

Incidence, Prevalence, and Economic Burden of Granulomatosis With Polyangiitis and Microscopic Polyangiitis in the United States

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Introduction

- Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a group of rare diseases associated with small- to medium-sized blood vessel inflammation that can lead to life- and organ-threatening disease. The two most common subtypes of AAV are granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA).^{1,2}
- However, due to the rarity of the diseases, the epidemiology and economic burden of GPA/MPA are not well characterized.



Objective

To assess the incidence, prevalence, and economic burden of GPA/MPA in the US



Methods

Design: A retrospective, cross-sectional study

Data sources: IQVIA open-source pharmacy claims database and medical claims database and the IQVIA PharMetrics® Plus database from January 1, 2020, to December 31, 2023 for primary objectives and from January 1, 2020, to April 30, 2024 for secondary objectives

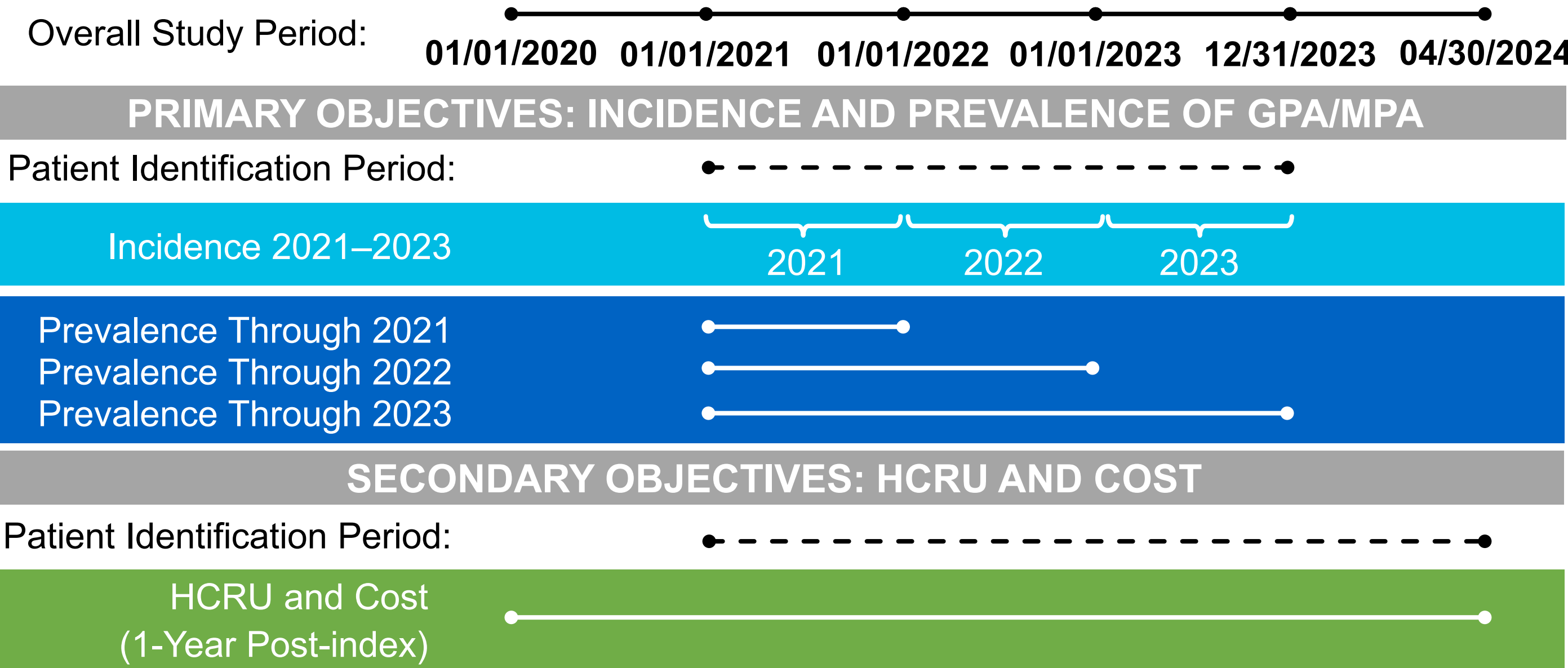
Key inclusion criteria:

