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Global Delphi consensus on treatment goals for generalized pustular psoriasis

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See Appendix 1 for details on the Partnering for Innovation and Excellence in Rare Skin Diseases (PIONEERS®) Working Group and the DELPHI Panel.

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Abstract

Background Generalized pustular psoriasis (GPP) is a chronic, systemic, neutrophilic inflammatory disease. A previous Delphi panel established areas of consensus on GPP, although patient perspectives were not included and aspects of treatment goals remained unclear.

Objectives To identify and achieve consensus on refined, specific treatment goals for GPP treatment via a Delphi panel with patient participation.

Methods Statements were generated based on a systematic literature review and revised by a Steering Committee. Statements were categorized into overarching principles, and short- and long-term treatment goals. A global panel of 30 dermatologists and 3 patient representatives voted in agreement or disagreement with each statement. Consensus was defined as ≥80% approval by the panellists.

Results Consensus was reached in the first round of voting and ≥90% agreement was reached for 23 of 26 statements. In summary, GPP requires a timely, tailored treatment plan, co-developed by patients and physicians, that involves a multidisciplinary approach and addresses the complexity, heterogeneity and chronicity of the disease. Short-term treatment goals should include pustule clearance within 7 days and prevention of pustule recurrence, reduction of cutaneous symptom burden (−4 or more points on the Itch and Skin Pain Numeric Rating Scale), improvement in systemic symptoms (e.g. resolution of fever within 3 days of treatment initiation and reduced fatigue), prevention of life-threatening complications and progressive improvement of inflammatory biomarkers. In patients with comorbid psoriatic diseases, treatment decisions should prioritize GPP. Long-term treatment goals should include minimizing disease activity through flare prevention and symptom control between flares, sustained disease control, management of comorbidities and improvement in quality of life (QoL). Small differences in perception between patients and physicians regarding the importance of certain treatment goals (e.g. avoiding hair and/or nail loss to improve QoL), reflect the complexity of assessing treatment goals and emphasize the need for a patient-centred approach.

Conclusions In the first global Delphi panel in GPP to include patient perspectives, consensus between dermatologists and patients was achieved on overarching principles of treatment, and short- and long-term treatment goals for GPP. These findings provide valuable insights for developing guidelines that consider the perspectives of patients and physicians in the treatment of GPP.

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Lay summary

Generalized pustular psoriasis ('GPP' for short) is a rare and life-long inflammatory disease that causes skin redness and blisters. People with GPP often experience a high temperature, tiredness and skin pain. The symptoms can suddenly become worse in episodes called 'flares'. Until now, doctors have not had standard treatment goals for GPP, or agreed ways to measure if a medicine is working well.

To determine what treatment goals are important to people with GPP and their doctors, a panel of 30 expert doctors and 3 patient representatives from 24 countries took part in a survey. The panel voted on 26 statements related to GPP treatment. The statements were prepared by a team of expert doctors. After 1 round of voting, the panel agreed on all the statements. Regarding GPP treatment, at least 32 out of 33 panellists agreed on each statement. The panel agreed that GPP is a complex, life-long disease, and that treatment should be started quickly, be tailored to each patient and involve doctors from other specialties, as well as dermatologists (skin doctors). Doctors and patient representatives gave different levels of importance to some treatment goals. For example, the 3 patient representatives all agreed that avoiding losing their hair/nails was important, but only 24 out of 30 doctors held the same view.

This particular type of survey on GPP treatment goals was the first to include patient representatives. Panellists agreed on all short-term, long-term and overall treatment goals for GPP.

What is already known about this topic?

- Generalized pustular psoriasis (GPP) is a chronic, systemic inflammatory disease.
- Patients with GPP experience a considerable disease burden, which affects their quality of life.
- Commonly used treatment goals for GPP often lack specificity and actionable outcome measures.

What does this study add?

- This global Delphi panel was the first to involve both physician and patient participation.
- Overarching treatment principles, and short- and long-term treatment goals achieved high levels of consensus among the panellists after one round of voting.
- These findings provide valuable insights into unmet needs in the treatment of GPP from the perspectives of both patients and physicians.

Psoriasis is a chronic inflammatory disease comprising different clinical phenotypes, of which plaque psoriasis is the most common, accounting for approximately 80% of cases.[1] A rare and severe subtype, generalized pustular psoriasis (GPP), is recognized as clinically, genetically and phenotypically distinct from plaque psoriasis (International Classification of Diseases, 10th Revision code L40.1).[2] GPP is a chronic, systemic, neutrophilic inflammatory disease associated with cutaneous and noncutaneous manifestations.[3–6] The clinical course is heterogeneous, with chronic manifestations and periods of flaring.[6,7] The unpredictability of GPP manifestations greatly affects patients' quality of life (QoL), causing fear and anxiety over the disease course, which add to short- and long-term impacts on QoL.[6–13]

Until recently, no treatment goals had been defined for GPP. Moreover, treatment recommendations were often based on therapies developed for the treatment of plaque psoriasis.[6,14] In contrast with plaque psoriasis, the interleukin (IL)-36 pathway plays a central pathogenetic role in GPP, driving pustule formation – a key manifestation of the disease.[5,6,15] Thus, treatments for plaque psoriasis are not designed to target the underlying pathogenesis of GPP. Using treatment guidelines and objectives for plaque psoriasis also fails to reflect the lived experience of patients with GPP. Studies have shown that GPP exerts a considerable burden on patients, with a greater impact on QoL and higher healthcare resource use and economic costs than plaque psoriasis.[16–18]

A global Delphi consensus on the clinical course, diagnosis, treatment goals and holistic management of GPP was carried out in 2022. It highlighted the need for approved treatment options that address the full nature of GPP in a targeted, effective and sustainable manner.[6] At the time of publication, achieving these goals was challenging with the available treatment options. Other consensus statements have been published, although none has provided comprehensive guidance on treatment goals. A 2023 consensus statement from the US National Psoriasis Foundation (NPF) emphasized the life-threatening nature of GPP flares and the need for urgent treatment, advocating for timely access to U.S. Food and Drug Administration (FDA)-approved therapies for GPP to reduce morbidity and mortality.[3] Moreover, in 2024, the International Psoriasis Council (IPC) established an international consensus on the definition and diagnostic criteria of GPP with a panel of 33 global GPP experts,[19] underscoring the value of collaboration in the field of rare diseases.

