

Prevalence of generalized pustular psoriasis in the USA: Results from multiple real-world databases

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Introduction

- Generalized pustular psoriasis (GPP) is a rare, neutrophilic skin disease characterised by episodes of widespread eruption of sterile, macroscopic pustules that can occur with or without systemic inflammation and with or without plaque psoriasis^{1,2}
 - The clinical course of GPP can be either relapsing, with recurrent flares and phases of skin clearance, or persistent mild symptoms with intermittent flares of increased severity^{3,4}
- The overall prevalence of GPP in the US is not well characterised⁵
- In 2015, a GPP-specific diagnosis code was included in the International Classification of Diseases 10th revision (ICD-10) to aid identification of GPP in claims databases
- In this study, GPP prevalence was assessed in two US claims databases, including prevalence by age and sex
- Prevalence of GPP in other geographical regions was also assessed using applicable databases

Study objectives

- To estimate the prevalence of generalized pustular psoriasis (GPP) in patients from two large US administrative claims databases in 2018 and 2019, respectively
- To understand the variability of GPP prevalence, prevalence was also assessed in Japan and Germany

Methods

- This study determined the prevalence of GPP in patients enrolled in the Optum[®] Clinformatics[™] Data Mart Research Database in 2019 (using data collected from 1 January 2019 to 31 December 2019) and the IBM[®] MarketScan[®] Research Databases in 2018 (using data collected from 1 January 2018 to 31 December 2018) (Figure 1)
- Patients were identified as having GPP if they had one inpatient or one outpatient ICD-10 diagnostic code of L40.1
 - An additional more stringent definition, including one inpatient or two outpatient codes (separated by 30–365 days), was used to enhance sensitivity
- Prevalence was calculated by dividing the number of patients with GPP in the respective database by the total number of patients enrolled in the study year (with continuous enrolment for 365 days; gaps of up to 30 days were permitted)
- Analyses were performed by Instant Health Data
- Prevalence of GPP in Japan (in 2018) using the Japanese Medical Data Center (JMDC) and the Medical Data Vision (MDV) databases was estimated (data were collected from 1 January 2015 to 31 August 2018) (Figure 1)
 - In the JMDC and MDV databases, patients with GPP were identified if they had at least one confirmed ICD-10 diagnosis code of L40.1 (inpatient or outpatient)
- Using the IQVIA[®] German EMR patient record database (data collected between 1 October 2019 and 30 September 2020), the prevalence of GPP in the total population of Germany was extrapolated (Figure 1)
- Overall, prevalence of patients with GPP in Germany was calculated as the number of patients with at least one confirmed diagnosis code of L40.1 during the study period divided by the total population of Germany on 31 December 2019

Results

Patient population and demographics

- In total, 1212 patients were identified in the Optum[®] Clinformatics[™] database and 1383 patients were identified in the IBM[®] MarketScan[®] Research database (Figure 2)

