One-Year Burden of Crises and Exacerbations in Myasthenia Gravis: Real-World Evidence from France (RELIEF Study)

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Introduction

- Myasthenia gravis (MG) can progress to generalized (gMG) form despite management strategies combining symptomatic treatments, immunosuppressive treatments (steroidal or non-steroidal), and rescue interventions (e.g., intravenous immunoglobulins (IVIg) or plasma exchange (PE)) (1).
- Acute events, which may arise and can be life-threatening, substantially contribute to the medical, quality-of-life, and economic burden of myasthenia gravis (MG) (2-6).
- Building on our findings of early disease progression in MG (7), we observed that initial exacerbations and crises predominantly occur within the first year after MG diagnosis, supporting the focus on early acute burden in this analysis.

Objective

The objective was to evaluate the one-year risk of MG crises and exacerbations after diagnosis of MG and gMG and characterize the nature of these acute episodes.

Methods

- **Study design:** retrospective cohort study
- **Data source:** French health claims database - Système National des Données de Santé (SNDS) (8)
- **Analyses period:** 01/01/2022 to 12/31/2023
- **Follow-up:** at least one year (except in case of death).

Selection of populations

- Specific algorithms based on diagnosis codes and treatment dispensations were applied to identify two populations between 01/01/2022 and 12/31/2022 : adults with a first MG diagnosis and adults with a first gMG diagnosis (see Table 1)

Definitions and statistical methods

- **Crises:** hospitalization with an associate diagnosis of MG AND respiratory failure (main or related diagnosis), intubation, ventilation, or enteral feeding.
- **Exacerbations :** crisis, acute IVIg or PE, hospitalization for MG (main diagnosis), or hospitalization for dysphagia (main or related diagnosis) with an associate diagnosis of MG. Exacerbations separated by less than 2 months were considered as a single event.
- **Analyses:** incidence rate of MG crises/exacerbations, number of patients with at least one MG crisis/exacerbation, number of MG crises/exacerbations among patients with at least one MG crisis/exacerbation .

Limitations

- The algorithms used to identify patients with MG and gMG as well as crises and exacerbations were developed with the study's Scientific Committee, based on the 2015 national diagnostic and care protocol (PNDS) (1) and published literature (9-11), ensuring methodological relevance. However, limited diagnostic precision and the lack of external validation may lead to false positives or negatives.
- Because this study is a retrospective longitudinal analysis using SNDS data, it is subject to potential selection and time-related biases.

Conclusion

- MG crisis is a life-threatening acute event whose incidence is not negligible in the first year.
- MG exacerbations are frequent in the first year following the diagnosis, particularly in gMG patients .
- These results highlight the importance of prompt therapeutic intervention in gMG patients .

Results

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Key results

Crises:

- Affect 6% of patients with MG and 10% of patients with gMG in the year following the diagnosis
- The incidence rate of crisis is 1,7 fold higher in gMG vs. MG

Exacerbations :

- Affect 57% of patients with MG and 72% of patients with gMG in the year following the diagnosis
- The incidence rate of exacerbations is 2 fold higher in gMG vs. MG

Acute burden is substantial the first year after diagnosis.

Characteristics

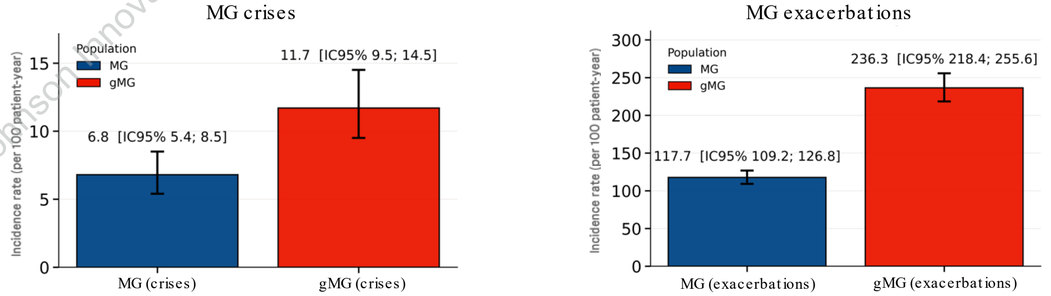
Table 1: Characteristics of adults with MG or gMG with a first diagnosis in 2022

