

# The NORD PRP Report

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## Pityriasis Rubra Pilaris

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## General Discussion

### Summary

Pityriasis rubra pilaris (PRP) is a rare skin disorder that causes inflammation of the skin, thickening of the nails and at times shedding of the hair. 4

The name means scaling (pityriasis), redness (rubra), and involvement of the hair follicles (pilaris). 4

Typically, PRP appears first as a small spot somewhere on the face and then spreads to the back and the rest of the body. 5

It may impact different parts of the body in different ways for unpredictable periods of time. 5

The inflammation may cover the entire body or just parts of the body such as the elbows, knees, palms, and soles. 6

The disease may progress and leave distinct areas of uninvolved skin, the so-called "islands of sparing" or "skip areas". 7

The classic adult type, the most prevalent subcategory, had previously been reported to resolve within three years. The largest case series to date, however, demonstrated that courses are often much longer than this. The pediatric type tends to be a more protracted course. 36

The peak onset years of PRP are in the first, sixth and seventh decades of life. While it most commonly affects adults, there is a significant proportion of pediatric patients affected. The disorder favors no gender.8

There are five types of PRP, which are classified based on age of onset and body areas affected. The sixth type of PRP, or HIV associated, has been more recently described but is still debated. PRP usually occurs at random, but some forms may be hereditary.9

### **Subdivisions of Pityriasis Rubra Pilaris**

- \* type 1 – classical adult onset
- \* type 2 – atypical adult onset
- \* type 3 – classical juvenile onset
- \* type 4 – circumscribed juvenile onset
- \* type 5 – atypical juvenile onset
- \* type 6 – HIV-associated

While the exact prevalence and incidence are unknown, there are an estimated 818 “active” patients in the U.S. and less than 1900 patients in Europe.

PRP is an ultra-rare skin disorder. In fact, it is considered an orphan disease. The rarity of PRP notwithstanding, the signs and symptoms of PRP often mimic those of eczema (31.6 million patients) and psoriasis (8 million patients). 10-11

PRP patients and their caregivers quickly learn that every case of PRP is unique.<sup>4</sup>

Unfortunately, there is no specific or consistently effective therapy for PRP. In fact, there are no treatments approved by the US Food and Drug Administration (FDA) or the European Medicines Agency (EMA) for use in PRP.<sup>12</sup>

Nevertheless, experts tend to use a multimodal approach including topical and systemic therapies to control the symptoms of the disorder. Topical therapies can reduce skin (cutaneous) inflammation and aid with itch (pruritus) and dryness/flaking (xerosis).

Systemic therapy can reduce inflammation and is generally required in the majority of patients with large body surface areas of involvement.<sup>12</sup>

## Introduction

When James Shooter was admitted to St. Bartholomew's Hospital in London, England in 1828, he unwittingly became the world's first recorded patient with what the medical community would eventually call pityriasis rubra pilaris. In 1828, however, Mr. Shooter's skin disorder did not yet have a name. Seven years passed before Claudius Tarral, a French dermatologist, wrote about the case in *Traite theorique et pratique des maladies de la peau* (Treatise on Skin Diseases) in 1835. Tarral saw it as a variant of psoriasis.<sup>13-14</sup>

It would take another 21 years for Marie Guillaume Alphonse Devergie, a dermatologist and forensic doctor at St. Louis Hospital in Paris, to publish the most complete description of PRP. In fact, it is considered to be the "original description" of PRP by the medical community. Devergie's article was published in the *Gazette Hebdomadaire de Medecine et de Chirurgie* in 1856.<sup>14</sup>

While Devergie saw the skin disorder as a combination of "follicular lesions and psoriasis palmaris, pityriasis capillitii and pityriasis rubra", it would take yet another 21 years before another Frenchman and dermatologist by the name of Richaud to recognize PRP as a distinct entity. Richaud published his paper "Etude sur le pityriasis pilaris" in 1877.<sup>15</sup>

When Ernest Besnier presented nine cases in a 120-page article published in 1889 – 12 years after Richaud, 43 years after Devergie, and 54 years after Tarral, he forever fixed the name of the disease as pityriasis rubra pilaris. Besnier too, was a Frenchman and dermatologist. He was also the medical director of the St. Louis Hospital in Paris – the same hospital as Devergie.<sup>16</sup> The name comes from three Latin words: pityriasis (scale-like), rubra (red) and pilaris (hair follicles).<sup>2</sup>

Like many rare disease, the PRP community laments the snail's pace at which PRP research progresses. What should we expect – it took 61 years just to pick the name.

