Chapter 4: Dermatology

Atopic dermatitis/eczema

Definition

Atopic dermatitis, or eczema, is a chronic inflammatory skin disease that causes dry skin, severe itching, and heightened sensitivity to various environmental stimuli

Risk factors/causes

- Genetic risk factors – A family history of atopy (eczema, asthma, or allergic rhinitis) is the strongest risk factor for atopic dermatitis
- Environmental Exposures – climate, urban dwellers, early exposure to non-pathogen microorganisms

Promotion/prevention

- Health education
- Avoid anything that causes the skin to itch, because scratching triggers flare-ups
- Moisturize the skin at least twice a day
- Use a gentle non soap cleanser
- Avoid irritants such as soaps, detergents, including shampoo, bubble bath, environmental factors or allergens such as cold and dry weather and more specific things like house dust mites, pet fur, pollen and moulds

Signs and symptoms

- Infants and young children
    - Marked by red, scaly, and crusty lesions on the scalp, cheeks, and body's extensor surfaces
    - The diaper area is usually spared
    - Severe cases may show vesicles, serous exudates, and crusting.
- Older children and adolescents
    - Appears as lichenified plaques in flexural areas, such as the antecubital and popliteal fossae, wrists, ankles, and neck
    - The neck may show reticulate pigmentation, known as "atopic dirty neck"

Investigations

- No specific investigations are required
- It is diagnosed based on clinical evaluation, including history, morphology and

distribution of skin lesions and associated clinical signs

Differential diagnosis

- Seborrheic dermatitis
- Ichthyosis
- Psoriasis
- Drug eruptions

Management

Effective management includes:

- Educating the patient
- Maintaining skin hydration
- Restoring skin barrier function
- Using pharmacological treatments to address skin inflammation
- Identifying and eliminating exacerbating factors

Primary level

Refer to management guidelines at Secondary Level

Secondary level

Mild-To-Moderate Atopic Dermatitis

- Patient / guardian education
  - Avoid known triggers.
  - Reduce bathing frequency and avoid using irritant soaps / products. Non-fragranced aqueous cream can be used as a soap substitute if necessary.
  - Rinse clothes thoroughly after washing to remove all soap residue.
  - Nonpharmacological therapy such as emollients administered as monotherapy.
  - Emollients need to be used liberally and multiple times a day to keep the skin moisturized always.
- Add topical pharmacotherapy if nonpharmacological therapy is insufficient such as topical steroids (preferred).
  - Topical steroids should not be used for prolonged periods of time, but should only treat acute flare-ups
  - Use low potency topical corticosteroids (TCS) on affected areas once daily for 2-4 weeks.
- Assess for concomitant skin infection (staph aureus) and treat as necessary.

Refer severe cases to tertiary level facility

Tertiary level

Moderate/Severe Atopic Dermatitis (with significant functional impairment)

- Patient education as above
- Consider alternative non-pharmacological therapy (e.g., wet wrap therapy with emollients
- Topical steroids
  - Use medium – to – high potency TCS on affected areas once or twice daily for 2 – 4 weeks
    Note: Only low potency steroids should be used on the face, neck and skin folds
- Assess for and treat concomitant infections.
  - If refractory disease, consult a dermatologist and consider:
    - Increasing potency of topical agents
    - Topical calcineurin inhibitors
    - Adding systemic therapy to topical agents if the disease is refractory.
    - Use of phototherapy (UV light)
      - Non-corticosteroid systemic immunomodulatory medications

Follow up

- Dermatology clinic every 1-3 months
- Ongoing education
- Review of medication and side effects

Miliaria

Definition

Eccrine miliaria, commonly referred to as "sweat rash," "prickly heat," or "heat rash," is a temporary skin condition that occurs when the eccrine sweat duct is obstructed. There are three types of eccrine miliaria, including:

- **Miliaria crystalline** – also known as sudamina, is very common in neonates. Incidence peaks at approximately one week of age
- **Miliaria rubra** – the most common type. It has been reported in 4 percent of neonates and in up to 30 percent of people of all ages
- **Miliaria profunda** – also known as tropical anhidrosis

Risk factors/causes

Anything that causes sweating can lead to miliaria including:

- A hot and humid environment
- Intense exercise or physical activity
- Febrile Illness
- Occlusion of the skin with non- porous dressings or synthetic clothing against the skin

