Series Editor: Camila K. Janniger, MD

Update on Pediatric Psoriasis, Part 1: Clinical Features and Demographics

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Pediatric psoriasis consists of infantile psoriasis, a self-limited disease of infancy; psoriasis with onset in childhood; and psoriasis with psoriatic arthritis. Approximately one-quarter to one-third of cases of psoriasis begin before 18 years of age. A variety of lesion types are seen in childhood, including plaque-type, guttate, nail-based, napkin, and erythrodermic disease. This article reviews current concepts in pediatric psoriasis. Part 2 will review therapeutics for psoriasis.

Cutis. 2010;86:118-124.

Psoriasis vulgaris is a common dermatologic disorder. The point prevalence was 1.5% in 2005 in a United Kingdom population[1] and 3.15% in a US population survey.[2] Between 22%[3] and 32% of cases of psoriasis begin in childhood[4]; 12.5% of cases occur before the age of 14 years,[5] and incidence is higher for both sexes in the second decade of life than in the first decade.[3] Psoriasis is a chronic T-cell mediated inflammatory disorder of the skin characterized by hyperproliferation of keratinocytes and consequent red scaly skin plaques.[5] Quality of life can be impaired in all age groups when psoriasis is active.[6] Pediatric-onset psoriasis often is linked to precipitants such as pharyngitis, stress, and trauma. Despite the differences in pediatric patients with psoriasis, the therapies used are essentially the same as those used in adulthood, with dosage and strength reductions calculated based on age, weight, and available formulations. This article provides a rational approach to the diagnosis and management of pediatric psoriasis, with a careful focus on those aspects of the disease that are unique to the pediatric patient.

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The author reports no conflict of interest.

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Demographics

Pediatric psoriasis can occur in infancy, which has a better prognosis for long-term remission, or in childhood.[7] Most children manifest with plaque-type psoriasis vulgaris (68.6%) with lesions localized to the scalp, postauricular region, face, diaper area, elbows, and knees. Guttate disease (psoriasis guttata), presenting with small round or oval plaques over the trunk, accounted for 28.9% of 277 children in a Chinese survey. Uncommon patterns observed in childhood include erythroderma, pustular disease including palmoplantar pustular psoriasis, and isolated mucosal disease/glossitis.[5] Diaper involvement is common in infancy. Unlike school-aged children and teenaged patients with psoriasis, infantile or neonatal psoriasis often clears within a few years. Inverse psoriasis with involvement of the folds of the skin (axillae, inner thighs) is another less common presentation. Nail psoriasis can be noted in the setting of plaque-type psoriasis vulgaris, psoriatic arthritis, or with isolated nail disease. Involvement of joints with psoriatic arthritis is less prevalent in younger patients; however, it does occur in childhood disease and should be considered in the differential diagnosis of pediatric arthritis.[8,9]

The annual incidence of psoriasis has been noted to double from 1970 to 2000, but there are no data looking at incidence trends in childhood.[10] The average age of onset appears to be as young as 11 years in one study[11] and as high as 27 years in another.[3] Onset may occur earlier for females and in the presence of a family history of psoriasis.[3]

An equal male to female ratio was noted in the largest survey of 1262 children from 1981 to 1995 in a children's hospital in Australia.[12] Women with psoriatic arthritis experience greater work disability, but there is

otherwise no difference for men and women in the morbidity of psoriasis. School- or activity-based disabilities are not uncommon in children with psoriatic arthritis; however, differences in disability by sex have not been documented in children.[13] One ethnic variation that has been noted in psoriasis vulgaris that has been described is a reduction noted among Inuit people, suggesting that diets high in omega-3 fatty acids may have a preventative effect on psoriatic onset.[14] In one population of Taiwanese patients, no cases of psoriasis were noted in children aged 6 to 11 years, demonstrating that pockets of populations with low psoriasis risk do exist in other locations.[15]

Pathogenesis

Psoriasis is a T-cell mediated immunologic disorder of the skin. The exact pathogenesis of psoriasis has not been completely elucidated; however, it is known to have a genetic basis, as 23% to 71% of children will have a family history of psoriasis,[12] and psoriasis is more common in identical versus fraternal twins (65%–72% vs 15%–30%).[16,17]

