Risk of myasthenia gravis exacerbation and level of healthcare resource utilization by Myasthenia Gravis Activities of Daily Living score

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Angela Ting[1], Minjee Park[2], Oshin Sangha[3], Wendi Huff[4], Mohita Kumar[1], Jean-François Ricci[2], Edward Lee[1]

[1]UCB Pharma, Atlanta, GA, USA; [2]Alira Health, Basel, Switzerland; [3]Alira Health, Vancouver, Canada; [4]Myasthenia Gravis Foundation of America, Westborough, MA, USA

Introduction

- MG is a rare, chronic, autoimmune disease of the neuromuscular junction, characterized by fluctuating chronic muscle weakness and fatigue[1]
- Many patients with MG continue to experience limitations in their daily activities, exacerbation of myasthenic symptoms, or life-threatening myasthenic crises, despite treatment with conventional therapies[2,3]
  - Patients with exacerbation of symptoms also have higher levels of HCRU compared with patients who do not have a history of myasthenic crises or exacerbation[4]
- The MG-ADL scale is a validated, eight-item, patient-reported outcome measure developed to assess MG symptoms and effects on patients' daily activities[5]
  - It is a standard outcome measure in clinical trials to assess the severity of MG symptoms[5]
- There are limited data available on the relationship between MG-ADL scores and risk of MG exacerbations and the characterization of exacerbation-related HCRU. Here, we report the results of a retrospective, cross-sectional, observational study of patient characteristics, exacerbation risk in relation to MG-ADL score, and level of HCRU following exacerbation, using data from the MGFA Global MG Patient Registry (MGFAPR)
  - The MGFAPR is a voluntary, online, patient-reported registry that captures the natural history of MG among patients in the USA, hosted on the Health Storylines™ platform (https://www.mgregistry.org/)

Methods

- This study included USA participants, aged ≥18 years, with a self-reported diagnosis of MG and complete MG-ADL data who enrolled in the registry between July 01, 2013, and September 30, 2022
  - Survey data at enrollment and at patients' first follow-up at 6 months were assessed
- In the MGFAPR, an MG exacerbation was defined as having new symptoms, or worsening of old symptoms, for a duration of at least 7 days and a gap of at least 30 days since the last exacerbation
- Exacerbation-related HCRU was defined as the use of IVIg or PLEX due to an exacerbation in the last 6 months, or overnight hospitalization due to 'exacerbation or worsening of MG' in the last 6 months
- Descriptive statistics for patient socio-demographics, disease characteristics and level of HCRU at enrollment were stratified by MG-ADL score: 0–1, 2–4, 5–7, 8–10, 11–13 and 14+
- Negative binomial regression was used to assess the relationship between MG-ADL score and exacerbation. Two regression models used MG-ADL score as either a count or categorical variable

Results

- Of 3416 patients identified from the MGFAPR, 2092 (61%) were female
- A greater proportion of patients in the higher MG-ADL score groups (MG-ADL >7) were female, younger at the time of MG diagnosis, African American, or unemployed compared with patients in the lower MG-ADL score groups (MG-ADL≤7) (**Table 1**)
- Overall, the proportion of patients who had an MG exacerbation increased in the higher MG-ADL score groups, with almost 80% of patients in the MG-ADL 14+ score group having had an exacerbation in the past 6 months (**Figure 1**)
- For each additional point in MG-ADL score, the rates of exacerbations increased by 13% (IRR: 1.13; 95% CI: 1.11, 1.15; p<0.001)
  - A similar result was observed when MG-ADL was used as a categorical variable, where, compared with MG-ADL scores of 0–1, adjusted exacerbation IRRs significantly increased with higher MG-ADL scores from 2.00 (MG-ADL score 2–4; p<0.001) to 7.22 (MG-ADL score 14+; p<0.001) (**Figure 2**)
- A greater proportion of patients in the higher MG-ADL score groups had comorbidities such as anxiety, depression, autoimmune diseases (ATD, IBD, SLE and rheumatoid arthritis), osteoporosis, asthma and COPD (p<0.001)
- Approximately 50% of patients with one exacerbation at enrollment (n=778) required treatment with IVIg, PLEX, or overnight hospitalization following their exacerbation (**Figure 3**)

