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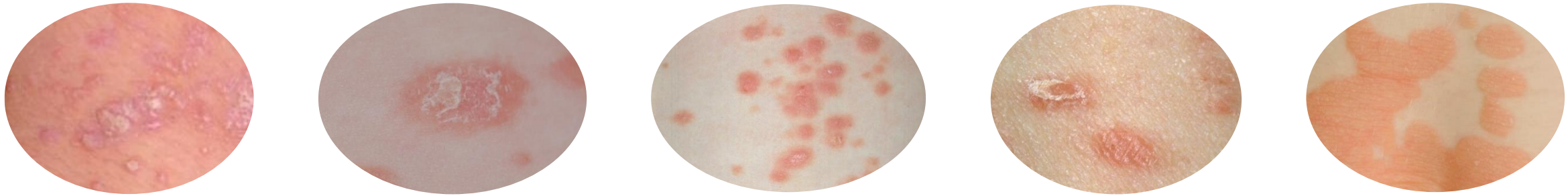
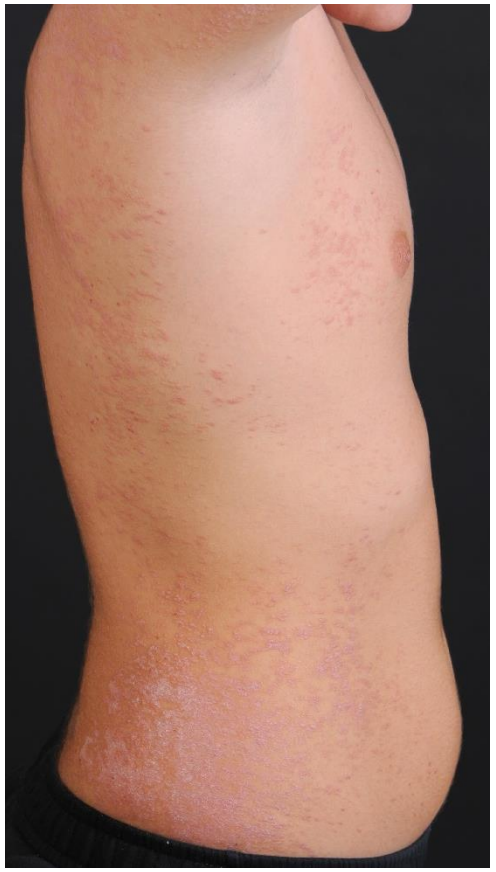
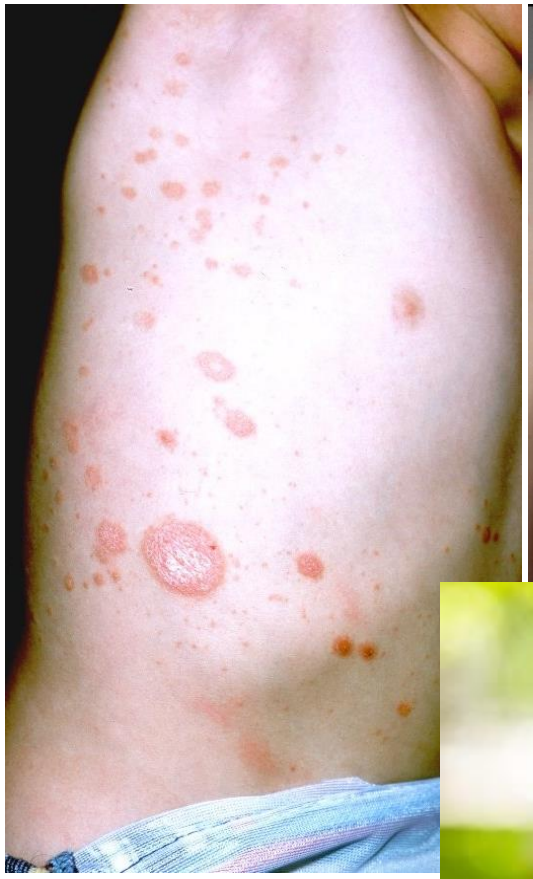
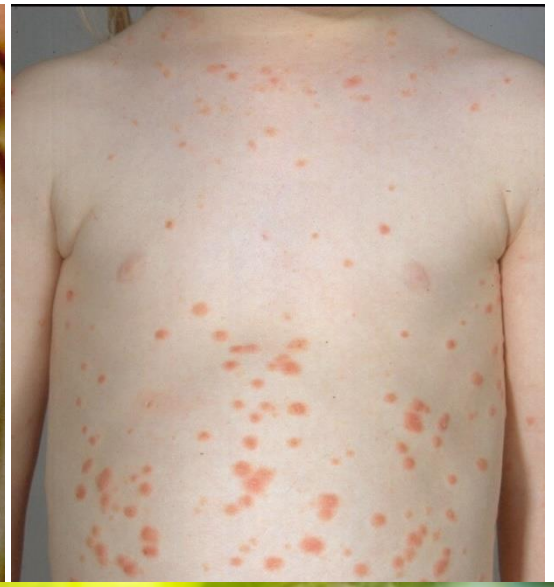
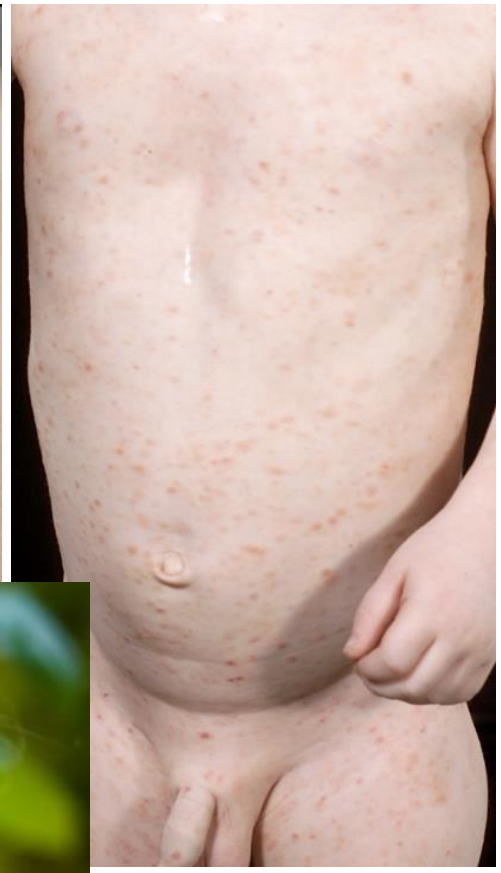
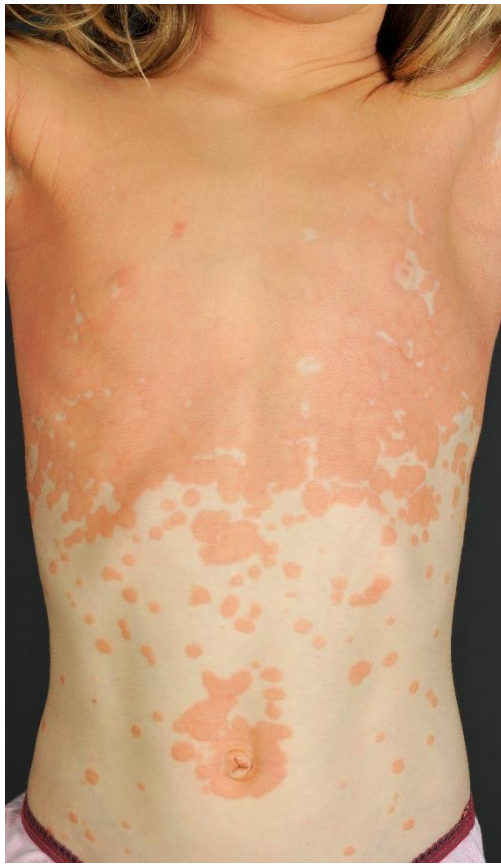
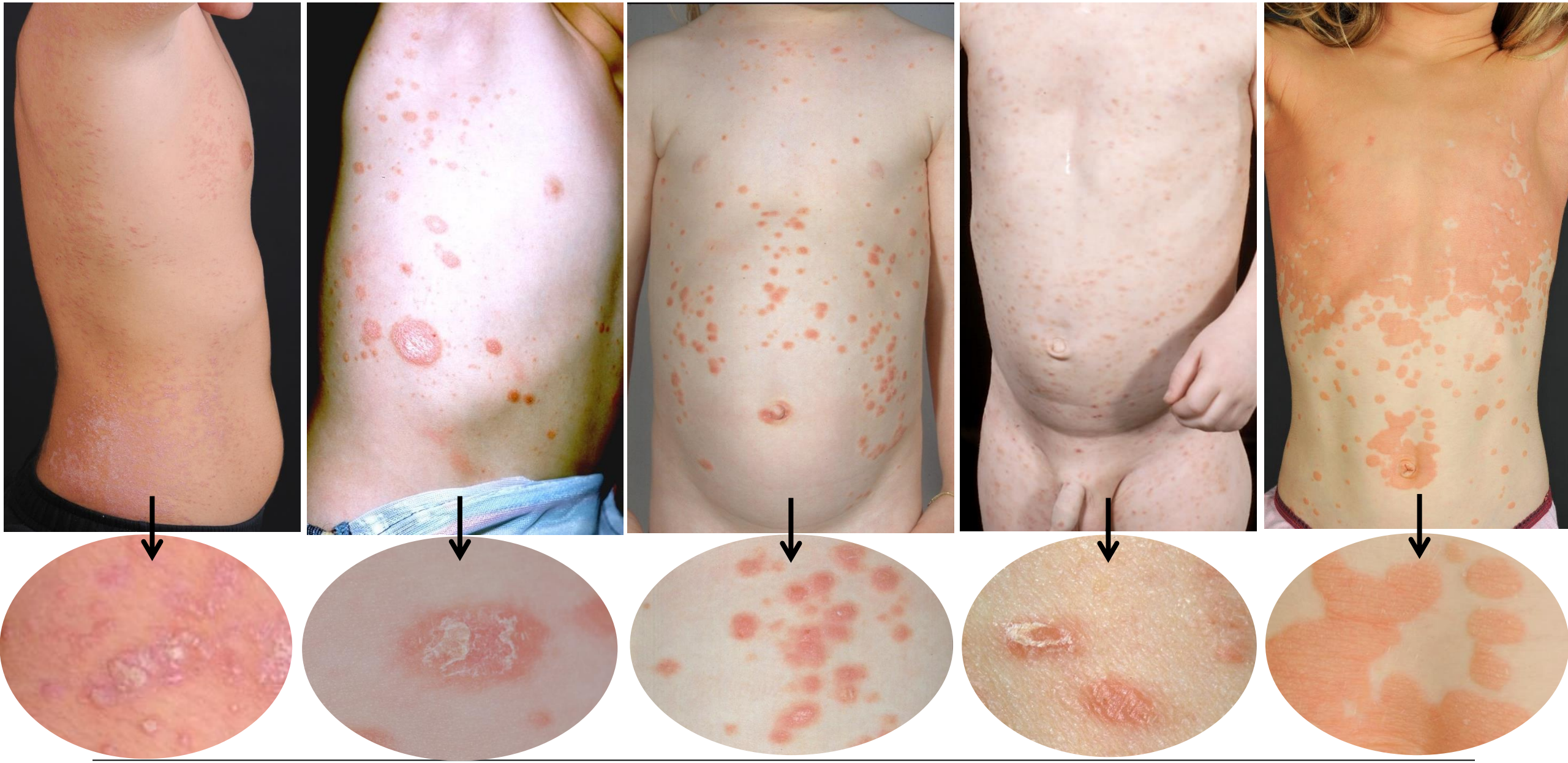
Psoriasis and other papulosquamous disorders



