Disease Burden Related to Eosinophilic Granulomatosis With Polyangiitis (EGPA): A Targeted Literature Analysis

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Background

- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as Churg-Strauss syndrome) is a rare, potentially organ- and life-threatening disease characterized by systemic eosinophilia, airway disease, and multisystem small-vessel vasculitis[1,2]

Objective

- To highlight the current evidence on disease burden and the path to diagnosis in EGPA from a broader targeted literature review to understand the epidemiology and management of EGPA

Methods

- A literature review was designed to search for real-world and randomized, controlled studies in EGPA using MEDLINE and EMBASE; the article search was performed on November 23, 2020, and was semisystematic, with a targeted search of the following:
  - MEDLINE and EMBASE for journal articles and conference abstracts (EGPA + real world evidence [RWE] and randomized controlled trial [RCT] study filter, in the English language, in the last 5 and 10 years, respectively)
  - Supplemented materials with searches of clinicaltrials.gov (EGPA + RCT + last 10 years) and Google (EGPA + clinical practice guidelines)
- Outcomes of interest for this literature search are shown in **Figure 1**
- Articles were screened according to PICOTS criteria:
  – Population | – Intervention | – Comparator | – Outcomes | – Timeframe | – Study type
- Numerical results are presented as a range or simple average, as reported from the relevant studies identified

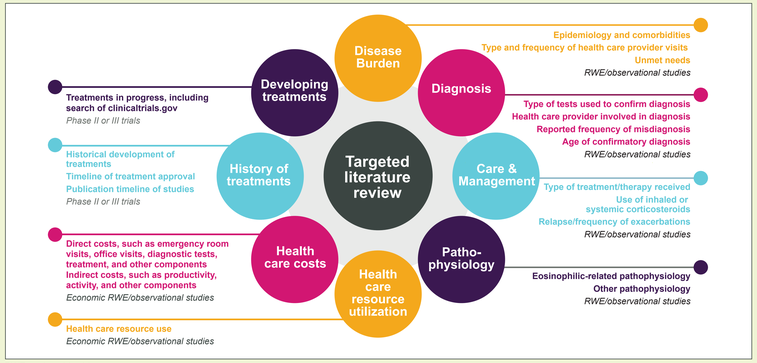
Results

- Of 1023 references identified, 526 potentially relevant articles were eligible for full-text screening; of these, 33 priority articles were selected for an intensive review (**Figure 2**)
- Significant evidence gaps exist in the understanding of the epidemiology, natural history, and economic burden of EGPA (**Figure 3**)
- More than 80% of patients with EGPA have comorbid asthma, sinusitis, or rhinitis; manifestations occurring in >15% of patients included polyneuropathy, ocular involvement, cardio-cerebrovascular disease, and nasal polyposis (**Figure 4**)
- Between 6% and 30% of patients with EGPA overlap with hypereosinophilic syndrome, and approximately one-third of patients with EGPA are antineutrophilic cytoplasmic antibody (ANCA) positive (**Figure 5**)
- Nonspecific presenting features lead to an evaluation by multiple specialists and delayed diagnoses (average [range] age of diagnosis: 53 [14-77] years) (**Figure 6**)
- Exposure to oral corticosteroids is high in EGPA patients, along with high annual hospitalization and emergency room (ER) visit rates (**Figure 7**)

Key Takeaways

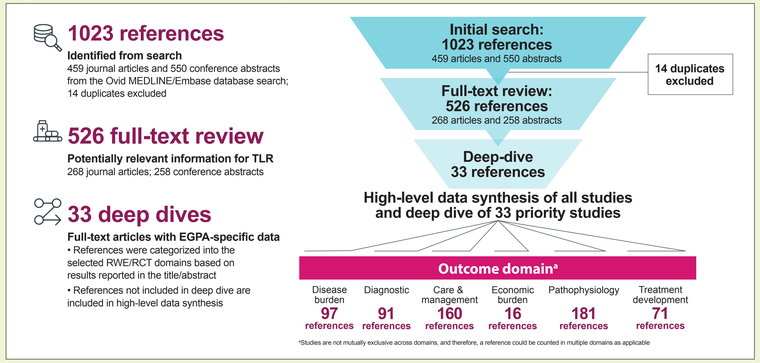
- The incidence and prevalence of EGPA is likely to be grossly underestimated
- Improved diagnostic approaches (e.g. predictive modeling and machine learning algorithms) are necessary to identify potential patients with EGPA before they experience serious health events that can lead to hospitalization and increased healthcare resource utilization
- Improved coordination among various medical specialties is necessary to refer, diagnose, and manage EGPA
- There is a need for increased awareness of the short and long term side-effects of steroid use

Figure 1. Outcomes of interest for targeted literature review



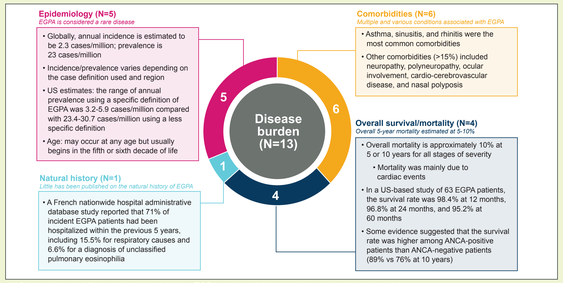
RWE, real world evidence.
[a]Studies are not mutually exclusive across domains; a reference could be counted in multiple domains, as applicable.

