Acceptance code: RWD95

Eosinophilic granulomatosis with polyangiitis: estimation of the clinical and economic burden in France - a real-world retrospective study based on the National Health Data System (SNDS)

Tauty S*[1], Brouquet A[1], Bugnard F[1], Bénard S[1], Cottin V[2], Taillé C[3], Guillevin L[4], Puéchal X[4], Terrier B[4]

[1] **stève** consultants, Oullins, France [2] *pneumology, hôpital Louis Pradel, Lyon, France* [3] *pneumology, AP-HP Bichat, Paris, France* [4] *internal medecine, AP-HP Cochin, Paris, France*

*stauty@steve-consultants.com

Rationale

Eosinophilic Granulomatosis with PolyAngiitis (EGPA) = Churg-Strauss syndrome

Systemic necrotizing vasculitis of small vessels, associated with anti-neutrophil cytoplasmic antibodies (ANCA)

Rare disease : prevalence 7.3-18 / 1,000,000 ; incidence 0.9-4 / year / 1,000,000

Diagnosis in adulthood, exceptional pediatric forms

Three-phase course:

1. **Asthma and allergic manifestations**
2. **Eosinophilia and pulmonary infiltrates**
3. **Systemic manifestations**

Treatment with **corticosteroids**, **immunosuppressants** and **interleukin-5 inhibitors**

Few comprehensive real-life data

Objectives

- Epidemiology
- Patient characteristics
- Treatment use
- Complications, relapses and death
- Healthcare resource use and associated costs

Methods

EGPA patients were identified in the SNDS by the presence of at least one of the following criteria, **between 2010 and 2019**:

- A **Long Term Disease** (LTD) with a diagnosis ICD-10 code **M30** (periarteritis nodosa and related conditions) or **M301** (periarteritis with pulmonary involvement [Churg-Strauss])
- A **hospitalization** with a diagnosis ICD-10 code **M301** (DP - main, DR - secondary, DAS - significant associated)

As the M30 code is not specific to EGPA, patients included only by a M30 LTD and presenting a hospitalization with a diagnosis of another vasculitis (M30x except M301) were excluded from the study population. Patients hospitalized with a code M317 (microscopic micropolyangiitis) were also excluded.

EGPA epidemiology was estimated between 2013 and 2019, in order to have at least 3 years of history for differentiating prevalent and incident cases. **Incident patients** were defined by the absence of LTD, hospitalizations or EGPA treatment in the historical period.

To assess the **disease burden**, patients newly diagnosed in 2012-2016 were followed from their index date (1st disease marker) until the end of 2019, ensuring a minimum follow-up of 3 years. Comorbidities and medical history were assessed within 2 years prior to the index date.

EGPA relapses were defined as a doubling of the corticosteroid doses between 2 consecutive dispensations OR dispensation of a new EGPA treatment OR hospitalization with a DP M301. Time to relapse was estimated using the Kaplan-Meier method.

EGPA hospitalizations were identified by an ICD-10 code M301 as DP/DR/DAS. **Reimbursed costs** were calculated per patient for each year after index date. Both inpatient and outpatient costs were included.

Results

Figure 1. Patient selection

SNDS extraction: patients with at least one of the following criteria between 2010 and 2019 : LTD M30 or M30x or hospitalization M301 **(n=13,334)**

- Non-valid records (n=3,025)
- *Among valid records:* LTD M30x except M301 (n=861)

Patients with valid records and with at least one of the following criteria between 2010 and 2019: LTD M30 or M301 or hospitalization M301 **(n=9,448)**

- Hospitalization M317 (n=465)
- *If included by LTD M30*: hospitalization M30x except M301 (n=1,270)
  - Deceased before 01JAN2012 (n=348)
  - *If incident*: previous dispensation of a treatment of interest (N=373)

EGPA patients 2012-2019 **(n=7,238)**

- Newly diagnosed EGPA patients 2012-2016 **(n= 1,650)** — *Longitudinal analyses*
- EGPA patients 2013-2019 **(n=7,084)** — *Epidemiology*

