

# Characteristics and Treatment of Generalized Pustular Psoriasis Flares in the US

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## BACKGROUND & STUDY OBJECTIVES

- Generalized pustular psoriasis (GPP) is a rare, but severe multisystem, chronic inflammatory disease characterized by sudden and widespread eruption of sterile pustules of varying severity.<sup>1-3</sup>
- Pustules during and after a GPP flare can often last for many weeks and can lead to life-threatening complications often requiring emergency care.<sup>1-4</sup>
- The most recent estimate of mortality is 4.2% from a nationwide study of hospitalized patients with GPP in Japan.<sup>5</sup>
- There is growing evidence focused on understanding the overall disease burden and treatment patterns in patients with GPP; however, there is minimal research documenting characteristics of GPP flares.<sup>4,6</sup>
- The objective of this study is to characterize flare episodes in GPP and their treatment.

## METHODS

- This retrospective descriptive study included adult patients with GPP (ICD-10 code L40.1) identified in the Optum<sup>®</sup> de-identified electronic health record (EHR) data between 1 July 2015-30 June 2020.
- The index GPP diagnosis was the first occurrence in the EHR with no coded history of GPP for at least six months prior.
- Only patients with at least 12 months of health care activity documented in the EHR after the index diagnosis and with notes available in the EHR were included in the study.
- EHR "terms" refer to signs, diseases, and symptoms (SDS) documented, and "attributes" are descriptions of the SDS terms. Categories were developed to capture like words of the same concept. Each health care visit beginning on or after the index date of GPP diagnosis with documented notes that met at least one of the following criteria were considered a documented flare in the EHR.
  - For visits with a primary diagnosis of GPP (ICD-10 code L40.1):
    - An encounter with place of service = 'EMERGENCY PATIENT', 'OBSERVATION PATIENT', 'INPATIENT', or 'URGENT'.
    - Any term in the "Flare" category
    - Any term in the "Pustule/Lesion" category plus any flare attribute
    - Any term in the "Rash" category plus any flare attribute
    - Any term in the "Other GPP symptoms" category plus any flare attribute
  - For dermatology-related visits (i.e., type of provider was a dermatologist or primary reason for visit was any skin-related diagnosis defined by ICD-10 L\* or R2\* code):
    - Any term in the "Flare" category AND any term in the "Pustule/Lesion" category
    - Any term in the "Pustule/Lesion" category plus any flare attribute
- Flare episodes were defined as consecutive days that a flare was documented in the EHR and were characterized by the frequency of occurrence per patient, the setting of care where they were identified, the type of specialist managing the episode, associated symptoms, and the treatments before, during, and after the episode.

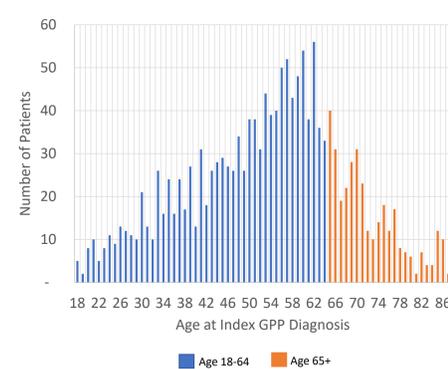
## RESULTS

- Of the 48.6 million patients with EHR notes available, 1,535 patients with GPP were identified, and 271 patients had at least one flare episode documented in their EHR and accounted for a total of 513 flare episodes during the study period. (Patients' characteristics - Table 1, Figure 1)