Table 1 Socio-demographic and disease characteristics by MG-ADL score group at enrollment

	MG-ADL score 0–1 (n=358)	MG-ADL score 2–4 (n=783)	MG-ADL score 5–7 (n=979)	MG-ADL score 8–10 (n=734)	MG-ADL score 11–13 (n=400)	MG-ADL score 14+ (n=162)
Age at diagnosis, years*						
Mean (SD)	**50.8** (18.9)	**51.8** (18.2)	**50.2** (17.4)	**47.6** (16.6)	**46.2** (15.3)	**45.8** (15.8)
Sex, n (%)*						
Female	**181** (50.6)	**398** (50.8)	**571** (58.3)	**499** (68.0)	**316** (79.0)	**127** (78.4)
Male	**177** (49.4)	**385** (49.2)	**408** (41.7)	**235** (32.0)	**84** (21.0)	**35** (21.6)
Race, n (%)†						
White	**324** (90.5)	**677** (86.5)	**842** (86.0)	**619** (84.3)	**332** (83.0)	**128** (79.0)
African American	**16** (4.5)	**37** (4.7)	**49** (5.0)	**34** (4.6)	**21** (5.3)	**11** (6.8)
Asian	**6** (1.7)	**12** (1.5)	**12** (1.2)	**8** (1.1)	**1** (0.3)	**1** (0.6)
Indigenous	**0** (0)	**1** (0.1)	**4** (0.4)	**3** (0.4)	**1** (0.3)	**0** (0)
Other	**12** (3.4)	**56** (7.2)	**72** (7.4)	**70** (9.5)	**45** (11.3)	**22** (13.6)
Employment status, n (%)*						
Unemployed	**167** (46.6)	**422** (53.9)	**491** (50.2)	**436** (59.4)	**277** (69.3)	**118** (72.8)
Full-time	**130** (36.3)	**286** (36.5)	**387** (39.5)	**220** (30.0)	**98** (24.5)	**29** (17.9)
Part-time	**55** (15.4)	**71** (9.1)	**97** (9.9)	**75** (10.2)	**25** (6.3)	**14** (8.6)
Unknown or missing	**6** (1.7)	**4** (0.5)	**4** (0.4)	**3** (0.4)	**0** (0)	**1** (0.6)
Treatment for MG, n (%)‡						
Pyridostigmine*	**196** (54.7)	**566** (72.3)	**751** (76.7)	**603** (82.2)	**338** (84.5)	**140** (86.4)
Prednisone	**143** (39.9)	**332** (42.4)	**446** (45.6)	**328** (44.7)	**171** (42.8)	**80** (49.4)
Mycophenolate	**99** (27.7)	**162** (20.7)	**203** (20.7)	**159** (21.7)	**92** (23.0)	**39** (24.1)
IVIg or SCIg*	**27** (7.5)	**83** (10.6)	**164** (16.8)	**148** (20.2)	**96** (24.0)	**51** (31.5)
Azathioprine	**55** (15.4)	**116** (14.8)	**141** (14.4)	**103** (14.0)	**54** (13.5)	**27** (16.7)
Disease duration, years*						
Mean (SD)	**8.9** (11.5)	**6.6** (9.6)	**5.5** (8.3)	**6.5** (10.2)	**6.0** (8.7)	**5.8** (8.6)

Significant association with increasing MG-ADL score: *p<0.001. †p=0.013. ‡For the 'Treatment for MG' category, only the top five highest percentages are shown and one patient may have received more than one treatment.
MG-ADL scores range from zero (normal) to 24 (most severe).

