

Burden of Illness Associated with Eosinophilic Granulomatosis with Polyangiitis (EGPA, formerly Churg-Strauss Syndrome) in the United States: Evidence from Two Administrative Claims Databases

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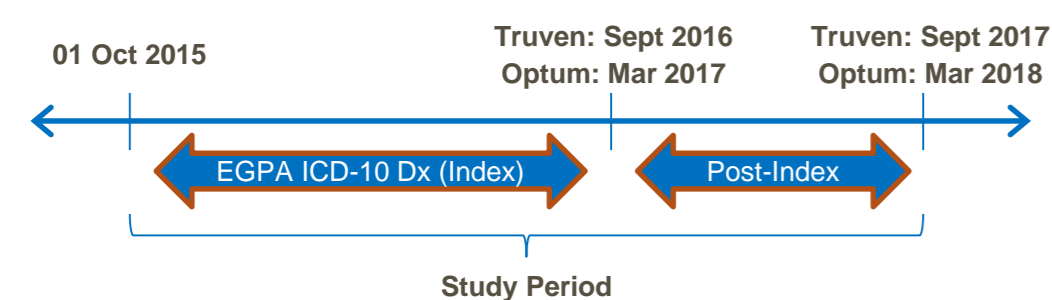
Aims

- Eosinophilic granulomatosis with polyangiitis (EGPA), formerly Churg-Strauss Syndrome, is a rare, complex multisystem inflammatory disorder
- EGPA is characterized by asthma, chronic rhinosinusitis, blood eosinophilia, and vascular inflammation that may lead to organ damage
- Global prevalence and incidence estimates for EGPA vary substantially:
 - Global prevalence range: 5.3 to 38 cases per million¹
 - Global incidence range: 0.5 to 3.7 cases per million¹
- In part due to the historical lack of EGPA-specific ICD-9 diagnosis codes and no regulatory-approved treatments for EGPA, published data on the clinical and economic burden of EGPA are lacking
- The introduction of the ICD-10 diagnosis code M30.1 (polyarteritis with lung involvement [Churg Strauss syndrome]) in October 2015 should facilitate the identification of an EGPA cohort for such an analysis
- The purpose of this study was to describe the clinical and economic characteristics of patients diagnosed with EGPA in the United States

Methods

- Retrospective analysis of two administrative claims databases was conducted, where the study period spanned from ICD-10 introduction through the most recent data availability (Figure 1):
 - Optum Clinformatics Data Mart: 01 Oct 2015 to 31 Mar 2018
 - Truven MarketScan: 01 Oct 2015 to 30 Sept 2017
- Index date was defined as the first medical claim with the presence of the ICD-10 diagnosis code M30.1 (polyarteritis with lung involvement [Churg Strauss syndrome])
- Inclusion criteria
 - ≥18 years of age
 - ≥1 medical claim with the ICD-10 diagnosis code M30.1
 - ≥12 months of continuous enrolment post-index
- Endpoints
 - Demographics, comorbidities, healthcare utilization (services and prescription medications) and costs for the 12-month post-index period (including index)

Figure 1. Study Schematic



Results

Study Sample Selection and Population Characteristics

- Less than 0.002% of the population in both databases had an ICD-10 EGPA diagnosis (Table 1). Demographic characteristics and comorbidities were similar across the two databases (Table 2)

Table 1. Study Sample Selection

Criteria	Optum Clinformatics		Truven MarketScan	
	N	%	N	%
Cohort Identification Period	01 Oct 2015 - 31 Mar 2017		01 Oct 2015 - 30 Sept 2016	
Total Sample	23,923,982	100.0%	32,633,801	100%
≥1 EGPA ICD-10 diagnosis	456	0.0019%	508	0.0016%
≥18 years of age	455	0.0019%	501	0.0015%
≥12 months continuous enrollment post-index	353	0.0015%	359	0.0011%

EGPA = eosinophilic granulomatosis with polyangiitis; ICD = International Classification of Diseases.