TABLE 1 – Race and Ethnicity of Patients

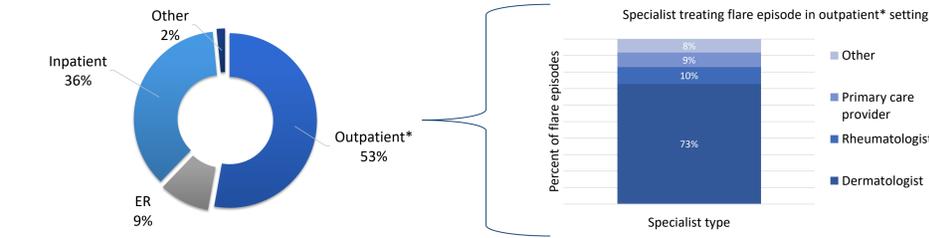
Race	Ethnicity	# Patients	% Patients
African American	Hispanic	4	0.3%
	Non-Hispanic	115	7.5%
	Unknown	3	0.2%
Asian	Hispanic	3	0.2%
	Non-Hispanic	23	1.5%
	Unknown	2	0.1%
Caucasian	Hispanic	47	3.1%
	Non-Hispanic	1,181	76.9%
	Unknown	55	3.6%
Other/Unknown	Hispanic	31	2.0%
	Non-Hispanic	40	2.6%
	Unknown	31	2.0%

FIGURE 1 – Number of Patients at Age of Index GPP Diagnosis



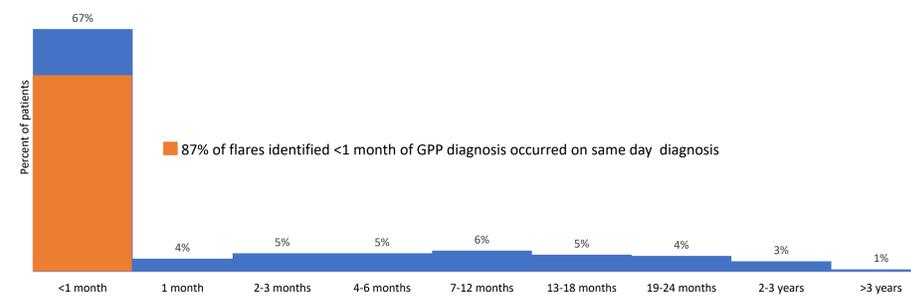
- Half of GPP flares were identified in the outpatient setting (53%), followed by the inpatient setting (36%), and ER (9%) (Figure 2)
- In the outpatient setting, most flares were treated by dermatologists (73%), followed by rheumatologists (10%) and primary care providers (9%) (Figure 2)

FIGURE 2 – Flare Episodes Identified Inpatient, Outpatient, and ER Settings



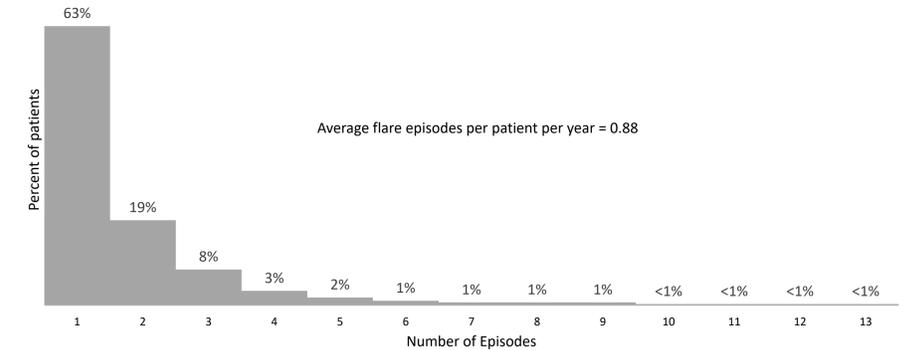
- Majority of flares occurred within one month of initial GPP diagnosis. (Figure 3)
- Over half of the flare episodes (58.3%) occurred on the same day as the index GPP diagnosis indicating that they were likely seeking care for a new GPP diagnosis based on a flare. (Figure 3)