Inclusion of the patient voice in GPP-specific consensus statements is important as previous studies have shown that a gap exists between physicians and patients in their perception of disease burden, treatment goals and treatment

satisfaction.[20] Incorporating patients' expertise and expectations could allow a more inclusive approach, improving treatment success.

Here we report findings from the first global Delphi panel in GPP to include both physicians and patient representatives. It was conducted to achieve consensus on key treatment goals for GPP.

Materials and methods

Statements relating to overarching statements, and short- and long-term treatment goals were developed following a systematic literature review (SLR) and discussion with a Steering Committee of clinical experts. These statements were evaluated within the framework of a Delphi panel of physicians and patient representatives. Details on the SLR methodology, Steering Committee and Delphi panellists are available in Appendix S1 and Table S1 (see Supporting Information), and Appendix 1, respectively.

Data from the Delphi panel were collected via two planned rounds of questionnaires, which were hosted on an online platform (IQVIA; https://www.iqvia.com/). Panellists were asked to rate their level of agreement or disagreement with each statement on a Likert scale [1 (strong disagreement) to 7 (strong agreement)]. A free-text field was also available to provide comments. Consensus was reached when ≥80% of the panel scores fell within either the region of disagreement (1–3) or agreement (5–7). If no consensus was reached for specific statements after round 1, these statements were to be discussed and revised by the Steering Committee and included in the questionnaire for round 2 of the survey.

A descriptive statistical analysis was conducted using SAS® software version 9.4 or later (SAS Institute, Cary, NC, USA). Continuous variables were described by number (of valid cases/missing values), mean (SD) and median (range). Categorical variables were described as the total number and relative percentage per category.

Results

Systematic literature review

The results of a previous SLR in GPP conducted by Puig *et al.* provided a comprehensive understanding of the current evidence and supported the generation of 26 statements to be evaluated by the Delphi panel (Table S2; see Supporting Information).[6]

Demographics of the Delphi panellists

The panel comprised 33 panellists from 24 countries, including 3 patient representatives from 3 different countries. Geographically, 36% of the panellists were based in Europe, 33% in Asia, 15% in the USA and Canada, 12% in South America and 3% in Africa, ensuring global representation (Figure S1; see Supporting Information). The sex ratio of panellists was largely balanced (49% women vs. 52% men) and all panellists were aged ≥40 years. All physicians in the panel were dermatologists, with more than half (57%) working in a hospital inpatient/outpatient practice setting; 33% were hospital- and office-based and 10% were office-based only. Eighty-three per cent of physicians had co-authored publications on GPP and 77% had participated in national or international working groups on GPP. Of the three patient representatives, two were from Asia and one was from the USA; all had been diagnosed with GPP for >10 years. One patient representative was a member of a GPP-specific patient advocacy group.

Delphi method findings

The round 1 questionnaire included 20 main statements, consisting of 18 standalone statements and 2 statements with 8 substatements (Figure 1). Consensus was achieved for all statements in round 1 (Tables 1–3); a second round was therefore not conducted (study timeline in Table S3; see Supporting Information). The following statements are listed in descending order of agreement.

Table 1 Statements on overarching principles in generalized pustular psoriasis (GPP)[a]

Statement	Overall level of agreement (%)	Physicians agreeing with statement (*n*=30)	Patient representatives agreeing with statement (*n*=3)
GPP is a complex, heterogeneous and chronic condition, with effective management requiring timely treatment and multidisciplinary collaboration to prevent escalation to life-threatening complications	100	30	3
Effective management of GPP requires a comprehensive treatment approach that addresses:			
• short-term goals, such as flare treatment	97	30	2
• long-term objectives, including the prevention of future flares	100	30	3
• minimizing disease activity	100	30	3
• optimizing functional status and improving QoL	100	30	3
• minimizing morbidity	97	29	3
• preventing or minimizing complications that may arise from untreated active disease	100	30	3
Tailored treatment plans should be created collaboratively between the patient and their HCPs	97	29	3
Patients should be seen promptly and offered regular evaluations by appropriate specialists, where treatment should be modified as necessary	97	29	3

HCP, healthcare professional; QoL, quality of life. [a]Statements are listed in descending order of agreement.