Promotion/Prevention

- Avoid excessive wrappings/woollen hats in infants
- Avoid tight clothing

Signs and Symptoms

- **Miliaria crystalline** – Presents as 1-2 mm superficial clear blisters that easily break leaving a bran like scale. There is no inflammation. The blisters are usually widely spread on the head, neck and upper trunk
- **Miliaria rubra** – Results in red 2-4 mm non-follicular papules and papulo-vesicles. They are very itchy. Background erythema is often present. It involves the trunk and the skin folds of the neck, axilla and the groin. Miliaria Pustulosa is a variant of miliaria rubra with pustules
- **Miliaria profunda** – Presents as asymptomatic deep papules. The flesh coloured 1-3mm diameter papules develop on the trunk and extremities

Investigations

- Clinical diagnosis.
- Tzanck smear can be taken from the vesicles to distinguish miliaria from herpes

simplex or toxic erythema of the newborn

Differential Diagnosis

- Herpes simplex
- Bacterial folliculitis
- Acute generalized exanthematous pustulosis (AGEP)

Management

Primary level See the tertiary-level guidance
Secondary level See the tertiary-level guidance
Tertiary level • Minimize heat and humidity exposure to reduce sweating and irritation of the skin • Calamine lotion • Emollients • Mild topical steroids • Reassurance - usually resolves within 2 days after changing to a cooler environment. Refer to a dermatologist if the condition persists

Follow up

- The condition is generally self-limiting thus will not require routine follow-up unless other concerns are flagged by the diagnosing physician

Urticaria

Definition

Refers to a group of conditions in which wheals (hives) or angioedema (swelling) develop in the skin. It is very common in children

Occurs as a result of mast cell and basophil activation causing release of histamine (which causes itching) and vasodilatory mediators (which cause localized swelling)

- A wheal is a superficial swelling, usually pale or skin coloured. It is often surrounded by an area of erythema and can last from a few minutes to 24 hours
- Angioedema is a deeper swelling from within the skin or mucous membranes. It usually looks red and puffy. The most common areas for angioedema in children are the lips, tongue and eyelids

Risk factors/causes

In many cases, no specific cause can be identified. Other potential causes are:

- Infections (viral and bacterial infections)
- Food allergy (such as eggs, milk, soy, peanut and wheat)
- Drug induced urticaria such as antibiotics, NSAIDs
- Bee or wasp sting

Prevention/promotion

- Choosing mild or fragrance-free soaps, skin creams, and detergents
- Antihistamines
- Keeping a record of any possible triggers, such as a food diary

Signs and symptoms

<u>Wheals</u>

- Appear on any part of the body
- Usually red, raised plaques with a central pale area and can be itchy
- Can be round, oval, or serpentine and vary in size from small to large

<u>Angioedema</u>

- It most commonly affects the face. The child may have a swollen tongue, eyelids, or lips
- Usually localized to a single area such as the hands, feet, and genitalia
- Is often tender or painful

Inducible urticaria

- Due to a physical stimulus: the wheals will be localized to the exposed site.
- Often comes on within minutes after exposure and resolves in less than an hour

Anaphylaxis (severe allergic response)

- Shortness of breath, wheezing, collapse, hypotension

Investigations

- Urticaria is usually diagnosed through history and physical examination
- If food or drug allergy is suspected skin prick or immunoglobulin E (IgE) tests can be helpful

Differential Diagnosis

- Insect bites – Presents on exposed sites, as asymmetrical clusters of itchy papules or wheals, often with a central fluid filled blister
- Contact dermatitis – Presents on areas in contact with a causative irritant or allergen with irregular red, blistered, scaly sometimes swollen plaques. Dermatitis persists for days or weeks, much longer than urticarial wheals
    - Erythema multiforme – Erythematous plaques usually located on acral sites. Target lesions (a pattern of concentric rings) are characteristic, sometimes with central blistering
    - Urticarial vasculitis (uncommon) – It resembles urticaria with the exception that the wheals last longer than 24 hours and are followed by bruise-like discoloration

Management

Primary level

Mild urticaria

- Initial treatment of new-onset urticaria should prioritize short-term relief of symptoms
- About two-thirds of cases self-resolve
- The itch may be reduced by cooling with a fan, ice pack or moisturizing lotion such as calamine lotion
- Antihistamine pharmacotherapy (cetirizine/promethazine/piriton). These are not curative, but often controls the itch and the spread of wheals until the urticaria settles on its own
- In cases that are unresponsive to antihistamines, a short course oral prednisolone 0.5 - 1mg/kg/day x 3 days

