Factors Associated with Myasthenia Gravis – Activities of Daily Living Score in a Real-World, United States Population Prescribed Standard of Care Therapy

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Objective

To explore the factors associated with increased MG-ADL score in a population treated with SOC therapies.

Key Takeaways

- **Amongst those treated with SOC, patients with MG-ADL scores ≥5 experienced significantly more lines of therapy and higher acute treatment usage, compared to those with an MG-ADL score ≤4.**
- **Patients with MG-ADL scores of ≥5 experienced more gMG-related clinical events and hospitalizations compared with patients scoring ≤4.**
- **These findings suggest there exists a high proportion of MG patients for whom advanced therapies should be considered to reduce symptoms and clinical events.**

Background

Generalized myasthenia gravis (gMG) is a subtype of autoimmune myasthenia gravis, a rare chronic condition characterized by muscle weakness and fatigue[1].

There is no cure for MG and despite the availability of novel treatments, many patients are still managed with traditional standard of care (SOC) therapies, including acetylcholinesterase inhibitors, steroidal and non-steroidal immunosuppressants and immunoglobulins, despite ongoing symptoms. The side effects of immunosuppressive agents make them intolerable for some gMG patients[2].

The Myasthenia Gravis Activities of Daily Living (MG-ADL) is a measure of functional status and is often used as an outcome tool to measure treatment response in clinical trials and observational studies and may be included in the restrictions for use of certain novel therapies[3].

Methods

- Data were drawn from the Adelphi gMG Disease Specific Programme™ (DSP)[4-7], a cross-sectional dataset of US-based neurologists reporting on their consulting gMG patients collected between January-August 2024.
- The MG-ADL was used to measure symptom severity and the impact of gMG on patients' activities of daily living (ADLs) and reported by physicians after a recent consultation with the patient[8]. Descriptive statistics were reported alongside bivariate comparisons of patients grouped by MG-ADL scores ≤4 and ≥5. Physicians also reported on their gMG patient's demographics, clinical characteristics, current treatment, myasthenic crises and exacerbations and hospitalisations.
- Physicians were eligible for inclusion if they were responsible for the management of at least one patient with a confirmed diagnosis of gMG and identified as having a primary specialty of neurology. For this analysis, physicians reported on patients with a confirmed diagnosis of gMG defined as a Myasthenia Gravis Foundation of America (MGFA) class II-IV and prescribed a SOC therapy at time of survey.

Results

- ***Forty-one physicians reported on 148 gMG patients prescribed SOC at time of survey.***
- ***Overall mean (standard deviation; SD) age was 55.3 (14.6), 50.7% were male, and 33.1% had MG-ADL scores ≥5 (Table 1).***
- ***There was a significantly higher mean (SD) number of treatment lines prescribed to patients with MG-ADL scores ≥5 (1.8 [1.0]) versus ≤4 (1.5 [0.7]; p<0.05) since diagnosis.***
- ***A higher proportion of patients with MG-ADL scores ≥5 were prescribed ≥3 treatment lines compared to those with MG-ADL scores of ≤4 (24.5% versus 5.1%, respectively [p<0.05]).***