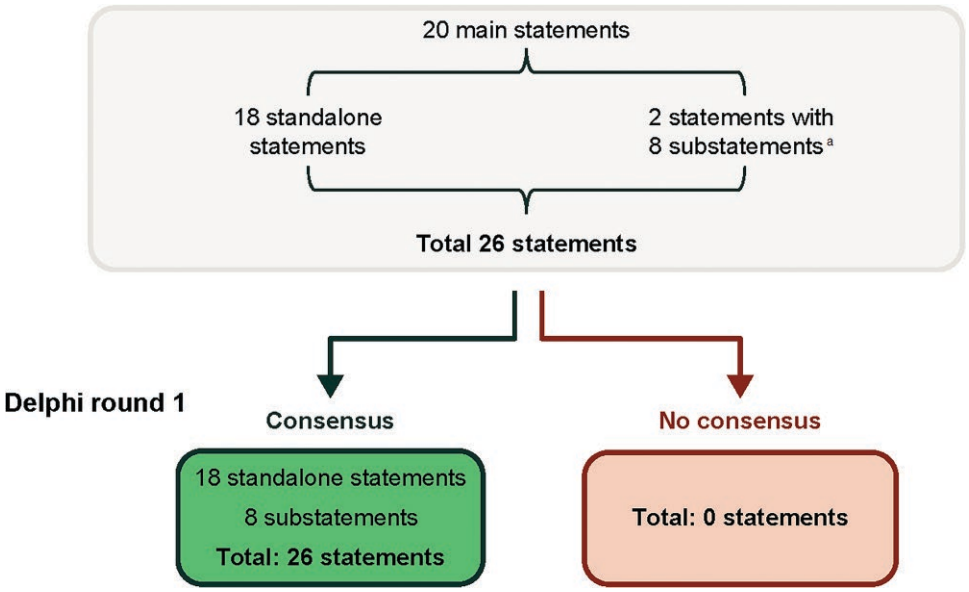

Figure 1 Flowchart of consensus on statements following Delphi round 1. [a]For statements with substatements, only the substatements were voted on.

Overarching principles

1. 'Generalised pustular psoriasis is a complex, heterogeneous and chronic condition, with effective management requiring timely treatment and multidisciplinary collaboration to prevent escalation to life-threatening complications' (100%).

Consensus was reached for all overarching principles of GPP management (Table 1). All panellists agreed with the characterization of GPP as a complex, heterogeneous and chronic condition, with effective management requiring timely treatment and multidisciplinary collaboration to prevent escalation to life-threatening complications. This echoes the Delphi consensus by Puig *et al.*,[6] in which these statements generated high levels of agreement. The consensus statement from the NPF also advocates for timely treatment with U.S. FDA-approved therapies for GPP in order to reduce morbidity and mortality in patients presenting with GPP.[3] GPP is associated with mortality rates of 2–16% due to severe complications such as multisystem organ failure and sepsis.[8] All panellists acknowledged that GPP can have fatal consequences, and life-threatening complications should be prevented via timely treatment and multidisciplinary collaboration.

2. 'Effective management of generalised pustular psoriasis requires a comprehensive treatment approach that addresses both short-term goals (such as flare treatment) and long-term goals (including the

Table 2 Statements on short-term treatment goals[a]

Short-term treatment goal	Overall level of agreement (%)	Physicians agreeing with statement ($n=30$)	Patient representatives agreeing with statement ($n=3$)
Substantial pain reduction (e.g. at least −4 points for the 0–10 Itch and Skin Pain NRS item)	100	30	3
Prevention of life-threatening complications	100	30	3
Evaluation of the effectiveness of treatment within 3–7 days of initiation	97	29	3
Resolution of fever within 3 days	97	29	3
Substantial improvement in fatigue	97	29	3
Prevention of the formation of new pustules; no new/fresh pustules observed within 2–3 days of treatment initiation	94	28	3
When managing patients with comorbid psoriatic diseases, treatment decisions should prioritize GPP	94	28	3
Achievement of pustular clearance; GPPGA pustulation subscore of 0 within 7 days of treatment initiation	91	27	3
Progressive improvement of inflammatory biomarkers (e.g. CRP and/or ESR)	91	27	3
Pustules should be the main metric for assessing response to treatment	82	25	2
Avoiding hair and/or nail loss	82	24	3

CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; GPP, generalized pustular psoriasis; GPPGA, Generalized Pustular Psoriasis Physician Global Assessment; NRS, numeric rating scale. [a]Statements are listed in descending order of agreement.

Table 3 Long-term treatment goals[a]

Long-term treatment goal	Overall level of agreement (%)	Physicians agreeing with statement ($n=30$)	Patient representatives agreeing with statement ($n=3$)
Sustained disease control is defined as continuous clear or almost-clear skin	100	30	3
Sustained improvement of QoL as measured by DLQI and/or other related PROs, as well as work productivity	100	30	3
Minimizing disease activity to the greatest extent possible, including but not limited to:			
• preventing flares, reducing frequency of flares and/or prolonging time between flares	100	30	3
• controlling signs and symptoms of GPP (e.g. pustules, erythema, pain, itching) between flares	97	29	3
Management of potential associated conditions	94	28	3
Clinicians and patients need to be educated that tapering or discontinuing therapy in patients who have achieved treatment goals may result in new episodes of flaring	88	26	3

DLQI, Dermatology Life Quality Index; GPP, generalized pustular psoriasis; PRO, patient-reported outcome; QoL, quality of life. [a]Statements are listed in descending order of agreement.

prevention of future flares, minimising disease activity, optimising functional status and improving quality of life, minimising morbidity and preventing or minimising complications that may arise from untreated active disease)' (97–100%).

There was a high level of agreement that effective management of GPP requires a comprehensive treatment approach that addresses short- and long-term treatment goals. In the absence of specific guidelines, treatments approved for plaque psoriasis have historically been used to treat GPP. Retinoids, ciclosporin and methotrexate are the most commonly used nonbiologic treatments for GPP, although none has been approved specifically for GPP.[14,21] The evidence for use of most biologics in the treatment of GPP (e.g. inhibitors of tumour necrosis factor and IL-17) stems mostly from uncontrolled clinical trials and case studies/reports,[21] and these treatments have been approved in limited markets only.[14,21] Only IL-36 receptor (IL-36R) inhibitors have data from double-blind placebo-controlled studies specifically for the short- and long-term treatment of GPP.[22] Spesolimab – a humanized monoclonal antibody that inhibits IL-36R – has been evaluated in the largest clinical programme designed for the treatment of patients with GPP.[23,24]

3. 'Tailored treatment plans should be created collaboratively between the patient and their healthcare providers' (97%).

