

# Flare characteristics of generalized pustular psoriasis in North America: Survey results from dermatologists in the Corrona Psoriasis Registry

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## Introduction

- Generalized pustular psoriasis (GPP) is a rare and potentially life-threatening systemic and chronic skin disease, characterised by relapsing and remitting flares consisting of a disseminated skin rash with sterile pustules<sup>1-3</sup>
- The severity and frequency of GPP flares varies between patients, and symptoms can last for months; common triggers include smoking, infections, stress, medication use and pregnancy<sup>1,4</sup>
- There is a lack of consensus over the precise definition of a flare, diagnosis, and the typical frequency, severity or duration
- There is limited published literature describing how physicians treat flares and post-flare disease activity<sup>5</sup>
  - Standard of care for GPP varies by region, and there are no therapies approved for the treatment of GPP in the USA or Europe<sup>6</sup>
- Given the limited data on GPP, a survey of dermatologists from the USA and Canada participating in the Corrona Psoriasis Registry was conducted
- The aims of this survey were to improve the understanding of the natural history of GPP, how dermatologists diagnose GPP and GPP flares, the treatment options for GPP currently prescribed by dermatologists and their perceived adequacy, and to establish the challenges and unmet needs faced in the treatment of GPP

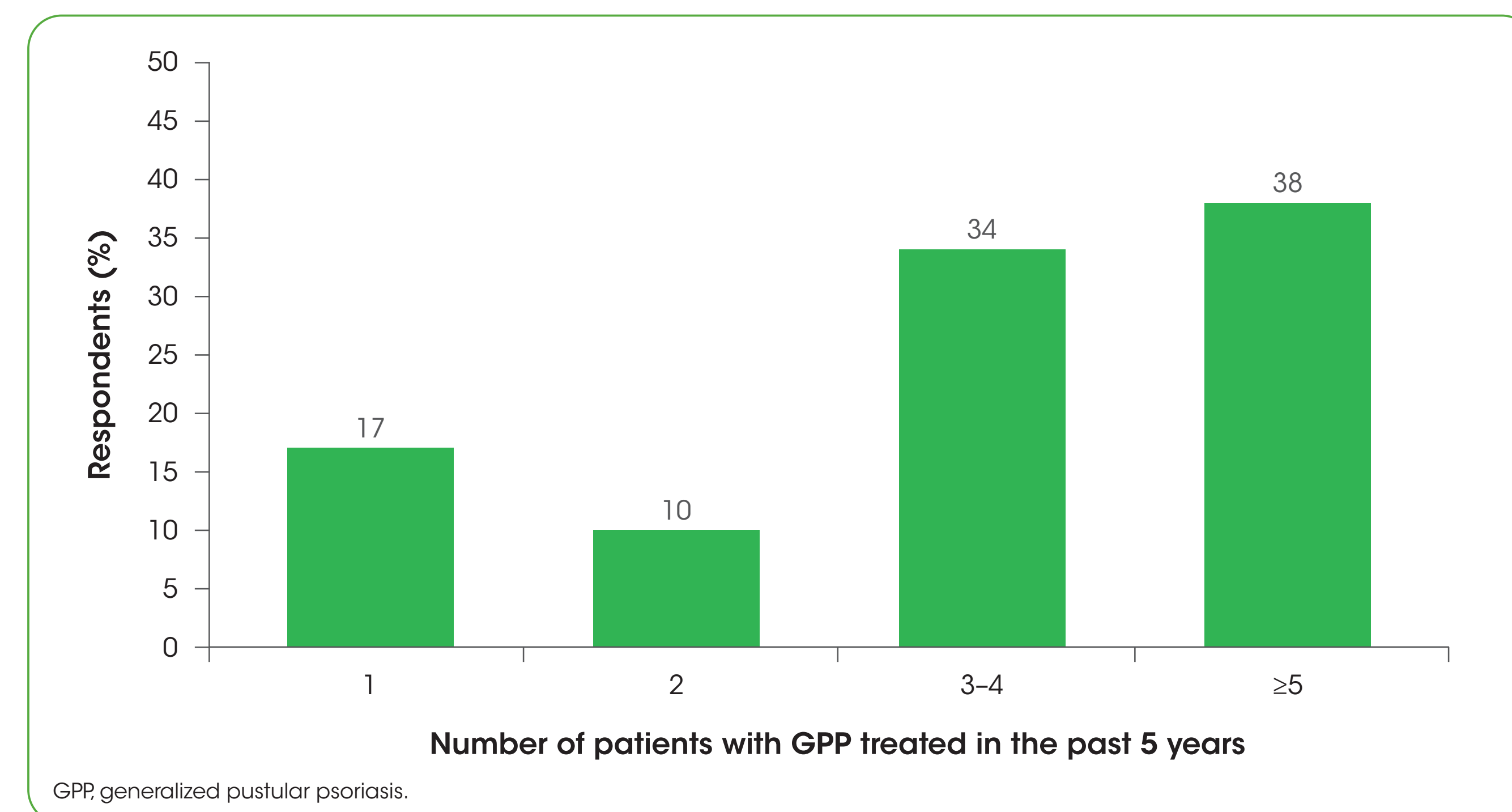
## Methods

- Dermatologists in the Corrona Psoriasis Registry (a collaboration with the National Psoriasis Foundation) who had treated adult patients (aged ≥18 years) with GPP within the past 5 years were eligible to participate in the survey
  - Dermatologists must have been at an active clinical centre as of 10 September 2019 (N=448) to be included
- Dermatologists in the Corrona Psoriasis Registry who had treated a patient with GPP (N=32) were invited to participate in an online survey hosted on SurveyMonkey
- The survey included 28 multiple choice questions exploring GPP flare onset and diagnosis, flare frequency and duration, treatment of flares, treatment of residual disease and physicians' overall experience of managing patients with GPP
- Respondents were asked to exclude patients with juvenile pustular psoriasis, localised forms of pustular psoriasis (e.g. palmoplantar pustulosis and acrodermatitis continua of Hallopeau), acute generalized exanthematous pustulosis, or pustulation restricted to psoriatic plaques
- A descriptive analysis of the responses was conducted

