

Demographics, Therapeutic Efficacy, and Comorbid Burden in Pityriasis Rubra Pilaris: Insights from a Retrospective Cohort

Alexandre Couto e Silva

Oregon Health and Science University

Heejo Keum

University of Texas Southwestern Medical Center

Amirali Veshagh

Oregon Health and Science University

Teri M. Greiling

Greiling@ohsu.edu

Oregon Health and Science University

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Abstract

Pityriasis rubra pilaris (PRP) is a rare, severe inflammatory skin disorder characterized by follicular papules, papulosquamous red-orange plaques, and palmoplantar keratoderma. Despite its clinical significance, epidemiological data on PRP remains scarce, there is no FDA-approved therapy, and comorbid conditions are unclear. This study analyzed a cohort of 74 patients with PRP at Oregon Health & Science University using a retrospective chart review from January 2013 to December 2023. Juvenile onset PRP (n = 12) represented 1 in 2,863 unique patients to dermatology, with a mean (SD) age at diagnosis of 9.0 (4.6) years. Adult onset PRP (n = 62) represented 1 in 1,798 unique patients seen by dermatology, with a mean (SD) age at diagnosis of 56.8 (15.2) years. Of the total cohort, 45/74 (60.8%) were male, and 29/74 (39.2%) were female sex (Fig. 1). Within the juvenile-onset group, 6/12 (50%) were male and 6/12 (50%) were female sex. Within the adult-onset group, 39/62 (62.9%) were male and 23/62 (37.1%) were female sex. The majority of patients self-identified as White. Half of juvenile-onset patients and over 90% of adult onset patients were treated with systemic therapy. Biologic therapies, particularly IL-17 inhibitors and IL-23 inhibitors demonstrated the highest treatment efficacy with good responses in adult patients of 57.1% and 64.7% respectively. The oral agents methotrexate and acitretin were commonly prescribed to adult patients but had lower efficacy, with good response rates of 26.3% and 29.2% respectively. Comorbidities included common conditions such as hypertension, hyperlipidemia, type 2 diabetes, obesity, anxiety, depression, and sleep disorders. It remains unclear whether these are more common in PRP or simply reflect the demographics of the cohort. This study provides insight into the prevalence, epidemiology, treatment outcomes, and comorbidities of PRP.

Introduction

Pityriasis rubra pilaris (PRP) is a rare and severe inflammatory skin disorder characterized by palmoplantar keratoderma and follicular papules that coalesce into well-demarcated reddish-orange plaques with nonadherent scale. Symptoms such as intense itch, skin tightness, burning, stinging, and pain significantly impair quality of life, often contributing to psychological distress, including depression and suicidal ideation [1–3]. Therapeutic approaches remain limited and challenging, although advancements in biologics targeting the IL-17/IL-23 pathway show promise in open label trials [4–6].

Despite its clinical significance, epidemiological data on PRP remains scarce. Reported prevalence rates vary widely, from 1 in 500 new patients in a Kenya dermatology practice [7], 1 in 5,000 new patients seen by British dermatologists [8], to 1 in 50,000 in Indian dermatology practices [9]. PRP prevalence in the United States unknown; this statistic is cited as 1 in 3,500–5,000 patients in multiple review articles without a primary source listed. PRP can occur at any age, with a bimodal incidence peaking in early childhood (ages 6–7) and the fifth decade of life [10]. Most studies indicate equal prevalence among males and females, though some suggest a slight male predominance [10]. While PRP is known to affect sleep and mental health [1–3], it is unclear whether other medical comorbidities are more common in PRP [1, 11].

This study aims to add additional insights into PRP demographics, treatment efficacy, and comorbidities by analyzing a retrospective cohort of 74 adult and juvenile patients seen over 10 years at Oregon Health & Science University (OHSU).

Methods

We conducted a retrospective cohort study using the Epic electronic health record (EHR) to identify patients at Oregon Health & Science University (OHSU) from January 2013 to December 2023 with a presumed diagnosis of Pityriasis Rubra Pilaris (PRP) as indicated by the ICD10 diagnostic code L44.0. 99 patient records were evaluated by a thorough chart review of clinical notes, pathology reports, and photographic documentation when available, which resulted in 74 confirmed cases of PRP. Charts were reviewed for demographic data, medications used with documentation of effectiveness (categorized into failed response, partial response, and good response), and comorbid conditions using all extracted ICD10 codes associated with 4 or more patients. This study was approved by the OHSU Institutional Review Board (#00027329). Patient consent was waived due to the retrospective nature of the study and the use of de-identified data.