Collaboration between patients and healthcare professionals (HCPs) to develop a tailored plan achieved a high degree of agreement, underlining the desire of both patients and physicians to involve patients in decision-making.

4. 'Patients should be seen promptly and offered regular evaluations by appropriate specialists, where treatment should be modified as necessary' (97%).

The majority of the panel agreed on the need for prompt evaluations by appropriate specialists, during which treatment should be modified as necessary. Although regular evaluations are recommended, patients may often initiate the consultation due to the unpredictable nature of flares.[6] In addition to dermatologists, specialists working in rheumatology, genetics, cardiology and high-dependency or intensive care units may be involved in the treatment of GPP.[25] It is important to define clear roles and responsibilities within the multidisciplinary team when creating an informed management plan for each patient.[25]

Short-term treatment goals

1. 'Substantial pain reduction [e.g. at least −4 points for the 0–10 Itch and Skin Pain Numeric Rating Scale (NRS) item]' (100%).

Consensus was reached for all statements regarding short-term treatment goals for GPP (Table 2). All panellists regarded itch and pain reduction to be an immediate priority. The significance of this is supported by the literature. In a 2019 patient workshop, 86% of participants said their daily lives were most commonly affected by itching.[26] More than 70% of patients with GPP rated the burden associated with pain as high, as reported by Reisner *et al.*[10]

2. 'Prevention of life-threatening complications' (100%).

GPP can be life threatening due to complications during a flare.[6] Some studies have shown that 35–64% of patients with GPP who experience flares require hospitalization.[8,27,28] Life-threatening complications include infections, and renal, hepatic, respiratory and heart failure.[6,8] Prevention of these events is key to preventing mortality, as indicated by the panellists.

3. 'Evaluation of the effectiveness of treatment within 3–7 days of initiation' (97%).

There was a high level of agreement that the effectiveness of treatment should be evaluated within 3–7 days of initiation, echoing the overarching principle of effective management requiring timely treatment, as well as the consensus from the NPF.[3]

4. 'Resolution of fever within 3 days' (97%).

Consensus was achieved for the resolution of systemic symptoms, including fever. Of note, a recent IPC consensus statement provided further guidance on the role of fever in GPP, stating that although GPP may manifest with or without systemic signs and symptoms such as fever, they are considered to be diagnostic criteria for GPP.[19] Furthermore, previous studies have shown that approximately 26–39% of patients develop a high fever during a flare.[20,29]

5. 'Substantial improvement in fatigue' (97%).

Almost all panellists agreed that resolution of fatigue, a systemic symptom of GPP, should be treated in the short term. As with fever, this aligns with the recent IPC consensus that fatigue and other systemic symptoms support a diagnosis of GPP.[19] In a patient survey, fatigue was ranked as one of the most common and bothersome symptoms of GPP.[12]

6. 'Prevention of the formation of new pustules; no new/fresh pustules observed within 2–3 days of treatment initiation' (94%).

The majority of panellists agreed on the importance of preventing new pustule formation. Pustules are the key manifestation of GPP symptoms and are observed both clinically and histologically.[30] Twenty-eight of 30 (93%) clinicians agreed that preventing the formation of new pustules should be a short-term goal; this achieved unanimous agreement from patient representatives.

7. 'When managing patients with comorbid psoriatic diseases, treatment decisions should prioritise generalised pustular psoriasis' (94%).

When managing patients with concomitant psoriatic diseases, panellists agreed that treatment decisions should give precedence to GPP. Approximately half of patients with GPP also present with plaque psoriasis,[31,32] despite the distinct nature of both diseases. Treatment of GPP should be prioritized to avoid life-threatening complications.

8. 'Achievement of pustular clearance; Generalized Pustular Psoriasis Physician Global Assessment pustulation subscore of 0 within 7 days of treatment initiation' (91%).

Twenty-seven of 30 (90%) clinicians agreed that pustular clearance, defined as a Generalized Pustular Psoriasis Physician Global Assessment (GPPGA) pustulation subscore of 0, should be achieved within 7 days of treatment initiation; this was unanimously agreed upon by the patient representatives. Randomized clinical trials for GPP have used a GPPGA pustulation subscore of 0 as an endpoint when developing novel therapies for GPP.[23]

9. 'Progressive improvement of inflammatory biomarkers (e.g. C-reactive protein and/or erythrocyte sedimentation rate)' (91%).

Improvement of systemic inflammation, indicated by C-reactive protein (CRP) and/or erythrocyte sedimentation rate (ESR), was agreed upon by the majority of panellists as a treatment goal. Laboratory abnormalities indicative of the systemic involvement seen in GPP include elevated CRP and ESR.[33] There is no global agreement on the cutoff values for CRP and ESR,[33] although recommendations from treatment guidelines in Japan have provided thresholds for severity.[4,34] In addition, the recently published IPC consensus states that elevated CRP supports a diagnosis of GPP.[19]

10. 'Pustules should be the main metric for assessing response to treatment' (82%).

Using pustules as the main metric for assessing response to treatment achieved consensus in the first round; of 30 clinicians, 25 (83%) agreed with this statement, and 2 of the 3 (67%) patient representatives agreed. The number of patient representatives who participated in the Delphi panel was limited; therefore, any potential differences in treatment priorities between patients and physicians should be interpreted with caution. A subset of physicians may consider other symptoms to be of greater or equal importance to pustules. Of note, the recent IPC consensus states that macroscopically visible sterile pustules are mandatory for the diagnosis of GPP.[19]

11. 'Avoiding hair and/or nail loss' (82%).

