140 Pediatric Dermatology: Scope and Challenges

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Skin is the largest organ of the body and a very complex and dynamic structure consisting of many parts and appendages. The actual condition of the skin may reflect not only skin-specific or skin-limited disorders, but – in most cases – is a mirror of either the somatic or psychological status of the patient.

The outermost layer of the skin surface – the epidermis – is an effective barrier to the penetration of irritants, toxins, and organisms, as well as a membrane that holds in body fluids. It acts also as an immunologically active organ responding to external and internal stimuli, communicating with other parts of the skin and, through signaling molecules, transmitters, and neural connections, with different body structures including the central nervous system.

Beside the main cellular component – keratinocytes – epidermis contains specific and sophisticated cells such as melanocytes, which are important for protection against the harmful effects of ultraviolet (UV) light, and the Langerhans cells, a part of the body's first-line immunologic defense. In its basal layer epidermis also contains multipotent stem cells, which in physiologic condition reassure the normal turnover of this skin layer.

The dermis, consisting largely of fibroblasts and collagen, is a tough, leathery, mechanical barrier against cuts, bites, and bruises. Its collagenous matrix also provides structural support for a number of cutaneous appendages.

Hair, which grows from follicles deep within the dermis, is important for cosmesis as well as for protection from sunlight and particulate matter. The bulb part of a hair follicle is also the place from which the epidermis and hair regenerate, being the part of the skin richest in stem cells.

Sebaceous glands arise as an outgrowth of the hair follicles. Oil produced by these glands helps to lubricate the skin and contributes to the protective function of the ceramide-rich epidermal barrier.

The nails are specialized organs of manipulation that also protect sensitive digits.

Eccrine sweat glands accomplish skin thermoregulation.

Glomus cells regulate changes in the cutaneous blood flow.

The skin also contains specialized receptors for heat, pain, touch, and pressure, and the input from these structures helps to protect the skin surface against environmental influence or trauma.

Below the dermis, in the subcutaneous tissue, fat is stored as a source of energy, a reservoir of biologically active substances, vitamins, and hormones, and also acts as a soft protective cushion.

The newborn and neonatal skin is of special interest not only because the transition from an aqueous and sterile atmosphere to a dry and pathogen-rich environment is a tremendous challenge to the young skin. The functioning epidermal barrier is of great importance for the prevention of water loss and for the interaction with numerous microorganisms that colonize the neonatal skin from the moment of birth.

The anatomy of the epidermal and dermal components of the newborn and neonatal skin is the same as that of older skin. However, the process of postnatal maturation and adaptation to environmental stimuli is crucial for the further physiological condition of the skin. It is also important to know that the young skin is much more permeable than the skin of an older person, which implicates the need for appropriate dose adaptation of topically used agents.

The importance of appropriate neonatal skin care and treatment to maintain the protective function of the skin will be discussed later in this section.

Disorders of the skin in infants and young children vary from the occurrence and presentation of the same symptoms in older children and adults. Both diagnosis and treatment may be influenced by the more sensitive pattern of reaction and, in many cases, therapeutic regiments differ from those of adults.

In the beginning of any diagnostic process, a careful and adequate patient and family history should be obtained followed by a detailed physical examination of the child's body. Easy visibility of the lesion may sometimes lead to cursory examination and hasty diagnosis.

Examination of the entire skin including the scalp, oral and anogenital regions, periorbital skin and conjunctiva,

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palms and soles, as well as nails and hair should be routinely performed to obtain a clue for final diagnosis.

Examination should always be performed in a well-temperatured room. Overwarming or overcooling of sensitive skin, especially that of newborns and neonates may lead to wrong impressions and misdiagnosis of physiological skin response to environmental stimuli as a specific pathologic condition (i.e., vascular phenomenon).

The morphology and configuration of cutaneous lesions are not always specific, but are of great importance to the classification and, consequently, correct diagnosis of skin diseases. Therefore, understanding dermatologic terminology is of great importance to a physician who is not a dermatologist. This also helps to eliminate barriers in communication and misunderstandings if specific dermatologic consultation is requested and when a dermatologist is responding to pediatrician consultation using his or her specific "skin-derived" language.

