

PEER REVIEWED

# Myasthenia Gravis, the Great Masquerader

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## Abstract

*Myasthenia gravis (MG) is often called “the great masquerader” because the initial presentation of the disease can be widely variable. We demonstrate a unique case in which a patient first presented with what was thought to be an isolated left fourth nerve palsy. Months later, he developed acute, severe ophthalmoplegia in the right eye and profound, bilateral ptosis. The patient was seen by neurology emergently, diagnosed with suspected MG, and started on pyridostigmine. He subsequently developed cholinergic toxicity from pyridostigmine. This case highlights an atypical initial presentation, as well as treatment complications that required coordination with neurology to prevent life-threatening sequelae.*

**Key Words:** *myasthenia gravis, ocular myasthenia gravis, pyridostigmine, fourth nerve palsy, bilateral ptosis*

## Background

A 76-year-old patient presented with a complaint of binocular diplopia that began months prior. A complete history, eye examination and review of systems were performed, after which he was diagnosed with a presumed, isolated left fourth nerve palsy. Given the patient’s systemic history of hypertension and hyperlipidemia, along with lack of other neurological signs or symptoms, the palsy was thought to be likely vasculopathic in nature. His condition was monitored for 5 months and eventually he was prescribed glasses with ground-in prism to neutralize the small, residual deviation. One month later, the patient presented with nearly complete ophthalmoplegia of his right eye and concurrent severe, bilateral ptosis of the upper eyelids. He was referred to neurology for same-day neuroimaging and evaluation as myasthenia gravis was suspected.

This case provides an example of why myasthenia gravis (MG) is often called “the great masquerader.” The initial presentation of the disease is highly variable and can mimic other conditions, such as cranial nerve palsies. Moreover, other conditions that can cause diplopia and eyelid ptosis, such as intracranial mass and aneurysm, can be life-threatening. Because of this, it is imperative for optometrists to have a sound understanding of myasthenia gravis and the many ways it can present so that timely referral and treatment can be achieved for optimal patient care. In addition, myasthenia gravis is a condition that provides a great opportunity for optometrists to collaborate with neurology and possibly other specialties in order to provide the best outcome for the patient. Lastly, this case details the possibility and presentation of cholinergic toxicity from pyridostigmine, a condition that can mimic myasthenic crisis. All optometrists should be familiar with this life-threatening complication so that it can be quickly identified and treated. This case report is intended for third- and fourth-year optometry students, optometry residents and practicing eyecare providers in a primary-care or tertiary-care setting.

## Case Description

A 76-year-old White male presented to the optometry clinic on Jan. 31, 2019, with a complaint of constant, binocular diplopia for months. He also reported painless, progressive vision loss in both eyes.

Both complaints were present for an unknown specific duration as the patient was a poor historian. His known ocular history included bilateral senile cataracts and dry eye syndrome. His systemic history included hypertension and hyperlipidemia. When asked, he stated that his diplopia was less bothersome in left gaze or with right head tilt. It resolved with closing either eye. In primary gaze, it seemed to be mainly vertical but had a small horizontal component. The patient denied any difficulty breathing or swallowing. He was oriented to time, person and place.

Upon examination, his visual acuities were 20/40 in each eye, with no improvement with pinhole or refraction. Pupils were normal in reaction OU without an afferent defect. Confrontation fields were full OU. Extraocular motility testing showed restriction of the left eye in right and down gazes. He had a left hypertropia (8 prism diopters) measured by cover test. Parks three-step test showed the left hypertropia worsened in right gaze and in left head tilt suggesting a left fourth nerve palsy. The patient denied recent trauma as well as any other neurological symptoms. He did not present with a noticeable head tilt, and one was not noted in his driver's license photograph that was examined that day for historical purposes. Given his age and known vascular risk factors of hypertension and hyperlipidemia, the condition was thought to be isolated and likely of a vasculopathic etiology. Neuroimaging was considered but not pursued at the initial visit given the patient's vascular risk factors, lack of other neurological symptoms, and report of the condition being relatively new (within the past 6 months). He did not complain of ptosis, and it was not noted in the clinical findings from that exam. Dilated exam showed nuclear sclerosis OU that was consistent with his vision. Optic nerves revealed healthy rims and distinct margins OU. Fundus exam was unremarkable in both eyes. The exam summary findings were senile nuclear cataracts OU and a new, presumed left fourth cranial nerve palsy. On trial frame, the patient was able to achieve fusion with 8 diopters of base-down prism in front of the left eye. An 8-diopter Fresnel prism was applied base-down over his left eyeglass lens and he was scheduled to be seen again in 1 month. He was educated that if he experienced any worsening of diplopia or new neurological symptoms, he should seek emergent care.

