Pediatric Papulosquamous and Eczematous Dermatoses

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Disclosure Statement

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Atopic Dermatitis

• Epidemiology:
  • Most common in infants and children
  • Affects 15-30% of children, ~1% of adults
  • 85% begin during the first year, 95% begin before 5 years
  • Up to 70% spontaneous remission before adolescence
  • Prevalence of 15-20% in industrialized nations during early childhood

• Pathogenesis:
  • Atopic dermatitis is a chronic, relapsing, intensely pruritic condition that is often associated with allergic rhinitis and/or asthma, often referred to as ‘the atopic march.’
  • Involves a skin barrier breakdown in addition to dysfunctional innate and adaptive immune response, including:
    • T-helper 2 cells and hyperimmunoglobulinemia E.
    • T-helper 2 cells stimulate the production of immunoglobulin E and eosinophilia by releasing interleukin-4, -5, and -13 as well as in decreasing protection against bacterial superinfection by releasing interleukin-10.

• Complications:
  • Bacterial infections: most commonly Staphylococcus aureus
  • Viral infections: HSV, molluscum contagiosum, HPV, eczema herpeticum, eczema coxsackium, eczema vaccinatum
  • HRQL impairment in generalized AD – exceeds that in asthma, epilepsy, and diabetes – comparable to that in renal disease or cystic fibrosis – equals (child) or exceeds (parent) that in psoriasis

❖ Beattie P, Lewis-Jones M; BJD 2006
• More than 50% develop Asthma
• About 75% develop Allergic Rhinitis
Infantile Type
Face, Scalp, Trunk, Extensor surfaces of extremities

Childhood Type
Flexural Folds of Extremities (Antecubital, Popliteal Fossa, neck and ankles)

Adult Types
Upper arms, back, wrists, hands, fingers, feet, toes

Source: Color Textbook of Pediatric Dermatology, 4th Edition
• **Treatment:**
  - **Lifestyle modification and trigger avoidance**
    - Environmental triggers, such as heat, humidity, inappropriate bathing habits, detergents/soaps, abrasive clothing, chemicals, smoke, and even stress, tend to aggravate the condition.
    - Latex allergy and nickel allergy occur more often in persons with atopic dermatitis.
  - **AAD guidelines endorse use in pediatric population:**
    - Topical corticosteroids, topical calcineurin inhibitors, tar, emollients
    - Systemics: Anti-histamines, cyclosporine, mycophenolate mofetil, azathioprine, methotrexate, short term corticosteroids
    - UV-therapy: safe and highly effective
  - **Recent Treatment Advances**
    - **Dupilumab**, a human monoclonal antibody that inhibits signaling of IL-4 and IL-13, showed efficacy in two phase 3 trials of adult patients with moderate to severe atopic dermatitis inadequately controlled by topical treatment
      - Dupilumab was reported to be effective in reducing eczema area and severity as well as pruritus, depression and anxiety, and improving quality of life.
    - **Crisaborole**, a phosphodiesterase 4 (PDE-4) inhibitor is applied topically twice daily as a 2% ointment, and received FDA in Dec 2016 for mild to moderate atopic dermatitis in patients over 2 years old.
      - Reported to be both safe and effective, with the majority of patients clear or almost clear after one month of use

  ❖ [Saporito RC, Cohen DJ](#) Apremilast Use for Moderate-to-Severe Atopic Dermatitis in Pediatric Patients.
Psoriasis

• a chronic, intermittently relapsing inflammatory disease characterized by sharply demarcated erythematous, silvery, scaly plaques most often seen on the scalp, elbows, and knees. Additional sites of involvement include the nails, hands, feet, and trunk.

• Epidemiology:
  • Psoriasis affects about **2% of the world's population** and can develop at any age and in both sexes.
    • Psoriasis incidence has a bimodal pattern, with one peak in childhood and a second peak in adulthood.
    • It occurs most frequently in individuals of Northern European descent.
  • Etiology: aberrant T-cell function and keratinocyte responses are believed to be major culprits in the pathogenesis of psoriasis.
    • Psoriasis is a polygenic disease: HLA-Cw6 influencing early onset, also associated with: HLA-B13, B17, and B27

• Skin compostion changes: increased involucrin, K6 & 16; decreased loricrin

• Removal of scale → pinpoint bleeding = **Auspitz sign**

• Trauma, including acupuncture, may induce local plaques of psoriasis (the Koebner phenomenon). Patients may interpret these lesions as "slow-healing wounds."
Psoriasis

- **Infantile psoriasis:**
  - <1% of infants by 1 year of age and 2% of infants by age 2. Infantile psoriasis resembles adult psoriasis. There is usually less white scale on the plaques of infants
  - Psoriasis in infancy typically involves the diaper area and face.
  - Nail findings of pitting, onycholysis, oil spots, and subungual hyperkeratosis are present in 10% of affected infants.
  - Not typically pruritic