Dr. med. Isabelle Luchsinger

Oberärztin Pädiatrische Dermatologie Universitäts-Kinderspital Zürich

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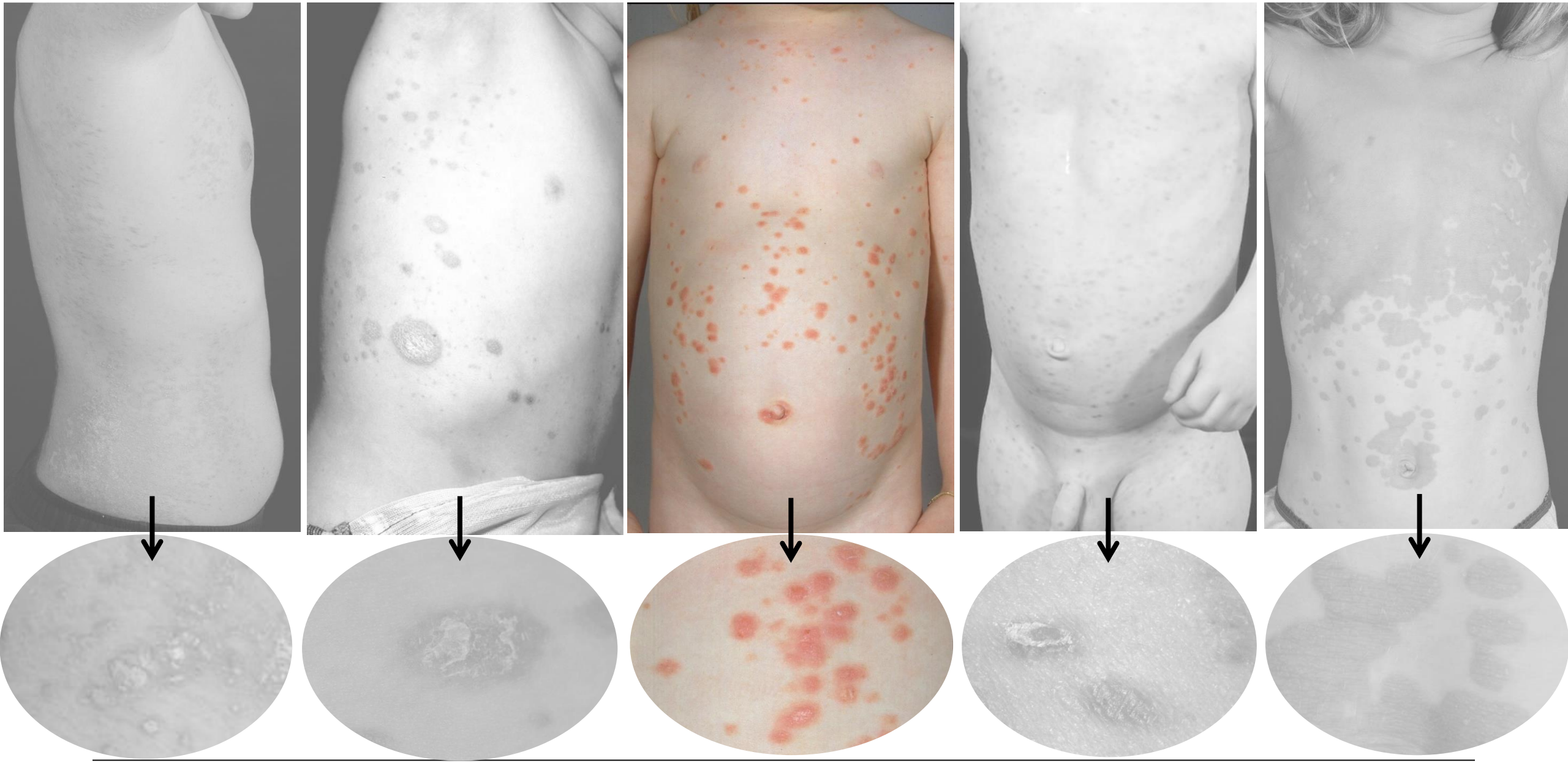

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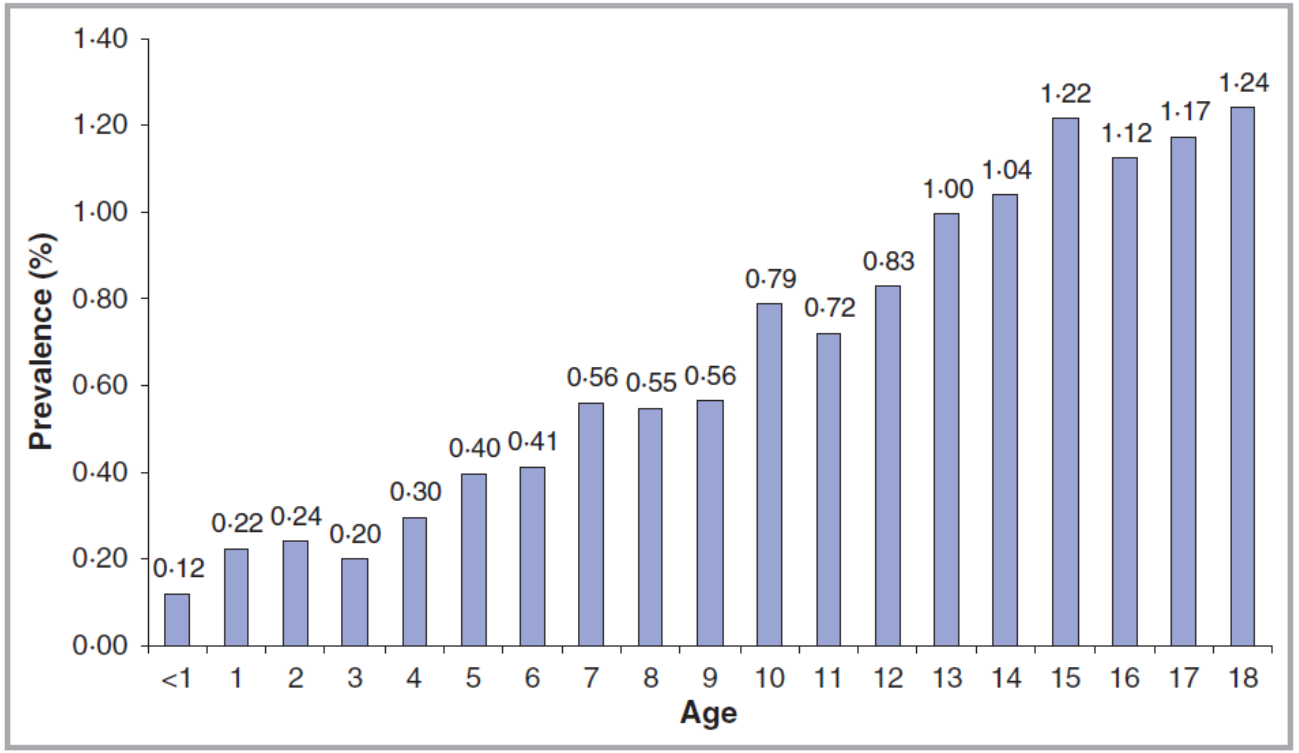
Pediatric psoriasis - epidemiology

- prevalence: 0 – 2.1% (Taiwan – Italy)
- linear increase by age: 0.2% at 2 years to 1.2% at 18 years
- ~4% of all dermatoses in children
- ~30% with psoriasis onset childhood/adolescence
- Caucasian (Europe!) >> Asian, African
- female > male (?)



database from German health insurance organization, n= 1.3 million non-selected individuals

Paller AS et al, J Drugs Dermatol. 2018
Burden-Teh et al, Br J Dermatol 2016
Augustin M et al, Br J Dermatol 2010

Pediatric psoriasis – clinical manifestations

Psoriasis variant	Relative frequency	Compared to adults
Plaque Facial Scalp	**~65%** (9 – 91.9%) ~20% (3.5 – 56.7%) 40-60%	= **>>** =
Guttate	**~20%** (1.6 – 48.2%)	**>>**
Inverse Diaper psoriasis	**~8%** (5 – 15%)	**>>** exclusively
Nail	**~20%** (2 – 39.3%)	=
Pustular	**rare**	**<**
Erythrodermic	**Extremely rare**	**<**

Mild >>> severe

Menter A et al, J Am Acad Derm 2020
Burden-Teh et al, Br J Dermatol 2016
Tollefson et al, J Am Acad Dermatol 2010
Cordoro KM, Adv Dermatol 2008

Facial Psoriasis

- more common in childhood
- involvement of the periorbital area is most typical
- plaques more clearly delineated than in atopic dermatitis, less pruritic, possible annular configuration
- 5% of pediatric patients show an eczema/psoriasis overlap (FA pos for AD and Pso)

- Treatment: Topical calcineurin-inhibitors!