Figure 1. Overall study design

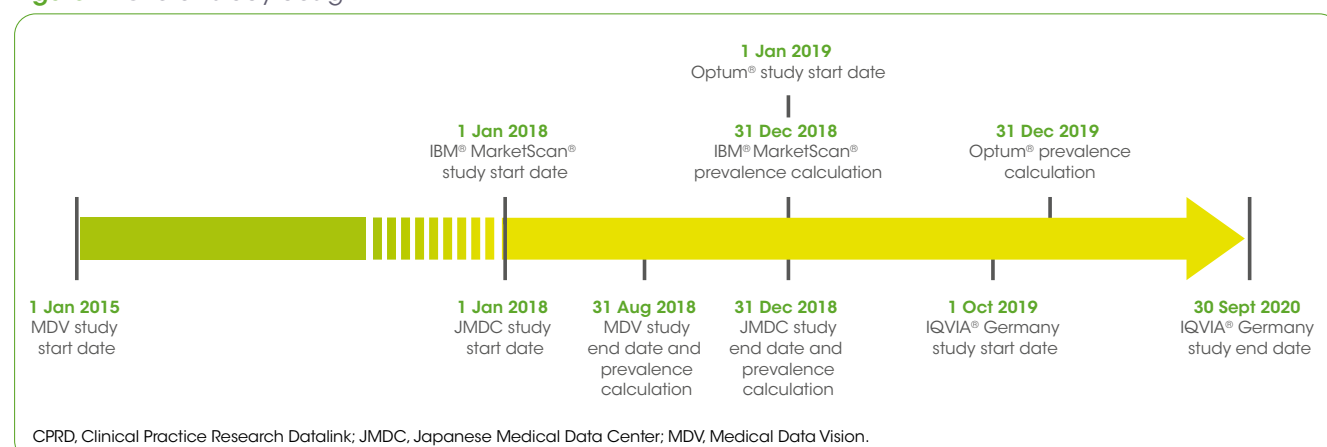
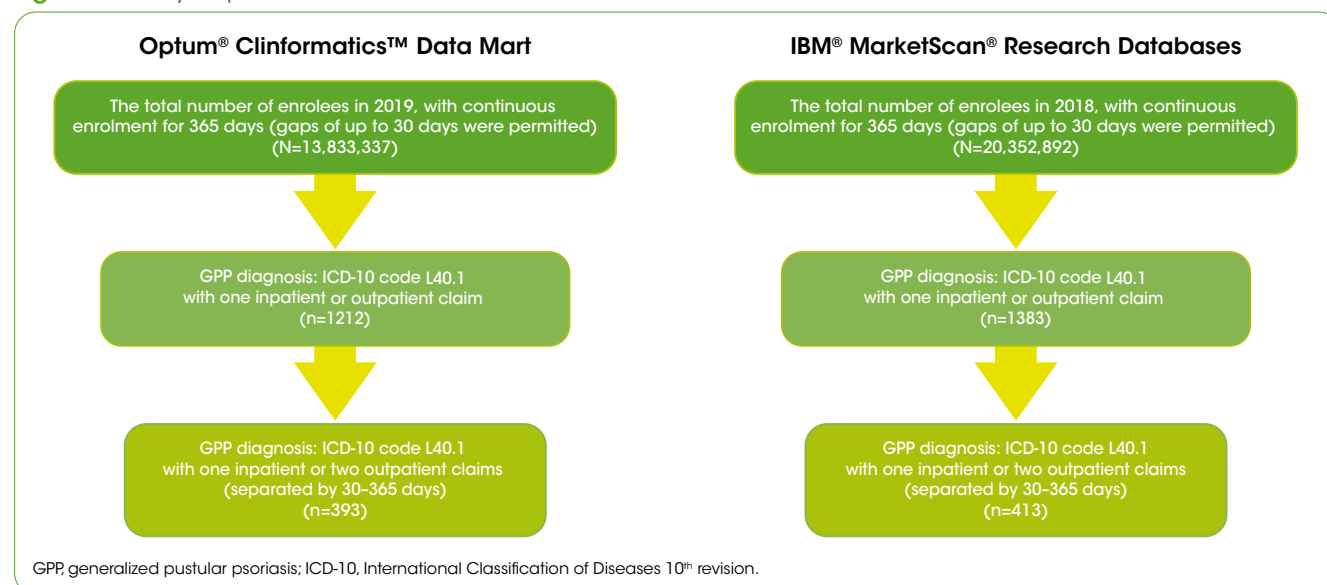


Figure 2. Study disposition



- In the Optum[®] and IBM[®] MarketScan[®] studies, patients with GPP were more likely to be female (64.7% and 61.2%, respectively; Table 1)

Table 1. Patient demographics

| | Patients with GPP in the Optum [®] Clinformatics [™] Data Mart database (N=1212) | Patients with GPP in the IBM [®] MarketScan [®] Research Databases (N=1383) |
|-----------------------|--|---|
| Female, n (%) | 784 (64.7) | 846 (61.2) |
| Age, years, mean (SD) | 60.8 (16.7) | 49.5 (15.1) |
| Age, n (%) | | |
| <18 years | 26 (2.1) | 63 (4.6) |
| 18–64 years | 596 (49.2) | 1211 (87.6) |
| ≥65 years | 590 (48.7) | 109 (7.9) |

GPP, generalized pustular psoriasis; SD, standard deviation.

