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Introduction

Generalized pustular psoriasis (GPP) is a rare, severe variant of psoriasis, characterized by recurrent flares of widespread sterile pustules which may occur either *de novo* or be provoked by triggers.

Globally, there is a scarcity of studies on the epidemiology of GPP, with no population-based studies in Southeast Asia.

Published GPP data are highly variable with prevalence ranging from 1.76 to 124 cases per million persons.

The aim of this study was to estimate, for the first time, the annual incidence and prevalence of GPP in multi-ethnic Malaysians in the Johor Bahru district and characterize frequency of flare episodes and trigger factors.

Methods

DATA SOURCE

We used routinely collected electronic health records from the Teleprimary Care (TPC) clinical information system, a locally developed database linking public primary and secondary care facilities.

STUDY POPULATION

Dermatologist-confirmed GPP patients were identified by ICD-10 diagnostic code.

The study population (denominator) comprised any patient who contributed ≥ 1 day in the TPC. Between 01/01/2010 and 31/12/2020, 1.16 million people in Johor Bahru were registered in TPC (~98% of the estimated population), with 230 with dermatologist-confirmed GPP.

STATISTICAL ANALYSIS

The person-time at risk was calculated from the start of each calendar year of interest or the day of registration until the index date, death, or end of the calendar year, whichever came first.

- Annual prevalence (per million) and incidence (per million person-years) were estimated and stratified by age, sex, ethnicity, and associated psoriasis vulgaris (PV).
- The distribution of age of psoriasis onset was examined by plotting psoriasis incidence against age.
- Mean flares per patient per year and frequency of trigger factors were calculated and stratified by presence of associated PV.

