Pediatric Papulosquamous and Eczematous Disorders

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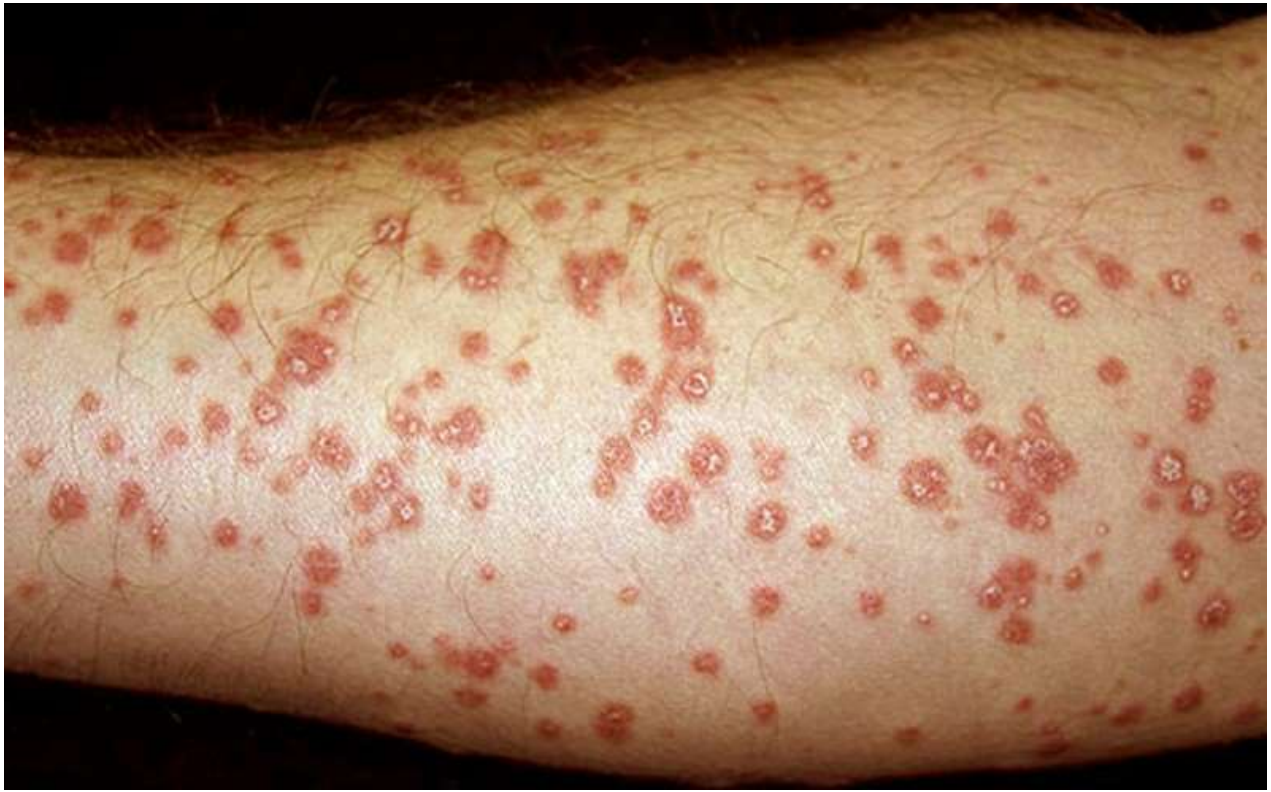
Pediatric Psoriasis



Epidemiology

- Psoriasis can first appear at any age, from infancy to the eighth decade of life
- The prevalence of psoriasis in children ages 0 to 18 years old is 1% with an incidence of 40.8 per 100,000 ppl
- ~ 75% have onset before 40 years of age

What causes psoriasis?

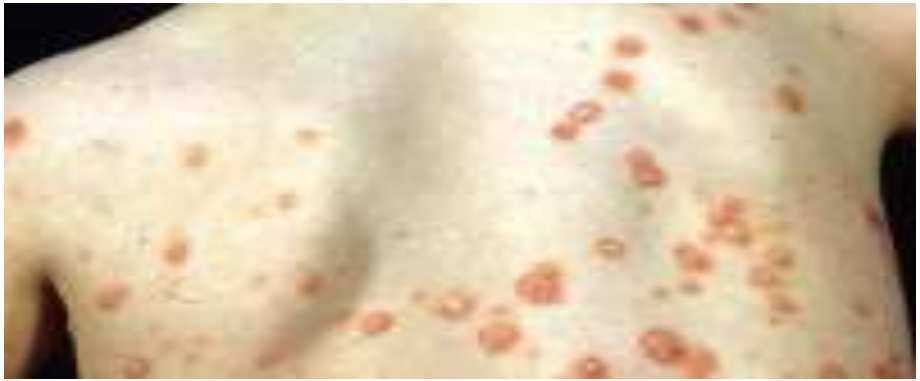
- Multifactorial
- Genetics
  - HLA associations (Cw6, B13, B17, B57, B27, DR7)
- Abnormal T cell activation
  - Th1, Th17 with increased cytokines IL 1, 2, 12, 17, 23, IFN-gamma, TNF-alpha
- External triggers:
  - Injury (Koebner phenomenon)
  - medications (lithium, IFNs, β-blockers, antimalarials, rapid taper of systemic corticosteroids)
  - infections (particularly streptococcal tonsillitis).

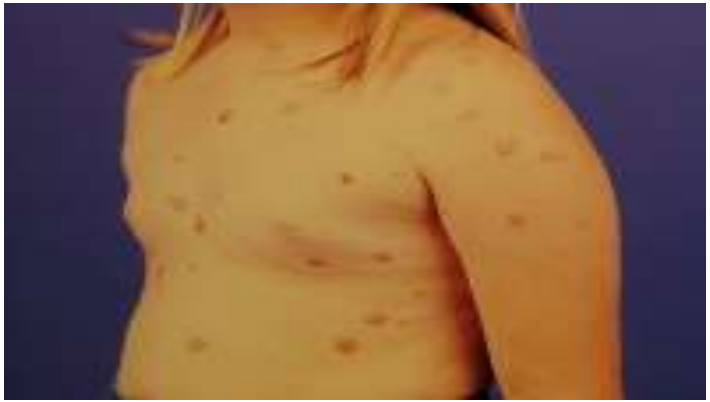
Pediatric Psoriasis

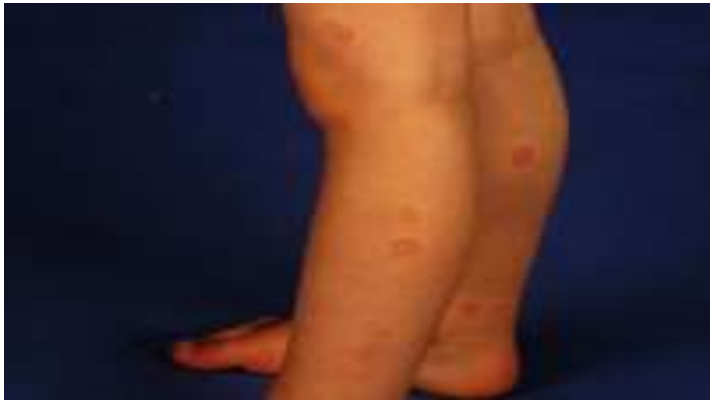
Types:

- Acute Guttate Psoriasis – Small erythematous plaques occurring after infection (MOST common in children)
  - **40% of patients with guttate psoriasis will progress to develop plaque type psoriasis**
- Chronic plaque Psoriasis – erythematous plaques with scaling
- Flexural Psoriasis – Erythematous areas between skin folds
- Scalp Psoriasis – Thick scale found on scalp
- Nail Psoriasis – Nail dystrophy
- Erythrodermic Psoriasis– Severe erythema covering all or most of the body
- Pustular Psoriasis – Acutely arising pustules
- Photosensitive Psoriasis – Seen in areas of sun exposure

Guttate Psoriasis







Psoriasis Treatment

- 1st line- Topical corticosteroids
- Topical calcipotriene (Vit D analogue)
- Phototherapy
- Retinoids like Acitretin can be used in children starting at 6 months of age
- Methotrexate is used as “rescue therapy” in children; important to not exceed 0.7 mg per kg per week
- Important to supplement MTX with Folate to minimize GI toxicity and possibility of bone marrow suppression
- Biologics

Psoriasis Treatment

Biologic Agents: Etanercept

- Soluble tumor necrosis factor receptor fusion protein
- Works by binding and inhibiting TNF, reducing inflammation and altering immune response
- Has the most evidence, including a placebo randomized trial and multiple case reports

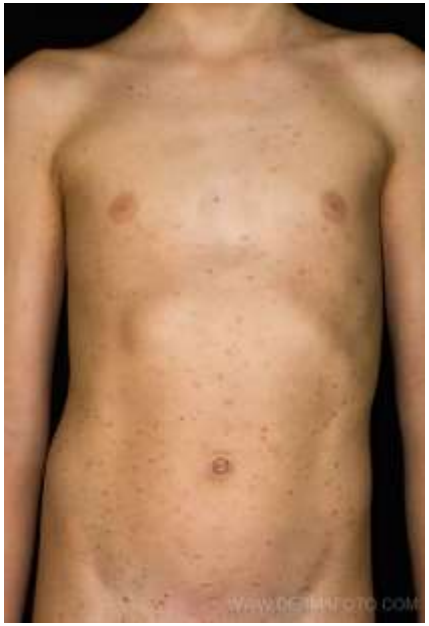
Psoriasis Treatment

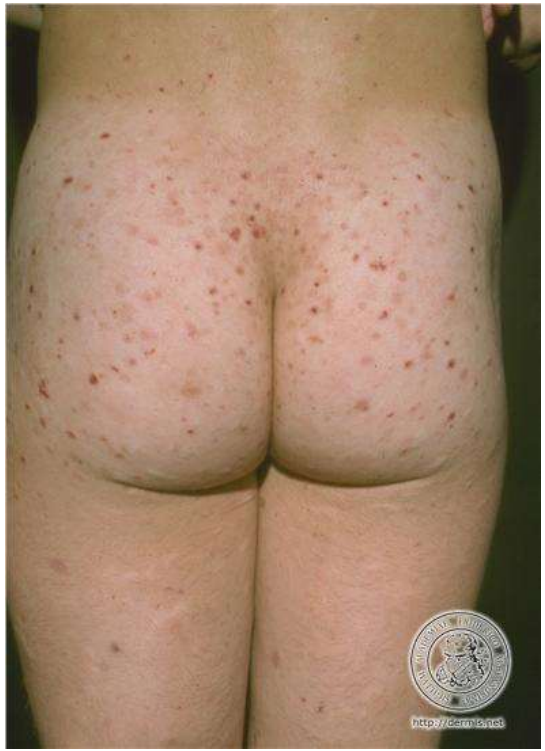
Etanercept:

