



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: scaling (pityriasis), redness (rubra) and involvement of the hair follicles (pilaris).

Several types of pityriasis rubra pilaris are now recognized:

- By far the most common is the 'classic adult type', which causes a widespread rash during adult life. A variation on this affects young children - 'the classic juvenile type'.
- Next most common is the 'circumscribed juvenile type' that affects the palms and soles, and points of the knees and elbows of children.
- Finally there are two other types of pityriasis rubra pilaris that are less typical – the 'atypical adult' and 'atypical juvenile' types. Both are too rare to be discussed further here.

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?

One type of pityriasis rubra pilaris (the 'atypical juvenile type' – see above) does run in families, however in most instances pityriasis rubra pilaris is 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 may occur if the rash is angry and widespread and loses a great deal of heat.

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 then spreads downwards to cover much of the trunk.
- The patches are dry and scaly, red with an orangey tinge, and have sharp edges. They can join together to cover large areas of skin, and 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 '*circumscribed juvenile type*' shows:

Sharply defined red patches, with plugging of the hair follicles, on the points of the knees and Sometimes there is a yellowish thickening of the palms and soles.

How will pityriasis rubra pilaris be diagnosed?

In some ways the different types of pityriasis rubra pilaris look rather like psoriasis and are often mistaken for it. Your doctor will probably refer you to a skin specialist who will make the diagnosis by recognising the special combination of features mentioned above. To confirm it, 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 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 go on for much longer. The '*circumscribed juvenile type*' tends to last for life.

How can pityriasis rubra pilaris be treated?

The '*classic adult type*' of pityriasis rubra pilaris is treated as follows:

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

- Steroid creams and ointments soothe but probably do little to get rid of 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 bad:

- Pityriasis rubra pilaris improves if it is treated with a derivative of vitamin A known as acitretin. However this carries with it a wide range of potentially

serious side effects, including the ability to damage an unborn child. Treatment with acitretin can only be undertaken by a dermatology department, and has to be monitored very carefully.

- Methotrexate, a strong drug that suppresses the immune system may help pityriasis rubra pilaris, but it too has many side effects.

The 'circumscribed juvenile type' needs only the applications to the skin mentioned above.

Where can I get more information about pityriasis rubra pilaris?

Link to patient support group:

www.prp-support.org

Web links to other Internet sites:

www.emedicine.com/derm/topic337.htm

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

(While every effort has been made to ensure that the information given in this leaflet is accurate, not every treatment will be suitable or effective for every person. Your own doctor will be able to advise in greater detail)

**BAD PATIENT INFORMATION LEAFLET
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