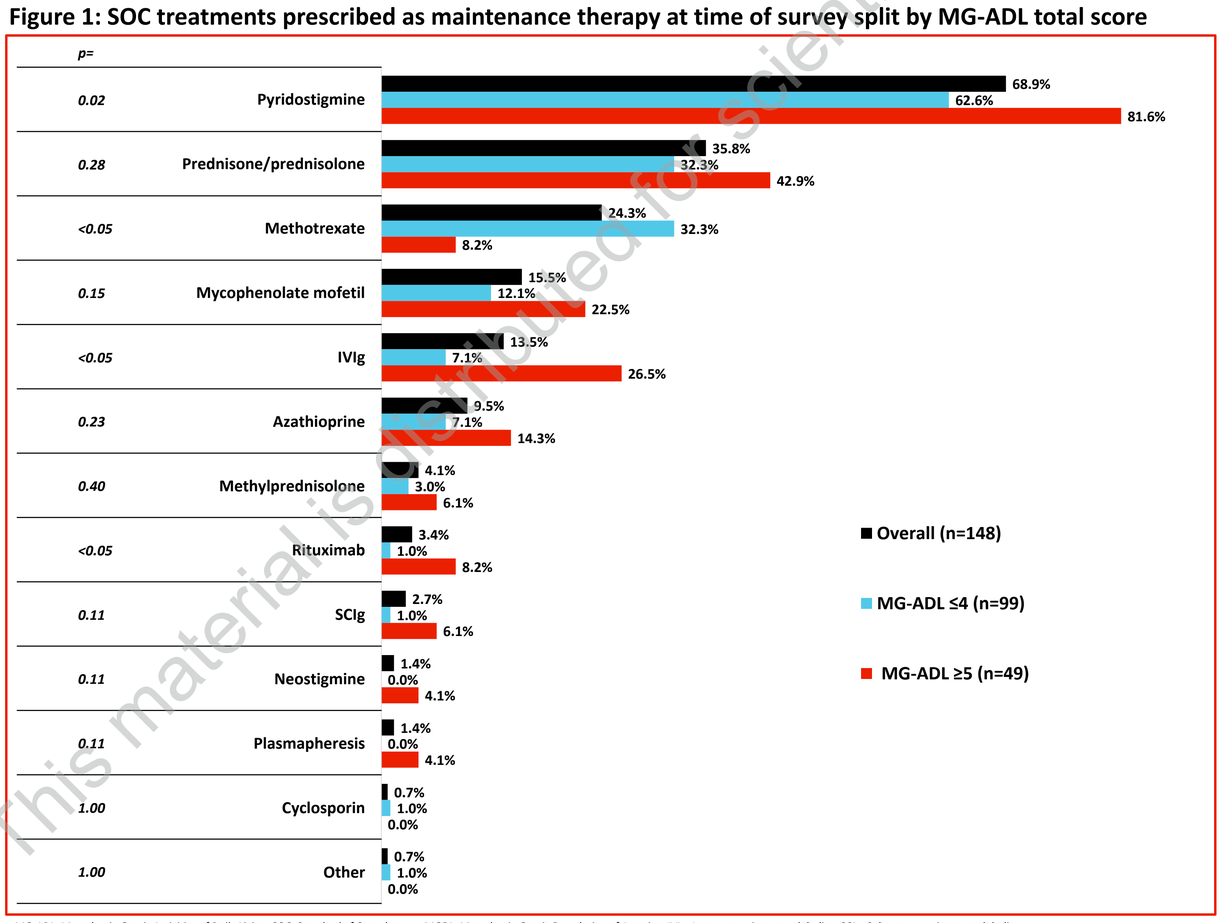
Table 1. Demographics at time of survey of patients with gMG

	Overall (N=148)	MG-ADL ≤4 (N = 99)	MG-ADL ≥5 (N = 49)	*p-value*
Demographics				
Age; years, mean (SD)	55.3 (14.6)	54.4 (14.0)	57.1 (15.7)	0.30
Male, n (%)	75 (50.7)	53 (53.5)	22 (44.9)	0.38
Time since diagnosis, n	132	93	39	
Time since diagnosis, years; mean (SD)	4.2 (5.3)	4.0 (4.2)	4.8 (7.2)	0.38
Current MGFA Class				
Class II, n (%)	134 (90.5)	92 (92.9)	42 (85.7)	
Class III, n (%)	12 (8.2)	7 (7.1)	5 (10.2)	<0.05
Class IV, n (%)	2 (1.4)	0 (0.0)	2 (4.1)	
Number of Treatment Lines, mean (SD)	1.6 (0.8)	1.5 (0.7)	1.8 (1.0)	
Number of Treatment Lines; 1, n (%)	85 (57.4)	61 (61.6)	24 (49.0)	
Number of Treatment Lines; 2, n (%)	46 (31.1)	33 (33.3)	13 (26.5)	<0.05
Number of Treatment Lines; ≥3, n (%)	17 (11.5)	5 (5.1)	12 (24.5)	

