

# Dermatomyositis and Cutaneous Lupus

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# Dermatomyositis

- Multisystem disease: skin, muscle, lung
- **Adult-onset:**
  - Classic DM
  - Classic DM + malignancy
  - Classic DM + overlap CT disease
  - **Clinically amyopathic DM** – amyopathic vs. hypomyopathic
- **Juvenile-onset:**
  - Classic DM
  - Clinically amyopathic DM – amyopathic vs. hypomyopathic

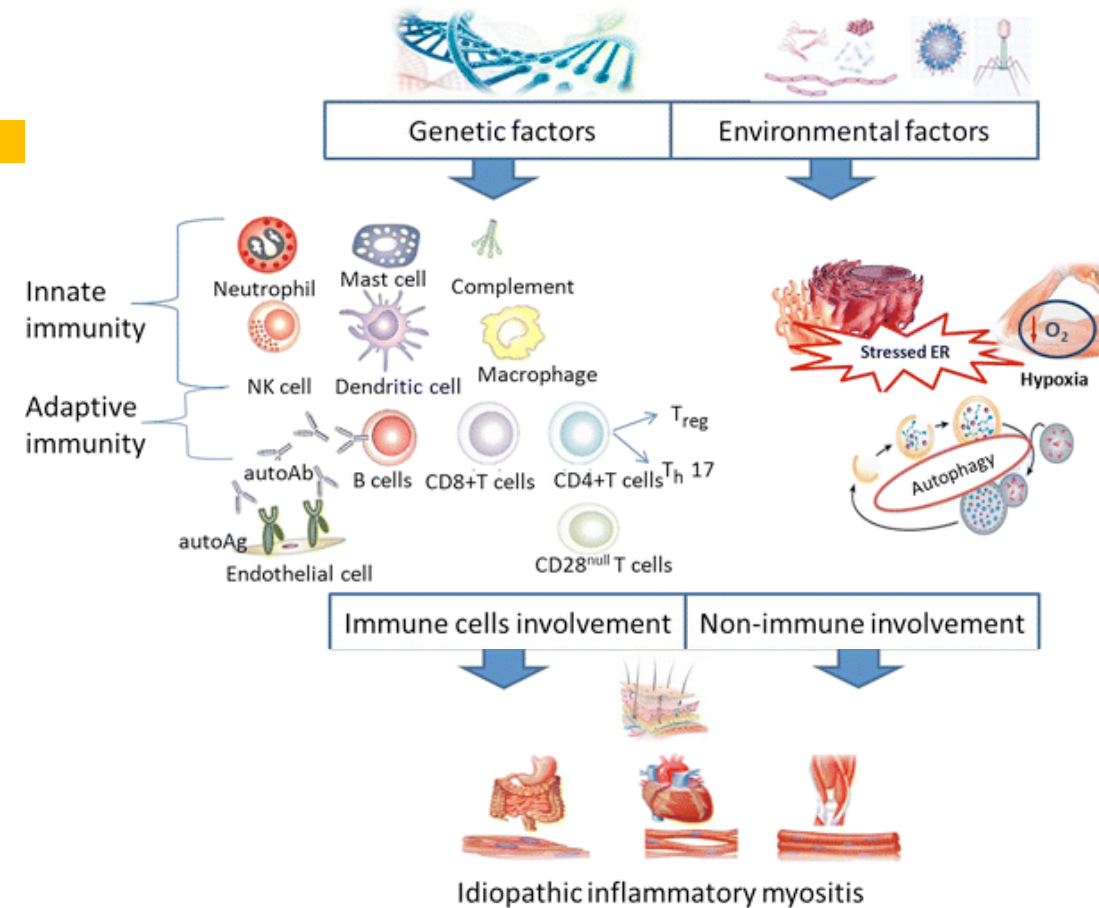


# Epidemiology

- Incidence: 2-9 / million
- Bimodal peak: kids & adults 40-65 yo
- F > M (2-3x)
- 4x more common in AA

# Pathogenesis

- Genetic predisposition: HLA-DR subtypes, TNF-308A polymorphism
- Environmental triggers: malignancy, drugs, infection
- Hypoxia related – hypoxia-inducible factor 1-a (HIF1a)
- Immune-mediated process
  - Cell-mediated immunity, **type I IFNs**, MHC Class I overexpression
  - **Humoral component**



# Classic Findings



## Heliotrope Sign



*Courtesy of Jennifer Abrahams, M.D.*

# Classic Findings



**Midfacial Erythema**

# Classic Findings



**Shawl Sign**



# Classic Findings

- **Holster Sign**



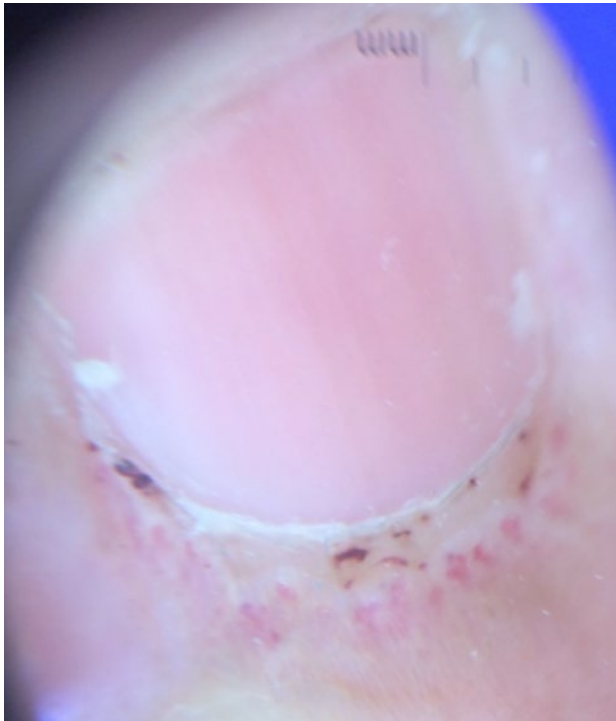
# Classic Findings



## Gottron Papules and Gottron's Sign

*Courtesy of Jennifer Ledon, M.D.*

# Classic Findings



**Periungual Changes**



**Mechanic's Hands**

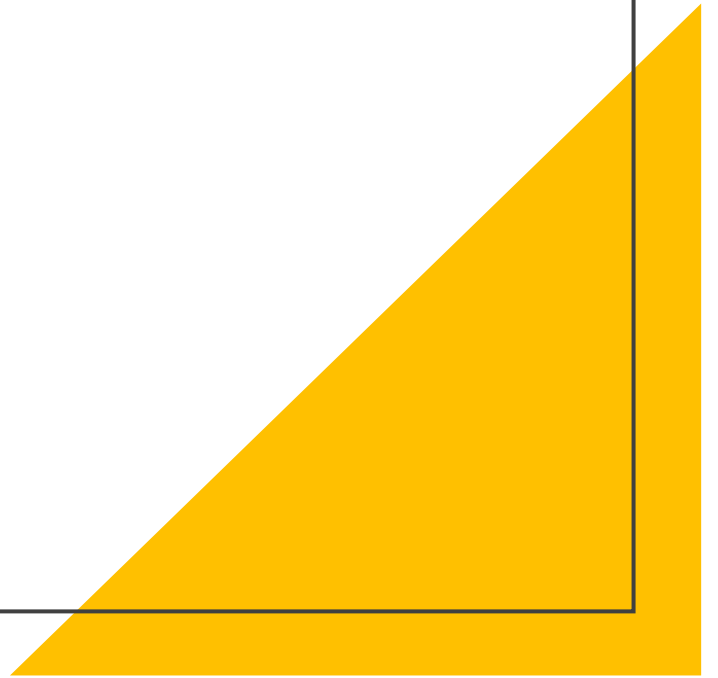
# Classic Findings



**Scalp Poikiloderma**

*Courtesy of Jeffrey Callen, M.D.*

# Variability in Disease Course



# Patient 1

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- 54 yo F, 3 y h/o bx-confirmed DM
- No ILD, malignancy or muscle dz
- Responded well to MMF and sun protection



