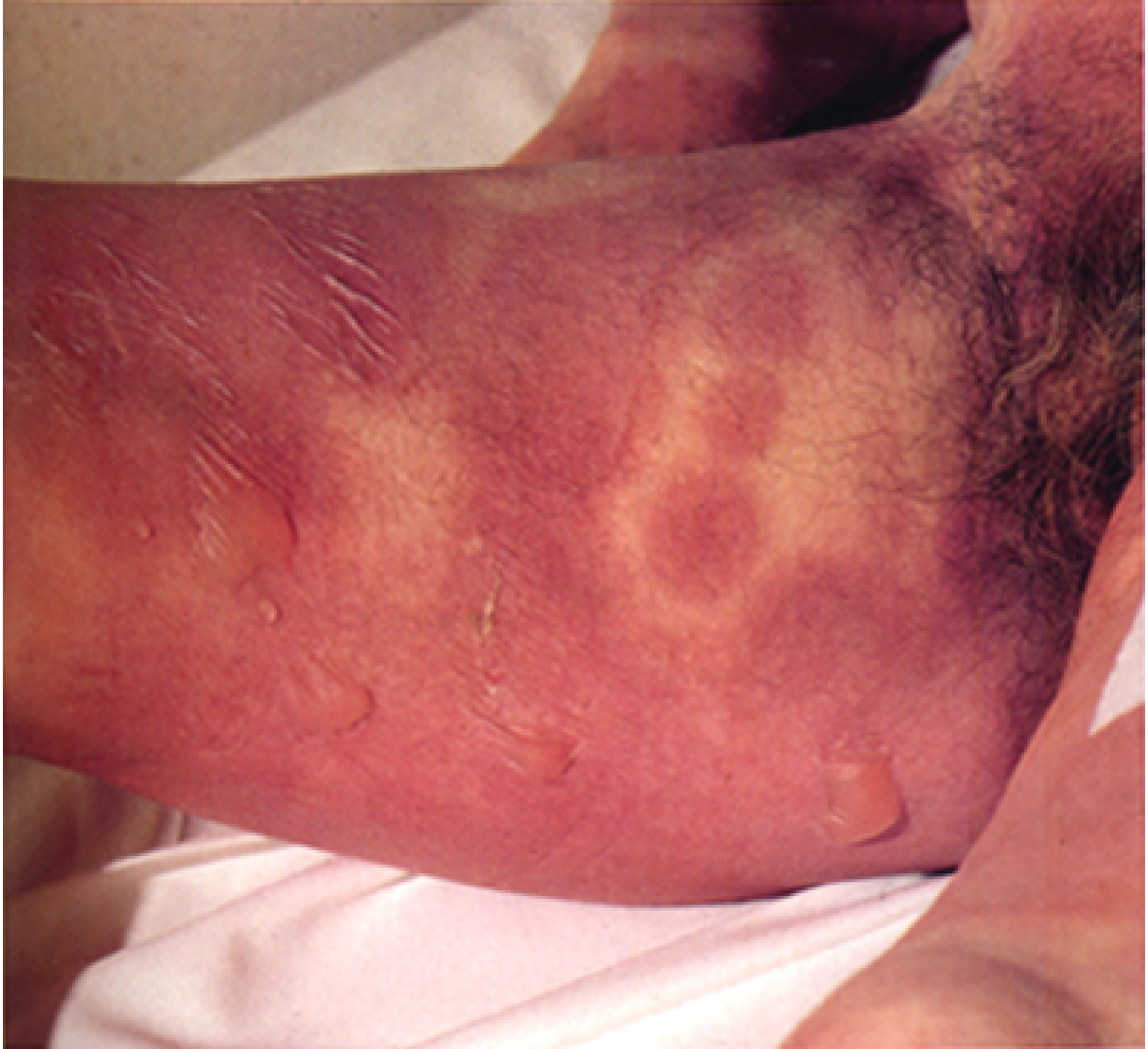
1. TRUE
2. FALSE

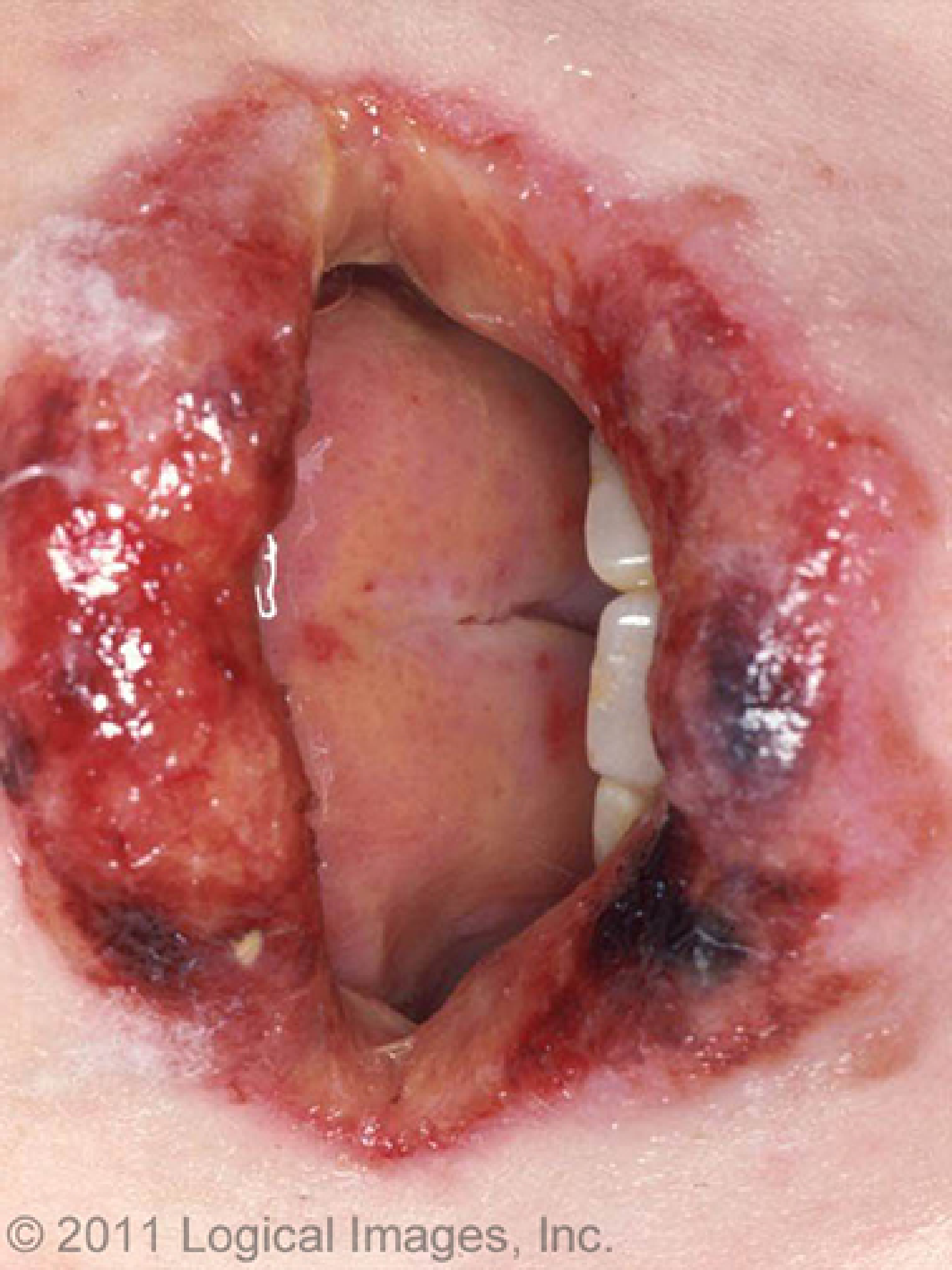
# STEVEN JOHNSON SYNDROME/TOXIC EPIDERMAL NECROLYSIS

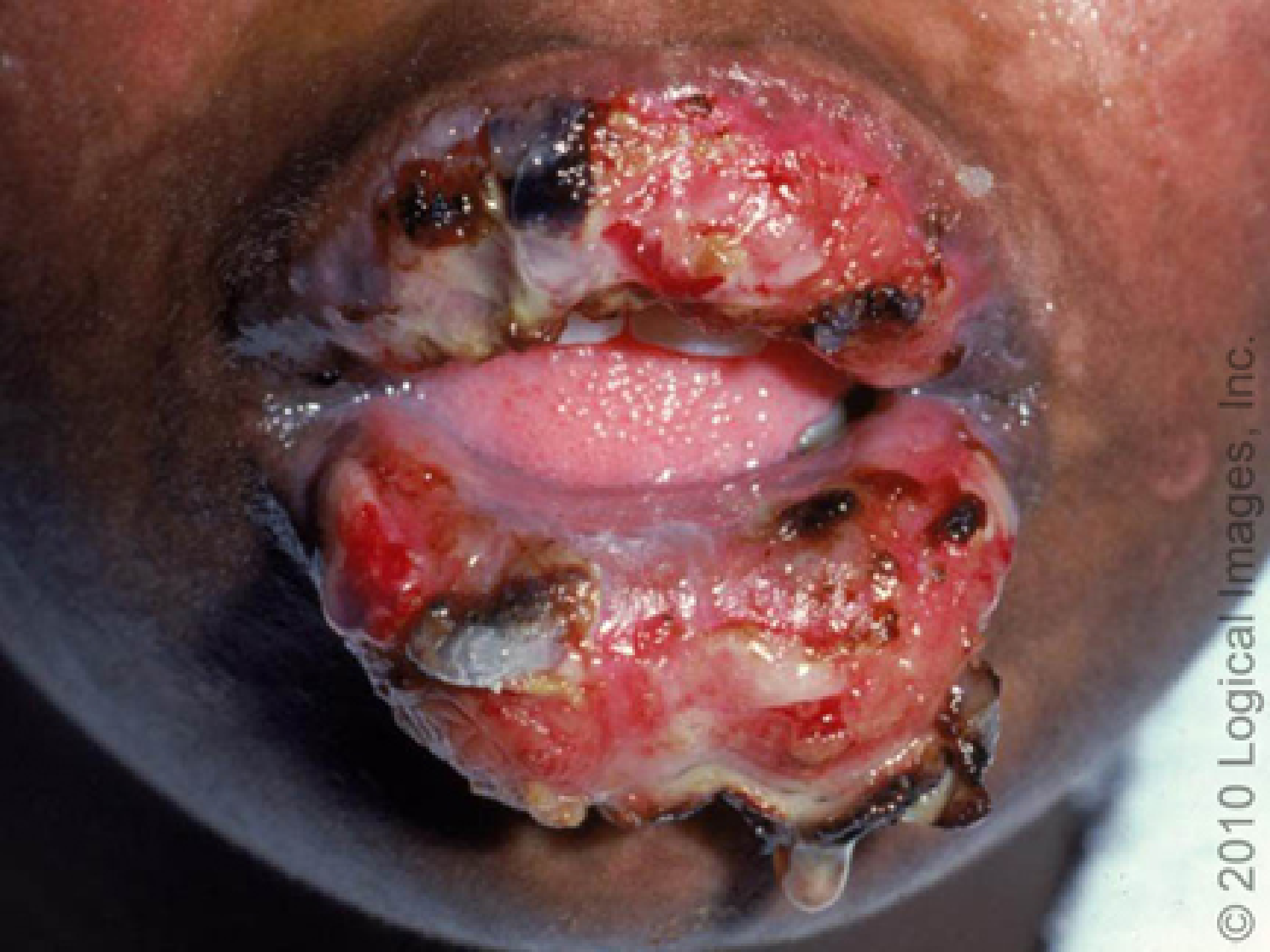
- MUCOCUTANEOUS DRUG REACTIONS
- DIFFUSE KERATINOCYTE APOPTOSIS
- PERCENT OF BODY SURFACE AREA (BSA) DETERMINES WHETHER IT IS SJS OR TEN
- LIKELY REPRESENT A SPECTRUM OF THE SAME RARE, ADVERSE IMMUNOLOGIC REACTION
- MORTALITY: SJS (1-5%), TEN (25-35%)
- DRUGS: SULFONAMIDE ANTIBIOTICS AND ANTICONVULSANTS