Follow-up examinations in March and April 2019 showed partial resolution of the presumed left fourth nerve palsy, with all other ocular findings stable. His left hypertropia was consistently neutralized with 6 diopters of prism. A 6-diopter Fresnel prism was applied base-down over his left eyeglass lens. No additional neurological signs or symptoms were noted at those exams. The patient was followed monthly until June 2019, at which time he was prescribed new glasses with 6 diopters of ground-in prism, base-down over the left eye, to correct the residual deviation. Given the stability of the condition and no new symptoms being reported, he was scheduled for a follow-up visit in 6 months.

The patient presented to the optometry clinic on Aug. 15, 2019, for an acute visit with complaints of severe bilateral upper eyelid drooping and worsening vertical and horizontal diplopia in all gazes over the previous month. Upon exam, the ptosis was complete in the right eye, with the palpebral aperture measuring zero. The patient was unable to open the right eye without manually lifting his upper eyelid. His left eye also had severe ptosis, with a palpebral aperture of 3 mm measured in primary gaze. In primary head position, extraocular motility testing showed near complete ophthalmoplegia of his right eye, with very limited movement in all directions of gaze. His left eye showed limited movement in down and right gazes, findings consistent to his previous exams. Pupils were normal in reaction OU without an afferent defect. Confrontation fields were grossly full OU. With his eyelids being held during testing, visual acuity was stable at 20/30 in each eye. Upon questioning, the patient stated that his ptosis was less severe in the morning but worsened throughout the day. He denied difficulty breathing but did state that swallowing and speech were sometimes labored, which he had also noticed over the previous month. The patient was diagnosed with severe bilateral upper eyelid ptosis and pupil-sparing ophthalmoplegia of both eyes, worse in the right than the left. Myasthenia gravis was the suspected etiology due to his ocular and systemic symptoms. Given the severity and acute worsening of his condition, it was deemed appropriate to forgo further clinical testing, such as the ice pack or rest test, and instead refer emergently for a neurologic workup. A routine consultation in the outpatient neurology

clinic would likely not be available for several weeks to months; therefore, neurology advised the patient to be seen in the emergency department that day.

Examination in the emergency department included neuroimaging and chest X-ray, which were both unremarkable. Diagnostic blood work was ordered. The only additional clinical finding from this exam was fatigable weakness of the shoulders and neck. Neurology diagnosed the patient with presumed generalized myasthenia gravis and prescribed pyridostigmine bromide 60 mg three times a day by mouth. It was noted that the patient was currently taking atorvastatin 40 mg daily, but no change or discontinuation of this medication was recommended.

Blood test results received a week later confirmed the condition. The patient's acetylcholine receptor (AChR) binding antibody serum level was highly elevated at 3.89 nmol/L (reference range 0.00-0.24). The patient was monitored by phone call 1 week later by his optometrist, at which time the patient reported noticeable improvement in symptoms. However, in the second week of using pyridostigmine, the patient called to complain of severe side effects after taking medication, including dysphagia, dysarthria and diarrhea. Neurology was consulted, and the patient was instructed to discontinue the medication and seek emergent care. When he presented to the emergency room several days later, he was noted to have lost 25 pounds in the previous month. Neurology diagnosed cholinergic toxicity from pyridostigmine as the cause of his severe reaction and side effects. The patient was taken off pyridostigmine and hospitalized so that he could be given a short course of intravenous immune globulin. He was also started on mycophenolate mofetil (MMF) 500 mg twice per day by mouth for myasthenia gravis. His condition improved dramatically during his 5-day hospitalization.

At a follow-up optometry appointment 1 month later, he showed improved ocular motility of his right eye and only mild ptosis of both eyelids. He again was measured to have a 6-diopter left hypertropia in primary gaze, stable to previous examinations since diagnosed with a left fourth nerve palsy nearly a year prior. The patient noted that his habitual glasses with ground-in prism worked well at times, but that he continued to experience diplopia in secondary gazes and when fatigued. At that visit, his optometrist frosted the right eyeglass lens of an older pair of his glasses. By phone call the next week, the patient reported great success with these glasses. At his next monthly follow-up visit in November, the patient stated that he no longer wore the frosted lens spectacles and had returned to using his habitual glasses with prismatic correction of 6 prism diopters base-down over the left lens.