# Patient 2



- 48 yo F w/ rash x 6 months, bx-confirmed
- Clinically **amyopathic**
- No ILD
- A/w **weight loss**, fatigue
- A/w metastatic ovarian CA
- Expired <6 months after dx

# Patient 3

- 31 yo F w/ 1 month h/o skin lesions, SOB
- **+ mild myositis**
- CT Chest: diffuse ILD
- Expired 2 months after onset from rapidly progressive ILD unresponsive to CS, IVIg, rituximab



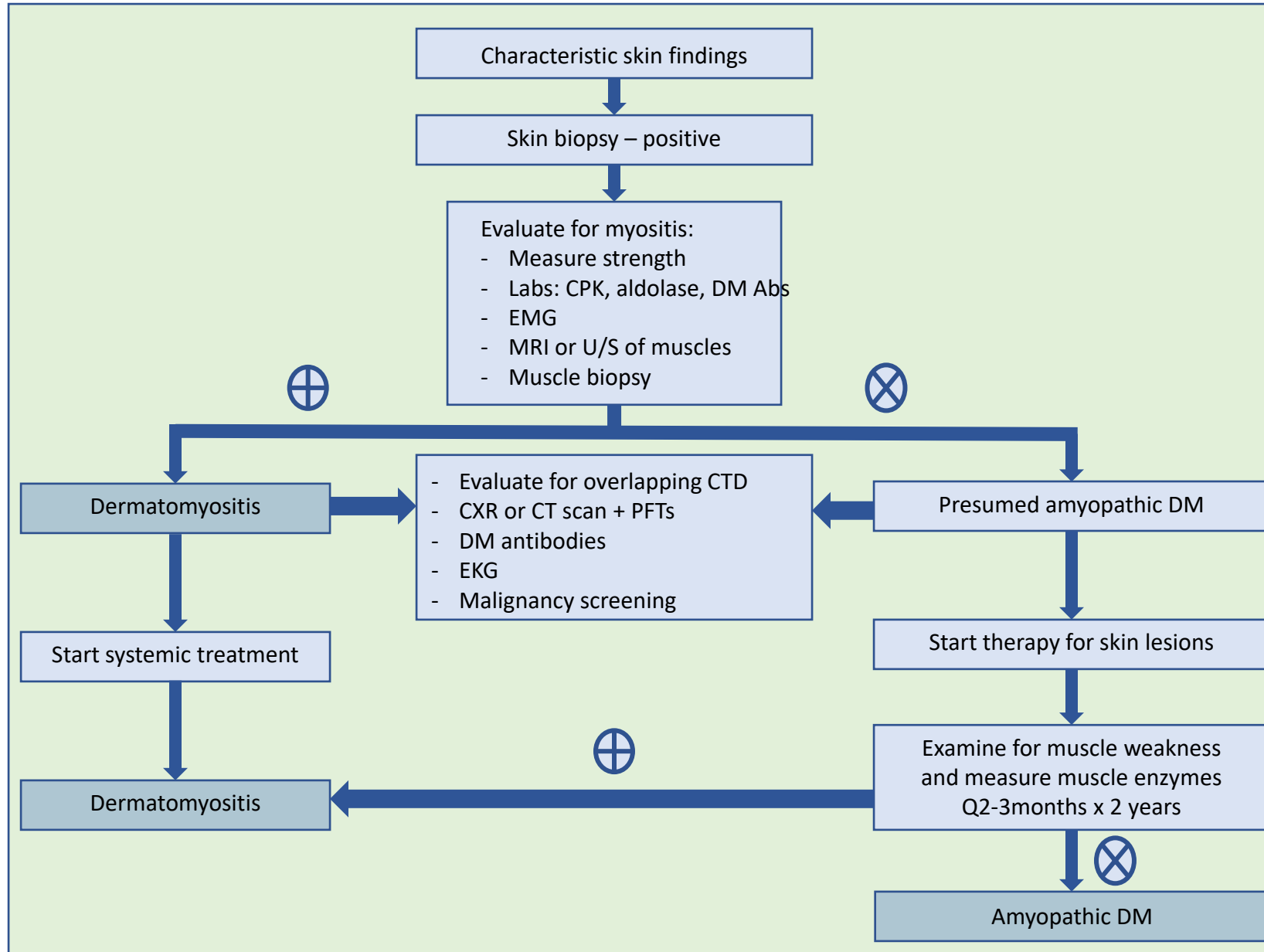
# Clinical Features - Systemic

- Muscle involvement
  - Symmetric involvement
  - Proximal extensor muscles
- Pulmonary involvement – 15-30%
  - Interstitial fibrosis
- Underlying malignancy 15-30%
  - Increased with TIF-1y Ab+
  - MC ovarian and colon CA



Work-up

# Diagnosis



# Autoantibodies in Dermatomyositis

- Important for diagnosis, prognosis
  - Unclear role in pathogenesis

- **Myositis-associated autoantibodies (MAAs)**

- Anti-PM/Scl, -U1/U2/U3/RNP, -Ku, -SSA/Ro, -SSB/La
- Typically seen in overlap

- **Myositis-specific autoantibodies (MSAs)**

- Anti-synthetase antibodies: **Jo-1**, PL-7, PL-12, EJ, OJ
- Others: **Mi-2**, TIF-1γ, NXP-2, SAE, MDA5, SRP

**Jo-1 + Mi-2:**

- Prevalence ~35%

**All MSAs:**

- Prevalence ~80%

- Specificity >90%

# Malignancy Screening

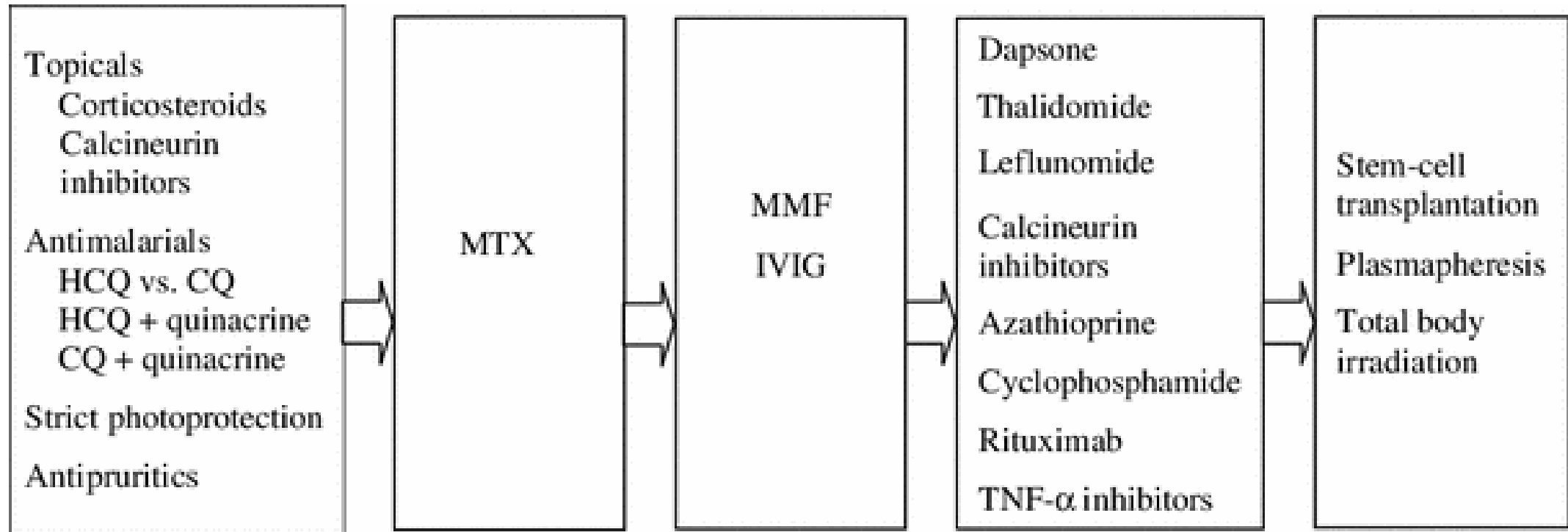
- Screen annually for 2-3 years:
  - Labs: U/A, FOBT, PSA (men), CA-125 (women)
  - Imaging:
    - Mammogram, TVUS (women)
    - CT C/A/P
    - Colonoscopy: if age-appropriate or IDA, +FOBT or sx
    - EGD if colonoscopy neg with IDA, +FOBT or sx

