

# Characteristics and management of generalized pustular psoriasis (GPP): Experience from the Central and Eastern Europe (CEE) GPP Expert Network

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**Our study, based in the CEE region, highlights the substantial clinical burden associated with GPP and the high unmet need for treatments that provide rapid and sustained skin clearance**

## PURPOSE

To determine the characteristics of GPP flares and explore the current treatment landscape among patients with GPP in the CEE region.

## INTRODUCTION

- GPP is a rare, potentially life-threatening, autoinflammatory skin disease, characterised by sudden flares of widespread sterile pustules on non-acral skin that can occur with or without systemic inflammation<sup>1-5</sup>
- Symptoms include pain, itching, fever and fatigue, all of which can severely affect patient quality of life, and patients often require treatment between flares to manage persistent skin lesions<sup>3, 6-9</sup>
- Current treatment options include cyclosporine, retinoids, methotrexate and biologics, but there are no GPP flare-specific treatments approved in the USA or Europe<sup>3, 11</sup>
- Approved biologics in Japan include anti-TNF- $\alpha$  therapies, IL-17/IL-17R inhibitors and IL-23 inhibitors; however, supporting evidence is weak, based on open-label, single-arm trials in small patient populations<sup>11, 12</sup>

## CONCLUSIONS

- During GPP flares, patients experienced moderate-to-severe cutaneous symptoms that affected at least 50% of the body, often accompanied by fever and other systemic symptoms; >75% of patients required treatment in hospital
- Although some current treatments may be effective in resolving GPP flares, flare resolution was often slow; these results highlight the high unmet need for treatments that provide rapid and sustained skin clearance
- Currently available biologics and investigational anti-IL-36R agents showed promising efficacy; data from additional patients are needed to fully understand their therapeutic potential

## METHODS

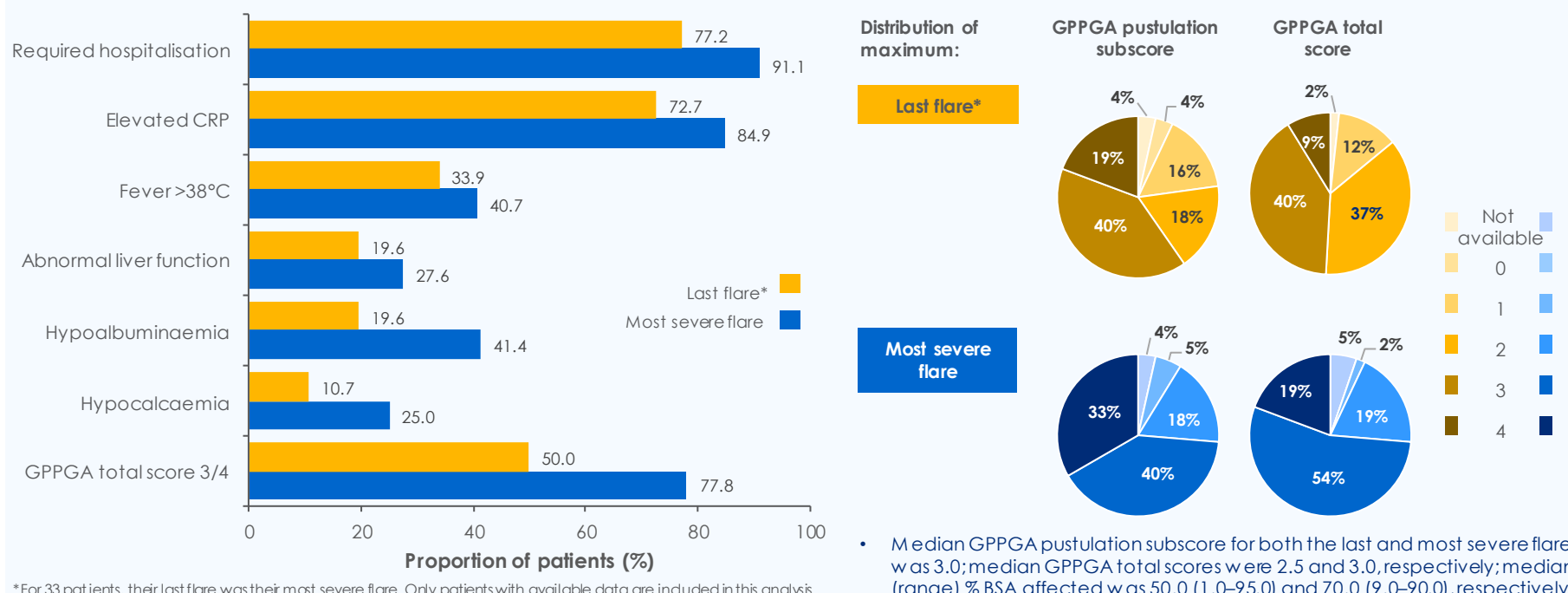
- Eligible cases (N=57) that met the GPP diagnosis criteria, as defined by the centre of excellence or ERASPEN, and had follow-up or treatment in the last 10 years were included from 10 centres across the CEE region
- Investigators used CRFs to collect patient demographics; clinical characteristics of the last and most severe flares; and past treatments and their respective outcomes
- Overview of CRF:**
  - Patient demographics at last observation, including concomitant diseases
  - GPP history: duration of GPP, number of flares since diagnosis, number of flares requiring hospitalisation or ICU admission and trigger factors
  - Clinical characteristics of the (1) last flare and (2) most severe flare, including:
    - Flare duration; hospitalisation/ICU care required; GPPGA component scores; total BSA affected; systemic inflammation symptoms; treatments received and duration of treatment; and time from treatment initiation to flare resolution