Infectious diseases that attract T cells to the skin have been linked with triggering psoriasis. It is understood that upper respiratory tract infections and inflammatory foci (38.6%)[18] including perianal and throat *Streptococcus* are common causes of disease onset in childhood, with 14.8%[9] of children having upper respiratory tract infections[18] and 21.3% being pharyngeal culture positive for group A β-hemolytic streptococci (*Streptococcus pyogenes*).[9] The guttate psoriasis subset may be linked to an inflammatory focus in about two-thirds of cases and is not caused by a specific subtype of group A β-hemolytic streptococci but rather by a host-specific response.[17-20] Cross-reactivity of keratinocyte antigens with streptococcal antigens is thought to initiate psoriatic disease in the setting of streptococcal infection.[20,21]

Other infections noted in psoriatic disease include the presence of staphylococcal superantigens[22] and human papillomavirus DNA.[23] Although it has not been noted in a larger series of children with psoriasis, I have noted 3 children (aged 4–8 years) who had molluscum contagiosum viral infection prior to the onset of psoriasis. Two of the 3 children also had a family history of psoriasis.

No single gene has been found to be responsible for psoriasis vulgaris. A series of genes have been isolated where mutations have been associated with psoriatic disease including *HLA-Cw6*; *IL12B9* (1p31.3); *IL13* (5q31.1); *IL23R* (1p31.3); signal transducer and activator of transcription 2 gene, *STAT2*, and *IL23A* (12q13.2); tumor necrosis factor α–induced protein 3 gene, *TNFAIP3* (6q23.3); and TNFAIP3 interacting protein 1 gene, *TNIP1* (5q33.1). These genes play a role in helper T cell (T_H2 and T_H17) activity as well as nuclear factor κB signaling, demonstrating both a role for T_H2 and T_H17 lymphocytes in the pathogenesis of psoriatic disease. T_H17 cells as well as collections of T_H2 and T_H1 cells have been noted in psoriatic lesions.[16,24]

Other susceptibility gene loci include *PSORS1* and *HLA-C*; the latter may interact with the *PSORS6* gene (19p13).[25] Additionally, *HLA-Cw6* may interact with caspase recruitment domain family, member 15, *CARD15*; cylindromatosis gene, *CYLD*; and transglutaminase 5, *TGM5*, susceptibility alleles.[26] Additional susceptibility has been linked to *SCL12A8*, a gene that belongs to the solute carrier gene family; filaggrin gene, *FLG*, and *TGM5*; epidermal differentiation genes *CARD15* and *CYLD*, which modulate the transcription factor nuclear factor κB; and *IL1RN*, which encodes an IL-receptor antagonist. Susceptibility loci for psoriasis may be shared with Crohn disease (IL12B and IL23R) and ulcerative colitis (IL23R). As a result, patients with Crohn disease are 5 times more likely to develop psoriasis than other members of the population.[16]

Relationship to Autoimmunity

Psoriasis is one of more than 80 autoimmune conditions that have been described. Autoimmunity encompasses a broad spectrum of medical diseases in which the body forms an immune reaction against a normal organ, the most common being thyroid disease.[27] Concurrent autoimmune illnesses have been reported in childhood including morphea[28] and vitiligo vulgaris.[29] Family history of psoriasis and other forms of autoimmunity can be noted in pediatric patients with psoriasis.[30]

The concurrent incidence of autoimmune thyroiditis has been controversial in the literature. One Italian study demonstrated excess thyroiditis in patients with psoriatic arthritis, especially males and patients with concurrent rheumatoid arthritis.[31] Another study did not demonstrate significant increases of thyroiditis markers in adults with psoriasis vulgaris when compared to age-matched controls.[32] Based on the conflicting data, annual thyroid screening is not warranted in children; thyroid screening and screening for autoimmune thyroid disease should be performed in the setting of psoriatic arthritis and when psoriasis occurs in the presence of other autoimmune diseases, such as vitiligo, that have a stronger association with thyroid disease.[33]

Clinical Characteristics and Diagnosis

Psoriasis vulgaris occurs in a variety of clinical types, most presenting with erythematous plaques over the extensor joints and scalp with overlying white

Table 1.