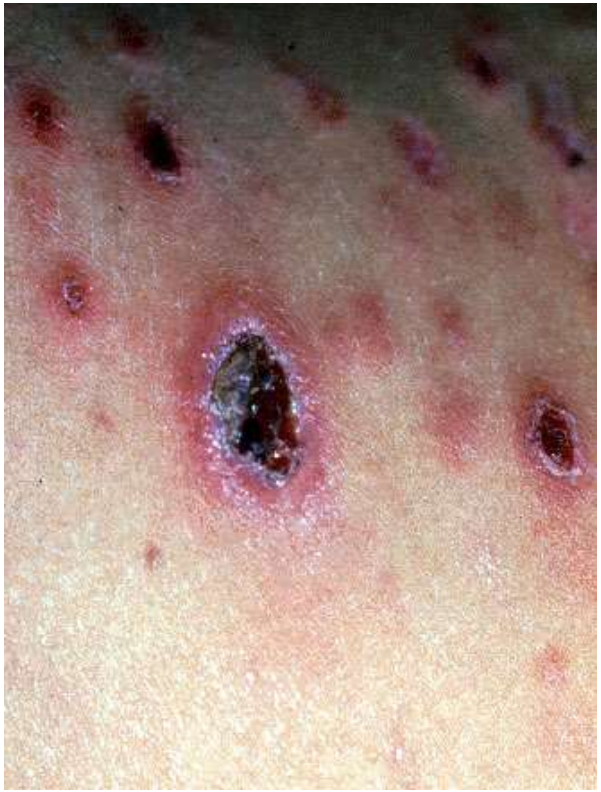
- Phase 3 randomized study showed statistically significant reductions in disease severity as early as week 2 of weekly treatment with Etanercept at 0.8 mg/kg in children and adolescents with moderate to severe psoriasis

Etanercept treatment for children and adolescents with plaque psoriasis, Paller et al., NEJM , January 2008

Pityriasis Lichenoides







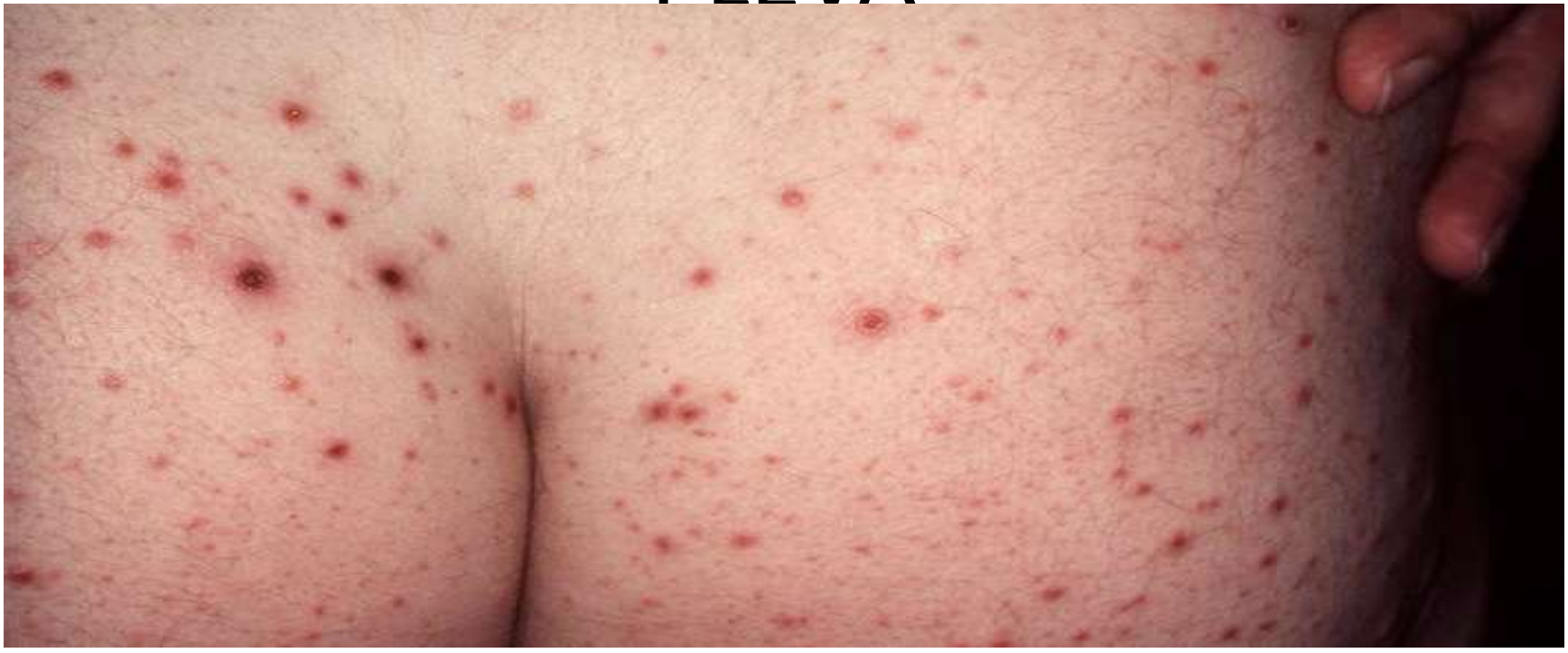
Pityriasis Lichenoides

- Unknown etiology
- Most often affects adolescents and young adults
- Males > females
- Acute: PLEVA
- Chronic: PLC

Pityriasis Lichenoides et varioliformis acuta (PLEVA)

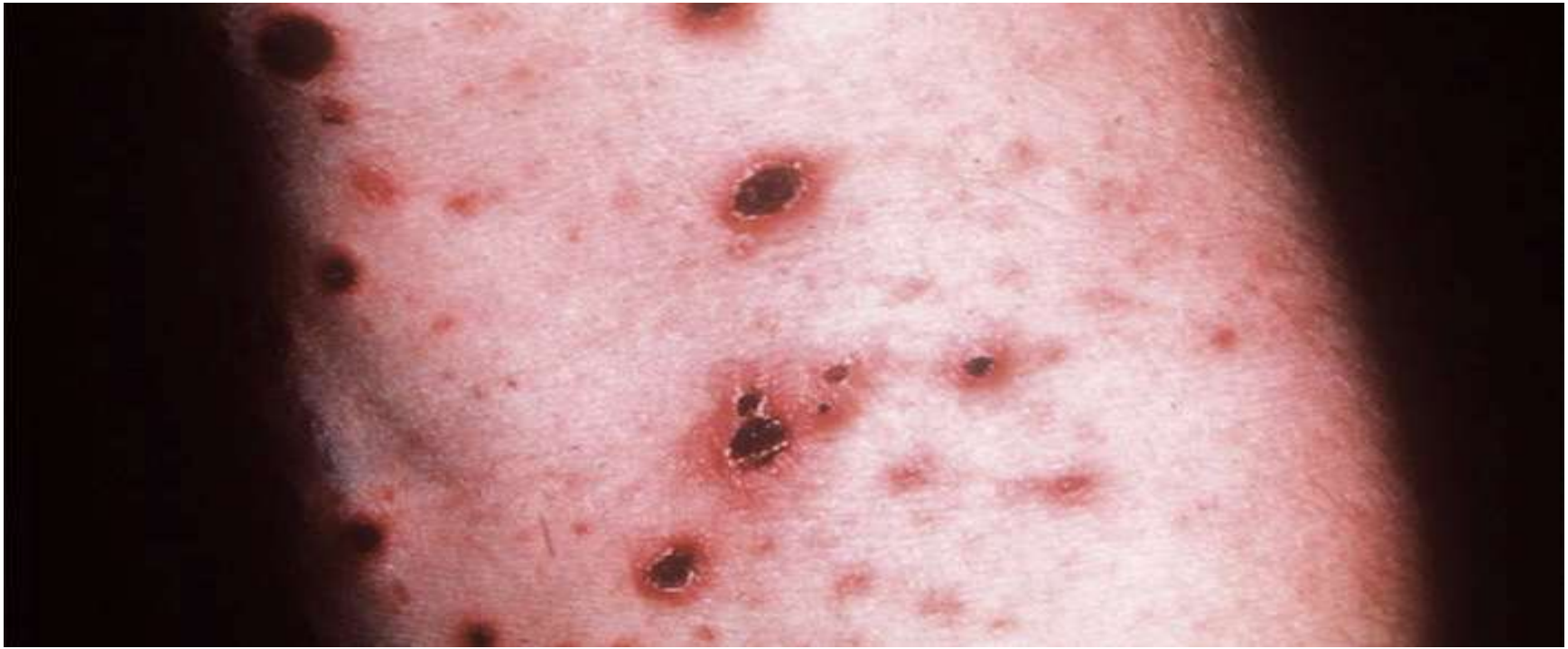
- Abrupt eruption of erythematous papules and vesicles with crusted or necrotic centers
- Lesions are painful and itchy
- Usually distributed over trunk, buttocks, and extremities, but sometimes may be widespread, covering any part of the body
- Involutes within weeks to months

PLEVA



Typical hemorrhagic crusted papules of pityriasis lichenoides et varioliformis acuta