Prevalence of GPP in the Optum[®] (in 2019) and IBM[®] MarketScan[®] databases (in 2018)

- Overall, prevalence of GPP for one inpatient or one outpatient claim was reported as 0.9 per 10,000 persons in 2019 in the Optum[®] database and 0.7 per 10,000 persons in 2018 in the IBM[®] MarketScan[®] database (Figure 3)
- Using the more stringent case definition, prevalence of GPP was estimated to be 0.3 per 10,000 persons in the Optum[®] database (in 2019) and 0.2 per 10,000 persons in the IBM[®] MarketScan[®] database (in 2018) (Figure 3)
- In both studies, when stratified by age, the prevalence of GPP was higher in those aged ≥65 years compared with those aged <18 years or 18–64 years (Figure 4)
- Prevalence was higher in females than in males in both studies (Figure 5)
 - Prevalence of GPP was higher in females compared with males across all age groups in the Optum[®] database, regardless of the case definition
 - Prevalence of GPP was higher in females across all age groups only using the one inpatient or one outpatient definition in the IBM[®] MarketScan[®] database
 - However, when the more stringent case definition was used, GPP was slightly more prevalent in females than in males aged 18–64 years in the IBM[®] MarketScan[®] database only (0.3 per 10,000 persons vs 0.2 per 10,000 persons, respectively; Table 2)

Figure 3. Overall prevalence of GPP in the Optum[®] database in 2019 and the IBM[®] MarketScan[®] database in 2018

Figure 4. Prevalence of GPP stratified by age group in the Optum[®] Clinformatics[™] Data Mart (2019) and IBM[®] MarketScan[®] Research (2018) databases

