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Consensus Document

Generalized Pustular Psoriasis: Review and Consensus of the Psoriasis Group of the Spanish Academy of Dermatology and Venereology

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ABSTRACT

Generalized pustular psoriasis (GPP) is an autoinflammatory disease characterized by primarily sterile pustules, with a widespread distribution, and flares that can be associated with life-threatening complications. Spesolimab (Spevigo®) is the only drug approved for treatment and prevention of GPP flares, and there are uncertainties that justify the development by the Psoriasis Group (GPs) of the Spanish Academy of Dermatology and Venereology (AEDV) of a Delphi consensus on the diagnosis and treatment of this rare disease. A panel of experts, starting from a literature search in PubMed (since 2014), designed a structured questionnaire with assertions that were evaluated (Likert scale from 1 to 7) by 38 members of the PWG with experience in GPP. Following two rounds, between October 2024 and January 2025, agreement (≥80% of participants) was reached on 50 out of 70 statements, including the definition of GPP outbreak, infection screening, medium- and long-term treatment goals, and criteria for initiation of maintenance treatment. This Delphi consensus is intended to support clinicians in the diagnosis and treatment of patients with PPG in our setting.

Introduction

Generalized pustular psoriasis (GPP) is a rare autoinflammatory disease characterized by the presence of primarily sterile pustules that frequently coalesce and arise on an erythematous base, with generalized distribution.[1] The prevalence of GPP varies across studies and reference populations, ranging from 1 to 180 cases per million inhabitants.[2] In Spain, the 5-year prevalence and incidence rate of GPP are estimated at 13 and 7 cases per million inhabitants, respectively.[3]

GPP follows a relapsing course, with flares that may be associated with systemic signs, laboratory abnormalities, and potentially life-threatening complications.[4] Up to 50% of patients require hospitalization during flares, with an mean duration of approximately 2 weeks.[5] In a Spanish study on GPP-related hospitalizations, the estimated incidence rate between 2010 and 2015 was 3.18 cases per million inhabitants per year, with a mean age at admission of 62.2 years; 6.1% of patients required admission to an Intensive Care Unit, and 4.8% died.[6] In a Japanese study of 1516 hospitalized patients, mortality was higher among those on corticosteroids only (9%) vs those on biologic agents (1%).[5]

The rarity of GPP limits clinicians' experience and complicates evaluation of therapeutic proposals. Although therapeutic alternatives used in moderate-to-severe plaque psoriasis have been tested in GPP, results have been inconsistent and generally inferior to those observed for approved indications.[7] The approval of spesolimab (Spevigo®) for the treatment and prevention of GPP flares has renewed interest in the disease and highlighted the need to improve dermatologists' knowledge and establish consensus-based management recommendations.[8] The objective of this work is to establish recommendations from the Psoriasis Working Group (PWG) of the Spanish Academy of Dermatology and Venereology (AEDV) for the diagnosis and treatment of GPP using a modified Delphi consensus.

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Methods

A scientific committee of 6 PWG members, based on a literature search (PubMed since 2014), designed a structured questionnaire that underwent two rounds of online consultation (between October 2024 and January 2025) with participation of PWG members who had treated at least 1 GPP patient in the preceding 5 years. Not all panelists had direct experience with spesolimab; therefore, responses to corresponding statements were based on published evidence and the collective experience of the PWG presented at annual Group meetings and published after the Delphi development.[9] Panelists evaluated the questionnaire statements using a 1–7 Likert scale with three response categories: disagree (1–2), neutral (3–5), and agree (6–7). Levels of agreement were defined as: unanimity (100% of experts rating within the same category), consensus (≥80%), or absence of consensus (<80%). Ampersand Consulting developed the web platform and served as scientific secretariat for the project, funded by the AEDV.

Results

All PWG members were invited to participate in the first Delphi round. A total of 39 experts, including the scientific committee, completed the first-round questionnaire, and 38 completed the second round. Panelists included 20 women and 18 men, representing 12 Autonomous Communities, accounting for 89% of the Spanish population. A total of 92% worked in centers with a dedicated psoriasis clinic or unit; 26% had treated fewer than 3 patients in the previous 5 years, while 32% had treated 5 or more during that period.

