PA648

Characterisation and burden of eosinophilic granulomatosis with polyangiitis in European patients by disease phase

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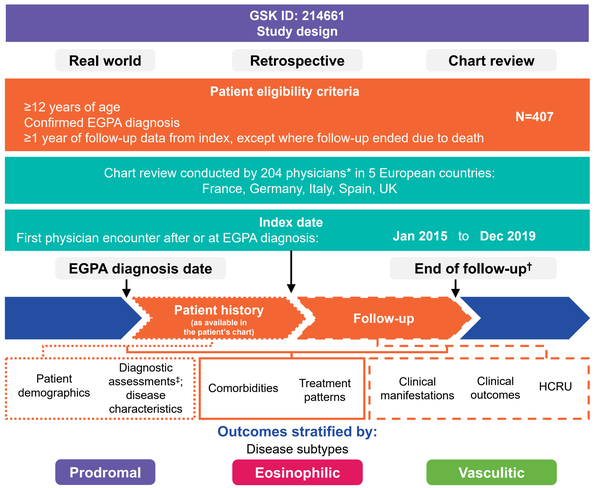
Digital poster
Supplemental data
Narrated summary

Aims

- EGPA is a rare, systemic and progressive inflammatory disease characterised by vasculitis of the small-to-medium-sized blood vessels, hypereosinophilia (defined as levels ≥1000 cells/μL) and asthma.[1–3]
- EGPA is typically characterised by 3 partially overlapping disease phases: prodromal (dominated by respiratory tract symptoms); eosinophilic (blood eosinophilia, eosinophilic tissue infiltration and eosinophil-induced organ damage) and vasculitic (systemic necrotising vasculitis).[4,5] However, these phases do not necessarily progress in the same order, and some patients may not develop vasculitic complications.[6]
- For remission induction, defined as the absence of the typical signs, symptoms or features of active disease, EGPA treatment guidelines recommend glucocorticoids, biologics or combinations of high-dose glucocorticoids and immunosuppressive/cytotoxic therapies depending on the severity of the disease. Maintenance of remission also includes immunosuppressive/cytotoxic therapies or biologics[7]; the former, as well as OCS, often lead to considerable adverse effects.[7–9]
- Our real-world data from a longitudinal chart review study on the treatment patterns, HCRU, clinical symptoms and outcomes for patients with EGPA in Europe have been presented previously[10]; and support the substantial burden of disease associated with EGPA.[11] However, the disease burden by EGPA disease phase is not well characterised.
- Here in this post hoc analysis of the same chart review study, we aimed to describe the demographics, clinical characteristics, treatment patterns and HCRU for patients with EGPA in Europe according to their disease phase.

Methods



*Recruited from targeted specialties of allergy (13%), immunology (6%), rheumatology (44%) and pulmonology (37%); †earliest of death, loss to follow-up or date of chart abstraction; ‡includes blood eosinophil counts; §as recorded in physician's notes based on the latest documentation prior to the index date

Results

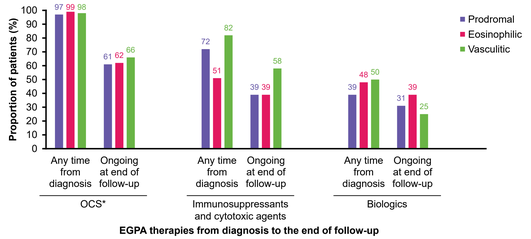
Table 1: **Demographics were generally similar between disease phases, although patients in the vasculitic group were more likely to have comorbidities and higher BEC than the other groups**