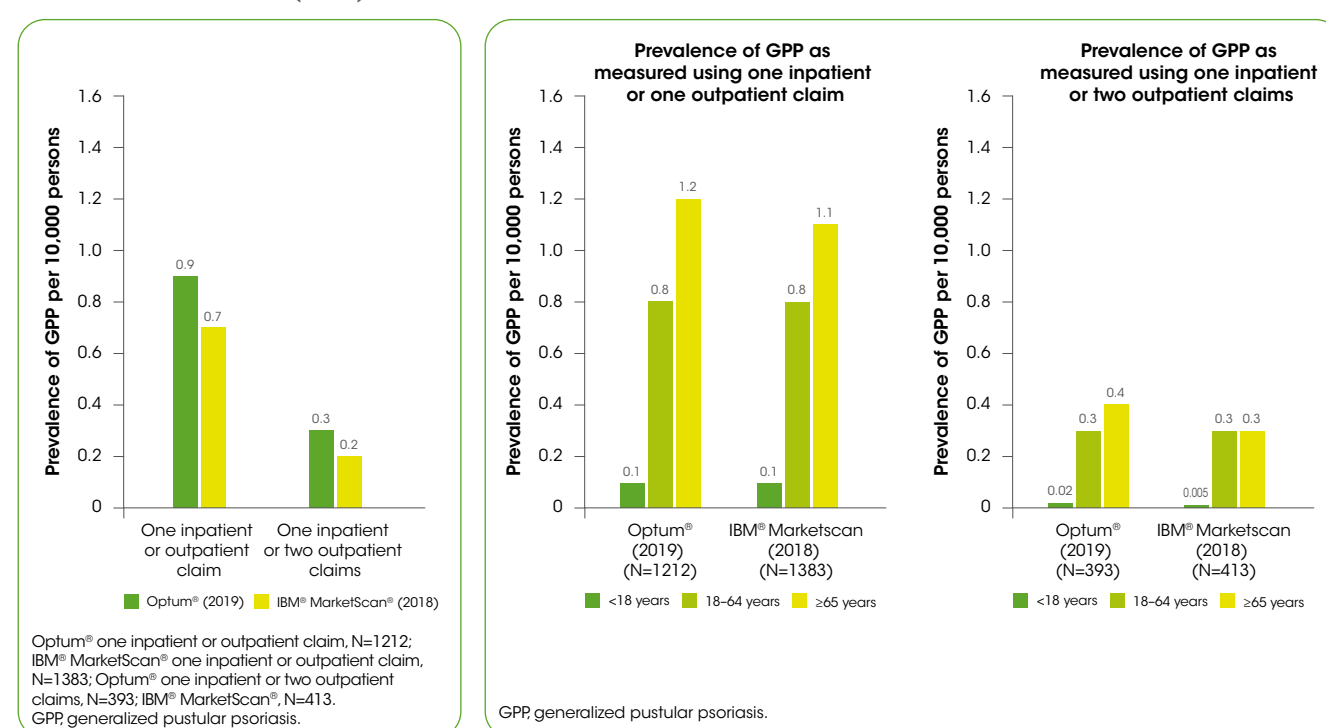


Figure 5. Prevalence of GPP stratified by sex in the Optum[®] (2019) and IBM[®] MarketScan[®] (2018) databases