Recently, the Research Group from Thomas Jefferson University's PRP Center of Excellence has compiled the world's largest cohort of PRP patients. With each day, new understanding of the diagnosis and management of PRP are gained; yet much work remains to be accomplished.

## **Signs & Symptoms**

The terms "sign" and "symptom" are not redundant. SIGN is an indication of a medical condition that can be objectively observed by others, including healthcare professionals. In contrast, a SYMPTOM is subjective, information that is shared by the PRP patient with the healthcare provider such as pain, itching, fatigue. The following is a list of signs and symptoms that define PRP.

### **Pre-onset Signs**

Mild signs include dandruff and crusty scalp, as well as limited red patches or scaling of the skin, (e.g., a "dime-sized red spot" on the forehead). The duration varies from patient to patient. At some point the patient determines that intervention of a healthcare professional is warranted, (eg, a red spot has doubled in size in less than two weeks).

### **Progression of Signs**

Depending on the advancement of inflammation, a general practitioner or dermatologist will see signs including pink, red, or orange-red scaly patches on the skin. These patches are usually itchy. Initially, PRP patients may have the scaly patches only on some parts of the body. Patches most often occur on the elbows, knees, hands, feet, and ankles. Skin on the palms and soles may also become red or waxy and thickened with a classic orange hue (palmoplantar keratoderma). The scaly patches may eventually spread over the entire body.<sup>17</sup> Islands of sparing can occur within the salmon-colored patches of inflammation.

Cracks (fissures) may develop within thickened skin which can be painful and make walking and using one's hands difficult. Nails may become thickened, discolored, ridged, cluttered with debris under the free-edge and may even

shed. Hair may shed considerably due to the disorder itself or some treatments used. Heat intolerance, protein loss and fluid imbalance can occur when the rash becomes widespread (erythroderma).<sup>4</sup>

Diagnosis is often delayed by the prolonged course of evolution of the disease and its ability to masquerade as other disorders, particularly eczema and psoriasis. Fortunately, many of the treatments for eczema, psoriasis and PRP are shared.<sup>5</sup>

### **Acute Stage Signs and Symptoms**

A dermatologist will be able to see the signs of a body engulfed by dry, red, and flaking skin, swollen feet and legs, and cracked and bleeding hands and feet. There may be serious issues related to impaired mobility, eyes and vision (caused by tightness and pulling of the eyelids), and dexterity. The PRP patient will see symptoms of PRP from a different perspective, e.g., pain of motion, unrelenting itch (pruritus), and heat intolerance. Pruritus, pain, and sleep disturbance are common with this disorder. The acute stage poses the greatest challenge to body, mind, and spirit and can last anywhere from less than a month or many months longer. This is the time in the PRP journey that PRP patients and caregivers should seek support from patient support groups. It is also a time to address issues of depression.<sup>5</sup>

### **Management Stage**

After the acute stage, the journey of a PRP patient takes on a new focus – mitigating and managing symptoms. Potential irritants are waiting on the roadside, such as joint pain, clogged ears, and disability claims, to name a few. While 90% of the PRP patient population can look forward to full remission within one to five years, the timetable is not certain. Those diagnosed with atypical adult onset and atypical juvenile onset, the chronic versions of PRP, must develop long-term coping skills. For everyone, the daily routine associated with medications, moisturizing, and dealing with the challenges of body, mind and spirit of this skin disorder cannot be ignored.

