

PSORIASIS: A SPECTRUM OF EXPRESSIONS

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SUMMARY

Psoriasis is a frequently occurring skin condition caused by hereditary factors and triggered by environmental factors. Psoriasis has a heterogeneous expression pattern. Psoriasis may occur as chronic plaque psoriasis, erythrodermic psoriasis, psoriasis inversa and psoriasis guttata. In many patients the nails are involved as well and in one out of ten patients arthropathy complicates the picture. Before considering a treatment, non-hereditary factors should be investigated and the patient should be informed about the factors, which may aggravate psoriasis. It is of importance that the patient is ascertained that psoriasis is not contagious and that the patients are informed about the risk of psoriasis in next generations. A treatment is selected based on the clinical expression of psoriasis, the acceptability of psoriasis for both the patient and his/her environment and the social circumstances of the patient.

KEYWORDS

epidemiology, epidermis, genetics, inflammation, proliferation, psoriasis, skin

INTRODUCTION

Psoriasis is a complex spectrum of expressions (1). Genetic factors as well as environmental factors determine the fate of psoriasis. In 1997 the possibilities for adequate treatment have been improved.

EPIDEMIOLOGY

Psoriasis occurs in 2% of the world population (2,3). However the occurrence of psoriasis is not distributed at random over the world (4,5). Psoriasis is relatively seldom in American Indians and in the

Table 1. Risk to get psoriasis (4)

Family members involved with psoriasis	Risk (%)
One parent, no brother or sister	10
No parent, one brother or sister	7
One parent, one brother or sister	16
No parent, two brothers or sisters	16
Both parents	50
Second grade family	4
Third grade family	1-2

native population in the Far East. The low prevalence of psoriasis in black Americans is in line with the low prevalence of psoriasis in the black population of Western Africa as opposed to the frequency of psoriasis in the East of Africa. In Java, the frequency of psoriasis in Europeans was 30 times increased compared to the native population of Java. According to a study, carried out in the USA, 36% of the psoriatic patients stated that one or more family members were also affected by psoriasis (6). The male/female ratio is approximately 1.

The genetics of psoriasis is not determined by a single gene. Psoriasis is a polygenic inherited disease. Table 1 summarizes an estimation of the risks of reoccurrences of psoriasis in families in which at least one person is suffering from the disease.

The age of onset of psoriasis varies considerably. Before the 25th year of age psoriasis had been expressed in the majority of patients (7,8). Psoriasis may occur already in children but may also occur for the first time at an advanced age. Environmental factors play an important role in the initiation of psoriatic skin manifestations in a genetically predisposed patient.

Table 2. Triggering factors

Triggering factors	
Systemic	Topical
<ul style="list-style-type: none"> • Infections • Stress • Hypocalcemia 	<ul style="list-style-type: none"> • Skin disorders • Trauma



Fig. 1. Chronic plaque psoriasis

Systemic factors may induce the expression of psoriasis after some days and weeks. Focal infections (tonsillitis, sinusitis, periapical granuloma, and cholecystitis) may induce psoriasis guttata, or may change the picture from chronic plaque psoriasis into unstable psoriasis. Bacterial superantigens may induce an immune response, responsible for the inflammatory changes at this stage of the disease (Table 2).

The relevance of psychological stress with respect to psoriasis has been debated.

Some authors are of the opinion that psychological stress is virtually unimportant in the initiation of psoriasis (1), but other authors state that psychological stress is a triggering factor in psoriasis in 40% of the patients (1). The relationship between disease-activity and psychological stress is difficult to assess. It is the opinion of the author that psychological stress is of major importance in the course of psoriasis.

