

Validation of the Generalized Pustular Psoriasis Physician Global Assessment (GPPGA) scale for assessing severity of GPP

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Intra- and inter-rater reliability analyses demonstrate that the GPPGA is a reproducible and reliable method for physician assessment of GPP severity, supporting its use as an endpoint in clinical trials

PURPOSE

To validate the accuracy and consistency of GPPGA scoring for measuring the severity of skin symptoms in patients with GPP, by assessing its reproducibility over time and reliability between different assessors.

INTRODUCTION

- GPP is a rare and potentially life-threatening skin disease characterized by the widespread eruption of sterile, neutrophilic pustules that can occur with or without systemic inflammation^{1,2}
- Currently, there are no agreed or validated measurements for assessing the severity of GPP. As a result, many different methodologies are used in clinical trials and clinical practice, which presents a challenge when comparing the efficacy of potential treatments
- Developed as a novel GPP-specific clinical endpoint, the GPPGA is a composite score adapted from the well-understood Physician Global Assessment widely used by dermatologists³

CONCLUSIONS

- The GPPGA is a reliable and robust clinical endpoint for assessing disease severity in patients with GPP
- International GPP-experienced dermatologists and expert raters were able to consistently score GPP severity, and the scores were reproducible by the same assessor over time
- The findings of this study support the use of the GPPGA as a suitable endpoint for future clinical trials in patients with GPP, and validate its use as a standard tool for the assessment of disease severity in clinical practice

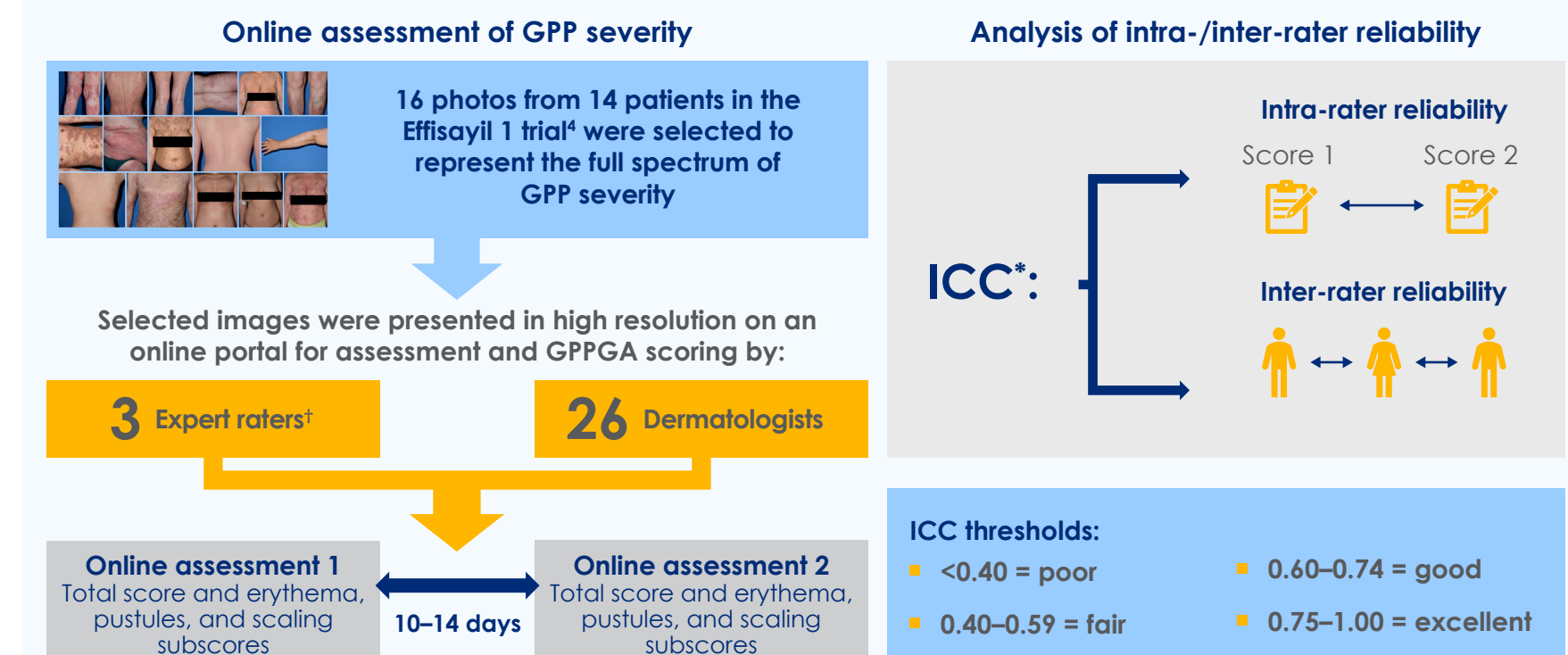
METHODS

GPPGA severity scoring

Score	Erythema	Pustules	Scaling
0 (clear)	Normal or post-inflammatory hyperpigmentation	No visible pustules	No scaling or crusting
1 (almost clear)	Faint, diffuse pink, or slight red	Low-density occasional small discrete pustules (noncoalescent)	Superficial focal scaling or crusting restricted to periphery of lesions
2 (mild)	Light red	Moderate-density grouped discrete small pustules (noncoalescent)	Predominantly fine scaling or crusting
3 (moderate)	Bright red	High-density pustules with some coalescence	Moderate scaling or crusting covering most or all lesions
4 (severe)	Deep fiery red	Very-high-density pustules with pustular lakes	Severe scaling or crusting covering most or all lesions