Results

Demographic Characteristics

The cohort included 74 patients with Pityriasis Rubra Pilaris (PRP), which represented 1 in 1,967 unique patients seen by OHSU dermatology over the 10-year time period. 12 patients were juvenile-onset (diagnosed before age 18) representing 1 in 2,863 unique patients to dermatology, with a mean (SD) age at diagnosis of 9.0 (4.6) years (range 2–16 years). 62 patients were adult-onset, representing 1 in 1,798 unique patients seen by dermatology, with a mean (SD) age at diagnosis of 56.8 (15.2) years (range 18–77 years). Of the total cohort, 45/74 (60.8%) were male, and 29/74 (39.2%) were female sex (Fig. 1). Within the juvenile-onset group, 6/12 (50%) were male and 6/12 (50%) were female sex. Within the adult-onset group, 39/62 (62.9%) were male and 23/62 (37.1%) were female sex.

The majority of patients self-identified as White (66/74, 89.2%). Other racial and ethnic groups represented in the cohort included 1 Asian (1.4%), 1 Native American (1.4%), 4 Hispanic (5.4%), 1 African American (1.4%), and 1 patient (1.4%) for whom no information was available. Of those with a documented PRP subtype, 43/56 (76.8%) were type I, 2/56 (3.6%) were type II, 2/56 (3.6%) were type III, 8/56 (14.3%) were type IV, and 1/56 (1.8%) were type V. There were no cases of HIV-associated (type VI) PRP.

Treatment Outcomes

Of the twelve pediatric patients, 6 were treated without systemic therapy and 6 were treated with systemic therapy, which included n = 1 each of acitretin (good response), isotretinoin (failed response),

methotrexate (failed response), ustekinumab (good response), ixekizumab (good response), and dupilumab (failed response).

Of the adults for whom treatment information was available, 2/61 (3.3%) were treated with topicals only, 3/61 (4.9%) with topicals and phototherapy, 56/61 (91.8%) were treated with systemic therapy, and 39/56 (69.6%) were treated with more than one systemic therapy. Systemic therapies included oral retinoids in 24/56 (42.9%), methotrexate in 19/56 (33.9%), IL17 inhibitors in 21/56 (37.5%), IL12/23 or IL23 inhibitors 17/56 (30.4%), and TNF inhibitors in 11/56 (19.6%).

When grouped, IL17 inhibitors achieved a 12/21 (57.1%) good response, 5/21 (23.8%) partial response, and 4/21 (19.0%) failure (Fig. 2). Grouped IL12/23 or IL23 inhibitors achieved a 11/17 (64.7%) good response, 4/17 (23.5%) partial response, and 2/17 (11.8%) failure (Fig. 2). TNF-alpha inhibitors had a 5/11 (45.5%) good response and 6/11 (54.5%) failure (Fig. 2).

Among oral systemic agents, methotrexate and oral retinoids showed similar efficacy, with good responses in 5/19 (26.3%) and 7/24 (29.2%) respectively; partial response in 5/19 (26.3%) and 7/24 (29.2%) respectively; and failure in 9/19 (47.4%) and 10/24 (41.7%) respective (Fig. 2). Cyclosporine had the lowest good response rate of 1/7 (14.3%) with the remaining patients having no response.

Associated Comorbidities

Comorbidities present in more than one juvenile patient with PRP included anxiety (F41.9) in 3/12 (25%), hypovitaminosis D (E55.9) in 3/12 (25%), feeding difficulties in a newborn in 2/12 (17%), and obstructive sleep apnea in 2/12 (17%) of patients.

Adults with PRP exhibited a range of common comorbid conditions, with cardiovascular, metabolic, and psychiatric disorders most prevalent. Essential hypertension (I10) in 19/62 (30.6%) and hyperlipidemia (E78) in 18/62 (29.0%) were the most common comorbidities. Type 2 diabetes mellitus (E11) in 12/62 (19.4%) and obesity (E66) in 10/62 (16.1%) were also prevalent. Psychiatric conditions, including anxiety disorders (F41) in 11/62 (17.7%), major depressive disorder (F33) 7/74 (11.3%), and adjustment disorders (F43) in 6/74 (9.7%), were frequently reported. Sleep disorders (G47) and arthralgias each affected 10/74 (16.1%) of adults. A full list of comorbidities present in more than 4 adult patients with PRP is in Table 1, as well as the mean age of patients with each comorbidity.