  →Data supporting long-term use of TCIs are robust, documenting safety and efficacy

Scalp Psoriasis

- commonly the initial site of psoriatic involvement
- girls > boys (brushing?)
- Sebopsoriasis in adolescent patients

- Treatment:

  Salicylic acid: be careful in young children!

  do not use in babys

  toddlers: max. 2% concentration, max. 10% KOF
  school age: 2-3 (-5)%, max. 20% KOF

  Topical corticosteroids (in the form of oils, solutions, or foams)

Diaper-area Psoriasis/ inverse Psoriasis

- sharply defined plaques, bright-red coloration, shininess, and large, dry scales
- scale may not be visible
- Many infants with psoriasiform lesions elsewhere
- Koebner phenomenon responsible

- Therapy:
  - Mild topical corticosteroids (class II), cave occlusion!
  - topical calcineurin-inhibitors
  - Antifungal therapy if Candida

Guttate psoriasis

- Acute rash with psoriasiform scaling
- occurs in children and young adults
- is often the first manifestation of psoriasis
- often triggered by group A streptococcal infection (pharyngitis or perianal area)
- 2/3 with upper respiratory tract infection 1 to 3 weeks before acute flare

Guttate psoriasis

Risk for lifelong disease:
40% progress to plaque type
often more severe psoriasis
than with initial plaque disease

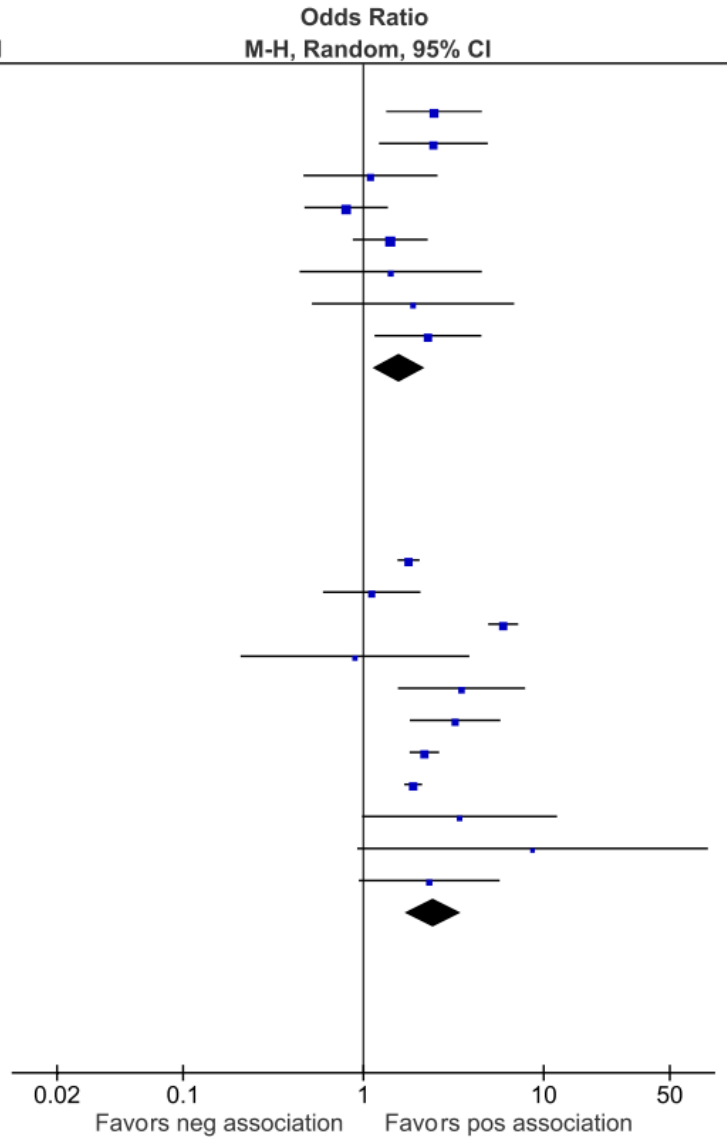
Nail involvement

- in 20% - 50% of pediatric patients with psoriasis
- more commonly during the second decade of life
- boys > girls
- associated with more severe disease

Pourchot D et al. Pediatr Dermatol 2017

Pediatric psoriasis – cardiovascular and metabolic comorbidities

Study or Subgroup	Psoriasis Events	Psoriasis Total	Control Events	Control Total	Weight	Odds Ratio M-H, Random, 95% CI
Overweight						
Boccardi 2009	42	96	24	100	15.2%	2.46 [1.34, 4.54]
Ergun 2016	43	267	11	151	13.1%	2.44 [1.22, 4.90]
Lee 2016	18	135	9	73	10.1%	1.09 [0.46, 2.58]
Mahe 2015	28	261	34	261	17.4%	0.80 [0.47, 1.37]
Paller 2013	72	409	27	205	19.1%	1.41 [0.87, 2.27]
Tom 2015	8	40	6	40	6.4%	1.42 [0.44, 4.53]
Torres 2014	7	20	6	27	5.4%	1.88 [0.52, 6.86]
Zhu 2012	52	332	11	146	13.4%	2.28 [1.15, 4.51]
Subtotal (95% CI)		**1560**		**1003**	**100.0%**	**1.58 [1.14, 2.19]**
Total events	270		128			
Heterogeneity: Tau² = 0.09; χ^2 = 11.86, df = 7 (P = 0.11); I² = 41%						
Test for overall effect: Z = 2.74 (P = 0.006)						
Obese						
Augustin 2010	214	2549	16256	331758	13.6%	1.78 [1.55, 2.05]
Ergun 2016	33	267	17	151	9.6%	1.11 [0.60, 2.07]
Kwa 2017	113	1506	65259	4857674	13.3%	5.96 [4.92, 7.22]
Lee 2016	5	135	3	73	4.1%	0.90 [0.21, 3.87]
Mahe 2015	26	261	8	261	7.9%	3.50 [1.55, 7.88]
Paller 2013	83	409	15	205	10.1%	3.22 [1.81, 5.75]
Paller 2019	170	7686	316	30744	13.4%	2.18 [1.80, 2.63]
Tollefson 2018	862	29957	463	29957	13.7%	1.89 [1.68, 2.12]
Tom 2015	11	40	4	40	5.0%	3.41 [0.98, 11.85]
Torres 2014	5	20	1	27	2.1%	8.67 [0.92, 81.34]
Zhu 2012	30	332	6	146	7.2%	2.32 [0.94, 5.70]
Subtotal (95% CI)		**43162**		**5251036**	**100.0%**	**2.45 [1.73, 3.48]**
Total events	1552		82348			
Heterogeneity: Tau² = 0.23; χ^2 = 133.43, df = 10 (P < 0.001); I² = 93%						
Test for overall effect: Z = 5.03 (P < 0.001)						



Comorbidity	Odds ratio
Overweight	1.58
Obesity	2.45
Diabetes	2.32
Hypertension	2.19
Hyperlipidemia	2.01
Metabolic syndrome	3.53
Ischemic heart disease/heart failure	3.15

Phan K et al, Pediatr Dermatol 2020

Other medical comorbidities in pediatric psoriasis

- Juvenile idiopathic arthritis/psoriatic arthritis (often starting prior to skin manifestations ≠ adults)
- Uveitis
- Crohn's disease, (ulcerative colitis)
- Vitiligo

→ rates of comorbidities correlating with severity of psoriasis

Edson-Heredia E et al, Adv Ther 2021

Comorbidity screening guidelines in pediatric psoriasis

Condition	Screening recommendation
Overweight/Obesity	Yearly from 2 years of age (BMI percentile)
Type 2 Diabetes Mellitus	Every 3 years from 10 years of age if obese/if overweight and risk factors for DM (fasting serum glucose)
Dyslipidemia	In all patients 9 to 11 and 17 to 21 years of age (fasting lipid panel)
Hypertension	Yearly starting at 3 years of age
NAFLD	If obese or risk factors for NAFLD, starting at 9 to 11 years of age (ALT)
IBD	Look for decreased growth rate, weight loss, other symptoms consistent with IBD
Arthritis	Regular review of systems and physical examination; if arthritis screening for uveitis recommended (ophthalmologist)
Mood disorders	Yearly for depression/anxiety at any age; substance abuse beginning at 11 years of age (e.g. Patient Health Questionnaire (PHQ)-4 tool)
Quality of life	Regularly (CDLQI)