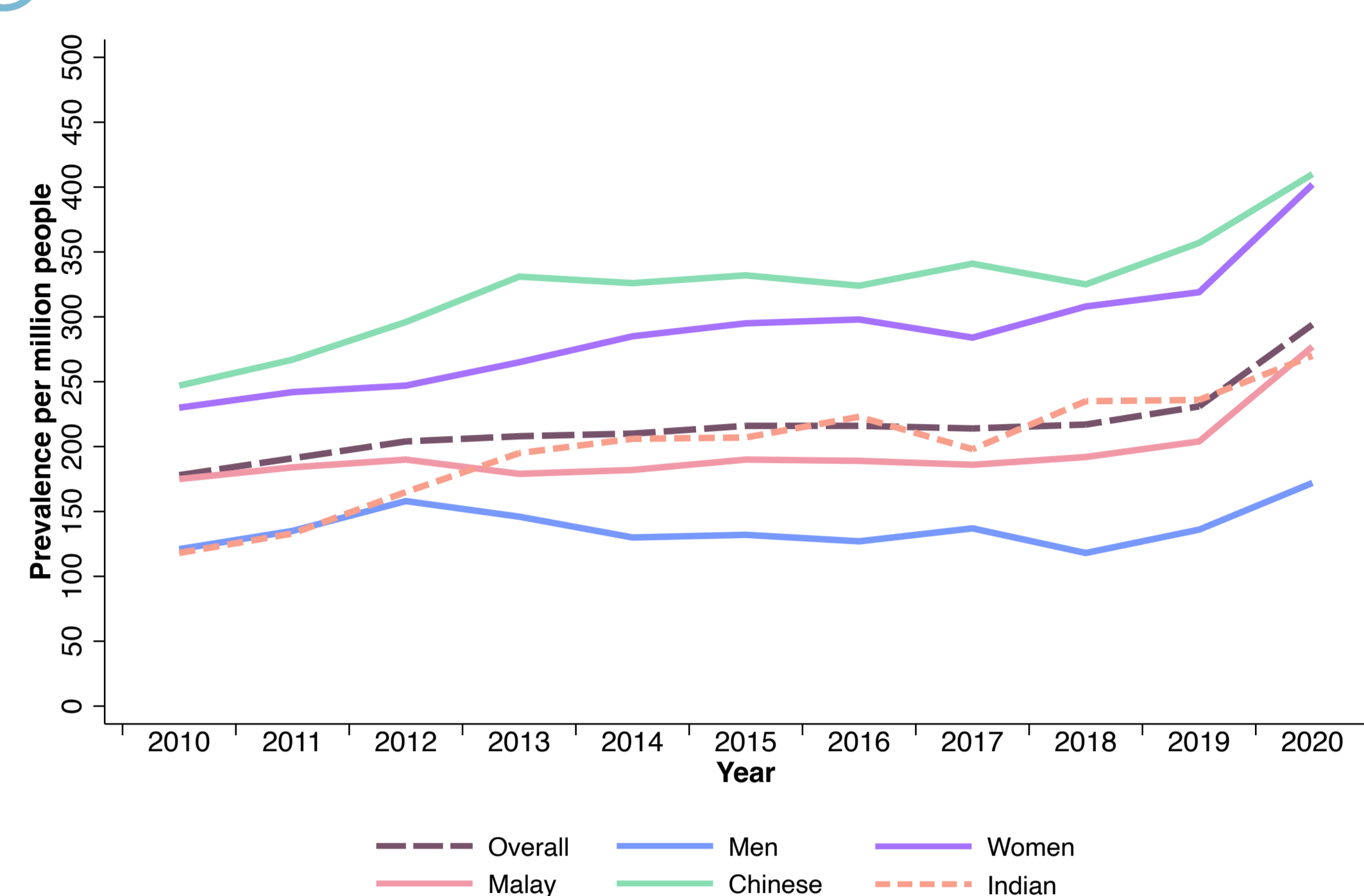
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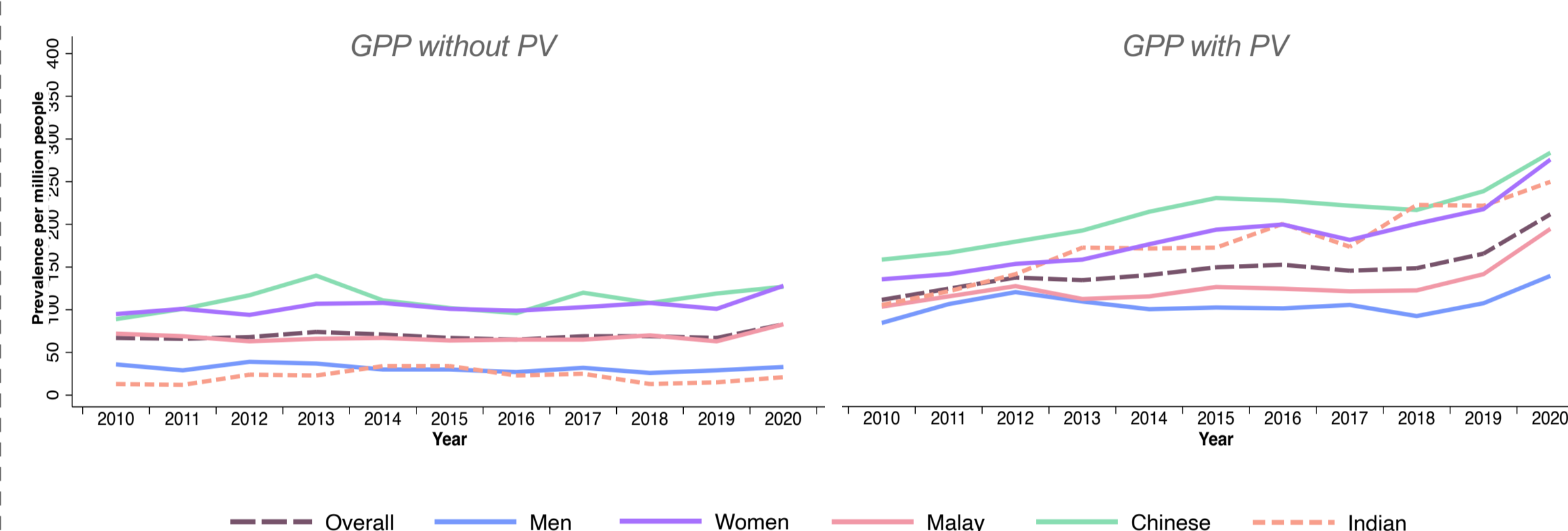
The Global Psoriasis Atlas has been supported by: The LEO Foundation, Abbvie, Almirall, Amgen, Janssen, Eli Lilly and Company, Novartis Pharma AG and UCB (2022/23)