Table 1

Comorbid Conditions in Adult Patients with Pityriasis Rubra Pilaris (PRP). Comorbid conditions were identified using ICD10 codes documented in the electronic health records of the study cohort (n = 62). Conditions associated with 4 or more patients were included in the table. Age represents the mean age of the patient with PRP who had each comorbid diagnosis.

Category	ICD10	Condition	Number	Percent	Mean age (yr)
Cardiovascular	I10	Essential hypertension	19	30.6%	69.7
	I50	Congestive heart failure	4	6.5%	80.3
	I48	Atrial fibrillation	4	6.5%	77.3
	I25	Coronary artery disease	4	6.5%	74.5
Metabolic/Endocrine	E78	Hyperlipidemia	18	29.0%	70.2
	E11	Type 2 diabetes mellitus	12	19.4%	67.3
	E66	Obesity	10	16.1%	60.2
	E03	Hypothyroidism	9	14.5%	63.8
	R73	Hyperglycemia/prediabetes	7	11.3%	59.1
	E55	Vitamin D deficiency	6	9.7%	62.2
	E88	Other metabolic disorders	5	8.1%	67.6
Psychiatric/Neurological	F41	Anxiety disorders	11	17.7%	58.5
	G47	Sleep disorders	10	16.1%	63.3
	F43	Adjustment disorders	6	9.7%	58.5
	F32/F33	Depressive Disorders	4	6.5%	52.7
Musculoskeletal/Pain	M25	Joint pain	10	16.1%	68.2
	M54	Back pain	9	14.5%	69.4
	M17/M19	Osteoarthritis of the knee/joints	8	12.9%	73.0
	M79	Soft tissue pain	7	11.3%	71.9
	M75	Shoulder and rotator cuff disorders	4	6.5%	66.0
Dermatological	L40	Psoriasis	10	16.1%	65.8
	L30	Dermatitis/eczema	4	6.5%	76.3
	L53	Erythroderma	4	6.5%	65.5

Category	ICD10	Condition	Number	Percent	Mean age (yr)
Allergy	J30	Allergic rhinitis	9	14.5%	53.9
Gastrointestinal	K21	Gastroesophageal reflux disease	7	11.3%	62.4
	K58	Irritable bowel syndrome	4	6.5%	49.5
	K76	Non-alcoholic fatty liver disease	4	6.5%	67.3
Genitourinary	N39	Urinary tract infections	6	9.7%	67.3
	N40	Benign prostatic hyperplasia	6	9.7%	70.3
	N52	Erectile dysfunction	5	8.1%	66.0
	N18	Chronic kidney disease	5	8.1%	82.4
	N17	Acute kidney injury	4	6.5%	75.0
Other Conditions	Z85	History of malignancy	7	11.3%	76.9
	R53	Fatigue/malaise	6	9.7%	78.6
	D64	Anemia	4	6.5%	74.3
	H61	Cerumen impaction	4	6.5%	74.3
	N95	Menopausal disorders	4	6.5%	66.3
	C44	Basal cell carcinoma	4	6.5%	74.5
	H90	Sensorineural hearing loss	4	6.5%	68.8
	H25	Cataracts	4	6.5%	78.5

Discussion

While the incidence and prevalence of PRP remain uncertain, we report the first data in the United States showing that the diagnosis of PRP represented approximately 1 in 2,000 patients seen by OHSU Dermatology, with the diagnosis being slightly more common in adults (1 in 1,798 adults vs. 1 in 2,863 juvenile patients). This incidence rate is somewhat similar to prior reports from Great Britain (1:5,000 patients) but quite different from prior reports from Kenya (1 in 500) and India (1 in 50,000). It remains uncertain whether the prevalence of PRP varies by country or whether this reflects publication or diagnostic bias. This cohort was largely white and was reflective of the racial and ethnic distribution of the population served by OHSU. The age and sex distribution in the OHSU cohort was similar to other reports in the United States [12]. Male and female sex were equal in the pediatric population, with male

sex have a slight predominance in adults with PRP. PRP was diagnosed across all ages with slight peaks in the first and sixth decade.

Treatment outcomes, while non-standardized, were similar in this cohort to published systematic reviews [13, 14]. Half of children and over 90% of adults required systemic therapy. Positive responses to oral retinoids and methotrexate were similar and relatively low (< 30% of patients), whereas psoriasis biologics such as IL17 and IL23 inhibitors had higher rates of positive responses, similar to that seen in systematic open label trials [4–6].

