

MYASTHENIA GRAVIS COMPOSITE SCALE

Introduction

The Myasthenia Gravis Composite scale (MGC) was designed to incorporate both patient-reported and examiner-determined items that could be administered within a relatively short time, about 5 minutes. All items are adopted from other validated scales.

The patient reported items (talking, chewing, swallowing, breathing) are derived from the Myasthenia Gravis Activities of Daily Living (MG-ADL) profile without modification. Although originally designed as an instrument to be administered by a trained examiner, growing consensus amongst experts is that the MG-ADL Score, as a patient reported outcome measure (PROM), could be self-reported by the patient after proper instruction (see separate instructions for administration of the MG-ADL).

The examiner-determined items (ptosis, double vision, eye closure, neck flexion/extension, shoulder abduction, hip flexion) are derived from the Quantitative Myasthenia Gravis (QMG) and Myasthenia Gravis Manual Muscle Testing (MG-MMT) instruments with some modifications. Maximum time for onset of diplopia and ptosis are reduced from >60 seconds in the QMG to 45 seconds in the MGC for both symptoms; this was done after construct validity testing showed that <1% of ocular score times occurred between 46 and 60 seconds.¹ Eye closure weakness was specifically defined (normal; can be forced open with effort; can be forced open easily; unable to keep eyes closed) instead of the MG-MMT use of four categories. Similarly, neck flexion, shoulder abduction and hip flexion muscle strength score was converted to 3 categories of weakness [mild; moderate (50% +/- 15%); severe] instead of quartiles from the MG-MMT.

Unlike the above scales, the MGC uses a weighted scoring system so that item scores do not all carry equal weight. Despite the weighting, the score can be summated to estimate an overall disease severity score. The maximum score is 50, and a 3-point change in the summated score is considered clinically meaningful.¹

General procedures for administration

- **Patient reported items:**
 - In most instances, subjects will have completed the MG-ADL profile prior to the MGC and item scoring can directly be imported from this into the MGC. In instances where the

MG-ADL has not been performed, the identical instructions from the MG-ADL should be used to inform subjects on how to score these items.

- Refer to the MG-ADL instructions for details on administration of the patient reported items of the MGC.

- **Examiner determined items:**

- Pyridostigmine needs to be held for at least 12 hours prior to these assessments, whereas long-acting (extended release) formulations should be held for at least 24 hours. The time and amount of the last dose taken should be recorded.
- If the QMG assessment has already been performed, item scores for ptosis and diplopia can be calculated from the QMG, provided exact times for diplopia and ptosis onset are recorded. Importantly, item score cut-offs are not identical between the QMG and MGC (Table).

Table. Duration of assessment for QMG and MGC

	QMG	MGC
Ptosis	0-61s	0-45s
Diplopia	0-61s	0-45s

- If the QMG has not been performed, please refer to the instructions for testing ptosis and diplopia in the QMG manual. The instructions for performing these assessments in the MGC and QMG are the same, only the scoring is different. Make sure to record the times to event.
- Instructions for performing eye closure, neck flexion/extension, shoulder abduction, and hip flexion as part of the MGC are included in the detailed instructions that follow.

ITEM-SPECIFIC MYASTHENIA GRAVIS COMPOSITE SCALE INSTRUCTIONS

1. Ptosis, upward gaze

Transfer **time** from QMG scale, and score based on MGC cut-offs.

If QMG not performed, or time not documented, then refer to the separate QMG instructions.

2. Double vision on lateral gaze, left or right

Transfer **time** from QMG scale, and score based on MGC cut-offs.

If QMG not performed, or time not documented, then refer to the separate QMG instructions.

3. Eye closure

Patient positioning and examiner instructions for the MGC eye closure and QMG facial muscles assessments are the same.

If the QMG has been performed, assess eye closure according to the MGC specific grading (normal (0); can be forced open with effort (0); can be forced open easily (1); or unable to keep eyes closed (2)).

If the QMG has not been performed, refer to the QMG instructions for facial muscles assessment and assess eye closure according to the MGC specific grading.

4. Talking

Transfer response from MG-ADL.

If MG-ADL not performed, then refer to the separate MG-ADL instructions.