Table 2. Demographic Characteristics

Outcome	Optum Clinformatics		Truven MarketScan	
	N	%	N	%
Study Period	01 Oct 2015 - 31 Mar 2018		01 Oct 2015 - 30 Sept 2017	
N	353		359	
Age, mean (SD)	60.8 (15.8)		55.2 (13.9)	
Female, N (%)	215 (60.9%)		220 (61.3%)	
Quan-CCI, mean (SD)	2.2 (2.0)		1.6 (1.5)	
AHRQ Level 1 Comorbidities, N (%)				
Musculoskeletal/connective tissue	353 (100.0%)		359 (100.0%)	
Symptoms/signs/ill-defined conditions	343 (97.2%)		340 (94.7%)	
Respiratory system	323 (91.5%)		328 (91.4%)	
Circulatory system	294 (83.3%)		279 (77.7%)	
Nervous system	296 (83.9%)		277 (77.2%)	
Asthma, N (%)	227 (64.3%)		238 (66.3%)	
Constitutional Symptoms, N (%)				
Arthralgia	131 (37.1%)		105 (29.3%)	
Fever	50 (14.2%)		34 (9.5%)	
Myalgia	41 (11.6%)		27 (7.5%)	
Skin Involvement, N (%)				
Purpura	27 (7.7%)		17 (4.7%)	
Urticaria	15 (4.3%)		11 (3.1%)	
Ear, Nose & Throat, N (%)				
Chronic sinusitis	102 (28.9%)		124 (34.5%)	
Allergic rhinitis	97 (27.5%)		97 (27.0%)	
Acute sinusitis	64 (18.1%)		71 (19.8%)	
Renal - Nephritis, N (%)	13 (3.7%)		14 (3.9%)	
Nervous System, N (%)				
Polyneuropathy	50 (14.2%)		51 (14.2%)	
Mononeuritis multiplex	11 (3.1%)		13 (3.6%)	
Stroke	13 (3.7%)		11 (3.1%)	
Chest, N (%)				
Pulmonary infiltrates	86 (24.4%)		87 (24.2%)	
Alveolar/pulmonary hemorrhage	15 (4.3%)		10 (2.8%)	
Cardiovascular, N (%)				
Congestive heart failure	42 (11.9%)		33 (9.2%)	
Cardiomyopathy	17 (4.8%)		22 (6.1%)	

CCI = Charlson Comorbidity Index; N = number; SD = standard deviation.

Prescription Medication Utilization (12-month post-index)

- The majority of patients had received at least one prescription medication: non-biologic respiratory prescription (64.3% to 71.3%), biologic or immunosuppressant prescription (82.2% to 90.3%), or corticosteroids (74.8% to 85.8%) (Table 3)
- More than 50% of patients had a prednisone-equivalent oral corticosteroid (OCS) dose >15 mg/day, increasing to >75% of patients when considering a threshold >7.5 mg/day

Table 3. Concomitant Prescription (Rx) Medications (12-month post-index)

Outcome	Optum Clinformatics		Truven MarketScan	
	N	%	N	%
Study Period	01 Oct 2015 - 31 Mar 2018		01 Oct 2015 - 30 Sept 2017	
N	353		359	
Unique Rx Medications, mean (SD)				
NDC	18.1 (13.5)		18.4 (11.4)	
HCPC	2.3 (3.6)		2.8 (4.0)	
Medication Dispensings, mean (SD)	55.2 (61.7)		52.8 (38.4)	
Non-Biologic Respiratory Rx, N (%)	227 (64.3%)		256 (71.3%)	
SABA	143 (40.5%)		173 (48.2%)	
ICS/LABA	132 (37.4%)		169 (47.1%)	
ICS	65 (18.4%)		81 (22.6%)	
LTRA	61 (17.3%)		63 (17.6%)	
LAMA	29 (8.2%)		22 (6.1%)	
Biologic Respiratory Rx, N (%)	22 (6.2%)		25 (7.0%)	
Biologic or I/S Rx, N (%)				
Any immunosuppressive	290 (82.2%)		324 (90.3%)	
Azathioprine	68 (19.3%)		69 (19.2%)	
Methotrexate	44 (12.5%)		53 (14.8%)	
Rituximab	37 (10.5%)		36 (10.0%)	
Mycophenolate mofetil	27 (7.7%)		34 (9.5%)	
Cyclophosphamide	14 (4.0%)		18 (5.0%)	
Corticosteroids, N (%)	264 (74.8%)		308 (85.8%)	
Oral corticosteroids (OCS)	241 (68.3%)		287 (79.9%)	
Injectable or IV corticosteroids	118 (33.4%)		136 (37.9%)	
OCS Utilization				
Average daily dose, mean (SD)	18.3 (15.7)		51.3 (219.0)	
Median daily dose	15.1		15.7	
≤4 mg/day	6 (2.5%)		19 (6.7%)	
>4 mg/day to ≤7.5 mg/day	43 (17.8%)		49 (17.3%)	
>7.5 mg/day to ≤15 mg/day	69 (28.6%)		72 (25.4%)	
>15 mg/day	123 (51.0%)		143 (50.5%)	

