Prevalence, comorbidities, and mortality of generalized pustular psoriasis (GPP): A literature review

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This literature review identified the major challenges to interpreting prevalence, comorbidity, and mortality data on GPP, with considerable variation observed in study design, diagnostic criteria, and patient populations across published studies

PURPOSE

To present a literature review on available GPP prevalence, comorbidity, and mortality data to aid better understanding of GPP among clinicians and healthcare providers and help manage patients with this rare condition.

INTRODUCTION

- GPP is a rare, autoinflammatory skin disease characterized by episodes of widespread eruptions of sterile, macroscopic pustules that can occur with or without systemic inflammation and symptoms[1,2]
- Currently, there is no globally accepted guidance for management of GPP flares or for long-term treatment of the disease. Furthermore, there are no GPP-specific therapeutic agents approved in the USA or Europe[3,4]
- Here, we present a literature review on GPP prevalence, comorbidity, and mortality estimates in key demographics, and discuss the factors that impact collection, interpretation, and comparison of published data

CONCLUSIONS

- No consistent trends in GPP prevalence were identified. The variation in estimates was likely caused by the considerable differences in study design and settings, which included variations in disease definition, data sources, sample size, and patient inclusion criteria
- Clear trends in GPP comorbidities were identified, the most common of which was plaque psoriasis. Arthralgia and arthritis were also reported as common comorbidities
- High mortality rates indicate that GPP flares are potentially life-threatening due to a range of complications, including sepsis and multisystem organ failure. These findings highlight the need for new, more effective treatments for GPP

METHODS

- A literature search was conducted using Embase, MEDLINE, and the Cochrane Library in April 2021, and supplemented with a bibliographic review of relevant literature, congress abstracts, and consensus studies identified in the electronic searches until October 2021

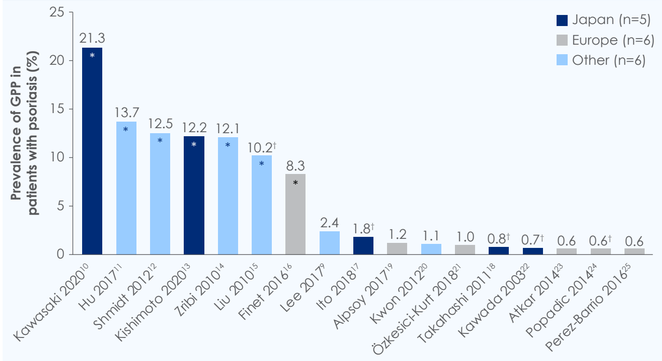
RESULTS

Prevalence of GPP by geographic location

Country	Study details	GPP diagnosis details	Patients with GPP	Estimated prevalence of GPP per million	Reference
China	A population-based study using data from the national database of Urban Basic Medical Insurance from 2012 to 2016, containing in- and out-patient data	ICD definitions (ICD10: 140.1 and ICD9: 694.3)	3401	14	Feng 2021[5]
France	A retrospective epidemiologic survey that collected in- and out-patient data from 112 dermatologic wards	Dermatologist's own criteria	99	1.76	Augey 2006[6]
Germany	Analysis of IQVIA German electronic medical records patient database containing in- and out-patient data from a 1-year period (2019 to 2020). Prevalence was extrapolated to the total population in Germany	≥1 confirmed diagnosis using ICD10 code of L40.1	11,236	140	Feldman 2021[7]
Japan	In-patient data obtained via questionnaires sent to 575 community center hospitals throughout Japan, seeking patients with GPP from 1983 to 1989	Dermatologist's own criteria	541	7.46	Ohkawara 1996[8]
Japan	In- and out-patient data from the Japanese Medical Data Vision database collected from 2018	≥1 confirmed diagnosis using ICD10 code of L40.1	106	20	Feldman 2021[7]
Japan	In- and out-patient data from the Japanese Medical Data Vision database collected between 2015 and 2018	≥1 confirmed diagnosis using ICD10 code of L40.1	676	30	Feldman 2021[7]
South Korea	In- and out-patient data from the Korean Health Insurance Review and Assessment Service database collected between 2011 and 2015	KCD-6 diagnosis code of L40.1	26,955	89–124	Lee 2017[9]
USA	In- and out-patient data collected from the Optum® Clinformatics® Data Mart in 2019	≥1 confirmed diagnosis using ICD10 code of L40.1	1212	90	Feldman 2021[7]