The 25 statements that did not reach consensus in the first round were re-submitted in the second round, either with identical wording or with changes/splitting to improve clarity and avoid ambiguity. Unanimity or consensus was achieved in 50 of 70 statements, corresponding to 52 items across 11 sections. The final Delphi consensus results are shown in Table 1.

Discussion

Definition, classification, and clinical features of GPP

Consensus was achieved for all statements in this section, aligning with recently published international guidelines and consensus documents.[10–13]

Definition of a flare

In the reviewed literature, a GPP "flare" is defined implicitly, lacking uniform criteria regarding dynamics or affected body surface area. In one clinical trial, an inclusion criterion was involvement of at least 5% body surface area with erythema and pustules.[14] The panelists' consensus matched this definition. In long-term follow-up studies, most patients (60–80%) experience a single flare, usually lasting 2–5 weeks, but one-third present persistent pustular lesions (localized, annular, or generalized) between flares despite maintenance of the therapy that resolved the flare. Most patients experience mild pustular recurrences when attempting treatment withdrawal or upon exposure to potential triggers.[4] Furthermore, consensus was reached in defining a flare as the sudden onset of grouped or confluent pustules, or an increase over days or weeks in pustule density or confluence, erythema intensity, and lesion extent/distribution, with or without systemic signs.

Diagnosis and differential diagnosis

Consensus or unanimity was achieved by panelists in all statements within this section, which is consistent with an international consensus.[11] Histopathologic examination is highly informative for diagnosing GPP, but in situations with potential life-threatening risk, treatment should not be delayed pending results.[12,13] Genetic testing is informative but not required for diagnosis.[12]

Treatment of GPP flares

Consensus was reached only regarding the indication of spesolimab (Spevigo®) for the treatment of GPP flares in adults and adolescents ≥12 years.[15] Details of the discussion are presented in the accompanying box (Table 2).[7,14,16–18]

Pre-treatment screening in GPP flares

The two statements in this section achieved consensus. Treatment with biologics or potentially immunosuppressive agents is contraindicated in patients with active infection. The role of IL-36 in defense against tuberculosis and other bacterial infections is well-established.[19] In patients with severe GPP flares, initiation of treatment should not be delayed while awaiting tuberculosis screening results, provided there is no evidence of active tuberculosis.[20]

Short-term treatment goals (1–4 weeks)

Unanimity was achieved in prioritizing rapid and sustained pustule clearance, considered the main parameter of clinical response[21]; crusts are considered residual lesions. Unlike a recently published global consensus,[21] our Delphi did not define response objectives at 2–3 days, a timeframe considered overly optimistic. Symptoms of GPP (pain, pruritus) were not included due to their subjective nature, and adnexal involvement is uncommon, although it may significantly impact patients' quality of life.

Maintenance treatment

There was consensus among the panelists on the need to personalize treatment when clinical activity of GPP persists after control of the flare, continuing whichever therapy had been effective; spesolimab is the only drug approved with this indication, and cyclosporine should be avoided. No maintenance treatments completely prevent GPP flares; 10% of patients on spesolimab at the approved dose for 48 weeks experienced a new flare (all within the first 4 weeks of treatment) vs 52% of patients on placebo.[22] There is insufficient information on the possible effect of prior therapeutic burden (e.g., maintenance with systemic or biologic treatments) regarding flare prevention with spesolimab.

Most patients with GPP have <1 flare per year[23]; therefore, maintenance therapy should be considered after the second flare or when the interval between flares is <1 year. Among patients undergoing maintenance therapy who experience a maximum of 1 intercurrent flare per year (partial effectiveness), there was consensus on the need to treat the GPP flare with IV spesolimab and switch maintenance therapy to subcutaneous spesolimab. In patients on maintenance treatment with a past medical history of >1 flare per year (treatment failure), it is recommended to change maintenance therapy to subcutaneous spesolimab without waiting for a new flare. There is still insufficient information on the response to IV spesolimab in patients who experience a flare despite subcutaneous spesolimab maintenance.

Treatment of concomitant plaque psoriasis

Consensus was reached on all statements in this section; treatment of plaque psoriasis (or localized acral pustular psoriasis, for which spesolimab has not demonstrated efficacy[24]) should be approached independently from GPP management.