	MG (N = 1,197)	gMG (N = 859)
Identification principle in the SNDS	Long-term disease for MG OR at least two hospitalizations for MG OR at least one hospitalizations for MG and two dispensing of acetylcholinesterase inhibitor	Immunosuppressive therapy OR second-line immunosuppressant [a] OR IVIg OR PE OR new generation therapy OR thymectomy
Male, n (%)	607 (50.7%)	429 (49.9%)
Age at diagnosis, mean (SD), (in years)	62.4 (18.0)	63.9 (17.4)
Comorbidities, n (%)*		
Type 1 and type 2 diabetes	124 (10.4%)	119 (13.9%)
Thymoma	20 (1.7%)	64 (7.5%)
Depressive episode	31(2.6%)	29 (3.4%)
Duration of disease prior to the diagnosis of gMG, mean (SD), (in years)	-	1.8 (4.9)

*Abbreviations: gMG : generalized myasthenia gravis;IVIg: intravenous immunoglobulins;MG : myasthenia gravis; PE: plasma exchange;SD: standard deviation. *Comorbidities were identified during the year preceding the first recorded diagnosis of MG or gMG, based on ICD-10-coded diagnoses from MCO hospital stays or LTD status. a. Except cyclophosphamide*

Crises and exacerbations

Figure 1: Description of 1-year incidence rate of MG crises and exacerbations in adults with MG or gMG with a first diagnosis in 2022



Crises and exacerbations

Table 2: Description of MG exacerbations and within one year following diagnosis in adults newly diagnosed with MG or gMG in 2022

	MG (N = 1,197)	gMG (N = 859)
Myasthenia gravis crises		
Number of patients with at least one MG crisis	76 (6.3%)	89 (10.4%)
Number of MG crises, among patients with at least one MG crisis, mean (SD) [95% CI]	1.1 (0.2) [1.0 ; 1.1]	1.1 (0.2) [1.0 ; 1.1]
Number of patients with at least one criterion for MG crisis		
Hospitalization for respiratory failure (main or related diagnosis)	34 (44.7%)	45 (50.6%)
Intubation	35 (46.1%)	40 (44.9%)
Mechanical ventilation	62 (81.6%)	69 (77.5%)
Enteral feeding	⩽ 10	⩽ 10
Myasthenia gravis exacerbations		
Number of patients with at least one MG exacerbation	685 (57.2%)	619 (72.1%)
Number of MG exacerbations, among patients with at least one MG exacerbation, mean (SD) [95% CI]	1.3 (0.5) [1.2 ; 1.3]	1.3 (0.6) [1.3 ; 1.4]
Number of patients with at least one criterion for MG exacerbation		
Acute immunoglobulin	433 (63.2%)	547 (88.4%)
Acute plasma exchange	59 (8.6%)	72 (11.6%)
Hospitalization with MG	607 (88.6%)	483 (78.0%)
Hospitalization for dysphagia	12 (1.8%)	12 (1.9%)
Crisis	76 (11.1%)	89 (14.4%)

Conflicts of Interest

Thierry Gendre: consulting fees from J&J, UCB, and Amgen; paid oral presentations from Argenx and CSL Behring; conference expenses from Alexion. Didier Theis and Jean-Baptiste Noury: consulting fees from J&J . Erika Guyot, Maeva Nolin, Manon Belhassen employees of Epimentis (PELyon). Ingrid Rodriguez, Julia Meijer, Laurène Gautier and Julien Dupin: employees of J&J . The study is sponsored by J&J .

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