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Quality of life and activities of daily living of myasthenia gravis patients in Hospital Seberang Jaya, Malaysia using MGQOL-15 and MGADL scores: A cross sectional study

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Abstract

Background & Objective: Myasthenia gravis (MG) is an immune mediated neuromuscular disease with a wide range of clinical symptoms causing fatigability, which can influence quality of life (QoL) and activity of daily living (ADL). Patient-derived quality of life, as assessed by questionnaires has proven to be an essential outcome measures when used in conjunction with clinical examination. The objective of this study is to explore factors associated with QoL and ADL of MG patients in Hospital Seberang Jaya, Malaysia. *Methods:* In this cross-sectional study 35 MG patients were recruited from the Neurology clinic of Hospital Seberang Jaya, Penang. An interview-administered questionnaire was utilized. The questionnaire consisted of 3 parts: Socio-demographics (age, gender, ethnicity), Clinical factors (MG type, MG crisis, Ach Antibody positivity, thymoma was recorded) and QoL assessment using validated MG-QoL-15 and MGADL questionnaires respectively. Descriptive and bivariate analysis was conducted using SPSS version 23. *Results:* Our sample were 57% males, and the mean age was 54.34 years, range 19 to 81 years. Higher score of MG-QoL-15 and MGADL indicates lower QoL of MG patients. In bivariate analysis, Non-Malays were 8 times more likely to have better QoL compared to Malays (OR=8, 95%CI =1.726-37.09, p = 0.005). Patients with poorer ADL score were 6.5 times more likely to have lower QOL compared to those with lower ADL score (OR = 6.5, 95% CI = 1.3-31.8, p = 0.016). Other clinical factors were not significantly associated with QoL.
Conclusion: This study showed that the non-Malay MG patients and those with lower grades of the disease (grade I to II) had better QoL.

Keywords: Myasthenia gravis, quality of life, myasthenia gravis-quality of life 15, myasthenia gravis activities of daily living, validation in Malay.

INTRODUCTION

Myasthenia gravis (MG) is an acquired autoimmune disease, the main feature of which is a production of IgG-class antibodies against postsynaptic nicotinic acetylcholine receptors (AChR). Neuromuscular transmission deteriorates due to the reduction of functional AChRs. MG is one of the relatively rare diseases of nervous system; there are 2-11 new cases of MG per 1 million population per year.[1-3]

In Malaysia, the prevalence of MG has not been well known until 30 years ago after a clinical survey was performed which reported a total of 62 cases seen from 1968 to 1979 at the University Hospital in Kuala Lumpur, giving an average of 5.16 new cases every year.[4] A study in 2010 have suggested that different ethnic groups may have different clinical presentations.[5]

Quality of life (QoL) is a measure of general well-being of individuals and societies, outlining negative and positive features of life. Its quantification enables assessment of impact of various diseases or therapeutic interventions from the patients' perspective. The World Health Organization (WHO) defined QoL as individuals' perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations,

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standards, and concerns. It is a broad-ranging concept affected in a complex way by the person's physical health, psychological state, level of independence, social relationships, and his/her relationship to salient features of the environment.[6] The core components include physical, psychological, emotional, social, and occupational domains.

Activities of daily living (ADLs), often termed physical ADLs or basic ADLs, include the fundamental skills typically needed to manage basic physical needs, comprised the following areas: grooming/personal hygiene, dressing, toileting/continence, transferring/ambulating, and eating. Basic ADLs are generally categorized separately from Instrumental Activities of Daily Living (IADLs), which include more complex activities related to independent living in the community (e.g., managing finances and medications). IADL performance is sensitive to early cognitive decline, whereas physical functioning is often a significant driver of basic ADL ability.[7]

Patient-derived quality of life, as assessed by questionnaires has proven to be an essential outcome measures when used in conjunction with clinical examination.[8] MG disease status can be established with Myasthenia Gravis Quality of Life - 15 (MG-QoL-15) and Myasthenia Gravis Activities of Daily Living (MGADL) questionnaires to measure patients' perception of MG-related dysfunction.[9]

The aim of this study was to assess the quality of life and activity of daily living in a sample of patients suffering from MG but with a stable clinical course while on regular treatment in Neurology clinic, Hospital Seberang Jaya using the validated Malay version MG-QoL-15 scale and MG-ADL scale. The effect of age, gender, and disease severity on MG-QoL-15 was analyzed.