## Remission & Healing Milestones

The medical community defines remission as the disappearance of signs and symptoms of disease, whether through the use of medication or naturally with time. Recovery is considered the restoration of health or function. The PRP community has adopted a more celebratory approach to disease recovery with recognition of healing milestones. The return of sweat, the first trip out of the house for groceries, and dark hardwood floors that don't need hourly vacuuming are all cause to celebrate. These milestones are symptoms of healing that PRP patients and caregivers feel and signs that everyone else observes.

## Causes

The specific underlying cause of PRP is unknown, although a combination of a genetic predisposition, environmental trigger, and other unknown causes is believed to play key roles. Vitamin A deficiency was once believed to be related to the disorder, however, this theory lacks sufficient evidence and treatment with Vitamin A has been less than effective.<sup>36, 38</sup>

## Affected Populations

Based on conversations within the PRP community, we can say with metaphysical certitude that PRP isn't a punishment for misbehavior or forgetting to put the toilet seat down. There are thousands of perfectly wonderful people who have – or had – PRP. Moreover, there are many very bad people who don't have it. Who then gets the short end of this rare disease stick?

**Prevalence:** In March 2003, Dr. Andrew Griffiths delivered a "Dowling Oration" to members of the British Association of Dermatology assembled in Liverpool, England. Dr. Griffiths reflected on 35 years of diagnosing, treating and researching pityriasis rubra pilaris. He unilaterally guessed the PRP prevalence rate at one in 400,000 persons. While the methodology used by Dr Griffiths is subject to debate, dermatologists worldwide have used his estimate due to a

lack of evidence-based studies of how often diseases occur in different groups of people and why (epidemiology).<sup>1</sup>

**Age:** Pityriasis rubra pilaris is a rare disorder that may develop during childhood or adulthood. Juvenile onset accounts for 45% of the “active” patient population while adult onset accounts for 55%.<sup>6</sup> Although PRP may occur at any age<sup>10</sup>, it most commonly affects those in their first, second, fifth, and sixth decades of life.<sup>24, 2</sup>

**Gender:** PRP appears to occur in males and females in relatively equal numbers. However, in childhood, the male to female ratio is 3:2.<sup>25</sup>

**Race:** Persons of any race may be affected.<sup>2, 7</sup>

**Acquired or Inherited:** PRP is usually sporadic (occurring randomly) but some forms may be hereditary.<sup>6, 2</sup>



## Related Disorders

Symptoms of the following disorders can be similar to those of pityriasis rubra pilaris. Comparisons may be useful for a differential diagnosis:

**Psoriasis:** According to the National Psoriasis Foundation (NPF), "Psoriasis is an immune-mediated disease that causes raised, red, scaly patches to appear on the skin. It typically affects the outside of the elbows, knees or scalp, though it can appear on any location. Some people report that psoriasis is itchy, burns and stings. Psoriasis is associated with other serious health conditions, such as diabetes, heart disease and depression." More information is available at the NPF website.<sup>26</sup>

**Atopic dermatitis (eczema & dermatitis):** There are different types of eczema that collectively affect more than 30 million Americans: atopic dermatitis, contact dermatitis, dyshidrotic eczema, hand eczema, neurodermatitis, nummular eczema and stasis dermatitis. For more information, contact the National Eczema Foundation.<sup>27</sup>

**Allergic reaction:** Most skin allergic reactions are minor, such as an eczema-like rash from poison ivy, or mosquito or other bug bites, or sneezing from hay fever. The type of reaction depends on the person's immune system response, which is sometimes unpredictable. For more information about allergic reactions, go to eMedicineHealth.<sup>28</sup>

**Pityriasis rosea:** Pityriasis rosea (PR) is a benign rash, a common skin disorder observed in otherwise healthy people, most frequently found in children and young adults, that is thought to be caused by a mild viral infection. It usually goes away without treatment after a few months. It can easily mimic types of similar skin eruptions including lichen planus, psoriasis, and pityriasis rubra pilaris. Pityriasis rosea has a very specific and recognizable rash. It does, however, begin with a "herald mark". Follow the link listed in the reference section for additional information at Medscape.<sup>29</sup>