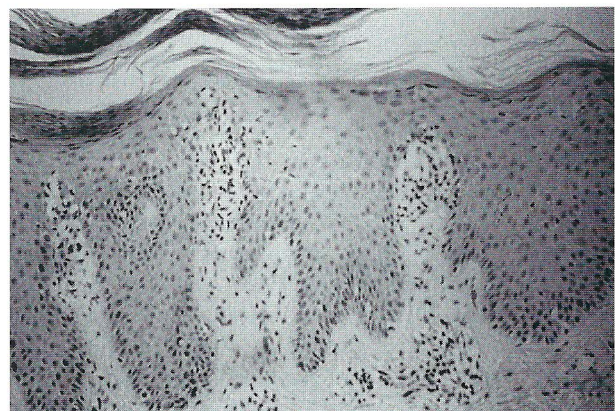


Fig. 2. Histological appearance of plaque psoriasis with prolonged rete ridges, parakeratosis and inflammatory infiltrate

Table 3. Local factors which may induce psoriasis

LOCAL FACTORS WHICH MAY INDUCE PSORIASIS
SKIN DISORDERS
Bites of insects
Burns
Dermatitis
Drug reactions
Excoriations
Photosensitivity
Herpes zoster
Incisions
Lichen planus
Lymphangitis
Miliaria
Vitiligo
Pityriasis rosea
TREATMENTS
Electrodesiccation
Liquid nitrogen
Pressure(belt)
Primary irritants
Radiation (UVR, X-ray)
Scarification
Skin tests (scratch, injection)
Vaccination

Various medications may induce psoriasis. Use of lithium carbonate, cardioselective, and non-cardioselective betablocking agents, antimalarial drugs, inhibitors of angiotensine converting-enzymes and non-steroid anti-inflammatory drugs. Also local factors aggravate psoriasis. Following injury of the skin psoriasis may occur within a few weeks. Various skin disorders induce psoriasis at the site of occurrence. Table 3 provides an overview of these external-triggering factors.

The induction of psoriasis following a local triggering factor has been described in 1883 for the first time by Heinrich von Koebner. The so-called "Koebner phenomena" occur in 25% of patients with psoriasis.

So far the genetics of psoriasis has not been resolved. Associations with HLA B13, B17, B27, B37, Cw6, and DR7 as well as the association with the apolipoprotein E-gene, the alpha-1 antitrypsine inhibitor gene and the gene of the interleukin-1 receptor antagonist suggest a polygenic inheritance (9). On the other hand, various authors suggest a monogenic inheritance. Traupe and co-workers proposed the concept of one major gene and genomic

imprinting, suggesting the relevance of some modifying genes (10). Recently a predisposing gene has been identified on chromosome 17 (11).

Inflammation, increased epidermal proliferation, and abnormal keratinization are the hallmarks of the psoriatic plaque. Inflammation in the psoriatic lesion is characterized by accumulation of polymorphonuclear granulocytes and mononuclear cells in dermis and epidermis. A wide spectrum of mediators of inflammation has been reported to occur in psoriatic scales, varying from arachidonic acid metabolites, interleukins, and platelet activating factors and complement factors. The pathogenetic relevance of these factors, so far, remains to be elucidated as we are not informed whether these factors are primarily or secondarily involved in the pathogenesis of psoriasis.

A T-cell response and the accumulation of polymorphonuclear granulocytes probably play a major role in the pathogenesis of psoriasis (12,13). The dermis in the psoriatic plaque is abnormal in many respects. The endothelium is activated (14) and the stroma contains fibroblasts with an increased proteoglycan synthesis (15). The dermis of the psoriatic plaque is characterized by changes of the extracellular matrix with increased expression of the molecule tenascin (16). The epidermis of the psoriatic lesion is hyperproliferative with increased recruitment of cycling epidermal cells from the resting G₀ compartment (17). The cell cycle of the epidermis in psoriasis is normal. A premature process of keratinization with absence of stratum granulosum and a stratum corneum with remnants of cell nuclei character-