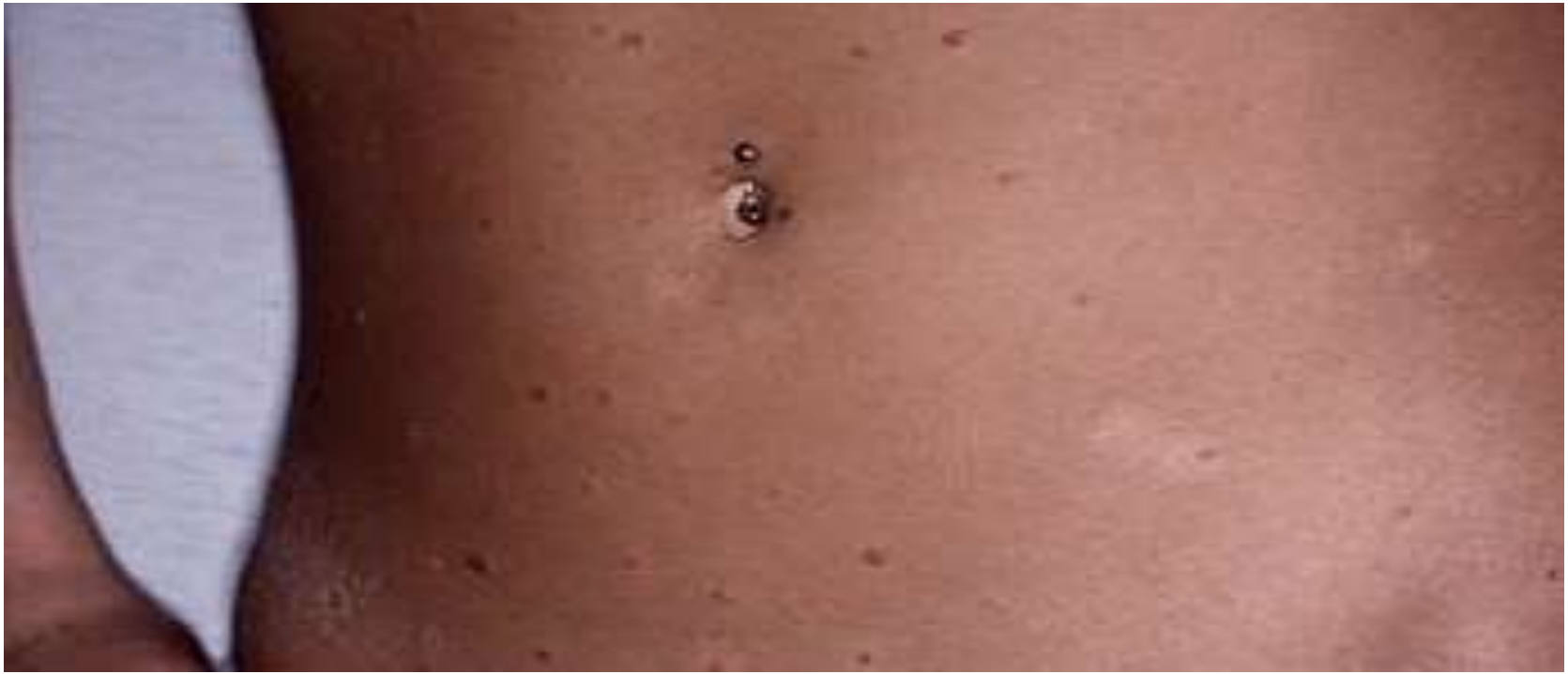
PLEVA



Pityriasis Lichenoides Chronica (PLC)

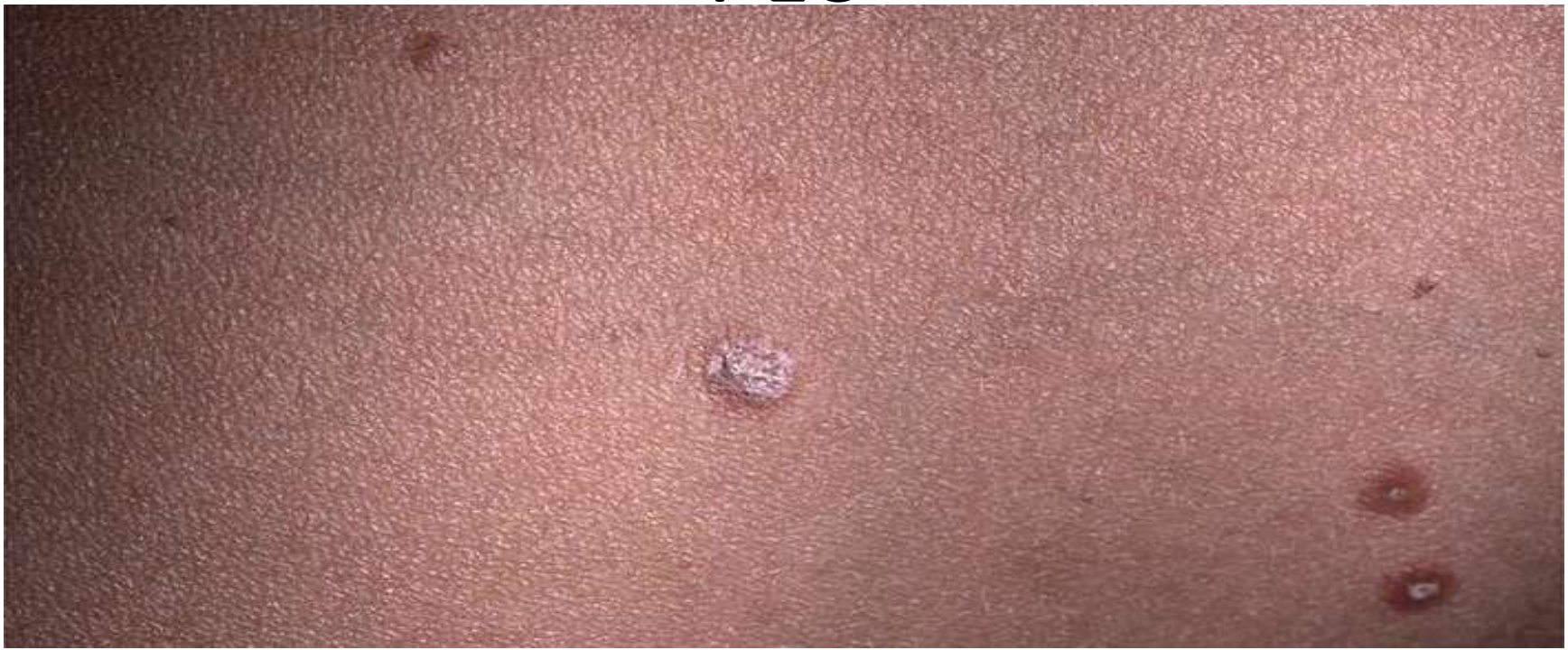
- Reddish brown papules with adherent scale
- Heals with PIH
- More chronic course lasting months to years with exacerbations and remission

PLC



Scaling papules of pityriasis lichenoides chronica

PLC



Note papules in different stages of evolution and the scale with frosted-glass appearance in the lower right-hand corner

PLEVA/PLC -Treatment: First line

- Symptomatic
- Local wound care for larger ulcerations
- Topical steroids
- Topical immunomodulators, tacrolimus, pimecrolimus
- Oral erythromycin (children) and doxycycline (adults)

PLEVA/PLC- Treatment

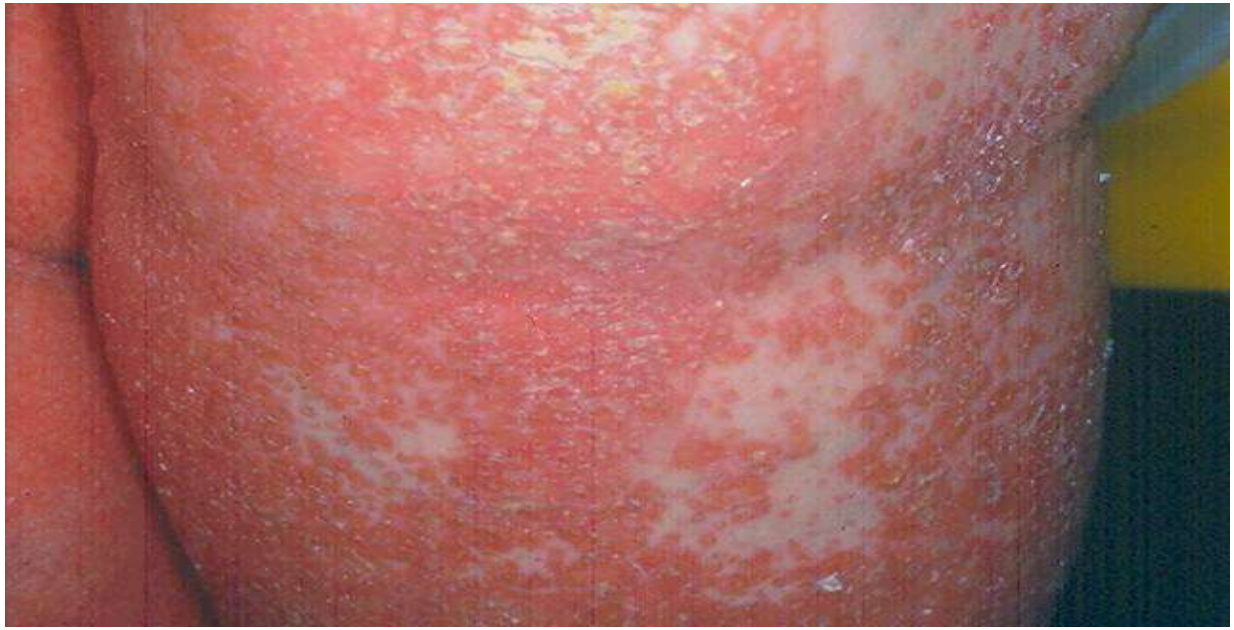
Second-line therapies include:

- Phototherapy – UVB or PUVA

Third-line therapies include:

- Systemic steroids
- Methotrexate orally or by IM injection
- Acitretin
- Dapsone
- Cyclosporine

Pityriasis Rubra Pilaris (PRP)

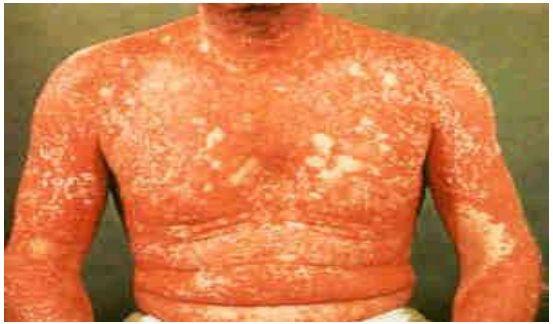


Pityriasis Rubra Pilaris (PRP) Background

- Chronic papulosquamous disorder of unknown etiology characterized by reddish orange scaly plaques, PPK and keratotic follicular papules.
- Etiology unknown. Nearly always acquired. Occasional familial cases described with AD inheritance recently linked to CARD gene.
- May be caused by immune response to antigen
- Cases described after strep infections

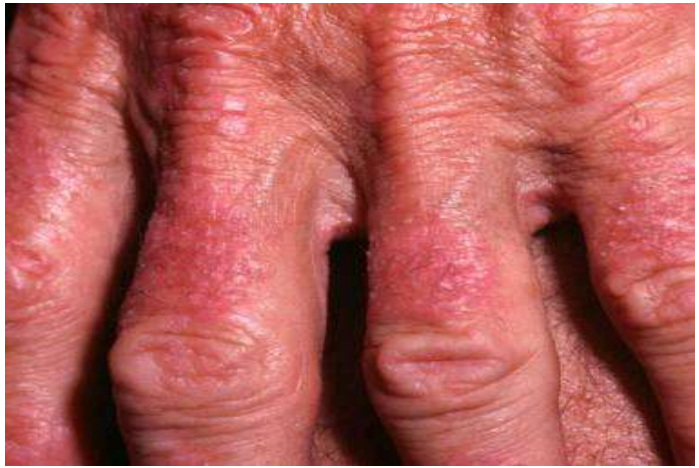
Pityriasis Rubra Pilaris (PRP) Presentation

- Orange-red or salmon-colored scaly plaques with sharp borders, may expand to cover entire body
- Areas of uninvolved skin referred to as islands of sparing



