

Global consensus on the clinical course, treatment and management of generalized pustular psoriasis (GPP)

Lluís Puig¹, Siew Eng Choon², Alice B. Gottlieb³, Slaheddine Marrakchi⁴, Jörg C. Prinz⁵, Ricardo Romiti⁶, Yayoi Tada⁷, Dorothea von Bredow⁸, Melinda Gooderham⁹

¹Department of Dermatology, Hospital de la Santa Creu I Sant Pau, Barcelona, Spain; ²Department of Dermatology, Hospital Sultanah Aminah, and Clinical School Johor Bahru, Monash University Malaysia, Subang Jaya, Malaysia; ³Icahn School of Medicine at Mount Sinai, New York, NY, USA; ⁴Department of Dermatology, Hedi Chaker Hospital, University of Sfax, Sfax, Tunisia; ⁵Department of Dermatology and Allergy, Ludwig-Maximilian-University Munich, Munich, Germany; ⁶Department of Dermatology, University of São Paulo, São Paulo, Brazil; ⁷Department of Dermatology, Teikyo University School of Medicine, Tokyo, Japan; ⁸IQVIA Commercial GmbH & Co., Munich, Germany; ⁹School of Medicine, Queen's University and Centre for Dermatology and Probiotic Medical Research, Peterborough, ON, Canada

Using a Delphi panel approach, we have established global consensus on the clinical course, diagnosis, treatment goals and management of GPP; the evidence-based algorithm we have subsequently developed will provide much needed guidance for physicians to implement in clinical practice

PURPOSE

To conduct a Delphi panel study to gain advanced insights into the clinical course, diagnosis, treatment goals and management of GPP.

INTRODUCTION

- GPP is a rare, neutrophilic skin disease, with a prevalence ranging from 0.02–1.4 per 10,000 people worldwide^{1–5}
- ERASPEN and JDA have published guidelines for the classification and diagnosis of GPP, respectively;^{1,2} however, the evidence base for these guidelines is limited
- Few clinical trials have been conducted in GPP due to the rarity of the disease and lack of international consensus on criteria for diagnosis and treatment goals
- As a result, there is a general paucity of information to inform optimal management of patients with GPP

CONCLUSIONS

- Global consensus among expert dermatologists was reached on:
 - Key clinical and histological features supporting GPP diagnosis and flare definition
 - GPP being distinct from plaque psoriasis, although both conditions may occur in the same patient
 - Treatment goals of rapid, sustained control of cutaneous and systemic symptoms, and long-term prevention of new flares
 - Multidisciplinary disease management and assessment tools for monitoring disease severity in clinical practice

METHODS

- An SLR was conducted to identify published literature and develop statements for four key domains of GPP:
 - Clinical course and flare definition
 - Diagnosis
 - Treatment goals
 - Holistic management of GPP
- The Delphi panel comprised 21 expert dermatologists
- Statements were rated on a Likert scale (1 [strong disagreement] to 7 [strong agreement]); consensus was reached when statements were agreed on by ≥80% of panellists