The importance of avoiding hair and/or nail loss as a short-term treatment goal was unanimously agreed upon by all three patient representatives. The level of agreement among physicians (80%) was less consistent, highlighting potential differences in priorities between patients and physicians regarding short-term treatment. Little has been published on this specific symptom of GPP and its treatment. There is evidence that in patients with psoriasis (including but not limited to plaque psoriasis), nail psoriasis is associated with a significantly greater impairment in QoL when compared with patients without nail involvement.[35]

Long-term treatment goals

1. 'Sustained disease control is defined as continuous clear or almost-clear skin' (100%).

All panellists agreed that sustained disease control is defined as continuous clear or almost-clear skin. A randomized trial for prevention of GPP flares included sustained disease control, defined as a GPPGA total score of 0 or 1 (clear or almost-clear skin) through to week 48 (end of study), as one of the endpoints,[36] providing evidence for this statement.

2. 'Sustained improvement of quality of life as measured by the Dermatology Life Quality Index and/or other related patient-reported outcomes as well as work productivity' (100%).

Consensus was reached by all panellists on sustained improvement in QoL as measured by the Dermatology Life Quality Index (DLQI) and/or other related patient-reported outcomes (PROs), as well as work productivity. Whether during or between flares, GPP is associated with reduced QoL through its symptoms, impact on activities of daily

living, and the resulting emotional and psychological burden placed on patients.[6,9,10] The impact of treatment on patients' QoL has been most commonly evaluated in studies of biologics,[22] with the DLQI being the most widely used instrument across studies.[37]

3. 'Minimising disease activity to the greatest extent possible, including but not limited to: preventing flares, reducing frequency of flares and/or prolonging time between flares, and controlling signs and symptoms of generalised pustular psoriasis (e.g. pustules, erythema, pain, itching) between flares' (97–100%).

The panel strongly agreed on the importance of minimizing disease activity as a long-term goal, including preventing and reducing the frequency of flares, and symptom control between flares. Flare frequency is highly variable, and although specific triggers have been identified (e.g. treatment with or withdrawal from systemic corticosteroids, infections, pregnancy and stress),[6] flares are often unpredictable and occur without warning. A survey of dermatologists from the CorEvitas Psoriasis Registry reported that 83% of patients with GPP still had chronic symptoms (e.g. minimal skin scaling/lesions and reduced erythema) after resolution of a flare.[11] Even in the absence of flares, patients have reported that GPP has a high impact on intimacy with a spouse or partner (23%), as well as the ability to exercise (21%), attend important life events (15%) and wear certain clothing or shoes (15%).[10]

4. 'Management of potential associated conditions' (94%).

There was agreement that management of comorbidities should be a long-term treatment goal. This reflects the consensus from the Delphi panel by Puig *et al.* that comorbidities such as plaque psoriasis, diabetes mellitus, hypertension and liver disease may affect treatment decisions.[6] Many patients with GPP suffer from comorbidities, which contribute to the ongoing burden for the patient and healthcare systems.[8] A French population-based study reported that in 1842 patients with GPP, comorbidities included hypertension (44%), ischaemic heart disease (26%), hyperlipidaemia (25%), congestive heart failure (24%) and type 2 diabetes (7%).[38]

5. 'Clinicians and patients need to be educated that tapering or discontinuing therapy in patients who have achieved treatment goals may result in new episodes of flaring' (88%).

This statement achieved a lower level of agreement (88%) than other long-term treatment goals. In the Delphi consensus by Puig *et al.*,[6] panellists achieved complete agreement in round 1 that treatment discontinuation was a potential trigger for flares. It is interesting to note that although consensus was reached, a lower percentage of physicians (n=26/30; 87%) agreed with the statement than patient representatives, who reached unanimous agreement. A possible explanation for this is that physicians may consider the risk to be well known. There are currently no existing outcome data in patients who have discontinued treatment following the achievement of treatment goals, and further research is required.

Discussion

A multinational panel of dermatologists and patient representatives reached consensus on all treatment goals related to GPP in the first round of this Delphi exercise, indicating a global understanding of the characteristics of GPP and a desire for specific, detailed goals for disease management. Building on an initial Delphi consensus on the clinical course, diagnosis, treatment goals and holistic management of GPP,[6] these findings add to the body of evidence and guidance for the treatment of GPP.

The Delphi panel reached ≥90% agreement on 23 of 26 statements. Regarding the overarching principles of GPP treatment, all panellists agreed that treatment should be timely and involve a multidisciplinary approach to address the complexity, heterogeneity and chronicity of the disease. Collaboration between patients and HCPs is necessary to develop a tailored treatment plan, with regular evaluations (initiated by both physicians and patients) and treatment modification as necessary. Short-term treatment goals should focus on rapid and substantial improvement using quantifiable measures (e.g. cutaneous symptom relief as assessed by at least a 4-point reduction in the itch and skin pain NRS). Post-treatment initiation, pustular clearance (a GPPGA pustulation subscore of 0) should be achieved within 7 days, prevention of new pustule formation within 2–3 days and fever resolution within 3 days. The effectiveness of treatment should be assessed within 3–7 days. Other goals include the prevention of life-threatening complications, and improvement in fatigue and inflammatory biomarkers. In addition, the treatment of GPP should be prioritized when managing patients with comorbid psoriatic disease, to avoid life-threatening complications and reduce the risk of mortality. For long-term treatment goals, the emphasis should be on minimizing disease activity through flare prevention and symptom control between flares, as well as sustaining disease control in the long term (i.e. maintaining clear or almost-clear skin, managing comorbidities and improving QoL, as measured by PROs such as DLQI).

