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MYASTHENIA GRAVIS AND CRISIS: EVALUATION AND MANAGEMENT IN THE EMERGENCY DEPARTMENT

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☐ **Abstract—Background: Myasthenia gravis (MG) is an uncommon autoimmune disorder affecting the neuromuscular junction and manifesting as muscle weakness. A multitude of stressors can exacerbate MG. When symptoms are exacerbated, muscle weakness can be severe enough to result in respiratory failure, a condition known as myasthenic crisis (MC). Objective: This review discusses risk factors, diagnosis, management, and iatrogenic avoidance of MC. Discussion: MC can affect any age, ethnicity, or sex and can be precipitated with any stressor, infection being the most common. MC is a clinical diagnosis defined by respiratory failure caused by exacerbation of MG. Muscle weakness can involve any voluntary muscle. MC can be differentiated from other neuromuscular junction diseases by the presence of normal reflexes, normal sensation, lack of autonomic symptoms, lack of fasciculations, and worsening weakness with repetitive motion. Treatment should target the inciting event and airway support. All acetylcholinesterase inhibitors should be avoided in crisis, including edrophonium testing and corticosteroids initially. Respiratory support can begin with noninvasive positive-pressure ventilation, as this has been successful even in patients with bulbar weakness. If intubation is necessary, consider avoiding paralytics or use a reduced dose of nondepolarizing agents. Conclusions: MC should be in the differential of any patient with muscular weakness and respiratory compromise. Emergency department management of MC should focus on ruling out infection and respiratory support. Strong consideration should be given to beginning with noninvasive positive-pressure ventilation for ventilatory support. Corticosteroids, depolarizing paralytics, and acetylcholinesterase inhibitors should be avoided in patients with MC in the emergency department. Published by Elsevier Inc.**

☐ **Keywords—myasthenia gravis; myasthenic crisis; neurology; neuromuscular; respiratory; weakness**

INTRODUCTION

Myasthenia gravis (MG) is a relatively uncommon autoimmune disorder of the neuromuscular junction (NMJ) that manifests as weakness of certain muscles that improves with rest. The incidence ranges between 1.7 and 21.3 per million inhabitants worldwide (1–3). Though rare, MG is fairly ubiquitous and can be found in multiple races, numerous geographic locations, and in both males and females of any age (1–4). Neonatal and juvenile forms also exist, with juvenile MG being more common in the Asian population (2,3,5,6). In Asians, 50% of patients with MG are diagnosed in childhood, while juvenile cases only represent 10% of patients with MG in whites (1,3,5). Before 50 years of age, new-onset MG has a female predominance, although the incidence is higher in men after 60 years of age (1,2,4).

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Approximately 85% of cases are caused by acquired antibodies against the acetylcholine receptors at the postsynaptic side of NMJs (1,7). Normally, acetylcholine release by a nerve fiber produces a postsynaptic motor end-plate potential that leads to contraction of the muscle fiber. In MG, the acetylcholine receptors are blocked by autoantibodies. In addition, the receptors themselves are destroyed via a complement-mediated mechanism that leads to a reduced total number of receptors. This results in failure of the muscle to properly contract. Repeated stimulation of the same muscle leads to fewer sites for acetylcholine to bind, resulting in fatigable muscle weakness (3,6–9).

In addition to acetylcholine, other receptor antibodies, such as muscle-specific tyrosine kinase and lipoprotein receptor-related protein 4 have been implicated in causing MG, and it is thought that seronegative patients have antibodies yet to be discovered (7,9,10). Clinical manifestations of MG vary depending on age of onset, antibody implicated, and thymus involvement. Because of this, there are a number of MG classes and subgroups (7,8,10). Both the Osserman and the Myasthenia Gravis Foundation of America classifications have 5 classes of disease based on severity, and the latter has subgroups for predominance of bulbar versus trunk and limb weakness. Class 1 of each manifests with only ocular involvement, while other classes demonstrate progressive severity. MG typically manifests with ptosis, diplopia, ophthalmoplegia, and weakness of the proximal extremity muscle groups, neck extensors, and facial or bulbar muscles (1,11). However, this is one disease entity where a "classic" presentation may not necessarily be the norm.