**Fungal infection:** Mycoses are fungal infections of the skin. They are common and generally mild. However, in very sick or otherwise immunosuppressed people, fungi can sometimes cause serious disease.<sup>30</sup>

**Lupus:** Lupus is a chronic autoimmune disease that can damage any part of the body including skin, joints and organs. "Chronic" means that the signs and symptoms tend to last longer than six weeks and often for a lifetime. In lupus, something goes wrong with the immune system. Normally our immune systems produce proteins called "antibodies" which protect the body from invaders. "Autoimmunity" means your immune system cannot tell the difference between these foreign invaders and your body's healthy tissues. As a result, it creates autoantibodies that attack and destroy healthy tissue. When someone has lupus, these autoantibodies cause inflammation, pain, and damage in various parts of the body.<sup>31</sup>

**Cutaneous T-cell lymphoma:** On occasion, the diagnosis of a PRP patient has been changed to cutaneous T-cell lymphoma (CTCL). Chronic inflammation in the skin occurs in PRP and other inflammatory skin diseases like psoriasis and eczema and can even trigger CTCL after many years. The PRP community recommends that appropriate tests which may include skin biopsy and blood work be performed to rule out CTCL. <sup>32</sup>

## Diagnosis

A medical diagnosis is based on information from sources such as findings from a physical examination, an interview with the patient, family or both, a medical history of the patient and family, and clinical findings as reported by laboratory tests and radiologic studies.

A differential diagnosis is a process of weighing the probability of one disease versus that of other diseases. It represents an alternative diagnosis based on the clinical and pathologic features of the patient.

Pityriasis rubra pilaris is not easy to diagnose. In March 2003, English dermatologist Dr. Andrew Griffiths reinforced that fact when he titled his Dowling Oration to the British Association of Dermatologists “Pityriasis Rubra Pilaris – The Scarlet Pimpernel”. Griffiths quoted author Baroness Orczy’s character who says: “They seek him here, they seek him there, that damned elusive Pimpernel.” We agree with Griffiths, “This disease remains an enigma.”<sup>1</sup>

### **The Diagnostic Role of the Dermatologist**

Clinical observation is where it all begins. What symptoms are visible to the dermatologist during the examination? A dime-sized red spot on a forehead can reasonably be diagnosed as seborrheic dermatitis. Similarly, a patient “in full bloom” presenting with 90% of the skin covered with redness and scale, islands of sparing, and other key indicators might be more than enough to awaken a memory of a grand rounds experience years earlier in medical school.

### **The Diagnostic Role of the Dermatopathologist**

The microscope is where the clinical observations are either supported or not. Biopsies sent to a dermatopathologist are often used to “rule out” specific skin maladies or causes. The PRP community has learned from shared experiences – albeit anecdotal – that when the dermatologist suspects PRP and instructs the dermatopathologist to “consider PRP,” that the findings support the clinical observations. Special tests can be performed on the skin biopsy to rule out the

possibility of cutaneous lymphoma which can mimic PRP at the microscopic level (histology).

Nevertheless, the signs of PRP under the microscope are neither sensitive (the ability to pick up a diagnosis when using a test) nor specific (not shared by many other disorders as well) to PRP. As such, the diagnosis of PRP remains largely a clinicopathologic correlation, that is to say piecing together the clinical and pathologic features to make the “best fit” of a diagnosis based on the patient’s presentation.