Figures

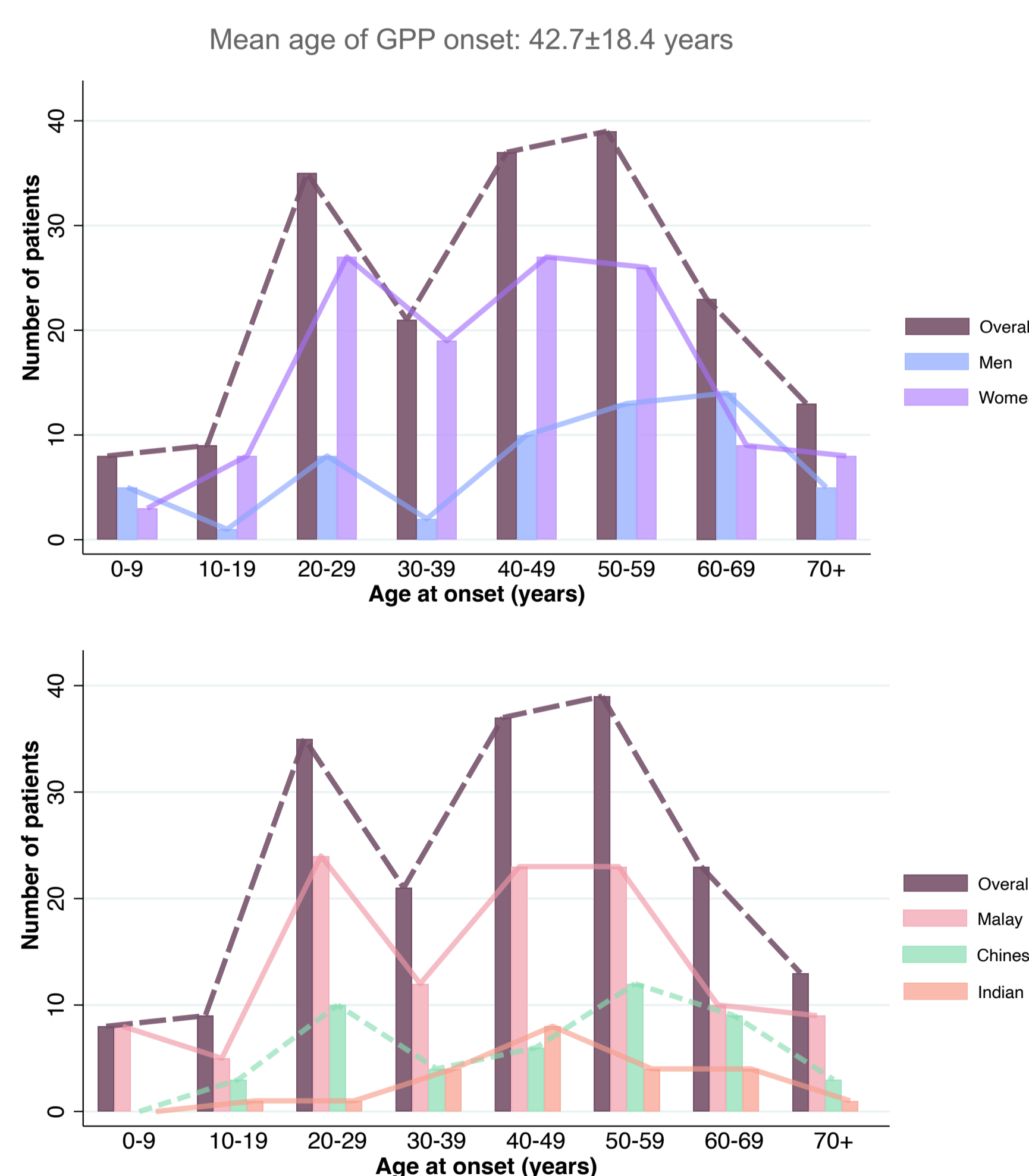
A Annual prevalence of GPP between 2010 and 2020