HCPC = Healthcare Common Procedure Coding System; ICS = inhaled corticosteroid; I/S = immunosuppressant; IV = intravenous; LABA = long-acting beta-agonist; LAMA = long-acting muscarinic antagonist; LTRA = leukotriene receptor antagonist; mg = milligram; N = number; NDC = National Drug Code; Rx = prescription; SABA = short-acting beta-agonist; SD = standard deviation.

Healthcare Utilization and Costs (12-month post-index)

- Healthcare utilization and costs are summarized in Table 4
- Patients appeared to be high utilizers of healthcare services at a substantial mean cost (\$43,046 and \$56,402 in the Optum and Truven databases, respectively, with the difference in cost estimates owing to the costing methodology employed within the respective databases)

Table 4. Healthcare Utilization and Costs* (12-month post-index)

Outcome	Optum Clinformatics		Truven MarketScan	
	N	%	N	%
Study Period	01 Oct 2015 - 31 Mar 2018		01 Oct 2015 - 30 Sept 2017	
N	353		359	
Healthcare Utilization, N (%)				
Inpatient admissions	93 (26.4%)		80 (22.3%)	
Emergency room visits	155 (43.9%)		124 (34.5%)	
Physician office visits	313 (88.7%)		351 (97.8%)	
Hospital-based outpatient visits	266 (75.4%)		289 (80.5%)	
EOS Laboratory Test Results				
Patients with test (e.g., CPT 85025), N (%)	311 (88.1%)		270 (75.2)	
If yes, number of tests, mean (SD)	4.6 (4.7)		4.6 (4.8)	
EOS level (cells/μL), mean (SD)	814.9 (1,645.1)		873.4 (1,074.9)	
EOS level (cells/μL), median	300		536	
Asthma-Related Exacerbations, N (%)	50 (14.2%)		64 (17.8%)	
Events, mean (SD)	1.6 (1.1)		1.8 (1.6)	
Total Healthcare Costs*, mean (SD)	\$43,046 (58,029)		\$56,402 (146,928)	
Pharmacy cost component	\$14,816 (28,427)		\$13,595 (20,848)	
Medical cost component	\$28,230 (43,003)		\$42,807 (143,886)	
Utilization and Costs Among Patients with ≥1 Service Utilization				
Healthcare Utilization, mean (SD)				
Inpatient admissions	1.6 (1.2)		2.0 (2.0)	
Length of stay	7.1 (9.5)		7.9 (16.6)	
Emergency room visits	2.4 (2.9)		2.5 (2.9)	
Physician office visits	17.8 (14.1)		19.9 (15.7)	
Hospital-based outpatient visits	11.6 (14.8)		8.6 (10.5)	
Healthcare Costs*, mean (SD)				
Inpatient admissions	\$34,024 (38,661)		\$100,842 (265,499)	
Emergency room visits	\$9,719 (17,282)		\$7,085 (11,510)	
Physician office visits	\$3,394 (3,308)		\$3,951 (4,496)	
Hospital-based outpatient visits	\$9,494 (16,321)		\$14,114 (44,839)	

*Caution when comparing costs between databases. The Optum database employs a standard costing methodology that may result in lower estimates with a tighter standard deviation. CPT = Current Procedural Terminology; EOS = eosinophil; μL = microliter; N = number; SD = standard deviation.

Conclusions

- This study characterized patients diagnosed with EGPA based on the ICD-10 diagnosis code M30.1; and, study results are generally consistent with demographic and clinical characteristics from previously published studies.
- This study demonstrated that healthcare resource utilization and costs associated with EGPA were substantial in the 12-month follow-up period
- Additional research is warranted to quantify the clinical and economic burden of EGPA, especially as compared with a control group (e.g., asthmatics)

References

- GlaxoSmithKline. Data on File. 2017N348628_00. 2017.

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