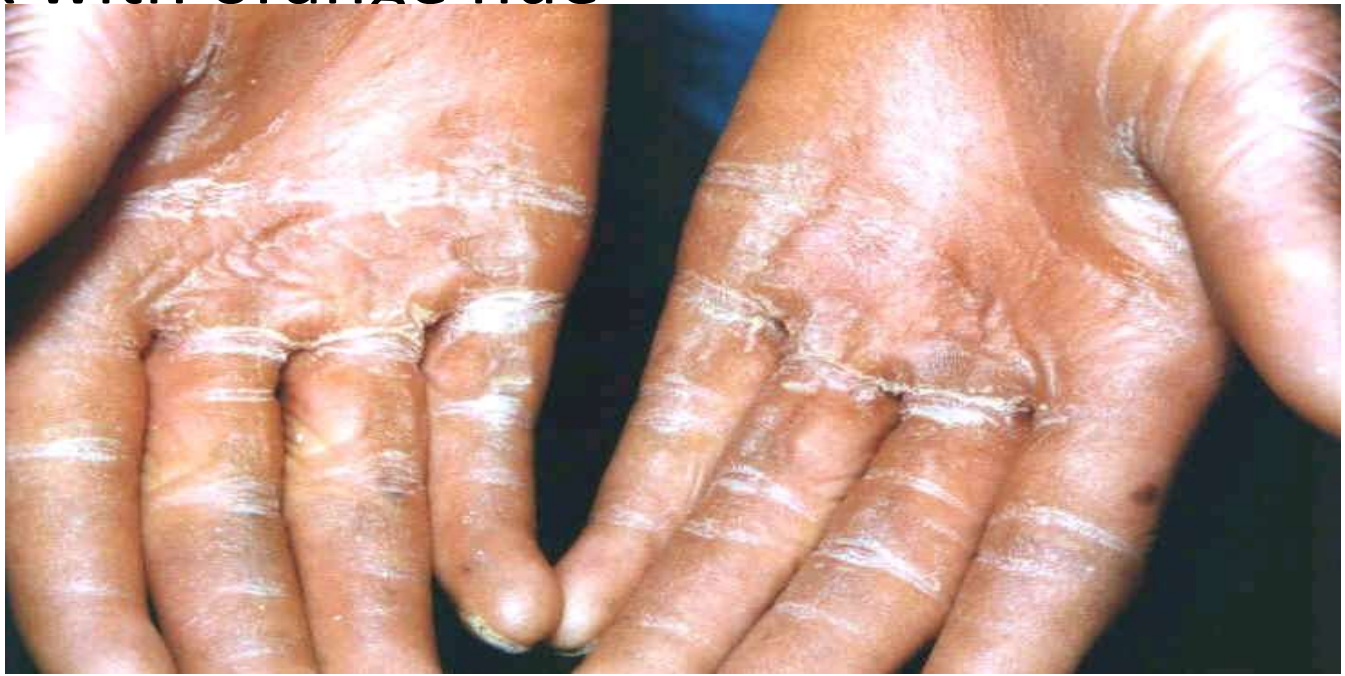
Pityriasis Rubra Pilaris (PRP) Presentation

- Follicular hyperkeratosis commonly seen on dorsal aspects of proximal fingers, elbows and wrists.



Pityriasis Rubra Pilaris (PRP) Presentation

- PPK with orange hue



Pityriasis Rubra Pilaris (PRP) Presentation

- Nail changes include subungual hyperkeratosis and nail plate thickening
- May rapidly evolve into erythroderma

Pityriasis Rubra Pilaris (PRP)- Adult Forms

- Type I (Classic Adult): More than 50% of cases, sudden onset of symptoms with duration 2-5 years
- Type II (Atypical Adult): about 5% of cases, slow onset with alopecia, localized lesions and more chronic course
- Type VI (HIV-associated PRP): Presents with acneiform lesions and elongated follicular plugs. Resistant to standard Tx, but may respond to antiretroviral therapy.

Pityriasis Rubra Pilaris (PRP)- Juvenile Forms

- **Type III (Classic Juvenile):** Resembles classic adult form, with early onset (first 2 years of life); most resolve within 3 yrs; 10% of cases
- **Type IV (Circumscribed Juvenile Form):** Most common in children (25% of cases), lesions on extensor surface and present in prepubertal years; about 50% may persist into adulthood
- **Type V (Atypical Juvenile Form**): Similar to Type III + scleroderma-like changes on hands and feet. This form accounts for about 5% of all cases and most familial cases. More chronic course.

Pityriasis Rubra Pilaris (PRP)
DDx

- CTCL
- Erythroderma
- Erythrokeratoderma Variabilis
- Psoriasis
- Seborrheic dermatitis

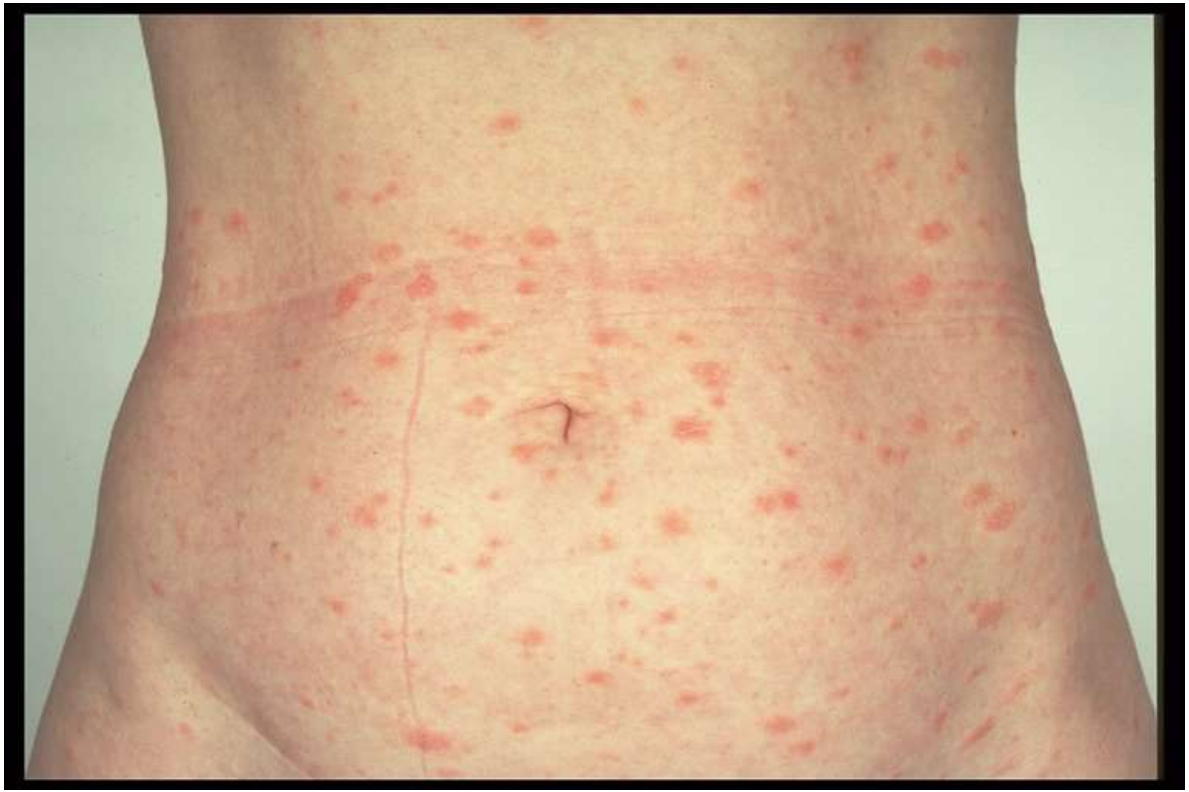
Pityriasis Rubra Pilaris (PRP) Workup

- Diagnosis based on correlation between clinical findings and histological findings
- No lab tests indicated

Pityriasis Rubra Pilaris (PRP)-Tx

- No set protocols, evidence for specific therapies sparse
- Topical steroids
- Tazarotene reported to improve Type IV
- Emollients, especially for hands
- NB-UVB
- Isotretinoin
- Cyclosporine, Azathioprine, Methotrexate
- TNF-alpha inhibitors
- Ustekinemab

Pityriasis Rosea



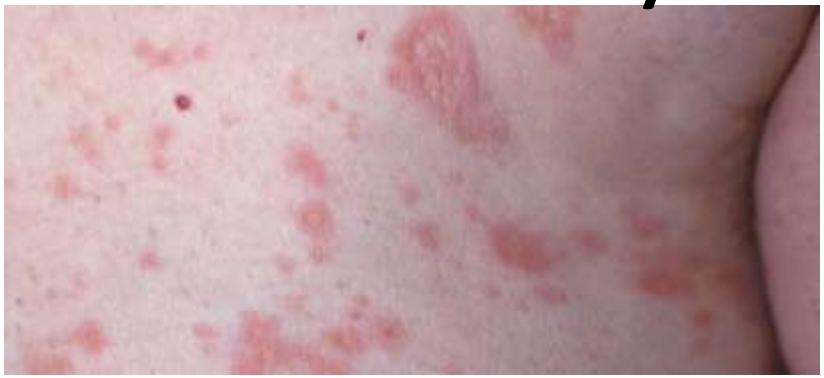
Pityriasis Rosea

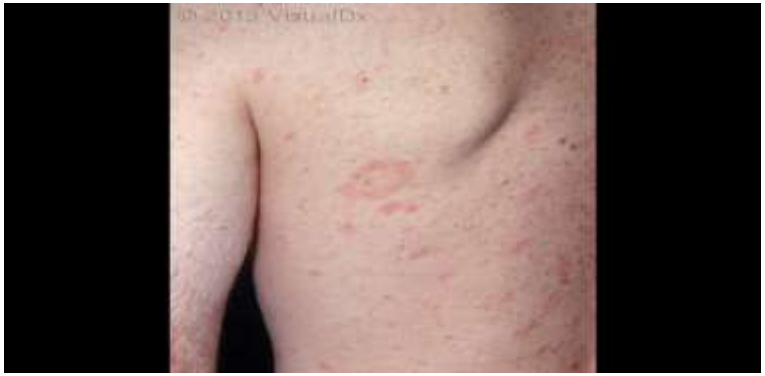
- Self limited papulosquamous eruption seen primarily in healthy adolescents and young adults
- First the rash begins with a solitary oval 2-5 cm scaly pink patch with a slightly raised advancing margin, classically on the trunk, which enlarges over several days
- Hours to days later similar smaller scaly patches appear on the trunk, but may also present on the proximal extremities and neck
- Usually a “Christmas tree” pattern is described on the back due to the long axis of the lesions following Langer’s lines of cleavage
- Patients may complain of upper respiratory symptoms prior to the outbreak
- Assoc. with HHV-7 and to a lesser extent HHV-6 infection (Drago et. al 2009)