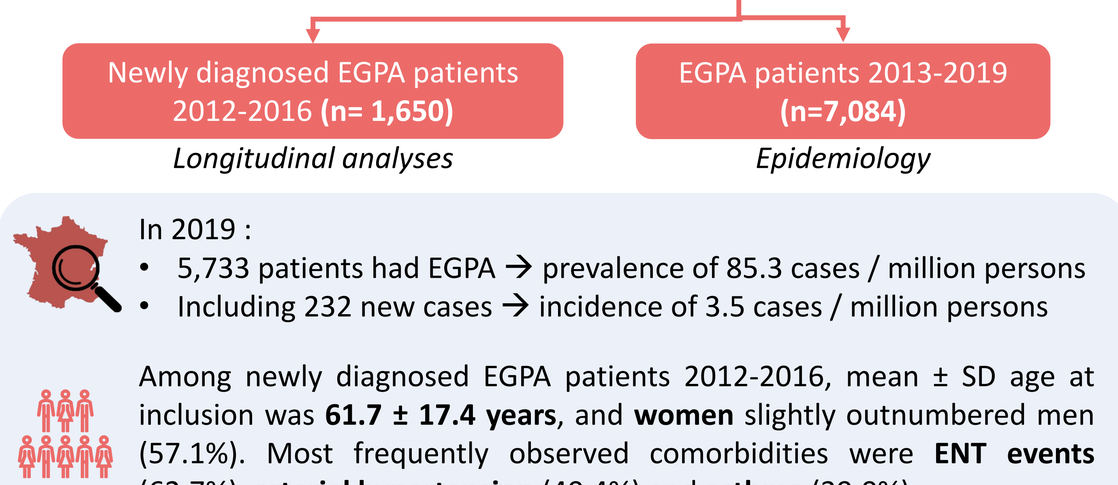
In 2019 :

- 5,733 patients had EGPA → prevalence of 85.3 cases / million persons
- Including 232 new cases → incidence of 3.5 cases / million persons

Among newly diagnosed EGPA patients 2012-2016, mean ± SD age at inclusion was **61.7 ± 17.4 years**, and **women** slightly outnumbered men (57.1%). Most frequently observed comorbidities were **ENT events** (62.7%), **arterial hypertension** (40.4%) and **asthma** (39.0%).

During the 1st year of follow-up, 72.0% of patients had at least one EGPA treatment dispensation and 78.1% had at least one asthma treatment

Figure 2. Mean monthly dose of oral corticosteroids (mg)



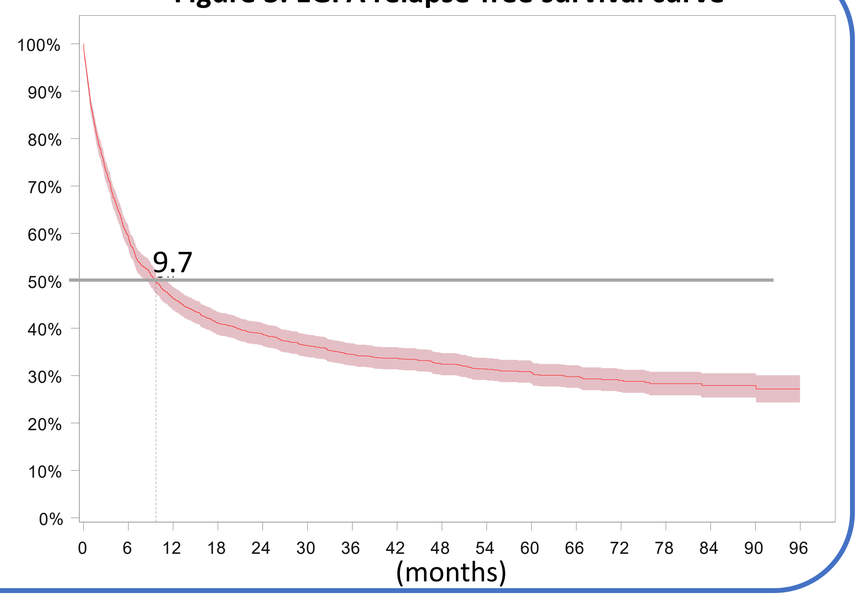
Regarding the development of comorbidities that may be related to chronic corticosteroid use

- 21.6% of patients developed **osteoporosis** during follow-up,
- 20.7% developed **arterial hypertension**,
- 10.9% developed **diabetes**.

EGPA **relapse** rate (% [IC95]) was

- 53.8% [51.3%-56.2%] at 1 year,
- 65.6% [63.2%-67.9%] at 3 years,
- 69.3% [66.9%-71.6%] at 5 years,
- With a median EGPA relapse-free survival of 9.7 [8.5-11.2] months.