# Management

# Treatment of Cutaneous DM

- Topicals as **adjunctive ONLY**
  - Topical steroids
  - Topical calcineurin inhibitors
  - Anti-pruritics (Sarna)

# Algorithm for Dermatomyositis



# Lupus Erythematosus

- Multi-systemic disease, with highly variable presentation
- Skin-limited (cutaneous lupus or CLE) vs systemic (systemic lupus or SLE)
- In SLE, MC affected organs:
  - Joints (#1)
  - Skin (#2) – seen in 75% pts with SLE
  - Hematologic, pulmonary, renal and CNS
- Main complaints: fever, wt loss, fatigue, myalgias / arthralgias, LAN

# Lupus Erythematosus

- Epidemiology:
  - Gender (F:M = 6:1)
  - Hormonal factor (women during child-bearing years)
  - Ethnicity (4x higher in African-American women)
    - AA → develop the disease at an earlier age and higher mortality rate



# Pathogenesis

- Environmental factors:
    - UV radiation
    - Meds
    - Possibly viruses
  - Genetic factors:
    - SCLE: HLA-B8-DR3
    - SLE: TYK2, IRF5, CTLA4
    - Familial chilblain: TREX1
  - Immune dysregulation
    - Innate immunity
    - Immune complex clearance
    - B- and T-cell function
    - Autoantibodies
-

# Cutaneous Lupus Subtypes



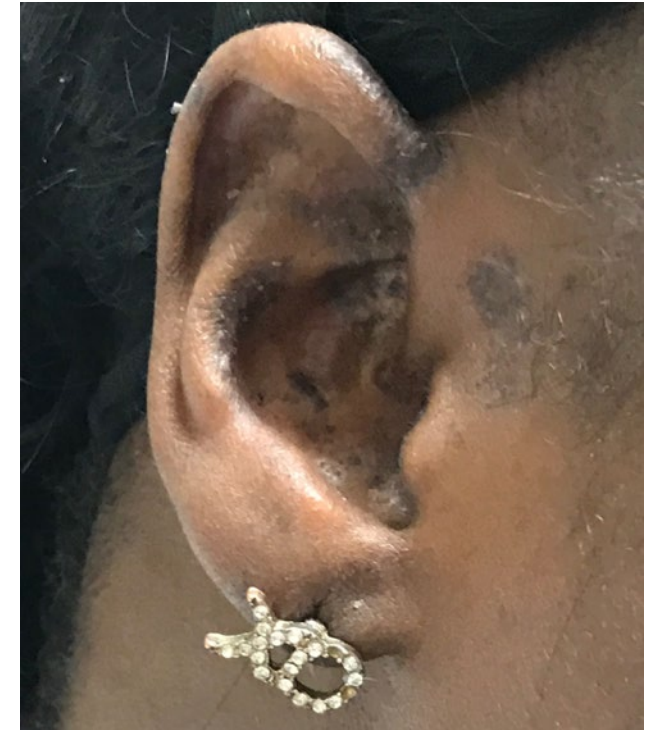
# Cutaneous Lupus



Acute (ACLE)



Subacute (SCLE)



Chronic (CCLE)

Risk of  
progression:

100%

15-50%

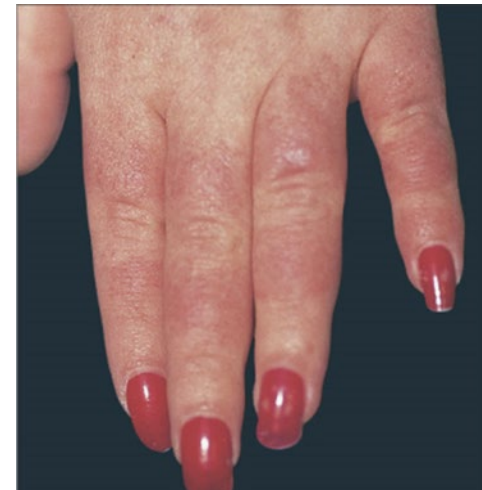
5-20%

Acute  
Cutaneous  
Lupus



# Acute Cutaneous Lupus (ACLE)

- ~100% a/w SLE



ACLE

Mimickers



# Rosacea





CLUES?



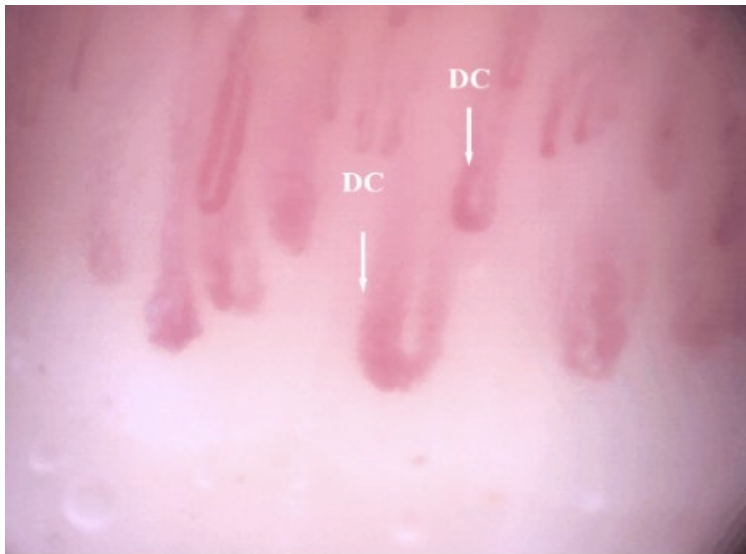
## Clues – Poikiloderma

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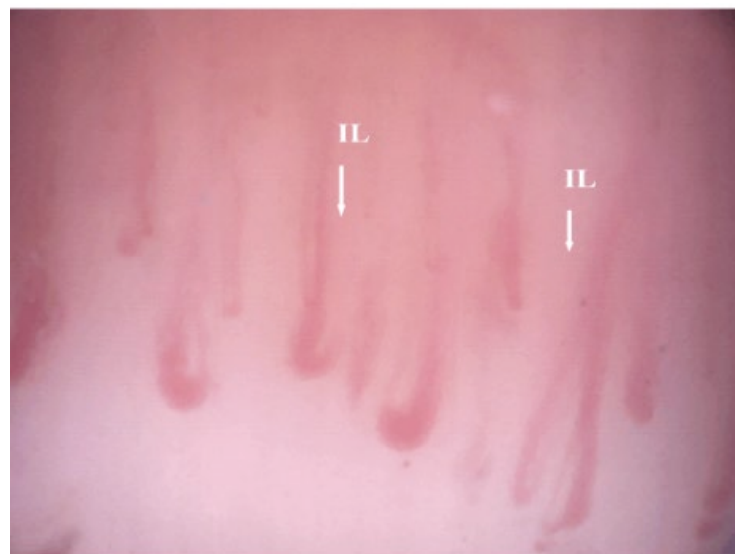
- Tetrad of hypo-, hyper-pigmentation, atrophy, telangiectasia
- **A/w AICTD**
  - Also CTCL, sun exposure, genodermatoses

