PATIENT INFORMATION LEAFLET

PITYRIASIS RUBRA PILARIS

WHAT ARE THE AIMS OF THIS LEAFLET?

This leaflet has been written to help you understand more about pityriasis rubra pilaris (PRP). 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?

PRP is a rare, long-term inflammatory skin condition. The name means scaling (pityriasis), redness (rubra) and involvement of the hair follicles (pilaris). It affects all races and sexes equally.

It is classified into six clinical types:

- Type I is the most common and is called 'classic adult type'.
- Type II is a variation of type I, known as atypical adult onset.
- Type III, IV and V represent classical, circumscribed and atypical juvenile forms.
- Type VI is PRP associated with HIV (human immunodeficiency virus) infection.

WHAT CAUSES PITYRIASIS RUBRA PILARIS?

The cause of PRP is not known. It is not an infection and therefore cannot be passed on to others.

IS PITYRIASIS RUBRA PILARIS HEREDITARY?

Usually, PRP is not hereditary but there have been reports of families with multiple affected members.

WHAT DOES PITYRIASIS RUBRA PILARIS FEEL AND LOOK LIKE?

PRP can look and feel different in people affected by it:

- The rash which can be itchy in its early stages
- Thick skin on the palms and soles that splits and becomes painful. Walking may be difficult
- Shivering, temperature dysregulation and fluid loss may occur if the rash covers large areas of skin.

The most common type of PRP (type I, classic adult) accounts for half of all cases and has the following features:

- The rash usually starts suddenly on the scalp and spreads to cover much of the chest and abdomen.
- The patches are dry, scaly and red with an orange tinge and have well-defined edges. Smaller patches may join together to cover large areas of skin. Occasionally, people with PRP can become red all over; this is called erythroderma.
- Areas of normal-looking skin where there is no rash, known as 'spared areas', can often be seen lying between larger, red patches.
- The hair follicles in the affected areas can feel rough to the touch because of build-up of scale at their base (follicle plugs).
- The skin on the palms and soles may become thickened and have an orange colour. The nails may thicken, be discoloured and sometimes shed.
- The nails can also be affected.

Type II (atypical adult) accounts for 5% of all PRP cases and does not follow the head-to-toe progression described above. Typically, it affects the legs with thickening of the skin on the palms and soles. It can also be associated with hair loss on the body.

Type III accounts for 10% of all cases. The presentation is similar to Type I, but the main difference is that it starts in childhood.

Type IV (circumscribed juvenile) accounts for 25% of cases. It affects children before puberty and is characterised by well-defined, red patches, hair follicle plugs on the knees and elbows, as well as thickening of the skin on the palms and soles.

Type V represents 5% of cases. It is commonly familial, i.e. there are other family members affected. It starts early in life with dryness, hair follicle plugs and sometimes thickened skin on the hands and feet.

Type VI is seen in people with HIV and is characterised by red, scaly spots around the hair follicles and may or may not involve the palms and soles.

HOW WILL PITYRIASIS RUBRA PILARIS BE DIAGNOSED?

The diagnosis is made by examination of the skin and nails by a doctor or nurse. A skin biopsy is not necessary but may be done to exclude other causes of widespread redness. Skin biopsy is where a small piece of skin is removed under a local anaesthetic and examined under a microscope. A referral to a skin specialist may be made by the doctor to confirm a diagnosis. There are no specific blood tests to confirm PRP. The different types of PRP may look like psoriasis and are often mistaken for psoriasis especially at the early stages.

CAN PITYRIASIS RUBRA PILARIS BE CURED?

The natural history of PRP depends on the type. Due to its rarity, no studies which include a large enough number of patients have been possible, and there is no consensus on treatment. Multiple topical and systemic (oral) treatments can be used and there is a tendency, in the majority of cases, towards a natural, sudden clearance.

The 'classic adult type' has the best prognosis with up to 80% clearing spontaneously within 3 years.

HOW CAN PITYRIASIS RUBRA PILARIS BE TREATED?

There are no treatment guidelines for PRP. Topical and systemic (oral) treatments, alone or in combination, are commonly used.

Creams or ointments applied to the skin may be all that is required if the PRP is affecting small areas of the body:

- Steroid creams and ointments can improve the redness and scaling, but probably do not alter the duration of the rash.
- Emollients (moisturisers) recommended by the doctor or specialist are a very important part of treatment to help moisten dry skin and restore the barrier function of the skin. The emollient should be applied liberally and regularly.

CAUTION:
This leaflet mentions 'emollients' (moisturisers). Emollients, creams, lotions and ointments contain oils. When emollient products get in contact with dressings, clothing, bed linen or hair, there is a danger that they could catch fire more easily. There is still a risk if the emollient products have dried. People using skincare or haircare products should be very careful near naked flames or lit cigarettes. Wash clothing daily and bedlinen frequently, if they are in contact with emollients. This may not remove the risk completely, even at high temperatures. Caution is still needed. More information may be obtained at *https://www.gov.uk/guidance/safe-use-of-emollient-skin-creams-to-treat-dry-skin-conditions*.

Oral medications may be needed if the PRP is extensive. These medications should only be prescribed by a dermatologist after skin assessment and confirmation of the diagnosis. The most commonly used medications are acitretin and methotrexate.

Phototherapy (light therapy) is also used in some cases, sometimes in combination with acitretin.

Biologic medications are injectable treatments that may be used. These are especially helpful when the treatments above have not been successful in keeping PRP under control. Biologic medications reduce skin inflammation by blocking the activity of chemical messengers known as cytokines in the body. Examples of biologic medications used in PRP include infliximab, ustekinumab, secukinumab and adalimumab.

WHERE CAN I GET MORE INFORMATION ABOUT PITYRIASIS RUBRA PILARIS?

Web links to other relevant resources:

www.pcds.org.uk/clinical-guidance/pityriasis-rubra-pilaris

prpalliance.com

www.prpsurvivalguide.org
www.rareconnect.org/en/community/pityriasis-rubra-pilaris

Please note that the British Association of Dermatologists provides web links to additional resources to help people access a range of information about their skin condition. The views expressed in these external resources may not be shared by the Association or its members.

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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