It is important to understand that most dermatologic terms are descriptions of the morphology of the lesion and applicable to many different diseases. Appropriate description of the lesion, timeline of its appearance, changes over time, and associated symptoms (i.e., itching or burning versus asymptomatic course) are crucial in distinguishing between different pathologic conditions.

Annular, nummular, circinate, or *ring-shaped lesions* (ringworm-like) may most often be found in superficial fungal infections. However, ringed lesions can be seen in seborrheic dermatitis, nummular eczema, pityriasis rosea, erythema anulare, erythema migrans, erythema multiforme, lupus erythematosus, urticaria, lupus vulgaris, leprosy, tinea versicolor, secondary syphilis, and many others.

Lesions that have an arc-like configuration are named *arciform* or *arcuate*; if they show a tendency to merge they are described as *confluent* and most commonly occur in exanthemas, pityriasis rosea, urticaria, and erythema exsudativum multiforme.

Distribution of the lesion over specific dermatome (single or multiple) is described as *dermatomal* because it is most commonly seen in HSV-3 infection.

Lesions that are solid, infiltrated, and discus-shaped (solitary or multiple) are named *discoid*, as in lupus erythematosus, discoid eczema, granulomas.

If skin lesions are drop-like, infiltrated, and showing a covering scale, they are called *guttate* and are often seen in childhood psoriasis or in adolescents and often follow acute respiratory tract infections (i.e., early pityriasis rosea).

Lesions showing spiral-like, coiled, or twisted appearance are described as *gyrate* (i.e., in urticaria, erythema anulare).

If such lesions have more than one kind of shape, they are considered to be *multiform* or *polycyclic* (i.e., urticaria, erythema multiforme, polymorphous light eruption).

Serpentine or snake-like morphology is named *serpiginous* and is most commonly seen in larva migrans and elastosis perforans serpiginosa.

Lesions presenting with a central depression and shaped like an umbilicus are termed *umbilicated*, as is the case in molluscum contagiosum, varicella, variola, eczema herpeticum, or HSV-III infection.

Vesicular and papular lesions localized in clusters are referred to as *grouped*, as in cases of HSV infection (HSV-I, -II or -III), insect bites, bullous dermatosis of childhood, and less commonly in contact dermatitis.

Iris-like or *target* lesions are those having a concentric, ringed appearance most characteristic of erythema exsudativum multiforme and Stevens-Johnson syndrome.

Lesions having line or stripe form are defined as *linear* or *band-like* and are most commonly seen in epidermal nevi, linear morphea, lichen striatus, incontinentia pigmenti, hypomelanosis of Ito, acanthosis nigricans, etc.

Lesions demonstrating a *net-like* or *murmur-like* pattern are described as reticulated – i.e., in livedo reticularis, curis marmorata, and cutis telangiectatica congenital.

An important pattern of skin morphology often indicating a specific disease is called *isomorphic response* or *Koebner phenomenon*. This term describes the appearance of new lesions along the site of a superficial injury. The most prominent example is a linear development of new lesions in psoriasis vulgaris, lichen planus, or keratosis folicularis. Linear spreading of warts or molusca contagiosa after scratch inoculation is also an example of this phenomenon.

The morphological configuration and regional distribution of skin lesions, presence or absence of mucous membrane involvement, or coincidence of skin and nail lesions may also be of importance in the finding of correct diagnosis.

Dependence from exposure to UV light (i.e., sun exposure areas) is important in estimating a possible photosensitive background or establishing a relationship between medication and the appearance of skin lesions.

Coexistence of itching and aggravation of symptoms in a warm environment (e.g., nights at bedtime) together with specific localization (i.e., intertriginous areas, genitalia, interdigital spaces) and coexistence of similar symptoms in other family members in close contact with each other is specifically suggestive of infestations such as, scabies. However, in infants and small children this diagnosis may be difficult because the lesions are typically

localized on the palms and soles and often on the neck and the head. A change of skin color frequently helps in making the correct diagnosis.

Brown or brownish discolorations are often associated with postinflammatory hyperpigmentation, pigmented nevi, incontinentia pigmenti, café-au-lait spots, epidermal nevi (together with hyperkeratotic, villous-like structure), photodermatitis and phytophotodermatitis, acanthosis nigricans, or as iatrogenic-induced hyperpigmentation by use of local irritants or concentrated fragrances (melasma, chloasma).