Myasthenic Crisis

Myasthenic crisis (MC) is a clinical diagnosis defined by respiratory failure in a patient with MG. When symptoms of MG are exacerbated, weakness of respiratory or upper airway muscles can be profound causing difficulty swallowing or breathing, resulting in respiratory compromise (7,8,12–15). Crisis occurs in 15–30% of patients with MG (4,7). MC can occur at any time in the disease course but is more common early after diagnosis (in the first 2–3 years, median 8 months) (1,4,7,11,12). Although most patients with MC will have a previous diagnosis, there are case reports of patients presenting to the emergency department (ED) in crisis at onset (16). Almost any form of stress—physical or emotional—can trigger MC. Infection has been shown to cause >30% of MG crises, with respiratory infections being the most common (7,8,12). Aspiration pneumonitis is the second most common cause (12–14). Many myasthenic patients are immunocompromised at baseline, and others struggle to adequately protect their airways because of fluctuating muscle weakness (7). Crisis can be also precipitated by a number of other causes, including medications, poor control of underlying disease, pregnancy, surgery, acute elevation in body temperature, or even emotional upset (7,8,12,15,17). Even the medications most commonly used to manage MG—corticosteroids and cholinesterase inhibitors—may precipitate MC in patients with severe disease (7,12–14).

Differential Diagnosis

MG has earned the moniker "the great imitator" because of its ability to mimic the symptoms of other disorders, and its numerous subgroups and various antibodies implicated in the disease often make its presentation heterogeneous (7–9). The differential diagnosis for MC is vast and includes any disease that causes muscular weakness, especially bulbar or respiratory muscle weakness. Weakness can occur because of a malfunction in the NMJs or in other areas of the body. Issues resulting in failure of impulse transmission across a NMJ are myasthenic syndromes. Aside from MG, other entities that affect the NMJ include neonatal MG, Lambert–Eaton myasthenic syndrome (LEMS), D-penicillamine–induced autoimmune disease, and toxic causes (botulism, organophosphate and carbamate poisoning, tick or snake neuroparalytic envenomation, and anticholinesterase overdose) (7,17).

Acquired myasthenic syndromes can be broken into autoimmune and toxic forms. Autoimmune MG has been described in detail above. Neonatal MG is likely acquired by passive transfer of maternal antibodies and is self-limiting (9,18,19). LEMS is an autoimmune disorder characterized by antibodies directed against the voltage-gated calcium channels in the presynaptic side of the motor end plate. It is most often associated with small cell lung cancer, although a few cases have been associated with autoimmune MG. Patients with LEMS may experience a transient or mild involvement of bulbar muscles, but respiratory failure caused by LEMS alone is extremely rare. With LEMS, repeated stimulation causes an increase in the amount of acetylcholine in the synaptic cleft, leading to improved motor activity. This is the opposite of what occurs in MG. The drug D-penicillamine (used in Wilson disease or in rheumatic diseases) has also been implicated in an induced form of MG caused by an autoimmune mechanism, but symptoms usually completely disappear after withdrawal of the drug. Toxic paralysis of NMJ transmission can be seen in botulism, neuroparalytic envenomation (e.g., tick and snake bites), organophosphate poisoning, and acetylcholinesterase inhibitor overdose (the latter two present as cholinergic crisis). These may cause severe muscle weakness and respiratory failure (7,17).

In addition, there are several disorders not affecting the NMJ that can cause simultaneous motor weakness and

respiratory distress, including (but not limited to) primary muscle disease (e.g., hypothyroid myopathy, myositis, rhabdomyolysis, and muscle dystrophy, etc.) peripheral nerve disorders (e.g., Guillain–Barré syndrome [GBS] and porphyria), critical illness, motor neuropathy, and central nervous system disorders (e.g., stroke, spinal cord injury, infection, such as tetanus, rabies, or recreational drug overdoses) (7,17). Distinguishing these entities can be difficult. Effort should be taken to distinguish between toxic disorders, central and peripheral nervous system disorders, and MC through history and physical examination. The next discussion will focus on history and examination findings, with differentiating neurologic conditions as a significant component.

DISCUSSION

ED Evaluation and Management

Obtaining a complete history and physical examination can help diagnose MC or determine the precipitant. The patient in extremis may require airway support, and every effort should be made to provide bridging support to obtain a history and focused examination.