METHODS

This hospital-based study was carried out in Hospital Seberang Jaya (HSJ), which is a tertiary care referral center for neurological cases in Seberang Perai district, Penang. HSJ is the cluster lead hospital in Seberang Perai district among the four hospitals in this district. The other non-lead cluster hospitals are Hospital Bukit Mertajam, Hospital Sungai Bakap and Hospital Kepala Batas. HSJ has 393 bed occupancy capacity and 2 intensive care units accommodating both adults and pediatrics services.

Data collection

This cross-sectional, hospital-based study was conducted in the Neurology Clinic, HSJ. Patients with MG were recruited between April 2017 and June 2017. Myasthenia gravis was diagnosed by the neurologist based on the clinical presentation, presence of fluctuating weakness of skeletal muscles and unequivocal improvement in muscle strength following administration of intramuscular neostigmine and positive result of acetylcholine receptor antibody. The clinical status was assessed using the Myasthenia Gravis Foundation of America (MGFA) task force guidelines[10] Genkin and Osserman classification.[11] Only those patients who were stable on regular treatment were recruited after obtaining a written informed consent. The study was registered with National Medical Research Registry (NMRR) of Malaysia (NMRR-17-110-33970).

All patients were evaluated by personal face-to-face interview by study investigators in the clinic. Consent is obtained from the patients. A data collection form consisting of demographics details of patient, clinical presentation at onset, diagnostic history and investigation was used. The demographic variables included age, gender, ethnicity and employment status, whereas clinical presentation domains consisted of year of onset, clinical symptoms at onset, current Osserman grade, history of thymoma or thymectomy, family history, comorbidity and type of MG. Diagnostic investigations such as MRI brain and acetylcholine receptor antibody positivity was documented. MRI brain was performed to rule out mass lesions compressing the cranial nerves in ocular MG.[16]

Quality of Life (QoL) of MG patients was assessed by using the validated Malay and English version MG-QoL-15 scale.[12] This is a self-administered disease-specific questionnaire consisting of 15 items. Response to each item was scored on a scale of "0," "1," "2," "3," and "4" representing "not at all," "a little bit," "somewhat," "quite a bit," and "very much," in.[12] The total score is 60 representing the QoL of the MG patients. There are four dimensions in this questionnaire: mobility (8 items), symptoms (3 items), general contentment (2 items) and emotional wellbeing (2 items). The study subjects read the questionnaire and marked the most appropriate response to each question. In case the subjects were illiterate, the items of the questionnaire were read out to them in their local language and their responses marked. Adequate time was provided to each subject to complete the questionnaire. When the total score

of MGQoL-15 is higher it indicates that the QoL of MG patient is poorer.

Activity of Daily Living (ADL) of MG patients was assessed by study investigators using the validated Malay and English version MG-ADL scale.[12] There are 8 items in this MGADL form. The items are talking, chewing, swallowing, breathing, impairment of ability to brush teeth/ comb hair, impairment of ability to rise from a chair, double vision and eye lid drooping. The above items are graded with a score of 0,1,2,3 where the score 0 is normal and continued with worsening condition as the score increases. The total score is 24. Higher score in MGADL indicates worse ADL.

Inclusion and exclusion criteria

The following inclusion criteria were: (1) Adult patients more than 18 years old diagnosed with MG attending neurology clinic follow up and mentally competent; (2) Able to give written informed consent directly or through legally acceptable representative. Exclusion criteria were: (1) Patients being admitted in hospital during study period; (2) Patients passed away during the study period; (3) Patients with significant co-existing diseases that may affect QoL, such as poorly controlled diabetes mellitus, stroke, renal failure, and asthma.

Statistical analysis

The data was analyzed using the Statistical Package for the Social Sciences (SPSS) version 23. Frequency and percentage expressed as mean ± standard deviation (SD) unless otherwise stated. Bivariate analysis One-way ANOVA was used for analysis of normally distributed variables. Kruskal-Wallis ANOVA was used for non-normally distributed data. Categorical data was analyzed using Chi-square or Fisher's exact test. A value of $P < 0.05$ was considered as statistically significant. Multivariate logistic regression analysis was used too.