Severe/life-threatening anaphylaxis (urticaria with angioedema)

- Remove inciting allergen if still present
- Place patient in recumbent position and elevate the lower extremities
- 100% Oxygen therapy via non-rebreather mask at 15 litres/min
- Give adrenaline 0.01mg/kg IM STAT (1:1000 solution)
- Repeat dose in 5 - 15 minutes if not responding
- Normal Saline rapid bolus 10mls/kg. Repeat if necessary to maximum 40mls/kg
- Intramuscular promethazine 0.5mg/kg IM TDS (maximum 25mg/dose) for children more than 2 years old
- Refer
    - patients with severe anaphylaxis,
    - those requiring more than one dose of adrenaline,
    - those who received adrenaline only after a significant delay (>60 minutes)

Secondary level

See the tertiary-level guidance below

Tertiary level

- Manage mild/moderate as above
- Severe/life threatening anaphylaxis:
- As above but can add:
  - Salbutamol nebuliser 2.5 - 5 mg if bronchospasm is not responding to IM epinephrine - can repeat every 15 minutes if required
  - H1 antihistamine
    - IV Diphenhydramine 1 mg/kg over 5 minutes (maximum 50mg/dose)
      or
    - IV Cetirizine 2.5mg (6mo-5yrs) or 5 - 10 mg (6-11 yrs) over 2 minutes
      or
    - IM Promethazine 0.5 mg/kg (max 25mg/dose) for children > 2 years old
  - Glucocorticoids
    - Methylprednisolone 1 mg/kg (maximum 125mg)
      or
    - IV hydrocortisone 1 - 2 mg/kg
      or
    - Prednisone 1-2 mg/kg PO

- Refractory anaphylaxis
  - Intubation and admission to ICU if airway compromise.
  - Adrenaline infusion 0.1-1 mcg/kg/min, titrated to effect if refractory shock
  - Some patients may require a second vasopressor in addition to epinephrine if not responding

Follow up

Patients with a history of life-threatening anaphylaxis should be followed up bi-annually for:

- Ongoing education / counselling on avoidance of triggers
- Anaphylaxis emergency action plan including EPIPEN dosage and administration technique
- Detect and manage co-morbidities

Psoriasis

Definition

- Psoriasis is an immune-mediated disease that causes red patches with silvery scales.
- It can occur at any age and in different forms, but chronic plaque psoriasis is common in children

Risk factors/causes

- Genetic risk factors – i.e. early-onset psoriasis (onset under the age of 40 years) is the human leukocyte antigen (HLA) type Cw6 (PSOR1)
- Environmental exposures
  - Skin trauma (Koebner phenomenon), sun exposure, certain medications
- Infections
  - Streptococcal, staphylococcal, varicella zoster
- Autoimmune/inflammatory diseases
- Psychological and physical stress

Promotion/prevention

- Use emollient/moisturizing creams
- Use gentle cleansers and perfumes
- Avoid dry, cold weather
- Avoid medications that cause flare-ups
- Avoid scrapes, cuts, bumps and infections
- Stress management

Signs and symptoms

- **Diaper area psoriasis** – Presents with shiny, red patches in diaper area with minimal or absent scaling. Affect infants
- **Chronic plaque psoriasis** – Presents as red, scaly patches with well-defined edges, typically on the elbows, knees, scalp and lower back. Auspitz sign may be observed when scales are removed. Children may have thinner scales, less distinct edges and may develop psoriasis in facial, intertriginous and diaper areas
- **Guttate psoriasis** – Presents with numerous, small, "drop-like," erythematous papules and plaques, particularly when involving the trunk and proximal extremities. Plaques are erythematous
- **Generalized pustular psoriasis and erythrodermic psoriasis** – Acute generalized pustular psoriasis presents with widespread erythema, pustules and scales. Erythrodermic psoriasis presents with widespread erythema and scale.