History

As discussed, most patients presenting in MC will have a known diagnosis of MG, although patients may present initially with respiratory distress (16). Any neurologic symptoms improving with rest, such as extremity or neck muscle weakness, double vision, dysphagia, or difficulty speaking can suggest MG. A focused review of systems is important, with attention to infectious sources, respiratory symptoms, and toxin or drug exposures (12). A current medication list, with close attention to any new medications or recent medication changes, can assist (7,8). In addition, knowledge of recent trauma, surgery, or a new life stressor can be useful because these may also precipitate MC (7,8,12).

Physical Examination

A physical examination can be used to diagnose MC, find the precipitant of crisis, and help differentiate MG from other neuromuscular diseases. While a complete examination is important, this article will focus on neurologic and pulmonary examinations because these are unique to crisis patients.

Pulmonary Examination

The pulmonary examination in patients with MC can vary greatly. Some patients with MG will present in obvious distress with accessory muscle use, retractions, abnormal lung sounds, or hypoxia. However, the examination in other crisis patients can be more obscure, making diagnosing respiratory failure more difficult. These patients can present in "quiet distress," with clear lung sounds that are diminished at the bases with little to no accessory muscle use. Accessory muscle use can be blunted or absent late in crisis because of poor air movement from accessory muscle weakness (8). Physicians must pay careful attention to respiratory rate, difficulty with phonation, a quiet voice, or weak neck muscles, which can signify respiratory distress or impending failure.

MC is a clinical diagnosis. Pulse oximetry and blood gases are not useful in diagnosing crisis because the partial pressure of CO_2 (pCO_2) rises and oxygen saturation drops late in crisis. A normal oxygen saturation or pCO_2 does not exclude MC (7,8,12–16). Pulmonary function testing and trending values every 2 hours may be useful if physical examination does not suffice. The patient needs ventilatory support or close monitoring if the patient demonstrates vital capacity <10–20 mL/kg or negative inspiratory force < –20 to –30 cm H_2O as shown in Table 1 (7,8,12,13). Despite the wide acceptance of use these values in patients with MG, they are not derived from studies on patients with MG but rather from studies in patients with GBS (20–22). One small observation study of 10 patients with MG suggested that vital capacity does not correlate with the need for mechanical ventilation (23). However, given the lack of larger studies on MG specifically and other studies showing correlation with other neuromuscular disease, these values are generally accepted. Clinicians should focus on the respiratory status of the patient, including respiratory rate, work of breathing, oxygenation, phonation, and trends in these variables, rather than relying on absolute numbers.

Many of these pulmonary function tests are not readily available in the ED, and a single breath test will likely be more useful. The single breath test is a measurement of how many words can be spoken in one breath. The patient takes one breath in and then counts aloud as high as possible before requiring another inspiratory breath. A patient able to count to 50 with one breath correlates

Table 1. Ventilatory Support Threshold*

Pulmonary Test	Threshold for Considering Ventilatory Support	Normal Value
Single-breath test	<15–20 words	40–50 words
VC	<10–20 mL/kg	>60 mL/kg
NIF	<–20 to –30 cm H_2O	−60

NIF = negative inspiratory force; VC = vital capacity.
* Data taken from multiple sources (7,8,11,12,20,24,25).

with good pulmonary function. A patient who can only count to 15 or 20 or less correlates with poor respiratory status, as shown in Table 1 (20,24,25).

Neurologic Examination

A neurologic examination can help differentiate MG from other neuromuscular diseases.

On cranial nerve evaluation, patients may have ptosis that is either unilateral or bilateral (11,26). Pupillary examination should be normal, but unilateral or bilateral extraocular muscles weakness is common (11). Facial muscles may be profoundly weak (7,11). Patients with bulbar palsy can present with dysphonia, dysarthria, or dysphagia. This can be evaluated by listening to the patient speak. Dysphagia can be tested in patients not imminently close to intubation by having the patient drink a few ounces of water (1,7,11,12,27). Muscle strength testing will demonstrate the greatest abnormalities in the proximal limbs and muscles of the neck. Weakness of the neck flexors and extensors can be evaluated by having the patient lift and keep his/her head off the bed, or maintain a neutral position if sitting up, without any drooping of the head (1,8,11,27,28). Sensation and reflexes will be normal, and patients will demonstrate normal Babinski response and clonus (1,2,7).