RESULTS

The study comprised of 35 subjects, 57.1% were males and 42.9% were females with age ranging from 19 to 81 years (mean=54.34 years, SD=16.46 years). There were 15 cases referred from the private hospitals. Chinese patients were half of the total study population (19, 54.3%). More than half of the study population present with ocular type MG (20, 57.1%). Most of the patients are more in a stable, milder in severity of the disease. Fifteen patients (43%) were in Osserman grade I. Six patients (17.1%) had experienced myasthenic crisis at least once in their life time. Table 1 shows the clinical demographics characteristic of the study sample.

None of the patients had family history of MG. The most common symptoms seen in MG patients was ptosis presenting as the initial complaint, in 28 patients (80%). As expected, the majority of ocular MG patients (90%) presented with ptosis. This was followed by fatigability (12,34.2%), dysphagia and limb weakness (10 each, 28.57%) patients, diplopia (9,25.7%), dysarthria and unsteady gait (3each, 8.57%) patients, facial weakness and blurring of vision (2each, 5.71%) patients.

Table 2 shows the determinants of MGQoL-15 score in patients with MG. The cut off point for MG-QoL-15 is taken as 5 as it is the mean score among the study sample, whereas the cut off point for MGADL taken as 2. The worst MGQoL score of 44, was scored by an elderly man with comorbidities. The best MGADL point, 0, was scored by 10 of the patients (29%).

In bivariate analysis, non-Malay were 8 times more likely to have better QoL compared to Malay patients (OR=8, 95%CI =1.726-37.09, p = 0.005). Patients with poorer ADL score were 6.5 times more likely to have lower QoL compared to those with lower ADL score (OR = 6.5, 95% CI = 1.3-31.8, p = 0.016). Clinical factors were not significantly associated with QoL.

DISCUSSION

In this study we report the MG-QoL-15 score of 35 patients of MG with a stable course who visited the neurology clinic. In western literature it is stated that females are predominant whereas in this study males predominate.[13] Majority of the patients that were involved in this study were older patients with mean age of 54 years, up to 81 years of age along with other medical comorbidities. Hypertension and diabetes are the commonest comorbidities among these older patients. Two (5.7%) patients presented with hypothyroidism.

The current study showed that ethnicity (non-Malay) and MG ADL (≤2) has significant higher odds ratio towards the better quality of life in patients with MG. Other characteristics such as employment, duration of illness, types of MG, MG crisis, thymoma, thymectomy and AChR antibody did not show any significant contribution to the MG-QoL. These findings are comparable with the results in other studies[18] except the study

Table 1: Clinical demographics characteristics of the sample (n=35)

Variables	Values [n (%)]
Gender: Male Female	 20 (57.1%) 15 (42.9%)
Mean Age	54.34(SD:16.46years)
Disease duration (mean ± SD)	7.85 ± 7.2
Ethnicity	Malay: 14 (40.0%) Chinese: 19 (54.3%) Indian: 1 (2.9%) Others: 1 (2.9%)
Type of MG	Ocular: 20 (57.1%) Generalized: 15 (42.9%)
Current Osserman grade I/II/IIA/IIB/III/IV	 15/1/7/7/3/2
Anti AChR antibody positivity	20 (57.1%)
Thymectomized	5 (14.3%)
Myasthenic crisis	6 (17.1%)
Comorbid	Diabetes: 6 (17.1%) Hypertension: 14(40.0%) Hypothyroid: 2(5.7%) IHD: 3(8.6%) CKD: 2(5.7%)

IHD: Ischemic heart disease, CKD: Chronic kidney disease

from Brazil[19] which showed a significant reduced QoL in those who have had MG crisis before.

This study showed that ethnicity was significantly associated with QoL of MG patients. In particular, the non-Malays have better QoL compared to Malay patients. This could be related to the finance and social cultural factors of the Malay group or compliance of medications. However, lack of finance, social cultural information and medication compliance information were not captured in this study. The study results also offered an explanation which is the correlation of poor ADL with worse QoL and our patients have poorer ADL. Only one-fourth of the study patients (25%) were employed with 3 of them are having a lower quality of life with MG-QoL-15 score of more than 5. Three-fourths of the study population were not employed in view of their age and ADL dependency.