GPPGA is a 5-grade numerical scale ranging from 0 (clear) to 4 (severe) that is used to assess the severity of erythema, pustules, and scaling. Composite mean score = (erythema + pustules + scaling)/3; total GPPGA score given is 0 if mean is 0 for all three components, 1 if 0<mean<1.5, 2 if 1.5<mean<2.5, 3 if 2.5<mean<3.5, 4 if mean ≥3.5.

Study methodology



*Intra-rater reliability was assessed by ICC using a two-way mixed-effects model for absolute agreement. Inter-rater reliability was assessed by ICC using a two-way random-effects model for absolute agreement; ⁵expert raters were three GPP clinical leaders who were involved in the original development of the GPPGA severity score.

Abbreviations

CI, confidence interval; GPP, generalized pustular psoriasis; GPPGA, Generalized Pustular Psoriasis Physician Global Assessment; ICC, intra-class correlation coefficient; SD, standard deviation.

References

1. Navarini AA, et al. *J Eur Acad Dermatol Venereol* 2017;31:1792–1799; 2. Fujita H, et al. *J Dermatol* 2018;45:1235–1270; 3. Choon SE, et al. *BMJ Open* 2021;11:e043666; 4. Bachelez H, et al. *New Engl J Med* 2021;385:2431–2440.

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RESULTS

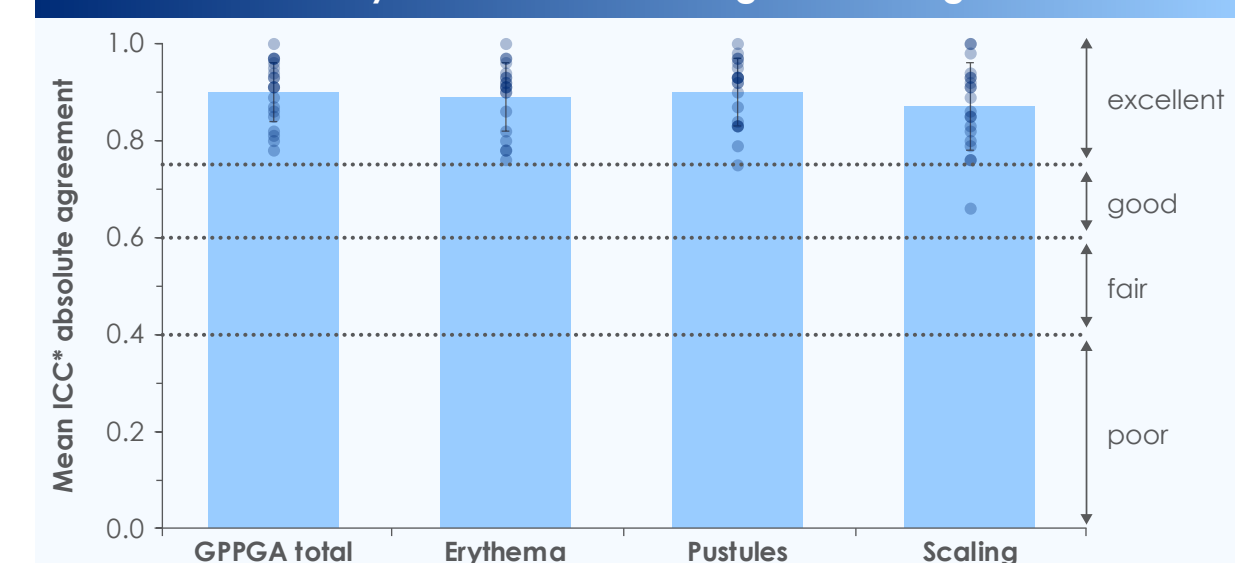
Intra-rater reliability of the GPPGA among expert raters