- Aged ≥ 18 years on the index date **AND**
- At least two medical claims with an appropriate GPA/MPA/AAV diagnosis **AND**
- Continuous data availability during the calendar year for incidence/prevalence assessment **AND**
- Continuous enrollment for 12 months pre- and post-index (earliest diagnosis) for economic burden assessment

Study outcomes:

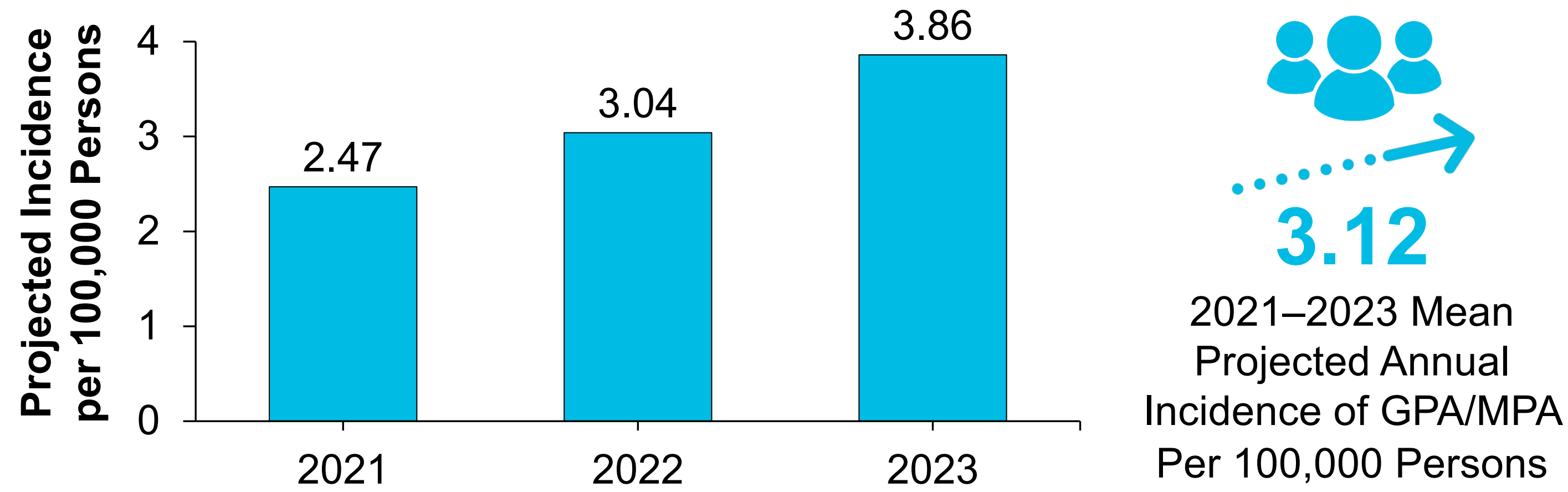
- GPA/MPA incidence and prevalence:** Projected using patient counts (per calendar year), stratified by age (0–17, 18–64, and 65+ years), and projected to the US Census Bureau’s annual Current Population Survey (2021–2023)
- Direct all-cause healthcare resource utilization (HCRU) and costs over the 1-year post-index period:** Data were stratified by relapse (defined as an inpatient/emergency department [ED] visit for GPA/MPA or related conditions accompanied by an increased glucocorticoid dosage) and by end-stage kidney disease (ESKD) diagnosis (identified using a diagnosis of ESKD in any position or procedure code for dialysis)