Osier E et al, JAMA Dermatol 2017

Quality of life in children with psoriasis

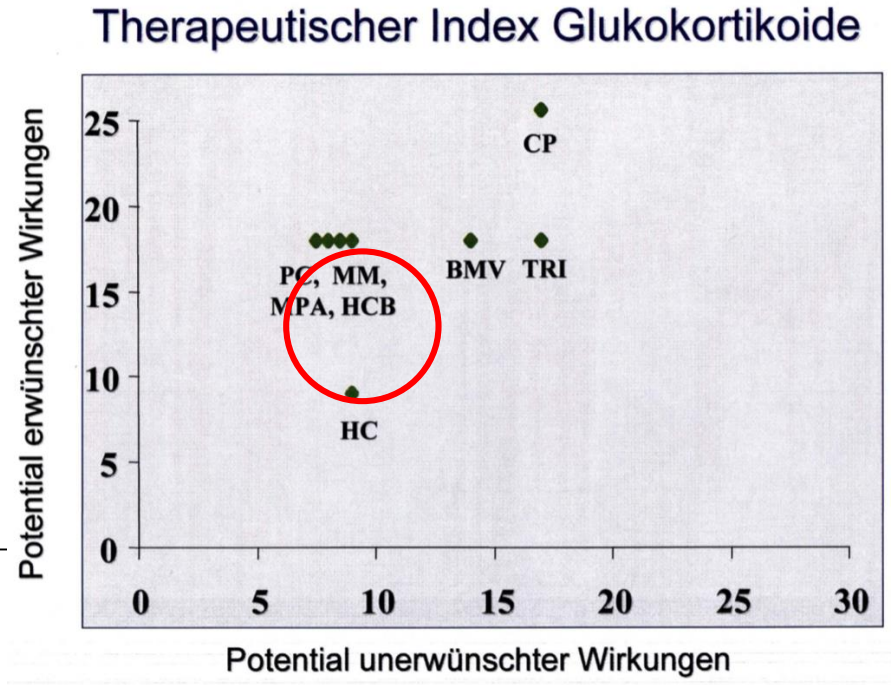
- QoL is **moderately impaired** in children and adolescents with psoriasis
- quality of life of **both** the pediatric patient and the parents negatively affected
- higher QoL impairment
  - patients diagnosed at an older age
  - more severe psoriasis
  - children with arthritis
- QoL impairment in different childhood diseases:

  Diabetes < Arthritis = **Psoriasis** = Asthma < psychiatric diseases

Randa H et al. Acta Derm Venereol 2017
Tollefson MM et al. J Am Acad Dermatol 2017
de Jager MEA et al. Br J Dermatol 2010

Treatment of pediatric psoriasis

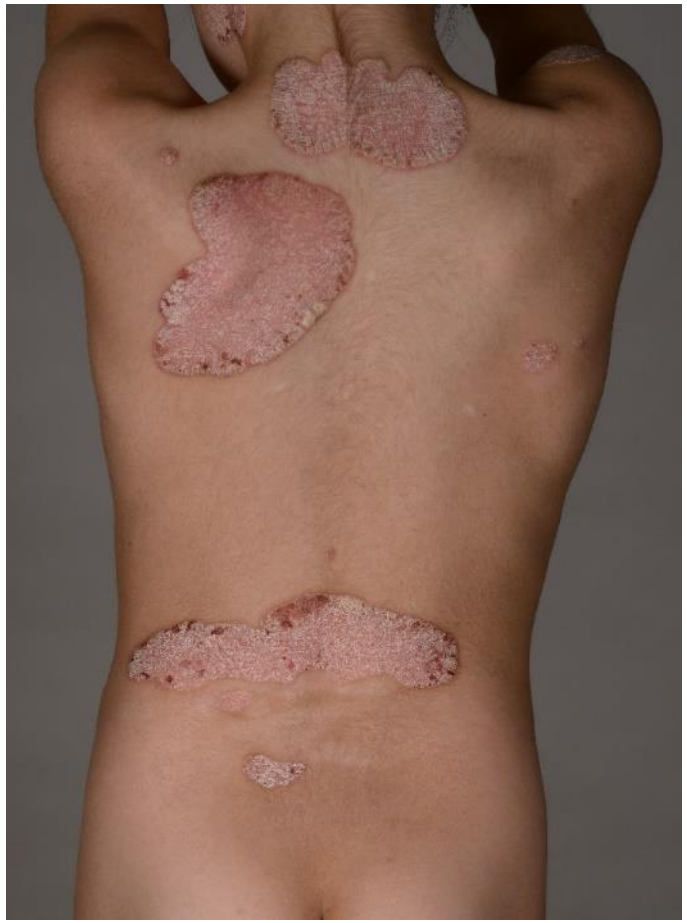
- Patients and parents must understand the chronicity
- Inform about Koebner phenomenon
- Removal of potential trigger factors (medications, etc.)
- rapid intervention for streptococcal infection