# Clues – Capillaroscopy

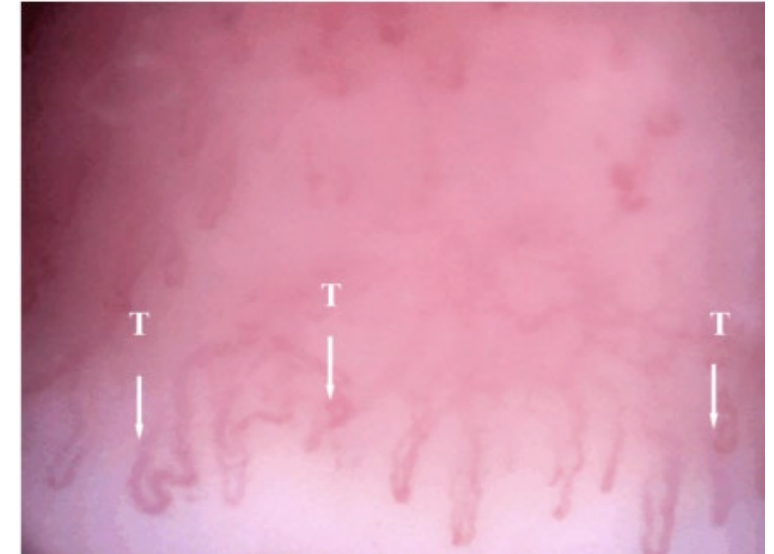
- Less specific, prevalence 75-95%



Regularly dilated

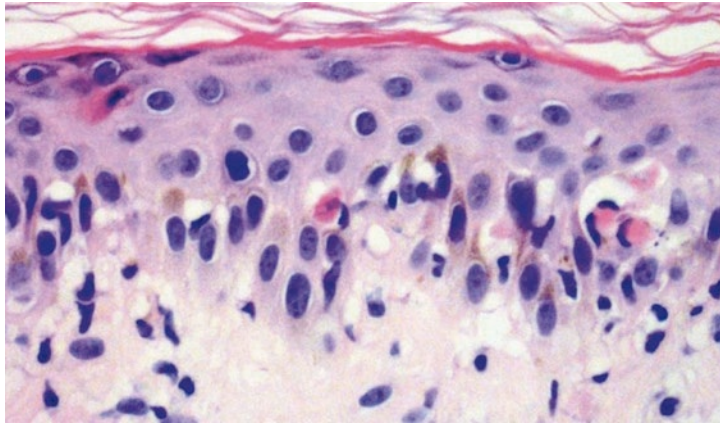


Increased length



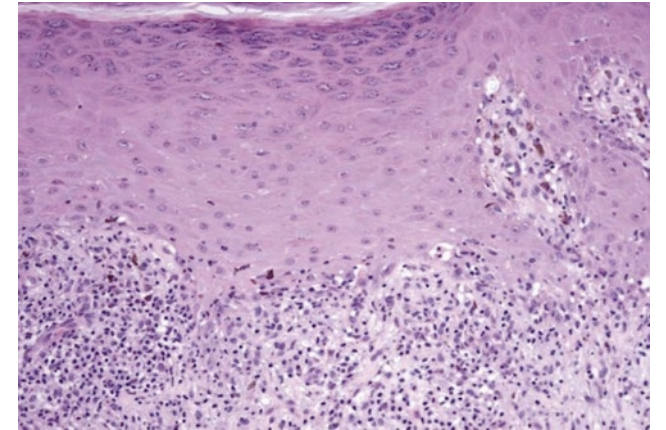
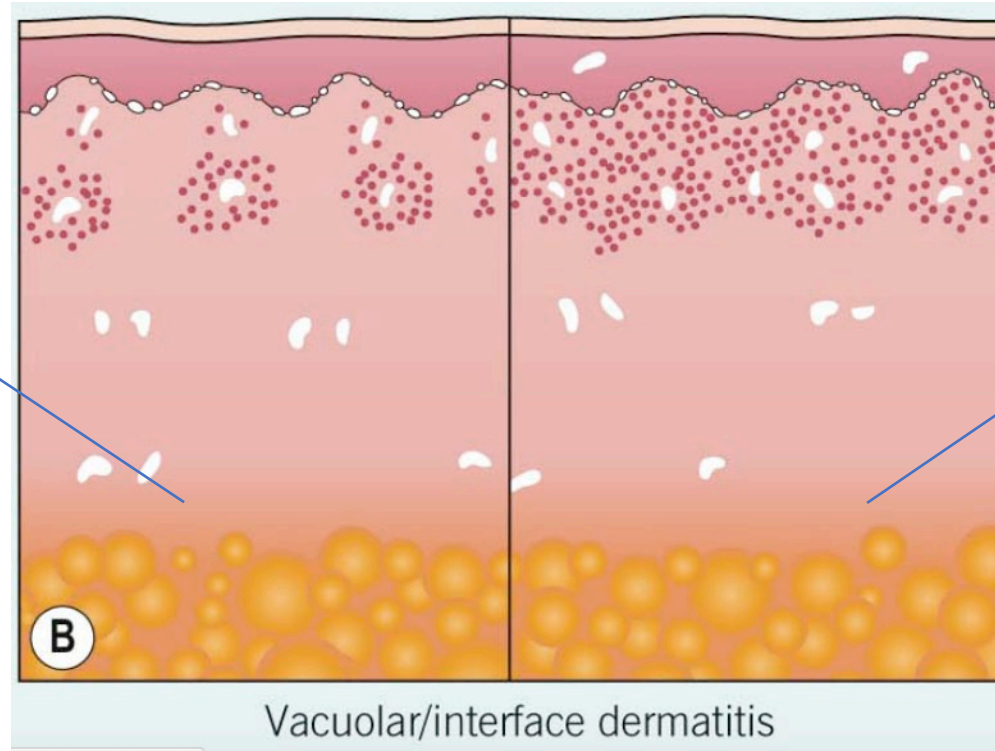
Increased tortuosity

# When all else fails... Biopsy!




Interface, vacuolar

## INTERFACE DERMATITIS



Interface, lichenoid

Vacuolar/interface dermatitis

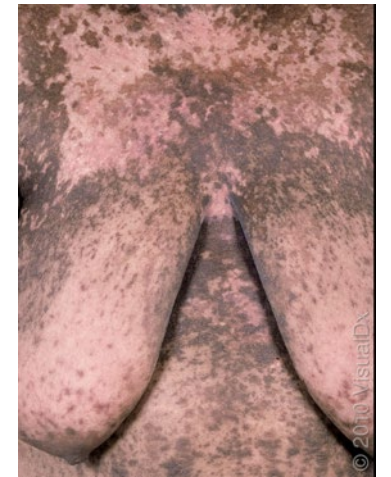


# Subacute Cutaneous Lupus (SCLE)



# Subacute Cutaneous Lupus (SCLE)

- A/w SLE 15-50%
- Clinical: polycyclic / **annular** red scaly plaques, psoriasiform
  - Photo-exposed sites
    - Face often spared
  - No scarring
- A/w + **SSA/SSB**



# Subacute Cutaneous Lupus (SCLE)

- 20-33% drug-induced
- Main culprits
  - **HCTZ**, NSAIDs, terbinafine, diltiazem, **TNF-Is, PPIs**
- May or may not clear after D/C
- Consider drug-induced in **older pts**



# SCLE Mimickers

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# Psoriasis



# Tinea Corporis



# Tinea Corporis





# Chronic Cutaneous Lupus



# Discoid Lupus (DLE)

- **MC form** of cutaneous lupus
- A/w SLE in 5-10% (localized) to 20% (disseminated)



# Discoid Lupus (DLE)



**Plugging (follicular)**



**Pigment**

**PASTE**

# Discoid Lupus (DLE)



Pigment



Plugging (follicular)