In MG-QoL-15 questionnaire, the symptoms that were most significantly abnormal in a validation study were related to psychological, social and occupational domains like "frustrated due to MG", "limits hobbies and fun activities", 'limits social activity" and "have to make plans around MG".[9] These issues need to be examined during routine clinical review. In this MG-QoL-15 score among the study patients, we noted that first question "frustrated due to MG" and seventh question "have to makes plans around my MG" were the most affected domains among the MG patients. Eighteen of responded to these domains each and mostly in the scale of '1' meaning they were slightly depressed and overwhelmed by the disease. This shows that MG patients have effective coping skills and psychological adjustment among our patients as observed by Paul *et al*.[15]

Overall, this MG-QoL-15 is user friendly and a disease specific scale for Health Related QOL in MG patients. We found it is user friendly, quick and disease specific scale that measures direct manifestations.

The limitation of this study is the small sample size. As MG is a rare disease, and HSJ do not have a large referral base.

In conclusion, this study was conducted to assess Quality of Life of Myasthenia Gravis patients by using validated MG-QoL-15 and MG-ADL questionnaires. Study results showed that

Table 2: Determinants of myasthenia gravis-quality of life-15 score in patients with MG (n=35)

Characteristics	Quality of Life		OR	95% CI	p value
	≤ 5	> 5	(odds ratio)	(confidence interval)	
1. Gender:					
Female	9 (60.0%)	6 (40.0%)			0.767
Male	11 (55.0%)	9 (45.0%)	1.22	0.316-4.762	
2. Age:					
≤ 55 years	9 (64.3%)	5 (35.7%)	1.64	0.408-6.561	0.486
> 55 years	11 (52.4%)	10 (47.6%)			
3. Ethnicity:					
Non-Malay	16 (76.2%)	5 (23.8%)	8.00	1.726-37.09	0.005
Malay	4 (28.6%)	10 (71.4%)			
4. Employment:					
Yes	6 (66.7%)	3 (33.3%)			0.503
No	14 (53.8%)	12 (46.2%)	1.72	0.351-8.403	
5. Duration of illness:					
≤ 6 years	12 (60.0%)	8 (40.0%)	1.75	0.427-7.171	0.435
> 6 years	6 (46.2%)	7 (53.8%)			
6. Type of Myasthenia Gravis:					
Ocular	12 (60.0%)	8 (40.0%)	1.31	0.339-5.076	0.693
Generalized	8 (53.3%)	7 (53.8%)			
7. MGFA grade:					
Grade I/II	18 (90.0%)	12 (80.0%)			0.411
Grade III/IV	2 (10.0%)	3 (20.0%)	2.27	0.064-3.070	
8. Myasthenic Gravis Crisis:					
Yes	5 (83.3%)	1 (16.7%)			0.154
No	15 (51.7%)	14 (48.3%)	4.76	0.484-45.455	
9. Thymoma:					
Yes	5 (71.4%)	2 (28.6%)			0.729
None	15 (53.6%)	13 (46.4%)	2.15	0.358-13.158	
10. Thymectomy:					
Done	3 (60.0%)	2 (40.0%)			0.889
Not done	17 (56.7%)	13 (43.3%)	1.15	0.167-7.874	
11. AChR Antibody:					
Positive	14 (70.0%)	6 (30.0%)			0.076
Negative	6 (40.0%)	9 (60.0%)	3.5	0.856-14.286	
12. ADL:					
≤ 2	17 (70.8%)	7 (29.2%)	6.48	1.318-31.831	0.016
> 2	3 (27.3%)	8 (72.7%)			

ethnicity and ADL were significant to the QoL of MG patients.