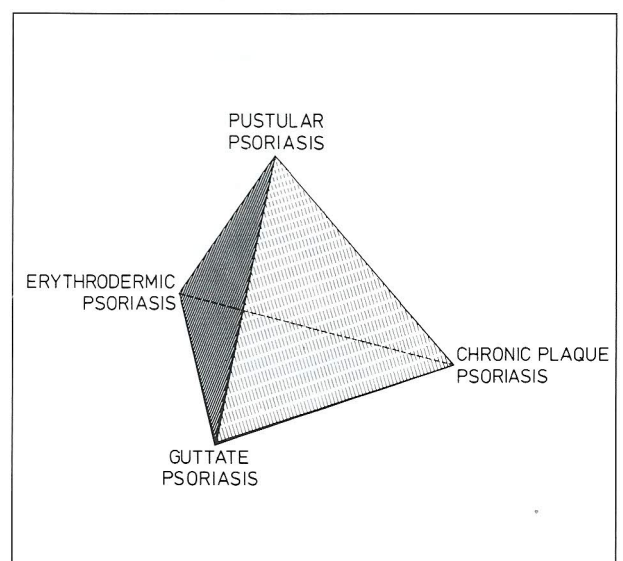


Fig. 3. Spectrum of manifestations

rizes the psoriatic lesion.

Inflammation, epidermal proliferation, and abnormal keratinization are expressed at the macro level as erythema, induration, and scaling.

PATHOLOGY AND CLASSIFICATION

The symptomless skin of a psoriatic patient has a normal histological appearance. The clinically involved skin (Fig. 2) shows deviations at various levels. The expression of each of these features can be different comparing individual patients. If the inflammatory changes are dominating, macropustules filled with polymorphonuclear granulocytes can be formed within the epidermis. The clinical picture in such a situation is psoriasis pustulosa.

The extent of the body surface involved with psoriasis may vary from patient to patient. In some patients, psoriasis is restricted to some sharply demarcated erythematous squamous plaques. This picture is designated as chronic plaque psoriasis.

In other patients small erythematous squamous papules are distributed over the body surface. This picture is designated as psoriasis guttata. In patients with psoriasis guttata it is of importance to search for systemic triggering factors. In erythrodermic psoriasis the skin as a whole is involved.

In fact four prototypes identify the extremes of the psoriasis spectrum:

- (i) Chronic plaque psoriasis
- (ii) Psoriasis guttata
- (iii) Erythrodermic psoriasis
- (iv) Psoriasis pustulosa

CLINICAL MANIFESTATIONS AND THEIR PROGNOSSES

Every patient has his own psoriasis with unique characteristics with respect to presentation and treatment response. The individual position of a patient with psoriasis is between the extremes of a three dimensional spectrum which can be constructed between the above mentioned prototypes (Fig. 3).

- (i) Chronic plaque psoriasis

This manifestation is the most frequent one in adults. It occurs in 90% of the patients. Predilection areas are the extensor surfaces of the elbows (Fig.

1), the knees, the sacrum, and the scalp. Lesions may occur on all sites. The face is involved relatively seldom. The demarcation of the chronic plaque lesions is remarkably sharp.

If psoriasis is triggered, and if new lesions occur, the patient may experience itch. Instable psoriasis is characterized by multiple papules and papulo-pustules around the psoriatic plaques.

Chronic plaque psoriasis may occur at "difficult" localizations. These include the regio genitalis, including the mucous surfaces of the genitalia, the palms and soles and the flexures. Often a bacterial overgrowth or a mycosis is a complicating factor, which triggers flexural psoriasis. In case of psoriasis of the scalp a loss of hair may occur. The course of chronic plaque psoriasis is chronic over years. In 10% of the patients remission periods of 5 years or longer may occur (18).

- (ii) Psoriasis guttata

Guttate psoriasis is characterized by multiple itchy erythematous squamous papules, distributed over the body (Fig. 4). Guttate psoriasis occurs predominantly in children. According to literature between 44-95% of the psoriatic children have this manifestation of psoriasis. Also at adult age, this variant may be seen in combination with other manifestations of psoriasis. As a rule systemic triggering factors are responsible for the occurrence of guttate psoriasis. Following elimination of systemic eliciting factors skin manifestations resolve spontaneously in a few weeks time.