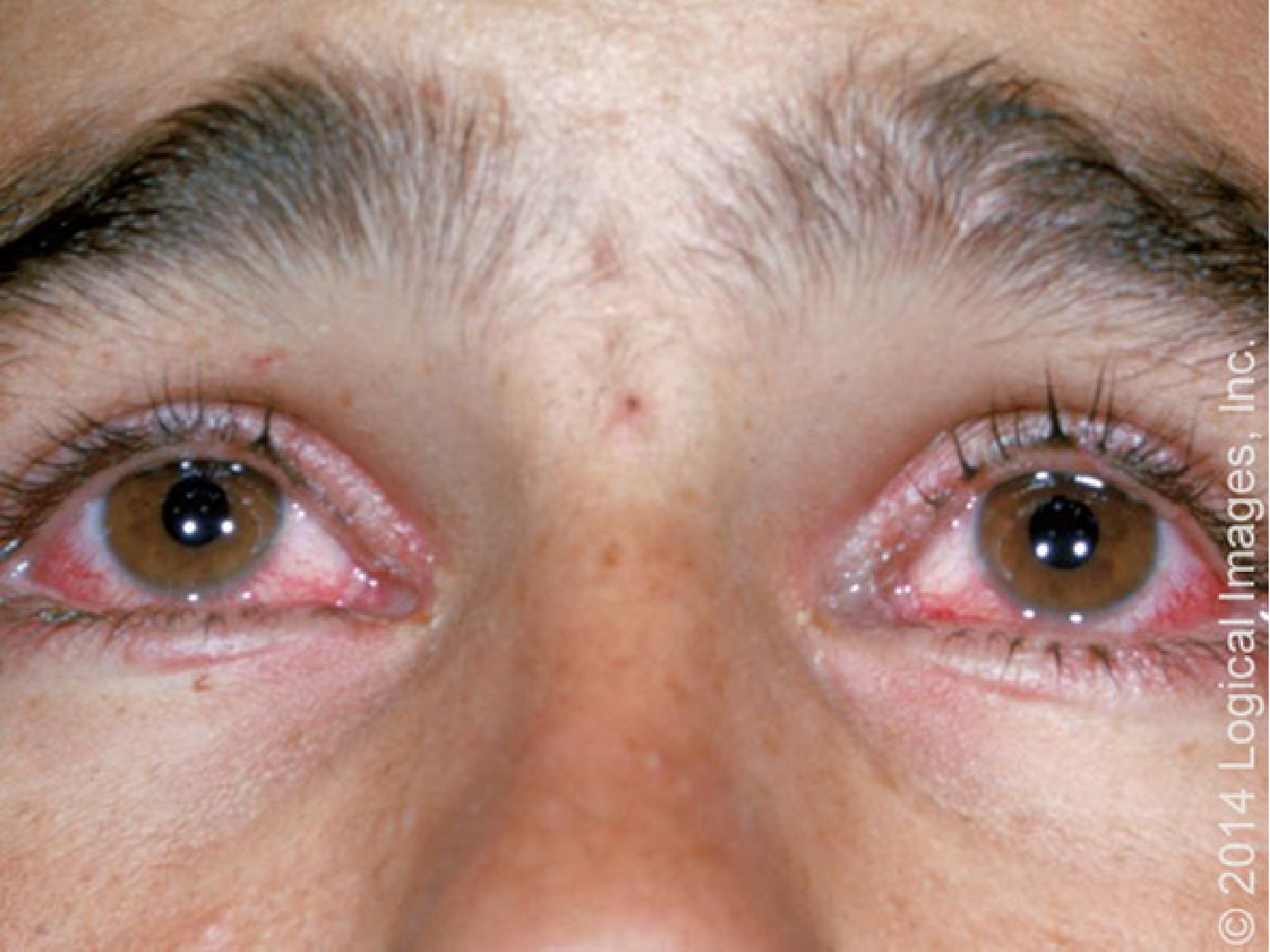
# STEVEN JOHNSON SYNDROME/TOXIC EPIDERMAL NECROLYSIS

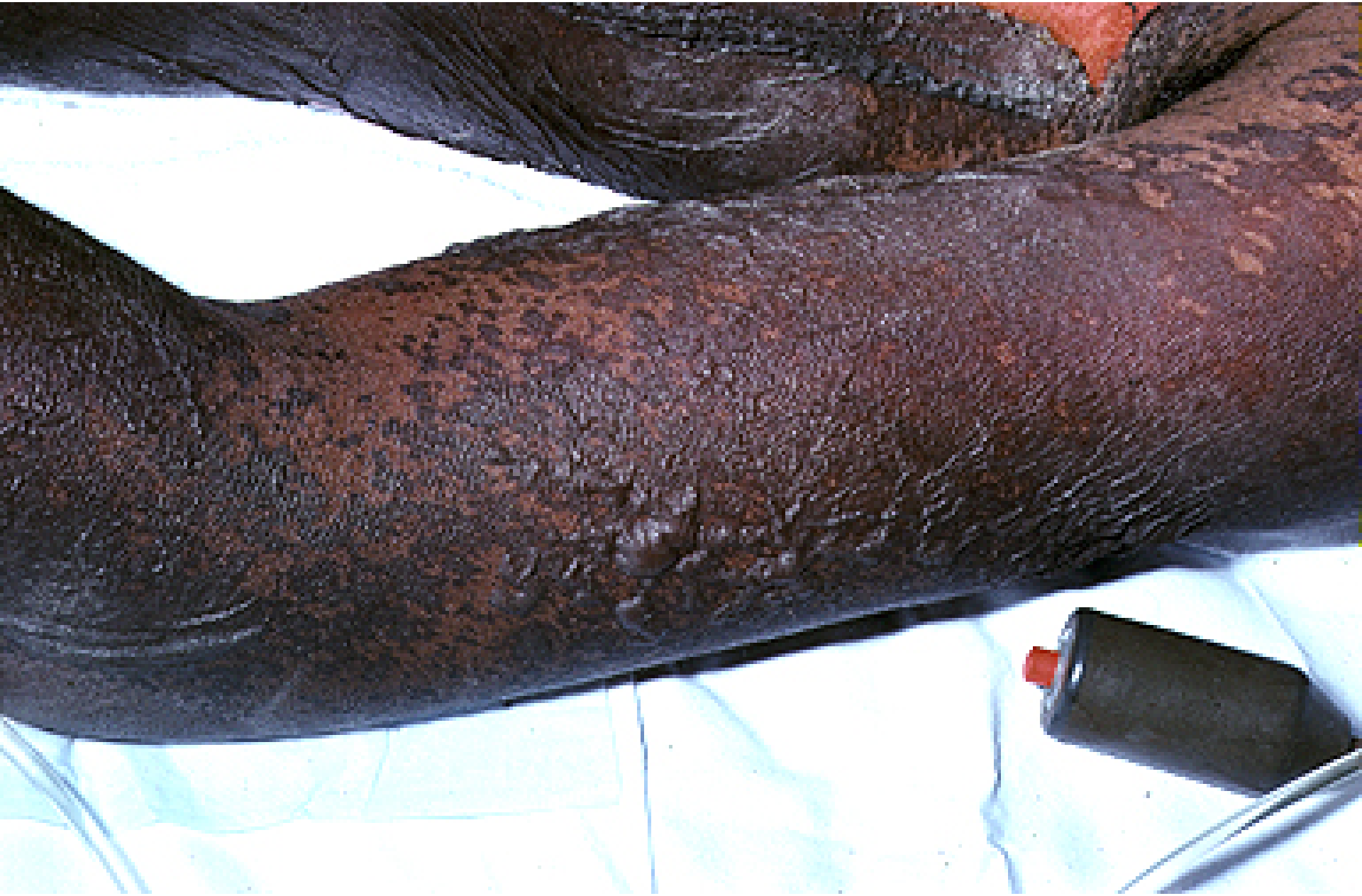
- OCCURS 1-3 WEEKS AFTER STARTING OFFENDING MED
- INITIAL: FEVER, DYSPHAGIA, BURNING SENSATION OF EYES
- DIFFUSE SKIN PAIN
- DUSKY, ATYPICAL TARGETOID SKIN LESIONS AND MUCOSAL EROSIONS (AT LEAST TWO)
- BULLAE AND EPIDERMAL SLOUGHING
- AIRWAY INVOLVEMENT CAN OCCUR