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DISCLOSURE

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REFERENCES

1. Somnier FE, Keiding N, Paulson OB. Epidemiology of myasthenia gravis in Denmark. A longitudinal and comprehensive population survey. *Arch Neurol* 1991;48:733-9. doi: 10.1001/archneur.1991.00530190081019.
2. Beekman R, Kuks JB, Osterhuis HJGH. Myasthenia gravis: diagnosis and follow-up of 100 consective patients. *J Neurol* 1997;244:112-8. doi: 10.1007/s004150050059.
3. Robertson NP, Deans J, Comspton DAS. Myasthenia gravis:a population based epidemiological study in Cambridgeshire, England. *J Neurol Neurosurg Psychiatry* 1998;65:492-6. doi: 10.1136/jnnp.65.4.492.
4. Tan CT, Loh TG. Myasthenia gravis – a clinical survey in Malaysia. *Med J Malaysia* 1980; 35:144-9. PMID: 7266408
5. Shahrizaila N, Lee SC, Goh KJ. The clinical features of myasthenia gravis in a Malaysian population. Abstracts of ICCN 2010: 29th International Congress of Clinical Neurophysiology. *Clin Neurophysiol* 2010; 121(1): S1-S346.
6. Skevington SM, Lotfy M, O'Connell KA; WHOQOL Group. The World Health Organization's WHOQOL-BREF quality of life assessment: Psychometric properties and results of the international field trial. A report from the WHOQOL group. *Qual Life Res* 2004;13:299-310. doi:10.1023/B:QURE.0000018486.91360.00.
7. Boyle PA, Cohen RA, Paul R, Moser D, Gordon N. Cognitive and motor impairments predict functional declines in patients with vascular dementia. *Int J Geriatr Psychiatry* 2002; 17(2): 164-9. doi:10.1002/gps.539.
8. Slevin ML, Plant H, Lynch D, Drinkwater I, Gregory WM. Who should measure quality of life, the doctor or the patient? *Br J Cancer* 1988; 57(1):109-12. doi:10.1038/bjc.1988.20.
9. Burns TM, Grouse CK, Wolfe GI, Conaway MR, Sanders DB, MG Composite & MG QOL15 Study Group. The MG QOL15 for following the health related quality of life of patients with myasthenia gravis. *Muscle Nerve* 2011; 43(1):14-8. doi:10.1002/mus.21883.
10. Jaretzki A 3rd, Barohn RJ, Ernstoff RM, *et al*. Myasthenia gravis: Recommendations for clinical research standards. Task Force of the Medical Scientific Advisory Board of the Myasthenia Gravis Foundation of America. *Neurology* 2000;55:16-23. doi:10.1212/wnl.55.1.16.
11. Osserman KE. Myasthenia gravis. New York: Grune and Stratton, 1958. doi:10.1186/1750-1172-2-44
12. Mohd Thabit AA, Rosli NA, Solehan HN, *et al*. Validation study of the Malay version of the Myasthenia Gravis Quality of Life (MGQOL)15 and Myasthenia Gravis Activities of Daily Living (MGADL) questionnaires: *Neurol Asia* 2016;21(1);33-9.
13. Rastenytė D, Vaitkus A, Neverauskas R, Pauza V. Demographic-clinical profile of the patients with myasthenia gravis. *Medicina* 2002;38(6):611-6. PMID: 12474667.
14. Leonardi M, Raggi A, Antozzi C, *et al*. The relationship between health, disability and quality of life in myasthenia gravis: Results from an Italian study. *J Neurol* 2010;257:98-102. doi:10.1007/s00415-009-5279-z.
15. Paul RH, Nash JM, Cohen RA, Gilchrist JM, Goldstein JM. Quality of life and well-being of patients with myasthenia gravis. *Muscle Nerve* 2001;24:512-6. doi: 10.1002/mus.1034.
16. Jowkar AA. Myasthenia gravis workup. *Medscape* Updated: Aug 27, 2018
17. Kumar R, Nagappa M, Sinha S, Taly AB, Rao S. MG-QoL-15 scores in treated myasthenia gravis: Experience from a university hospital in India. *Neurol India* 2016;64;405-10. doi: 10.4103/0028-3886.181542.
18. Szczudlik P, Sobieszczuk E, Szyluk B, Lipowska M, Kubiszewska J, Kostera-Pruszczyk A. Determinants of quality of life in myasthenia gravis patients. Front. Neurol 2020; 11:553626. doi:10.3389/fneur.2020.553626.
19. Mourão AM, Gomez RS, Barbosa LSM, *et.al*. Determinants of quality of life in Brazilian patients with myasthenia gravis. *Clinics (*Sao Paulo) 2016; 71(7): 370-74. doi: 10.6061/clinics/2016(07)03.