MG-ADL, Myasthenia Gravis Activities of Daily Living; MGFA, Myasthenia Gravis Foundation of America; gMG, generalized Myasthenia Gravis

- ***Physicians reported 81.6% of patients with MG-ADL scores of ≥5 were prescribed Pyridostigmine, compared to 62.6% for those with MG-ADL scores of ≤4 (Figure 1).***
- ***There was a significantly higher prescription of IVIg, rituximab and Methotrexate in patients with MG-ADL scores of ≥5 compared to patients with MG-ADL scores of ≤4 (Figure 1).***

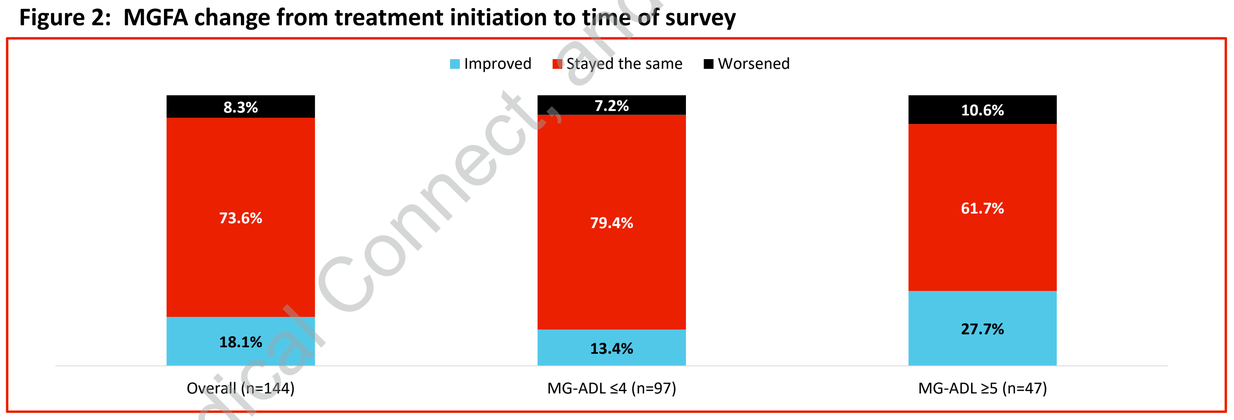
Figure 1: SOC treatments prescribed as maintenance therapy at time of survey split by MG-ADL total score



MG-ADL, Myasthenia Gravis Activities of Daily Living; SOC, Standard of Care therapy; MGFA, Myasthenia Gravis Foundation of America; IVIg, Intravenous immunoglobulins; SCIg, Subcutaneous immunoglobulins.

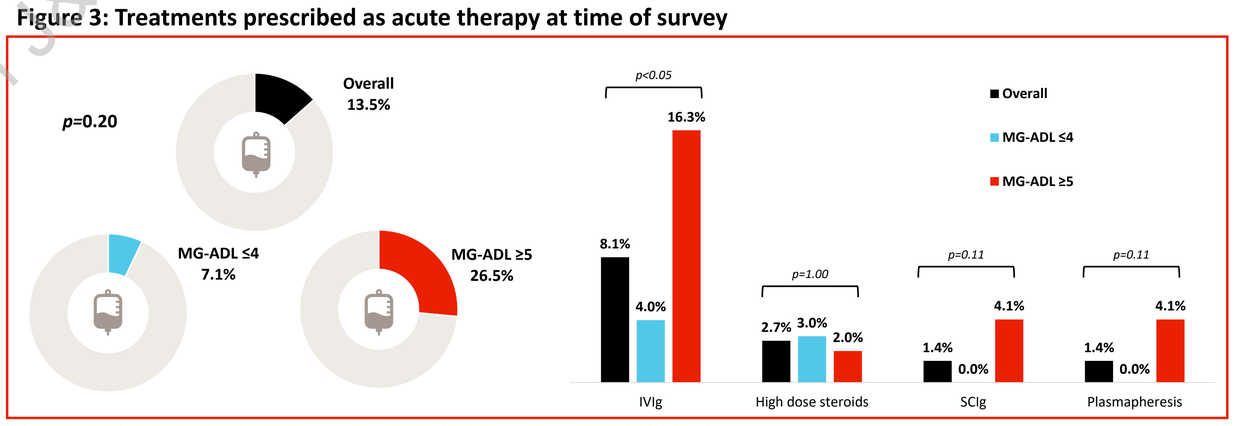
- ***Physicians reported that 73.6% of patients' MGFA class had stayed the same since the initiation of their current treatment (Figure 2).***
- ***There was a higher proportion of patients with an MG-ADL score of ≥5 whose MGFA class had worsened since treatment initiation (10.6%), compared to those with MG-ADL scores of ≤4 (7.2% [p=0.20]) (Figure 2).***
- ***Physicians reported 69.4% of patients with MG-ADL scores ≥5 were currently experiencing shortness of breath versus 27.3% of those with MG-ADL scores ≤4 (p<0.05).***
- ***There was a significantly higher percentage of patients with MG-ADL scores ≥5 experiencing difficulty chewing and difficulty swallowing (71.4% and 81.6%, respectively) compared to those with MG-ADL scores ≤4 (p<0.05).***