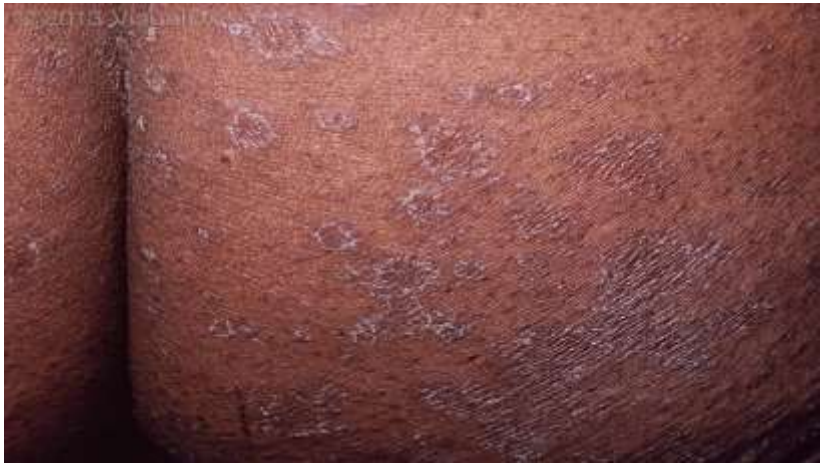
Pityriasis Rosea

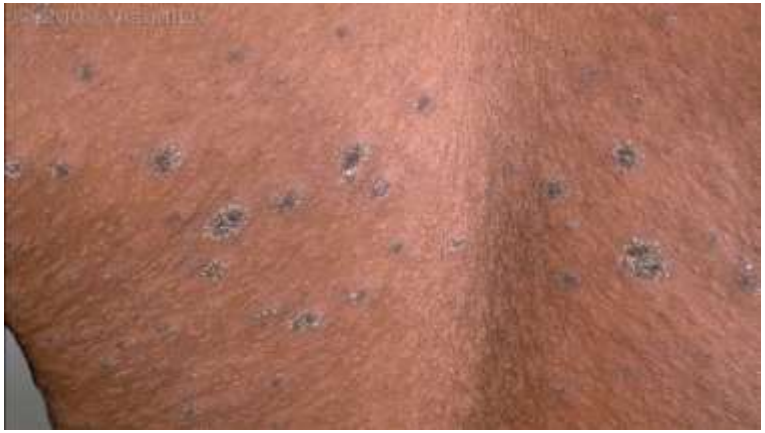
- May or may not be pruritic
- Typically resolves spontaneously in approximately 6 weeks
- Usually does not recur
- Treatment: Antipruritic lotions, low to medium strength topical steroids and antihistamines for symptomatic relief from itch
- More severe cases: UVB therapy
- No evidence that Azithromycin is effective (Pandhi et. al 2015)
- No evidences that antivirals are effective (Chuh et. al 2005)

Pityriasis Rosea









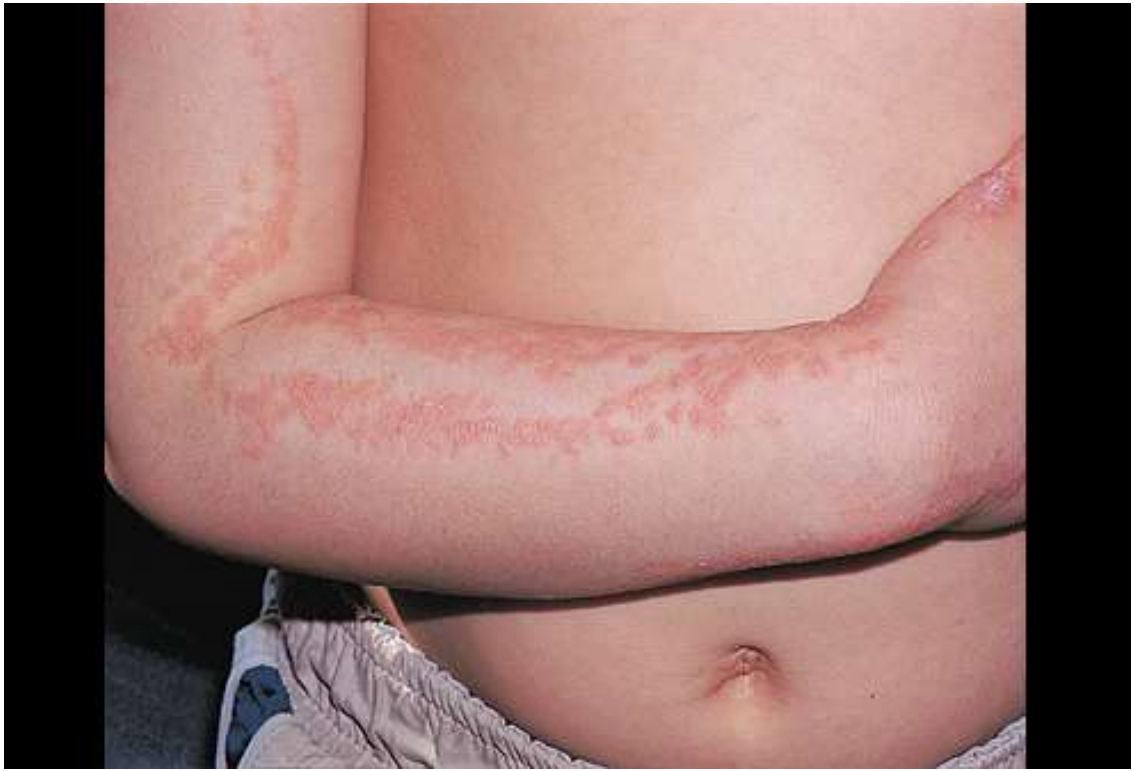
Pityriasis Rosea Variants

- Papular PR - young kids and darker skinned patients
- Inverse pattern - flexural accentuation
- Vesicular
- Pustular
- Urticarial

Pityriasis Rosea
Differential Diagnosis

- **Drug eruption**
- **Secondary syphilis**
- **Pityriasis lichenoides**
- **Nummular dermatitis**
- **Guttate psoriasis**
- Tinea corporis
- Tinea versicolor
- Parapsoriasis
- Erythema multiforme
- Urticaria
- Erythema dyschromicum perstans (ashy dermatosis)

Lichen Striatus



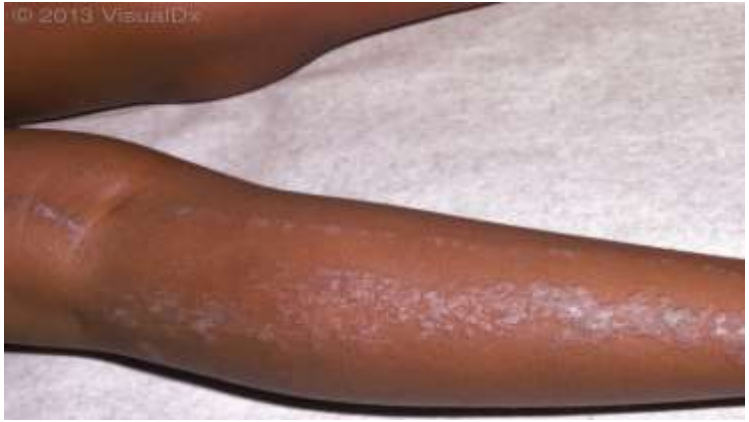
Lichen Striatus

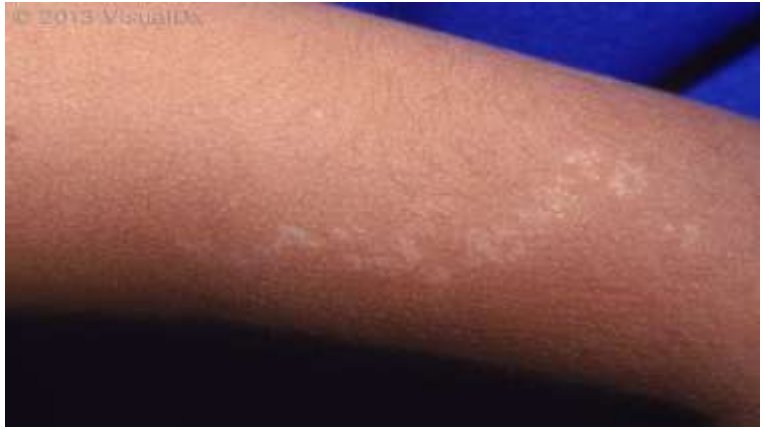
- Uncommon self-limited skin disorder of younger children of unknown etiology
- Has been reported in children as young as 3 months
- Presents with linear bands of slightly scaly, pinpoint, and lichenoid papules that follow the lines of Blaschko
- The onset it usually fairly rapid and may reach maximal involvement within a few days to weeks

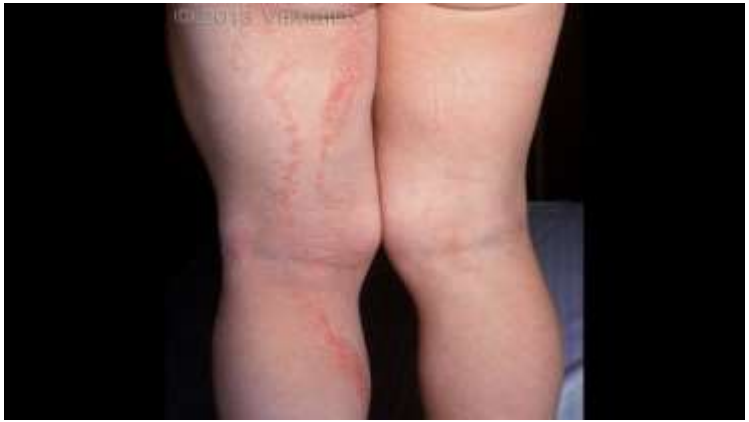
Lichen Striatus

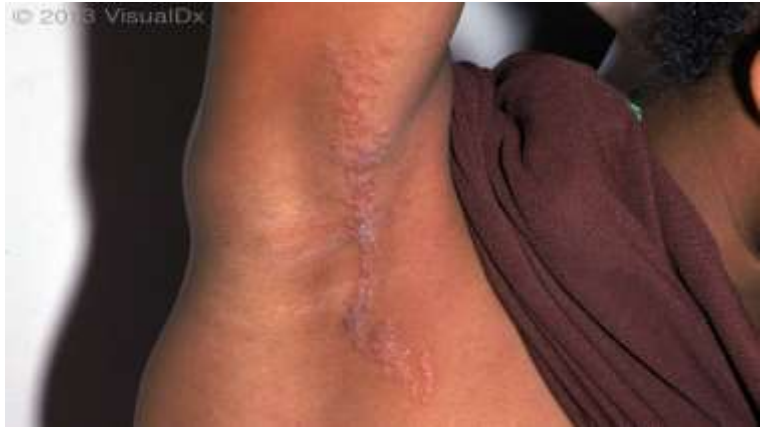
- Asymptomatic, rarely pruritic
- Lesions are usually on an extremity but can occur anywhere
- Often subtle and resolve leaving hypopigmentation or hyperpigmentation
- Treatment: Disease is self-limited, so aggressive treatment is not indicated
- Topical steroids for pruritus
- Typically resolves spontaneously within 1-2 years
- In adults, studies show good results with tacrolimus

