



Letter to the Editor

Response to “Letter to the editors in regard to the article ‘Predictive score for oral corticosteroid-induced initial worsening of seropositive generalized myasthenia gravis’”


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Dear Editor:

We would like to thank Dr. Pike-Lee for the inquiry regarding our study [1]. We would like to explain the factors included in our score, especially early-onset myasthenia gravis (EOMG) or thymoma-associated myasthenia gravis (TAMG) and the incorrect number of patients.

We reported that risk factors significantly associated with the occurrence of initial worsening were non-late-onset myasthenia gravis (LOMG), TAMG, upper limb weakness, initial prednisolone dose, and white blood cell count and potassium concentration after prednisolone administration; these factors were identified using univariate logistic regression analysis and Fisher's exact test [1]. Subsequently, stepwise multi-logistic regression revealed only three independent risk factors of initial worsening: non-LOMG, upper limb weakness, and initial prednisolone dose. Moreover, myasthenia gravis can be categorized into three subgroups (EOMG, LOMG, and TAMG) on the basis of onset age and thymus tissue [2]; therefore, we described non-LOMG as EOMG or TAMG in our study.

As Dr. Pike-Lee noted, the number of patients with initial worsening was 50 (42 patients in class II, 6 patients in class III, and 2 patients in

class IV) in the Myasthenia Gravis Foundation of America (MGFA) classification (original Table 1) [1]. We erroneously missed excluding four myasthenia gravis patients without acetylcholine receptor antibody in MGFA class II. In fact, the number of patients with MGFA class II was 38, and the *p* value relating to MGFA was 0.511. Accordingly, we have corrected Table 1 and made.

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Declaration of Competing Interest

None.

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Table 1
Comparison of patients' characteristics with or without initial worsening.

	Total (n = 62)	Initial worsening group (n = 16)	Non-initial worsening group (n = 46)	P value
Age at the time of therapy, years	59.4 ± 16.8	53.3 ± 15.5	61.5 ± 16.9	0.096
Age at onset, years	58.4 ± 16.9	52.3 ± 15.6	60.5 ± 16.8	0.097
Sex (male/female)	26/36	6/10	20/26	0.759
Disease duration, months	4.1 (2.5–12.4)	5.1 (1.7–12.8)	3.9 (3.0–12.8)	0.897
E-L-T classification				0.002
Early-onset MG, n (%)	12 (19%)	4 (25%)	8 (17%)	0.487
Late-onset MG, n (%)	33 (53%)	3 (19%)	30 (65%)	0.008
Thymoma-associated MG, n (%)	17 (27%)	9 (56%)	8 (17%)	0.018
Thymoma histopathology				0.217
Microscopic thymoma				
AB	2 (12%)	1 (11%)	1 (13%)	
B1	2 (12%)	2 (22%)	0 (0%)	
B2	9 (53%)	5 (56%)	4 (50%)	
B3	2 (12%)	1 (11%)	1 (13%)	
MGFA, n (%)				0.511
II	49 (79%)	11 (69%)	38 (83%)	
III	10 (16%)	4 (25.0%)	6 (13%)	
IV	3 (5%)	1 (6.2%)	2 (4%)	
AChR titer, nmol/L (median [IQR]), n = 62	40 (11–80)	41 (22–118)	39 (11–69)	0.567
White blood cell counts before prednisolone, /mm ³ (mean ± SD)	5798 ± 1391	5518 ± 1031	5895 ± 1494	0.339
White blood cell counts after prednisolone /mm ³ (mean ± SD)	7414 ± 3201	6243 ± 1619	7821 ± 3515	0.045
Potassium concentration before prednisolone, mEq/L (mean ± SD)	4.1 ± 0.4	4.0 ± 0.2	4.1 ± 0.4	0.260
Potassium concentration after prednisolone, mEq/L (mean ± SD)	3.8 ± 0.4	3.6 ± 0.2	3.9 ± 0.4	0.045
Quantitative MG Score before treatment, points (mean ± SD)	13.2 ± 5.8	14.6 ± 7.2	12.8 ± 5.3	0.336
Ocular symptoms, n (%)	60 (97%)	16 (100%)	44 (96%)	1.000
Facial palsy, n (%)	37 (60%)	7 (44%)	30 (65%)	0.150
Bulbar palsy, n (%)	18 (29%)	5 (31%)	13 (28%)	1.000
Dyspnea, n (%)	20 (32%)	6 (38%)	14 (30%)	0.752
Neck weakness, n (%)	53 (85%)	13 (81%)	40 (87%)	0.683
Upper limb weakness, n (%)	33 (53%)	12 (75%)	21 (46%)	0.039
Lower limb weakness, n (%)	38 (61%)	10 (63%)	28 (61%)	1.000
Initial prednisolone dose, mg/day	17.6 ± 13.3	24.1 ± 17.1	15.3 ± 11.0	0.029
Immunosuppressant, n (%)	7 (11%)	0 (0.0%)	7 (15%)	0.175
Plasmapheresis, n (%)	14 (23%)	7 (44%)	7 (16%)	0.035
Intravenous immunoglobulin, n (%)	5 (8%)	1 (6%)	4 (9%)	1.000

AChR: acetylcholine receptor, IQR: interquartile range, MG: myasthenia gravis, MGFA: Myasthenia Gravis Foundation of America, MuSK: muscle-specific tyrosine kinase, SD: standard deviation.

References

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- [2] H. Murai, M. Masuda, K. Utsugisawa, et al., Clinical features and treatment status of adult myasthenia gravis in Japan, *Clin Exp Neuroimmunol.* 5 (2014) 84–91.

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