Figure 6. Prevalence of GPP in other geographical regions

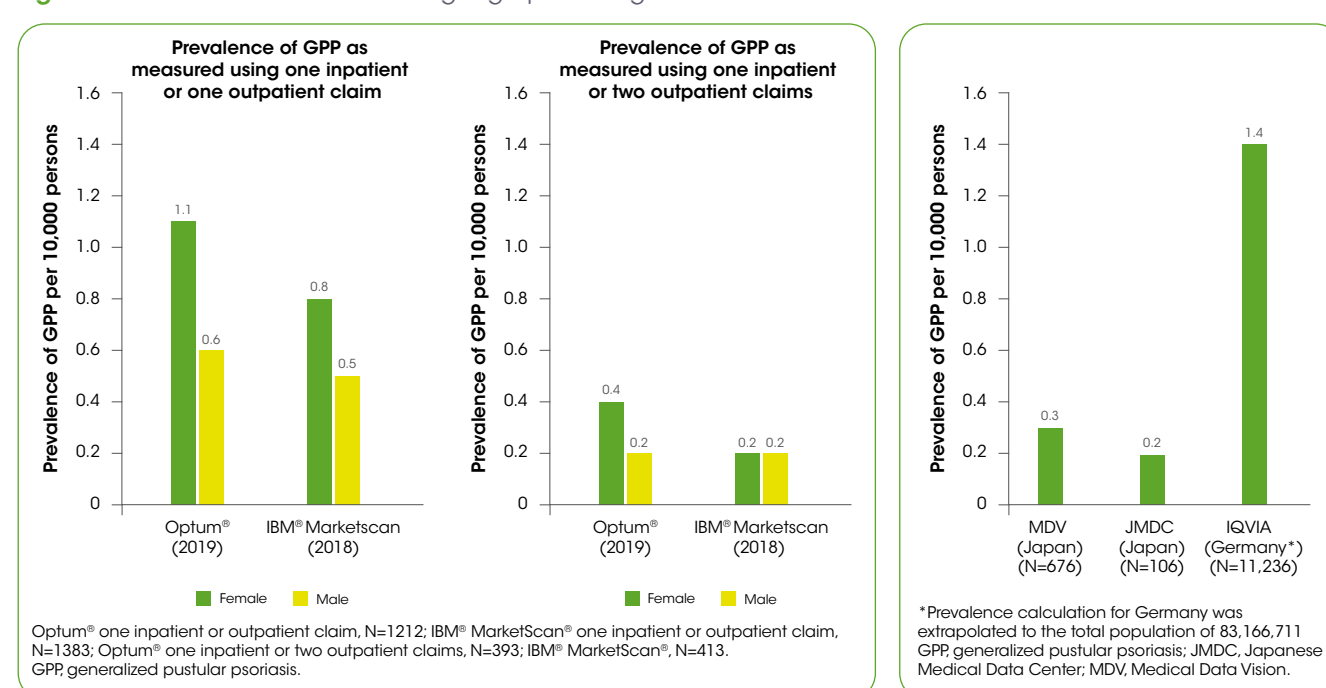


Table 2. Prevalence of GPP in females and males across different age groups

| Patient characteristics | GPP prevalence reported in the Optum [®] Clinformatics [™] Data Mart database* | | | | GPP prevalence reported in the IBM [®] MarketScan [®] Research database* | | | |
|-------------------------|--|----------------------------|--------------------------|----------------------------|--|----------------------------|--------------------------|----------------------------|
| | Female | | Male | | Female | | Male | |
| Sex | Original case definition | Additional case definition | Original case definition | Additional case definition | Original case definition | Additional case definition | Original case definition | Additional case definition |
| <18 yrs | 0.2 | 0.02 | 0.1 | 0.01 | 0.2 | 0.009 | 0.1 | N/A |
| 18–64 yrs | 1.1 | 0.4 | 0.6 | 0.2 | 1.0 | 0.3 | 0.6 | 0.2 |
| ≥65 yrs | 1.4 | 0.4 | 0.9 | 0.3 | 1.2 | 0.3 | 0.9 | 0.3 |

*Per 10,000 persons. Original case definition: one inpatient or one outpatient claim for ICD-10 code L40.1. Additional case definition: one inpatient or two outpatient claims for ICD-10 code L40.1. GPP, generalized pustular psoriasis; ICD-10, International Classification of Diseases 10th Revision; N/A, not applicable; yrs, years

Prevalence of GPP in other geographical regions

- The prevalence of GPP in the Japanese databases (identified by one diagnosis code of L40.1, inpatient or outpatient) was 0.3 per 10,000 persons (MDV database, 2015–2018) and 0.2 per 10,000 persons (JMDC database, 2018), in line with previous estimates⁶ (Figure 6)
- Between 2019 and 2020, the prevalence of GPP in Germany (identified by one diagnosis code of L40.1) was 1.4 per 10,000 persons as calculated from a total population of 83,166,711 (Figure 6)

Conclusions

- This study shows that GPP is more prevalent in older patients than in younger patients^{7–9}
- In line with published literature, GPP is also shown to be more frequent in females than in males, although there are variations in reported male-to-female ratios in the literature⁷
- This study also shows that the prevalence of GPP in the USA is comparable to the prevalence of GPP in Japan and that prevalence of GPP in Germany is slightly higher than in the USA and Japan
- A limitation to this study is that these data have not been extrapolated to analyse the prevalence of GPP in the US population and are limited to the population included in the respective databases, therefore prevalence may be underestimated
 - This limitation is also present in the prevalence calculations for Japan; however, GPP prevalence estimates in Germany were extrapolated to the total population
 - In addition, there is no validated algorithm to positively identify GPP in claims databases; the two case definitions reported herein demonstrate that, depending on the stringency of the definition, the prevalence of GPP can vary
- GPP is a rare disease, as shown by the prevalence estimates of patients included in the Optum[®] and IBM[®] MarketScan[®] databases; however, the impact of GPP on the lives of patients in the US is still not well characterised

Disclosures

This study was funded by Boehringer Ingelheim. N Kotowsky, R Gao and KG Brodovicz are full-time employees of Boehringer Ingelheim. SR Feldman is employed by the Wake Forest School of Medicine. C Leonardi is employed by Central Dermatology. A Menter is employed by Baylor Scott and White.

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