Lichen Striatus





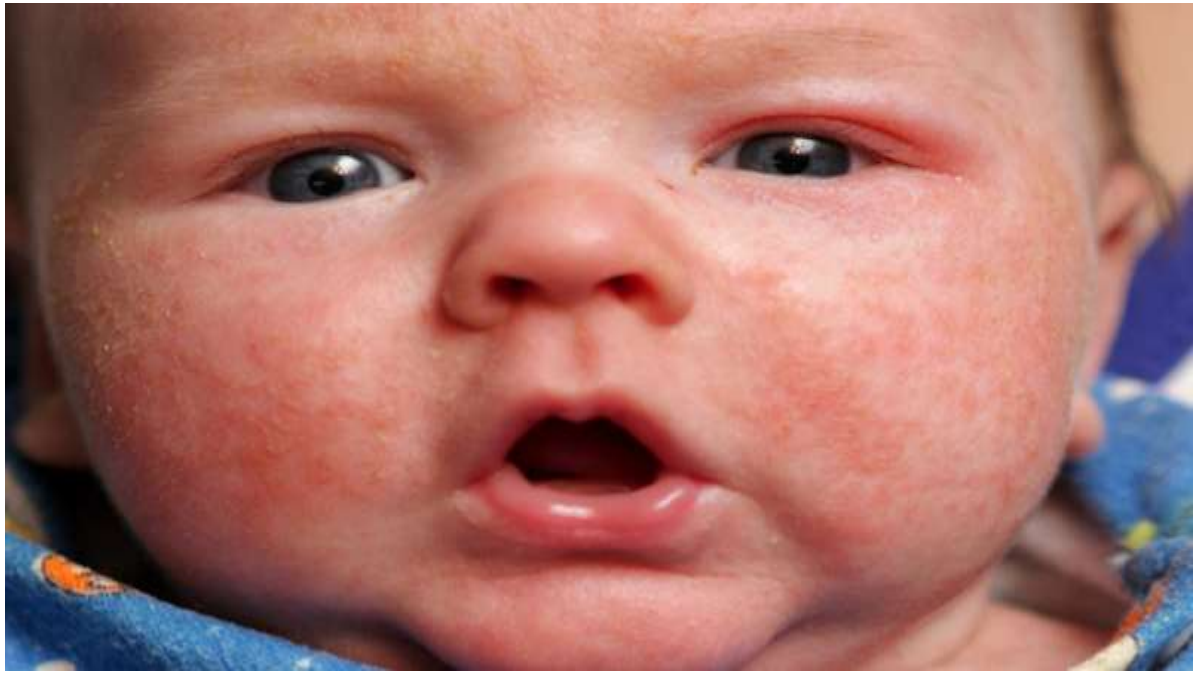




Lichen Striatus
Differential Diagnosis

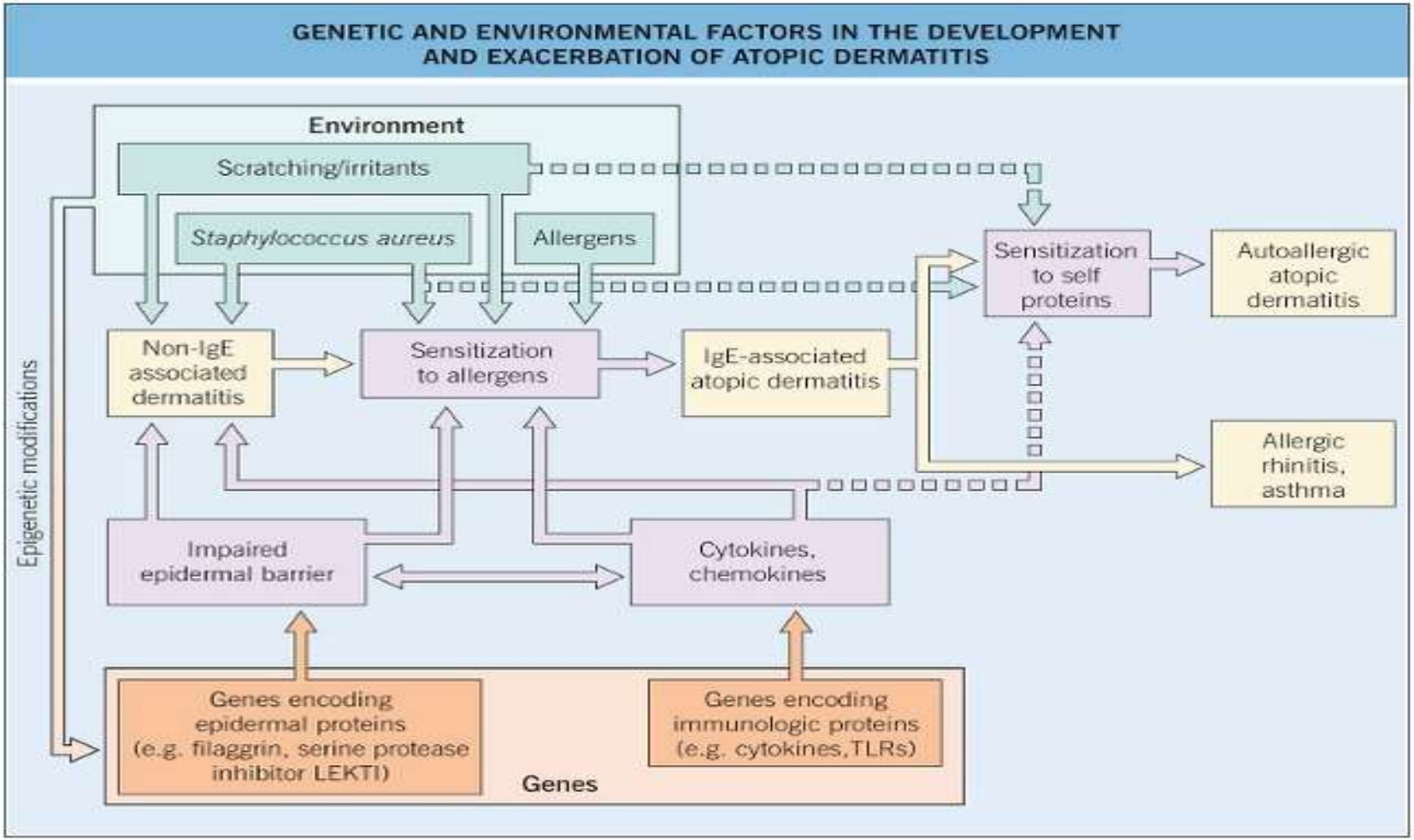
- Linear epidermal nevus
- Inflammatory epidermal nevus
- Linear Darier's disease
- Linear porokeratosis
- Incontinentia pigmenti
- Linear lichen planus

Atopic Dermatitis



Atopic Dermatitis

- Common inflammatory skin condition that typically begins during infancy, but occasionally in adulthood
- Occurs in 10-15% children
- Characterized by intense pruritus and a chronic or chronically relapsing course
- Th2 cytokine profile during the acute phase but transitions into a Th1 cytokine profile during the chronic phase



Bolognia, 3rd Edition

Filaggrin (FLG gene)
*fil*ament-*agg*regating prote*in*

- FLG is expressed in the granular layer of the stratum corneum. Encodes a protein that aggregates keratin filaments during terminal differentiation of the epidermis
- Encoded within the epidermal differentiation complex on chromosome 1q21 (Brown, 2008)
- Mutation responsible for Ichthyosis Vulgaris and in up 20-60% AD, depending on study (Elias, 2011)
- Various other genes that lead to increased susceptibility of AD, include KLK7, SPINK5, and CSTA..many others; FLG remains the most prominent (Cork, 2009)
- Presence correlated with AD that's early onset, relatively severe, persists into adulthood

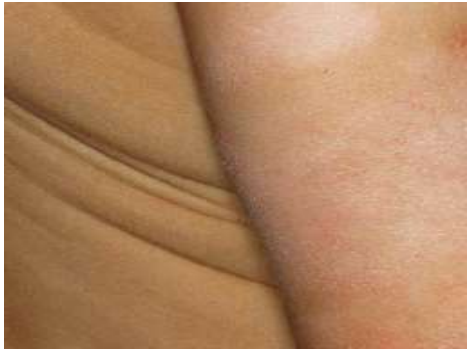
Atopic Dermatitis Clinical Features

- Infants: usually 2nd-3rd mo of life involving cheeks (often sparing central face), scalp, neck and extensor aspects of the extremities and trunk

- Children: shifts to more chronic inflammation with lichenification and a predilection for flexural sites; classic - antecubital and popliteal fossae, neck, wrists, ankles

- Adults: Also flexural. May present with hand derm, face (eyelid)

Atopic Dermatitis



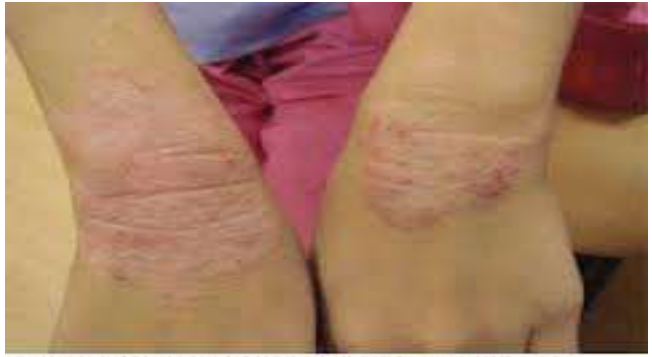
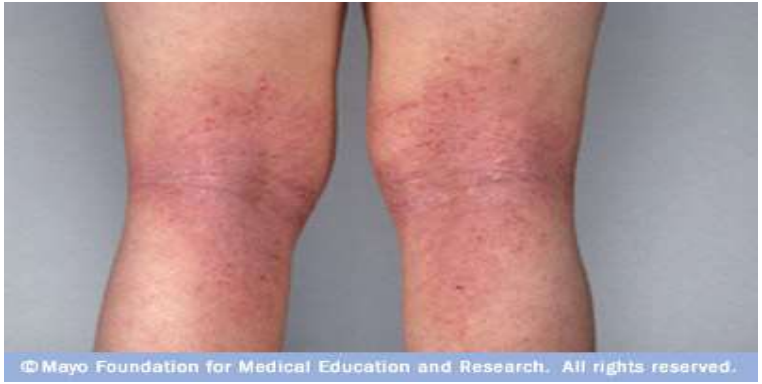
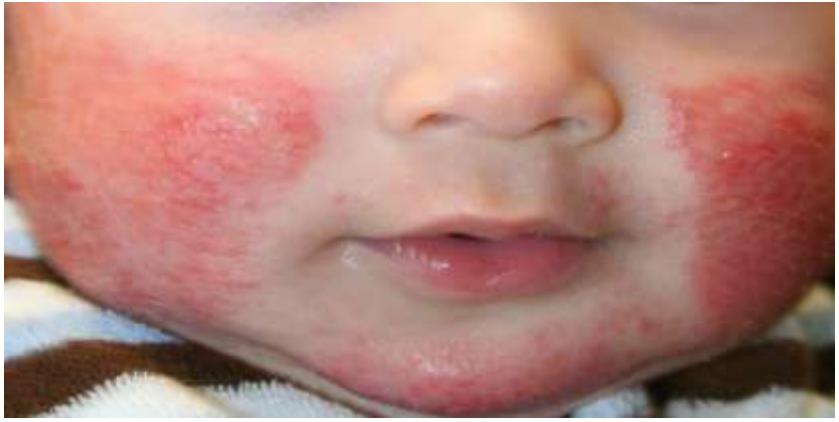


Figure 2: Childhood eczema with features of chronicity including induration and lichenification.