Systemic treatment in pediatric psoriasis

MTX

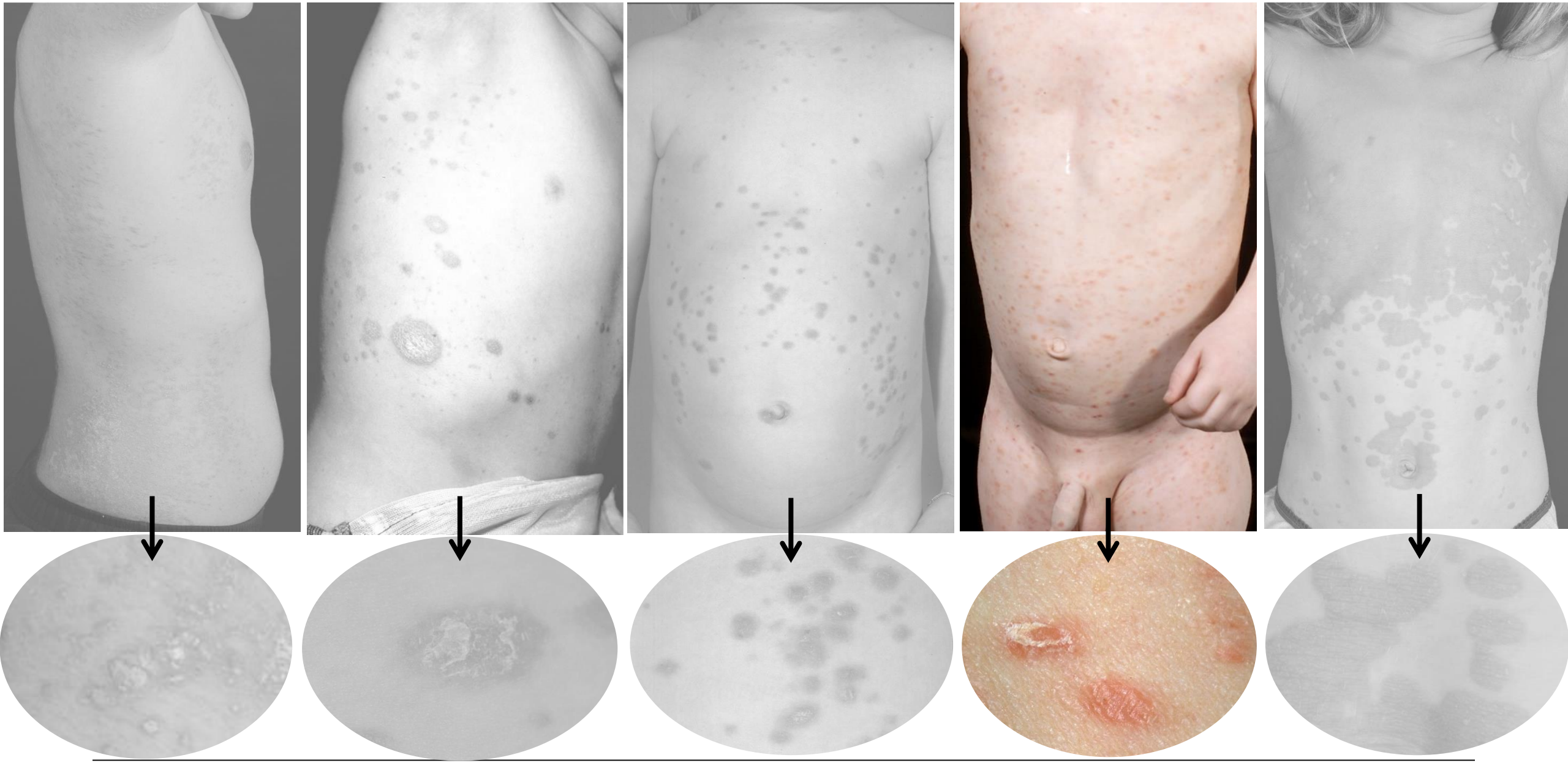
anti-TNF



anti-IL 12/23

anti-IL 17

UVBnb



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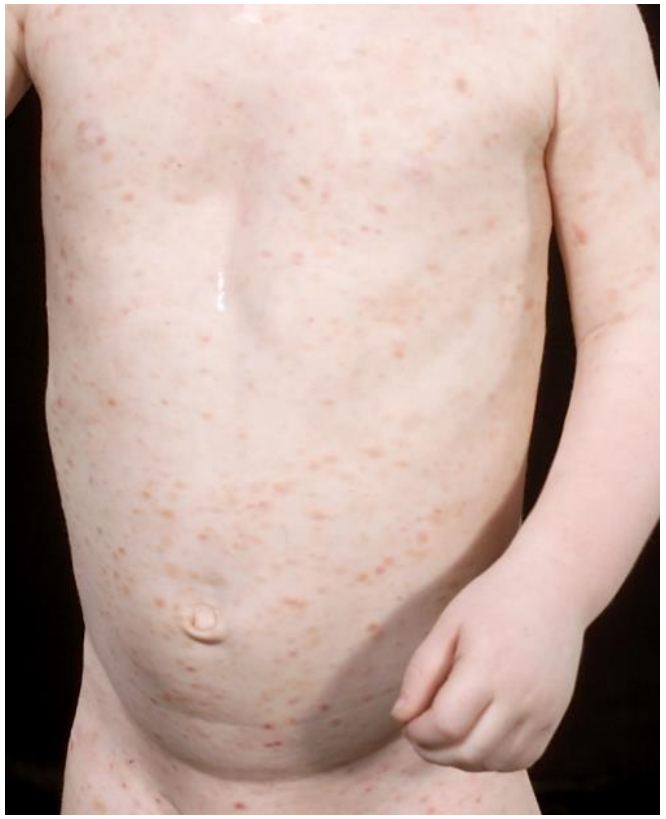
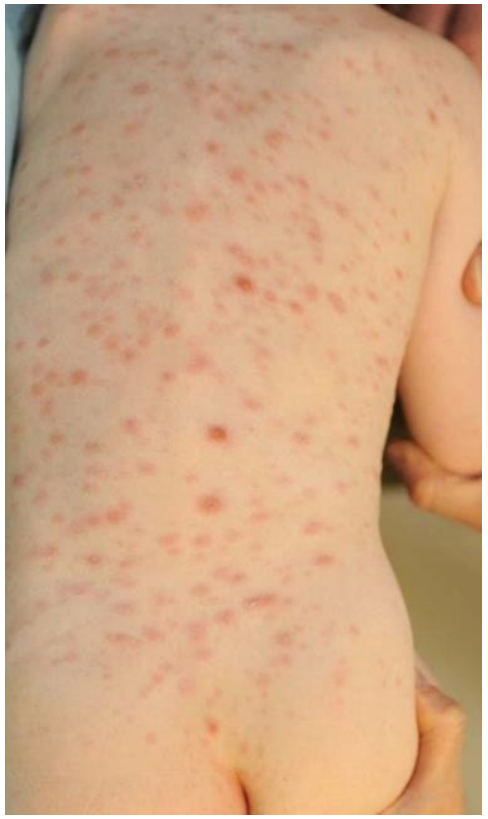
Pityriasis lichenoides

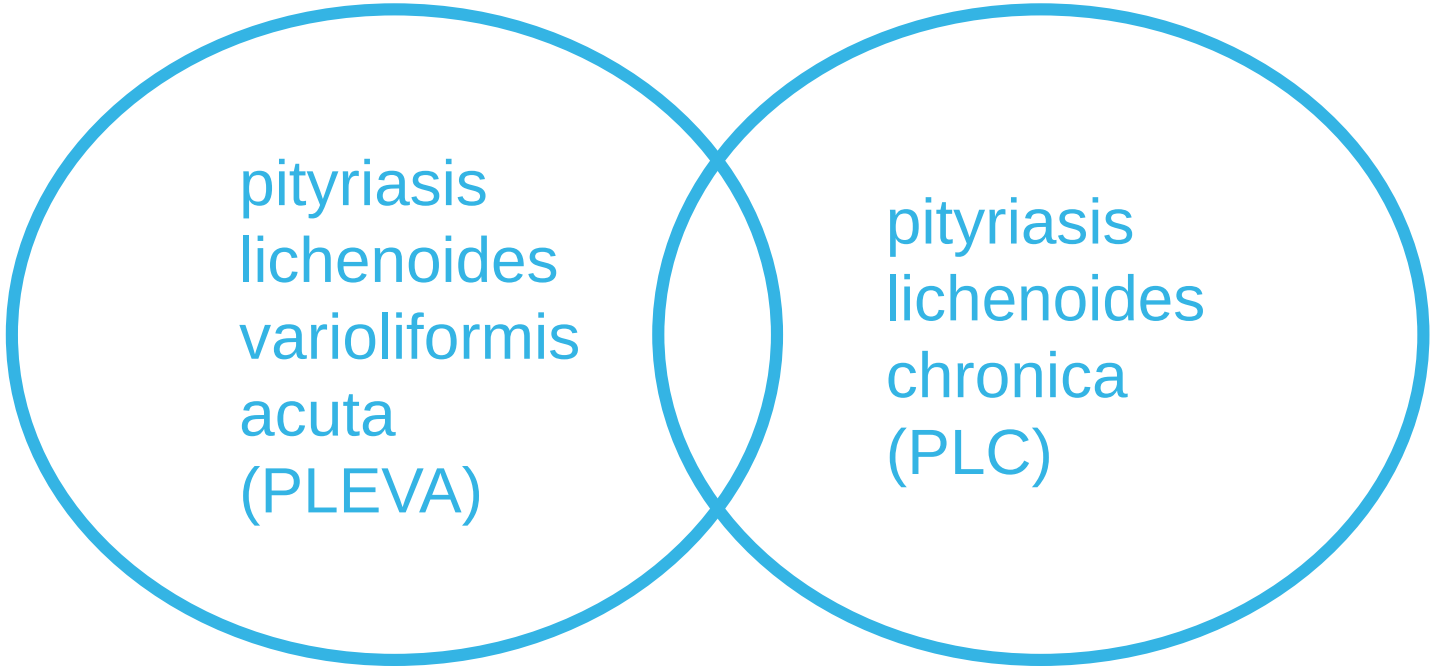
- benign lymphoproliferative process
- association of preceding exposure to viral infection (parvovirus or HHV-8) or immunization (MMR) → abnormal immune response to a viral antigenic trigger
- predominantly pediatric disorder
  – mean age at onset 5-7 years
  – boys : girls = 3 : 1





Geller L et al. Pediatr Dermatol 2015
Wahie S et al. Br J Dermatol 2007

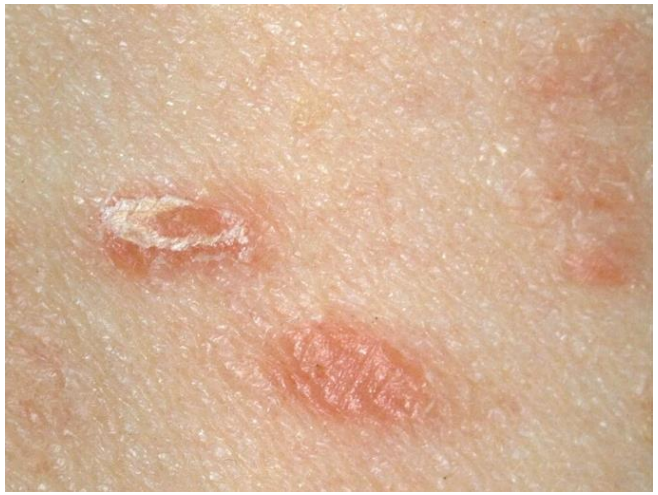
Pityriasis lichenoides: spectrum of inflammatory skin diseases



Pityriasis lichenoides varioliformis acuta (PLEVA)

- acute-to-subacute
- polymorphous eruption: oval or round, reddish-brown macules and papules → successive crops, vesicular, necrotic, and sometimes purpuric lesions
- Symmetric distribution on the trunk, buttocks and proximal extremities (flexors)
- Varioliform scars and post-inflammatory hyper- and hypopigmentation
- Symptoms include burning and pruritus
- mean disease duration of 35 months

Pityriasis lichenoides chronica (PLC)



- may begin de novo or may evolve from PLEVA
- erythematous-brownish papules with central scaling
- lesions often leave a hyper- and hypopigmentated macule
- trunk, rarely hand, feet and head
- usually asymptomatic, no pruritus
- a mean duration of at least 78 months

Pityriasis lichenoides: therapy

- relapsing course with long periods of remission
- treatment response limited, better in PLEVA than PLC
- most effective treatments:
  - phototherapy (47% response rate)/heliotherapy (33%)
  - topical corticosteroids (27%)
  - antibiotics (25%) (erythromycin, azithromycin, tetracycline)
  - methotrexate, cyclosporine, TNF-alpha Inhibitors

- topical corticosteroids and/or oral antihistamines may be helpful for pruritus and insomnia, but do not hasten resolution