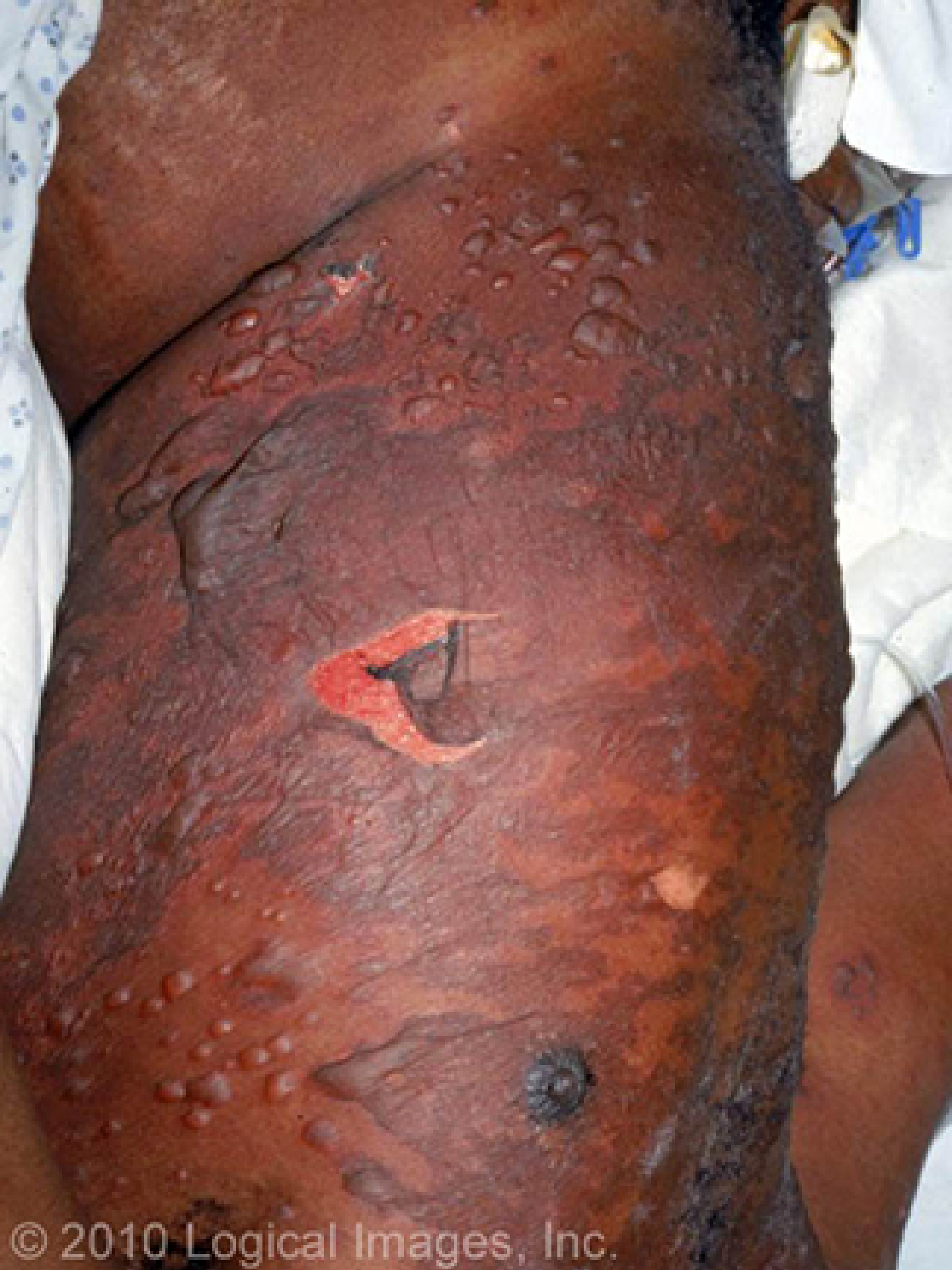


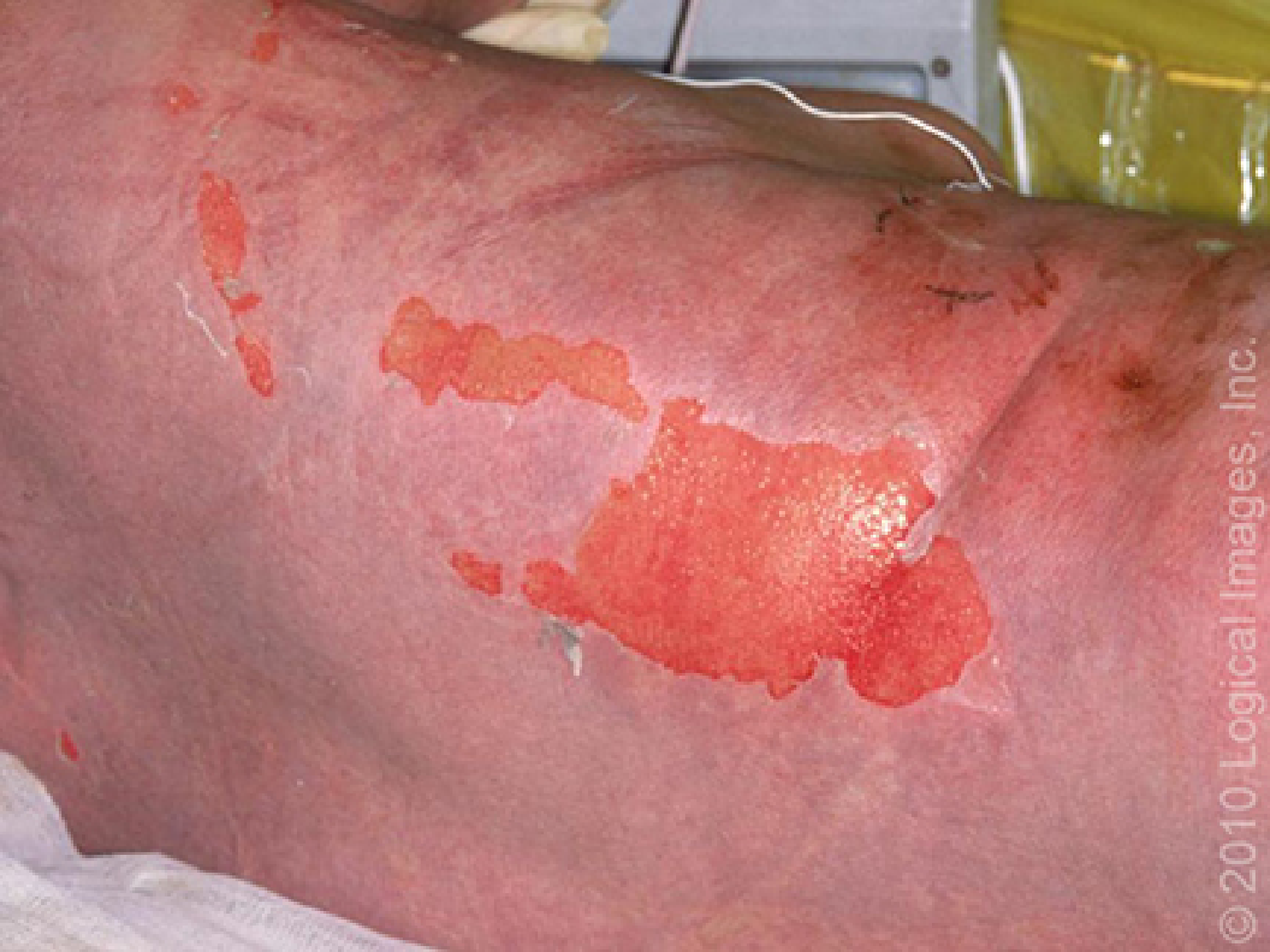












# STEVEN JOHNSON SYNDROME/TOXIC EPIDERMAL NECROLYSIS

- CLINICOPATHOLOGIC DIAGNOSIS
- NIKOLSKY'S SIGN
- SKIN BIOPSY (FROZEN SECTION PROCESSING OR OVERNIGHT PERMANENT): FULL THICKNESS EPIDERMAL NECROSIS WITH MINIMAL DERMAL INFLAMMATION
- LABORATORY WORK-UP BASED ON PATIENT'S UNDERLYING MEDICAL HISTORY, PHYSICAL EXAM AND CONCERN FOR OTHER ORGAN INVOLVMENT

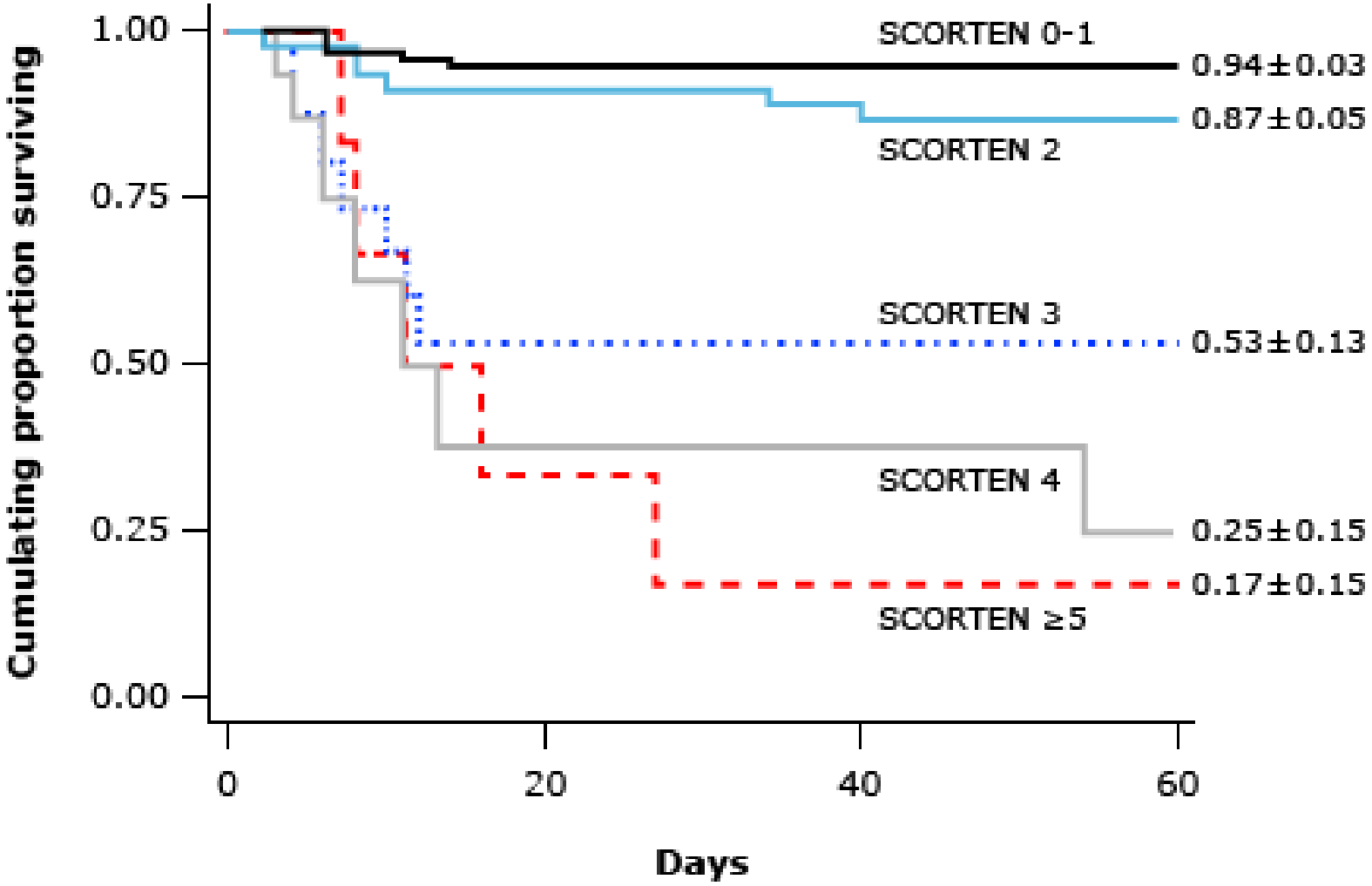
## SCORTEN score for Stevens-Johnson syndrome/toxic epidermal necrolysis

| Independent prognosis factors | | Weight |
|---|---|---|
| Age | ≥40 years | 1 |
| Malignancy* | Yes | 1 |
| Body surface area detached | ≥10% | 1 |
| Tachycardia | ≥120/min | 1 |
| Serum urea | >10 mmol/L | 1 |
| Serum glucose | >14 mmol/L | 1 |
| Serum bicarbonate | <20 mmol/L | 1 |
| **SCORTEN#** | | **7** |

SCORTEN: score of toxic epidermal necrolysis.
* Malignancy: evolving cancer and hematologic malignancies.