Figure 3. EGPA relapse-free survival curve



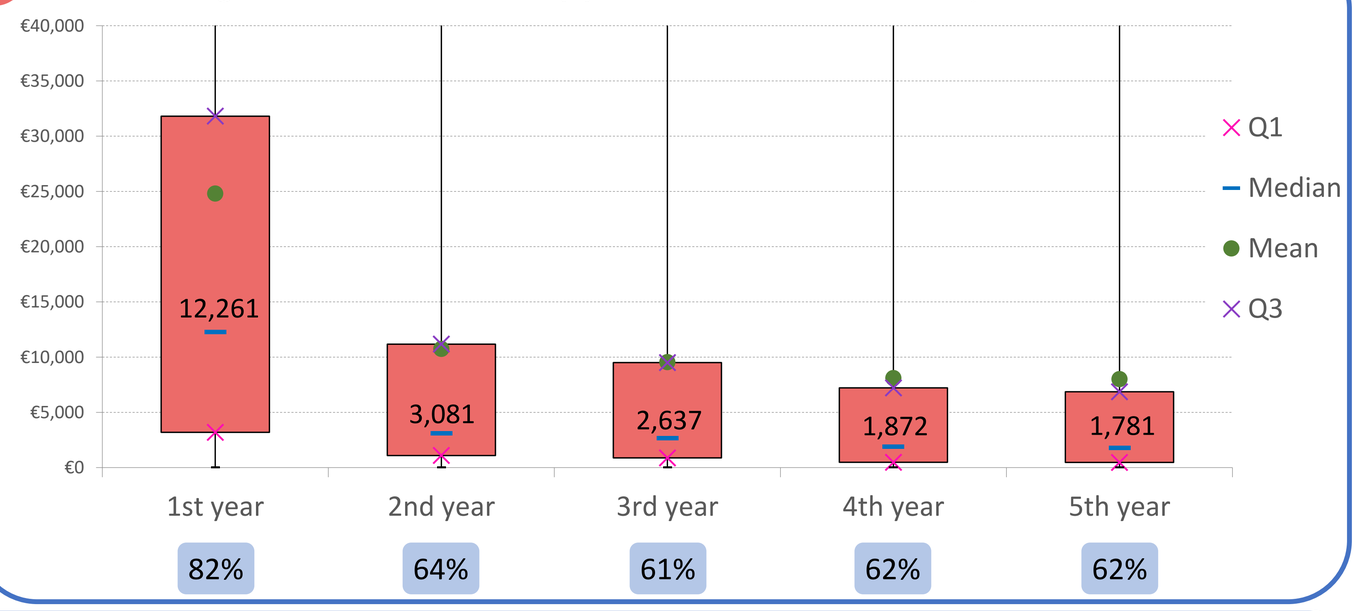
Among newly diagnosed EGPA patients 2012-2016, the mean annualized number of hospitalizations and the associated cumulative length of stay were:

- 2.9 all-cause hospitalizations for 23.4 days
- 0.6 hospitalizations related to EGPA for 4.5 days
- 1.3 hospitalizations related to infections / cardiovascular disease / osteoporotic fracture for 21.1 days

Table 1. Annualized number and cumulative length of stay of hospitalizations

	Number		Cumulative length of stay	
	Mean (SD)	Median (Q1-Q3)	Mean (SD)	Median (Q1-Q3)
All-cause	2.9 (9.1)	1.2 (0.5 - 2.7)	23.4 (59.2)	5.4 (1.5 – 16.7)
Related to EGPA	0.6 (2.3)	0.0 (0.0 - 0.5)	4.5 (19.3)	0.0 (0.0 – 2.0)
Related to infections / cardiovascular disease / osteoporotic fracture	1.3 (5.7)	0.3 (0.0 - 1.1)	21.1 (74.1)	2.3 (0.0 – 11.1)

Figure 4. Reimbursed costs (€) and % of costs linked to hospitalizations



Conclusion

This SNDS study assesses the epidemiology of EGPA in the overall French population. It presents the drug management of EGPA patients, with corticosteroid doses no longer decreasing from the 3rd year onwards, which can lead to significant undesirable effects, and provides recent data on the clinical burden, characterized by frequent and rapid relapses. The study also highlights the economic burden, with high costs during the first year, mainly due to hospitalizations. The identification of EGPA by the non-specific M30 code is a limitation of this study. Matching the SNDS with the *Groupe Français d'Etude des Vascularites* (GFEV) registry will provide additional clinical data and the opportunity to refine the identification of EGPA in the SNDS.

We would like to thank the members of the GFEV registry, especially Pr Benjamin Terrier, and AstraZeneca for their investment in this project.