Clinical Variants of Psoriasis Vulgaris in Childhood

Type of Psoriasis	Typical Clinical Appearance	Diagnostic Features and Tests	Comorbidities	Differential Diagnosis	Treatment Considerations
Plaque type	Erythematous plaques with micaceous scale in typical areas: scalp extending to forehead, postauricular region, elbows, knees, umbilical area, and buttocks	Clinical diagnosis generally can be made; nail pitting can be noted as a clue to the diagnosis of psoriasis in children; biopsy when needed will demonstrate thickened epidermis with neutrophils in the horny layer, spongiform pustules of Kogoj, and subcorneal microabscess of Munro (collections of neutrophils in the epidermis)	Recent pharyngitis, specifically with *Streptococcus*, may precipitate disease; pharyngeal bacterial culture or ASO testing can be helpful; secondary infection with *Staphylococcus aureus* can be noted; other forms of autoimmunity can be seen; in the setting of obesity, disease severity can be a marker of cardiovascular risk	Nummular dermatitis, tinea capitis, id reaction, pityriasis rubra pilaris, lichen planopilaris, atopic dermatitis (overlap rarely can be seen)	Treatments must be tailored to the age of the patient, quality-of-life issues, and body surface area affected; systemic agents and phototherapy may be needed in moderate to severe disease
Psoriasis guttata	Annular, localized, erythematous to salmon-colored plaques with hyperkeratosis, sometimes micaceous; commonly noted on the trunk, abdomen, and back	Clinical diagnosis is possible; biopsy similar to plaque-type psoriasis	Recent pharyngitis, specifically with *Streptococcus,* may precipitate disease; pharyngeal bacterial culture or ASO testing can be helpful	Nummular dermatitis, pityriasis rosea, id reaction, tinea corporis, pityriasis rubra pilaris	Oral antibiotics often are initially used because of presumptive infectious precipitating factors and anti-inflammatory capabilities (ie, erythromycin, cephalosporin); systemic agents and phototherapy may be needed in moderate to severe disease

Type of Psoriasis	Typical Clinical Appearance	Diagnostic Features and Tests	Comorbidities	Differential Diagnosis	Treatment Considerations
Inverse psoriasis	Localization of erythematous, sometimes macerated, thick plaques to the folds of the skin, including the axillae and groin; can be associated with plaque-type psoriasis in other sites	Although clinical diagnosis often is possible, similarity to other diseases may require biopsy for differentiation	Secondary infection with *Candida* and/or *Streptococcus* may require cutaneous culture and usage of topical anti-infectives	Intertrigo, erythrasma, tinea corporis	Topical medications should be nonsteroidal or low-potency topical corticosteroids to avoid atrophy of the occluded skin; oral or topical anti-infectives should be added where appropriate
Nail psoriasis	Classic: pitting, oil spots, and subungual hyperkeratosis; trachyonychia: extensive pitting and subungual hyperkeratosis	Ruling out tinea infection or secondary candidal infection of the nail bed is needed when subungual hyperkeratosis is present; fungal culture and nail plate biopsy	Secondary infections should be ruled out including *Pseudomonas*, onychomycosis, or candidal infections	Onychomycosis, lichen planus, pityriasis rubra pilaris	After treatment of any fungal superinfection, topical corticosteroids with tazarotene or calcipotriene can be applied to the paronychial skin; intralesional triamcinolone also can be used in the same region to reduce subungual inflammation
Napkin or diaper psoriasis	Macerated shiny erythema of the groin region, including the folds and the genital skin	Biopsy may need to be performed	Bacterial and fungal cultures may be needed for suspected secondary infections or perianal bacterial dermatitis	Diaper dermatitis, candidal diaper dermatitis, allergic contact dermatitis	Mild topical corticosteroids with or without topical anti-*Candida* agents can be helpful; barrier therapy with zinc oxide pastes reduces secondary irritant reactions