*Adapted with permission from: Guégan S, Bastuji-Garin S, Poszepczynska-Guigné E, et al. Performance of the SCORTEN during the first five days of hospitalization to predict the prognosis of epidermal necrolysis. J Invest Dermatol 2006; 126:272. Copyright © Macmillan Publishers Ltd.*

# SJS/TEN SCORETEN CRITERIA



# STEVEN JOHNSON SYNDROME/TOXIC EPIDERMAL NECROLYSIS

- SJS <10% BSA
- SJS/TEN OVERLAP 10-30% BSA
- TEN >30% BSA
- TREATMENT:
  - DISCONTINUE OFFENDING MEDICATION AND SUPPORTIVE CARE
  - CONSIDER BURN UNIT, BUT NEEDS TO BE FAMILIAR WITH THIS CONDITION
    - THERMAL REGULATION, WOUND CARE, FLUID REPLACEMENT, NUTRITION
  - OPHTHALMOLOGY
  - IVIG, PREDNISONE, CYCLOSPORINE

# Niklosky's sign is positive in Staphylococcal Scalded Skin Syndrome.

1. TRUE
2. FALSE

# STAPHYLOCOCCAL SCALDED SKIN SYNDROME

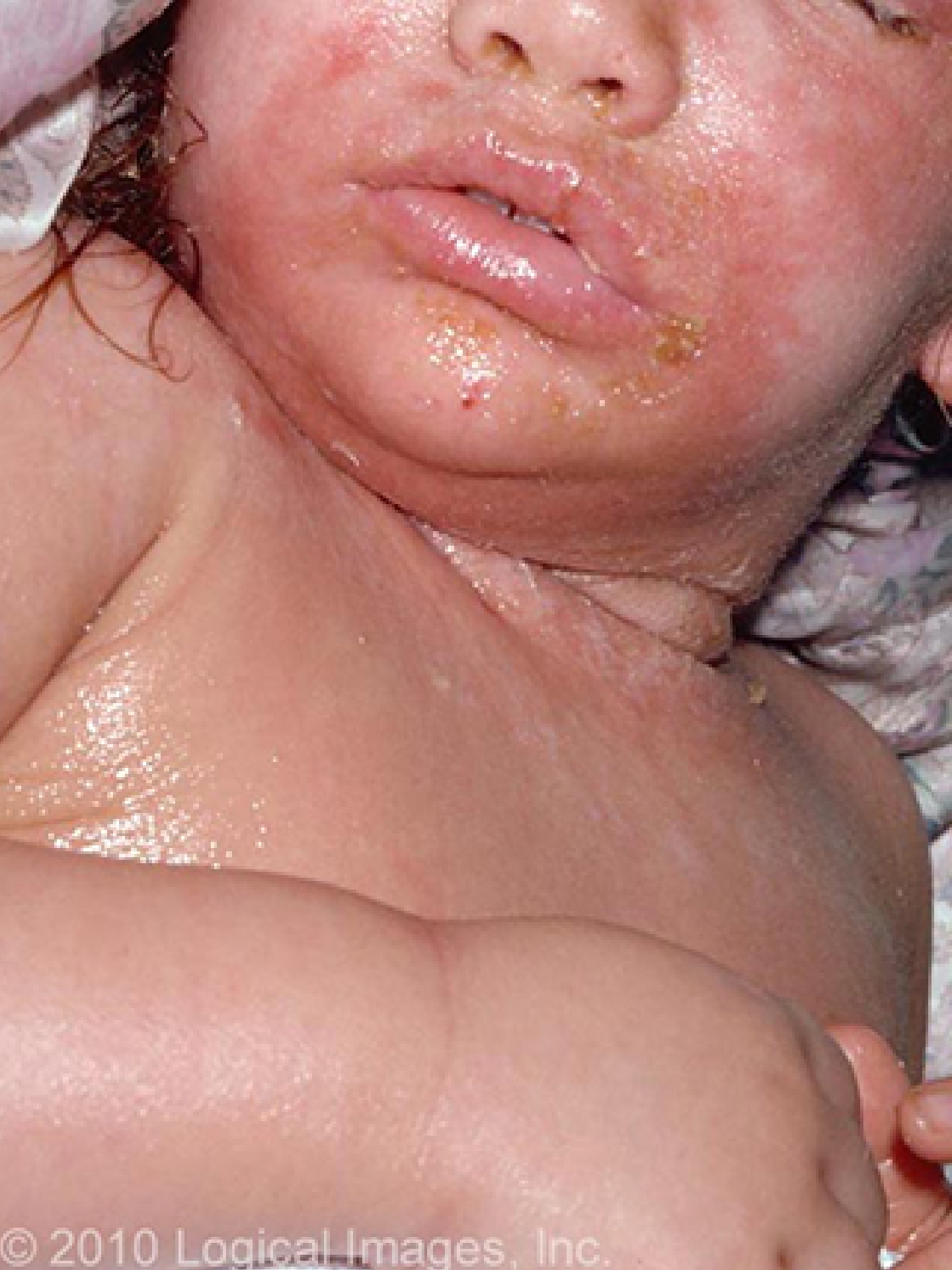
- AKA. RITTER'S SYNDROME
- SUPERFICIAL, BLISTERING SKIN DISEASE CAUSED BY LOCALIZED INFECTION WITH STAPH STRAIN THAT ELABORATES STAPYLOCOCCAL EXFOLIATIVE TOXIN (ET)
- PREVALENCE: NEONATES/CHILDREN >> ADULTS; MORTALITY: ADULTS >> CHILDREN/NEONATES

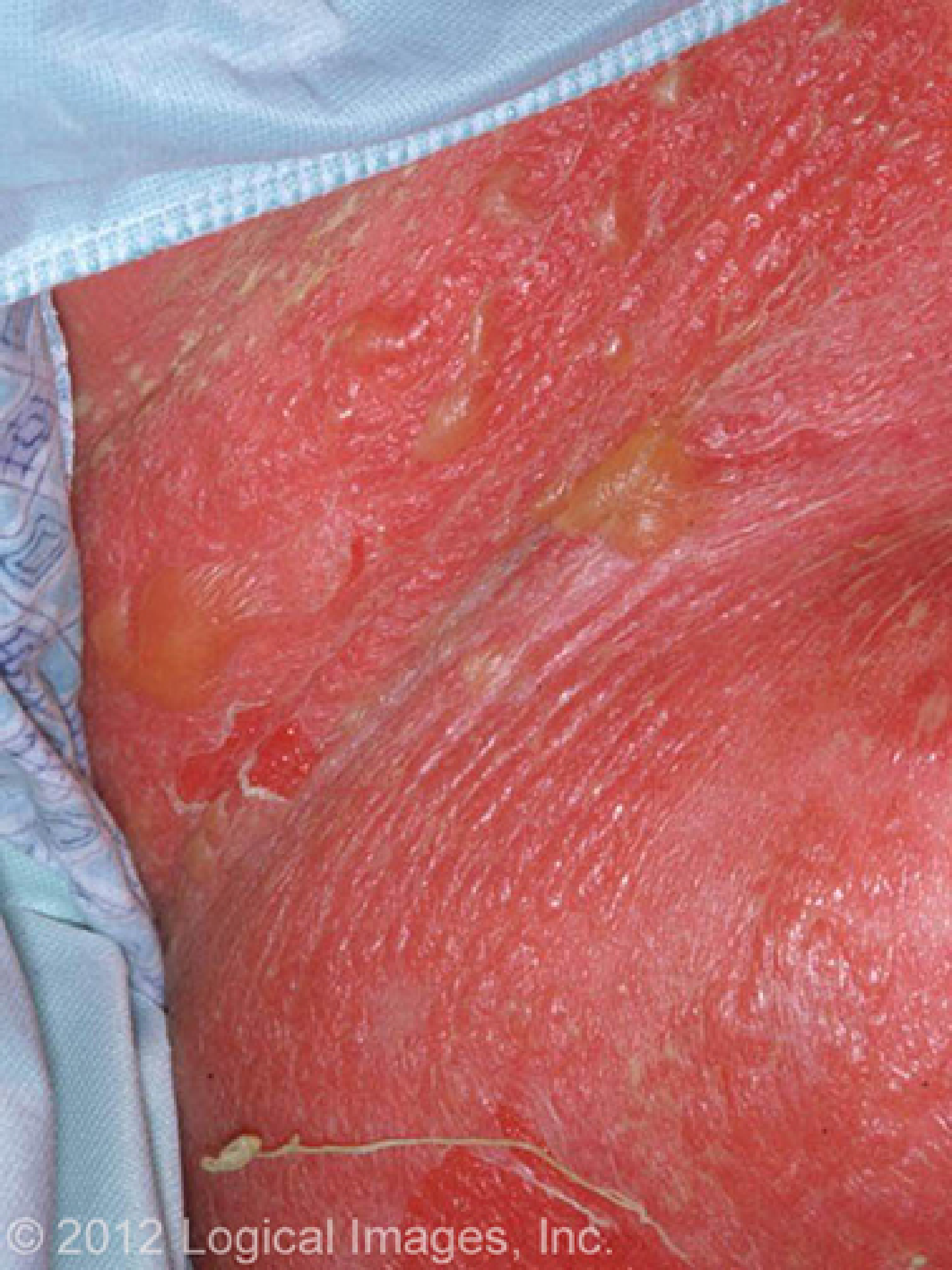
# STAPHYLOCOCCAL SCALDED SKIN SYNDROME

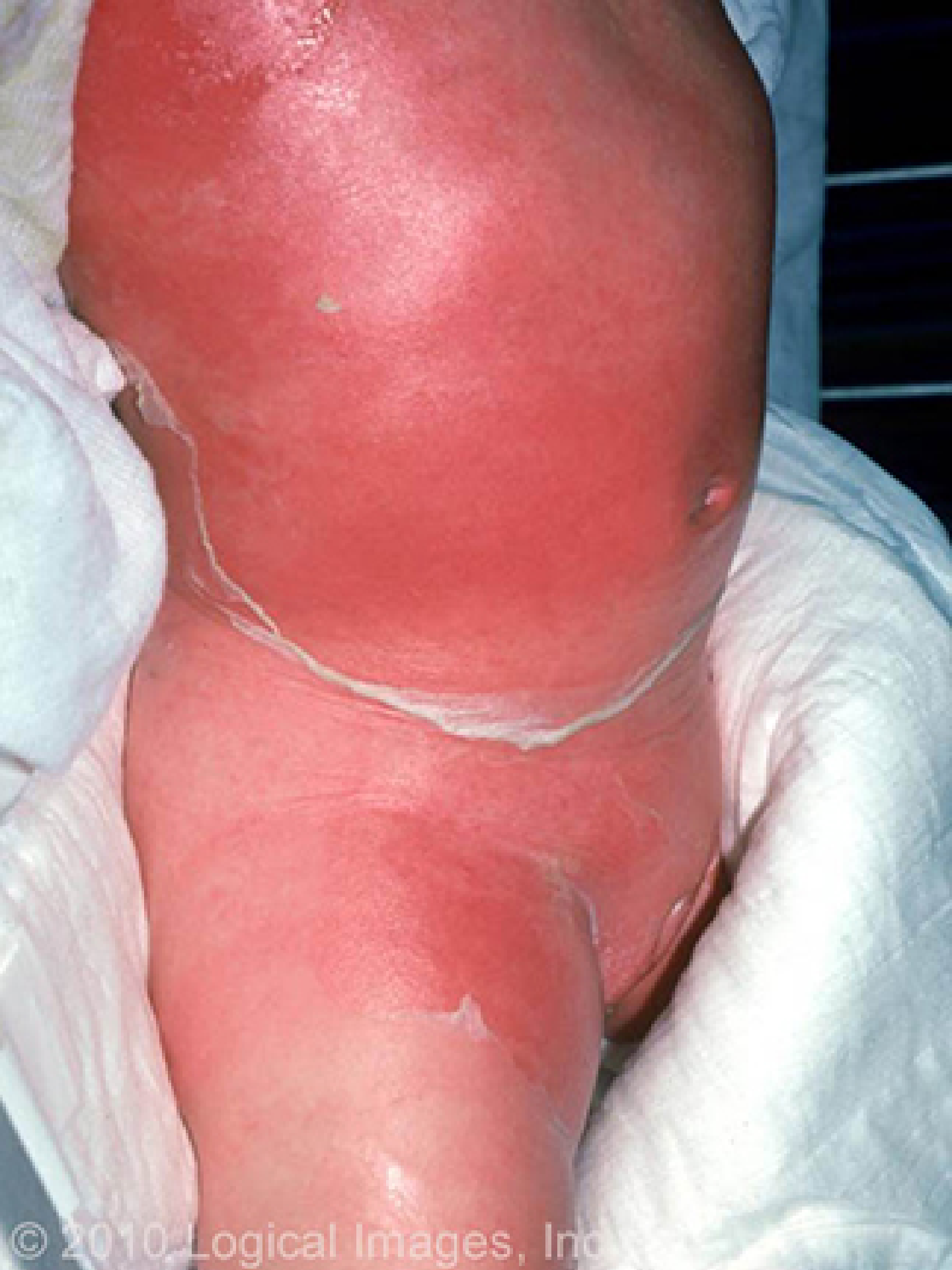
- HEMATOGENOUS DISSEMINATION OF ETs PRODUCED BY A LOCALIZED INFECTION WITH PHAGE GROUP II STRAINS OF *S. AUREUS*
- ETs ARE SERINE PROTEASES THAT BIND AND CLEAVE DESMOGLEIN 1 WHICH IS A COMPONENT OF DESMOSOMES IN THE EPIDERMIS (LOCATED IN GRANULAR LAYER) WHICH IS WHY THE BULLAE ARE SUPERFICIAL
- NIKOLSKY'S SIGN (DISLODGEMENT OF INTACT SUPERFICIAL EPIDERMIS WITH SHEARING FORCE) IS POSITIVE.