International experts from multiple organizations have reiterated that GPP is a chronic disease, including the European Rare And Severe Psoriasis Expert Network, the Japanese Dermatological Association, the IPC, the NPF and the Delphi consensus by Puig *et al.*[3–6,19] All panellists in this Delphi consensus agreed that GPP is a complex, heterogeneous and chronic disease, and that treatment approaches should target all aspects of the clinical course, not just flares. The heterogeneity of GPP also requires treatment goals be tailored to each patient, as agreed by the majority of panellists.

Despite achieving consensus on all statements, some differences in opinions were also found. While aiming for a global representation, regional variations in clinical presentations, genetic mutations and available treatments may have impacted the consensus.[14,39–41] The majority of physicians believed that pustules should be the main outcome metric for assessing treatment response (n=25/30), although a numerically lower proportion of patient representatives agreed with this statement (n=2/3). This potentially

highlights the difference in treatment priorities between physicians, as well as between physicians and patients. It is likely that physicians manage patients at various timepoints during a flare (e.g. when their pustules are resolving); as such, using pustules as the main metric for treatment response may not be applicable. Patients considered the alleviation of other symptoms beyond the skin, specifically avoiding hair and/or nail loss, to be of high importance due to the psychological and emotional impact on their daily lives. The impact of hair and/or nail loss was noted by one of the patient panellists, who explained that 'obviously this will affect our self-esteem'. However, physicians may perceive hair and/or nail loss as rare in GPP. This emphasizes the importance of incorporating patients' expectations as part of a holistic approach to the treatment of GPP. Overall, patients appeared to put equal emphasis on cutaneous manifestations and symptoms beyond the skin (e.g. fever, fatigue and potential hair and/or nail loss).

Patient participation is essential for the development of treatment goals to reflect the aspects of disease that have the greatest impact on patients' QoL. Patients possess a unique knowledge of their condition and their own treatment priorities, especially in rare diseases.[42] A deeper understanding of what is a higher priority for patients could address any disconnect between HCPs and patients. This is reflected by a survey of patients with GPP and dermatologists, which showed that only 35% of patients discussed treatment goals with their dermatologists.[20] Empowering patients can improve medication adherence and, by extension, treatment success – poor adherence to and persistence with treatments are known issues in managing dermatological conditions.[43,44]

One limitation of this Delphi panel study was the potential for selection bias, which is inherent in any study based on volunteer participation. As the survey results were restricted to panellists who were willing/able to answer an online questionnaire, the findings may not be representative of the opinions of all dermatologists and patients with GPP. However, every effort was made to include a global representation of physician and patient panellists who had significant experience of treating and living with the disease. As GPP is a rare disease, statements were developed based on the limited evidence available and experts' opinions. As more data become available from clinical trials and real-world practice, the understanding of GPP and GPP-specific treatment goals will continue to evolve.

This global Delphi panel, comprising physicians and patient representatives, achieved universal consensus on treatment goals spanning the clinical course of GPP, which is chronic, heterogeneous and unpredictable in nature. The critical need for timely, multidisciplinary management to prevent life-threatening complications was highlighted as an overarching principle and supported by all panellists. Short-term treatment goals should emphasize rapid flare control, including substantial alleviation of cutaneous manifestations, preventing new pustule formation and resolution of systemic symptoms. This should be achieved by evaluation of treatment effectiveness within a week of initiation, as well as using quantifiable measures such as GPPGA pustulation subscore, and itch and skin pain NRS. Long-term treatment goals established by the panel included sustained disease control, improvement of QoL and minimizing disease activity to prevent flares, while controlling signs and symptoms between flares. These findings provide actionable and specific measures of treatment success for HCPs and patients with GPP. This is vital for informed decision-making regarding treatment options and effective monitoring throughout the disease course. Furthermore, the consensus underscores the importance of personalized treatment plans developed collaboratively between patients and HCPs, reflecting a holistic approach to GPP management that prioritizes comprehensive symptom control and functional wellbeing.

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Data availability

To ensure independent interpretation of clinical study results and enable authors to fulfil their role and obligations under the International Committee of Medical Journal Editors criteria, Boehringer Ingelheim grants all external authors access to relevant clinical study data. In adherence with the Boehringer Ingelheim Policy on Transparency and Publication of Clinical Study Data, scientific and medical researchers can request access to clinical study data, typically 1 year after the approval has been granted by major regulatory authorities or after termination of the development programme. Researchers should use the https://vivli.org/link to request access to study data and visit https://www.mystudywindow.com/msw/datasharing for further information.

Ethics statement

The study followed the applicable regulatory and ethical requirements. The study complied with module VIII of the good pharmacovigilance practices. As the study was about collection of opinions rather than healthcare data, it was technically considered market research. IQVIA Commercial GmbH & Co OHG followed the European Pharmaceutical Marketing Research Association code of conduct guidelines for the study.

Patient consent

Not applicable.

Supporting Information

Additional Supporting Information may be found in the online version of this article at the publisher's website.