Study Overview

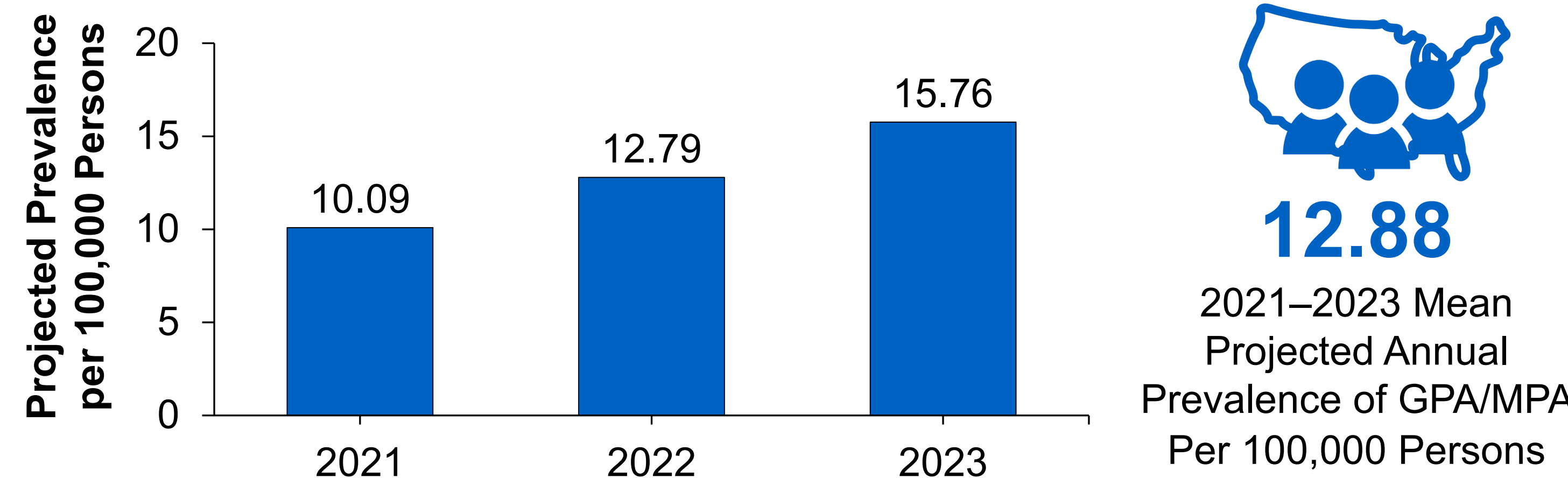


Results

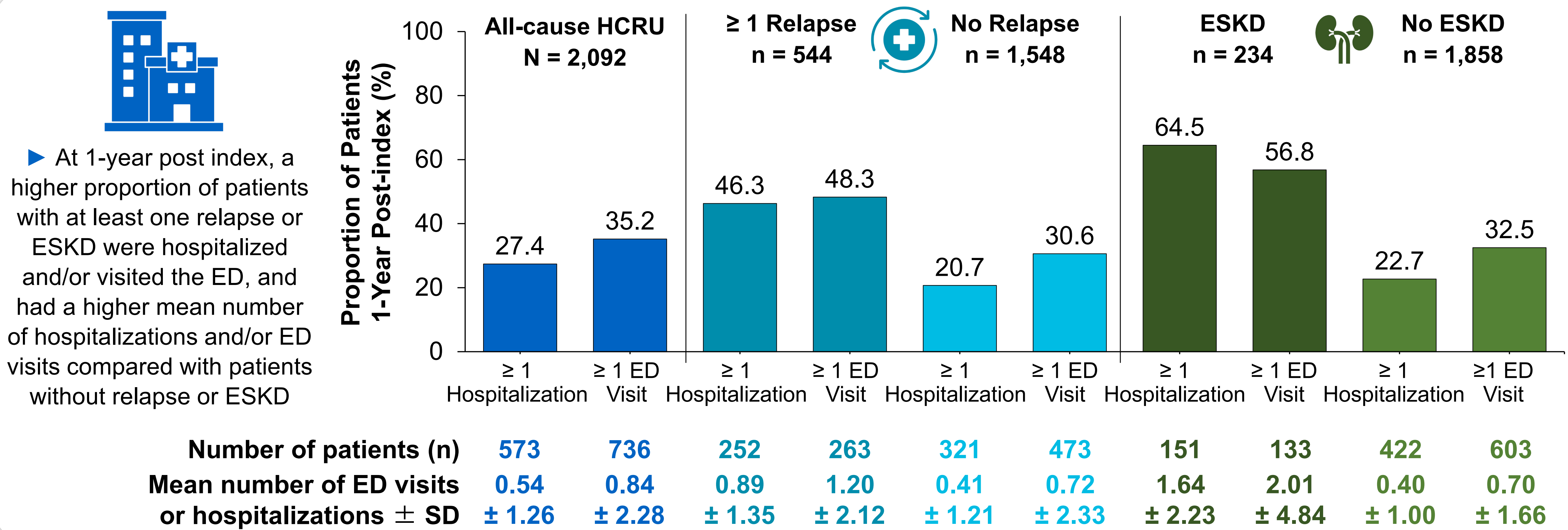
Projected Annual GPA/MPA Incidence: 2021–2023