Zang JB et al. Pediatr Dermatol. 2018

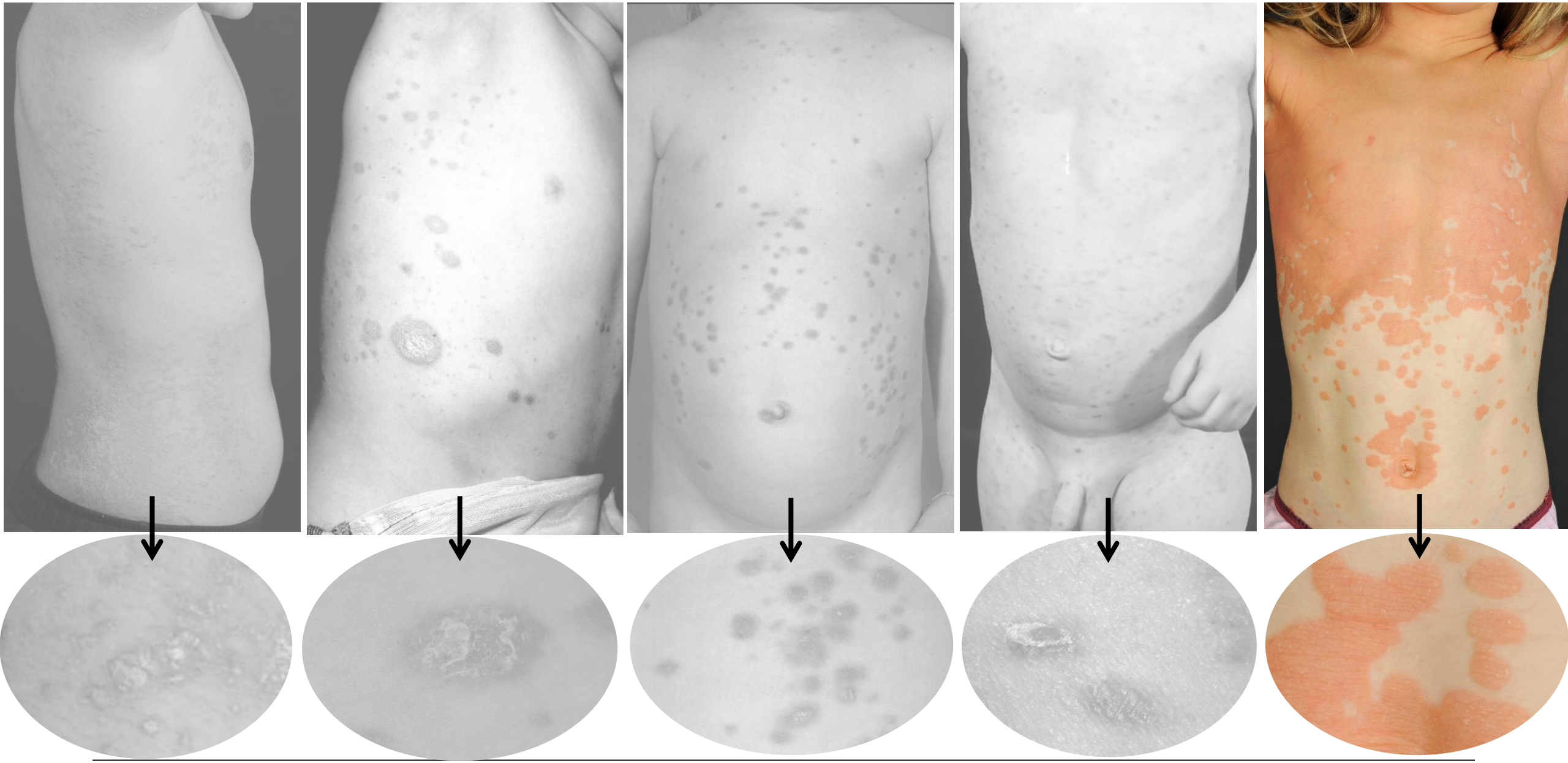
Follow-up of PL patients

– cutaneous T-cell lymphoma (mostly mycosis fungoides)
  – Age at CTCL diagnosis, 10.5 years median
  – Duration of disease before CTCL presentation, 6 years median
    →true transformation
    or
    →Lymphomatoid papulosis (associated with MF) mimicking PLC

– it may be important to monitor patients when the disease is persistent

→Biopsy especially when
  →prolonged disease duration (> 5 years)
  →Changing morphology (nodules, hypopigmentation or poikiloderma)
  →localization to sun-protected areas

Sibbald C. et al. British Journal of Dermatology 2016



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Pityriasis rubra pilaris

- Chronic skin disorder
- small follicular keratotic papules surrounded by **salmon-colored erythema**
- disseminated symmetrical yellowish-pink scaly plaques surrounding **islands of normal skin**
- Pruritus only occasionally

Pityriasis rubra pilaris

Hyperkeratosis of palms and soles: in the majority

- Can be present before or after the appearance of other features
- “keratodermic sandal”: sharply demarcated border
- Associated edema

Pityriasis rubra pilaris

- cephalic involvement 40%: cape-like configuration
- nails: dystrophic 13% (thickening, onycholysis, transverse striations, and subungual keratosis)

Pityriasis rubra pilaris

- many cases cannot easily be fit into any of these classifications
- onset in children acute or begins on the scalp and forehead and extends caudally
- Biopsy can aid in diagnosis (characteristic follicular keratosis, epidermal parakeratosis and dermal mononuclear infiltrates, particularly surrounding the hair follicle)

Type		Incidence	Clinical Features
I	Classic adult	Most adults	Follicular keratotic papules, first on face and extending caudally; progresses to generalized keratoderma with islands of sparing; palms and soles usually involved; generally clears within 3 years
II	Atypical adult	Rare	More ichthyosiform scaling; coarse palmoplantar keratoderma; long duration
III	Classic juvenile	14% to 35% of children	Same as type I
IV	Circumscribed juvenile	Most common type in children	Thick plaques on knees, elbows; palms, and soles involved
V	Atypical juvenile	Rare, familial; onset in first years of life	"Sclerodermatous" changes on palms and soles; follicular hyperkeratosis
VI	**PRP** in association with human immunodeficiency virus	Only described in 1 4 year old child	Classified as type IV based on distribution

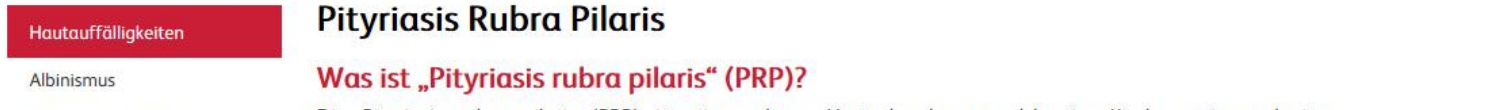
Griffiths WA. Pityriasis rubra pilaris. Clin Exp Dermatol. 1980;5(1):105–12.

Pityriasis rubra pilaris

- clinical course variable
- 50% clearance within 6 months
- two-thirds of patients with type III and IV juvenile PRP: protracted course lasting longer than 3 years.
- prognosis does not correlate with acute versus gradual onset or extent of involvement
- spontaneous remissions and exacerbations
- some children with change of phenotype to psoriasis

Pityriasis rubra pilaris: treatment

- Emollients, topical corticosteroid, tazarotene, keratolytic agents
- face: calcineurin inhibitor
- More extensive disease: systemic retinoid therapy, methotrexate, cyclosporine, azathioprine
- TNF-alpha inhibitors +/- retinoids
- shared pathogenic inflammatory pathway with psoriasis: IL-23–TH17-axis: Ustekinumab
- Ixekizumab, Secukinumab
- psychological support
- support group