## Results

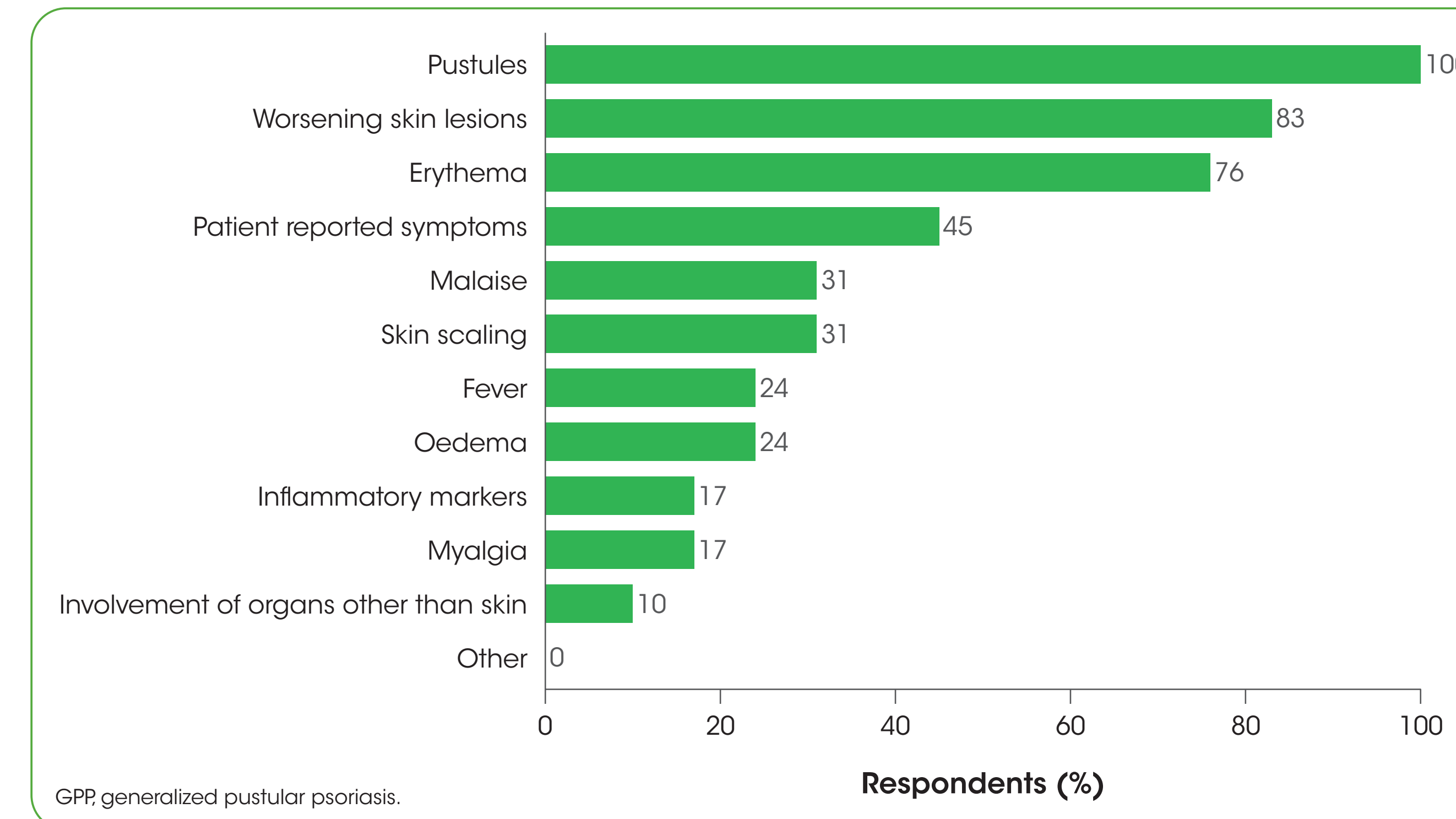
- Of the 32 invited dermatologists, 30 met the eligibility criteria. Of these 30 eligible dermatologists, 29 responded (97% eligible response rate)
  - Most respondents reported treating multiple patients with GPP in the past 5 years, with 72% reporting ≥3 patients (Figure 1)

Figure 1. Number of patients with GPP treated in the past 5 years by respondents (N=29)