Most patients in MC will not require immediate intubation, though almost 60% are intubated hours to days after admission (12). However, if the patient's respiratory status is too tenuous to allow a thorough neurologic examination, a rapid 30-s neurologic examination can help differentiate MC from other neuromuscular disease, such as cholinergic crisis, GBS, polymyositis, botulism, motor neuron disease, or spinal cord disease. Testing extraocular muscles; pupil reactivity; sensation in each extremity; the ability to lift the arms, legs, and head (test neck muscles) against resistance; Babinski reflex; and patellar reflex while observing for ptosis, extremity fasciculation, grossly increased secretions, and vital signs can be performed quickly and help differentiate MG from other neuromuscular disease. Table 2 shows the 30-s neurologic examination.

Table 2. Focused Neurologic Examination

30-Second Neurologic Examination
Test
Extraocular muscles
Pupil reactivity
Sensation in each extremity
Reflexes: Babinski, patellar
Strength against resistance: arms, legs, and head
Observe for:
Ptosis
Fasciculation
Grossly increased secretions
Vital signs

Ice Test

The ice pack test is more sensitive and specific than repetitive nerve stimulation and can be used on patients with ophthalmoparesis to help diagnose MG. The ice pack test is done by placing an ice pack over a patient's eye that is affected with either ptosis or ophthalmoparesis for 2 min and reassessing (29,30). If there is 2-mm improvement, the test is positive. In a retrospective cohort study, Farkiri et al. found the ice pack test was 92% sensitive and 79% specific for MG, and other studies have found higher sensitivity and specificity (29,31,32). When compared to repetitive nerve stimulation, the ice test is more sensitive and has similar or better specificity (29–31). Unfortunately, 22% of all patients with MG presenting to the hospital will not have ptosis, which limits its use (11).

Other Aspects of the Examination

The remainder of the physical examination is of lower yield, but examination of the head, eyes, ears, nose, and throat, as well as the skin, may help identify infectious precipitants (12). Neck evaluation may reveal an enlarged thyroid (7,8). The cardiac examination will demonstrate a regular rate and rhythm for most patients, although some degree of tachycardia may be present, particularly if a patient is anxious because of air hunger or septic from infection. Significant autonomic dysfunction (such as irregular cardiac rhythms, tachycardia, bradycardia, hypotension, hypertension, urinary overflow or retention, diaphoresis, or anhidrosis) is unlikely (7). Dysrhythmias may occur, and they have been known to precipitate crisis and have been induced by acetylcholinesterase inhibitors (AChIs) used in the treatment of MG (12). Table 3 shows the examination findings associated with several neuromuscular and neurologic disorders.

The examination features shown in Table 3 can be used to differentiate causes of neuromuscular weakness and respiratory failure. A patient with GBS who has progressed to respiratory difficulty would not be expected to have normal deep tendon reflexes. Both botulism and MC can cause external ophthalmoplegia, but autonomic dysfunction is less likely in MC (7,8).

Cholinergic Crisis

Edrophonium testing in MC is no longer recommended, and AChIs are not commonly used in the treatment of MC (7,8). If a patient with known MG is in respiratory distress with neurologic symptoms, MC is most likely. An 11-year retrospective study evaluating patients with MC found no cases of cholinergic crisis (33). A patient with cholinergic crisis is much more likely to have

Table 3. Examination Features of Neuromuscular and Neurologic Conditions

Clinical Assessment	Myasthenic Crisis	Cholinergic Crisis	GBS	Polymyositis	Botulism	Motor Neuron Disease	Spinal Cord Disease
External ophthalmoplegia	70%*	–	Only + in Miller Fischer variant	–	++	–	–
Pupils	Normal, reactive	Small, reactive	Normal, may be nonreactive	Normal, reactive	Dilated, sluggish	Normal, reactive	Small, reactive
Ptosis	78%*	–	+/–	–	+/–	–	+/–
Muscle weakness	100%*	++	+++	+++	+++	+++	+++
Limb fasciculations	–	+++	+/–	+	–	+++	+/–
Patellar reflexes	++	++	–	+	–	+++ or –	+++ or –
Babinski reflex (toe extension)	–	–	–	–	–	+	+/–
Sensation	Normal	Normal	Sometimes lost	Normal	Change	Normal	Sensory level
Autonomic symptoms	–†	+++	+	–	+++	–	+