Table 1. (continued)

Type of Psoriasis	Typical Clinical Appearance	Diagnostic Features and Tests	Comorbidities	Differential Diagnosis	Treatment Considerations
Erythroderma	Generalized erythema and thickening of the skin, sometimes with hyperkeratosis	Two biopsies generally are required from separate sites that differentiate psoriatic erythroderma from other causes of erythroderma in childhood	Fever, chills, and malaise can accompany erythroderma, making bacteremia a possible comorbidity that should be ruled out conclusively	Causes of erythroderma in childhood include atopic dermatitis, pityriasis rubra pilaris, mycosis fungoides, staphylococcal scalded skin syndrome	Topical anti-inflammatory agents can be used; however, systemic anti-inflammatory agents (never steroids orally) often are required to control severe and extensive disease
Pustular psoriasis	Erythroderma accompanied by sterile pustule formation, sometimes localized to the distal extremities, sometimes generalized; history of oral steroid usage may be noted	Biopsy often is needed and bacterial culture of the pustules; ASO titers can be drawn to rule out streptococcal precipitant	Secondary bacterial infection or tinea should be ruled out	Blistering distal dactylitis, tinea infection with *Trichophyton mentagrophytes*, herpes whitlow	Topical corticosteroids often are ineffective and systemic agents or topical psoralen plus UVA may be needed
Mucosal/glossitis	Annular plaques on the tongue may be noted in patients with psoriasis	Generally a clinical diagnosis	Rarely has any morbidity	Geographic tongue, aphthosis, lichen planus, white sponge nevus	No therapy usually is needed; however, topical medicaments in an oral base can be used when needed

Abbreviation: ASO, antistreptolysin.

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Table 2.

Calculating the Psoriasis Area and Severity Index Score

The individual is divided into 4 areas: head, arms (upper limbs), body, legs (lower limbs)
For each section, the amount of skin involved is measured as a percentage and assigned a score (0=0%; 1=<10%; 2=10%=−29%; 3=30%−49%; 4=50%–69%; 5=70%−89%; 6=90%−100%)
Severity is measured for 3 parameters: erythema (redness), desquamation (scaling), and induration (thickness)
For each section, severity is measured on a 5-point scale (0=none; 1=some; 2=moderate; 3=severe; 4=maximum)
The sum of all 3 severity parameters is then calculated for each section of skin, multiplied by the area score for that section, and multiplied by weighting of respective section (0.1 for head; 0.2 for arms; 0.3 for body; 0.4 for legs)

micaceous scale (Table 1). Presence of the following clinical features is supportive of the diagnosis of pediatric psoriasis: (1) the isomorphic response or Köbner phenomenon, which is occurrence of lesions in areas of trauma; (2) altered pigmentation with lesional clearance; (3) the Auspitz sign/pinpoint bleeding at the base of scale that has been removed; and (4) nail pitting.[35]

Severity grading for psoriasis usually is based on body surface area (BSA), impairment of quality of life, and comorbid psoriatic arthritis. The psoriasis area and severity index can be used to assess severity (Table 2) and is the current scoring system used in clinical trials. A Web site to calculate the psoriasis area and severity index can be accessed.[36] Others will classify disease as mild if less than 3% BSA is affected, moderate with 3% to 10% BSA, and severe with greater than 10% BSA. This oversimplification may minimize some of the disability or stigma of psoriasis when present on cosmetically obvious or functionally significant areas such as the palms.[37,38] Facial disease (seen in 38% of children) is particularly more common in children and involves a limited BSA but may cause extensive emotional distress.[12]

Differential Diagnosis

The differential diagnosis of psoriasis includes other papulosquamous disorders of childhood including lichen planopilaris, psoriasiform id reactions, nummular dermatitis, pityriasis rosea, mycosis fungoides, and pityriasis rubra pilaris. Biopsy can be helpful in differentiating psoriasis from these other illnesses.

Conclusion

Psoriasis vulgaris of childhood is a complex immunologic disease. Part 2 will review newer therapeutic options.

This article is the first of a 2-part series. The second part will appear in a future issue of Cutis®.

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