Atrophy / Alopecia

**PASTE**

# Discoid Lupus (DLE)



Pigment



Plugging (follicular)



Atrophy / Alopecia



Scar / Scale

**PASTE**

# Discoid Lupus (DLE)



Pigment



Plugging (follicular)



Atrophy / Alopecia



Scar / Scale

**PASTE**



**Telangiectasias**

# Discoid Lupus (DLE)



Pigment



Plugging (follicular)



Atrophy / Alopecia



Scar / Scale

**PASTE**

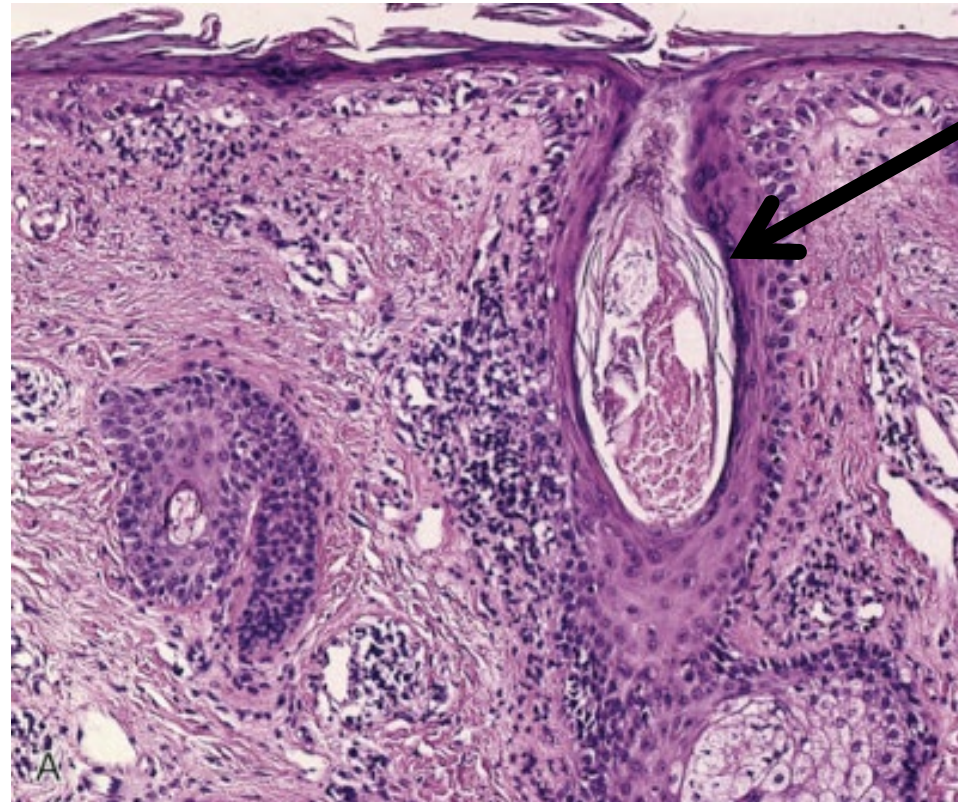
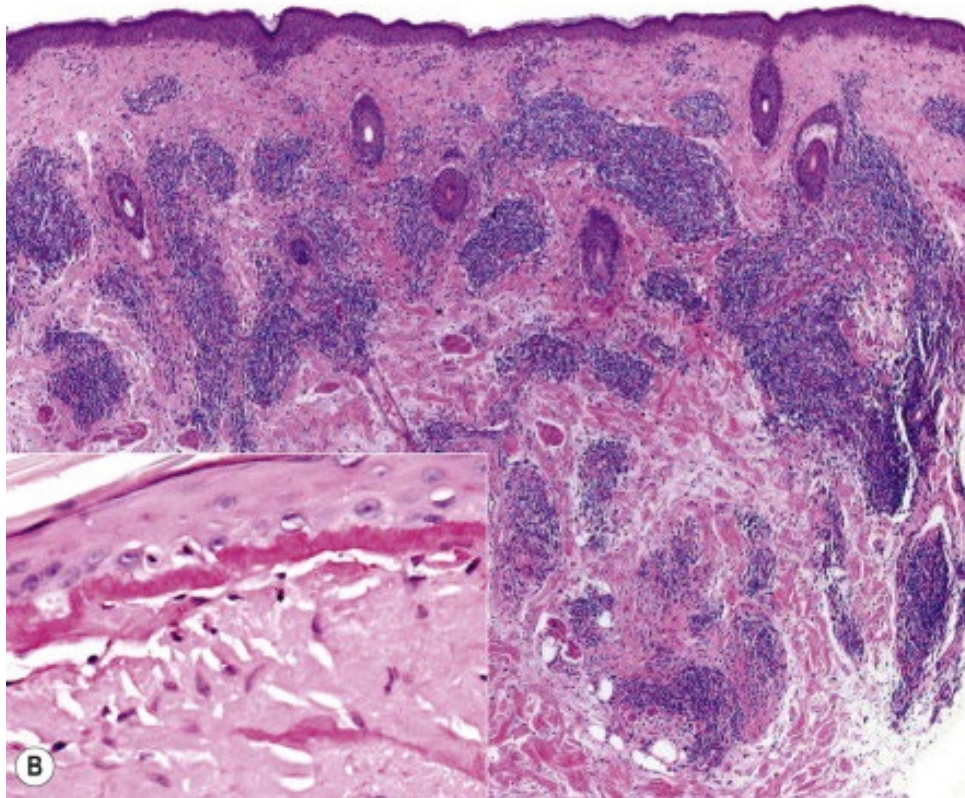


Telangiectasias



Erythema

# H&E – DLE



Follicular  
plugging

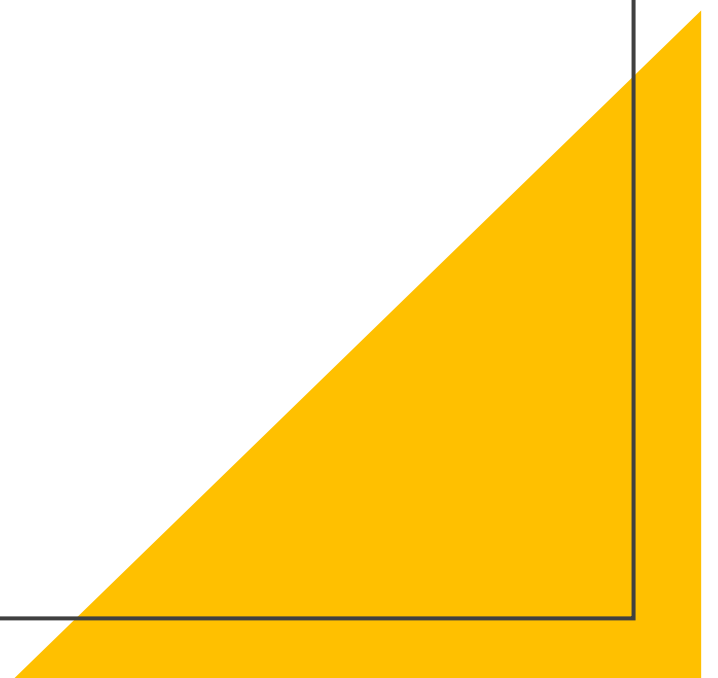
# Lupus Profundus

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- Form of CCLE
  - A/w SLE 10-15%
- **Panniculitis** = inflammation of fat
- Clinical: depressed, SQ nodules
  - Face, upper arms, breasts, buttocks, thighs
  - **Discoid lesions** overlying



Work-up



# Workup of Cutaneous Lupus

- Clinical (H&P)
  - Thorough ROS – don't forget h/o miscarriage, thrombosis, Raynaud's
- Biopsy
  - Classic findings on H&E
  - DIF: Granular deposits of IgG +/- IgM with complement at BMZ
- What labs should you check?
  - CBC, CMP, U/A, C3, C4, ANA, Ro/La, dsDNA, Sm, antiphospholipid Abs