Figure 2. Literature search and study identification



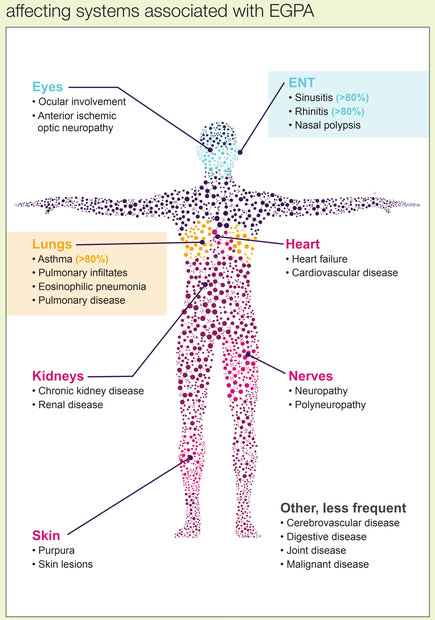
TLR, targeted literature review.
[a]Studies are not mutually exclusive across domains; a reference could be counted in multiple domains, as applicable.

Figure 3. Summary of literature about EGPA disease burden



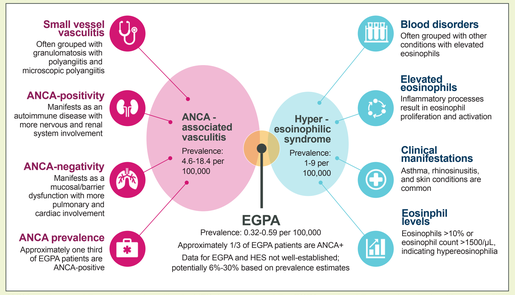
ANCA, antineutrophilic cytoplasmic antibody; EGPA, eosinophilic granulomatosis with polyangiitis.

Figure 4. Comorbidities and manifestations affecting systems associated with EGPA



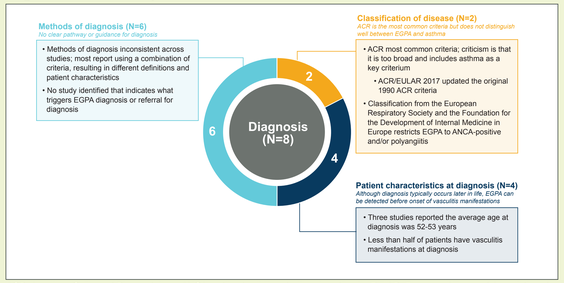
EGPA, eosinophilic granulomatosis with polyangiitis; ENT, ear, nose, and throat.

Figure 5. Overlap of EGPA with ANCA-associated vasculitis and hypereosinophilic syndrome[3-5]



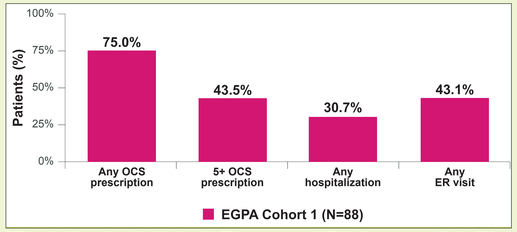
ANCA, antineutrophilic cytoplasmic antibody; HES, hypereosinophilic; EGPA, eosinophilic granulomatosis with polyangiitis.

Figure 6. Summary of literature about EGPA diagnosis



ACR, American College of Rheumatology; ANCA, antineutrophilic cytoplasmic antibody; EULAR, European Alliance of Associations for Rheumatology; EGPA, eosinophilic granulomatosis with polyangiitis.

Figure 7. OCS use and HCRU in a cohort of patients with EGPA[a]



EGPA, eosinophilic granulomatosis with polyangiitis; ER, emergency room; HCRU, health care resource utilization; OCS; oral corticosteroids.
[a]Case definition with higher specificity (>PPV).

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Disclosures

Hitesh Gandhi and Yen Chung: employees and may own stock—AstraZeneca. Kristina Lindsley and Jennifer Uyei: employees—IQVIA. IQVIA received research funding from AstraZeneca to complete this study.

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