GBS = Guillain–Barré syndrome.
* Data from Osserman and Genkins (11).
† Patients with myasthenic crisis may have autonomic symptoms from other causes, such as tachycardia from sepsis, fever, or air hunger. Autonomic symptoms include irregular cardiac rhythms, tachycardia, bradycardia, hypotension, hypertension, urinary overflow or retention, diaphoresis, or anhidrosis.

experienced toxic exposure to organophosphates than to have overdosed on pyridostigmine (7). While both cholinergic crisis and MC share respiratory failure and muscle weakness, as shown in Table 3, physical examination can distinguish these two entities in an undifferentiated patient.

Cholinergic crisis presents with muscle fasciculation and autonomic symptoms, such as bradycardia, miosis, diarrhea, nausea, vomiting, diaphoresis, and tearing (13,14,33). The presence or absence of secretions may not be as diagnostically useful. While increased secretions are more commonly found in cholinergic crisis, oral secretions from a patient in MC who cannot swallow because of bulbar weakness could cause confusion.

Airway Support

Patients with appropriate mentation and secretion management may be appropriate for a trial of noninvasive positive pressure ventilation (NIPPV) (34–36). Research has shown that NIPPV can reduce days of ventilatory support, time in the intensive care unit (ICU), and duration of hospital stay for patients with MC (34–36). Even in patients with bulbar weakness, NIPPV does not increase pulmonary complications when compared to endotracheal intubation (35,36). Seneviratne et al. conducted a retrospective cohort study reviewing 24 patients with MC who were initially treated with bilevel positive airway pressure (BiPAP) (26). Fourteen patients avoided intubation. In their analysis, $pCO_2 > 45$ was a predictor of NIPPV failure (36). Rabinstein and Wijdicks found 7 of 11 patients with MC treated with bilevel positive airway pressure avoided intubation, and hypercarbia ($pCO_2 > 50$) predicted failure (34). In both studies, initial pulmonary function test did not predict NIPPV failure (34,36).

Decisions regarding endotracheal intubation methods in patients with MC are similar to other critically ill patients. We could not identify any studies indicating that patients with MG specifically have more difficult airways. If difficult anatomy is present in patients with MC, case studies of neuromuscular disease, including MG, show success with awake intubation (37–40). To date, there are no studies on preoxygenating patients with MC with bilevel positive airway pressure; however, given the success with NIPPV in MC, it is a reasonable choice. Similarly, there have not been any studies on delayed sequence intubations for myasthenic patients, although it could be considered in patients with poor oxygenation (34–36,41–43).

Rapid Sequence Intubation Medications

Induction agents possess several unique characteristics in this population. Effects of all paralytic agents can be

expected to last two to four times longer (44). Patients with MG are less sensitive to depolarizing agents, requiring higher doses than usual (44,45). These patients have an unpredictable response to succinylcholine, and its use should be avoided (44).

If a paralytic is given, nondepolarizing agents, such as vecuronium or rocuronium, are preferred (44). Patients with MC are more sensitive to nondepolarizing agents (27,46). The published literature suggests that a reduced dose should be used (13,44,45,47–49). However, there have been no studies performed to determine the dose necessary for rapid sequence intubation (RSI) medications in patients with MC. Eisenkreaft et al. found that approximately half the dose of vecuronium was necessary for paralysis of patients with MG during surgery (47). Sungur Ulke et al. began with half the regular dose of rocuronium for paralysis in patients with MG when studying the effects of sugamadex (50). Several studies have shown success with half-doses in noncrisis patients, and even lower doses are required in crisis (44,46,49). It is reasonable to start with one-third to one-half the standard dose of nondepolarizing paralytics. For example, 0.3 to 0.5 mg/kg of rocuronium versus the standard 1-1.2 mg/kg usually used for RSI in the ED can be used.