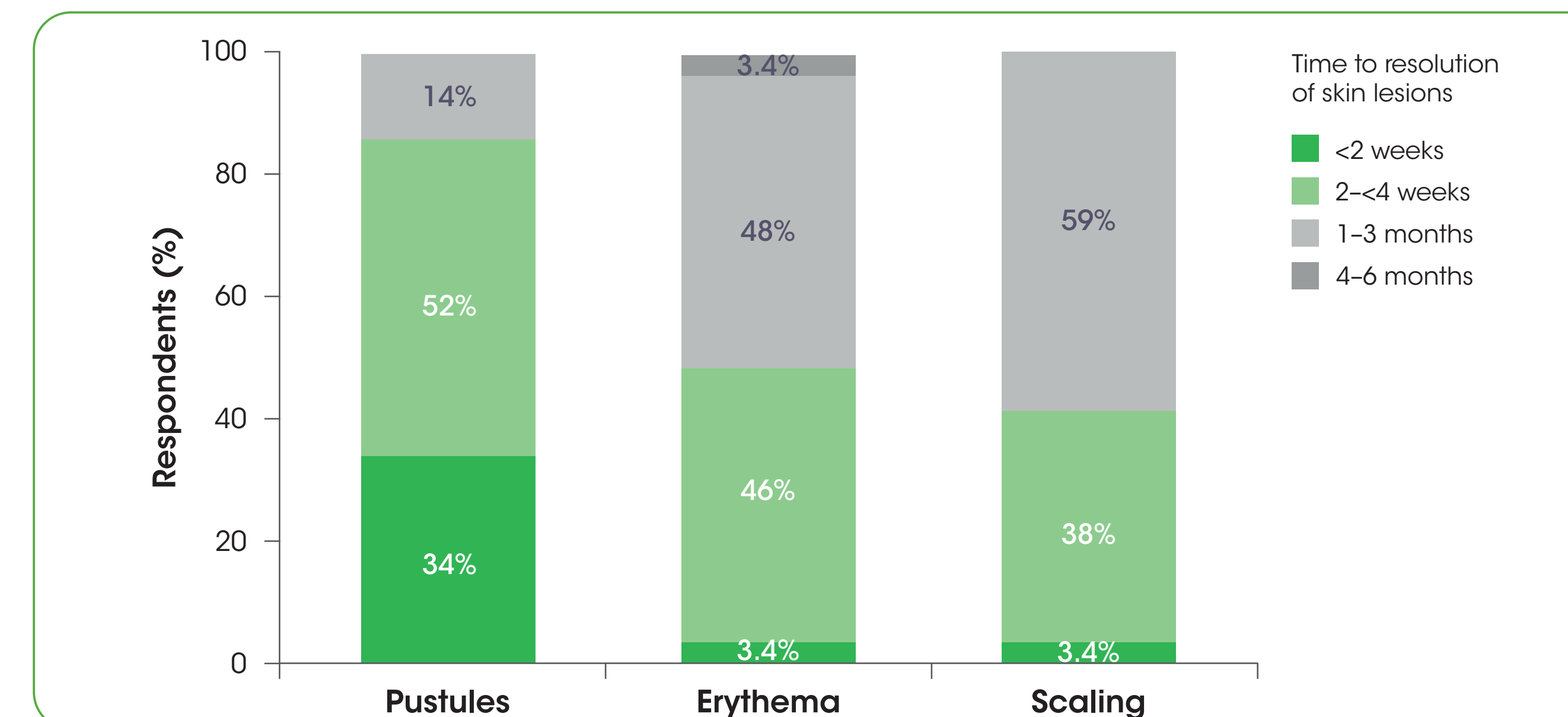
- All respondents stated that pustules were necessary to diagnose a GPP flare, followed by a worsening of skin lesions (83%) and erythema (76%; Figure 2)

Figure 2. A summary of the criteria used by responding dermatologists to diagnose GPP flares (N=29)



- Most respondents (69%) estimated that their patients had an average of 0-1 flare per year and 28% estimated 2-3 per year
  - Among the respondents whose patients averaged 0-1 flare per year, 20% of those respondents indicated that treatments did not adequately prevent flares, versus 67% among the respondents with patients averaging ≥2 flares per year
  - Steroid withdrawal (64%), infection (58%) and stress (50%) were cited as the most common triggering factors for GPP flares
- Over half (55%) of respondents reported that flares typically last 2-<4 weeks, and 41% reported 1-3 months
- The typical time to resolution of skin lesions was 2-<4 weeks (52%) for pustules, 1-3 months (48%) for erythema and 1-3 months (59%) for scaling (Figure 3)