Patients with these disorders often appear systemically ill

- **Inverse psoriasis** – Presents with well-demarcated, shiny, erythematous, thin plaques within skin folds and/or on anogenital skin. Scale is minimal or absent

Investigations

- The diagnosis of psoriasis in children usually can be made based upon the clinical features
- A skin biopsy is not necessary for the diagnosis of most patients

Differential diagnosis

- Atopic dermatitis
- Pityriasis rosea
- Pityriasis lichenoid chronica
- Pityriasis rubra pilaris

Management

Primary level

Refer to management guidelines at Tertiary Level

Secondary level

Refer to management guidelines at Tertiary Level

Tertiary level

- All **Patients** – Assess disease severity: e.g. based on estimated body surface area (BSA) affected and provide supportive care

Mild psoriasis (**below 3–5%** BSA involvement)

- Topical pharmacotherapy (e.g. corticosteroids, calcipotriene, retinoids) and/or targeted phototherapy
- Systemic agents if treatment response is insufficient

Moderate to severe psoriasis (**above 3–5%** BSA involvement)

- Systemic pharmacotherapy and/or phototherapy
- Narrowband UV-B therapy

All suspected psoriasis cases must be discussed with the dermatology department

Follow up

- Biannual follow up in dermatology clinic for long-term psoriasis

Oculocutaneous albinism

Definition

A collection of congenital disorders of melanin synthesis resulting in hypopigmentation There are two main distinctions:

- Partial albinism: A genetic condition characterised by the partial absence of melanin pigment from melanosomes in the body due to defects in the biosynthesis of melanin. Ocular albinism (OA) only affects the visual system (e.g. iris, retina), but not the skin or hair
- Total albinism: A genetic condition characterised by the complete absence of melanin pigment in melanosomes. Oculocutaneous albinism (OCA) is characterised by a lack of melanin in the skin, hair, and iris

Risk factors/causes

- Inheritance is autosomal recessive trait

Promotion/prevention

- Albinism is a genetic condition that is inherited and cannot be prevented
- It is not an illness
- While it presents unique physical characteristics, acceptance, understanding and love among family members can be fostered
- Health education to decrease co-morbidities associated with albinism
- Increase awareness and understanding to create safer lives for those living with this condition
- A heightened awareness of albinism is necessary for early diagnosis and management, providing the best opportunity for the child's vision to develop to its fullest potential

Signs and symptoms

Ocular albinism

- Eyes are translucent with hypo-pigmented blue, grey, or green irides
- Marked photophobia, with decreased visual acuity
- Other ocular manifestations such as strabismus, nystagmus and amblyopia
- Abnormalities of the optic nerve (e.g. hypoplasia, abnormal crossing of optic fibres at the optic chiasm)

Oculocutaneous albinism

- Along with ocular plus milky white skin colour which is photosensitive and
- sunburns easily

- Predisposition to skin cancer

Investigation

- Diagnosis based on physical examination including comprehensive ophthalmologic examination
- Molecular testing using a multigene panel or comprehensive genome sequencing can be done for precise diagnosis

Differential diagnosis

Disorders associated with hair and skin hypopigmentation

- Waardenburg syndrome type II
- Tietz albinism-deafness syndrome
- Chediak-Higashi syndrome

Management

Primary level

Refer to management guidelines at Secondary Level

Secondary level

- Lifelong photoprotection
  - Seeking shade and avoiding ultraviolet exposure during the peak hours of sunlight
  - Use of protective clothing, such as wide-brimmed hats, ultraviolet protective factor (UPF)-labelled clothing, shirts with a collar, long sleeves, long pants, and socks
  - Liberal and frequent (every two hours) application of sunscreen of at least sun protection factor (SPF) 30 when in the sun
  - Avoiding medications that increase photosensitivity whenever possible. NSAIDS, antihistamines, oral contraceptives, Sulphonylureas, Thiazide diuretics and Tetracycline
- **Skin cancer surveillance**
  - Skin examination at 6- to 12-month intervals starting in adolescence
  - Educated about the importance of skin self-examination including awareness of concerning skin lesions, such as new lesions in sun-exposed areas, non- healing lesions or lesions undergoing changes, and lesions associated with symptoms like pain, itching, or bleeding
- **Management of eye abnormalities**
  - Optometry and/or ophthalmology review for correction of refractive errors and provision of low vision aids

Refer to tertiary level for review by ophthalmologist/dermatologist
Tertiary level • Manage as above • Review by dermatologist and ophthalmologist

Follow up

- Dermatology review every 6-12 months for early cancer detection
- Ophthalmology follow-up in the first 2 years of life, then annually during school age
- On-going education, psychosocial counselling and social support

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