- **Guttate psoriasis**—An acute generalized eruption of small, discrete, raindrop-like papules with fine scale. This may occur 2-3 weeks after an URI – typically *streptococcal*
  - Is most common in children.
  - The prognosis in children is typically *better than in adults*, with many children spontaneously clearing within several weeks to months.
  - Nevertheless, most patients will go on to develop chronic psoriatic disease several years later.
What you eat can drastically affect psoriasis and oftentimes has as much effectiveness as the toxic alternatives.
Recent Treatment Advances

• **Biologics:**
  - **Secukinumab, an IL-17A inhibitor,** effective in moderate to severe plaque psoriasis
    - 300 mg by subcutaneous injection for weeks 0, 1, 2, 3 and 4, followed by 300 mg every 4 weeks.
  - **Ixekizumab, an IL-17A inhibitor,** effective in moderate to severe plaque psoriasis – 160 mg by subcutaneous injection is given on week 0 followed by 80 mg on weeks 2, 4, 6, 8, 10, 12. After week 12, 80 mg is given every 4 weeks.
    - Inflammatory bowel disease may flare or initiate during therapy.

• **New mechanisms**
  - **Apremilast (Otezla),** an oral PDE-4 inhibitor, is FDA-approved to treat adults with active psoriatic arthritis and moderate to severe plaque psoriasis.
    - The FDA is requiring a pregnancy register.
    - Titrated up to 30mg BID to minimize AEs.

  • **Oral tofacitinib, a JAK3 inhibitor, at 5-10mg BID,** has shown efficacy in phase 3 studies and an ongoing open-label long-term extension study in patients with moderate to severe plaque psoriasis.
    - **Baricitinib, a reversible JAK1 / JAK2 inhibitor,** has also been studied as a treatment for moderate to severe psoriasis.

  • **Ponesimod,** an oral, selective, reversible modulator of sphingosine 1-phosphate receptor 1, in patients with moderate to severe chronic plaque psoriasis is under study.
https://www.aad.org/public/diseases/rashes/pityriasis-rosea
Pityriasis Rosea

- Etiology is controversial, but likely due to a viral pathogen with a flu-like prodrome.
  - +/- HHV6 and HHV7
- Commonly begins with a solitary pink-flesh colored scaly plaque “herald patch,” followed by oval patches and plaques with a fine ‘collarette’ scale along the Langer’s lines of cleavage.
  - Typically the face, scalp and distal extremities are spared.
  - Inverse and papular patterns are more common in younger and darker-skinned individuals.
- Self limited and usually clears within 6 weeks.
  - Supportive treatment includes: topical corticosteroids and anti-histamines. UV light is thought to hasten resolution and reduce pruritus
Lichen Planus

• Etiology
  • Unclear but viruses, medications and contact allergens have been proposed
  • Autoreactive T cells attack basal keratinocytes.
  • **Hepatitis C** has **not** been associated in the U.S. with pediatric cases, but cases have been described after **hepatitis B vaccination**.
  • Drugs causing LP-like reactions include: ACEi, beta-blockers, thiazides, anti-malarials, griseofulvin, NSAIDs, tetracycline, carbamazepine, phenytoin, and penicillamine.

• Clinical
  • **Purple, planar, polygonal, and pruritic papules** commonly on the flexures, ankles, wrists and genitalia. White papules forming an annular or lace-like patterns may be found on the buccal mucosa but are less common in pediatric cases
  • Patients may spontaneously resolve in several months, or – more likely - follow a chronic course.

• Treatment
  • Limited disease can be treated with topical corticosteroids and oral antihistamines. For extensive or recalcitrant cases, the addition of a 2- to 6-week course of systemic corticosteroids (1 mg/kg per day) can be helpful, also UV-therapy, metronidazole, retinoids, and systemic steroid-sparing agents may be used.
Paller AS, Mancini AJ. Hurwitz clinical pediatric dermatology: a textbook of skin disorders of childhood and adolescence. Chapter 4, Papulosquamous and Related Disorders.
Pityriasis Rubra Pilaris

• Etiology
  • unknown, but likely a disorder of abnormal keratinization

• Clinical:
  • PRP starts as small follicular papules which coalesce into yellowish-pink, scaly plaques with **islands of sparing**.
  • Yellow waxy hyperkeratosis of the palms and soles is common.
  • **Hyperkeratosis of the elbows, knees, ankles, and Achilles tendon** commonly affects pediatric cases.
  • **Types III-V** occur in children with **type IV** being the most common.
  • Clinical course is variable.