Figure 2: MGFA change from treatment initiation to time of survey



- ***Acute treatments were prescribed alongside SOC in 13.5% of patients overall and more frequently to patients with MG-ADL ≥5 (26.5%) vs MG-ADL ≤4 (7.1%, p<0.05).***
- ***IVIg was most frequently prescribed acute treatment (overall 8.1%, MG-ADL ≥5; 16.3% vs MG-ADL ≤4; 4.0%, p<0.05, Figure 3).***

Figure 3: Treatments prescribed as acute therapy at time of survey



MG-ADL, Myasthenia Gravis Activities of Daily Living; IVIg, Intravenous immunoglobulins; SCIg, Subcutaneous immunoglobulins. P=0.20 in Figure 2.

- ***Since diagnosis, 32.6% of the overall sample, 53.2% of MG-ADL ≥5 patients and 22% of MG-ADL ≤4 patients had experienced MG crisis or exacerbation (p<0.05, Figure 4).***
- ***In the 12 months prior to the survey, 50% of patients with MG-ADL scores ≥5 experienced at least one MG crisis vs 20% of those with MG-ADL scores ≤4 (p<0.05, Figure 5).***
- ***In the same time period, 17.8% of patients with MG-ADL ≥5 were hospitalized at least once vs 12.3% of patients MG-ADL ≤4 (p<0.05, Figure 5).***

Figure 4: Since diagnosis, has the patient ever had either a myasthenic crisis or exacerbation of symptoms