References

1 Bachelez H. Pustular psoriasis: the dawn of a new era. *Acta Derm Venereol* 2020; **100**:adv00034.

2 Zheng M, Jullien D, Eyerich K. The prevalence and disease characteristics of generalized pustular psoriasis. *Am J Clin Dermatol* 2022; **23**:5–12.
3 Armstrong AW, Elston CA, Elewski BE *et al.* Generalized pustular psoriasis: a consensus statement from the National Psoriasis Foundation. *J Am Acad Dermatol* 2024; **90**:727–30.
4 Fujita H, Terui T, Hayama K *et al.* Japanese guidelines for the management and treatment of generalized pustular psoriasis: the new pathogenesis and treatment of GPP. *J Dermatol* 2018; **45**:1235–70.
5 Navarini AA, Burden AD, Capon F *et al.* European consensus statement on phenotypes of pustular psoriasis. *J Eur Acad Dermatol Venereol* 2017; **31**:1792–9.
6 Puig L, Choon SE, Gottlieb AB *et al.* Generalized pustular psoriasis: a global Delphi consensus on clinical course, diagnosis, treatment goals and disease management. *J Eur Acad Dermatol Venereol* 2023; **37**:737–52.
7 Zema CL, Valdecantos WC, Weiss J *et al.* Understanding flares in patients with generalized pustular psoriasis documented in US electronic health records. *JAMA Dermatol* 2022; **158**:1142–8.
8 Choon SE, Navarini AA, Pinter A. Clinical course and characteristics of generalized pustular psoriasis. *Am J Clin Dermatol* 2022; **23**:21–9.
9 Augustin M, Gottlieb AB, Tada Y *et al.* Generalized pustular psoriasis (GPP) continues to adversely impact quality of life between flares, as per Dermatology Life Quality Index scores: baseline data from the Effisayil 2 trial. Presented at the *American Academy of Dermatology (AAD) Annual Meeting*, San Diego, CA, USA, 8–12 March 2024; DV 43702.
10 Reisner DV, Johnsson FD, Kotowsky N *et al.* Impact of generalized pustular psoriasis from the perspective of people living with the condition: results of an online survey. *Am J Clin Dermatol* 2022; **23**:65–71.
11 Strober B, Kotowsky N, Medeiros R *et al.* Unmet medical needs in the treatment and management of generalized pustular psoriasis flares: evidence from a survey of Corrona registry dermatologists. *Dermatol Ther (Heidelb)* 2021; **11**:529–41.
12 Lebwohl M, Eyerich K, Pink A *et al.* Symptom and impact experience of adults with generalized pustular psoriasis (GPP) or palmoplantar pustulosis (PPP): an international web survey and qualitative interview study. Presented at *World Congress of Dermatology*, Singapore, 3–8 July 2023; abstr. 2324.
13 Bachelez H, Massol J, de Pouvourville G *et al.* Characterization of flares in patients with generalized pustular psoriasis: a population-based study from the French National Health Data System database (SNDS). *J Am Acad Dermatol* 2021; **85**:AB20.
14 Krueger J, Puig L, Thaçi D. Treatment options and goals for patients with generalized pustular psoriasis. *Am J Clin Dermatol* 2022; **23**:51–64.
15 Johnston A, Xing X, Wolterink L *et al.* IL-1 and IL-36 are dominant cytokines in generalized pustular psoriasis. *J Allergy Clin Immunol* 2017; **140**:109–20.
16 Strober B, Kotowsky N, Medeiros RA *et al.* Patient-reported outcomes from a large, North American-based cohort, highlight a greater disease burden for generalized pustular psoriasis vs plaque psoriasis: real-world evidence from the Corrona Psoriasis Registry. *J Am Acad Dermatol* 2021; **85**:AB108.
17 Strober B, Zimmerman TM, Lavasani L *et al.* Real-world treatment patterns in patients with generalized pustular psoriasis: results from a US claims analysis. Presented at the *American Academy of Dermatology (AAD) Annual Meeting*, San Diego, CA, USA, 8–12 March 2024.
18 Bachelez H, Barker J, Burden AD *et al.* Generalized pustular psoriasis is a disease distinct from psoriasis vulgaris: evidence and expert opinion. *Expert Rev Clin Immunol* 2022; **18**:1033–47.
19 Choon SE, van de Kerkhof P, Gudjonsson JE *et al.* International consensus definition and diagnostic criteria for generalized pustular psoriasis from the International Psoriasis Council. *JAMA Dermatol* 2024; **160**:758–68.
20 Yagi N, Sasaki T, Tada Y. Patient–dermatologist gap in perception of disease burden and treatment of generalized pustular psoriasis in Japan. *Future Rare Dis* 2023; **3**:FRD36.
21 Kodali N, Blanchard I, Kunamneni S, Lebwohl MG. Current management of generalized pustular psoriasis. *Exp Dermatol* 2023; **32**:1204–18.
22 Puig L, Fujita H, Thaçi D *et al.* Current treatments for generalized pustular psoriasis: a narrative summary of a systematic literature search. *Dermatol Ther (Heidelb)* 2024; **14**:2331–78.
23 Bachelez H, Choon S-E, Marrakchi S *et al.* Trial of spesolimab for generalized pustular psoriasis. *N Engl J Med* 2021; **385**:2431–40.
24 Morita A, Strober B, Burden AD *et al.* Efficacy and safety of subcutaneous spesolimab for the prevention of generalised pustular psoriasis flares (Effisayil 2): an international, multicentre, randomised, placebo-controlled trial. *Lancet* 2023; **402**:1541–51.
25 Strober B, Leman J, Mockenhaupt M *et al.* Unmet educational needs and clinical practice gaps in the management of generalized pustular psoriasis: global perspectives from the front line. *Dermatol Ther (Heidelb)* 2022; **12**:381–93.
26 Burden AD, Mrowietz U, Skalicky AM *et al.* Symptom experience and content validity of the Psoriasis Symptom Scale (PSS) in patients with generalized pustular psoriasis (GPP). *Dermatol Ther (Heidelb)* 2022; **12**:1367–81.
27 Choon SE, Lebwohl MG, Turki H *et al.* Clinical characteristics and outcomes of generalized pustular psoriasis flares. *Dermatology* 2023; **239**:345–54.
28 Noe MH, Wan MT, Mostaghimi A *et al.* Evaluation of a case series of patients with generalized pustular psoriasis in the United States. *JAMA Dermatol* 2022; **158**:73–8.
29 Bellinato F, Gisondi P, Marzano AV *et al.* Characteristics of patients experiencing a flare of generalized pustular psoriasis: a multicenter observational study. *Vaccines (Basel)* 2023; **11**:740.
30 Fujita H, Gooderham M, Romiti R. Diagnosis of generalized pustular psoriasis. *Am J Clin Dermatol* 2022; **23**:31–8.
31 Prinz JC, Choon SE, Griffiths CEM *et al.* Prevalence, comorbidities and mortality of generalized pustular psoriasis: a literature review. *J Eur Acad Dermatol Venereol* 2023; **37**:256–73.
32 Gooderham MJ, Van Voorhees AS, Lebwohl MG. An update on generalized pustular psoriasis. *Expert Rev Clin Immunol* 2019; **15**:907–19.
33 Rivera-Díaz R, Daudén E, Carrascosa JM *et al.* Generalized pustular psoriasis: a review on clinical characteristics, diagnosis, and treatment. *Dermatol Ther (Heidelb)* 2023; **13**:673–88.
34 Umezawa Y, Ozawa A, Kawasima T *et al.* Therapeutic guidelines for the treatment of generalized pustular psoriasis (GPP) based on a proposed classification of disease severity. *Arch Dermatol Res* 2003; **295**:S43–54.
35 Stewart CR, Algu L, Kamran R *et al.* The impact of nail psoriasis and treatment on quality of life: a systematic review. *Skin Appendage Disord* 2021; **7**:83–9.
36 Gordon KB, Lebwohl M, Barker JN *et al.* Effect of spesolimab on achieving sustained disease remission in patients with generalized pustular psoriasis: results from the Effisayil 2 study. Presented at *European Academy of Dermatology and Venereology (EADV) Congress*, Berlin, Germany, 11–15 October 2023.
37 Choon SE, De La Cruz C, Wolf P *et al.* Health-related quality of life in patients with generalized pustular psoriasis: a systematic literature review. *J Eur Acad Dermatol Venereol* 2024; **38**:265–80.
38 Viguier M, Bentayeb M, Azzi J *et al.* Generalized pustular psoriasis: a nationwide population-based study using the National Health Data System in France. *J Eur Acad Dermatol Venereol* 2024; **38**:1131–9.
39 Choon SE, Elewski BE, Fujita H *et al.* Diversity in the clinical presentation of generalized pustular psoriasis (GPP): a series of case vignettes from around the world. *Exp Dermatol* 2023; **32**:1284–95.