Table 1
Results obtained in the Delphi rounds.

	Round	Median	%D	%U	%A	Result
A. Definition, classification, and clinical characteristics of GPP						
A1. GPP is a rare autoinflammatory disease characterized by primarily sterile pustules that frequently coalesce and arise on an erythematous base.	1	7	0	0	100	Unanimity
A2. GPP may present acutely with disseminated pustules or subacutely with an annular phenotype, with or without association with other forms of psoriasis.	1	7	0	3	97	Consensus
A3. GPP follows a relapsing course with systemic signs and laboratory abnormalities.	1	7	0	0	100	Unanimity
A4. GPP differs from psoriasis cum pustulatione* by its extent (>3% BSA) and involvement of multiple topographic areas.	2	7	3	11	87	Consensus
A5. GPP may coexist with localized acral pustular psoriasis.	2	7	0	5	95	Consensus
A6. Acral pustular lesions may be present during GPP flares and do not exclude the diagnosis.	1	7	0	8	92	Consensus
B. Definition of "flare"						
B1. In a diagnosed GPP patient, a flare is defined by grouped or confluent pustules in >1 topographic area or >3% BSA.	2	5	11	42	47	No consensus
B2a. In general, in a patient diagnosed with GPP, a flare is defined by the presence of clustered or confluent pustules in more than one topographic location, or by involvement of >5% of the BSA.	2	7	3	8	89	Consensus
B2b. In general, in a patient diagnosed with GPP, a flare is defined by the presence of clustered or confluent pustules in more than one topographic location, or by involvement of >10% of the body surface area. *Note: Given the lack of consensus on statement B1 (>3% of the surface) and observing that the percentage of agreement increased with statement B2 (>5% of the surface) in the first round, a new statement with an even higher percentage (>10% of the surface) was proposed in the second round.	2	7	13	8	79	No consensus
B3. Sudden appearance of grouped/confluent pustules (with or without systemic symptoms) in a previously stable patient indicates flare.	1	6	0	15	85	Consensus
B4. In minimal disease activity, increased pustule density, confluence, erythema, or lesion extent over days–weeks indicates flare.	1	7	0	10	90	Consensus
C. Diagnosis and differential diagnosis						
C1. Diagnosis of GPP is clinical and must be complemented with histopathology, lab tests, and microbiology to rule out alternatives.	1	7	3	3	95	Consensus
Lab tests should include	1	7	0	0	100	Unanimity
C2a. Complete blood count (leukocytosis with neutrophilia).						
C2b. ESR.	1	7	0	8	92	Consensus
C2c. Protein electrophoresis (hypoalbuminemia).	2	7	3	3	95	Consensus
C2d. Standard biochemistry including renal and hepatic function.	1	7	0	0	100	Unanimity
C2e. Electrolytes (hypocalcemia).	1	7	0	10	90	Consensus
C2f. C-reactive protein.	1	7	0	0	100	Unanimity
C2g. Urinalysis.	2	7	0	5	95	Consensus
C3. Genetic testing is useful but not necessary for the diagnosis of GPP or for initiating treatment.	1	7	0	8	92	Consensus
C4. Bacteriological and mycological cultures of pustules are useful for diagnostic confirmation or to identify superinfection, in order to rule out an infection in a febrile patient with neutrophilia.	1	7	5	13	82	Consensus
C5. Blood cultures and procalcitonin level determination are useful for diagnostic confirmation or to identify superinfection to rule out an infection in a febrile patient with neutrophilia.	2	7	0	3	97	Consensus
C5a. Blood cultures are useful for diagnostic confirmation or to identify superinfection to rule out an infection in a febrile patient with neutrophilia.	2	7	0	0	100	Unanimity
C5b. Procalcitonin level determination is useful for diagnostic confirmation or to identify superinfection to rule out an infection in a febrile patient with neutrophilia.	2	7	3	13	84	Consensus
C6. In GPP flares possibly triggered by medications, the differential diagnosis should include AGEP and paradoxical psoriasis induced by tumor necrosis factor (TNF) inhibitors and other biologic agents.	1	7	0	3	97	Consensus
D. Treatment of GPP flare						
D1. First-line therapy for the first episode while awaiting diagnostic confirmation is oral corticosteroids.	2	6	11	16	74	No consensus
D2a. First-line therapy of GPP flare is cyclosporine.	2	6	18	21	61	No consensus
D2b. First-line therapy of GPP flare is methotrexate or acitretin.	2	4	32	34	34	No consensus
D3a. The first option for treating a GPP flare in patients with plaque psoriasis is the use of biologic therapies approved for plaque psoriasis, even if used off-label.	2	5.5	13	37	50	No consensus
D3b. The first option for treating a GPP flare in patients without plaque psoriasis is the use of biologic therapies approved for plaque psoriasis, even if used off-label.	2	4	32	39	29	No consensus