- (iii) Erythrodermic psoriasis

In erythrodermic psoriasis the skin as a whole is involved with erythema and scaling. In case of erythroderma, systemic complications may occur. The erythrodermic patient is poikylotherm with hypo- and hyperthermia, protein loss, abnormal water-salt equilibrium, kidney insufficiency and cardiac insufficiency. Erythrodermic psoriasis may occur early or late in the course of chronic plaque psoriasis. The course of erythrodermic psoriasis is variable. Goeckerman and O'Leary described the natural course in 10 patients suffering from erythrodermic psoriasis. In 5 patients chronic plaque psoriasis developed, in 2 patients erythroderma relapsed and in 3 other patients remissions occurred for 9 years. Eliciting factors are of major importance in the induction of erythrodermic psoriasis. In this respect it is of importance that antipsoriatic treatments with an



Fig. 4. Psoriasis guttata



Fig. 6. Pustulosis palmoplantaris



Fig. 5. Generalized pustular psoriasis

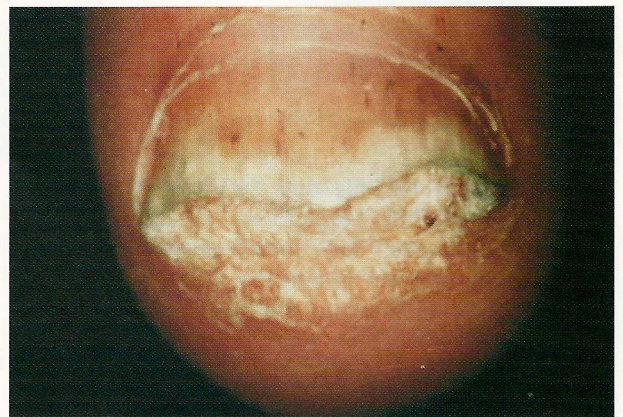


Fig. 7. Nail psoriasis. Subungual hyperkeratosis and distal onycholysis

irritant potential such as photo(chemo)therapy and dithranol, may induce erythroderma. Discontinuation of systemic and topical corticosteroids may also induce erythrodermic psoriasis.

(iv) Psoriasis pustulosa

Psoriasis pustulosa may occur as generalized pustular psoriasis or localized pustular psoriasis. The pustules are sterile. Generalized pustular psoriasis is a relatively seldom condition characterized by episodes of erythema and pustule formation (Fig. 5,6) with fever and feeling of illness. This phase may occur for a few days and is followed by a phase of scaling.

In an investigation of 104 patients in Great Britain, 62 patients proved to have either chronic plaque psoriasis or psoriasis guttata before the occurrence of pustular psoriasis (19). Triggering factors which may induce generalized pustular psoriasis are pregnancy, corticosteroid treatment, hypocalcemia, irritation (dithranol, phototherapy) and focal infections. The localized manifestations of psoriasis pustulosa can be divided into three subgroups.

PSORIASIS ARTHROPATHICA

In 5 - 10% of patients with psoriatic manifestations arthropathy may occur. For a detailed review on psoriasis arthropathica the reader is referred to the literature (1).

NAIL PSORIASIS

Psoriasis of the nails is the result of changes in the nail matrix and nail bed. The nail matrix is the origin of the nail plate. If the psoriatic process affects the nail matrix, sharply punched out pits occur in the nail plate (Fig. 7). The extrusion of protein rich fluid under the nail plate causes brownish discoloration (oil spot phenomenon), and formation of hyperkeratoses under the nail plate together with distal onycholysis of the nail plate from the nail bed. Fig. 7 demonstrates these features of subungual hyperkeratosis at distal onycholyses. The occurrence of nail-changes in patients with psoriasis varies between 10 - 55%.

DIFFERENTIAL DIAGNOSIS

Chronic plaque psoriasis may be difficult to differentiate from seborrhoeic dermatitis. Involvement of the scalp and flexures may be rather similar in both conditions. A yellowish scaling is in favour of seborrhoeic dermatitis whereas the occurrence of psoriatic plaques elsewhere on the body is in favour of psoriasis. Psoriatic plaques with an elevated scaling border are suggestive of epidermomycoses. If the erythematous plaque is not sharply demarcated parapsoriasis or lymphoma of the skin should be considered. Lues II should be considered in case of dissemination of erythematous plaques on the trunk and involvement of palms and soles.

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