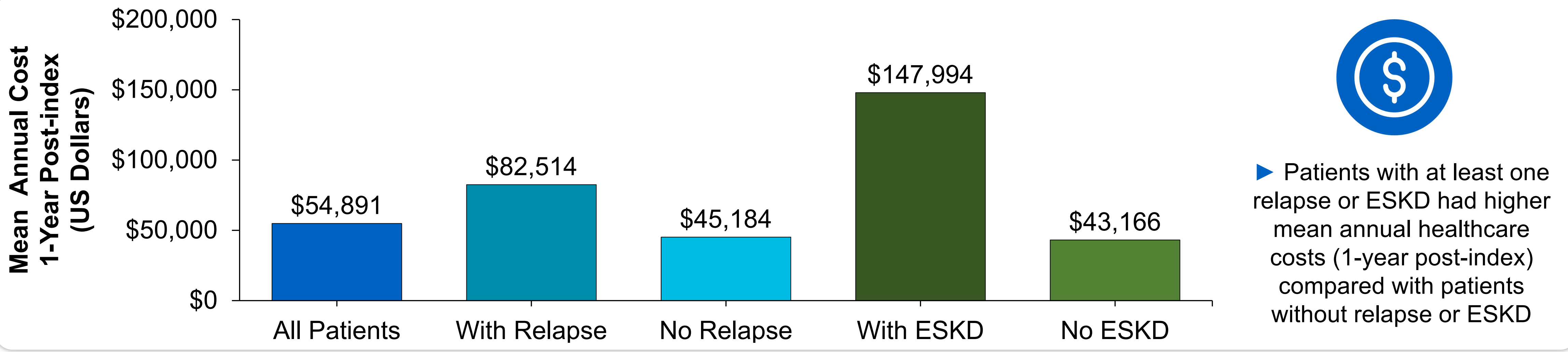
Projected Annual GPA/MPA Prevalence: 2021–2023



HCRU Among Patients With GPA/MPA



All-Cause Healthcare Costs in Patients with GPA/MPA



Key Takeaways

- The mean projected annual incidence and prevalence of GPA/MPA is 3.12 and 12.88 per 100,000 persons, respectively
- Patients with GPA/MPA and at least one relapse or ESKD had higher HCRU and mean annual costs compared with those without relapse or ESKD
- Targeted interventions aiming to reduce relapses and slow the progression of kidney disease may decrease the HCRU needs of patients with GPA/MPA



Scan the QR code for additional methods, results (incidence/prevalence by age), limitations, abbreviations, acknowledgments, funding, references, and disclosures.

Additional Methods

List of Patient Eligibility Criteria: Incidence of GPA/MPA

Objective	Details
Primary: Incidence of GPA/MPA	Inclusion Criteria
	Patients with continuous data availability in the LRx/Dx (proxy for CE) between January 1 and December 31 of the calendar year of interest
	LRx
	<ul style="list-style-type: none">Pharmacy stability: At least one pharmacy the patient visited during the study (January 1, 2021, to December 31, 2023) consistently supplied data during the calendar year of interestPatient pharmacy activity: At least one pharmacy claim between January 1 and June 30 and at least one pharmacy claim between July 1 and December 31 during the calendar year of interest
	Dx
	<ul style="list-style-type: none">Provider stability: At least one provider the patient visited during the study years (January 1, 2021, to December 31, 2023) consistently supplied data during the calendar year of interestPatient provider activity: At least one medical claim between January 1 and June 30 and at least one medical claim between July 1 and December 31 during the calendar year of interest
	Exclusion Criteria
	<ul style="list-style-type: none">Patients with a GPA/MPA/AAV diagnosis code (ICD-10-CM) prior to the calendar year of interest using all available history in the databasePatients with data quality issues (eg, missing sex)From numerator only: For patients with only the AAV diagnosis code observed during the calendar year of interest (ie, no diagnosis code for GPA or MPA during the calendar year of interest), those with at least one inpatient claim or at least two outpatient claims separated by ≥ 7 days with a diagnosis code for EGPA or at least one claim for mepolizumab during the calendar year of interest<ul style="list-style-type: none">The second outpatient claim can occur in the next calendar year