Comorbidities in this adult cohort with PRP included common medical diagnoses such as hypertension, hyperlipidemia, type II diabetes mellitus, and obesity. It remains unclear whether this reflects the demographics of the cohort, or whether these conditions are more common in individuals with PRP, as they are in psoriasis [15]. PRP and psoriasis share dysregulation in systemic inflammatory pathways like the IL17/IL23 axis [16], which may contribute to the higher rates of metabolic dysfunction. Psychiatric comorbidities such as anxiety, depression, adjustment disorder, and sleep disorders were common in the PRP cohort, reflecting the significant psychological burden of PRP [1–3].

This study has several limitations, including its retrospective design, reliance on EHR data, and small sample sizes for some therapies. These factors may limit the generalizability of our findings.

Nonetheless, this retrospective cohort of 10-years of PRP patients seen at OHSU dermatology appears to match the broader age, sex, and treatment response demographics previously reported in PRP, and provides initial data on better understanding the prevalence and impact of PRP in the United States.

Statements and Declarations

None of the authors have competing interests relevant to this study.

Author Contribution

AC and TG designed the work. AC and AV acquired the data. AC and HK analyzed the data. AC and TG interpreted the data and drafted the manuscript. All authors revised it critically for important intellectual content; approved the version to be published; and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Data Availability

All data supporting the findings of this study are available within the paper. Any additional questions can be directed to the corresponding author.

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Figures

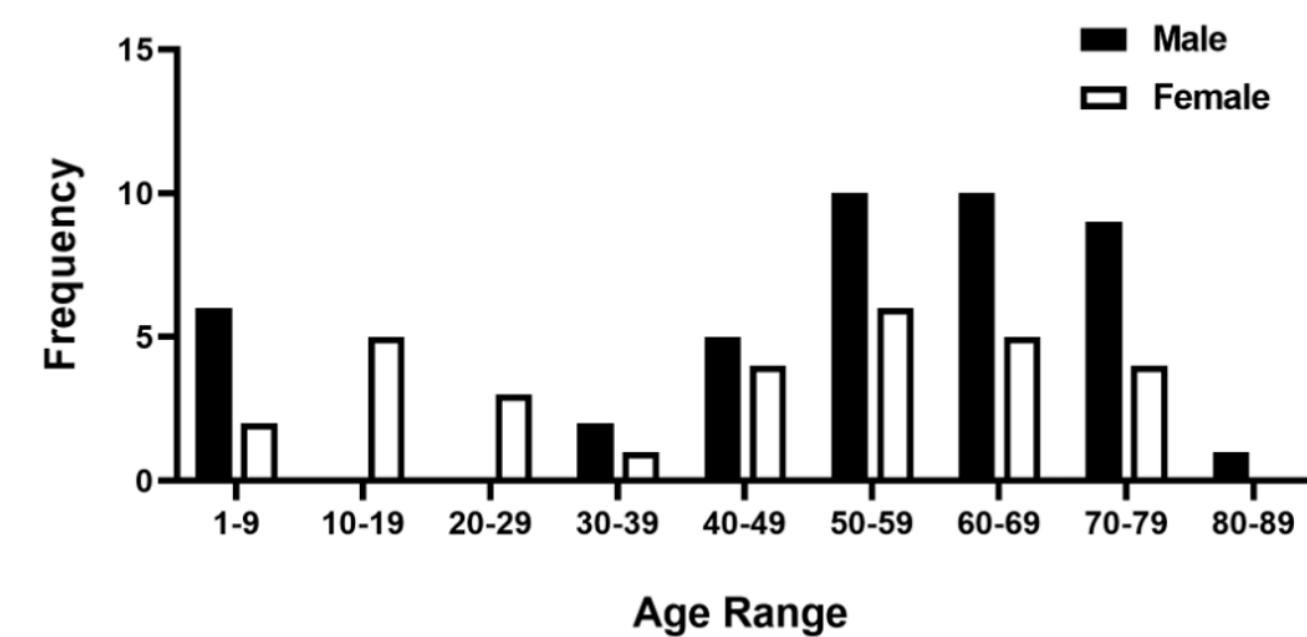


Figure 1

Age and Sex Profile of PRP Cohort. Histogram depicting the age and sex distribution of patients at the time of diagnosis with PRP. Age ranges are represented on the x-axis and frequency on the y-axis.

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Figure 2

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