5. Chewing

Transfer response from MG-ADL.

If MG-ADL not performed, then refer to the separate MG-ADL instructions.

6. Swallowing

Transfer response from MG-ADL.

If MG-ADL not performed, then refer to the separate MG-ADL instructions.

7. Breathing

Transfer response from MG-ADL.

If MG-ADL not performed, refer to further instructions below.

8. Neck flexion or extension

The subject should be positioned supine for testing neck flexion, and prone for testing neck extension. If the subject is unable to lay in these positions, the reason(s) should be clearly documented in the case report form. You may place a pillow under the patient's knees for comfort. While you are giving the instructions and demonstration, you may keep a pillow under their head, but this should be removed prior to the start of the testing. Strength should be graded as follows, and the weaker movement used to score item:

- Mild weakness = Medical Research Council (MRC) grade 4+ to 5-
- Moderate weakness = MRC grade 4 to 4-
- Severe weakness = MRC grade 3 or less

9. Shoulder abduction

The subject should be sitting upright, feet planted on ground, and without back support. If testing cannot be performed on one side (e.g., rotator cuff injury), assume the same score as the contralateral side. Document the reason for the inability to test the side. If there is asymmetry in the weakness, the side with the worst weakness should be recorded. Grade strength as follows:

- Mild weakness = MRC grade 4+ to 5-
- Moderate weakness = MRC grade 4 to 4-
- Severe weakness = MRC grade 3 or less

10. Hip flexion

Hip flexion testing should be performed with the subject in the supine position and with the knee flexed. Document the testing position. If testing cannot be performed in the recommended position, the reason for this should be clearly documented (e.g., orthopnea). If testing cannot be performed on one side (e.g., back pain), assume the same score as the contralateral side. Document the reason for the inability to test the side. If there is asymmetry in the weakness, the side with the worst weakness should be recorded.

Grade shoulder abduction strength as follows:

- Mild weakness = MRC grade 4+ to 5-
- Moderate weakness = MRC grade 4 to 4-
- Severe weakness = MRC grade 3 or less

DRAFT – NOT FOR USE

Once all responses have been recorded, the item scores can be added to obtain the total score.

REFERENCES:

1. Burns TM, Conaway M, Sanders DB. The MG Composite: A valid and reliable outcome measure for myasthenia gravis. *Neurology*. 2010;74:1434-1440.
doi:10.1212/WNL.0b013e3181dc1b1e

MG composite scale

Ptosis, upward ease (physician examination)	>45 seconds = 0	11-45 seconds = 1	1-10 seconds = 2	Immediate = 3
Double vision on lateral Gaze, left or right (physician examination)	>45 seconds = 0	11-45 seconds = 1	1-10 seconds = 3	Immediate = 4
Eye closure (physician examination)	Normal = 0	Mild weakness (can be forced open with effort) = 0	Moderate weakness (can be forced open easily) = 1	Severe weakness (unable to keep eye closed) = 2
Talking (patient history)	Normal = 0	Intermittent slurring or nasal speech = 2	Constant slurring or nasal but can be understood = 4	Difficult to understand speech = 6
Chewing (patient history)	Normal = 0	Fatigue with solid food = 2	Fatigue with soft food = 4	Gastric tube = 6
Swallowing (patient history)	Normal = 0	Rare episode of choking or trouble swallowing = 2	Frequent trouble swallowing, for example necessitating change in diet = 5	Gastric tube = 6
Breathing (thought to be caused by MG)	Normal = 0	Shortness of breath with exertion = 2	Shortness of breath at rest = 4	Ventilator dependence = 9
Neck flexion or extension (weakest) (physician examination)	Normal = 0	Mild weakness = 1	Moderate weakness (i.e., ~50% weak, ±15%) = 3	Severe weakness = 4
Shoulder abduction (physician examination)	Normal = 0	Mild weakness = 2	Moderate weakness (i.e., ~50% weak, ±15%) = 4	Severe weakness = 5
Hip flexion (physician examination)	Normal = 0	Mild weakness = 2	Moderate weakness (i.e., ~50% weak, ±15%) = 4	Severe weakness = 5
<i>TOTAL</i>				_____

