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Telemedicine visits in myasthenia gravis: Expert guidance and the Myasthenia Gravis Core Exam (MG-CE)

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CONFLICT OF INTEREST
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ETHICAL STATEMENT
We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

SUPPORTING INFORMATION
Additional supporting information may be found online in the Supporting Information section at the end of this article.

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Abstract

Introduction/Aims: Telemedicine may be particularly well-suited for myasthenia gravis (MG) due to the disorder's need for specialized care, its hallmark fluctuating muscle weakness, and the potential for increased risk of virus exposure among patients with MG during the coronavirus disease 2019 (COVID-19) pandemic during in-person clinical visits. A disease-specific telemedicine physical examination to reflect myasthenic weakness does not currently exist.

Methods: This paper outlines step-by-step guidance on the fundamentals of a telemedicine assessment for MG. The Myasthenia Gravis Core Exam (MG-CE) is introduced as a MG-specific, telemedicine, physical examination, which contains eight components (ptosis, diplopia, facial strength, bulbar strength, dysarthria, single breath count, arm strength, and sit to stand) and takes approximately 10 minutes to complete.

Results: Pre-visit preparation, remote ascertainment of patient-reported outcome scales and visit documentation are also addressed.

Discussion: Additional knowledge gaps in telemedicine specific to MG care are identified for future investigation.

Keywords

COVID-19; myasthenia gravis; outcome measures; physcial examination; telemedicine

1 | INTRODUCTION

The coronavirus disease 2019 (COVID-19) pandemic, due to the severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), has made telemedicine utilization an urgent necessity in myasthenia gravis (MG) care to provide continuation of care when routine outpatient operations may have been altered or reduced, and many patients are choosing telemedicine to minimize perceived risk of entering the healthcare setting.[1] However, guidance on reliably assessing MG status through telemedicine does not exist.[2] While remote neurological and neuromuscular physical examinations are outlined,[2–5] existing examination techniques will not adequately or feasibly evaluate weakness in a broad population of patients with MG. Traditionally, diagnosis and management of neuromuscular disorders, which relies on a nuanced physical examination, has been slower to incorporate telemedicine compared with other neurological subspecialties.[2,6] However, a disease-specific telemedicine encounter has potential to provide continuation of care during the pandemic. Telemedicine may also augment care of MG in the future by providing additional assessment of fluctuating weakness, added patient monitoring and education, and reduced burden and costs of healthcare services. Additionally, telemedicine has the potential to increase access to specialized neuromuscular care and clinical trials for patients with MG, especially in rural areas.[7] This paper outlines key components of telemedicine visits for MG and describes a new Myasthenia Gravis Core Exam (MG-CE), whose components can be performed via telemedicine.

2 | METHODS

During April–May 2020, MG experts, neuromuscular trainees, and researchers and clinicians with expertise in telemedicine and clinical trials had Web-based meetings to discuss the merits and limitations of standard MG-specific exam techniques in the telemedicine setting. These weekly discussions occurred iteratively over 4 weeks while clinicians were gaining experience in telemedicine visits using institutional systems and commercially available platforms at seven MG centers in the United States (US). Authors reached agreement through discussion and unblinded voting regarding most feasible and high-yield telemedicine assessment tools in MG.

3 | MG SPECIFIC TELEMEDICINE CONTENT

Telemedicine visits tasks are outlined in Figure 1.

3.1 | Preparation for patients

Best preparation for virtual visits has been reviewed elsewhere.[8,9] This paper will focus discussion on MG-specific considerations. Office staff can provide instructions to patients and sometimes troubleshoot technologic issues. In addition to standard preparation, it is important that patients have a well-lit area, which avoids background glare from windows or bright backlighting. Optimal lighting and camera position at the patient's eye level are critical to reliably observe nuances of ocular, facial, and bulbar weakness in MG. Comfort with technology, fast Internet connections, and adjustments at the beginning of the encounter optimize assessments. Tablets or laptop computers are easiest and provide best adjustable camera angles. Smartphones with robust Internet or cellular connections are also feasible, particularly if there is a hands-free stand or an assistant to direct the phone's camera (Figure 1).

Vital signs are an important component of the physical examination for patients with MG. Some treatments are weight-based (eg, immunoglobulins [intravenous or subcutaneous], corticosteroids and azathioprine) or may cause weight gain or hypertension, which require monitoring (eg, corticosteroids). Temperature, heart rate, blood pressure, and weight measurements are recommended if the patient has equipment and is able to obtain them reliably. Vital sign assessment can be performed by patients prior to the visit on the same day.