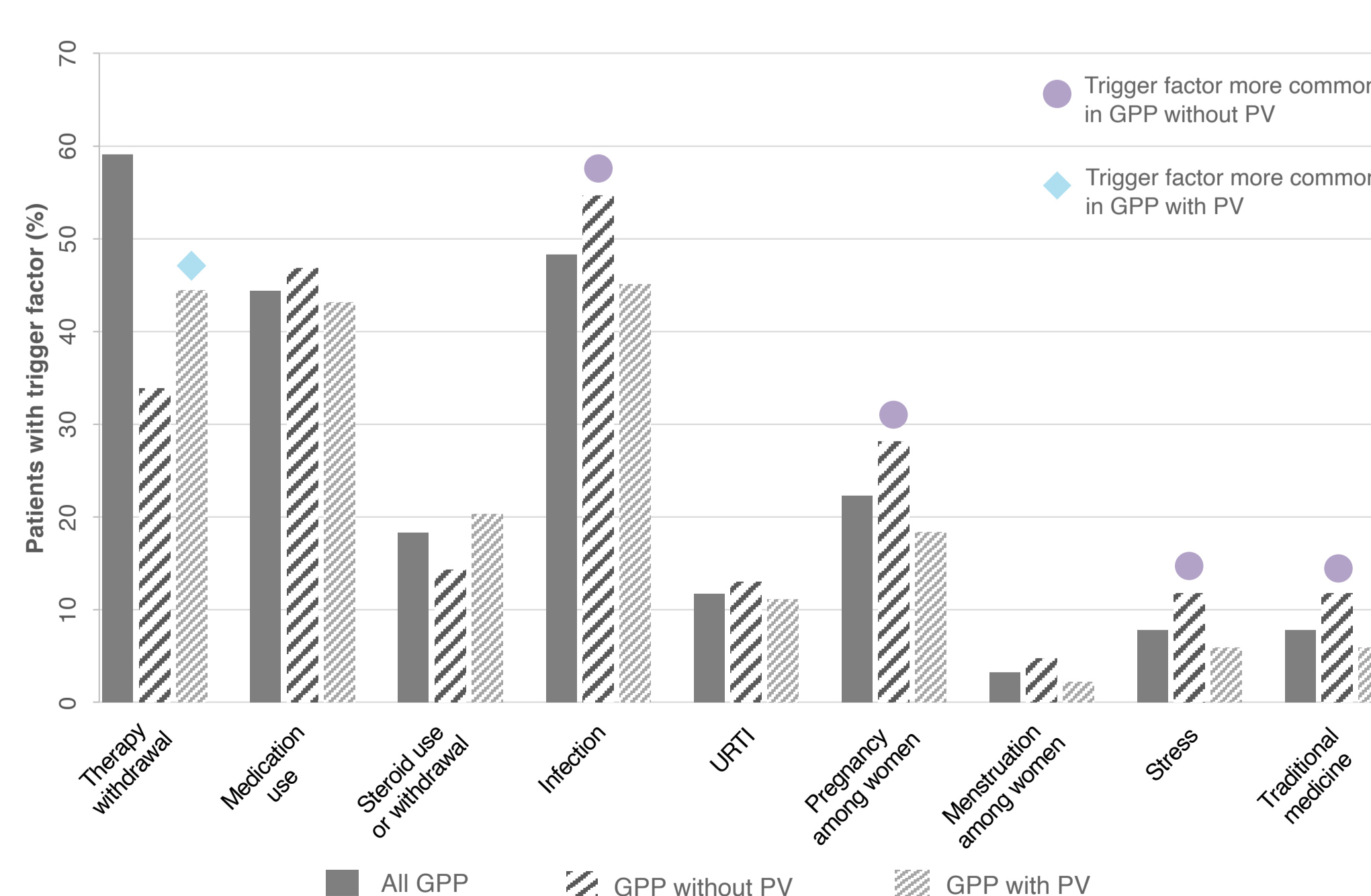
B Prevalence of GPP in people without and with associated PV



C Age distribution of incident GPP



D Trigger factors for GPP flares



Results

Period prevalence rate of GPP was 198 per million; 127/million in males, 267/million in females 186/million in Malays, 271/million in Chinese, 179/million in Indians

- Prevalence consistently higher in females than males and highest amongst Chinese, followed by Indians and Malays (Fig A).

Period prevalence of GPP without associated PV was 66 per million and GPP with associated PV was 132 per million;

- 67% of GPP patients had associated PV.
- Females and Malays more likely to have GPP without PV (Fig B).
- Males and Indians more likely to have GPP with PV (Fig B).

Period incidence of GPP was 27.2 (95% CI 22.8-31.6) per million person-years;

- No consistent sex or ethnic differences in incidence rates or trends over time.

Bimodality in age of GPP onset, with peaks at ages 20-29 & 50-59 years of age (Fig C).

- GPP onset ~7 years earlier in GPP patients without PV than with PV (mean age; 37.5 ± 20.7 vs 44.9 ± 17.0 years).

Patients experienced, on average, 1.33 ± 0.70 flares per year

- Malays & Chinese experienced more flares/patient/year on average than Indians.
- Flares occurred more frequently in GPP patients without than with PV (mean flares/patient/year 1.35 ± 0.77 vs 1.25 ± 0.58).

Common triggers of flares were withdrawal of therapy (mainly systemic steroids), infections (predominantly URTI), use of medications (mainly steroids and traditional medicines) and pregnancy (Fig D);

- Common triggers differed between patients with and without PV; Fig D.

Discussion

This study provides information for the first time on the burden of GPP in Southeast Asia, contributing to the global mapping of this disease.

GPP is more common in Johor Bahru than reported in other countries and varies by age, sex and ethnicity.

Patients with GPP alone were more likely to be female, Malay, have significantly earlier disease onset, greater frequency of flares and different trigger factors than GPP patients with PV.

These findings should help inform healthcare planning, resource allocation and management of patients with GPP in Malaysia.