40 Setta-Kaffetzi N, Simpson MA, Navarini AA *et al.* AP1S3 mutations are associated with pustular psoriasis and impaired Toll-like receptor 3 trafficking. *Am J Hum Genet* 2014; **94**:790–7.
41 Wang TS, Chiu HY, Hong JB *et al.* Correlation of IL36RN mutation with different clinical features of pustular psoriasis in Chinese patients. *Arch Dermatol Res* 2016; **308**:55–63.
42 McCray AT, LeBlanc K. Patients as partners in rare disease diagnosis and research. *Yale J Biol Med* 2021; **94**:687–92.
43 Feldman SR, Bohn RL, Gao R *et al.* Poor adherence to and persistence with biologics in generalized pustular psoriasis: a claim-based study using real-world data from two large US databases. *JAAD Int* 2024; **15**:78–83.
44 Lo A, Lovell KK, Greenzaid JD *et al.* Adherence to treatment in dermatology: literature review. *JEADV Clin Pract* 2024; **3**:401–18.

Appendix 1

Partnering for Innovation and Excellence in Rare Skin Diseases (PIONEERS®) Working Group and the DELPHI Panel

Dr Gabriel Magariños (University of Salvador, Argentina), Professor Dr Adam Reich (University of Rzeszów, Austria), Dr Juliana Nakano (Santa Casa de São Paulo, Brazil), Dr Xinghua Gao (China Medical University, China), Dr Furen Zhang (Shandong Academy of Medical Science, China), Dr Mahira El Sayed (Ain Shams University, Egypt), Professor Dr Julia Welzel (Augsburg University Hospital, Germany), Professor Paolo Gisondi (University of Verona, Italy), Yuzlina Binti Che Yaacob (patient representative, Malaysia), Professor Francesca Prignano (University of Florence, Italy), Professor Yayoi Tada (Teikyo University School of Medicine, Japan), Professor Seong Jin Jo (Seoul National University College of Medicine, Korea), Professor Peter van der Kerkhof (Radboud University, Netherlands), Emmylou Casanova (Patient representative, Philippines), Dr Raquel Rivera (Complutense University of Madrid, Spain), Professor Marcus Schmitt-Egenolf (Umeå University, Sweden), Professor Tsen-Fang Tsai (National Taiwan University Hospital, Taiwan), Dr Emel Bulbul Baskan (Bursa Uludağ University, Turkey), Professor David Burden (University of Glasgow, UK), Professor Arash Mostaghimi (Brigham and Women's Hospital, Harvard Medical School, USA), Dale Reisner (patient representative, USA), Professor Kilian Eyerich (Technical University of Munich, Germany), Professor Alexandros Stratigos (Andreas Sygros Hospital, Greece), Professor Dr Lev Pavlovsky (Rabin Medical Center, Israel), Dr Delfina Guadalupe Villanueva Quintero (Center for the Care of Inflammatory Diseases, Mexico), Professor Shinichi Imafuku (Fukuoka University, Japan), Dr Jennifer Becker (University of Ottawa, Canada), Dr Azura Affandi (Hospital Kuala Lumpur, Malaysia), Dr Carolina Cortés (Centro Medico Dali, Colombia), Dr Jose Manuel Carrascosa (Universitario Germans Trias i Pujol, Spain), Dr Marni Wiseman (University of Manitoba, Canada), Professor Manuelle-Anne Viguier (Hôpital Robert Debré, France), Dr Tina Bhutani (University of California, USA).