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Validity, reliability, and responsiveness of the individual Pityriasis Rubra Pilaris Area and Severity Index (iPRPASI)

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Keywords

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Abstract

Pityriasis rubra pilaris (PRP) is a rare inflammatory cutaneous condition characterized by widespread erythematous scaly plaques and palmoplantar keratoderma.[1] PRP skin involvement can cause functional impairments and emotional distress[2] leading to decreased quality of life. There is no clinical tool explicitly developed for PRP. Clinical trials[3,4] that have shown the efficacy of IL-17A inhibitor therapy have used the Psoriasis Assessment and Severity Index (PASI)[5] to measure the clinician-reported outcome of disease severity and the Dermatology Life Quality Index (DLQI)[6] to measure the patient-reported health-related quality of life. Despite PRP and psoriasis sharing similar clinical features, the PASI does not capture unique aspects of PRP. The Individual Pityriasis Rubra Pilaris Area and Severity Index (iPRPASI) was developed as a patient-reported outcomes measure to capture the more nuanced features of PRP, such as the high prevalence of palmoplantar keratoderma. An assessment of the validity, reliability, and responsiveness of the iPRPASI is conducted in this study. The correlation between the iPRPASI and the PASI, as well as the iPRPASI and the DLQI, was examined to establish the validity of the iPRPASI. Significant correlations were found between iPRPASI and PASI ($r2 = 0.902$, $p < 0.001$), as well as between iPRPASI and DLQI ($r2 = 0.810$, $p < 0.001$). The iPRPASI demonstrated reliability through a high Cronbach alpha value of 0.931. In measuring responsiveness to changes in disease severity over time, the iPRPASI showed a higher correlation with changes in PASI scores ($r2 = 0.880$, p 0.001) than changes in DLQI scores ($r2 = 0.609$, p 0.001). The iPRPASI is comparable to the currently used PASI in terms of validity, reliability, and responsiveness. Future uses of the iPRPASI could include clinical trials that include remote visits, and clinical practice to measure response to therapy by clinicians who lack expertise in this rare disease.

References

1.Griffiths WAD. Original article pityriasis rubra pilaris-an historical approach. 2 Clinical features. Clin Exp Dermatol. 1976;1(1):37–50.

2.Velasco, R., et al. (2023). "The comprehensive impact of pityriasis rubra pilaris on physical symptoms, mental health, and activities of daily living: A mixed methods study." JEADV Clinical Practice 2.

3.Haynes D, Strunck JL, Topham CA, et al. Evaluation of Ixekizumab Treatment for Patients With Pityriasis Rubra Pilaris: A Single-Arm Trial. JAMA Dermatol.

4.Boudreaux BW, Pincelli TP, Bhullar PK, Patel MH, Brumfiel CM, Li X, Heckman MG, Pittelkow MR, Mangold AR, Sluzevich JC. Secukinumab for the treatment of adult-onset pityriasis rubra pilaris: a single-arm clinical trial with transcriptomic analysis. Br J Dermatol. 2022 Nov;187(5):650-658. doi: 10.1111/bjd.21708. Epub 2022 Jul 15. PMID: 35701384.

5.Fredriksson, T. and U. Pettersson, Severe psoriasis--oral therapy with a new retinoid. Dermatologica, 1978. 157(4): p. 238-244.

6.FinlayAY,KhanGK.DermatologyLifeQuality Index (DLQI)—a simple practical measure for routine clinical use. Clin Exp Dermatol. 1994;19(3): 210-216. doi:10.1111/j.1365-2230.1994.tb01167.x