# STAPHYLOCOCCAL SCALDED SKIN SYNDROME

- PRIMARY INFECTION: CONJUNCTIVA, EAR, URINARY TRACT OR SKIN
- FEVER AND MALAISE FOLLOWED BY ERYTHRODERMA
- 24-48 HOURS: FLACCID BULLAE IN FLEXURAL SURFACES AND AROUND ORIFICES (ESP. EYES AND MOUTH); BULLAE ARE VERY THEN SO IT MAY PRESENT AS “CRINKLED” SKIN OR DESQUAMATION
- SUPERFICIAL BLISTERING = NO SCARRING







# STAPHYLOCOCCAL SCALDED SKIN SYNDROME

- DDX INCLUDES SJS/TEN, SCARLET FEVER AND STAPH TOXIC SHOCK SYNDROME
- DIAGNOSIS:
  - 1) APPROPRIATE CLINICAL PRESENATION WITH EITHER ERYTHRODERMA, SKIN DESQUAMATION, OR BULLAE
  - 2) ISOLATION OF S. AUREUS STRAIN THAT PRODUCES ET
  - 3) CHARACTERISTIC HISTOPATHOLOGY OF INTRAEPIDERMAL CLEAVAGE AT THE GRANULAR LAYER

# STAPHYLOCOCCAL SCALDED SKIN SYNDROME

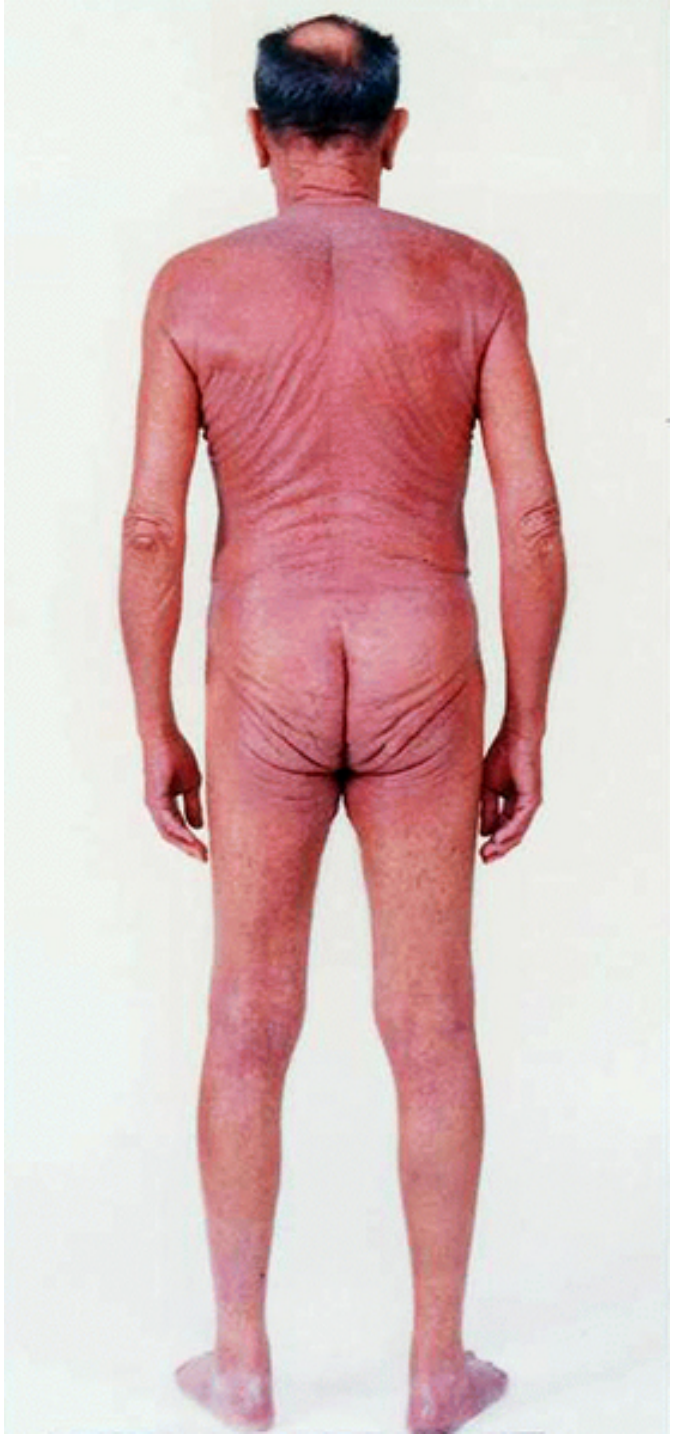
- TREATMENT:
  - HOSPITALIZATION
  - SYSTEMIC ANTIBIOTICS—IV PENICILLINASE-RESISTANT PENICILLIN
  - LOCAL WOUND THERAPY WITH BARRIER OINTMENTS AND COMPRESSES
  - IF MULT. OCCURENCES, CONSIDER COLONIZATION WORK UP AND SUBSEQUENT DECOLONIZATION THERAPY

# Erythroderma is typically a primary skin disease.

1. TRUE
2. FALSE

# ERYTHRODERMA

- GENERALIZED SKIN REDNESS AND SCALING
- POTENTIAL PRESENTATION OF A VARIETY OF DISEASES
- IT IS AN EMERGENCY IN CERTAIN DUE TO THE FOLLOWING:
  - FLUID SHIFTS
  - TACHYCARDIA
  - DISTURBANCES IN THERMOREGULATION
  - CARDIAC FAILURE



# ERYTHRODERMA

- STAPHYLOCOCCAL SCALDED SKIN SYNDROME
- CONTACT AND AUTOSENSITIZATION DERMATITIS
- ATOPIC DERMATITIS
- LEUKEMIA/LYMPHOMA/PARANEOPLASTIC
- DRUG (INCLUDES DRESS)
- PSORIASIS
- IDIOPATHIC
- MYCOSIS FUNGOIDES (CUTANEOUS T-CELL LYMPHOMA)—SEZARY SYNDROME
- PITYRIASIS RUBRA PILARIS

# ERYTHRODERMA

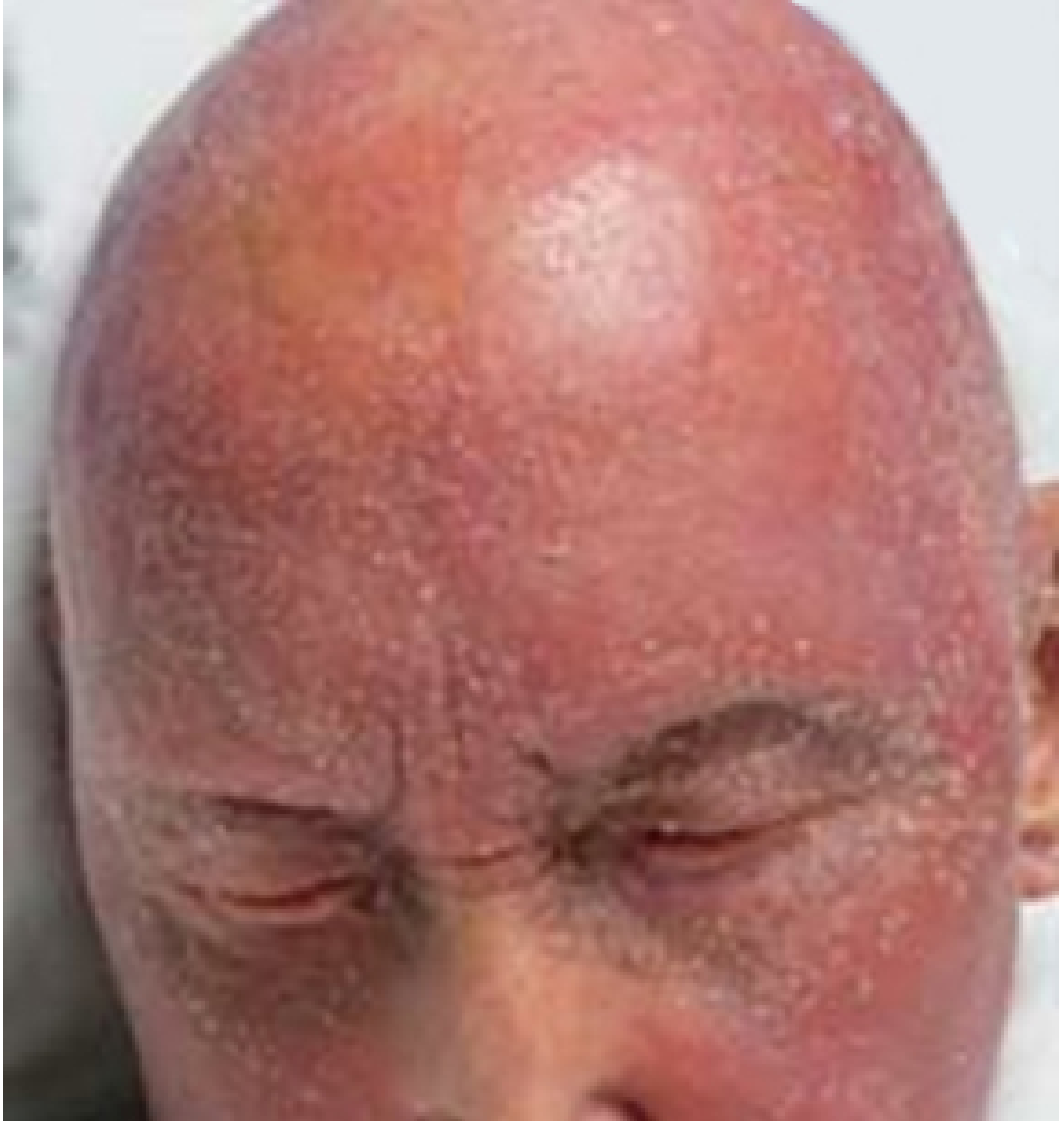
- TREATMENT REGARDLESS OF UNDERLYING DISEASE
  - NUTRITIONAL ASSESSMENT
  - CORRECTION OF FLUID AND ELECTROLYTE IMBALANCES
  - HYPOTHERMIA PREVENTION
  - TREATMENT OF SECONDARY INFECTIONS