### **Differential Diagnosis**

A differential diagnosis is the method by which a physician determines what disease process has caused a patient’s symptoms. The physician considers all relevant potential causes of the symptoms and then eliminates alternative causes based on a physical examination, clinical tests and a thorough case history.<sup>33</sup>

The International Center for Toxicology and Medicine states, “A differential diagnosis is a quest for a diagnosis. What is wrong with the patient internally? It is not, inherently, a search for the ultimate cause (critical to liability) of that disease process or disorder.”<sup>34</sup> It is “the process of weighing the probability of one disease versus that of other diseases possibly accounting for a patient’s illness.”<sup>35</sup>

## **Standard Therapies**

### **Treatment**

Treatment of pityriasis rubra pilaris is mainly anecdotal, based on case reports and case series, a feature shared by many disorders in dermatology due to their rarity. The fleeting nature of the large proportion of PRP symptoms also makes it difficult to study in standardized, long-term therapeutic studies. As controlled trials are lacking, the effectiveness and safety of treatments is

unclear. Thus, there is low quality evidence supporting treatment strategies of PRP. Currently there are no treatments approved by the US Food and Drug Administration or the European Medicines Agency for use in PRP.<sup>9</sup>

PRP tends to follow a natural waxing and waning course, with episodes in which there is periodic worsening (exacerbation) or cessation (remission) of symptoms. Thus, according to many researchers, it may be difficult to evaluate the effectiveness of particular therapies.<sup>9</sup>

The value of treatment is difficult to assess, as the clinical course is so variable for each of the different types of PRP. Patients with classical adult onset PRP, for example, may present with intense and widespread reddening of the entire skin surface (erythroderma). Hospital admission for skin care, fluid replacement and other supportive care may be warranted.<sup>6</sup>

From the patient perspective, there are two major objectives in the treatment of pityriasis rubra pilaris:

- \* relieving symptoms as they present
- \* achieving long-term remission, if possible. The mantra heard within the PRP community is simple but deafening: What works for one doesn't work for all.

### **Treatment Options**

Management of PRP often involves systemic and topical therapies combined. Topical therapies can help with the symptoms and may be enough for people with mild PRP. Topical treatments are usually combined with systemic therapy for PRP that affects a large part of the body. Most PRP patients need systemic therapy to control the condition.<sup>9</sup>

Treatment options will vary based on age, geography, and cost to the patient. Moreover, laboratory tests are important to monitor the effects of medications on the body – especially the liver – and to manage and monitor the side effects of drugs.

Some of the medications used to treat PRP can harm a developing fetus and are not recommended for use right before or during pregnancy.<sup>9</sup>

People seeking information about specific treatment options for themselves or family members should speak with their health care provider.<sup>9</sup>

## **RETINOIDS**

Oral retinoids are derivatives of vitamin A that slow the growth and shedding of skin cells. Treatment options include acitretin / Soriatane<sup>®</sup> and isotretinoin / Accutane<sup>®</sup>, though researchers at the PRP Center of Excellence prefer acitretin over isotretinoin and etretinate. Oral retinoids (synthetic vitamin A derivatives) are the first line systemic treatment for PRP.<sup>9</sup> A scientific survey of patients with PRP performed by researchers at Thomas Jefferson University published that oral retinoids were helpful in approximately 60% of patients with PRP.<sup>36</sup>

## **IMMUNOSUPPRESSANTS**

Immunosuppressants slow down the body's immune system. These can be used in combination when oral retinoids are ineffective. Treatment options (oral and injection) include methotrexate, cyclosporine, TNF-alpha inhibitors, IL-12/23 inhibitors, among others. Methotrexate was reported to be helpful in approximately 50% of patients with PRP.<sup>36</sup>

## **BIOLOGICALS**

Biologicals are also immunosuppressants. Biologicals are injectable or intravenous (IV) medications that target various pathways of inflammation in the body. With generally fewer side effects, biologicals are targeted to reduce inflammation. Treatment options include adalimumab / Humira<sup>®</sup>, etanercept / Enbrel<sup>®</sup>, infliximab / Remicade<sup>®</sup>, ustekinumab / Stelara<sup>®</sup>, secukinumab / Cosentyx<sup>®</sup>, ixekizumab / Taltz<sup>®</sup>, brodalumab / Siliq<sup>®</sup>, guselkumab / Tremfya<sup>®</sup>, and apremilast / Otezla<sup>®</sup>. These biologic medications are FDA-approved for psoriasis but improvement or remission for some patients with PRP have been published in the medical literature.

