

Prevalence and Treatment Patterns of Generalized Pustular Psoriasis (GPP) in Germany – Results of a Delphi Consensus Study

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Estimates of GPP prevalence in Germany were consistent with recently published estimates. According to experts, biologics are often used as first-line therapy, however, a proportion of patients show non-response

PURPOSE

This study aimed to determine the prevalence and treatment patterns of GPP in Germany.

INTRODUCTION

- GPP is a rare, severe, relapsing/remitting neutrophilic skin disease characterised by the sudden onset of erythematous cutaneous eruptions with sterile pustules^{1–4}
- While estimates of prevalence have ranged from 500–2,000 patients per year, there are no official records on incidence rates, nor disease-specific guidelines, for GPP in Germany^{1,5}
- In addition, due to the rarity of the disease, there are limited published data and few guidelines available on treatment of GPP flares⁵
- Consequently, there is a need for a better understanding of prevalence and treatment patterns for GPP flares in the German population

CONCLUSIONS

- A three-round Delphi consensus panel estimated that GPP prevalence in Germany is approximately 1.2 per 100,000 persons, with most patients experiencing moderate or severe disease
- The GPP disease course is usually classified by the severity of the most recent flare and assessed using BSA, which is considered the most important tool
- Consensus on GPP flare characteristics suggested a substantial disease burden
 - Experts estimated that patients experience on average 1.5 GPP flares per year
- Biologics may be frequently used for the treatment of GPP flares, however, some patients show non-response
- Therefore, a need for a fast-acting and effective GPP treatment remains
- Lastly, experts estimated that more than half of patients with GPP are eligible for maintenance therapy

METHODS

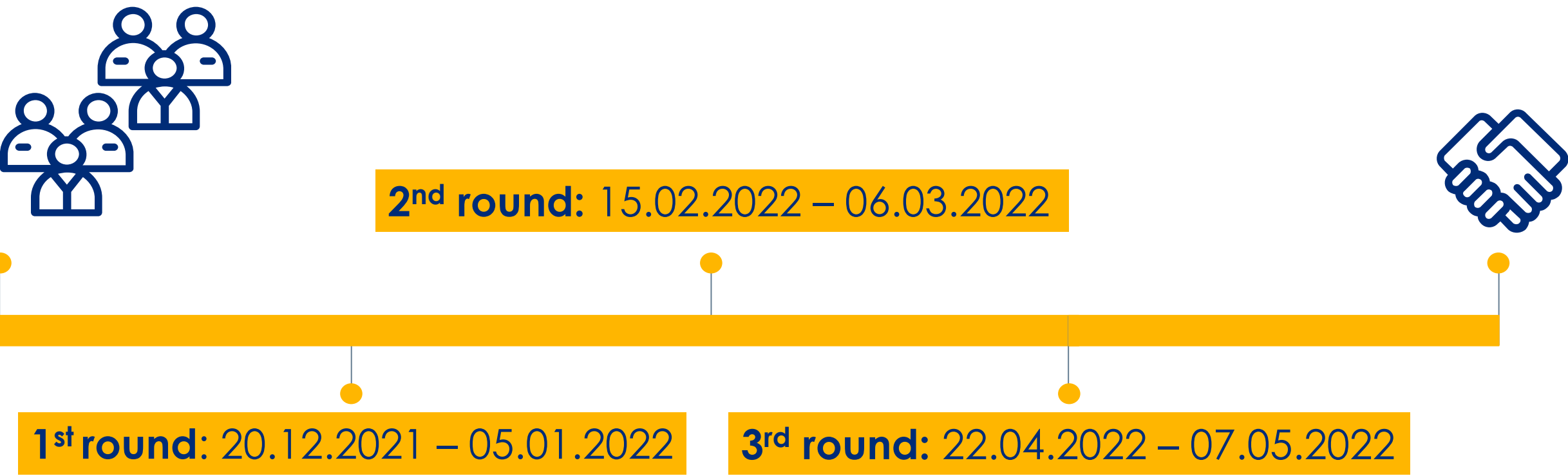
- A three-round Delphi consensus panel with six German clinical experts was held using online questionnaires between December 2021 and May 2022

- Six* experts completed three rounds of online questionnaires, answering questions relating to GPP prevalence; disease course; and flare severity, characteristics and treatment

- Consensus was reached when 75% of experts were in agreement

- Five of the six experts were employed in a university hospital with >10 years of experience treating patients with GPP
- Each expert was treating an average of three patients with GPP at the time of the study

*One expert dropped out after the 1st round, leaving five panellists in the 2nd and 3rd rounds.



RESULTS

GPP prevalence and disease course

Proportion of patients with psoriasis who have GPP

0.75% have GPP

All patients with psoriasis

Severity of GPP disease course

21% Mild, 46% Moderate, 33% Severe

GPP flare duration

60% <4 weeks, 25% 4–8 weeks, 11% 8–12 weeks, 4% >12 weeks

~1000 patients with GPP in Germany

GPP prevalence was estimated as 1.2 per 100,000 persons, or ~1000 patients with GPP in Germany, with the majority (79%) experiencing moderate to severe disease. Experts estimated that patients experience on average 1.5 flares per year

Distribution of first-line treatment options and treatment pathways in patients with GPP

Distribution of first-line treatment options

32% Systemic corticosteroids, 29% Biologics, 18% Immunosuppressants, 22% TNF-α inhibitors, 25% IL-17/IL-17A inhibitors, 14% IL-23, IL-12/IL-23 and IL-1β/IL-12 inhibitors, 1% Other, 2% Topical corticosteroids, 7% Cytostatic drugs, 7% Combination therapy, 4% Phototherapy/PUVA

Response rates to first-line therapy and distribution of treatment pathways in case of non-response

70% Response, 30% Non-response

44% Change to second-line therapy, 35% Adaption/adjustment of first-line therapy dosage, 14% Add-on therapy

The most commonly used first-line GPP treatments were agreed to be systemic corticosteroids and biologics; TNF-α inhibitors were the most frequently used biologics. Of the patients who did not respond to first-line therapy, the largest proportion went on to receive a second-line therapy. More than half of patients (56%) were considered to be eligible for maintenance therapy

Abbreviations
BSA, body surface area; GPP, generalized pustular psoriasis; IL, interleukin; PUVA, psoralen ultraviolet A; TNF, tumour necrosis factor.

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