Recommended: Patients may also complete the MG Revised Quality of Life (MG-QOL-15r) Scale,[10] which is available through the MyMG app or the Myasthenia Gravis Foundation of America (MGFA) website.[11] Physicians or trained ancillary staff can also elicit responses from patients with support of smartphrases or flowsheets in the electronic medical record before or during the visit (Figure 1).

3.2 | Visit components

1. **History:** MG-specific elements in addition to standard elements of the neurological history are important. The MG Activities of Daily Living (MG-ADL) scale is easily performed by telemedicine and provides key elements of the MG history.[12] Authors created a Smartphrase for the MG history and MG-ADL to facilitate insertion into the clinical note. MG medications, dosages, and potential side effects are queried in detail.

2. **General Examination:** This remarks on mental status and general appearance including any cushingoid features or skin changes, such as skin thinning or bruising for patients taking corticosteroids, and keratotic lesions for those on azathioprine.

MG-CE: The following eight core elements are recommended for all telemedicine assessments of MG: ptosis, diplopia, facial strength, bulbar strength, counting to 50, arm strength, single breath count, and sit-to-stand. The assessment takes approximately 10 min. We outline components of the MG-CE and illustrations of how to perform (Figures 2 and 3, Supporting Information Figures S1–S3, which are available online) and grade them (Table 1).

3. **Optional MG strength examination components:**

   a. Neck flexion assessment, which is part of the Quantitative Myasthenia Gravis (QMG) scale, can be incorporated into selected telemedicine visits, particularly when patients report neck weakness or respiratory symptoms. Assessment involves the requirement of a flat surface such as a bed or a long, firm couch, within the video frame of the examiner. The patient lies supine and lifts the head with the neck flexed to 45° and attempts to maintain this position for 120 seconds. This exam maneuver can provide valuable added information in some patients; however, the requirement to have a surface to lie on, to be able to maneuver to this area and be in view of the camera is not feasible for all patients with MG

   b. Assessment of swallowing by observing the patient swallow 4 oz (1/2 cup) of water can also be performed. For safety reasons and concern for managing any acute difficulty swallowing, it is best done in patients who do not report choking or nasal regurgitation. For this reason, it was considered optional by this group and was not included in the MG-CE

   c. Testing of casual gait for several feet in an open space can sometimes provide added information for assessment. Typically, gait dysfunction in patients with MG is related to other causes, such as musculoskeletal disorders. Therefore, assessment may be illustrative if the patient has gait dysfunction, to be able to distinguish the contribution from MG to the patient's symptoms

4. **Documentation:** Virtual visit documentation includes platform used, patient and physician location, who was in attendance, duration in minutes, and that consent for both the specific platform and billing was obtained. It is important to specify whether any recordings or photographs of the telemedicine visit were made. Documentation of the MG-ADL and MG-QOL-15r scales, if used, as well as the MG disease summary and the physical exam are recommended. Scoring of the MG-CE is outlined (Table 1)

5. **Visit follow-up:** Providing written instructions via electronic medical record functions is recommended, particularly when titration of medications is involved. Prescriptions, local laboratory testing, or infusion orders are then transmitted electronically. Identifying a route to and arranging local laboratory testing is an added task of the telemedicine visit. Customary time frames for follow-up may need to be adjusted based on the desired follow-up type (ie, telemedicine or in-person).

4 | DISCUSSION

The MG-CE is adapted from standard examination techniques in MG and evaluates the most common and critical areas of weakness in MG. It can be performed rapidly and generates essential data to triage severity of MG weakness and to follow elements of weakness in an individual patient through both telemedicine and in-person visits. Virtual visits, including the MG-CE, when combined with key elements of the history, and patient-reported outcome measures, such as the MG-ADL and MG-QOL-15r scores, have allowed for remote evaluation and management when in-person assessment has not been possible or optimal.

The MG-CE has several strengths. Its components were derived from validated MG specific scales,[13] and for that reason, the authors think it will be feasible and reliable when performed by neuromuscular specialists who are accustomed to performing these scales. Limitations of the MG-CE include that this is an expert-derived objective scale and validation is needed in particular with respect to additional previously validated MG outcomes. Further study is also required to assess the MG-CE intra-rater and inter-rater reliability between specialists and trainees or general neurologists. A second limitation is that assessment of bulbar, neck, and limb strength is particularly challenging remotely. Additional examination maneuvers are typically included in telemedicine visits when these areas are affected clinically. These include tests of facial strength (ie, raising eyebrows and smiling in addition to eye closure and cheek puff), and neck strength in a supine position, swallowing, and gait (as described above). They can be added to the visit based on the examiner's discretion. Without a trained on-site evaluator, formal manual muscle testing, such as is needed for the MG Manual Muscle Test (MMT) scale,[14] and detection of more subtle weakness in these regions is not feasible.

