

PITYRIASIS RUBRA PILARIS

What are the aims of this leaflet?

This leaflet has been written to help you understand more about pityriasis rubra pilaris. It tells you what it is, what causes it, what can be done about it, and where you can find out more about it.

What is pityriasis rubra pilaris?

Pityriasis rubra pilaris is the name given to a group of uncommon skin disorders that look rather similar, but which are probably different conditions. Their name means that they share some of the same features, e.g. scaling (pityriasis), redness (rubra) and involvement of the hair follicles (pilaris).

Several types of pityriasis rubra pilaris are now recognised:

- By far the most common is the 'classic adult type', which causes a widespread rash during adult life. A variation of this affects young children, and this is known as the 'classic juvenile type'.
- The next most common is the 'circumscribed juvenile type' that affects the palms and soles, the fronts of the knees, and the backs of the elbows of children.
- Two other atypical (*not* typical) rare types are also known.

What causes pityriasis rubra pilaris?

The cause of pityriasis rubra pilaris is not known. It is not an infection and cannot be passed on to others.

Is pityriasis rubra pilaris hereditary?

No, the common pityriasis rubra pilaris types are not hereditary.

What are the symptoms of pityriasis rubra pilaris?

- The rash can be itchy in its early stages.
- Thick skin on the palms and soles can split and become painful. Walking may be sore.
- Shivering, heat and fluid loss may occur if the rash is angry and widespread.

What does pityriasis rubra pilaris look like?

The main features of the 'classic adult type' of pityriasis rubra pilaris are as follows:

- The rash usually starts suddenly on the scalp and spreads to cover much of the trunk.
- The patches are dry, scaly and red with an orange tinge, and have well defined edges. Patches may join together to cover large areas of skin; occasionally, people with pityriasis rubra pilaris become red all over.

- ‘Spared areas’, where there is no rash, can often be seen lying inside the main patches of redness.
- Rough dry plugs in the hair follicles can often be seen or felt within the red patches, and also in the ‘spared areas’.
- The palms and soles thicken and turn orange. The nails may thicken too and go brown; sometimes they are shed.
- The features of the ‘circumscribed juvenile type’ are well-defined red patches with hair follicle plugs on the fronts of the knees and a yellowish thickening of the palms and soles.

How will pityriasis rubra pilaris be diagnosed?

The different types of pityriasis rubra pilaris may look like psoriasis and are often mistaken for it. Your doctor will probably refer you to a skin specialist; to confirm the diagnosis a small piece of skin (a biopsy) can be removed under a local anaesthetic and examined under the microscope. There is no blood test for pityriasis rubra pilaris.

Can pityriasis rubra pilaris be cured?

The outlook in pityriasis rubra pilaris depends on its type. Treatment helps but it cannot be guaranteed to clear either of the two most common types. The ‘classic adult type’ often goes away after two or three years, but it may persist for longer. The ‘circumscribed juvenile type’ tends to last for life.

How can pityriasis rubra pilaris be treated?

Applications to the skin are all that is needed if pityriasis rubra pilaris is mild:

- Steroid creams and ointments soothe but probably don’t alter the rash.
- Greasy applications (emollients), such as soft white paraffin, reduce the splitting of the thickened palms and soles.
- Medication by mouth may be needed if the pityriasis rubra pilaris is severe. These medications should only be undertaken by a dermatologist as monitoring is necessary
- Pityriasis rubra pilaris often improves if treated with retinoid drugs such as acitretin.
- Methotrexate, which is an immunosuppressant drug, may help pityriasis rubra pilaris.
- Occasionally, other oral or injected medicines may be used if the condition is resistant to standard treatment.
- The ‘circumscribed juvenile type’ needs only the applications to the skin

Where can I get more information about pityriasis rubra pilaris?

Web links to detailed leaflets:

www.dermnetnz.org/dna.psoriasis/prp.html

Links to patient support groups:

Pityriasis Rubra Pilaris Support Group

Web: www.prp-support.org