Of the 407 patients for whom data were collected, 381 could be subtyped and 26 had unknown disease subtype			
Patient demographics and disease characteristics	**Prodromal (N=36)**	**Eosinophilic (N=220)**	**Vasculitic (N=125)**
Male, n (%)	19 (53)	**127 (58)**	**73 (58)**
Age at EGPA diagnosis, mean (SD), years	42.7 (15.8)	42.8 (15.1)	**43.3 (14.7)**
≥18 years of age, n (%)	33 (92)	**206 (94)**	**118 (94)**
EGPA diagnosis, n (%)			
Before study (≤2014)	**4 (11)**	**25 (11)**	13 (10)
During study (2015–2019)	32 (89)	195 (89)	**112 (90)**
Time from asthma diagnosis to EGPA diagnosis, median (IQR)*, years	**3.0 (0.3, 8.9)**	1.5 (0.1, 4.3)	2.9 (0.1, 6.1)
Proportion of patients with asthma, n (%)	21 (58)	164 (75)	**95 (76)**
Length of follow-up, mean (SD), years	2.6 (1.4)	2.7 (1.5)	**2.8 (1.4)**
Country, n (%)			
France	**9 (25)**	47 (21)	21 (17)
Germany	4 (11)	40 (18)	**33 (26)**
Italy	4 (11)	44 (20)	**28 (22)**
Spain	**9 (25)**	48 (22)	20 (16)
UK	**10 (28)**	41 (19)	23 (18)
BEC			
n (%)	**33 (92)**	196 (89)	**115 (92)**
Median (IQR), cell/μL	1200 (120, 2000)	1500 (560, 3300)	**1800 (1000, 5000)**
Comorbidities, n (%)			
Vasculitis	8 (22)	87 (40)	**92 (74)**
Hypertension	10 (28)	81 (37)	**61 (49)**
Anxiety or depression	12 (33)	70 (32)	**54 (43)**
Lower respiratory disease(s)†	6 (17)	42 (19)	**29 (23)**
Osteoporosis	6 (17)	34 (16)	**27 (22)**
Glomerulonephritis	1 (3)	26 (12)	**39 (31)**
Obesity	6 (17)	34 (16)	**23 (18)**
Diabetes	**5 (14)**	15 (7)	13 (10)
Rheumatoid arthritis	0 (0)	9 (4)	**10 (8)**
Liver disease	**1 (3)**	**6 (3)**	**4 (3)**
Other	**2 (6)**	11 (5)	**8 (6)**
Cancer (any)‡	0 (0)	4 (2)	**4 (3)**

*The highest values (or proportions) in each row are shown in bold. *Among 154 patients who had asthma diagnosed before EGPA; †other than asthma and COPD; ‡reported cancer types included: lung (3), breast (1), colon (1), leukaemia (1), rectum (1), skin (1)*

Figure 1: **At any time from diagnosis, OCS use was the most common treatment received among all disease subtypes; patients in the vasculitic phase were most likely to receive each treatment type. By the end of follow-up, fewer patients overall were receiving treatments**

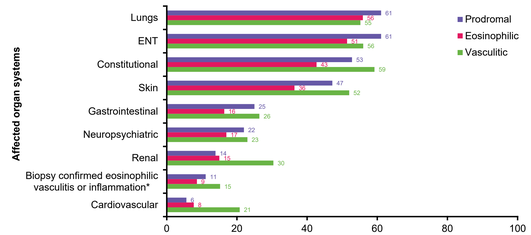
EGPA therapies	Prodromal (N=36)	Eosinophilic (N=220)	Vasculitic (N=125)
Number of distinct EGPA therapies used per patient, mean (SD)†	3.8 (2.1)	3.7 (1.8)	4.4 (1.7)
Proportion of patients using ≥3 distinct EGPA therapies, n (%)†	23 (64)	157 (71)	108 (86)
Maximum daily dose of OCS (mg), mean (SD)‡,§	37.8 (19.5)	29.1 (19.6)	29.8 (19.7)



For maintenance therapy; †receipt of 1 or multiple oral corticosteroid drugs was counted as a single therapy; ‡in patients with a reported maximum daily dose available for maintenance therapy (prodromal n=29 [81%], eosinophilic n=171 [78%] and vasculitic n=97 [78%]); §for patients who received ≥1 prescription for a drug, the maximum dosage was reported as the maximum dosage across all prescriptions

Figure 2: **Patients with vasculitic disease generally had a higher number of distinct clinical manifestations than patients with prodromal or eosinophilic disease; lungs, ENT, constitutional and skin were the most commonly affected organ systems**

Clinical manifestations	Prodromal (N=36)	Eosinophilic (N=220)	Vasculitic (N=125)
Number of distinct clinical manifestations, median (IQR)	3.0 (1.5, 6.0)	3.0 (1.0, 5.0)	4.0 (2.0, 7.0)
Patients with ≥6 distinct clinical manifestations, n (%)	10 (28)	48 (22)	46 (37)



The biopsy sites were skin (20), lung (9), kidney (3), muscle (2), alveoli (1), lower extremity (1), intestine (1), peripheral nerve (1), and unknown (4)