Additional Methods

List of Patient Eligibility Criteria: Prevalence of GPA/MPA

Objective	Details
Primary: Prevalence of GPA/MPA	Inclusion Criteria
	Patients with continuous data availability in the LRx/Dx (proxy for CE) between January 1 and December 31 of the calendar year of interest
	LRx
	<ul style="list-style-type: none">Pharmacy stability: At least one pharmacy the patient used from January 1, 2021, to December 31, 2023, consistently supplied data during the calendar year of interestPatient pharmacy activity: At least one pharmacy claim between January 1 and June 30 and at least one pharmacy claim between July 1 and December 31 during the calendar year of interest
	Dx
	<ul style="list-style-type: none">Provider stability: At least one provider the patient visited from January 1, 2021, to December 31, 2023, consistently supplied data during the calendar year of interestPatient provider activity: At least one medical claim between January 1 and June 30 and at least one medical claim between July 1 and December 31 during the calendar year of interest
	Exclusion Criteria
	<ul style="list-style-type: none">Patients with data quality issues (eg, missing sex)For patients with only the AAV diagnosis code observed during the calendar year of interest and in previous years (ie, no diagnosis code for GPA or MPA prior to or during the calendar year of interest), those with at least one inpatient claim or at least two outpatient claims separated by ≥ 7 days with a diagnosis code for eosinophilic GPA or at least one claim for mepolizumab during the calendar year of interest or in previous years<ul style="list-style-type: none">The second outpatient claim can occur in the next calendar year

Additional Methods

List of Patient Eligibility Criteria: Direct Economic Burden of GPA/MPA

Objective	Details
Secondary: Direct economic burden of GPA/MPA	Inclusion Criteria
	<ul style="list-style-type: none">Patients with at least two nonancillary medical claims with a diagnosis code for GPA/MPA/AAV separated by ≥ 30 days during the index period (January 1, 2021, to April 30, 2024) in the database (LRx/Dx and PharMetrics Plus)<ul style="list-style-type: none">Date of the first GPA/MPA/AAV diagnosis during this period is the index dateAged ≥ 18 years on the index dateA minimum of 12 months of CE pre-indexA minimum of 12 months of CE post-index
	Exclusion Criteria
	<ul style="list-style-type: none">Patients with data quality issues (eg, missing sex)For patients with only the AAV diagnosis code observed during the study period (ie, no diagnosis code for GPA or MPA), those with at least one inpatient claim or at least two outpatient claims separated by ≥ 7 days with a diagnosis code for EGPA or at least one claim for mepolizumab during the study period

List of ICD-10-CM diagnostic codes for identifying patients with GPA, MPA, and AAV

Code Type	Codes	Name/Description	Flag
ICD-10-CM	M31.3	Wegener’s granulomatosis	GPA
ICD-10-CM	M31.30	Wegener’s granulomatosis without renal involvement	GPA
ICD-10-CM	M31.31	Wegener’s granulomatosis with renal involvement	GPA
ICD-10-CM	M31.7	Microscopic polyangiitis	MPA
ICD-10-CM	I77.82	ANCA vasculitis	AAV

Additional Methods

Cost Standardization (HCRU and Cost): Study Outcomes

- Total all-cause HCRU and costs were stratified by relapse and ESKD status and reported overall and summarized by medical (not including office-administered drugs) and pharmacy claims (including office-administered drugs) over the 1-year post-index period
 - Specific categories of utilization were reported as follows (total costs were the sum of the below components):
 - Inpatient hospitalizations
 - ED visits
 - Outpatient physician office visits
 - Outpatient nonphysician office visits
 - Outpatient pharmacy (including drugs identified via pharmacy claims and office-administered drugs identified in medical claims)
- Costs were reported individually for each utilization category detailed above and converted to 2023 US dollars using the healthcare component of the Consumer Price Index

Statistical analyses

Descriptive statistics: For all relevant study measures, overall and for patients with at least one relapse or with ESKD

Categorical measures: Frequency (number of patients [n]) and percentage (%) of total study patients observed in each category