## RESULTS

### Baseline demographics and medical history

Characteristic	Patients (N=57)
Female, n (%)	34 (59.6)
Median age, years (range)	63 (16-92)
Median age at disease onset, years (range)	57 (0-84)
Presence/history of plaque psoriasis, n (%)	33 (57.9)
Presence/history of psoriatic arthritis, n (%)	14 (24.6)
Median disease duration, years (range)	8 (0.1-51)
Median total number of flares (range)	3 (1-25)
Median annual flare frequency	0.7
<b>Comorbidities, n (%)</b>	
Hypertension	27 (47.4)
Hyperlipidaemia	20 (35.1)
Cardiovascular disease	15 (26.3)
Diabetes	11 (19.3)
Obesity	9 (15.8)
Depression	9 (15.8)
Hypothyroidism	7 (12.3)
Hyperuricaemia	7 (12.3)
Chronic kidney disease	5 (8.8)
History of hepatitis B	4 (7.0)

### Clinical characteristics of last and most severe flares



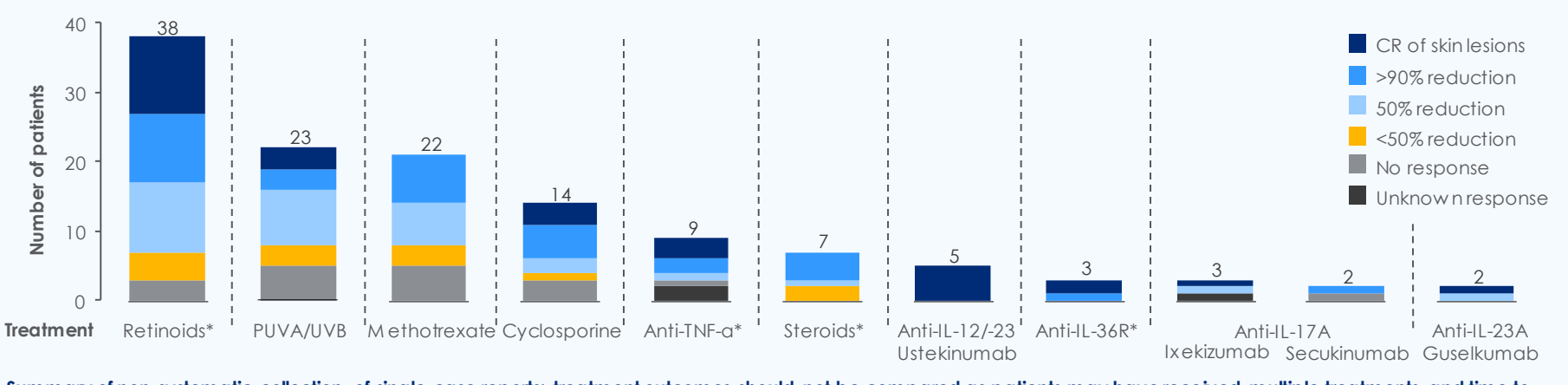
Median age was 63 years and median disease duration was 8 years; 57.9% of patients had presence/history of plaque psoriasis

Compared with the last flare, the most severe flare typically had more severe cutaneous symptoms affecting a higher %BSA and was more frequently accompanied by systemic symptoms; however, disease severity was also high for the last flare, and this resulted in >75% of patients requiring hospitalisation

### Outcomes for all treatments received for GPP, either as single agents, sequentially and/or in combination

Parameter	Last flare*	Most severe flare
Median duration of flare, weeks (range)	4.0 (1.0-48.0)	6.0 (1.0-30.0)
Median duration of hospitalisation, weeks (range)	2.0 (0.1-4.0)	2.0 (0.1-28.0)
Median time from treatment initiation to flare resolution, weeks (range)	4.0 (0.1-52.0)	4.0 (1.0-52.0)

\*For 33 patients, their last flare was their most severe flare. Only patients with available data are included in this analysis.



Summary of non-systematic collection of single case reports; treatment outcomes should not be compared as patients may have received multiple treatments, and time to achieve skin clearance and duration of response are not considered.

\*Retinoids: acitretin or isotretinoin; anti-TNF- $\alpha$ : adalimumab, etanercept, or infliximab; steroids: methylprednisolone or prednisone; anti-IL-36R: spesolimab or imsiglimab.

Median flare duration was typically longer for the most severe flare than the last flare; median time from treatment initiation to flare resolution varied widely

Treatment with retinoids, PUVA/UVB and methotrexate completely resolved or reduced skin lesions by >90% in 55.3%, 26.1% and 31.8% of cases, respectively; anti-TNF- $\alpha$  agents and other biologics were effective in 33.3-100% of cases, with individual therapies used in 2-5 patients each

**Abbreviations**  
BSA, body surface area; CEE, Central and Eastern Europe; CR, complete resolution; CRF, case report form; CRP, C-reactive protein; ERASPEN, European Rare and Severe Psoriasis Expert Network; GPP, generalized pustular psoriasis; GPPGA, Generalized Pustular Psoriasis Physician Global Assessment; ICD, International Classification of Diseases; IL, interleukin; IL-17, interleukin-17 receptor; IL-36R, interleukin-36 receptor; PUVA, psoralen and ultraviolet A; TNF, tumour necrosis factor; UVB, ultraviolet B.

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