Conclusion

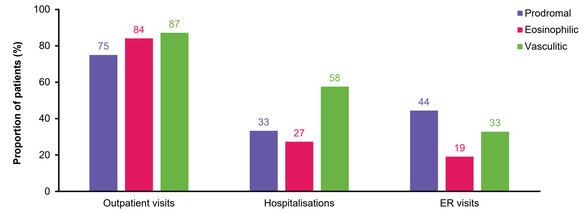
- This sub-analysis of a retrospective longitudinal chart review study demonstrated that EGPA significantly impacts both patients and healthcare systems in Europe, highlighting the need for improved EGPA diagnosis and management, particularly among patients in the vasculitic disease phase.
  - Asthma was common amongst patients with EPGA (lowest among prodromal); the most common comorbidities were vasculitis, hypertension, and anxiety or depression, all of which were more frequently observed in patients in the vasculitic phase compared with those in the prodromal and eosinophilic phases.
  - Most patients received multiple EGPA therapies to manage their disease; patients with vasculitic disease were more likely to be receiving ≥3 distinct therapies.
  - Patients experienced manifestations in multiple organ systems; those in the lungs and ENT were highest for patients in the prodromal phase in line with the description of the phases in the reported literature.[4,5] Constitutional was highest for patients in the vasculitic phase followed by skin and renal; neuropsychiatric was also higher in this group, all of which are as expected based on the literature.[12]
  - Not all patients experienced remission during the follow-up period; overall, there was a median delay of 13–16 months between diagnosis and first remission. Relapses were also common, particularly for those patients in the vasculitic phase (almost one-third [29%]).
  - Most patients had multiple outpatient visits; hospitalisations were highest for those patients in the vasculitic phase, though ER visits were more frequent for those in the prodromal phase.

Table 2: **Patients in the vasculitic phase were more likely to relapse; remission rates were similar between eosinophilic and vasculitic patients**

Clinical outcomes	Prodromal (N=36)	Eosinophilic (N=220)	Vasculitic (N=125)
Patients who experienced remission*, n (%)	17 (47)	133 (61)	79 (63)
Duration of remission(s), median (IQR) cumulative months†	16.0 (7.5, 28.5)	10.6 (4.6, 20.5)	13.4 (7.0, 24.7)
Time from diagnosis to first remission, median (IQR) months	14.3 (7.9, 31.6)	15.9 (8.1, 27.2)	12.5 (6.9, 24.2)
Patients who experienced a relapse‡, n (%)	8 (22)	32 (15)	36 (29)
Annualised number of relapses (among patients with a relapse), PPPY, median (IQR)	0.4 (0.3, 0.6)	0.6 (0.3, 1.0)	0.6 (0.4, 1.0)

Remission was defined by the physician. Definitions used included BVAS=0, OCS dose ≤4 mg/day and other; †the number of remissions were reported among the patients who had ≥1 remission. For patients who had >1 remission, the duration of remission was calculated as the sum of durations of remission for all reported remissions; ‡relapse was defined as an increase in OCS dose, an increase/change in dose of immunosuppressive therapy, or hospitalisation. Indication of relapse based on other definitions was also included

Figure 3: **Patients in the vasculitic phase had a higher HCRU than those in the prodromal or eosinophilic phases; ER visits were highest for those patients in the prodromal phase**



Abbreviations

BEC, blood eosinophil count; BVAS, Birmingham vasculitis activity score; COPD, chronic obstructive pulmonary disease; EGPA, eosinophilic granulomatosis with polyangiitis; ENT, ear, nose and throat; ER, emergency room; HCRU, healthcare resource utilisation; IQR, interquartile range; OCS, oral corticosteroids; PPPY, per person per year; SD, standard deviation.

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Disclosures

This study was funded by GSK (GSK ID: 214661).
On behalf of all authors, an audio recording of this poster was prepared by **LB**, who did not receive any payment for this recording.
LB, **JH**, **RA-C** and **WA** are employees of GSK and holds stocks/shares in GSK. **LH**, **AK** and **MSD** are employees of Analysis Group, Inc., which received funding from GSK to conduct the study.
Editorial support (in the form of writing assistance, including preparation of the draft poster under the direction and guidance of the authors, collating and incorporating authors' comments for each draft, assembling tables and figures, grammatical editing and referencing) was provided by Hayley Butler, PhD, and Sarah Farrar, PhD, at Fishawack Indicia Ltd, UK, part of Fishawack Health, and was funded by GSK.

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