# Diagnostic Criteria – 2012 SLICC

## Clinical Criteria

Acute cutaneous lupus  
Chronic cutaneous lupus  
Oral ulcers  
Non-scarring alopecia  
Synovitis  
Serositis  
Renal  
Neurologic  
Hemolytic anemia  
Leukopenia or lymphopenia  
Thrombocytopenia

## Immunological Criteria

ANA  
Anti-dsDNA  
Anti-Sm  
Anti-phospholipid antibodies  
Low complement  
Direct Coombs test

## Diagnosis:

4 criteria (at least 1 clinical and 1 immunologic)

OR

Biopsy-proven nephritis + ANA or ds-DNA



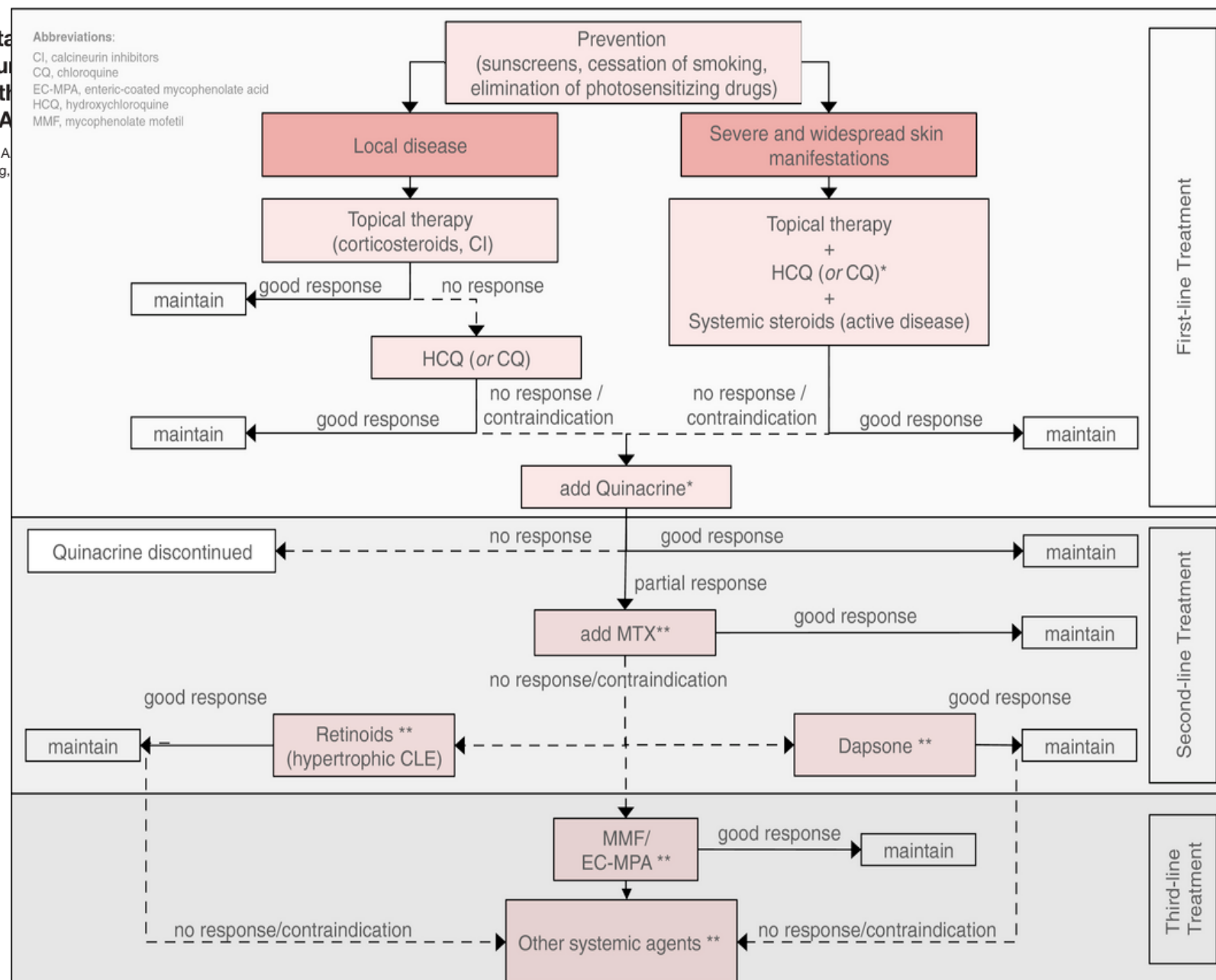
# Management Pearls



## GUIDELINES

## S2k guideline for treatment of cutaneous lupus erythematosus – guided by the European Dermatology Forum (EDF) in cooperation with the European Society of Dermatology and Venereology (ESVD)

A. Kuhn,<sup>1,2,\*</sup> E. Aberer,<sup>3,†</sup> Z. Bata-Csörgő,<sup>4,†</sup> M. Caproni,<sup>5,†</sup> A. H.-W. Klotgen,<sup>9,†</sup> A. Landmann,<sup>2</sup> B. Marinovic,<sup>10,†</sup> F. Nyberg,<sup>11</sup> J.C. Szepietowski,<sup>14,†</sup> B. Volc-Platzer<sup>15,†</sup>



modified after Kuhn A et al. *J Am Acad Dermatol* (2011): Part I: 65: e179–193, Part II: 65: e195–213; \*if patient shows any sign of systemic disease or the risk for the development of systemic disease is high, antimalarials should be continued. \*\*marked agents should not be continued in addition to further second- or third-line treatment options.

# Prevention – Sunscreen



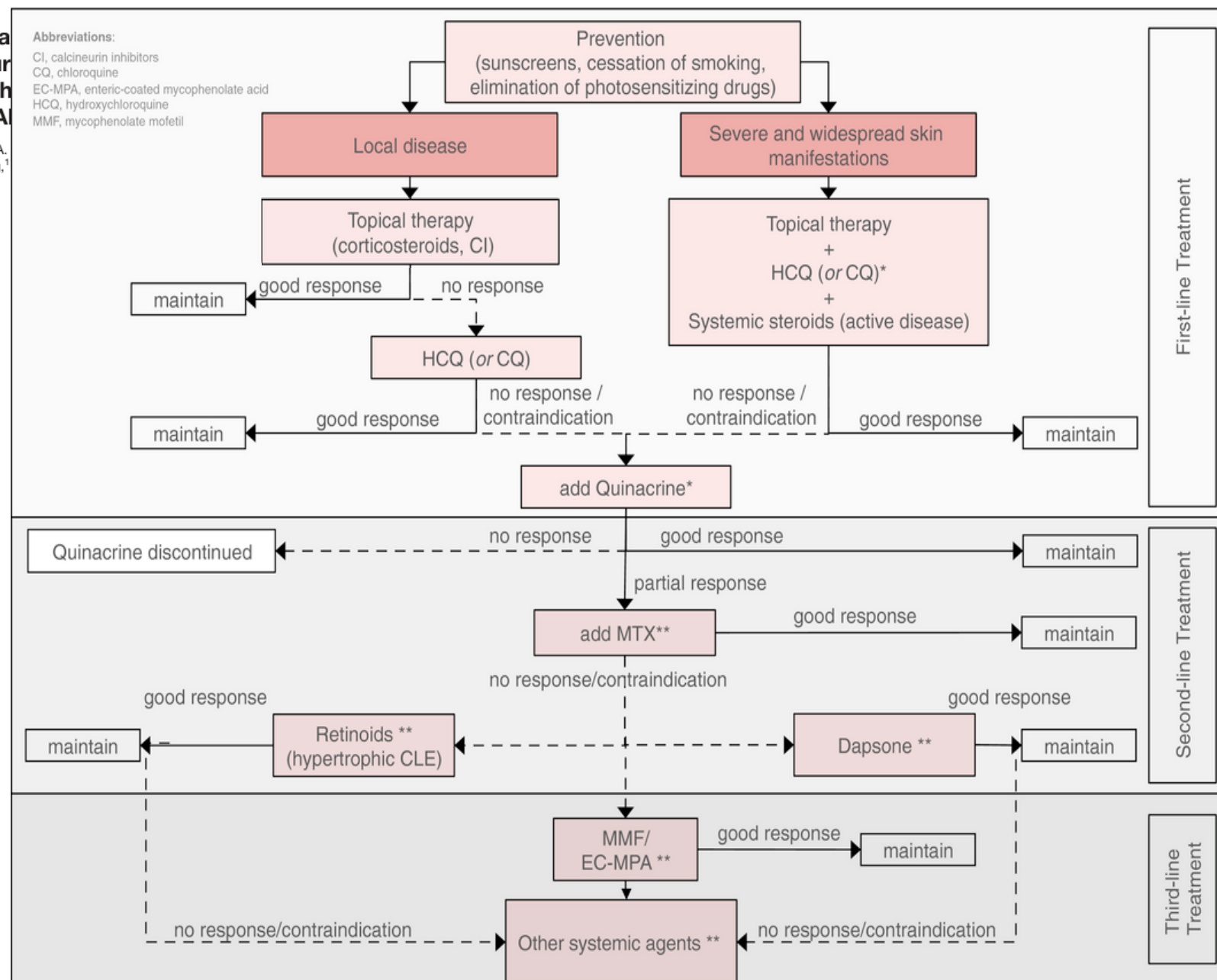
- Broad-spectrum sunscreen – SPF 30-45+
- UVB > UVA > Visible light activates CLE
  - **Physical blockers** (visible light): Zinc oxide, titanium dioxide
- Most important is compliance!!
- **Polypodium leucotomas**