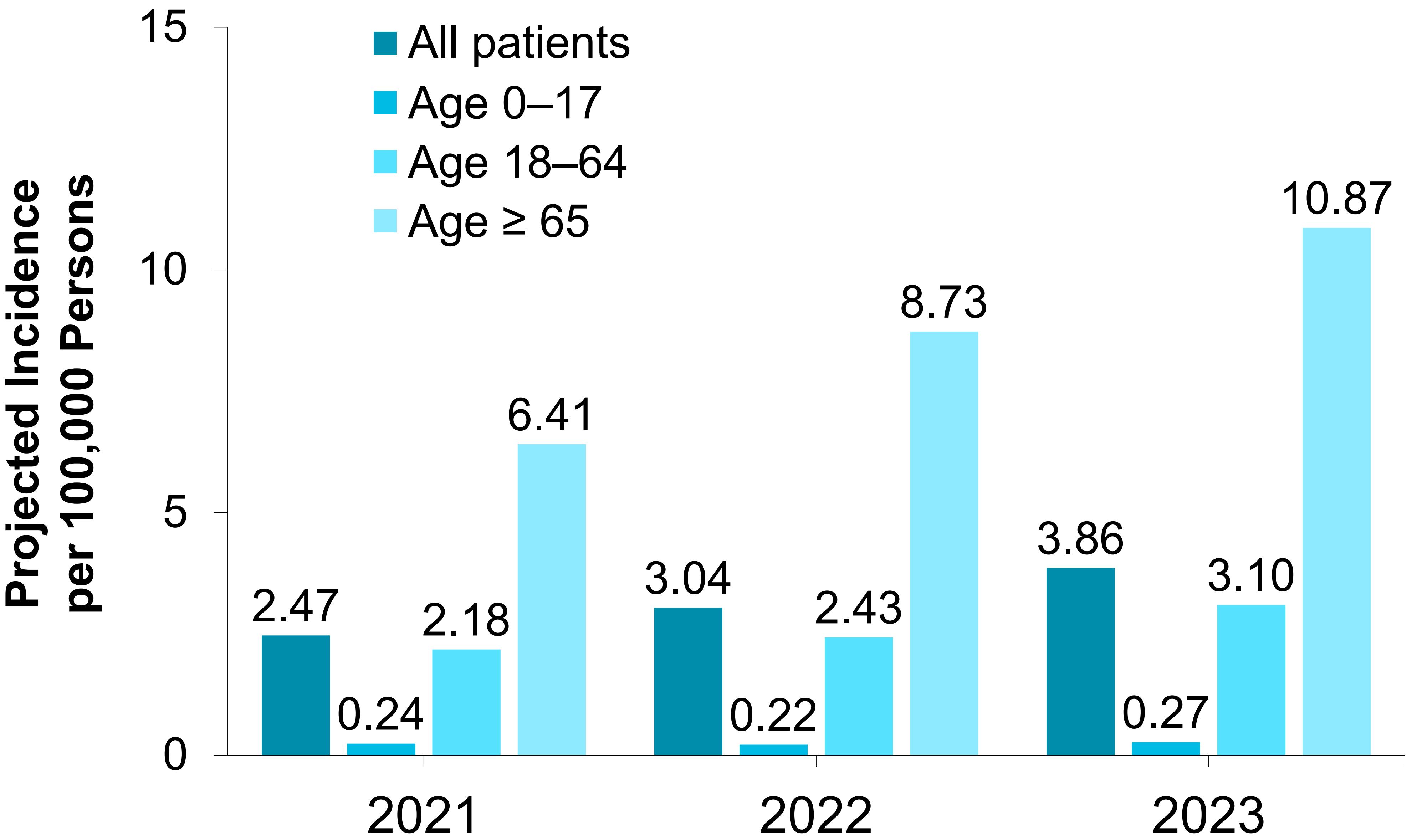
Continuous and count variables: Mean, SD, or median as relevant; continuous variables were also categorized into intervals