Summary and conclusions

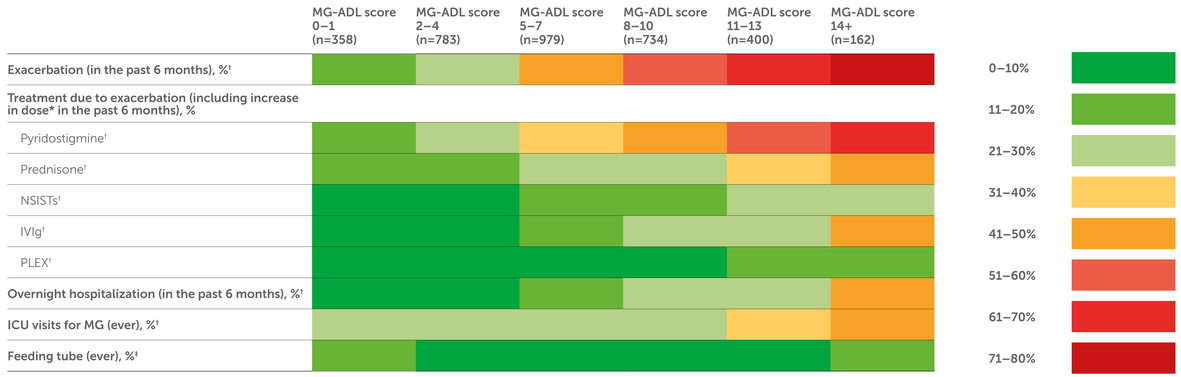
This study evaluated the socio-demographics, disease characteristics and HCRU of patients with MG stratified by categorized MG-ADL scores in a real-world MG population

There were socio-demographic disparities in disease severity at enrollment

Higher MG-ADL scores were associated with an increased risk of exacerbation; the observed relationship between MG-ADL scores and MG exacerbations highlights the importance of using MG-ADL to assess outcome measures

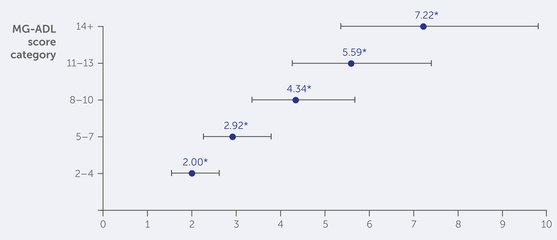
This study identified that there are patients with uncontrolled MG in the USA, resulting in exacerbations and increased levels of HCRU; personalized approaches, especially for socio-demographic groups at risk of increased disease severity, are necessary to alleviate the overall burden of MG

Figure 1 Heat map of MG treatment, HCRU, and MG exacerbations at enrollment by MG-ADL score group



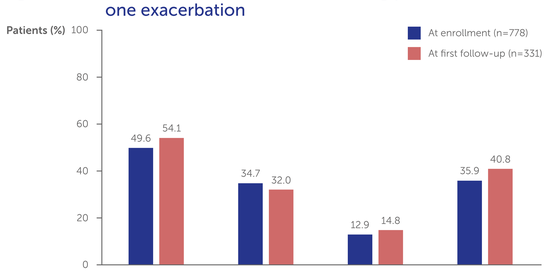
*Applicable for prednisone, immunosuppressive and pyridostigmine treatments. †p<0.001 and ‡p=0.017 for differences between MG-ADL score group.
% denotes the percentage of patients in the respective MG-ADL score group.

Figure 2 Adjusted risk of exacerbation by MG-ADL score at enrollment



*p<0.001 compared with the MG-ADL 0–1 score category. Adjusted negative binomial regression with MG-ADL score as a categorical variable; the unadjusted IRRs also demonstrated significance (p<0.001) across all MG-ADL score categories when compared with the MG-ADL 0–1 score category (data not shown).

Figure 3 Exacerbation-related HCRU among patients with one exacerbation



Abbreviations: ATD, autoimmune thyroid disease; CI, confidence interval; COPD, chronic obstructive pulmonary disease; HCRU, healthcare resource utilization; IBD, inflammatory bowel disease; ICU, intensive care unit; IRR, incidence rate ratio; IVIg, intravenous immunoglobulin; MG, myasthenia gravis; MG-ADL, Myasthenia Gravis Activities of Daily Living; MGFA, Myasthenia Gravis Foundation of America; MGFAPR, Myasthenia Gravis Foundation of America Global Myasthenia Gravis Patient Registry; NSISTs, non-steroidal immunosuppressants; PLEX, plasma exchange; SCIg, subcutaneous immunoglobulin; SD, standard deviation; SLE, systemic lupus erythematosus.

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