As such, the MG-CE as part of a comprehensive telemedicine visit will likely have most utility for assessing patients with a well-established diagnosis of MG, a recent in-person examination, and for whom patient reported severity measures closely track with abnormalities on MG-specific neurological examinations. Telemedicine will likely be less useful for new patients, patients in whom the diagnosis of MG is uncertain and in whom the reported symptoms tend to markedly overestimate or underestimate findings on examination. For most patients, in-person assessment will still provide more accurate and complete information in MG. Some of these limitations in the MG-CE could be addressed in the future if broader telehealth applications are desired with an on-site evaluator trained in manual muscle testing or added supplemental technologies to measure these additional areas of weakness remotely.

Using the approach outlined in the paper has allowed us to continue providing care to patients with MG during the COVID-19 pandemic. For patients who are requiring frequent visits during the COVID-19 pandemic, alternating in person and virtual visits has been a preferred strategy. Additionally after the pandemic, we anticipate that a mix of in-person and telemedicine assessments will be needed to provide a balance of added convenience and feasibility for patients and the detailed and accurate assessments required to best manage MG.[15]

We anticipate that future application of the MG-CE to serve areas without access to neuromuscular disease experts or by MG experts assessing patients remotely will assist in optimizing MG care. However, we will have to ensure that telemedicine in MG does not further the digital divide and that tools are able to be leveraged for use by a representative group of our patients.[16] When patients do not have the Internet capabilities or computer literacy to participate in video telemedicine, telephone visits can be performed. These telephone encounters are further limited in scope. Vital signs, the MG-QOL-15r, and the MG-ADL, along with objective assessment of dysarthria and breathing through the counting out loud and single breath count components, can be performed via phone-only encounters.[17] Expanding access to the technology and improving patient readiness for telemedicine and the accessibility of the interface should be considered into the development of future programs. For these reasons, additional study of telemedicine feasibility, acceptability, and outcomes in MG both during and after the COVID-19 pandemic is planned.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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DATA AVAILABILITY STATEMENT

Data available on request from the authors

Abbreviations:

COVID-19	coronavirus disease 2019
MG	myasthenia gravis
MG-ADL	Myasthenia Gravis Activities of Daily Living
MG-CE	Myasthenia Gravis Core Exam
MGFA	Myasthenia Gravis Foundation of America
MG-QOL-15r	Myasthenia Gravis Revised Quality of Life
MMT	Manual Muscle Testing
QMG	Quantitative Myasthenia Gravis (score)
SARS-CoV-2	severe acute respiratory syndrome coronavirus 2

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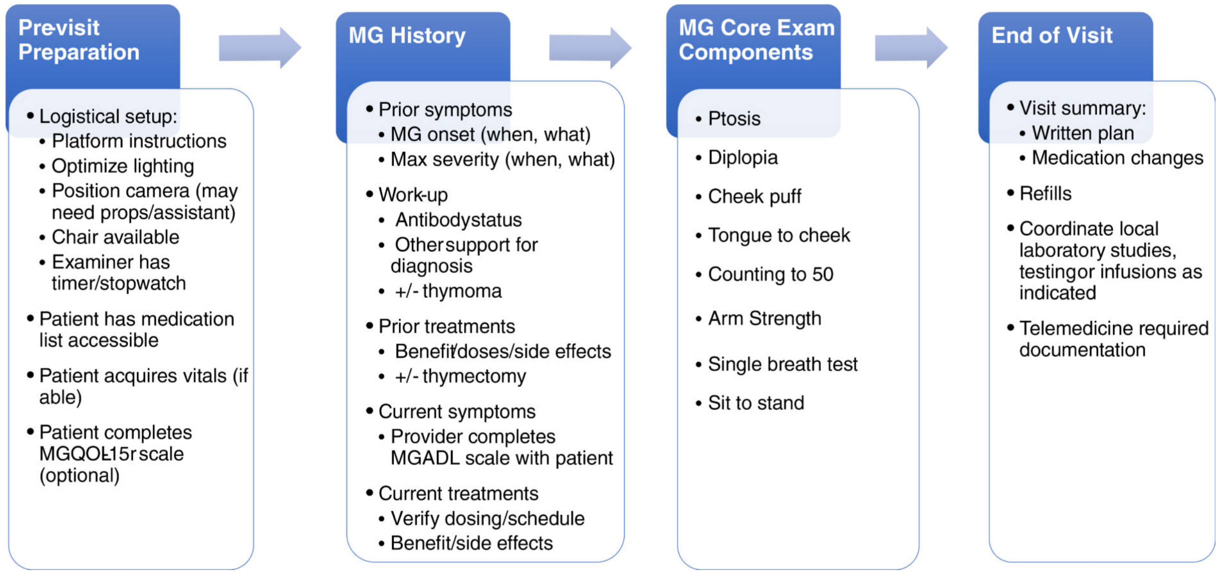

FIGURE 1.