Limitations

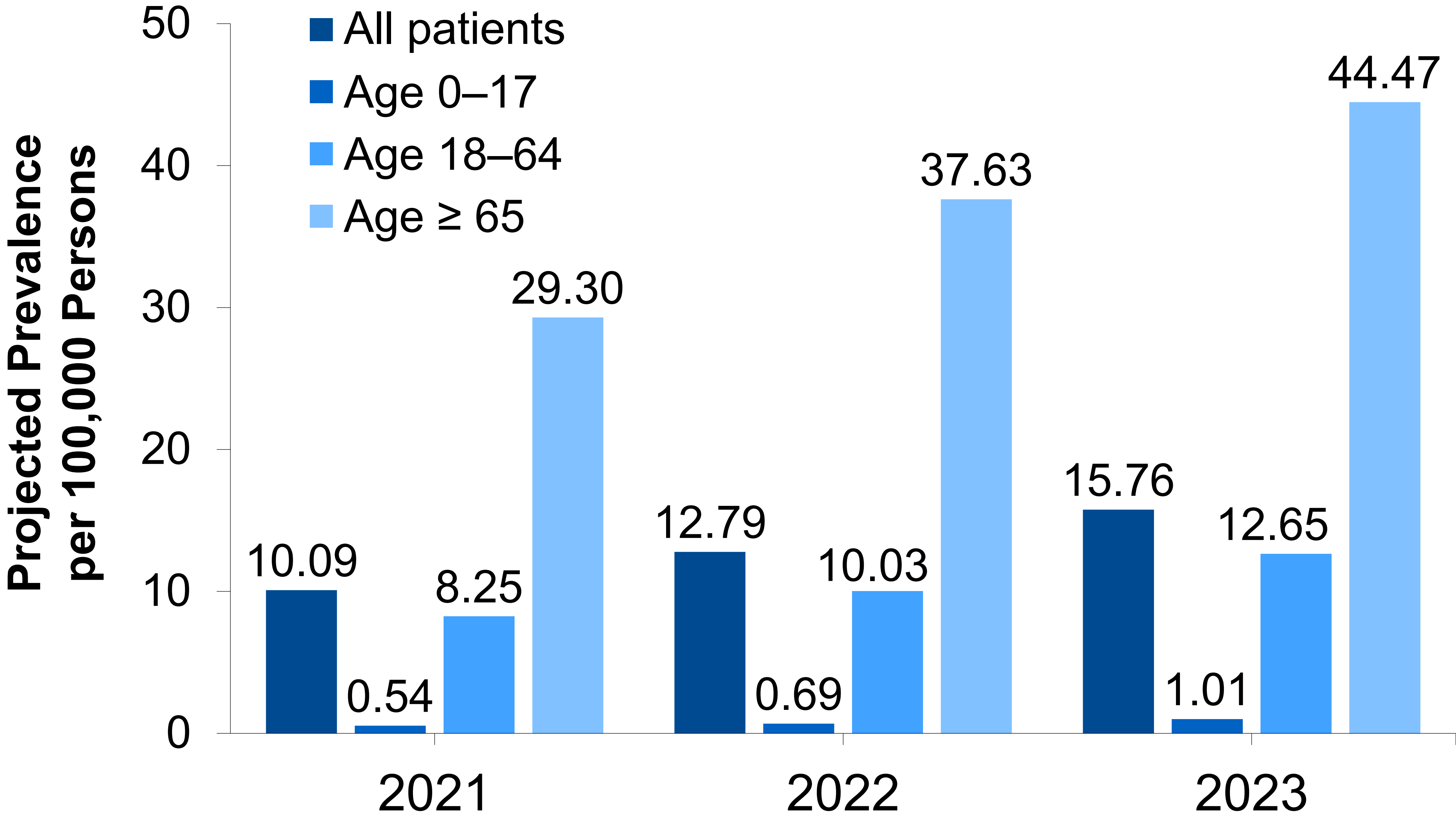
- Coding inaccuracies are possible when collecting claims data
- Variation may exist across the healthcare systems from which data were collected
- ICD-10-CM diagnostic codes documented in claims data are used primarily for reimbursement rather than for research purposes

Additional Results

Projected Annual GPA/MPA Incidence:
2021–2023



Projected Annual GPA/MPA Prevalence:
2021–2023



Additional Information

Abbreviations

AAV, ANCA-associated vasculitis; **ANCA**, antineutrophil cytoplasmic antibody; **CE**, continuous enrollment; **Dx**, Medical Claims Database; **ED**, emergency department; **ESKD**, end-stage kidney disease; **GPA**, granulomatosis with polyangiitis; **HCRU**, healthcare resource utilization; **ICD-10-CM**, International Classification of Diseases, Tenth Revision, Clinical Modification; **LRx**, IQVIA Longitudinal Prescription Claims Database; **MPA**, microscopic polyangiitis.

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References

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Disclosures

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