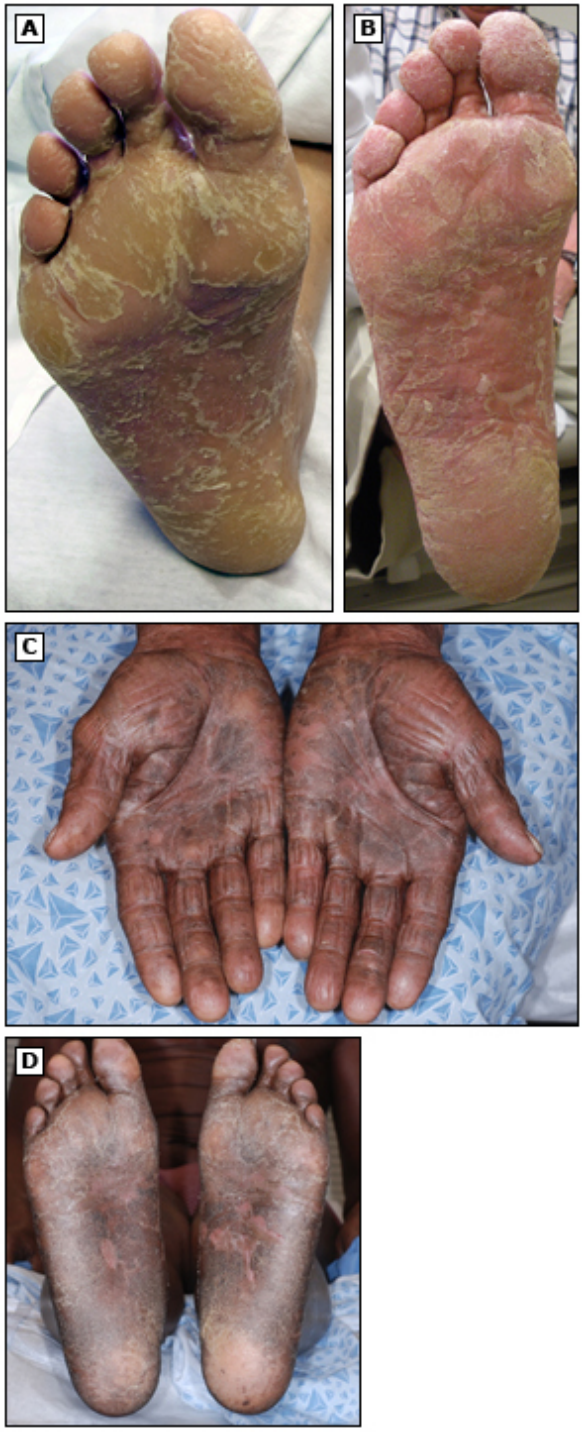
# Sezary Syndrome is secondary to an underlying B-cell lymphoma.

1. TRUE
2. FALSE

# SEZARY SYNDROME

- AGGRESSIVE LEUKEMIC VARIANT OF CUTANEOUS T-CELL LYMPHOMA (AKA. MYCOSIS FUNGOIDES)
- TRIAD
  - ERYTHRODERMA
  - LYMPHADENOPATHY
  - SEZARY CELLS (ATYPICAL LYMPHOCYTES) IN THE PERIPHERAL BLOOD




A
B
C
D

# SEZARY SYNDROME

- STILL QUESTION WHETHER THIS CAN OCCUR DE NOVO OR IT IS PROGRESSION OF EXISTING CTCL
- WORK-UP INCLUDES:
  - CBC, CMP, LDH, MANUAL SEZARY COUNT, FLOW CYTOMETRY
  - SKIN BIOPSY
  - CT SCANS OF CHEST/ABDOMEN/PELVIS FOR STAGING PURPOSES

# SEZARY SYNDROME

- VERY POOR PROGNOSIS WITH A 5-YEAR SURVIVAL OF 25%; MOST PATIENTS DIE OF AN OPPORTUNISTIC INFECTION FROM IMMUNOSUPPRESSION
- TREATMENT IS EXTRACORPOREAL PHOTOPHORESIS ALONE OR IN COMBINATION WITH OTHER MODALITY (CHEMOTHERAPY, RETINOIDS, METHOTREXATE, INTERFERON, DENILEUKIN DIFTITOX)

# Pain out of proportion to the clinical exam is more common with necrotizing fasciitis than with cellulitis.

1. TRUE
2. FALSE

# NECROTIZING FASCIITIS

- RARE WITH 1,000 CASES ANNUALLY IN THE U.S.
- HOWEVER, THERE IS SIGNIFICANT MORBIDITY AND MORTALITY WITH THE LATTER APPROACHING 60%

# NECROTIZING FASCIITIS

- TRAUMATIC/IATROGENIC INOCULATION OR HEMATOGENOUS SEEDING OF THE FASCIA
- CLASSIFICATION BASED ON ORGANISM
  - TYPE I: POLYMICROBIAL
  - TYPE II: GROUP A STREP PYOGENES
  - TYPE III: MARINE VIBRIO
- RISK FACTORS
  - ADVANCED AGE
  - DIABETES
  - IMMUNNOSUPPRESSION
  - PERIPHERAL VASCULAR DISEASE
  - SURGERY
  - TRAUMA

# NECROTIZING FASCIITIS

- TRUNK, GROIN, HEAD, NECK, EXTREMITIES
- EARLY SIGNS: FEVER, ERYTHEMA EDEMA
- TYPICAL DDX IS CELLULITIS WHICH CAN LEAD TO DELAY OF APPROPRIATE TREATMENT; IT IS IMPORTAN TO MAKE THE DISTINCTION

# EARLY SIGNS TO DIFFERENTIATE NECROTIZING FASCIITIS AND CELLULITS

**NECROTIZING FASCIITIS**

- PAIN OUT OF PROPORTION TO EXAM
- RAPIDLY PROGRESSING ERYTHEMA
- MYALGIA
- HISTORY OF A PUNCTURE INJURY
- BULLAE

**CELLULITIS**

- MILD TO MODERATE PAIN
- SLOWLY PROGRESSING ERYTHEMA
- MYALGIA NOT COMMON
- BULLAE DO NOT OCCUR EARLY

# LATE SIGNS TO DIFFERENTIATE NECROTIZING FASCIITIS AND CELLULITS

**NECROTIZING FASCIITIS**

- CREPITUS
- HYPOESTHESIA OF SKIN
- SKIN NECROSIS

**CELLULITIS**

- CREPITUS NOT SEEN
- PAIN MORE TYPICAL
- SKIN NECROSIS DOES NOT OCCUR

# NECROTIZING FASCIITIS

- DIAGNOSIS
  - SURGICAL DEBRIDEMENT WITH TISSUE BIOPSY AND CULTURE
  - MRI
  - LABORATORY RISK INDICATOR FOR NECROTIZING FASCIITIS
    - C-REACTIVE PROTEIN
    - WHITE BLOOD CELL COUNT
    - HEMOGLOBIN
    - SODIUM
    - CREATININE
    - GLUCOSE

# NECROTIZING FASCIITIS

- TREATMENT
  - URGENT AND EXTENSIVE SURGICAL DEBRIDEMENT WITH POSSIBLE LIMB AMPUTATION
  - APPROPRIATE ANTIBIOTIC THERAPY
    - CLINDAMYCIN (GROUP A STREP)
    - AMPICILLIN AND GENTAMICIN (GRAM NEGATIVE)

# Skin lesions of meningococcemia favor dependent areas of the body.

1. TRUE
2. FALSE

# MENINGOCOCCEMIA

- CAUSED BY GRAM-NEGATIVE, ENCAPSULATED DIPLOCOCCUS *NEISSERIA MENINGITIDES*

- CHILDREN>>ADOLESCENTS>YOUNG ADULTS; MALES > FEMALES
- MORTALITY 10-14%

# MENINGOCOCCEMIA

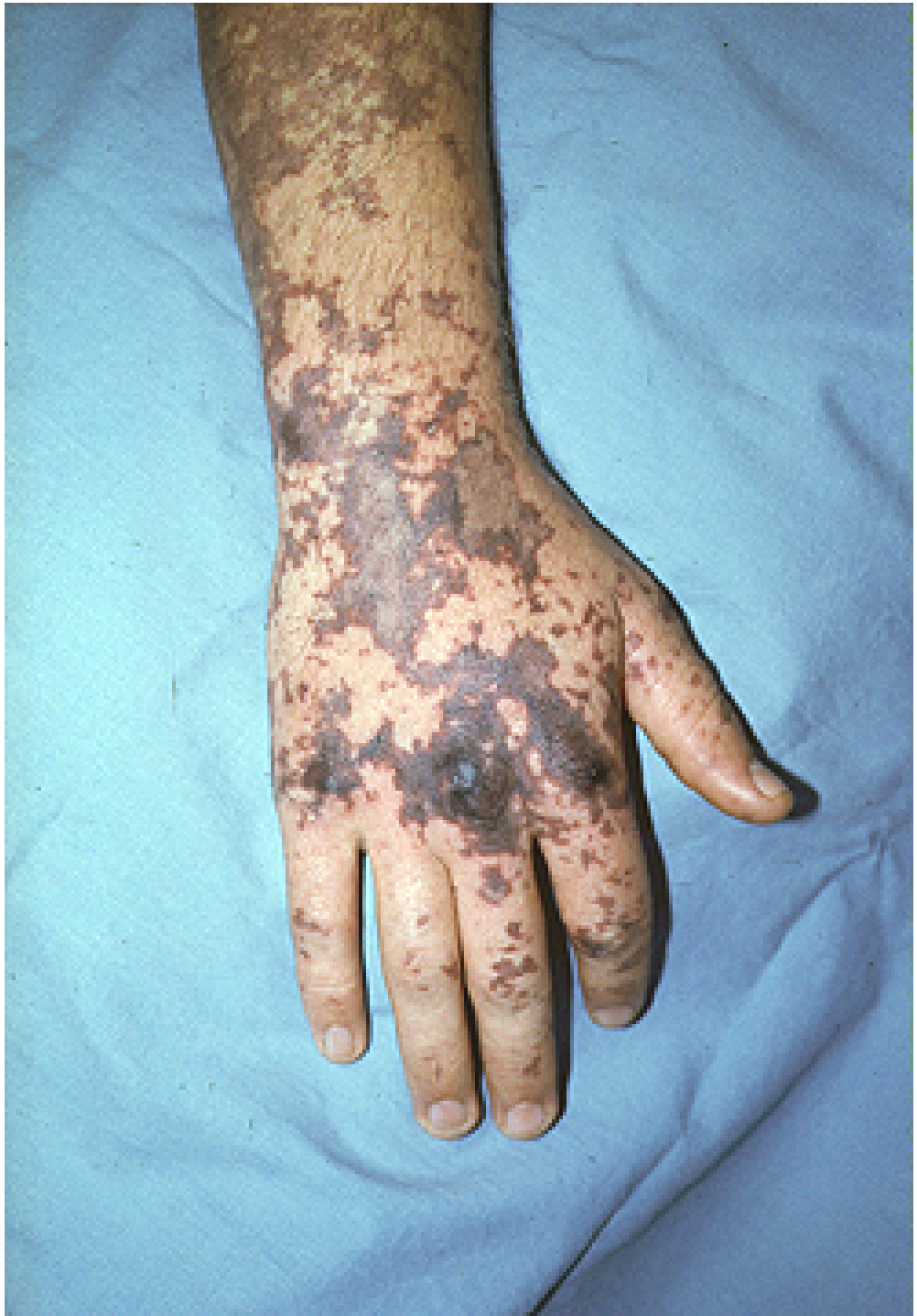
- TRANSMISSION: RESPIRATORY
- RISK FACTORS
  - CROWDING
  - SMOKING
  - COMPLEMENT DEFICIENCY
  - ASPLENIA
- SEROTYPES OF N. MENINGITIDES
  - A, C: EPIDEMICS
  - B: SPORADIC