Table 1
(*Continued*)

	Round	Median	%D	%U	%A	Result
D4. The first option for treating a GPP flare is spesolimab.	1	7	3	15	82	Consensus
D5. The approved option for treating a GPP flare is spesolimab.	1	7	0	3	97	Consensus
E. Pre-treatment screening						
E1. In patients for whom treatment of a GPP flare is being considered, the same baseline assessment should be performed as in patients with moderate-to-severe psoriasis who are eligible for systemic therapy.	1	7	3	13	85	Consensus
E2. In a patient with a severe GPP flare (requiring urgent management due to potential life-threatening risk), it is not necessary to wait for the result of latent tuberculosis testing (e.g., IGRA) before initiating systemic treatment. *Note: The median time to latent tuberculosis reactivation with infliximab may be around 12 weeks; therefore, delaying treatment initiation by one week is not critical. Keane J. Rheumatology 2005; 44:714–720: • IFX: Median time to diagnosis since initiation of TNFi: 12 weeks. • ADA: Median time to onset of reactivation since initiation of TNFi: 16–24 weeks. • ETN: Median time to onset of tuberculosis: 46 weeks. The aim is to determine whether the IGRA (Quantiferon) result would in any case change the treatment of the GPP flare and whether it is therefore necessary to wait for the result or not.	2	7	3	5	92	Consensus
F. Short-term (1–4 weeks) treatment goals						
F1. Achieve rapid and sustained clearance of erythema.	2	7	0	5	95	Consensus
F2. Achieve rapid and sustained clearance of crusts.	2	6	13	16	71	No consensus
F3. Achieve rapid and sustained clearance of pustules.	1	7	0	0	100	Unanimity
F4. Rapid relief of systemic symptoms.	1	7	0	3	97	Consensus
G. Maintenance treatment						
G1. Decision to start maintenance therapy must be individualized based on evolution after first flare.	1	7	0	8	92	Consensus
G2. Maintenance therapy should be considered when clinical activity persists.	1	7	0	3	97	Consensus
G3. If a biologic controls a flare, it may be continued as maintenance, with periodic assessment.	1	7	3	3	95	Consensus
G4. Acitretin is the best maintenance option.	2	5	13	50	37	No consensus
G5. Cyclosporine is the best maintenance option.	2	2	84	8	8	Consensus (disagreement)
G6. Methotrexate is the best maintenance option.	2	3	47	39	13	No consensus
G7. The best option for maintenance therapy is the use of biologic treatments approved for plaque psoriasis.	2	6	8	26	66	No consensus
G8. The best option for maintenance therapy is treatment with spesolimab.	2	7	0	5	95	Consensus
G9a. In patients on maintenance therapy who experience a flare, the flare should be treated with IV spesolimab, and the maintenance treatment should be switched to an alternative therapeutic option.	2	6	5	18	76	No consensus
G9b. In patients on maintenance therapy who experience a flare, the flare should be treated with IV spesolimab, and the maintenance treatment should be switched to subcutaneous spesolimab.	2	7	0	16	84	Consensus
G9c. In patients on maintenance therapy who experience a flare, the flare should be treated with IV spesolimab, and the current maintenance therapy should be continued.	2	6	18	16	66	No consensus
G10. In patients on maintenance therapy who have experienced >1 flare per year, it is recommended to switch to subcutaneous spesolimab (with induction), without waiting for another flare.	2	7	0	13	87	Consensus
G11. In patients on maintenance therapy who have experienced >2 flares per year, it is recommended to switch to subcutaneous spesolimab (with induction), without waiting for another flare.	1	7	0	13	87	Consensus
G12. In patients on maintenance therapy with <1 flare per year, it may be preferable to wait for a flare to occur, treating it with IV spesolimab, followed by maintenance therapy with subcutaneous spesolimab (without induction).	2	7	0	5	95	Consensus
G13. Maintenance therapy should control GPP and prevent flares.	1	7	3	0	97	Consensus
H. Treatment of concomitant plaque psoriasis						
H1. The presence of plaque psoriasis does not alter the response of GPP to treatment with spesolimab, but there are no data regarding the efficacy of this treatment on plaque psoriasis.	1	6	0	15	85	Consensus
H2. Treatment of concomitant plaque psoriasis (or localized acral pustular psoriasis) should be appropriate to the severity of that disease, regardless of the activity of GPP.	2	7	3	0	97	Consensus
H3. When combined treatment is required due to the coexistence of plaque psoriasis or localized acral pustular psoriasis with GPP, safety considerations (risk of infections, comorbidities, or patient frailty) should be given particular attention.	1	7	0	3	97	Consensus