## **OTHER THERAPIES**

- Topical creams that contain urea or ammonium lactate decrease scaling and flaking of the skin. Topical corticosteroid creams decrease skin inflammation. These are applied directly to the skin.<sup>8</sup>
- Oral vitamin A. This may be helpful in some people, but only in very high doses that may cause toxicity. Retinoids (synthetic derivatives of vitamin A) are safer and more effective and more commonly used than high-dose vitamin A.<sup>8</sup>
- Traditional Chinese Medicine and other Alternative Medicines with varying degrees of success.

## **REFERRALS**

Depending on the severity, duration and array of signs and symptoms, PRP patients seek the expertise of specialized healthcare professionals:

- \* Ophthalmologist: ectropion (eyelids are turned outward) and impaired vision
- \* Podiatrist: impaired mobility
- \* Otorhinolaryngologist (ENT specialist): impaired hearing, removal of ear wax (cerumen) from ear canal.
- \* Hepatologist: monitor impact of PRP treatment on the liver.
- \* Psychiatrists/Clinical Psychologist: depression and mental wellness
- \* PRP patient support resources: (see Resources) further information on patient care and tools see <http://prpalliance.com/survival-guide/>.

## Investigational Therapies

The Department of Dermatology and Cutaneous Biology located at Sidney Kimmel Medical College at Thomas Jefferson University (Philadelphia, PA) began genetic research in October 2012 studying CARD14 gene mutations in relation to PRP. Dr Jouni Uitto, Professor and Chair of Dermatology, was part of an earlier research effort in Tel Aviv that did not find a causal relationship between these mutations and PRP, but did discover a “genetic basis” for PRP. In July 2014, the genetic analysis research effort was expanded to include a clinical analysis component. The PRP Alliance helped recruit a cohort of over 100 PRP patients and Thomas Jefferson University has the full cooperation and support of the PRP community. Thomas Jefferson University is also seeking separate funding to build a PRP Patient Registry. No start date has been established. For information about PRP research, contact: <http://prpalliance.com/survival-guide/> .37

- \* Information on current clinical trials is posted on [www.clinicaltrials.gov](http://www.clinicaltrials.gov). All studies receiving U.S. government funding, and some supported by private industry, are posted on this government web site.
- \* For information about clinical trials being conducted at the NIH Clinical Center in Bethesda, MD, contact the NIH Patient Recruitment Office: Tollfree: (800) 411-1222; TTY: (866) 411-1010; Email: [prpl@cc.nih.gov](mailto:prpl@cc.nih.gov)
- \* Some current clinical trials also are posted on the following page on the NORD website: <https://rarediseases.org/for-patients-and-families/information-resources/news-patient-recruitment/>
- \* For information about clinical trials sponsored by private sources, contact: [www.centerwatch.com](http://www.centerwatch.com)
- \* For information about clinical trials conducted in Europe, contact:  
\* <https://www.clinicaltrialsregister.eu/>



## **NORD Member Organizations**

### EURORDIS- Rare Diseases Europe

Plateforme Maladies Rares

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Paris, 75014 France

Phone: 33156535210

Email: [eurordis@eurordis.org](mailto:eurordis@eurordis.org)

Website: <http://www.eurordis.org>

### Foundation for Ichthyosis & Related Skin Types

2616 N Broad Street

Colmar, PA 18915

Phone: (215) 997-9400

Toll-free: (800) 545-3286

Email: [info@firstskinfoundation.org](mailto:info@firstskinfoundation.org)

Website: <http://www.firstskinfoundation.org>

Other Organizations

### Genetic and Rare Diseases (GARD) Information Center

PO Box 8126

Gaithersburg, MD 20898-8126

Phone: (301) 251-4925

Toll-free: (888) 205-2311

Website: <http://rarediseases.info.nih.gov/GARD/>

### PRP Alliance, Inc.

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Website: <http://www.prpAlliance.com>

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