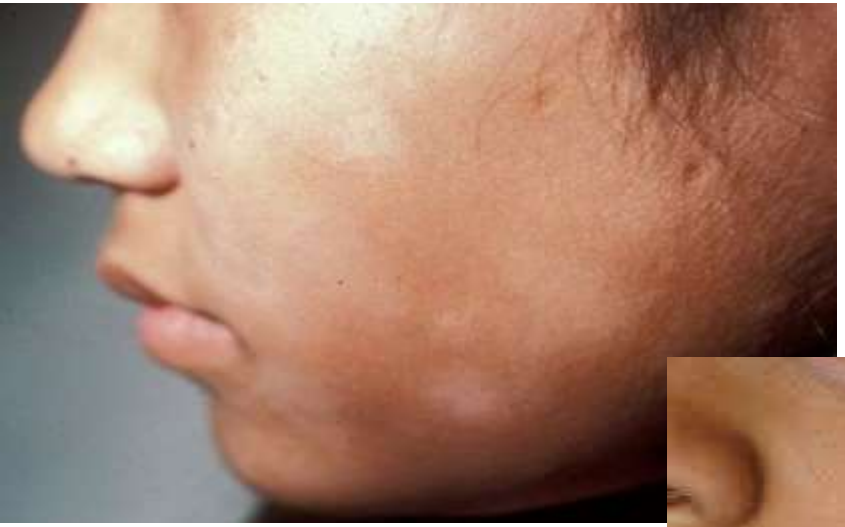


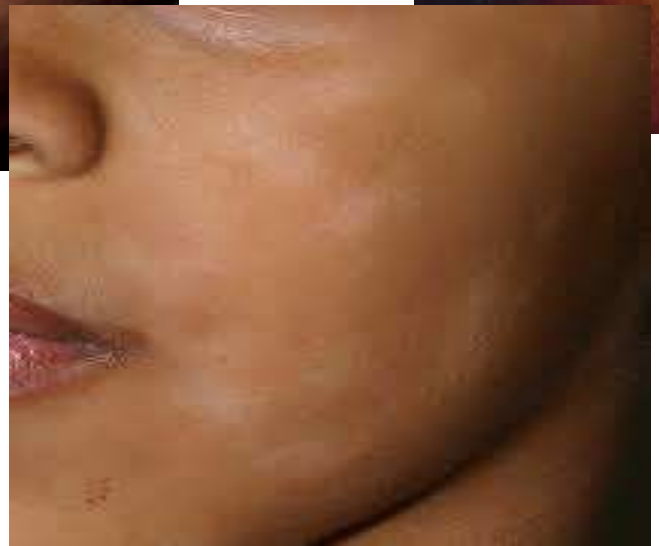


Pityriasis Alba

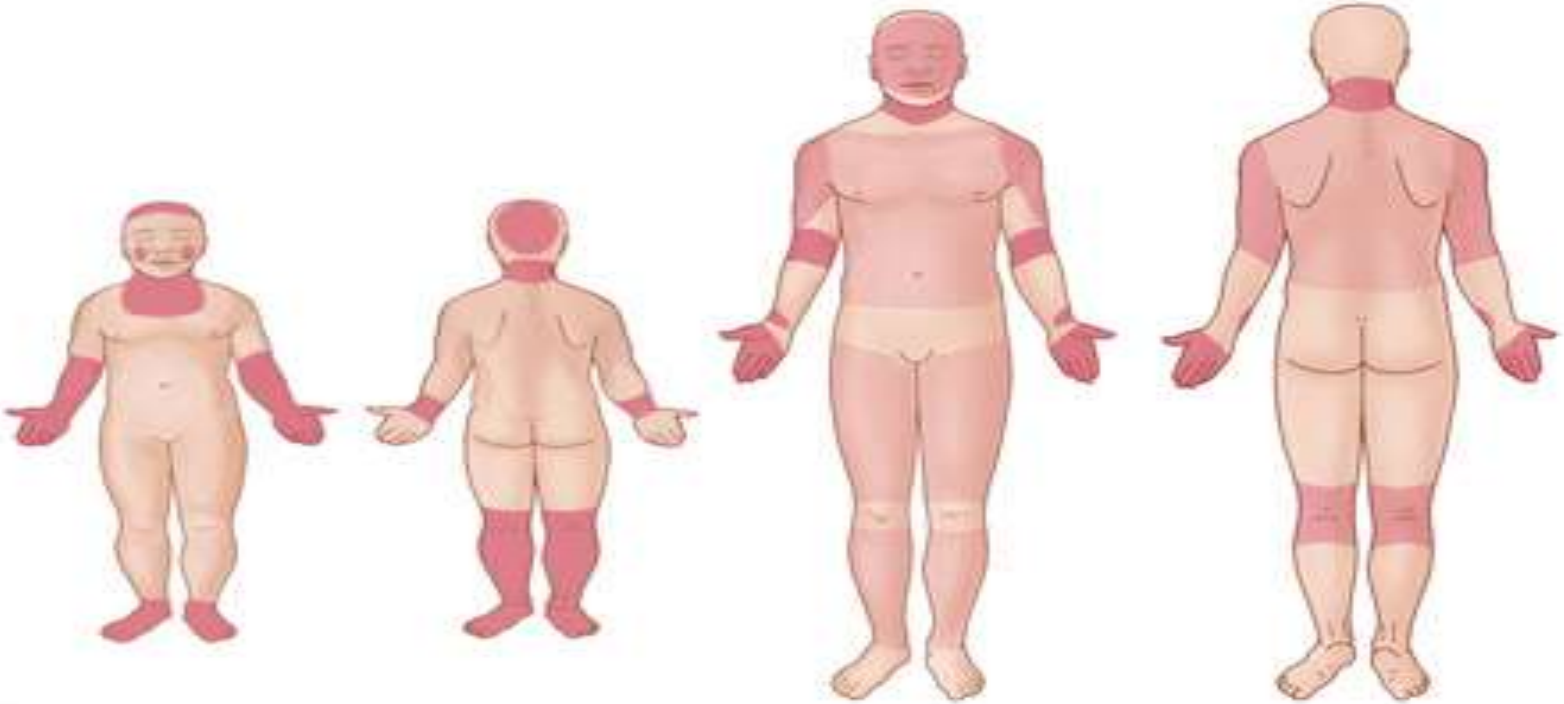
- Frequently affects children and adolescents with AD
- Characterized by multiple ill defined hypopigmented patches with fine scaling
- Typically face, but can occur on shoulders, arms
- Most obvious in individuals with darkly pigmented skin and or following sun exposure
- Thought to result from a low grade eczematous dermatitis that disrupts the transfer of melanosomes from melanocytes to keratinocytes

