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Pityriasis rubra pilaris

What is pityriasis rubra pilaris?

Pityriasis rubra pilaris (PRP) is the name given to a group of rare skin disorders that present with reddish–orange coloured scaling patches with well defined borders. They may cover the entire body or just parts of the body such as the elbows and knees, palms and soles.

Often there are areas of uninvolved skin, particularly on the trunk and limbs, which are referred to as "islands of sparing".

The palms and soles are usually involved and become diffusely thickened and yellowish (palmoplantar keratoderma). PRP is often initially mistaken for another skin condition, usually [psoriasis](#).

Why does it occur and who is at risk?

The cause of PRP is unknown. PRP is usually sporadic but in some forms it may be partially inherited. It may occur in any race, and males and females are equally affected.

There are several forms of PRP that are classified as follows:

Type	Features
Classical adult onset	<ul style="list-style-type: none"> • Accounts for 50% of all cases • Good prognosis with 80% of patients going into spontaneous remission within 3 years • After remission, relapses are uncommon • May be associated with underlying malignancy
Atypical adult onset	<ul style="list-style-type: none"> • Accounts for 5% of all cases • May persist for 20 years or more • Often has an eczematous component
Classical juvenile onset	<ul style="list-style-type: none"> • Accounts for 10% of all cases • Usually occurs between 5 and 10 years old • Often follows an acute infection of any type • Spontaneous remission within 1 year
Circumscribed juvenile	<ul style="list-style-type: none"> • Accounts for 25% of all cases • Occurs in prepubertal children • Usually confined to palms, soles, knees and elbows • Long term outcome unclear but possible improvement in late teens
Atypical juvenile onset	<ul style="list-style-type: none"> • Accounts for 5% of all cases • Sometimes inherited; may overlap with types of ichthyosis • Occurs at birth or early in childhood and is very persistent

HIV-associated

- Disease tends to be resistant to standard therapies

Juvenile pityriasis rubra pilaris



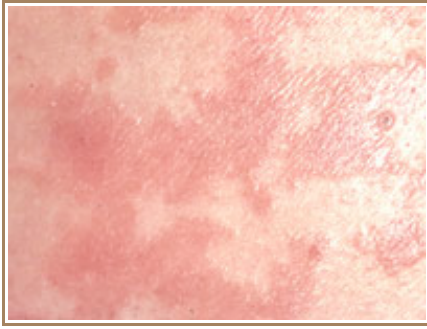
What does it look like?

Classical adult-onset PRP most often starts on the head, neck and upper trunk as a red scaly rash. Often there is a solitary lesion but within a few weeks multiple patches appear and they join together to form groups of reddish-orange lesions. Over a few weeks these spread downwards and may cover most of the body (erythroderma). Rough, dry plugs can be felt within the patches and are due to plugged hair follicles, often most obvious on the backs of the fingers. Patients may also complain of itching in the early stages of the disease.

The palms and soles become thickened and yellow coloured. Cracks may develop which can be painful and make walking and using the hands difficult. The nails may become thickened and discoloured at the free nail edge and may show linear black streaks (splinter haemorrhages). The hair may thin considerably.

Adult pityriasis rubra pilaris





What treatments are available?

The value of treatment is difficult to assess, as the clinical course is so variable for each of the different types of PRP. In general, the inherited forms of PRP tend to persist throughout life whilst the sporadically acquired forms tend to resolve spontaneously eventually.

Topical medications such as [emollients](#) may be used to relieve symptoms of dryness and cracking. In severe cases [acitretin](#) or [isotretinoin](#) may be useful. Methotrexate has been of limited value.

Related information

References:

Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

Other websites:

- [Support group](#)
- [Pityriasis rubra pilaris](#) - emedicine dermatology, the online textbook

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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