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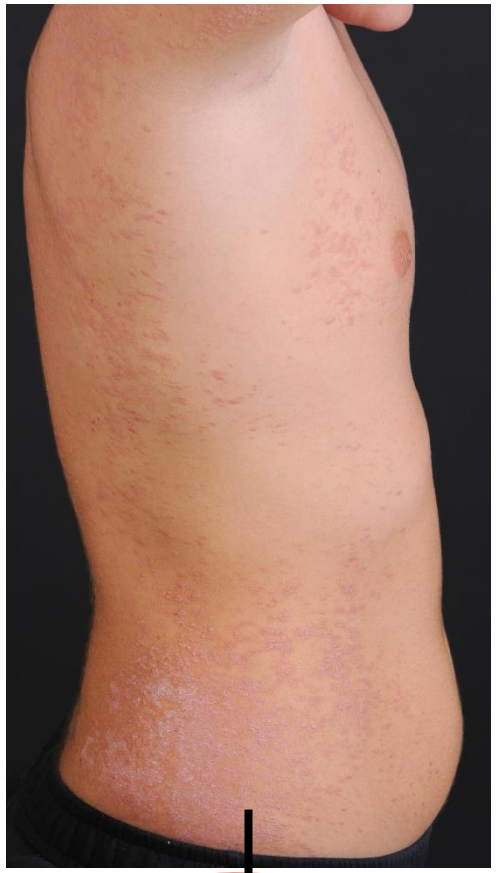
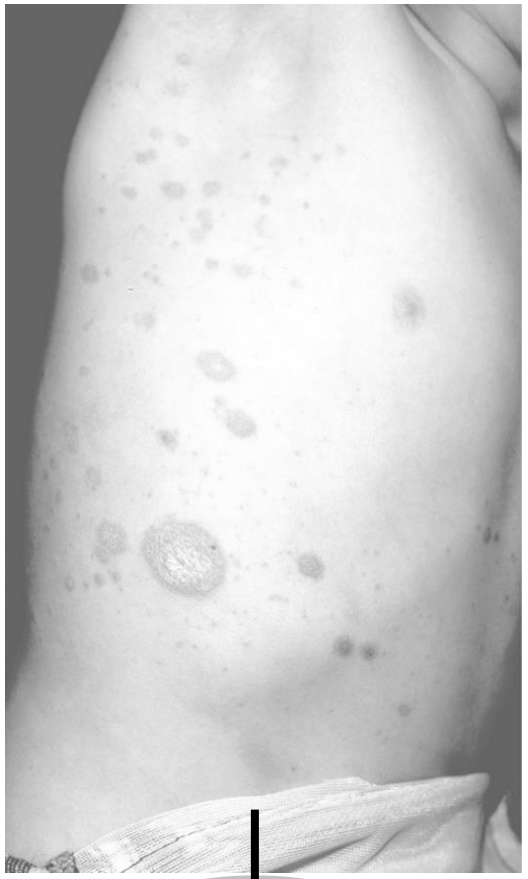
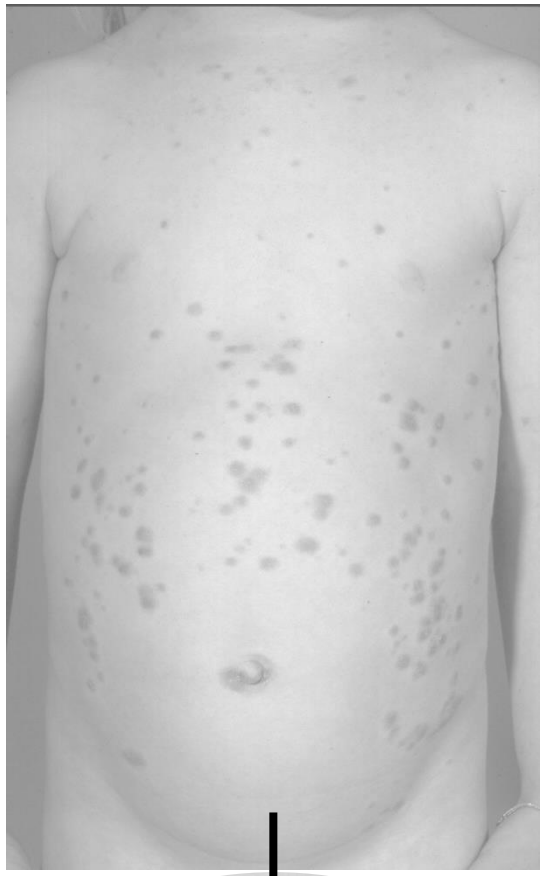
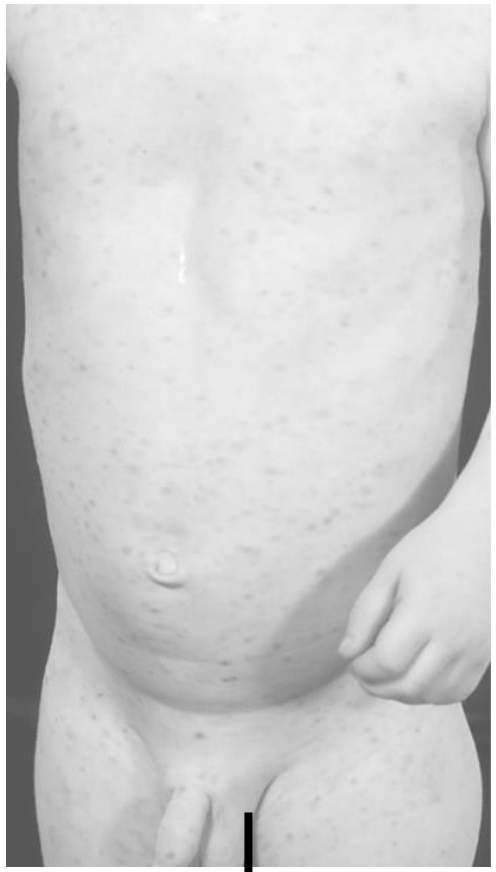
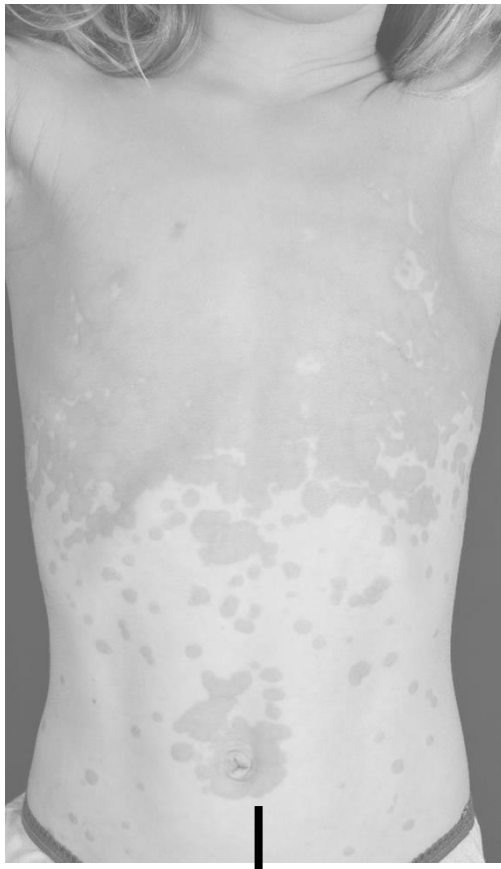
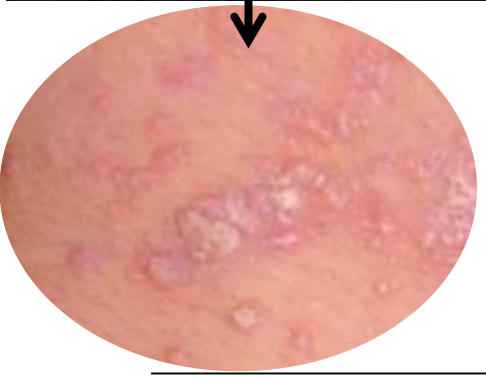
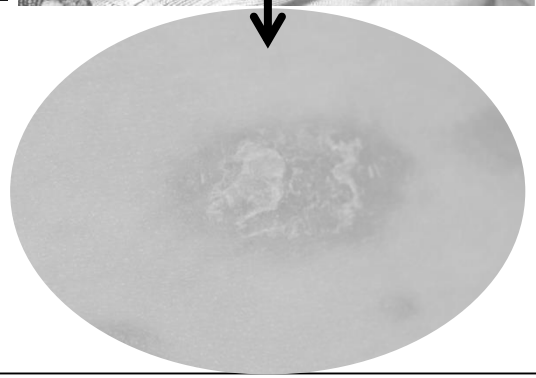
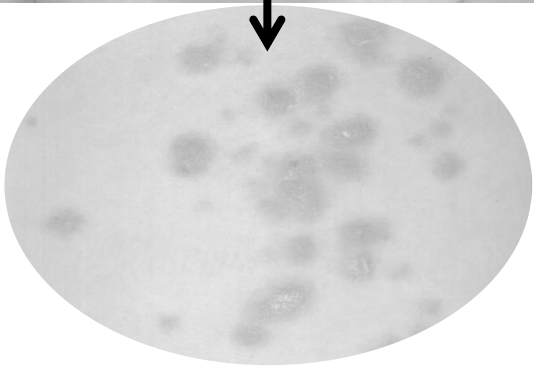
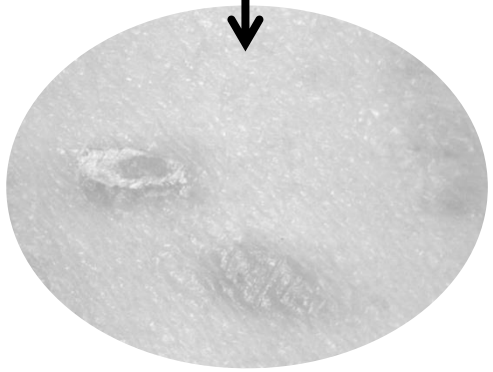
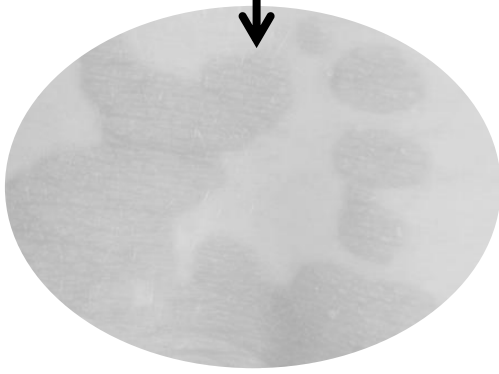
Davis KF et al. Arch Dermatol 2007
Petrof G et al. J Eur Acad Dermatl Venereol 2013
Feldmeyer L et al. *JAMA Dermatol*. 2017