Table 1
(*Continued*)

	Round	Median	%D	%U	%A	Result
I. Long-term treatment goals						
I1. Absence of pustules.	1	7	0	0	100	Unanimity
I2. Absence of erythema.	1	7	0	5	95	Consensus
I3. Complete absence of lesions.	1	7	0	13	97	Consensus
I4. Prevention of flares.	1	7	0	3	97	Consensus
I5. In patients with comorbidities, multidisciplinary management is necessary.	1	7	3	8	90	Consensus
J. Immunogenicity						
J1. Based on current evidence, immunogenicity affects long-term efficacy/safety of intermittent spesolimab.	2	6	8	21	71	No consensus
K. Special populations						
K1. In pregnant women, biologic use should be restricted to 1st and 2nd trimesters.	2	7	8	8	84	Consensus
K2. Biologic therapy during the 3rd trimester requires delaying live vaccines in newborns until 6 months.	1	7	0	5	95	Consensus
K3a. GPP flares in children <12 should be treated with psoriasis-approved treatments.	2	6	3	18	79	No consensus
K3b. GPP flares in children <12 should be treated with off-label spesolimab.	2	5	26	26	47	No consensus

%D, % disagree; %U, % undecisive; %A, % agree; SC, subcutaneous; IV, intravenous; AGEP, acute generalized exanthematous pustulosis; BSA, body surface area; IGRA, interferon-gamma release assay; PPG, generalized pustular psoriasis; ESR, erythrocyte sedimentation rate.

Table 2
Panelists' comments and discussion on the treatment of GPP flares.

In the first episode of the disease, while awaiting diagnostic confirmation, oral corticosteroids (prednisone 0.5–1.0 mg/kg/day or equivalent) may be used; however, this statement did not reach consensus (74% agreement) as the first-line treatment option. Consensus was also not achieved either on the use of biologic agents approved for plaque psoriasis and (in some countries) for the treatment of GPP flares[7]; other systemic therapies are not recommended due to safety concerns[16] or the slow onset of potential therapeutic effects. In any case, most published studies correspond to case series or open-label trials, without standardized outcome measures.[16]
The concept of "flare" is not specifically defined in the prescribing information for spesolimab, but in the EFFISAYIL-1 clinical trial, inclusion criteria required a moderate-to-severe flare, defined as a total GPPGA score ≥3, the appearance or worsening of pustules with a pustule subscore ≥2, and ≥5% body surface area with erythema and pustules.[14] In Spain, reimbursement is restricted to patients with a global GPPGA score ≥3 at treatment initiation.[17] For the treatment of GPP flares, spesolimab is administered as a 900 mg intravenous infusion, which may be repeated after 1 week "if flare symptoms persist".[15] In the clinical trial, a second 900 mg dose could be given on day 8 when both the global GPPGA and the pustule subscore were ≥2.[14]
In the EFFISAYIL-1 study, 19 of 35 patients on spesolimab had no pustules one week after infusion,[14] and 21 patients were free of pustules at week 12: 15 of the 23 who received only 1 dose, and 6 of the 12 who received a 2nd dose 1 week later.[18] Six of the 53 patients included in the study experienced a new flare during the 12-week follow-up; 5 of these were treated with an additional infusion of spesolimab, achieving complete clearance of pustules.[18]

Long-term treatment goals

There was broad consensus or unanimity in defining the long-term goals as the complete absence of GPP elementary lesions and the prevention of flares.