Some authors advise against using any paralytic in RSI in the ED because of the lack of electromyogram or similar monitoring in departments and unpredictable sensitivity (44,45,48). There are several case reports of successful intubation without paralytics (40,51,52). One case reported using lidocaine, propofol, and remifentanil (a short-acting opioid), a second used propofol and remifentanil, and another used only propofol (40,51,52). It is reasonable to intubate a patient in crisis using propofol and analgesics, such as fentanyl or remifentanil (40). Awake intubation with conscious sedation has been successful as well for both MC and other neuromuscular diseases (37–40). Awake intubation using dexmedetomidine, propofol, fentanyl/remifentanil, lidocaine, benzodiazepines, or ketamine can also be considered (53–55).

Laboratory and Radiology Evaluation

Once a patient has been stabilized from a respiratory standpoint, further evaluation can narrow the differential and evaluate for possible MC precipitants. Infectious evaluation may include a chest radiograph, complete blood cell count, urinalysis, and blood cultures (if indicated, such as in cases of severe sepsis or endocarditis). Lipase, a toxicologic screen, and a brain natriuretic peptide test can be useful if the history and physical examination suggest pancreatitis, toxic ingestion, or congestive heart failure, respectively (12). Chest radiography is an important evaluation that may reveal a mediastinal mass (thymoma is strongly associated with MG) (9,12,56). Metabolic and electrolyte abnormalities or anemia can result in crisis, and laboratory assessment, including magnesium, phosphate, calcium, thyroid function, and hemoglobin, is recommended. Pregnancy can precipitate crisis, and beta-human chorionic gonadotropin levels should be obtained on females of childbearing age (57,18,19). Creatine kinase levels can evaluate for other myopathies. Electrocardiography is useful in evaluating for cardiac dysrhythmia or may suggest a toxicologic etiology (12). Arterial or venous blood gases are not particularly useful in diagnosing crisis but can help with ventilator management and predicting NIPPV failure (9,34,36).

While laboratory values can help find crisis precipitants, there are no useful laboratory values to diagnose MC or MG in the ED. Both PaO_2 and pCO_2 are normal early in crisis, but blood gas values can help determine the severity of crisis. In general, hypercarbia will precede hypoxia (8). As the severity of MG progresses, the likelihood of hypoalbuminemia increases. Hypoalbuminemia is not diagnostic of MC but is common in MC (58). Antibody tests are useful in the diagnosis of MG, but the results are unlikely to be returned prior to the patient's disposition from the emergency department. In addition, 10–15% of generalized MG and 40–71% of patients with ocular MG are antibody-negative (8,10,59).

Treat precipitating event. If an infectious source is suspected, empiric antibiotics should be given. If medications are found to be contributing to crisis, these medications should be discontinued. Any electrolyte imbalance should be repleted, but extreme caution is warranted when repleting magnesium, because overcorrection can precipitate crisis (8,12,13,60).

Edrophonium and Pyridostigmine

AChIs should not be used in the ED for crisis patients. When given intravenously, these medications have been associated with fatal cardiac dysrhythmia (7,12). Edrophonium is a short-acting AChI that can be used to test for MG but is not recommended in patients suspected to be in crisis. This medication in excess may worsen a patient's respiratory status, and because of the complexity of crisis, the test can have false positives or negatives (7). Pyridostigmine, another AChI, is currently used for maintenance therapy and was historically given for crisis (8,9,12,33). Its use in crisis is no longer recommended because of its minimal efficacy and worrisome side effects (7,8,13,14). In addition to dysrhythmia, it also promotes secretions, which can further compromise the airway (7,9,12–14).

Corticosteroids, Intravenous Immunoglobulin, and Plasmapheresis

While corticosteroids are an effective immunosuppressant and are used successfully both in maintenance therapy and in crisis, they should not be started in the ED (7,9,56,61). Corticosteroids can precipitate crisis or cause further decompensation in patients with MC (7,9,12,13,56,61). Worsening symptoms are seen in 30–50% of patients with MG after steroid initiation (9,12,56,61). Therefore, steroid treatment in crisis should be started after plasmapheresis or intravenous immunoglobulin treatment (8,9,12,13,61).

Plasmapheresis and intravenous immunoglobulin are the mainstay of MC treatment (7–9,12,13,56). There is much controversy over which is more effective in treatment of crisis, with a positive trend for plasmapheresis (7,12,13,56). Both treatments take days to weeks for full effect (13). Immunoglobulin A deficiency screening is recommended prior to intravenous immunoglobulin treatment, and plasmapheresis is technically difficult in the ED (12,13). It is not necessary to begin these treatments in the ED, and they may wait until transfer to the ICU.