# MENINGOCOCCEMIA

- TYPICAL: FEVER, HEADACHE, FLU-LIKE SYMPTOMS
- 50% PRESENT WITH FEATURES OF FRANK MENINGITIS
- MYALGIAS IS OFTEN INTENSE
- CUTANEOUS SIGNS ARE OFTEN EARLIEST
  - PETECHIAL LESIONS OF THE TRUNK, LOWER EXTREMITIES AND PRESSURE AREAS SUCH AS THE WAISTLINE

# MENINGOCOCCEMIA

- PROGRESSION CAN BE RAPID AND DIFFUSE
- PETECHIA PROGRESS TO CLASSIC MORPHOLOGY OF PURPURA WITH AN ANGULATED, ERYTHEMATOUS BORDER WITH A “GUN METAL GRAY” COLOR CENTRALLY.
- DIFFUSE DISEASE IS PURPURA FULMINANS AND IS ASSOCIATED WITH DISSEMINATED INTRAVASCULAR COAGULATION AND MULTI-ORGAN FAILURE.



# MENINGOCOCCEMIA

- DDX:
  - VASCULITIS
  - CATASTROPHIC ANTIPHOSPHOLIPID ANTIBODY SYNDROME
  - TOXIC SHOCK SYNDROME
  - RMSF (REMEMBER DISTRIBUTION/PROGRESSION OF THE RASH)
  - ENDOCARDITIS

# Mortality from DRESS is a typically a result of kidney failure.

1. TRUE
2. FALSE

# DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS)

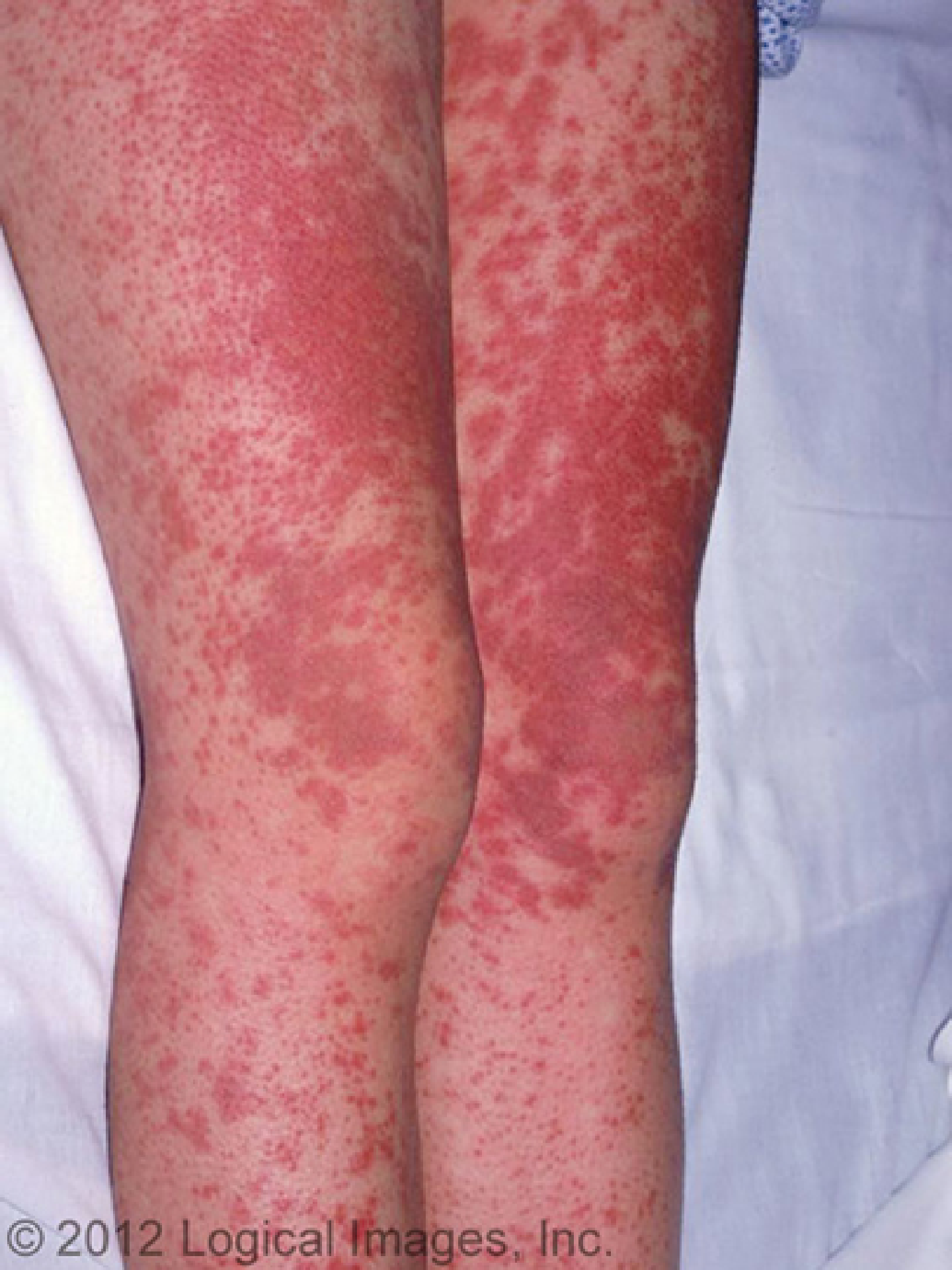
- AKA. DRUG-INDUCED HYPERSENSITIVITY SYNDROME
- IDIOSYNCRATIC DRUG REACTION THAT AFFECTS THE SKIN AND VARIOUS INTERNAL ORGANS; CAN BE FATAL.
- 1 IN 1,000 TO 1 IN 10,000 DRUG EXPOSURES; TYPICALLY FIRST EXPOSURE.

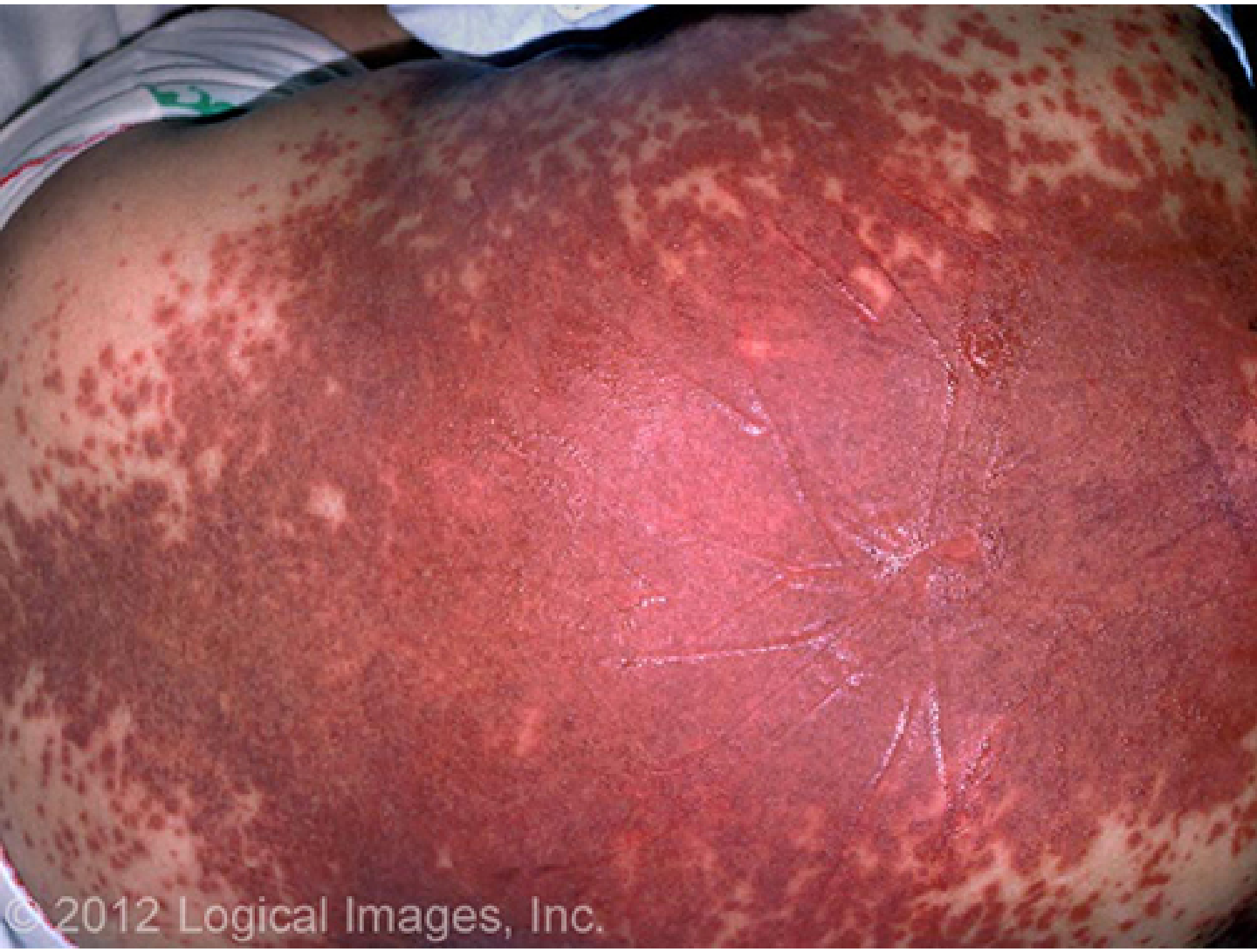
# DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS)