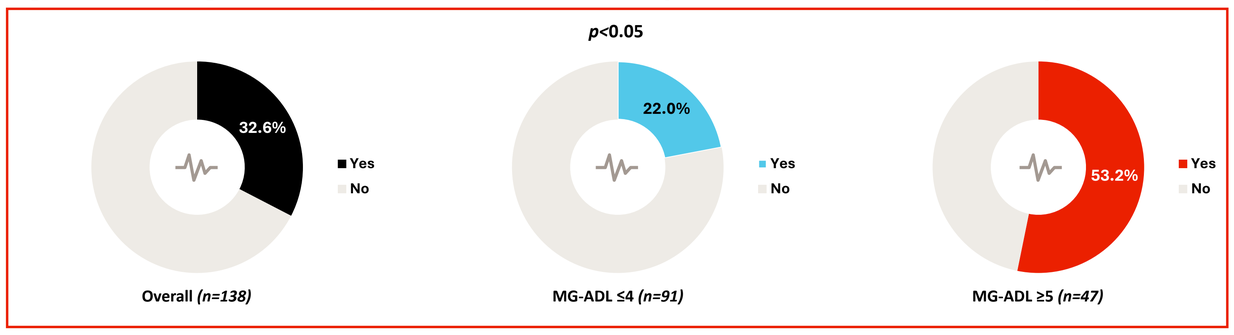
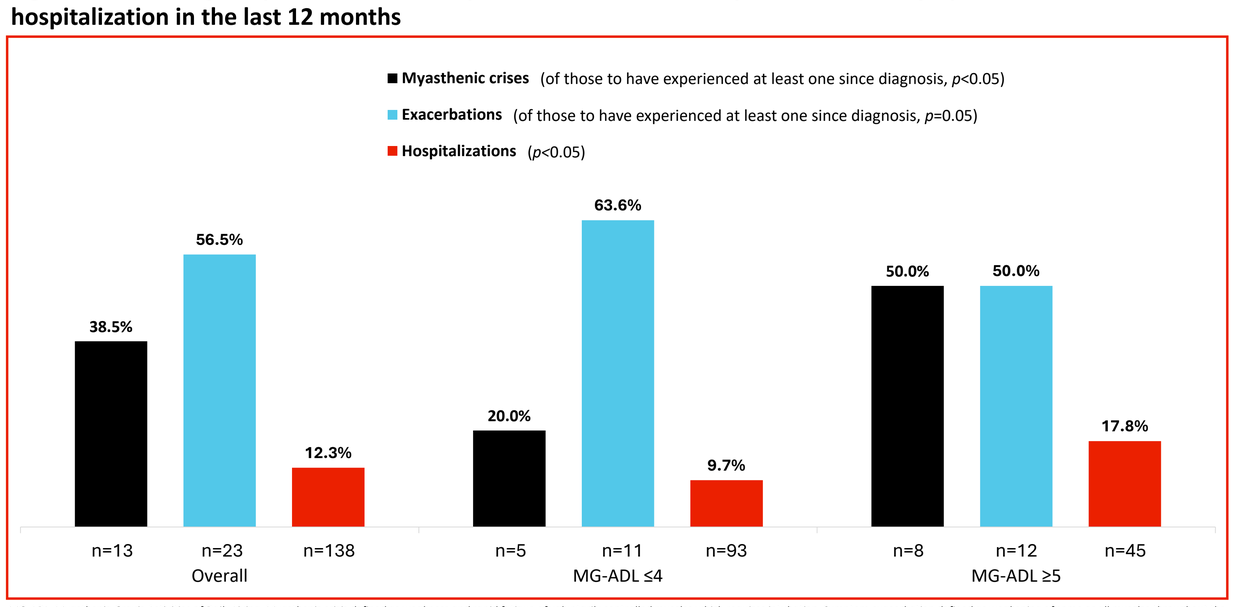


Figure 5: Percentage of patients to have experienced at least one myasthenic crisis, symptom exacerbations or hospitalization in the last 12 months



MG-ADL, Myasthenia Gravis Activities of Daily Living. Myasthenic crisis defined as weakness and rapid fatigue of voluntarily controlled muscles which requires intubation. Symptom exacerbation defined as weakening of some or all muscles throughout the body but assistance is not required for breathing for the purpose of this study.

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- ***REFERENCES:** 1. Phillips WD, Vincent A, F1000Res. 2016;5:F1000 3. Menon D, et al. Front Neurol. 2020;11:538. 4. Muppidi S, et al. Muscle Nerve. 2022;65(6):630-639. 5.Anderson P, et al. Current Med Res Opin. 2008;24:3063-3072. 6. Babineaux SM, et al. BMJ Open. 2016;6(8):e010352. 7. Higgins V, et al. Diabetes Metab Syndr Obes. 2016;9:371-380. 8. Anderson P, et al. Curr Med Res Opin. 2023;39:1707-1715. 9. Wolfe G.I, et al. Neurology. 1999;52:1487–1489*
- ***LIMITATIONS:** The DSP was not based on a true random sample of physicians or patients. While minimal inclusion criteria governed the selection of the participating physicians and patients, participation was influenced by willingness to complete the survey. The quality of the data obtained relies on how accurately physicians and patients were able to recall and report information, and therefore there is a potential for information bias and recall bias.*
- ***DISCLOSURES:** JP, LJ, AK and NC are employees of Johnson & Johnson. GG, HC, SLB and BP are employees of Adelphi Real World. RG is on the advisory board for Argenx, UCB and Janssen and serves as speaker for Argenx and Alexion.*
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