## GUIDELINES

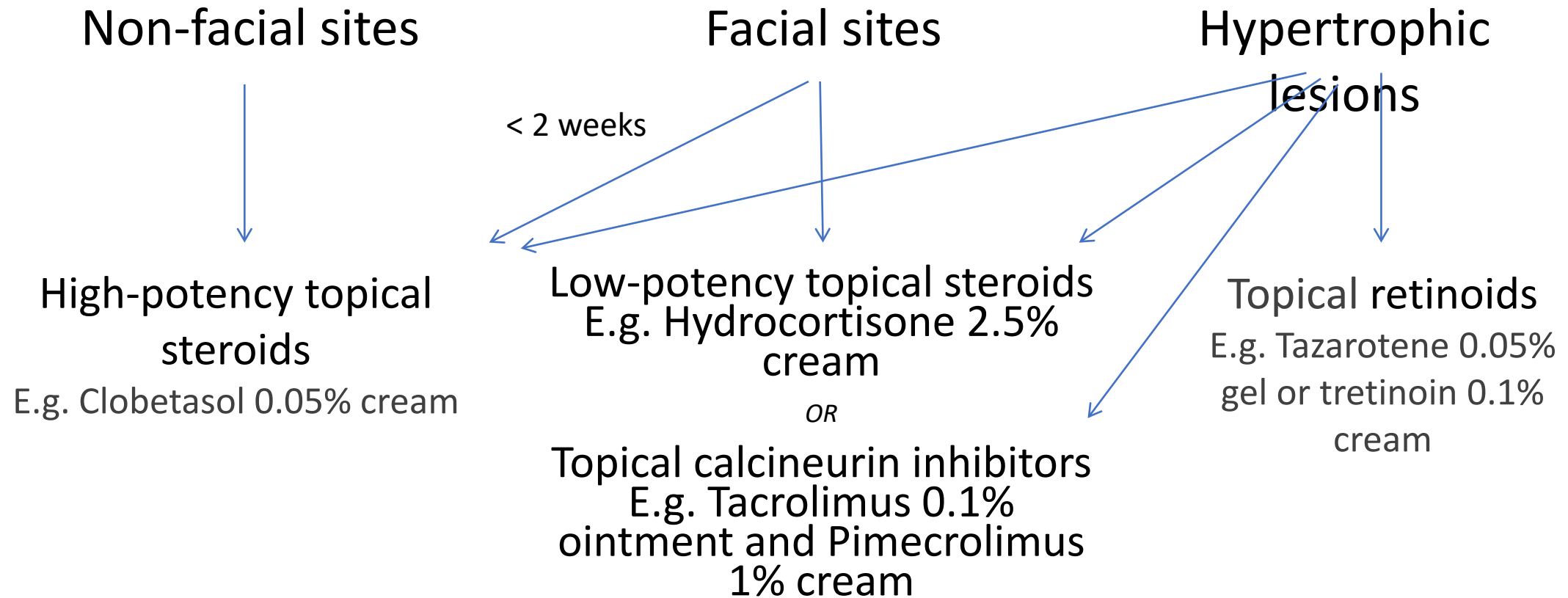
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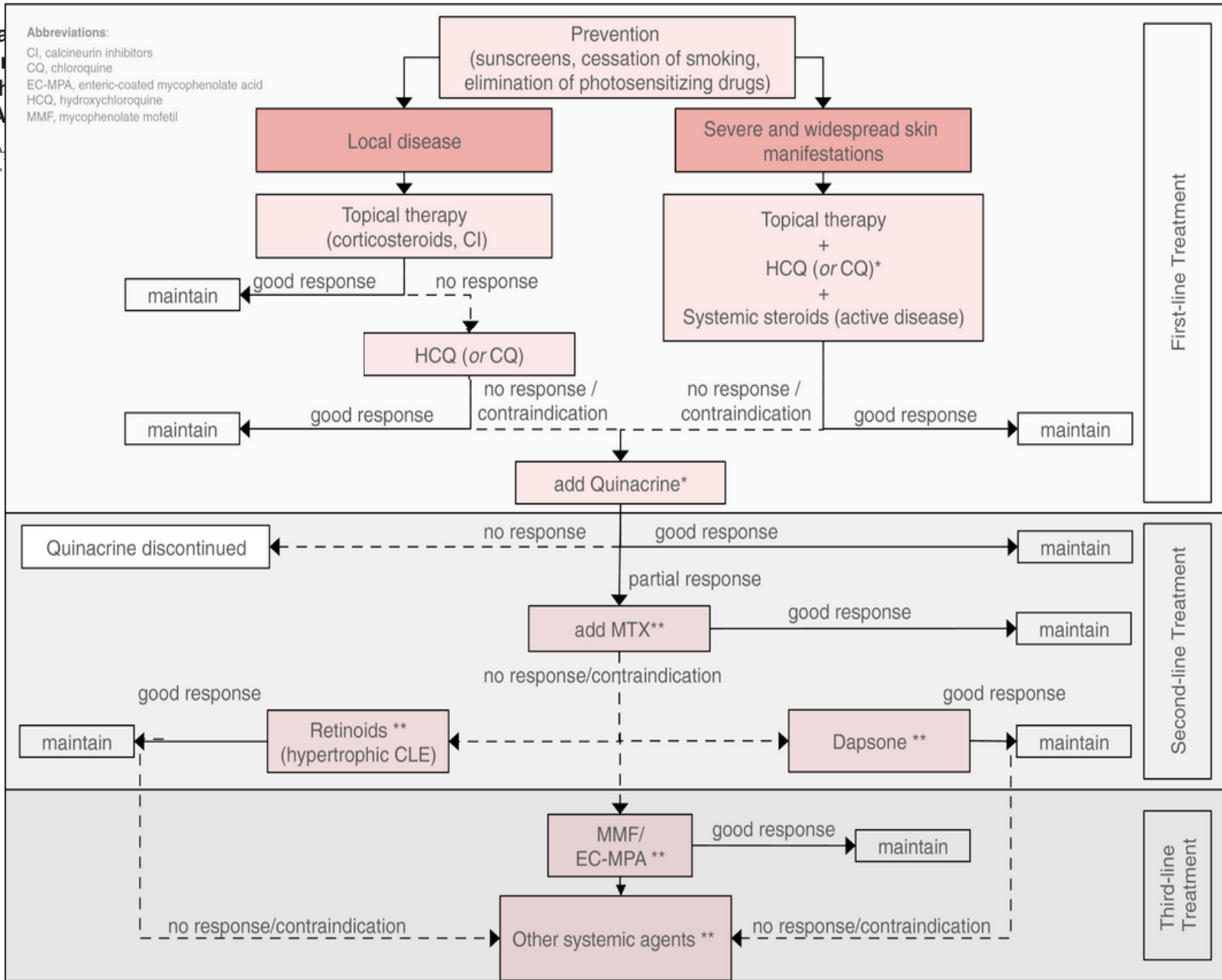
# Topical Therapies