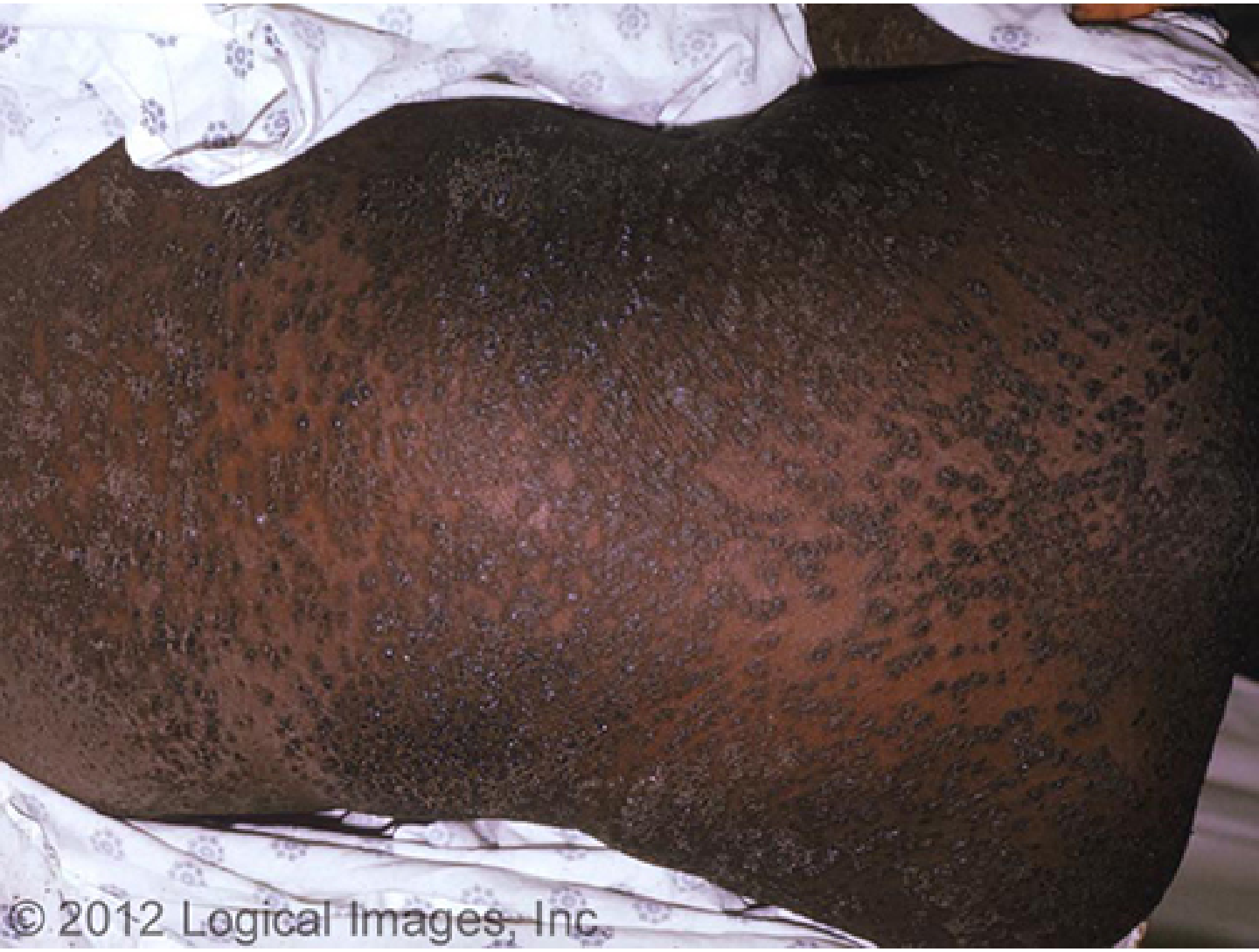
- CLASSICALLY ASSOCIATED DRUGS:
  - AROMATIC ANTICONVULSANTS
    - CARBAMAZEPINE, PHENYTOIN, PHENOBARBITAL
  - XANTHINE-OXIDATE INHIBITORS
    - ALLOPURINOL
  - SULFONAMIDES ANTIBOTICS
    - TRIMETHOPRIN-SULFAMETHOXASOLE
  - OTHER ANTIBIOTICS
    - MINOCYCLINE, DAPSONE
  - AMINOSALICYLATES
    - SULFASALZINE
  - REVERSE TRANSCRIPTASE INHIBITORS
    - ABACAVIR

# DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS)

- DRESS HAS A LONG LATENCY PERIOD (>2-3 WEEKS) FROM START OF MEDICATION TO ONSET OF SYMPTOMS
- CLASSIC TRIAD
  - 1. RASH (75%)
    - CENTRAL FACIAL ERYTHEMA AND EDEMA
  - 2. FEVER (85%)
  - 3. INTERNAL ORGAN INVOLVEMENT
    - GENERALIZED LAD, LEUKOCYTOSIS, ABNORMAL LFTs, PERIPHERAL EOSINOPHILIA (>50% BUT NOT REQUIRED FOR DIAGNOSIS)






© 2012 Logical Images, Inc.

# DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS)

- LIVER IS MOST COMMON END ORGAN INVOLVED AND FULMINANT HEPATITIS IS RESPONSIBLE FOR MAJORITY OF DEATHS DUE TO DRESS

# DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS): regiSCAR DIAGNOSTIC CRITERIA

- REQUIRED
  - 1. HOSPITALIZATION
  - 2. ACUTE RASH
  - 3. SUSPICION OF A DRUG REACION
- PLUS 3 OF 4
  - 1. FEVER >38 deg C
  - 2. AT LEAST TWO SITES OF LYMPHADENOPATHY
  - 3. AT LEAST ONE INTERNAL ORGAN INVOLVED
  - 4. HEMATOLOGIC ABNORMALITIES (LOW OR HIGH WBCs, EOSINOPHILIA AND LOW PLATELETS)

# DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS)

- TREATMENT
  - STOP OFFENDING MEDICATION
  - LONG-TAPER OF SYSTEMIC CORTICOSTEROIDS
  - IF DUE TO AROMATIC ANTI-CONVULSANT, SWITCH TO NONAROMATIC DUE TO CROSS-REACTIVITY
- LONG-TERM MONITORING FOR AUTOIMMUNE DISEASE, ESP. THYROID DISEASE

# The classic rash of Rocky Mountain Spotted Fever progresses in a central-to-distal fashion.

1. TRUE
2. FALSE

# ROCKY MOUNTAIN SPOTTED FEVER

- 90% OF CASES OCCUR BETWEEN APRIL AND SEP
- 1,900 CASES IN THE U.S. PER YEAR
- TICK BITE FROM *Dermacentor variabilis* AND LESS COMMONLY *Amblyomma americanum* AND *Dermacentor andersoni*
- BACTERIA: *Rickettsia rickettsi*

# IF YOU HAVE THE TICK AND REALLY WANT TO KNOW WHAT TYPE IT IS.

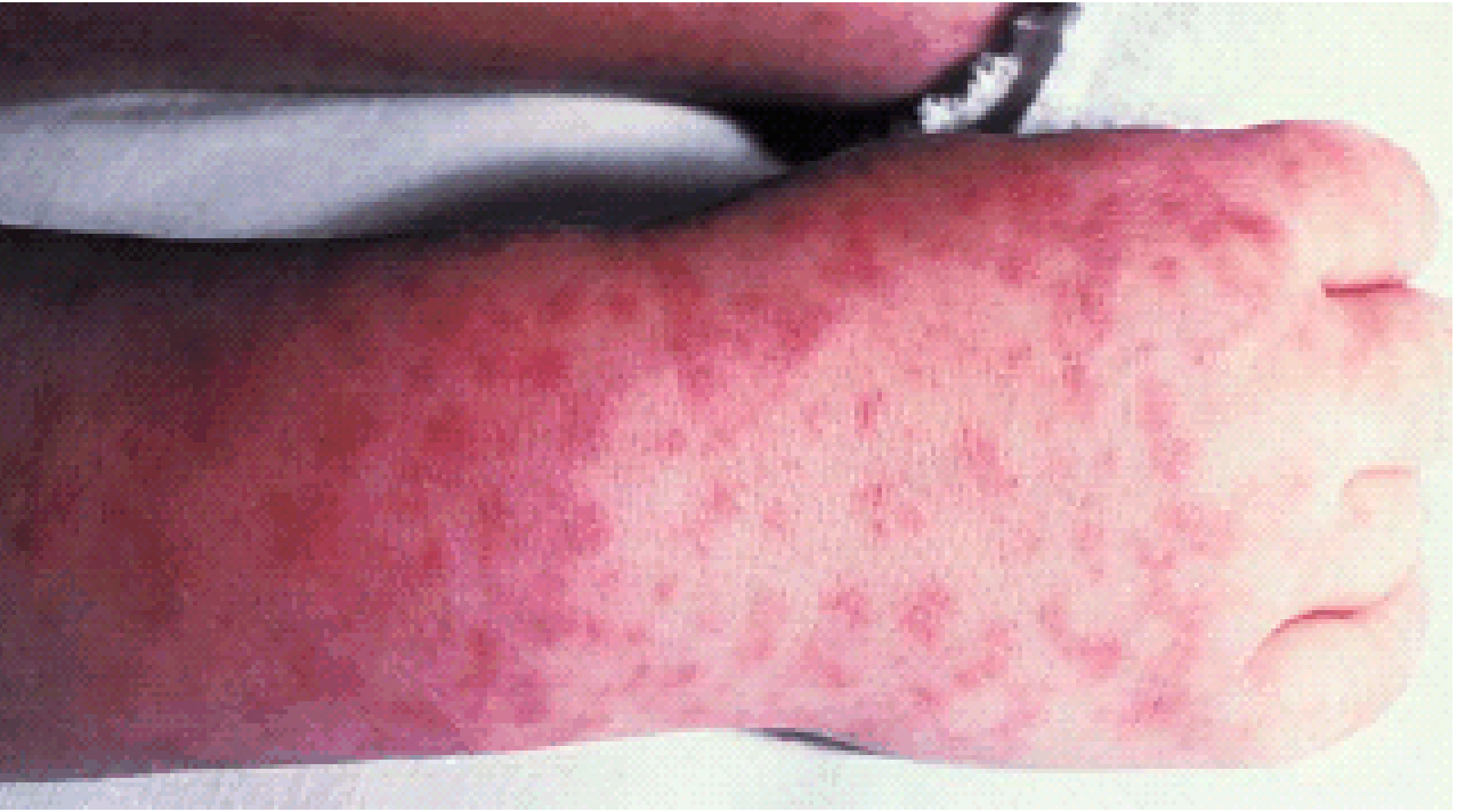
- https://phc.amedd.army.mil/topics/environhealth/epm/Pages/HumanTickTestKitProgram.aspx

# ROCKY MOUNTAIN SPOTTED FEVER

- BACTERIA INFECT ENDOTHELIAL CELLS AND CAUSES INCREASED VASCULAR PERMEABILITY LEADING TO:
  - EDEMA
  - HYPOVOLEMIA
  - HYPOTENSION
- MOST COMMON CAUSE OF DEATH IS END ORGAN DAMAGE TO THE LUNGS OR BRAIN

# ROCKY MOUNTAIN SPOTTED FEVER

- SYMPTOM ONSET = 1-14 DAYS, AVG. 5 DAYS
- FEVER, CHILLS, SEVERE HEADACHE, MALAISE
- SKIN RASH TYPICALLY OCCURS 2-5 DAYS AFTER FEVER ONSET
  - DISTAL (WRISTS, ANKLES, FOREHEAD) ERYTHEMATOUS MACULES
  - PROGRESSION TO PURPURA IN A DISTAL-TO-CENTRAL MANNER—KEY TO RMSF
- FATAL IN TREATED (5%) AND UNTREATED (25%)



# ROCKY MOUNTAIN SPOTTED FEVER

- TREATMENT: DOXYCYCLINE UNLESS PREGNANT OR UNDER AGE OF 9 IN WHICH CHLORAMPHENICOL IS USED
- PROMPT INITIATION OF TREATMENT IS IMPORTANT EVEN IN LIGHT OF ABSENCE OF DIAGNOSTIC TESTING AS ANY DELAY > 5 DAYS SIGNIFICANTLY INCREASES MORTALITY

# QUESTIONS OR COMMENTS.