MG telemedicine visit workflow [Color figure can be viewed at wileyonlinelibrary.com]

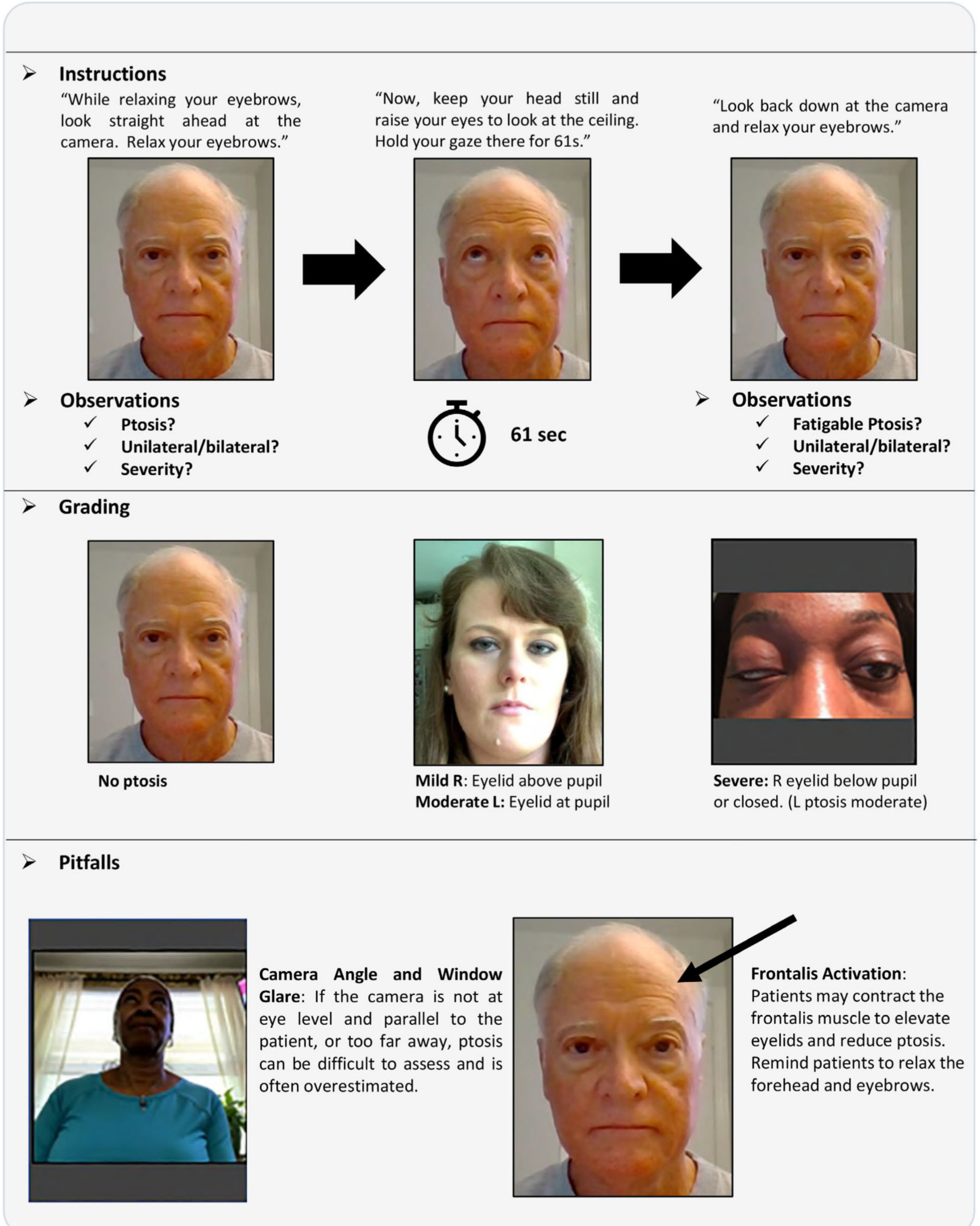

FIGURE 2.