Immunogenicity

Consensus was not achieved in this section. When biologic therapies are used, the immunogenicity of the drug, depending on route of administration, may affect maintenance of efficacy, particularly in the context of intermittent or on-demand treatment.[25] In the case of spesolimab, anti-drug antibodies have been detected in 46% of patients who received at least 1 IV dose, with a median onset of 2.3 weeks.[14] Although significant reductions in plasma spesolimab concentrations have been detected in patients with ADA titers ≥4000, the potential impact on clinical safety and efficacy remains unclear.[26,27] There are also insufficient data regarding the subcutaneous route.

Use in special populations

There was consensus to avoid the use of biologic therapy (including spesolimab) during pregnancy and to restrict its use – when necessary due to disease severity – to the 1st and 2nd trimesters.

No consensus was reached regarding treatment in pediatric patients up to 12 years of age, although most panelists (79%) favored the use of psoriasis biologics with pediatric indications rather than off-label spesolimab.

Conclusions

This document presents the Psoriasis Working Group's (PSW) consensus, based on a Delphi process, on the diagnosis, differential diagnosis, clinical course, and treatment of GPP.

Consensus was achieved on the definition of a GPP flare based on body surface area affected or changes in the intensity of clinical signs.

Screening for infections and comorbidities prior to treatment of GPP should follow the same protocol as in moderate-to-severe plaque psoriasis. Active infections and latent tuberculosis should be ruled out, but potential treatment of latent tuberculosis should not delay initiation of GPP therapy. Treatment with spesolimab in pregnant women and pediatric patients outside of approved indications should be individualized.

Spesolimab is the only approved maintenance treatment for GPP; this consensus defined the criteria for its initiation.

This study presents limitations inherent to the Delphi methodology, in which consensus relies primarily on the experience and knowledge of participants – who in this case consisted of many hospital-based specialists representing 12 Autonomous Communities. Application of the consensus recommendations must be individualized according to each patient's context and potential differences in health care system access.

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Conflicts of interest

LP has participated in clinical trials and/or received consulting fees or conference sponsorships from AbbVie, Almirall, Amgen, Biogen, Boehringer Ingelheim, Bristol Myers Squibb, Fresenius-Kabi, Horizon, J&J Innovative Medicine, Leo-Pharma, Lilly, Novartis, Pfizer, Samsung-Bioepis, STADA, Sun-Pharma, and UCB.

J-M C has participated in clinical trials and/or received consulting fees or conference sponsorships from AbbVie, Almirall, Amgen, Biogen, Boehringer Ingelheim, Bristol Myers Squibb, Galderma, J&J Innovative Medicine, Leo-Pharma, Lilly, Novartis, Pfizer, Sanofi, Sandoz, and UCB.

RR has received honoraria for advisory roles, lectures, and/or has participated in sponsored clinical trials for: AbbVie, Almirall, Amgen, Boehringer Ingelheim, Bristol Myers Squibb, Incyte, Janssen, LEO Pharma, Lilly, Novartis, Pfizer, Sandoz, and UCB.

EV has participated in clinical trials and/or received consulting fees or conference sponsorships from AbbVie, Acelryn, Almirall, Amgen, Biofrontera, Boehringer Ingelheim, Bristol Myers Squibb, Galderma, Incyte, J&J Innovative Medicine, Leo-Pharma, Lilly, Moonlake, Novartis, Pfizer, Sandoz, and UCB.

PC has served as advisor and/or investigator and/or speaker for the following pharmaceutical companies: AbbVie, Almirall, Amgen, Astellas, Beiersdorf, Biogen, BMS, Boehringer, Celgene, Gebro, Johnson & Johnson, LEO Pharma, Lilly, MSD, Novartis, Pfizer, Roche, Sandoz, Sanofi, SVR, Takeda, and UCB.

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