Estimates of GPP prevalence in the general population varied considerably from 1.76 to 140 per million

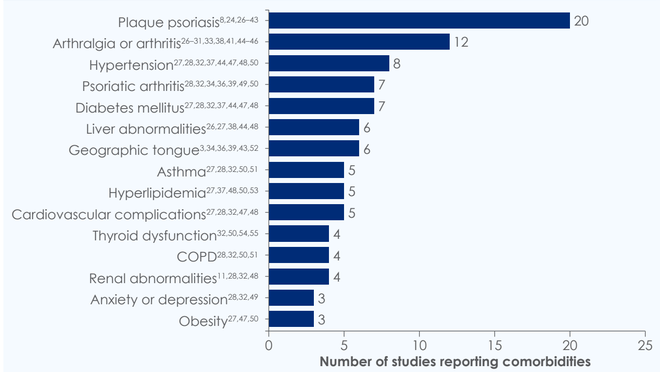
GPP prevalence in patients with psoriasis by geographic region



Studies are ordered from highest to lowest prevalence. "Other" comprises studies from China[11,13], South Korea[9,20], the USA[12], and Tunisia[14]. *Heterogeneous studies are those that included only hospitalized patients, psoriasis subtypes, or patients treated with specific drugs; †Prevalence of acute GPP (von Zumbusch type), all other studies report overall GPP prevalence.

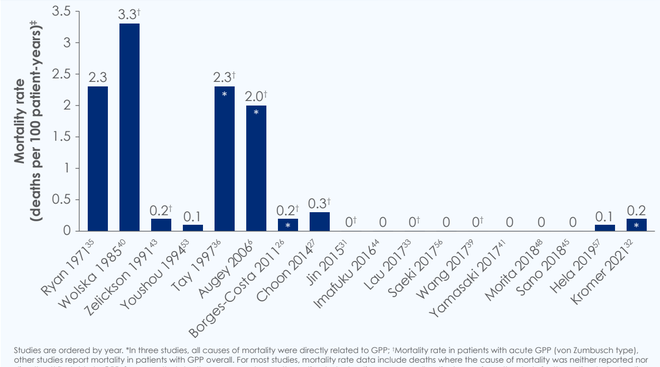
GPP prevalence among patients with psoriasis ranges between 0.6% and 21.3%; however, if heterogeneous studies are excluded, the range is between 0.6% and 2.4%

Studies reporting GPP-specific comorbidities



Clear trends in comorbidities were identified in patients with GPP. The most frequent was plaque psoriasis (percentage range of patients affected: 15.4–83.0%). Geographic tongue was reported more frequently in Asian (4 studies[33,36,39,52]) versus non-Asian studies (2 studies[34,43])

Mortality rates in patients with GPP



Studies are ordered by year. *In three studies, all causes of mortality were directly related to GPP; †Mortality rate in patients with acute GPP (von Zumbusch type), other studies report mortality in patients with GPP overall. For most studies, mortality rate data include deaths where the cause of mortality was neither reported nor directly attributable to GPP; ‡Assumes that deaths occur evenly over the entire study duration; assumes all patients remain on the study for the entire study duration.

Mortality rates in patients with GPP varied considerably from 0 to 3.3 deaths per 100 patient-years. In studies that measured GPP-related causes of death, sepsis/septic shock and cardiovascular complications were the most common cause of death

Abbreviations
COPD, chronic obstructive pulmonary disease; GPP, generalized pustular psoriasis; ICD, International Classification of Diseases; KCD, Korean Standard Classification of Disease.

References
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