MG Core Exam – Ptosis [Color figure can be viewed at wileyonlinelibrary.com]

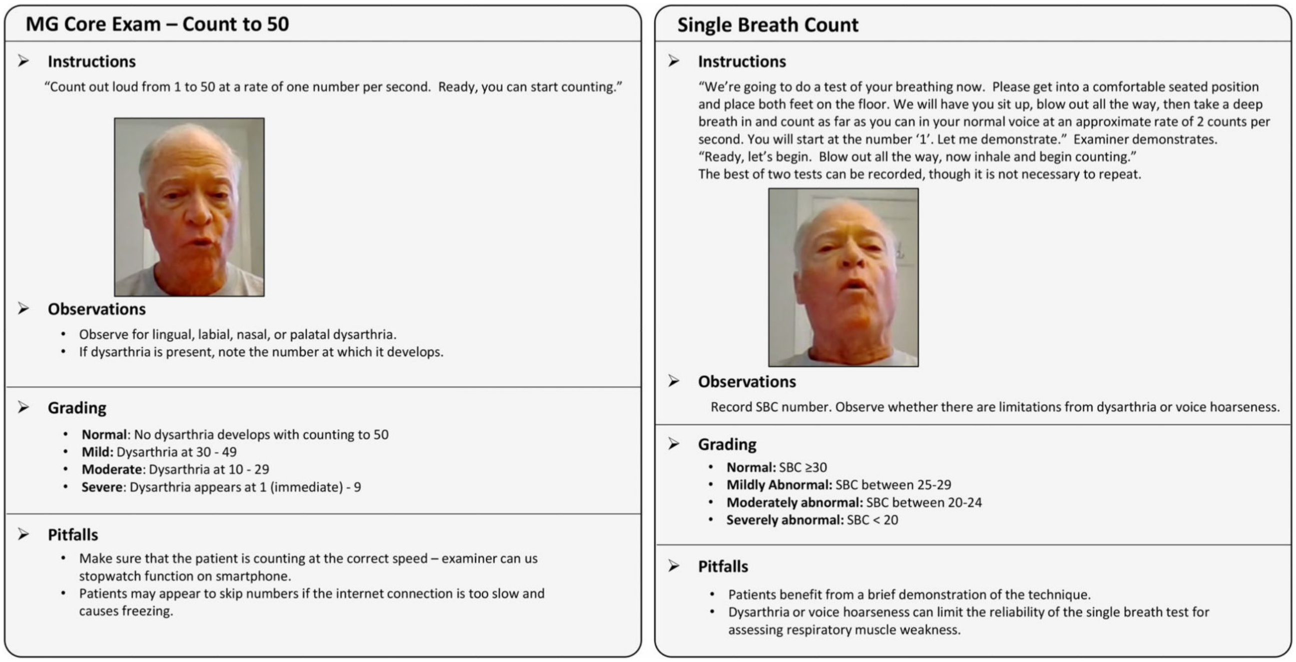

FIGURE 3.
MG Core Exam - Count to 50 and single breath count [Color figure can be viewed at wileyonlinelibrary.com]

TABLE 1

MG Core Exam Scoring

	Normal	Mild	Moderate	Severe	Item Score
Ptosis (61 seconds(s) upgaze)*	0 No ptosis	1 Eyelid above pupil	2 Eyelid at pupil	3 Eyelid below pupil	
Diplopia*	0 No diplopia with 61 s sustained gaze	1 Diplopia with 11–60 s sustained gaze	2 Diplopia 1–10s but not immediate	3 Immediate diplopia with primary or lateral gaze	
Cheek puff	0 Normal "O" seal	1 Transverse pucker	2 Opposes lips but air escapes	3 Cannot oppose lips or puff cheeks	
Tongue to cheek*	0 Full convex deformity in cheek	1 Partial convex deformity in cheek	2 Able to move tongue to cheek but not deform	3 Unable to move tongue into cheek at all	
Counting to 50	0 No dysarthria at 50	1 Dysarthria at 30–49	2 Dysarthria at 10–29	3 Dysarthria at 1–9	
Arm strength (shoulder abduction)*	0 No drift during > 120 s	1 Drift 90–120 s	2 Drift 10–89 s	3 Drift 0–9 s	
Single breath count	0 SBC ≥ 30	1 SBC 25–29	2 SBC 20–24	3 SBC < 20	
Sit to Stand	0 Sit to stand with arms crossed, no difficulty	1 Slow/extra effort	2 Uses arms/hands	3 Unable to stand unassisted	
					Total Score : / 24

* When test is performed on right and left, score the more severe side