Disposition

Patients requiring ventilatory support and those with pulmonary function tests below the discussed threshold should be managed in the ICU (1,7–9,12). If patients do not require mechanical ventilation but have bulbar weakness or another indication of impending respiratory failure, admission for cardiopulmonary monitoring in the ICU or step-down unit, pulmonary function testing every 2 h, and neurology consultation are recommended because of the risk of severe pulmonary complications and decompensation (12,61).

Inpatient Management: Items to Anticipate

Understanding the details of inpatient treatment of MC is useful to aid in disposition and provide information to patients and their families. The average patient may need ventilatory support for more than a week and will likely have a several-week hospital stay (7,12–14,33,34,36). In the hospital, a member of the neurology department will follow the patient, and after crisis has resolved, pyridostigmine and additional immunosuppressive mediations such as azathioprine, mycophenolate, cyclosporine, methotrexate, rituximab, or tacrolimus may be started (7,9,13). Thymectomy may also be considered after the crisis has resolved (7–9,56).

If the diagnosis is uncertain, additional diagnostic procedures, such as lumbar puncture, electromyography, or obtaining a muscle biopsy specimen may be performed. Serum antibodies can help monitor therapy success, while future toxicologic analysis, metabolic screens, and gene mutation analysis can assist in evaluating for other conditions (1). Computed tomography or magnetic resonance imaging may also help evaluate for other diagnoses, as well as thymoma (1,8,14).

ED Management Pitfalls (and How to Avoid Them)

Although MC management is the primary focus of this article, patients with a history of MG who are not exhibiting signs of respiratory failure—or who present to the ED for a complaint that is completely unrelated to their MG—should be regarded cautiously, as there are a few common ED therapies and medications that could result in iatrogenic precipitation of a crisis. Common medications linked to exacerbation are shown in Table 4.

Antibiotics

A variety of antibiotics are associated with exacerbating MG (2,62,63). High-risk antibiotics should be avoided. Most exacerbations to antibiotics will occur in the first 24–48 h, and therefore 24–48 h observation should be considered depending on the risk of exacerbation and patient social factors (63).

High Risk

Aminoglycosides. These drugs are known to prevent binding of acetylcholine by blocking the acetylcholine

Table 4. Commonly Used Medications in the Emergency Department that May Worsen Myasthenia Gravis

Antimicrobials	Aminoglycosides
	Quinolones
	Macrolides
	Antimalarials
Anticonvulsants	Phenytoin
	Carbamazepine
Antipsychotics	Chlorpromazine
	Prochlorperazine
Cardiovascular agents	Beta-blockers (including topicals)
	Calcium channel blockers
	Class I antidysrhythmics (lidocaine and procainamide)
Neuromuscular-blocking agents	Succinylcholine
Muscle relaxants	Long-acting benzodiazepines
	Baclofen
Others	Iodinated radiocontrast agents
	Magnesium sulfate and citrate

Data from multiple sources (2,6,7,12,44,56,62).

receptor. This class is well known to worsen muscle weakness in patients with MG (2,63–65).

Fluoroquinolones. Fluoroquinolones have been associated with worsening dyspnea, generalized weakness, dysphagia, diplopia, and ptosis (2). These drugs are currently contraindicated given a black box warning against their use in patients with MG (63,65–71).

Moderate Risk

Both macrolides and polymyxin B are considered moderate risk drugs in patients with MG (2,63,72–76).

Lower Risk

Penicillins, carbapenems, cephalosporins, nitrofurantoin, clindamycin, sulfonamides, doxycycline, and tigecycline all carry a risk of exacerbating MG, but the risk is lower than antibiotics previously discussed (62,63,75–77).

In addition to antibiotics, several other medications have been implicated in worsening disease. The goal is to avoid any unnecessary medications. Common ED medications that may precipitate crisis or worsen muscle weakness are discussed below.

Magnesium. Patients should be cautiously treated with magnesium if repletion is required, and magnesium for headaches or dysrhythmia should be avoided if possible. Magnesium competitively blocks calcium entry at the motor nerve terminal and interferes with neuromuscular transmission by inhibiting release of acetylcholine. Patients with underlying neuromuscular disorders can experience magnesium-induced muscle weakness (2,60,61,78–82).