Yellow discoloration, especially in infants, is often connected with the presence of carotene derived from food, particularly yellow vegetables and carrots. Examination of sclera helps in distinguishing this phenomenon from an icterus.

Red or purple lesions are usually of vascular origin like superficial hemangiomas, naevi flamei, or spider telangiectasias.

Rose color is often characteristic for active inflammation or in development of inflammatory lesions such as atopic dermatitis or psoriasis. The latter often show superficial scaling and pinpoint bleeding known as the Auspitz phenomenon.

Lesions with decreased pigmentation (hypopigmentation or depigmentation) may be seen in several pathologic conditions. Superficial tinea, pityriasis versicolor may be easily distinguished by Wood's lamp. Localized depigmentation in different shape and form may be seen in vitiligo, piebaldism (clinical picture, family history, and dynamics of lesion are important distinguishing factors), and chemically induced pigment loss or progressive macular hypomelanosis.

Postinflammatory lesions may also appear hypopigmented. In children with atopic constitution or atopic dermatitis, multiforme hypopigmentation is described as pityriasis alba. Generalized decrease in skin pigmentation may be seen in albinism or untreated phenyloketonuria.

Ethnic variations need to be taken into consideration in assessment of skin changes. Erythema or inflammation may be difficult to see in children with black skin; postinflammatory hyperpigmentation or hypopigmentation is usually much more prominent in black, Mediterranean, Arabic, and Asian populations.

Hyperpigmentation of flexure areas (neck, axilla, inguinal region, perianal region) are considered to be physiologic in children with colored skin. However, physicians may be confronted with the pressure of parents who are not willing to accept that their children have such normally occurring skin discoloration, even if the parents have the same skin symptoms.

Qualities of hair may also differ among individuals of different ethnic origins. African-American and African hair tends to be extensively curly, to tangle when dry, and becomes matte when wet. Due to the naturally curly or spiral nature of the hair and appearance of so-called bushy hair, folliculitis and pseudofolliculitis are more common in children of African, African-American and in some groups of Asian descent.

Prolonged and continuous traction on hair results more commonly in traction alopecia in African, Asian, and Arabic populations.

Wound healing complications, especially development of keloids and hypertrophic scars after surgery or burns, are reported more often in children of African, Asian, or Arabic origin. Similarly, keloid complications of acne or other follicular disorders are more prominent in children and adolescents with colored skin.

Skin is not just a cover of the human body. It helps to adapt to environmental conditions; communicate with the surrounding world in a physical, chemical, and immunologic manner; and is an organ of interpersonal communication.

Most important for appropriate skin function is to keep the skin barrier intact and to restore it as quickly as possible if the barrier function is impaired.

Interruption of disruption of the barrier may lead not only to skin dysfunction, but also to impaired function of other organs, i.e., to immunologic defects. Skin and mucous membranes are the targets of airborne and contact allergens.

There is a common belief that the best way to keep the skin barrier and skin itself in good shape is to make it moist and fat. However, this widely believed thinking is not correct. Use of fat lubricants (i.e., vaseline or liquid paraffin) may seal the skin surface, which increases the accumulation of water and heat below the applied product and leads to increased itching and worsening of skin lesions. This is of particular importance in hot and humid climate areas.

Many nondermatologists may be hesitant to use some specific agents in younger patients because of safety concerns.

Two groups of topical skin medication are of special interest: topical corticosteroids and emollients.

Emollients and moisturizers are of particular interest because fragile, young skin needs them more than adult skin and – because of high permeability – it may absorb applied ingredients in much higher concentrations.

Children's skin should be kept "not too dry" and "not too moist (fat)," and achieving this balance has become a science in itself.

In this section, dermatologic conditions most relevant in a pediatrician's daily practice are reviewed. Our goal is to give a practical overview and to help in the assessment and management of common skin diseases in neonatal, child, and adolescent populations. The intent is to not to deeply discuss etiologic, genetic or therapeutic issues, especially if they are in an experimental stage of medical knowledge or if only a single clinical report describes specific clinical outcome.

For detailed reviews and further studies, pediatric dermatology textbooks and monographs are recommended. A short list of such fundamental recent books is given below.

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