	Online assessment 1, mean (SD)	Online assessment 2, mean (SD)	ICC* (2,1) absolute agreement [95% CI]
Expert rater 1			
GPPGA total score	2.3 (1.1)	2.1 (1.1)	0.90 [0.75–0.96]
Erythema	2.4 (1.1)	2.2 (0.9)	0.86 [0.69–0.94]
Pustules	2.6 (1.2)	2.1 (1.3)	0.81 [0.47–0.93]
Scaling	1.9 (1.2)	1.8 (1.3)	0.88 [0.74–0.95]
Expert rater 2			
GPPGA total score	1.8 (1.2)	1.9 (1.3)	0.94 [0.85–0.97]
Erythema	1.9 (1.3)	1.9 (1.3)	1.00 [–]
Pustules	1.6 (1.5)	2.1 (1.6)	0.86 [0.66–0.94]
Scaling	1.9 (1.3)	1.9 (1.3)	0.98 [0.96–0.99]
Expert rater 3			
GPPGA total score	2.4 (1.1)	2.2 (1.2)	0.93 [0.83–0.97]
Erythema	2.6 (1.3)	2.3 (1.2)	0.88 [0.64–0.96]
Pustules	2.5 (1.6)	2.4 (1.5)	0.94 [0.86–0.97]
Scaling	1.9 (1.2)	1.9 (1.0)	0.85 [0.67–0.93]

All three expert raters completed both assessments. *ICC using a two-way mixed-effects model for absolute agreement. ICC <0.40 = poor; 0.40–0.59 = fair; 0.60–0.74 = good; 0.75–1.00 = excellent.

Intra-rater reliability for the three expert raters was 'excellent' for the GPPGA total score and for all three subscores (N=16 photos at each online assessment; ICC, 0.81–1.00)

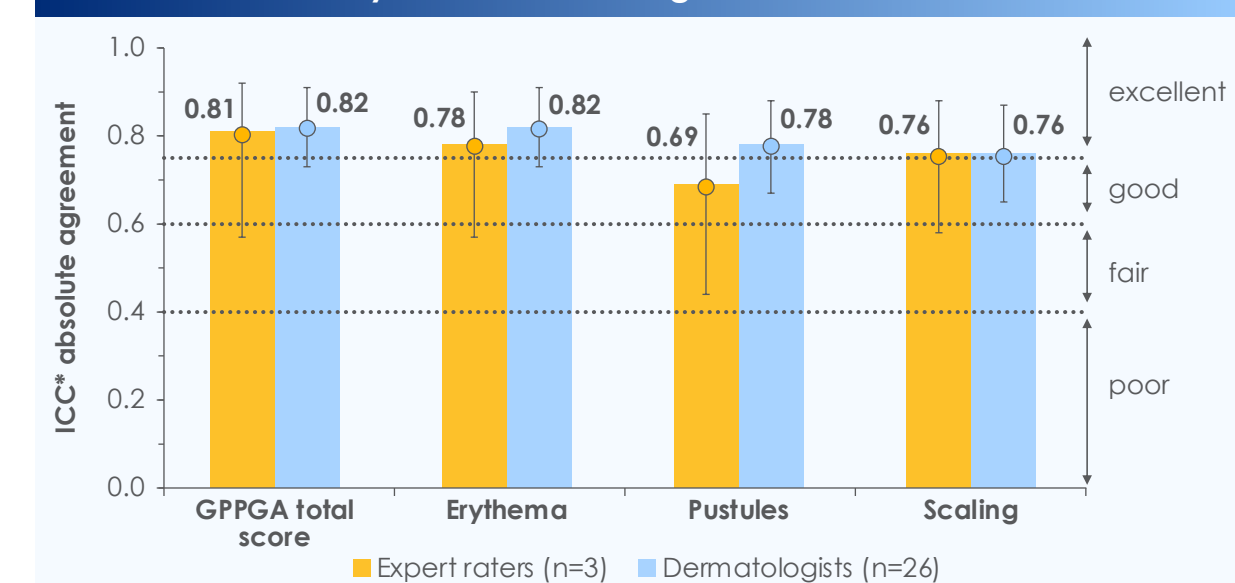
Intra-rater reliability of the GPPGA among dermatologists



20/26 dermatologists completed both assessments; 6/26 dermatologists completed one assessment. Error bars show the SD. *ICC using a two-way mixed-effects model for absolute agreement. ICC <0.40 = poor; 0.40–0.59 = fair; 0.60–0.74 = good; 0.75–1.00 = excellent.

Intra-rater reliability for dermatologists who completed both online assessments (n=20) was 'excellent' for the GPPGA total score and for all three subscores (N=16 photos at each online assessment; mean ICC, 0.87–0.90). All individual ICC values were within the 'excellent' range except for one dermatologist who recorded a scaling ICC of 0.66 (95% CI 0.29–0.85)

Inter-rater reliability for all raters using the first online assessment



Error bars show the 95% CI. *ICC using a two-way random-effects model for absolute agreement. ICC <0.40 = poor; 0.40–0.59 = fair; 0.60–0.74 = good; 0.75–1.00 = excellent.

Absolute agreement was 'excellent' for all items among the 26 dermatologists (ICC, 0.76–0.82). For the three expert raters, absolute agreement was 'excellent' for GPPGA total score and erythema and scaling subscores (ICC, 0.76–0.81); agreement for pustules subscore was 'good' (ICC, 0.69)



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