Beta blockers. The exact mechanism by which beta blockers reduce transmission across the NMJ is unknown, but all beta blockers have been implicated in worsening muscular weakness, even topical (ophthalmologic) preparations (2,65,83–86).

Corticosteroids. This is somewhat counterintuitive, because immunomodulators are a mainstay of maintenance therapy for patients with MG (6,7,56). However, it has been shown that ≤50% of patients with MG who receive high-dose corticosteroids will have an early exacerbation. In 10% of patients, this exacerbation is severe enough to require mechanical ventilation (6,12,56). The mechanism for this is unclear.

Radiographic contrast agents. It was thought new low-osmolality contrast was safe to use in patients with MG, as case reports and studies showing contrast-induced MG exacerbation were completed with high-osmolality contrast material (2,63,87–92). A study by Frank et al. also argued contrast was safe in patients with MG because the incidence of adverse outcome from contrast in MG was the same as the adverse outcomes in the general population for high osmolality contrast (93). However, an exacerbation of MG is more life-threatening than the majority of contrast reactions (92,94,95). With high-osmolality contrast, the incidence of adverse reaction was 4–13%, while with the new low-osmolality contrast it is 1–3% (94,95). Frank et al.'s argument does not hold true with low-osmolality contrast, because the incidence of low-osmolality contrast reactions in the general population is now lower than the incidence of reaction in patients with MG.

A retrospective cohort study by Somashekar et al. found a 5–6% exacerbation frequency with low-osmolality contrast, including weakness, respiratory distress, and death (92). Their study over 16 years suggests that low-osmolality contrast can exacerbate MG and result in complications. Exacerbation frequently was significantly different from those not receiving contrast for 24 h. Caution should be used when administering contrast, and 24-h observation should be considered after administration (92).

Neuromuscular-blocking agents. As previously discussed, depolarizing agents (succinylcholine) should not be used and nondepolarizing agents should be avoided if possible or used in low doses because they have been shown to cause prolonged paralysis in patients with NMJ disease (see Airway section) (44,62).

CONCLUSIONS

MC should be considered in patients with muscular weakness, particularly those in respiratory distress. A focused physical examination is vital in these patients, paying close attention to a patient's work of breathing. Respiratory compromise can present subtly, especially in patients with severe MC. A thorough neurologic examination can help differentiate this disease from other neuromuscular diseases and should be obtained prior to intubation, if possible. NIPPV has been shown to be successful in preventing intubation in crisis patients and can also be considered as a bridging therapy while obtaining valuable historical and physical examination information. If paralytics are necessary for intubation, low dose nondepolarizing agents should be used and depolarizing paralytics avoided. Corticosteroids and AChIs, such as edrophonium and pyridostigmine, should be avoided in the initial management of MC. For patients with a history of MG who present to the ED without evidence of respiratory distress, physicians

should avoid medications and therapies that can precipitate crisis.

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ARTICLE SUMMARY

1. Why is this topic important?

Myasthenia gravis (MG) is an autoimmune disease that affects the neuromuscular junction. Most commonly manifesting as weakness, several stressors may result in exacerbation potentially severe enough to result in respiratory failure, known as myasthenic crisis (MC).

2. What does this review attempt to show?

This review evaluates MG and MC pathogenesis, risk factors and causes, evaluation, diagnosis, and management.

3. What are the key findings?

MC affects those with MG and can be caused by any stressor, although infection is the most common. The presence of normal reflexes, normal sensation, lack of autonomic symptoms, lack of fasciculations, and worsening weakness with repetitive motion is characteristic of MG, with respiratory failure a severe complication in MC. Focused history and evaluation of the respiratory and neurologic systems are essential, with treatment targeting the inciting event. Airway support may be needed. Acetylcholinesterase inhibitors should be avoided in patients with MC, as should edrophonium testing.

4. How is patient care impacted?

Emergency physicians should consider MC in patients with muscular weakness and respiratory distress. Evaluation for underlying stressors including infection is required, with respiratory support provided. Corticosteroids, depolarizing paralytics, and acetylcholinesterase inhibitors should be avoided in patients with MC in the emergency department.