• Treatment:
  • Topical Emollients, Corticosteroid, retinols and keratolytics for limited disease. If extensive consider systemic retinoid therapy, UVB or vitamin A (>100 000 U/day).
Cutaneous T-cell Lymphoma

• Epidemiology
  • Lymphoma cutis is rare in children but the most common form is cutaneous t-cell lymphoma (CTCL)
  • 0.5-5% of cases occur in childhood
  • better outcomes in children secondary to limited disease at the time of presentation

• Clinical
  • Most commonly presents as erythematous scaly patches, papules, and plaques on the trunk and buttocks.
  • Lesions can morph into a variety of shapes and become scaly
  • Hypopigmented CTCL is the most common subtype in children. This presentation commonly mimics: tinea versicolor, pityriasis alba, post-inflammatory hypopigmentation.
  • Pathology shows a predominance of CD8+ Tcells in contrast to classic CTCL

• Differential:
  • psoriasis, atopic dermatitis, tinea corporis, contact dermatitis, and seborrheic dermatitis
  • If seemingly benign conditions are refractory to standard treatment, a biopsy (or a series of biopsies) is warranted

Paller AS, Mancini AJ. Hurwitz clinical pediatric dermatology: a textbook of skin disorders of childhood and adolescence. Elsevier Health Sciences; 2015 Sep 25. Figures 2.19, 3.4 and 3.35
Seborrheic dermatitis

• Clinical
  • Characterized by well demarcated erythematous plaques with overlying yellow, greasy scale and crust
  • Typical locations: scalp, face, post-auricular, pre-sternal, and intertrigenous areas.

• Epidemiology
  • Most common in infants (3rd-4th month) and adolescents. If infant seborrheic dermatitis persists past 6-12 months, concomitant atopy is more likely.

• Etiology:
  • largely unknown, but linked to sebaceous gland activity
  • increased incidence during times of heightened hormonal control
  • Adult disease is more closely linked to Pityrosporum ovale (Malassezia ovalis)

• Leiner’s Disease ‘phenotype of immunodeficiency’:
  • Severe seborrheic dermatitis, failure to thrive, and diarrhea. This phenotype is liked to compliment deficiency, severe combined immunodeficiency (Omenn syndrome), hypergammaglobulin E syndrome, and X-linked agammaglobulinemia
• Differential:
  • atopic dermatitis, psoriasis, diaper dermatitis, Langerhans cell histiocytosis, scabies, tinea corporis/capitis, contact dermatitis, LE, and CTCL

• Treatment:
  • **Infantile seborrhea** will heal in 3-4 weeks without treatment. If thick scale is present, the use of “no tears” shampoo and baby oil along with a gentle brush for exfoliation will suffice.
  • For non-scalp areas, low potency steroid cream and topical antifungals can be used.
  • Traditional anti-seborrheic shampoos are favored for **adolescent cases**. Monitor for secondary candida or bacterial infection and treat appropriately.
Lymphomatoid Papulosis

- Benign, recurrent disorder with histologic features similar to lymphoma

- Clinical:
  - presents with widespread papules, papulovesicles, or even nodules with necrotic centers. Often asymptomatic.
  - Individual lesions typically resolve within a few months, however the disorder can last for years
  - **Up to 9% of pediatric patients go on to develop Non-Hodgkin’s Lymphoma**, which makes long term monitoring important

- Histopathologic:
  - a **heavy CD30+** T cell infiltrate is seen that can resemble Hodgkin’s disease (Type A), mycosis fungoides (B), or anaplastic large T-cell lymphoma

- Treatment
  - Often unsatisfactory, but includes corticosteroids (topical and systemic pulse therapy) and PUVA/NBUVB
Pityriasis Lichenoides

• Idiopathic, papulosquamous eruption marked by two overlapping forms, which exist on a spectrum
  • Pityriasis Lichenoides et Varioliformis Acuta (PLEVA)
    • Acute form with widespread papulovesicles that ulcerate and can occasionally occur with constitutional symptoms
  • Pityriasis Lichenoides Chronica (PLC)
    • Scaly papules and plaques that can arise from PLEVA or de novo that can last 6 months to 2 years; lesions heal with dyschromia

• Etiology:
  • Idiopathic. Both believed to be a reactive lymphoproliferative disorder with a possible viral trigger

• Treatment
  • Systemic antibiotics (azithromycin), NBUVB (most effective), and topical corticosteroids mainly for control of pruritus
Reactive Arthritis

• Etiology:
  • Most common after enteric infections in children (Salmonella, Yersinia, Shigella)

• Clinical:
  • Reactive disorder most commonly presenting due to abnormal immune response following infection of certain antigenic pathogens
  • Complete triad of conjunctivitis, urethritis, and arthritis rarely seen
  • Skin manifestations include circinate balantis, keratoderma blenorrhagica, and geographic tongue
  • Arthritis most often asymmetric and affects weight bearing joints of hip, knee, ankle
  • sacroilliac involvement rare in children despite most being HLA-B27 positive

• Prognosis in children is better than adults and most resolve with administration of NSAIDS and bed rest.

• Treatment:
  • Systemic immunosuppressants including methotrexate and cyclosporine for resistant cases.
  • Ophthalmology and rheumatology referrals recommended
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