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 (double vision, eye closure, neck flexion/extension, shoulder abduction, hip flexion) are derived from the Quantitative Myasthenia Gravis-Revised (QMG-R) and Myasthenia Gravis Manual Muscle Testing (MG-MMT) instruments with some modifications. The ptosis item is derived from the Quantitative Myasthenia Gravis (QMG) (note that the ptosis item in the original QMG measures time to onset of ptosis, whereas the QMG-R measures maximum severity of ptosis). Maximum time for onset of diplopia and ptosis are reduced from >60 seconds in the QMG-R 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 keeps eyes closed) instead of the MG-MMT use of four categories. Neck flexion, shoulder abduction and hip flexion muscle strength is scored based on the modified Medical Research Council (MRC) grading (see below for details).[1]

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.[2]

General procedures for administration

- **Patient reported items:**
  - In most instances, participants 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 participants 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-R assessment has already been performed, the item score for diplopia can be calculated from the QMG-R, provided the exact time for diplopia onset is recorded. Importantly, the maximum duration and item score cut-offs are not the same between the QMG-R and MGC (Table).

Table. Duration of assessment for QMG and MGC

	QMG	MGC
Diplopia	0-61s	0-45s

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

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ITEM-SPECIFIC MYASTHENIA GRAVIS COMPOSITE SCALE INSTRUCTIONS

1. Ptosis, upward gaze

The participant should be sitting with feet supported, approximately 3 feet (1 meter) from the examiner. Eyeglasses should be off. Contacts can remain in. If the participant has a head drop, you may have someone hold the head up to complete this test. The examiner should sit in front of the participant at approximately eye level.

Give the following instruction:
"Look straight ahead. Make sure that your forehead is relaxed."
If the participant has ptosis where the upper eyelid drops down midway through the pupil looking straight ahead, stop the test and record a time of 0.

If there is no ptosis or insufficient ptosis (i.e., upper lid does not cross midway through the pupil) while looking straight ahead, give the next instruction:
"When I ask, look up at the ceiling without moving your head. Keep looking up until I tell you to relax. You will look up for up to 45 seconds."

If the participant has ptosis instantaneously when looking up to the ceiling (again, upper eyelid drops down midway through the pupil), stop the test and record a time of 0. If not, start the timer. Stop the test if the upper eyelid drops down midway through the pupil or after 45 seconds. Record the time at which ptosis developed or the test was stopped.

Additional Considerations:

- You may ask the participant to look up your finger rather than the ceiling (some participants prefer to have a specific target to look at).
- If the subject cannot move their eyes, they should be given a time of 0.

2. Double vision on lateral gaze, left or right

Transfer **time** from QMG-R scale, and score based on MGC cut-offs.
If QMG-R has not been performed, or time not documented, then refer to the separate QMG-R instructions.

3. Eye closure

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

If the QMG-R 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 keeps eyes closed (2)).

If the QMG-R 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 has not been performed, then refer to the separate MG-ADL instructions.

5. **Chewing**

   Transfer response from MG-ADL.

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

6. **Swallowing**

   Transfer response from MG-ADL.

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

7. **Breathing**

   Transfer response from MG-ADL.

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

8. **Neck flexion or extension**

   The participant should be positioned supine for testing neck flexion, and prone for testing neck extension. If the participant is unable to lie in these positions, the reason(s) should be clearly documented on the case report form. The alternative position is seated. Arms should be at the participant's sides. 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. Grade strength using the modified Medical Research Council (MRC) grading system,* and the weaker of neck flexion vs. extension is used to score item:

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

9. Shoulder abduction

The participant should be sitting upright, feet planted on ground, and without back support. The arms should be abducted to 90 degrees and elbows should be flexed. Shoulder abduction should be tested bilaterally at the same time by putting downward pressure on upper arms above the elbows. 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 weaker side should be recorded. Grade strength using the modified MRC grading system:*

- Mild weakness = MRC grade 4+
- 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 participant in the supine position and with the knee flexed at 90 degrees. Testing is performed unilaterally by exerting resistance just proximal to the knee. If testing cannot be performed in the recommended position, the reason for this should be clearly documented (e.g., orthopnea). The alternative position is seated. In the seated position, testing is also performed unilaterally. Whichever position (supine or seated) is used at baseline, should ideally be used throughout the study. Ask the patient to raise their knee and then the examiner applies downward pressure on the thigh just proximal to the knee. Document the testing position on the case report form. 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 weaker side should be recorded. Grade strength using the modified MRC grading system:*

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

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

REFERENCES:

1. Medical Research Council. Aids to the examination of the peripheral nervous system. London, England: The White Rose Press; 1976; Memordanum No. 45. ISBN 0 11 450033 9.
2. 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

***** Modified MRC grades:

4+: muscle can be moved against gravity and strong resistance

4: muscle can be moved against gravity and moderate resistance

4-: muscle can be moved against gravity and slight resistance

3: muscle can be moved fully against gravity

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Participant ID: _____

Visit ID: _____ **Date of Evaluation: _____**

MG Composite Scale

Ptosis, upward gaze (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 eyes 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 = **2** ◯	Frequent choking necessitating changes 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 = **3** ◯	Severe weakness = **4** ◯
Shoulder abduction (physician examination)*	Normal = **0** ◯	Mild weakness = **2** ◯	Moderate weakness = **4** ◯	Severe weakness = **5** ◯
Hip flexion (physician examination)*	Normal = **0** ◯	Mild weakness = **2** ◯	Moderate weakness = **4** ◯	Severe weakness = **5** ◯
			TOTAL SCORE (Sum of items 1-10)	

Evaluator Signature: _____ Date: _____

*Modified MRC:

- Mild weakness = MRC grade 4+ (muscle can be moved against gravity and strong resistance)
- Moderate weakness = MRC grade 4 to 4- (muscle can be moved against gravity and slight/moderate resistance)
- Severe weakness = MRC grade 3 or less (muscle can be moved fully against gravity or less)