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Lichen planus

– Disorder primarily of adults, 2-5% of all cases in childhood

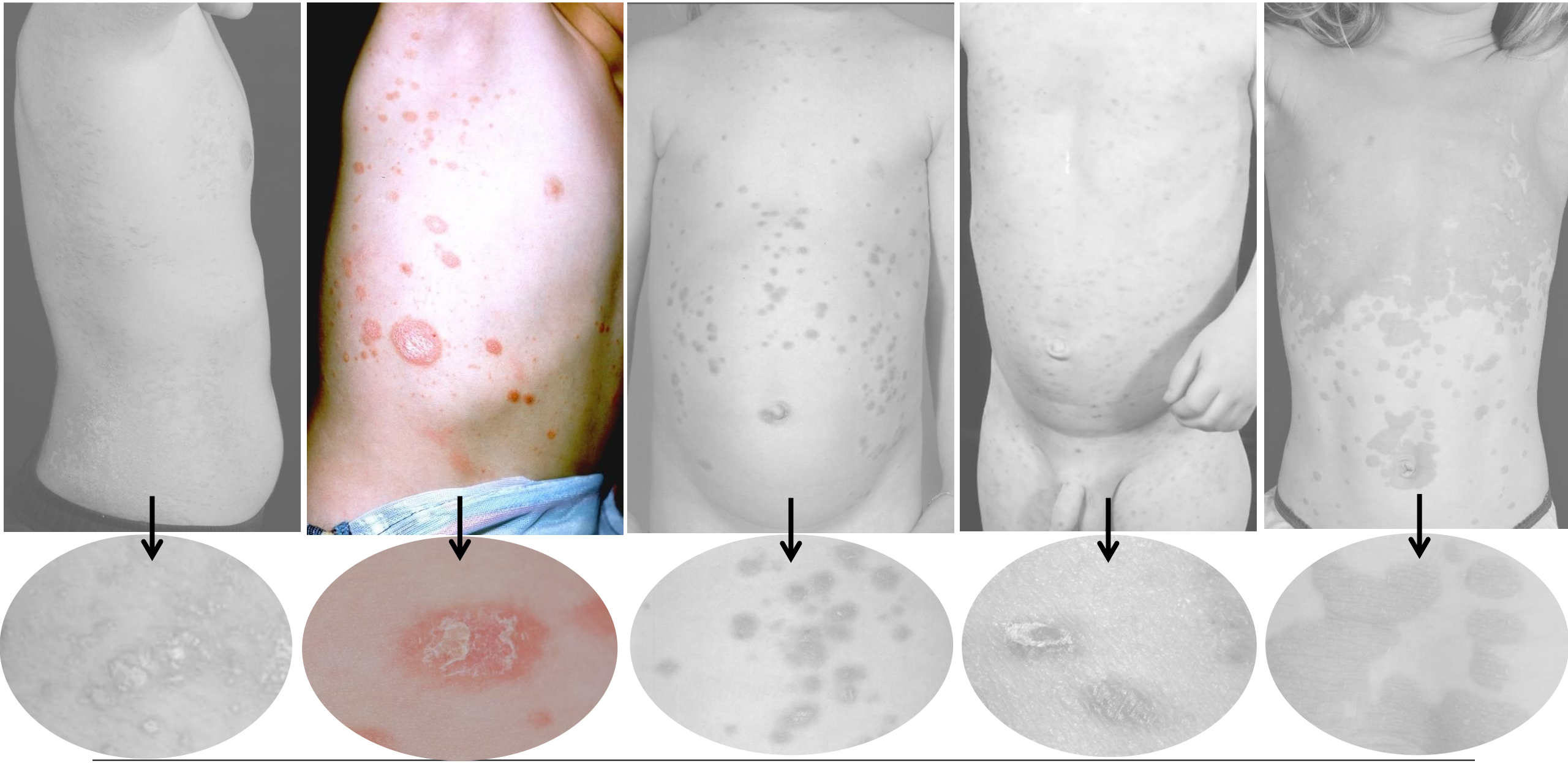
– 4 p: purple, polygonal, pruritic papules
– Reticulate white lines on the surface (→ Whickam's striae)
– Koebner phenomenon

– The etiology unknown: cytotoxic Th1-cell and plasmacytoid dendritic cell–mediated immune response
– Several cases after hepatitis B vaccination
– Familial cases rare (severity/chronicity +++)

Lichen planus in childhood

- most common clinical subtype: classic lichen planus
- rarely with nail or mucosal manifestations, no vulval LP
- no association with liver disase
- ↑ prevalence autoimmune disease and atopy in personal or family history
- more often in dark skin
- association with drugs rare

- therapy: potent topical steroids, oral steroids, oral retinoids; cyclosporin, phototherapy
- prognosis: most pediatric patients respond to treatment with full clearance over 1-6 months



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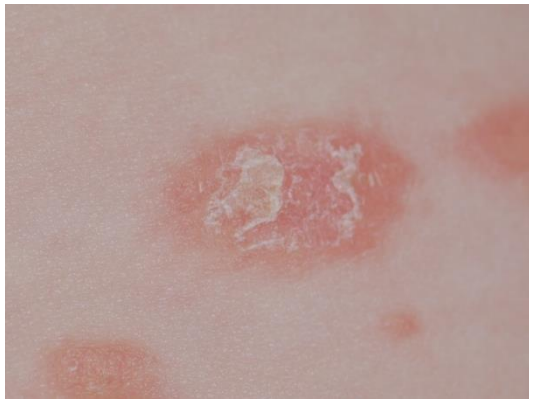
Pityriasis rosea

- Acute self-limiting parainfectious exanthema
- 50% before adulthood
- Reaction to HHV 6 or HHV 7?
- High circulating levels of TH17/22 cytokines described (as in Pso)
- Prodrome in 5%
- 25% with itching

Pityriasis rosea: clinics



- mother/herald patch in 50-70%
- after 2-14 days: generalized symmetrical eruption
- collarette scaling
- ovoid lesions parallel to the lines of cleavage
  - → christmas tree (back)
  - → upper chest V-shaped pattern
- 85% sparing the face, scalp and distal extremities
- 16% with oral lesions (asymptomatic erythematous patches)

Pityriasis rosea

Differences in children:

- More often inverse distribution (axilla and groin)
- Face and neck more often involved
- Black children: more extensive, more papular, resolve with dyspigmentation

Pityriasis rosea

Spontaneous resolution in 6-12 weeks, rarely 5 months

Differential diagnosis: in sexually active teenagers: look for Lues II (palmoplantar?)! Pso and PLC without Colarette

Treatment:

- reassurance
- topical antipruritics, antihistamins, mild topical corticosteroids
- Exposure to UV/sunlight
- Erythromycin, acyclovir, oral steroids: controversial

Terra firma-forme dermatosis

- "dry land«
- 2.19% prevalence in a group of 869 outpatient children and
- brown-grayblack patches, with a dirt-like appearance
- Surface verrucous or papillomatous
- pattern could be reticulated with interspersed islands of normal skin
- most frequently involved anatomic sites: neck, face, trunk, ankle, and navel
- No symptoms
- pathognomonic feature: persistence despite the use of ordinary soaps
- complete clearance after wiping with isopropyl alcohol 70%

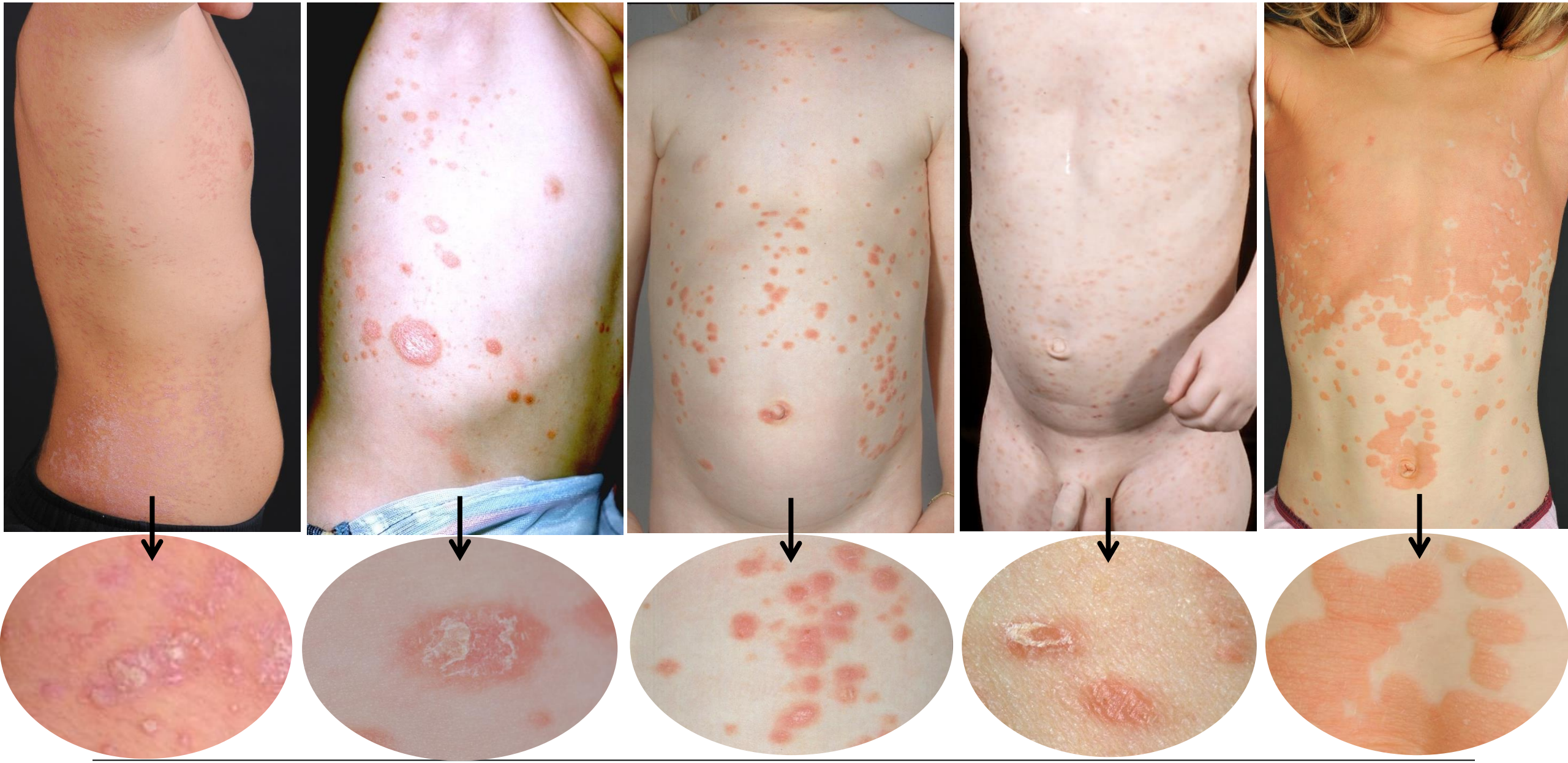
Sechi A. et al. International Journal of Dermatology 2021
Akcay G. et al. Van Med J 2019

Terra firma-forme dermatosis

- More often in summer
- correlation between TFFD and intense sun

- link between constitutional defects of the epidermal barrier and TFFD
- Significant association between TFFD and AD
- at least one allergic disorder, increase in IgE serum levels
- causative role of *Malassezia furfur* in the disease pathogenesis ?

- DD: Acanthosis nigricans, Epidermal nevi, Ashy dermatosis, frictional melanosis



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Vielen Dank für die Aufmerksamkeit

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