GUIDELINES

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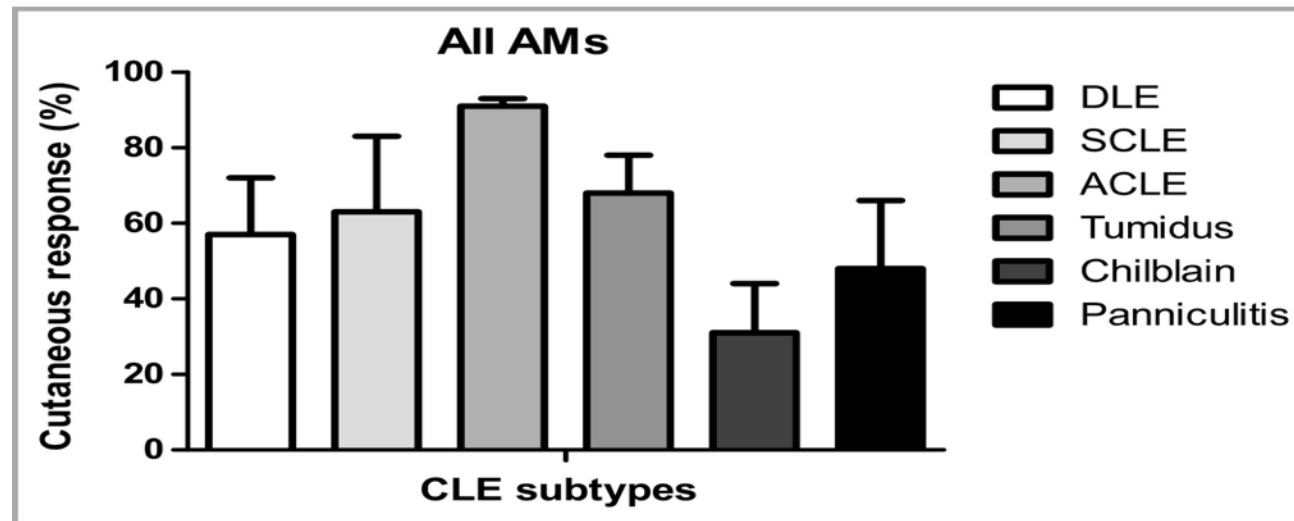
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# Antimalarials

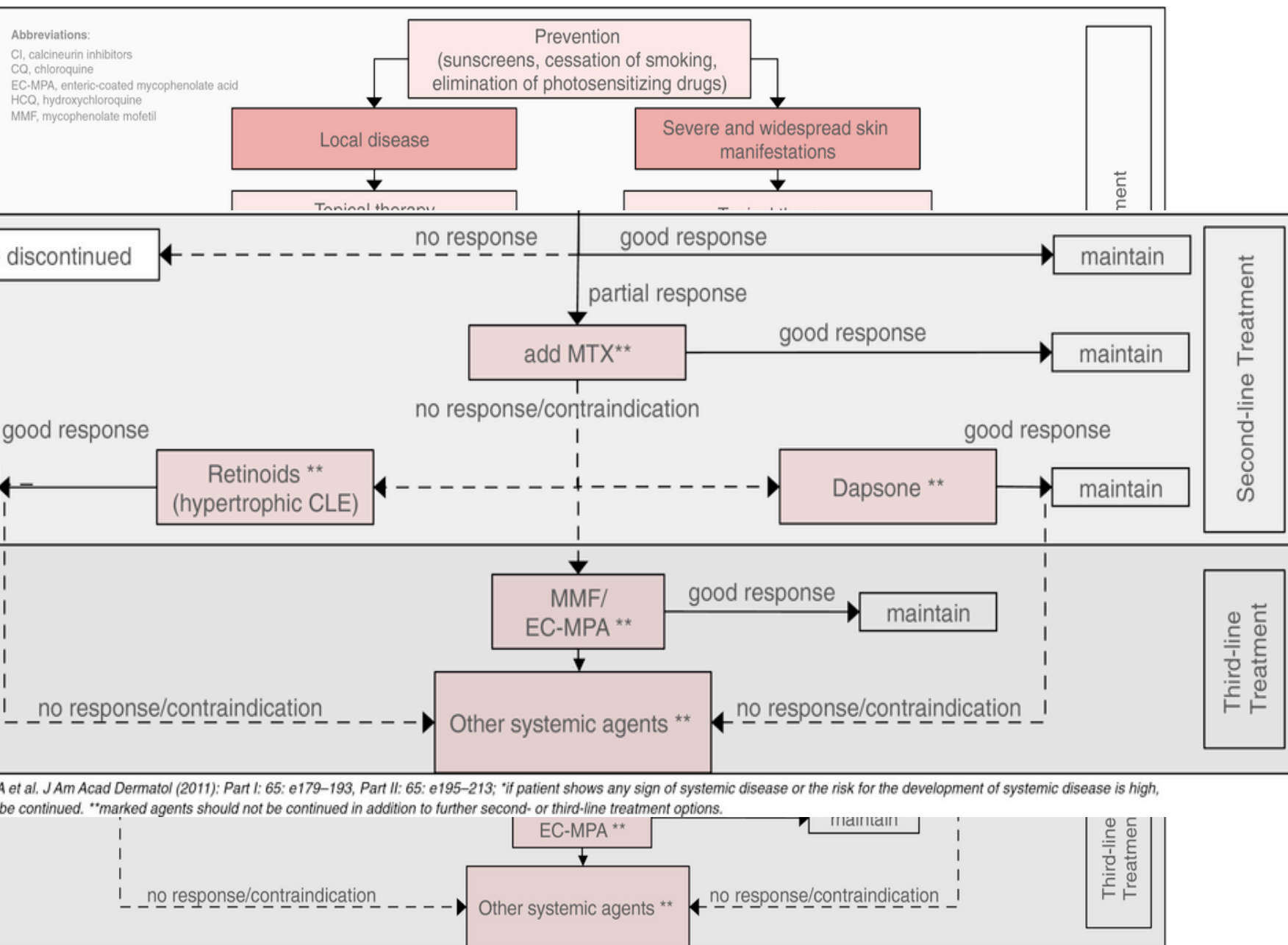
- Dosing?
  - HCQ up to 5 mg/kg/d of body weight PO daily
- Overall response rate: 63%



GUIDELINES

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# Take Home Points

- Dermatomyositis and lupus are photosensitive conditions that can herald underlying systemic disease
- Dermatomyositis presents with classic skin findings and can be a/w muscle disease, malignancy and lung disease
- Cutaneous lupus has several subtypes, with varying association with underlying SLE
- Photoprotection is of paramount importance for these patients
- Treatment is with topical and systemic therapy for disease control