

Myasthenia Gravis Impairment Index (MGII) Administration Manual

Introduction:

The MGII is a measure of disease severity based on the signs and symptoms of myasthenia gravis. It was developed using a patient-centered approach, and following current guidelines for outcome measure development, incorporated patient input throughout the different development phases.

The MGII has 22 patient reported items, administered as a questionnaire, and 6 examination items, performed by a rater. Scores are presented in a sum of all items for a total score, but also as an ocular and a generalized sub-score.

MGII Questionnaire, Administration Instructions:

The patient should be provided with the MGII questionnaire form that includes instructions for self-completion. The patient should complete the MGII without evaluator input or guidance. Similarly, no family members or other visit attendees should provide input to the patient on how to answer the items.

Evaluator Review:

The evaluator should review the MGII questionnaire once the patient has completed the form.

- If any MGII questionnaire item is not completed by the patient, inquire whether the omission was intentional or not. If omitted in error, then ask the patient to complete.
- If a patient asks for clarification about a question, please simply remind the patient to answer questions based on the impact of their MG-related symptoms. Do not help the patient to tease apart relative contributions from MG and non-MG causes.

Scoring instructions are detailed in page 4.

MGII- Examination Instructions

Below are the instructions for the items requiring physical examination. You only need a stopwatch. This part can take up to 8 minutes. Write the score in the examination sheet.

1. Diplopia:

Ask the patient to follow your finger in all primary directions (up, down, right and left). Ask the patient if they have diplopia in any direction, including in primary gaze. Record the number of gaze directions where diplopia is present.

A patient with monocular vision will score 0 in this item. A patient with severe restriction of extraocular movements will score 0 if they do not report diplopia.

2. Ptosis:

Ask the patient to hold their gaze upwards, for up to 60 seconds, and record the time to develop ptosis (in seconds). A patient with clear ptosis at baseline (i.e. lid touching the pupil) will score 2 points in this item, and no sustained upgaze test is needed.

If the patient has previously completed the QMGS and/or the MGC, you can transcribe the time in seconds of the ptosis item. Note that the scoring options are different for the QMGS and the MGII.

3. Lower Facial Strength:

Ask the patient to blow air into their cheeks and hold it against resistance. You can provide cues such as “like blowing a balloon”.

4. Arm Endurance:

Ask the patient to hold both arms out-stretched at a 90° angle with pronated forearms (max 180 seconds). Record the endurance time (in seconds) of the best arm. If the arms are falling, you can encourage the patient to lift them up only once. If the arms fall again, finish the test and record the time.

If the patient has shoulder pain (i.e. rotator cuff injury), only test the non-affected arm.

If the patient will be assessed with the QMGS too, complete the QMGS first as it has a longer maximum abduction time. Record the time for the best performing arm (right or leg) on the MGII scoring sheet. Note that the scoring ranges are different for the QMGS and the MGII

5. Leg Endurance:

Ask the patient to lie flat on their back and raise the dominant leg at a 45⁰ angle, with the knee straight and record the endurance time (max. 90 seconds).

If there are other conditions affecting the dominant leg (i.e hip arthritis), test the non-dominant leg. If the leg falls without touching the bed, you can encourage the patient to lift it up only once. If it falls again or if it touches the bed, finish the test and record the time.

If the patient will be assessed with the QMGS too, complete the QMGS first as it has a longer maximum abduction time. Record the time for the best performing leg (right or leg) on the MGII scoring sheet. Note that the scoring ranges are different for the QMGS and the MGII.

6. Neck Endurance:

Ask the patient to lie flat on the back and hold their head up at a 45⁰ angle. Record the endurance time (max. 60 seconds). If the head starts falling (without touching the bed), you can encourage the patient to lift it up only once. If it falls again or it touches the bed, finish the test and record the time.

If the patient will be assessed with the QMGS too, complete the QMGS first as it has a longer maximum neck endurance time. Record the time achieved on the QMGS on the MGII scoring sheet. Note that the scoring ranges are different for the QMGS and the MGII.

MGII- Scoring Instructions

- The MGII can be summarized as a total score and also as 2 sub-scores, reflecting an Ocular and a Generalized domain.
- The total score is the raw sum of all the items, including the clinical examination (E) and the patient-reported (PR) questionnaire.
- The ocular score is calculated by summing 8 items reflecting ocular impairments. These items are: PR items 1 to 6 and examination items 1 and 2.
- The generalized score is calculated by adding PR items 7 to 22 and examination items 3 to 6.
- The scoring sheet provided can be used for manually calculating sub-scores and can help visualize how items are summed. For larger datasets, we can provide an R script for calculating the sum-scores and imputing missing data.

Missing Data:

- If 2 boxes are marked, count the response indicating more severity. If a mark is placed outside of a box, count the response closer to the mark. If the mark is in the middle of two boxes, count the response indicating higher severity.
- For missing answers, impute the average score for the other items in the same domain or region (ocular or generalized).

Important: We consider the total sum-score reliable when there are ≤ 3 items missing ($<10\%$ missing responses). On the validation and responsiveness studies ($n>200$), less than 5% of patients had more than 3 missing items, so this scenario is infrequent.

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