



## **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. The name means scaling (pityriasis), redness (rubra) and involvement of the hair follicles (pilaris).

Several types of pityriasis rubra pilaris are currently recognised including:

- Type I. This is the commonest and is called 'classic adult type'. It causes a widespread red and scaly rash. A variation of this affects children, and is called 'classic juvenile type' (type III).
- Another type that affects children is called 'circumscribed juvenile type' (type IV) and causes a rash on the palms and soles, the front of the knees, and the back of the elbows.
- A rarer genetic form also occurs (type V).

### **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?**

Pityriasis rubra pilaris is usually not hereditary but there is a rare genetic type.

## **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 covers large areas of skin.

## **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; this is called erythroderma.
- '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 have an orange colour. The nails may thicken and may be discoloured and sometimes 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 diagnosis is made by examination of the skin and can be confirmed by a skin biopsy. Skin biopsy means that a small piece of skin is removed under a local anaesthetic and examined under the microscope. Your doctor will probably refer you to a skin specialist to confirm the diagnosis. There is no blood test for pityriasis rubra pilaris. The different types of pityriasis rubra pilaris may look like psoriasis and are often mistaken for psoriasis.

## **Can pityriasis rubra pilaris be cured?**

The outlook for pityriasis rubra pilaris depends on the type. Treatment helps but it cannot be guaranteed to clear either of the two most common types.

The 'classic adult type' often resolves after two or three years, but it may persist for longer. The classic juvenile type usually resolves on average in about one year while the circumscribed juvenile type tends to persist.

### **How can pityriasis rubra pilaris be treated?**

Creams or ointments applied to the skin may be all that is required if the pityriasis rubra pilaris is mild:

- Steroid creams and ointments soothe but probably do not alter the duration of the rash.
- Emollients recommended by your doctor or specialist are a very important part of treatment to help dry skin and restore the barrier function of the skin. The emollient should be applied liberally.

Tablet medication may be needed if the pityriasis rubra pilaris is severe. These medications should only be prescribed by a dermatologist as monitoring of blood tests is necessary:

- Pityriasis rubra pilaris often improves if treated with a retinoid drug such as [acitretin](#).
- [Methotrexate](#), an immunosuppressant drug, may help pityriasis rubra pilaris.
- The 'circumscribed juvenile type' usually only requires the topical applications mentioned above.

### **Where can I get more information about pityriasis rubra pilaris?**

*Web links to detailed leaflets:*

[www.dermnetnz.org/dna.psoriasis/prp.html](http://www.dermnetnz.org/dna.psoriasis/prp.html)

*Links to patient support groups:*

*Pityriasis Rubra Pilaris Support Group*

Web: [www.prp-support.org](http://www.prp-support.org)

*PRP Alliance, Inc.*

Web: [www.prpalliance.org](http://www.prpalliance.org)

*PRP Community Page on RareConnect*

Web: [www.rareconnect.org/en/community/pityriasis-rubra-pilaris](http://www.rareconnect.org/en/community/pityriasis-rubra-pilaris)

For details of source materials used please contact the Clinical Standards Unit ([clinicalstandards@bad.org.uk](mailto:clinicalstandards@bad.org.uk)).

**This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.**

*This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel*

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