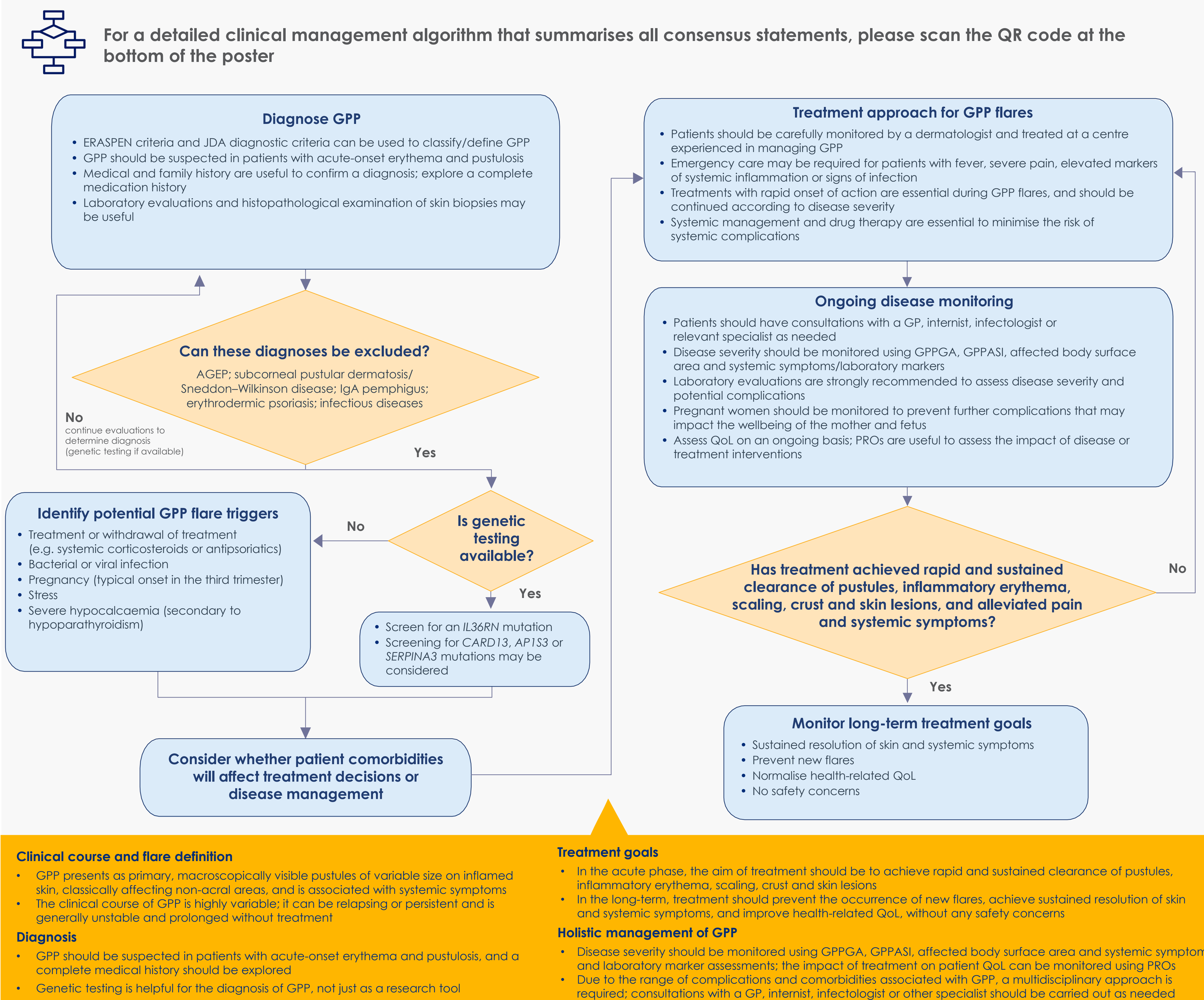
RESULTS

Consensus after two Delphi panel rounds

Domain/subdomain	Statements, n	Agreement, n (%)
Round 1		
Round 1 total	185	141 (76.2)
Domain 1: Clinical course and flare definition		
GPP definition/classification	21	16 (76.2)
Flare definition and GPP clinical course	9	9 (100.0)
Potential triggers and disposing factors	27	13 (48.1)
Prognosis	24	23 (95.8)
Domain 1 total	81	61 (75.3)
Domain 2: Diagnosis		
Criteria	2	2 (100.0)
Clinical diagnosis of GPP	3	3 (100.0)
Laboratory tests relevant for the diagnosis of GPP	15	9 (60.0)
Genetic screening in GPP diagnosis	2	1 (50.0)
Histopathologic features of GPP	5	4 (80.0)
Differential diagnosis	14	5 (35.7)
Domain 2 total	41	24 (58.5)
Domain 3: Treatment goals		
Flare/acute-phase treatment goals	9	9 (100.0)
Long-term goals	8	8 (100.0)
Domain 3 total	17	17 (100.0)
Domain 4: Holistic management of GPP		
Domain 4 total	46	39 (84.8)
Round 2		
Round 2 total	28	16 (57.1)
Domain 1: Clinical course and flare definition		
GPP definition/classification	3	0
Potential triggers and disposing factors	2	0
Domain 1 total	5	0
Domain 2: Diagnosis		
Laboratory tests relevant for the diagnosis of GPP	3	1 (33.3)
Genetic screening in GPP diagnosis	2	2 (100.0)
Histopathologic features of GPP	7	7 (100.0)
Differential diagnosis	2	0
Domain 2 total	14	10 (71.4)
Domain 4: Holistic management of GPP		
Domain 4 total	9	6 (66.7)
Rounds 1 and 2	Total	213
		157 (73.7)

- Overall, dermatologists reached consensus on 73.7% of statements, and these formed the basis of the clinical management algorithm
- All dermatologists reached consensus on statements on treatment goals, and high levels of agreement were reported for statements on holistic management
- More evidence is needed in areas with low consensus, such as potential triggers and disposing factors, laboratory tests relevant for the diagnosis of GPP and differential diagnoses

Clinical management flow diagram for GPP based on consensus statements



Abbreviations
 AGEP, acute generalized exanthematous pustulosis; ERASPEN, European Rare and Severe Psoriasis Expert Network; GP, general practitioner; GPP, generalized pustular psoriasis; GPPASI, Generalized Pustular Psoriasis Area and Severity Index; GPPGA, Generalized Pustular Psoriasis Global Assessment; Ig, immunoglobulin; JDA, Japanese Dermatological Association; PRO, patient-reported outcome; QoL, quality of life; SLR, systematic literature review.

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