Myasthenia gravis activities of daily living profile

Article abstract—The authors have developed an MG activities of daily living (ADL) profile (MG-ADL)—a simple eight-question survey of MG symptoms. In 254 consecutive encounters with established MG patients, the authors compared scores from the MG-ADL to the quantitative MG score (QMG)—a standardized, reliable scale used in clinical trials. The mean MG-ADL score was 4.89 ± 3.63. The mean QMG score was 10.80 ± 5.70. Pearson's correlation coefficient was 0.583 ($p < 0.001$). The MG-ADL is an easy-to-administer survey of MG that correlates well with the QMG and can serve as a secondary efficacy measurement in clinical trials.

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A number of grading systems have been developed to assess the degree of disease severity in MG and to monitor the response to therapy in clinical trials. Modified versions of Osserman's classification[1] are the most widely utilized, but have limitations from a clinical trial standpoint. These include vague descriptive terminology, a limited number of grades, and in some cases no category for remission. Because of these shortcomings, investigators have developed quantitative MG scoring systems (QMGs). Tindall et al.[2] used a 13-item QMG as the primary outcome measure in a double-blind, placebo-controlled trial demonstrating that cyclosporine is effective in MG. We recently determined that a modified version of Tindall's QMG has high interrater reliability in MG patients and normal control subjects.[3]

Increasing emphasis has been placed on scales that measure how neurologic disease impacts ADLs and quality of life.[4] We have developed a simple MG activities of daily living profile (MG-ADL) to assess the severity of MG symptoms. This eight-question survey is an expanded version of symptom-based test items from Tindall's scoring system and can be administered with patient instruction in less than 10 minutes. During this study we investigated the correlation between the MG-ADL and the QMG.

Methods. The MG-ADL and QMG scores were determined in 254 consecutive encounters with established MG patients. All MG patients were followed in our neuromuscular clinic and were diagnosed according to accepted clinical, electrophysiologic, and serologic standards.

A trained technician (L.H.) performed the QMG, and asked the questions and recorded the responses for the MG-ADL. The QMG consists of 13 objective items (figure 1), each scored from 0 (normal) to 3 (most severe). Total QMG scores range from 0 to 39. The MG-ADL is an eight-question survey of symptom severity, with each response graded from 0 (normal) to 3 (most severe). Two questions concern ocular, three oropharyngeal, one respiratory, and two extremity functions (figure 2). Cumulative MG-ADL scores range from 0 to 24.

The two scales were completed during the same patient encounter. Pearson's correlation coefficient was used to assess statistically the relationship between the two grading systems.

Results. A total of 156 MG patients (90 women, 66 men) were evaluated. Of the 254 consecutive encounters, 98 were repeat evaluations. The mean MG-ADL score was 4.89 (SD, 3.63; range, 0 to 18). The mean QMG score was 10.80 (SD, 5.70; range, 0 to 27). Pearson's correlation coefficient was 0.583 ($p < 0.001$). The 95% CI was 0.507 to 0.650.

Grade	0	1	2	3	Score
Double vision (lateral gaze) sec.	>60	11-60	1-10	Spontaneous	
Ptosis (upward gaze) sec.	>60	11-60	1-10	Spontaneous	
Facial Muscles	Normal lid closure	Complete, weak, some resistance	Complete, without resistance	Incomplete	
Swallowing	Normal	Occasional choking	Consistent choking	Cannot swallow	
Head, lifted (45° supine) sec.	>120	>30-120	>0-30	0	
Right arm outstretched (90° standing) sec.	>240	>90-240	>10-90	0-10	
Left arm outstretched (90° standing) sec.	>240	>90-240	>10-90	0-10	
Speech following counting aloud from 1-50 (onset of dysarthria)	None at #50	Dysarthria at #30-49	Dysarthria at #10-29	Dysarthria at #9	
Right leg outstretched (45° supine) sec.	>100	31-100	1-30	0	
Left leg outstretched (45° supine) sec.	>100	31-100	1-30	0	
Vital capacity (l): male	>3.5	>2.5-3.5	1.5-2.5	<1.5	
female	>2.5	>1.8-2.5	1.2-1.8	<1.2	
Rt hand grip: male	>45	>15-45	5-15	<5	
(Kg force) female	>31	>10-30	5-10	<5	
Left hand grip: male	>35	>15-35	5-15	<5	
(Kg force) female	>25	>10-25	5-10	<5	

Total score _____

Figure 1. Quantitative MG Scale.

Discussion. A number of grading systems have been developed to quantify the severity of MG symptoms and signs in the routine and experimental setting. Besinger et al.[5] introduced a quantitative MG scoring system consisting of eight items that assessed arm, leg, grip, and neck strength; vital capacity; and oropharyngeal function. The coefficient of variation between different examiners was less than 10%. Tindall et al.[2] expanded the scoring system to 13 items by adding scores for ptosis and diplopia and assessing limb strength bilaterally.

We modified Tindall's scoring system by removing chewing and swallowing assessments that were subjective and symptom based, replacing them with objective measures of dysarthria and dysphagia. The 13-item QMG has a high interrater reliability, with scores not differing from observed values by more than ±2.63 points at the 95% CI.[3] For placebo-controlled studies, this translates to a sample size of 17 patients per treatment arm to detect a difference using the QMG as the primary outcome variable at a power of 0.80. It takes approximately 30 minutes to complete the QMG. Technician training, a hand-held dynamometer, and a stopwatch are required.

Scales that measure functional status in neurologic disease are of increasing importance as more emphasis is placed on the impact of illness on everyday activities. Recently, the 136-item Sickness Impact Profile was administered to 524 ALS patients and was found to correlate closely with objective strength measurements.[4] Busch et al.[6] adapted a quality-of-life survey used in cancer studies to assess outcome in MG patients following thymectomy. Szobor[7] adapted Kurtzke's MS disability status scale for clinical research in MG. However, these scales have shortcomings in MG, including limited relevance, lengthy questionnaires, and complex classification schemes. We designed the MG-ADL as a simple, easy-to-administer profile of functional status pertinent to MG. In our study, the eight-question MG-ADL demonstrated a high correlation with the QMG. Unlike the QMG, the MG-ADL requires no special

Grade	0	1	2	3	Score
Talking	Normal	Intermittent slurring or nasal speech	Constant slurring or nasal, but can be understood	Difficult to understand speech	
Chewing	Normal	Fatigue with solid food	Fatigue with soft food	Gastric tube	
Swallowing	Normal	Rare episode of choking	Frequent choking necessitating changes in diet	Gastric tube	
Breathing	Normal	Shortness of breath with exertion	Shortness of breath at rest	Ventilator dependence	
Impairment of ability to brush teeth or comb hair	None	Extra effort, but no rest periods needed	Rest periods needed	Cannot do one of these functions	
Impairment of ability to arise from a chair	None	Mild, sometimes uses arms	Moderate, always uses arms	Severe, requires assistance	
Double vision	None	Occurs, but not daily	Daily, but not constant	Constant	
Eyelid droop	None	Occurs, but not daily	Daily, but not constant	Constant	

Total score _____

Figure 2. MG Activities of Daily Living Scale.

training or equipment and can be performed readily in most clinical settings. The MG-ADL can serve as a useful measure of MG symptom severity in routine practice and as a secondary outcome variable in clinical trials.

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