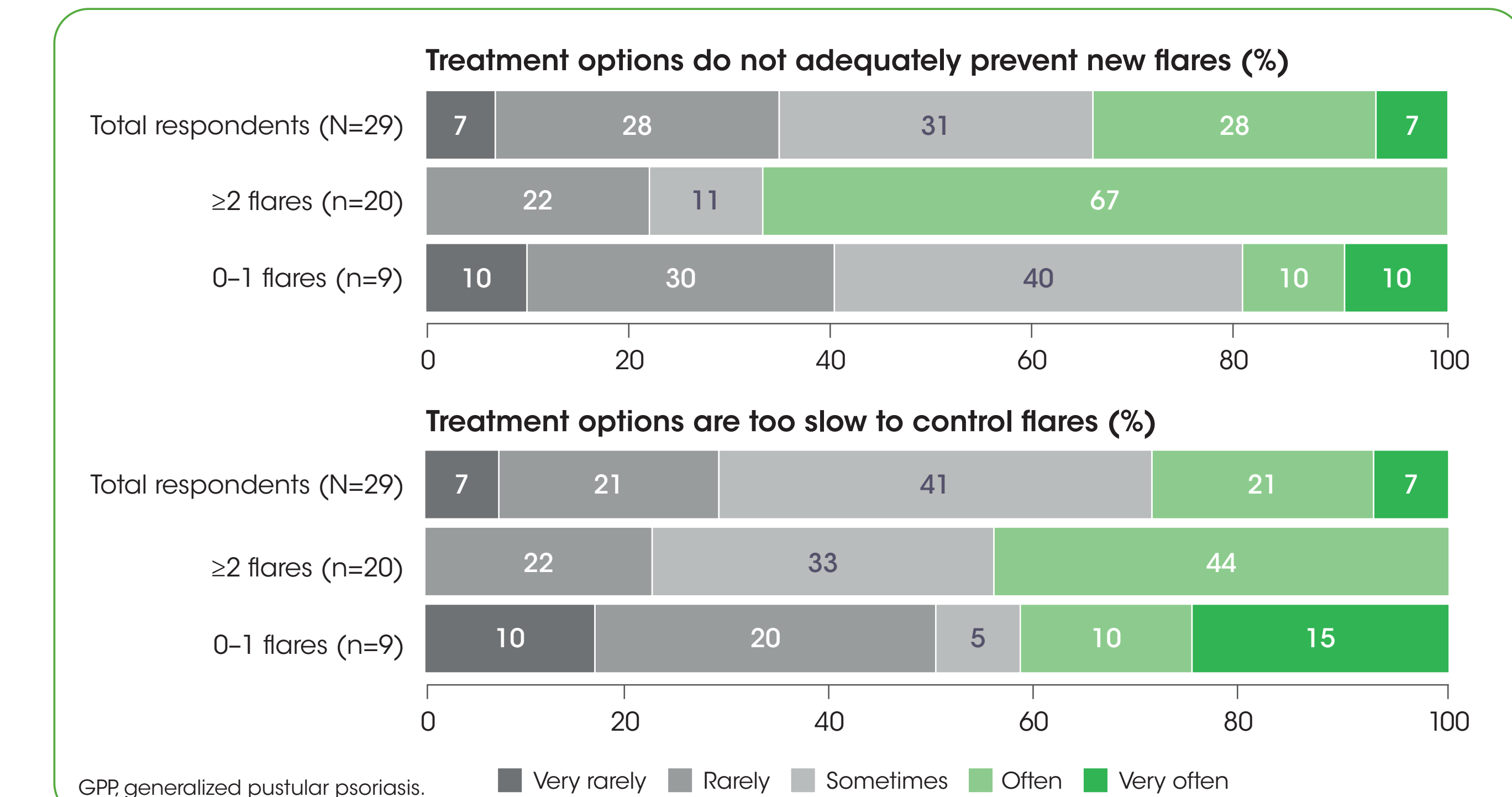
Figure 3. Time to resolution of skin lesions as reported by respondents (N=29)



- The most common criteria for flares to be considered resolved were few to no pustules (83%), reduced to no erythema (83%) and minimal to no skin lesions (66%)
- The frequency of flares requiring hospitalisation was reported as "somewhat", "very common" or "always" in 37% of respondents (17%, 17% and 3.4%, respectively), while the most common durations of hospitalisation were for 3-4 and 5-8 days (43% and 29% of respondents, respectively)
- Post-flare, 83% of respondents indicated that patients still had residual disease, most commonly: minimal skin scaling (76%), minimal skin lesions (66%) and reduced erythema (66%)

- Most dermatologists (67%) whose patients had flares ≥2 times per year indicated that treatments often did not adequately prevent new flares, and 78% considered available treatment options to be too slow to control flares "sometimes", "often", or "very often" (Figure 4)

Figure 4. Reported efficacy of current treatment options for GPP across all responding dermatologists and stratified by patient annual flare frequency (N=29)



## Conclusions

- This survey of dermatologists in the Corrona Psoriasis Registry suggests some consensus regarding dermatologists' opinions on the clinical presentation of GPP flares and the nature and frequency of GPP flares
- GPP flares may last up to 3 months and, despite treatment, most patients still have residual disease post-flare, demonstrating the chronic nature of the disease
- The average duration of hospitalisation for flares reported was 3-8 days, and while some therapies exist, the development of novel therapies that safely control acute GPP flares over a long-term period is warranted

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## Disclosures

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