FIGURE 3 – Flares Occurring After Initial GPP Diagnosis



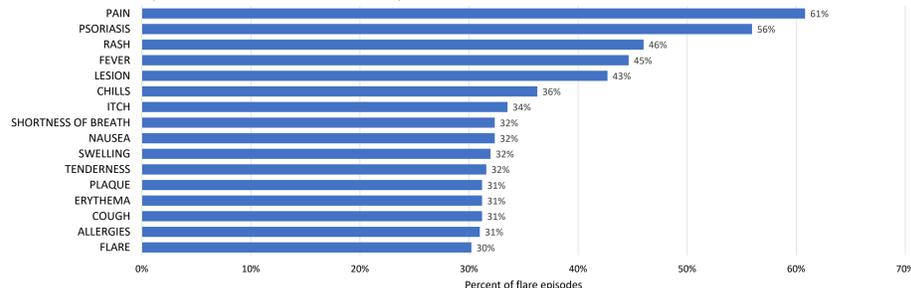
- Majority experienced one flare episode during follow-up (average follow up was 724.8 days [median=698 days]) (Figure 4)

FIGURE 4 – Flare Episodes Anytime After Initial GPP Diagnosis



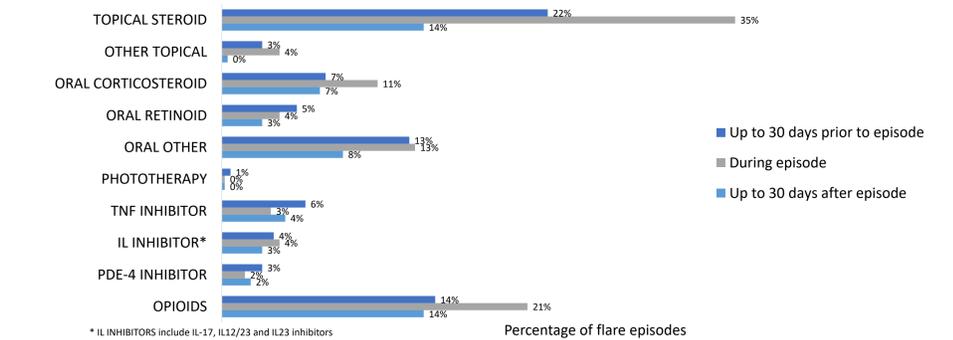
- The most common signs/symptoms documented during flare episodes were pain (61% of flare episodes) followed by rash (46%), and fever (45%). (Figure 5)

FIGURE 5 – Commonly Used Terms Documented in a Flare Episode



- The most common dermatologic treatment during a flare episode was topical steroids (35% of flare episodes) followed by other oral treatments, such as methotrexate, cyclosporine, and tacrolimus (13%), and oral corticosteroids (11%). Opioids were prescribed during 21% of flare episodes. (Figure 6)
- One-quarter of all flare episodes had no dermatologic treatment 30 days before, during or 30 days after a flare episode. (Figure 6)

FIGURE 6 – Common Treatments During Flare Episodes Documented in the EHR



## CONCLUSIONS

- There is significant unmet need for the treatment of GPP flares as evidenced by patients seeking treatment in inpatient and ER settings as well as the lack of advanced treatments beyond topical steroids.
  - Over one-third of GPP flares were identified in an acute care setting.
  - Most common treatment during a flare episode was topical steroids, and one-quarter of episodes had no documented dermatological treatment.
  - Treatment with opioids was common during flare episodes indicating the need for pain management.

## LIMITATIONS

- Due to the rarity and lack of awareness of GPP, it is likely that additional patients with true GPP remain undiagnosed or are miscoded with another form of psoriasis. In order to ensure that the study captured patients with GPP, only patients coded with a GPP diagnosis were included.
- The algorithm to identify GPP flares was intentionally conservatively designed to ensure that only flares due to GPP were identified. Thus, it is likely that the methodology underestimates the number of GPP episodes.
- Patient surveys suggest that patients do not always seek medical treatment for flares. This study only identifies patients with GPP flares documented in their EHR and underestimates the true number of patients with GPP who flare and the number of flare episodes. Also, documented flare episodes are likely to be higher severity than those not documented.
- In the EHR data, the treatments are based on prescriptions written or administered in the office/facility. Written prescriptions may not represent whether the prescription was filled and/or actually taken by the patient.
- Duration of flare episodes cannot be accurately determined in EHR data.

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