Pityriasis Alba

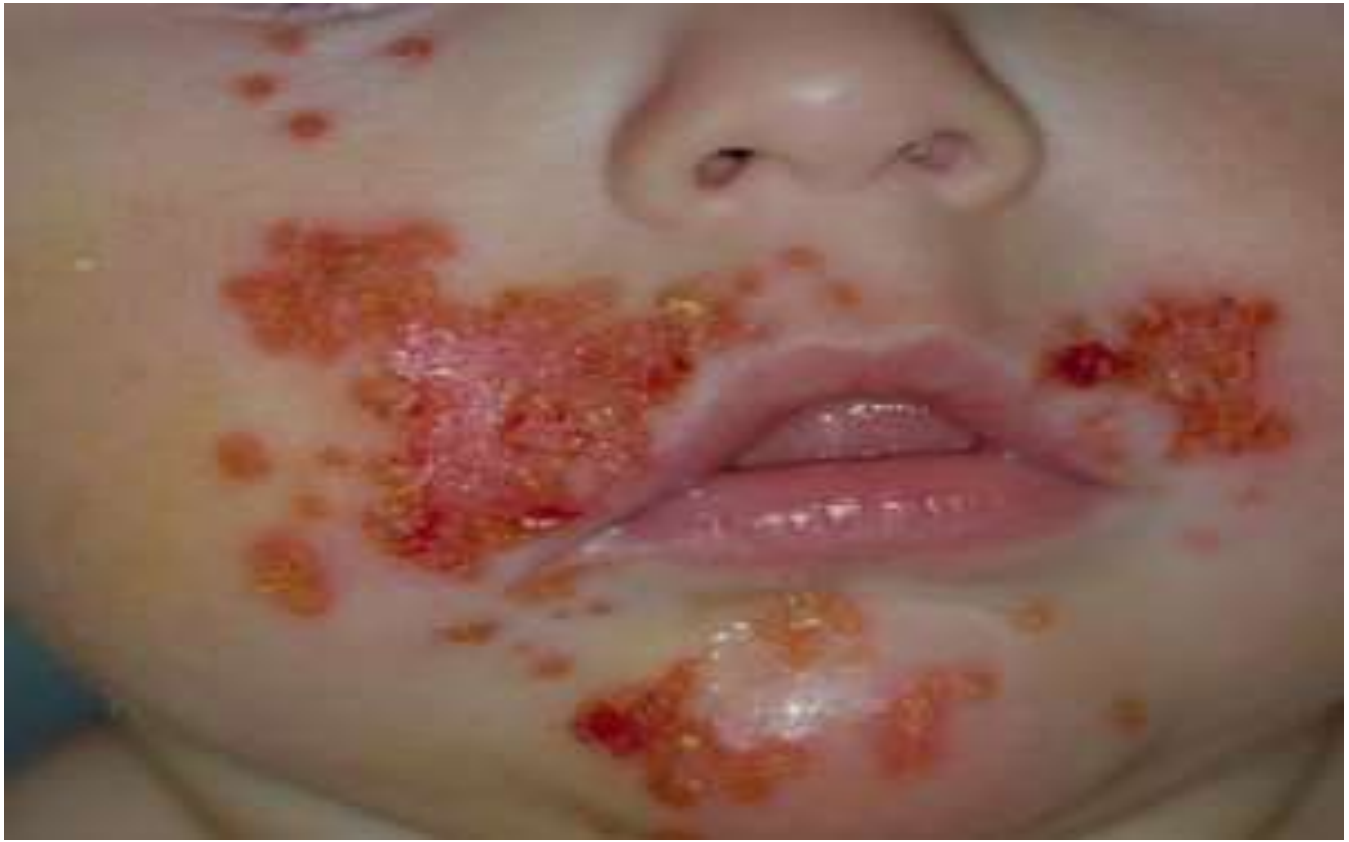




Common Sites of Eczema Outbreaks



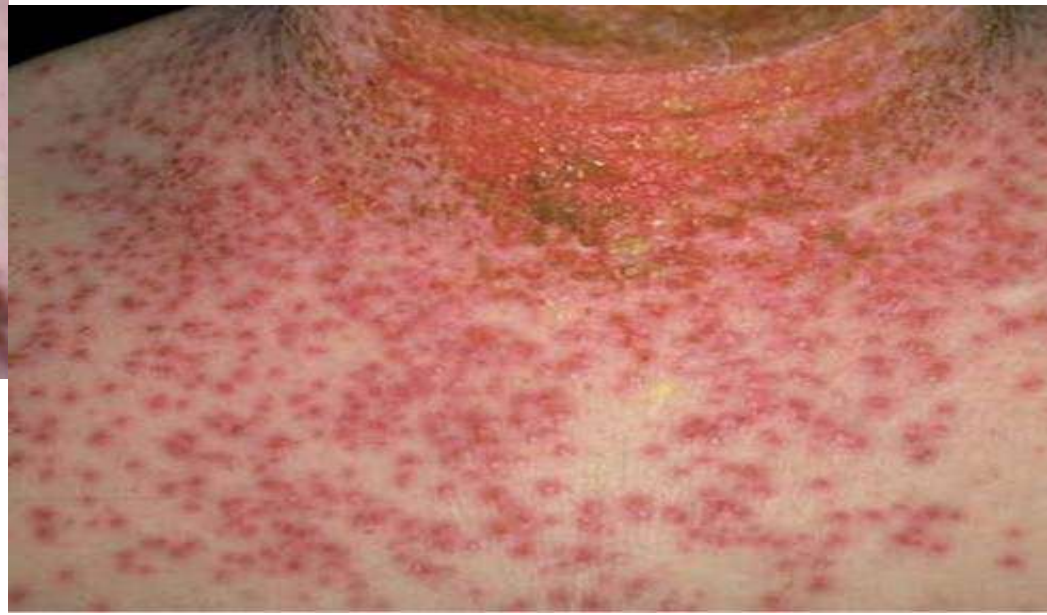
Complications: Impetigo



Complications: Impetigo

- Bacterial and viral infections represents the most common complication of AD.
- Considering that Staph Aureus colonizes the skin of majority of AD patients, its not surprising that impetigo occurs quite frequently.
- Bacterial infections may also exacerbate AD by stimulating the inflammatory cascade; such as through Staph exotoxins that act as superantigens.

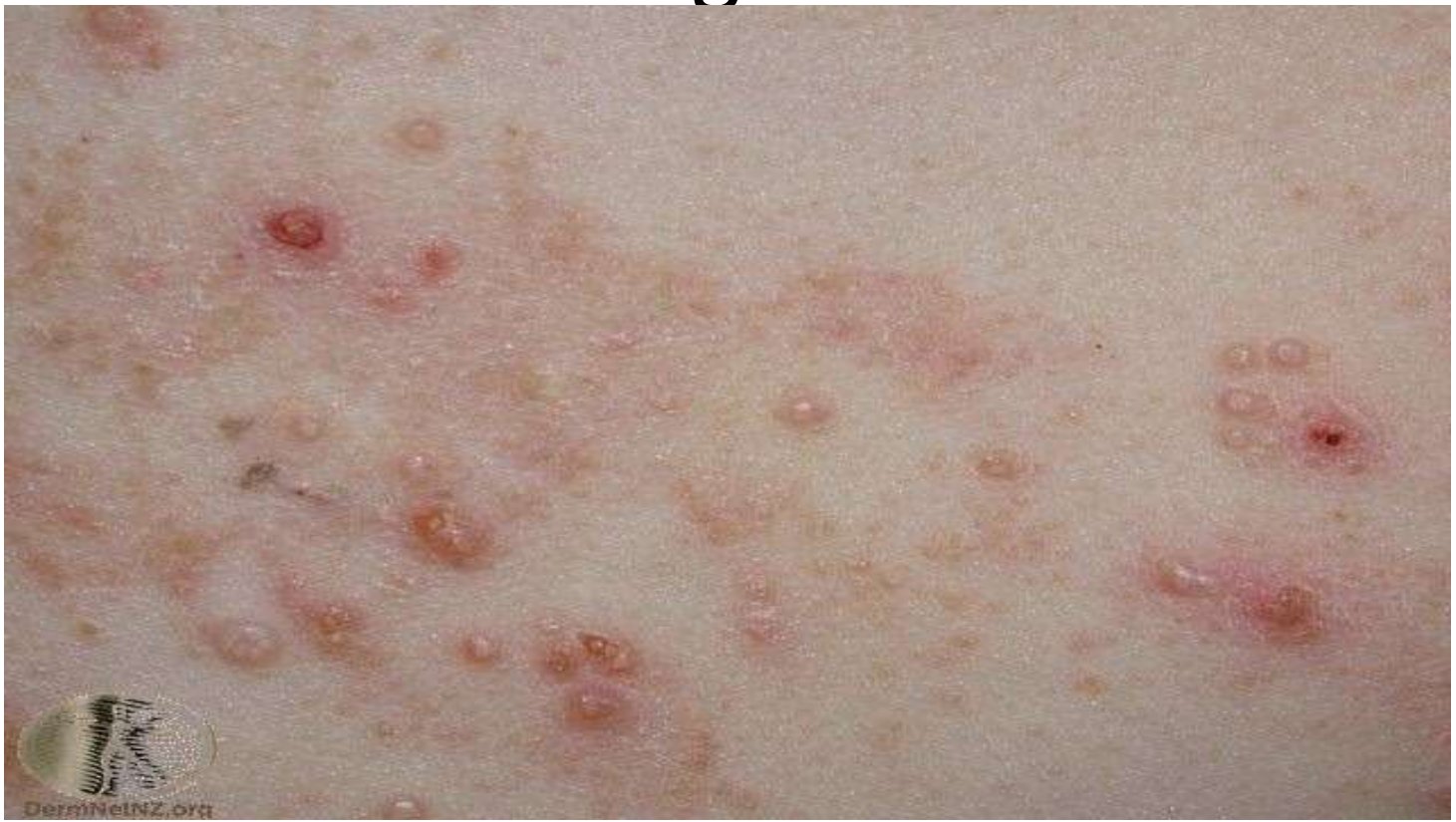
Complications: Eczema Herpeticum



Complications: Eczema Herpeticum

- Rapid dissemination of HSV over the eczematous skin of AD patients
- Present with vesicles, monomorphic punched out erosions with hemorrhagic crusting. Frequently widespread and may occur at any site, with a predilection for head, neck, and trunk.
- Often associated with fever, malaise, and LAD, and complications may include 2ndary bacterial infection.

Complications: Molluscum Contagiosum



Ocular complications:

- Keratoconjunctivitis – ocular itching, burning, tearing, discharge, blepharitis, eyelid dermatitis
- Subcapsular cataracts (anterior more specific to AD, posterior more common)
- Keratoconus
- Retinal detachment

AD Treatment

- The main idea…
  - a proactive approach to management is recommended, including avoidance of trigger factors, daily use of emollients, and anti-inflammatory therapy to control subclinical inflammation as well as overt flares

THERAPEUTIC LADDER FOR ATOPIC DERMATITIS	Evidence
Emollients (basic therapy)	1
Topical corticosteroids	1
Topical calcineurin inhibitors	1
UVB (narrowband>broadband), UVA-UVB, UVA1 or oral PUVA	1
Systemic corticosteroids (short term for severe acute flares; "rebound" exacerbations often occur upon discontinuation)	2
Cyclosporine (short/intermediate term)	1
Azathioprine	1
Mycophenolate mofetil/enteric-coated mycophenolate sodium	1*/2
Methotrexate	1*/2
Interferon-γ	**
IVIg	2†
Omalizumab	2†
Rituximab	2
Other (crude coal tar, hydroxychloroquine, extracorporeal photochemotherapy)	2–3
Adjunctive therapies	
Wet wraps, open wet dressings or soaks (combined with topical corticosteroids for acute flares) Dilute sodium hypochlorite (bleach) baths Treatment of associated bacterial, viral or fungal infections Oral antihistamines for antipruritic† and sedative effects Leukotriene antagonists† Sodium cromoglycate (topical or oral)† Probiotics† (may have efficacy in primary prevention)	

1= evidence based support from prospective controlled trial
2 =retrospective trial or large case series
3=small series or individual case reports

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Recommendations for nonpharmacologic interventions for the treatment of atopic dermatitis

(AAD Guidelines for AD care 2014)

- **Moisturizers** - **strong evidence** that their use can **reduce disease severity** and the need for pharmacologic intervention
- **Bathing** is suggested - there is **no standard for the frequency** or duration of bathing
- **Moisturizers should be applied soon after bathing** to improve skin hydration
- **Limited use** of nonsoap **cleansers** (that are neutral to low pH, hypoallergenic, and fragrance free)
- Addition of oils, emollients, and most other additives to bath water and the use of acidic spring water cannot be recommended at this time, because of insufficient evidence.
- Use of **wet-wrap therapy with or without a topical corticosteroid** can be **recommended for patients with moderate to severe AD**

Recommendations for the use of topical antimicrobials and antiseptics for the treatment of atopic dermatitis

(AAD Guidelines for AD care 2014)

- Except for bleach baths and intranasal mupirocin, no topical antistaphylococcal treatment has been shown to be clinically helpful in patients with AD, and is not routinely recommended.
- In patients with moderate to severe AD and clinical signs of secondary bacterial infection, bleach baths and intranasal mupirocin may be recommended to reduce disease severity.

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