The patient continues to experience improvement in symptoms as he is followed closely by neurology and optometry. Dysarthria resolved after nearly 3 months of treatment with MMF. The patient continues to take 500 mg of MMF twice per day by mouth to manage his symptoms. At the time of publication, the patient has no ptosis and no diplopia with his habitual prismatic correction.

## **Education Guidelines**

### *Learning objectives*

1. Understand the subtypes of myasthenia gravis and their typical presentations
2. Understand the clinical and diagnostic tests available
3. Understand available treatments and their possible side effects

### *Key concepts*

1. Providers should be aware of the many variable, initial presentations of myasthenia gravis, as well as common ocular and systemic signs and symptoms
2. Appropriate diagnostic tests should be ordered, and neurology referral made in a timely fashion to initiate therapy and rule out other concerning etiologies
3. The treatments available for myasthenia gravis are evolving, and these should be well-known to

optometrists so they can effectively educate patients and be a meaningful part of their multidisciplinary care

### *Discussion questions*

1. What are possible first symptoms and signs of myasthenia gravis?
2. What questions should we ask patients with suspected myasthenia gravis?
3. What clinical and diagnostic tests can be performed?
4. What medical treatment options for MG are available and how successful are they?
5. What side effects can patients experience from treatment?
6. What ophthalmic treatment options are available to manage ocular manifestations?

### *Literature review*

Myasthenia gravis is an autoimmune condition affecting the neuromuscular junctions and is characterized by fatigable muscle weakness.<sup>1,2</sup> The neurotransmitter acetylcholine (ACh) acts at neuromuscular junctions, which causes muscle contraction. In MG, ACh antibodies cause damage to acetylcholine receptors in striated muscle.<sup>1,2</sup> This condition causes weakness and fatigability of certain skeletal musculature and most often affects the limbs, facial expression, ocular movements, chewing and speech.<sup>1</sup> The eyelids and extraocular muscles (EOMs) are involved in more than 90% of cases.<sup>3</sup> Possible explanations for why the EOMs are so often affected in MG include that only slight weakness of extraocular muscles can result in diplopia, the high firing frequency of these muscles make them more susceptible to fatigue, and/or these muscles may be more vulnerable to neuromuscular blockade. The prevalence of MG is approximately 20/100,000 per year in the United States.<sup>1</sup> MG has a bimodal distribution, most commonly affecting women in their 20s and 30s and men in their 60s and 70s.<sup>3</sup> MG shows no racial or geographic predilection.

The condition can be subtyped by clinical features and serum antibodies. Most classify MG as either ocular myasthenia gravis (OMG) or generalized myasthenia gravis (GMG), with the former describing disease that affects only the orbicularis oculi, levator palpebrae superioris, and extraocular muscles causing ptosis and/or diplopia.<sup>1,2</sup> Generalized disease involves the facial, bulbar, lumbar and/or respiratory muscles.<sup>1,2</sup> It is estimated that 50-80% of patients with OMG will develop generalized symptoms, usually within 2 years.<sup>3,4</sup> The subtypes by antibodies include MG with AChR antibodies, MG with anti-muscle-specific kinase (MuSK) antibodies, MG with anti-lipoprotein receptor-related protein-4 (LRP4) antibodies, and seronegative MG.<sup>5</sup> More than 80% of cases are MG with AChR antibodies, 4% are with MuSK antibodies, and 2% are with LRP4 and seronegative disease.<sup>5</sup> Research in MG continues to examine whether these classifications affect prognosis and/or should guide treatment recommendations.<sup>1,5</sup>

The condition can also result from thymus hyperplasia and thymoma. With up to 15% of adult cases of MG having an associated thymoma, thoracic imaging is part of the recommended workup when MG is suspected.<sup>1,3</sup> Other autoimmune conditions such as hyperthyroidism and lupus erythematosus can occur in association with MG.<sup>1,3</sup> Lastly, the condition can be drug-induced or exacerbated by medications such as D-penicillamine, aminoglycosides, beta-blockers, statins and quinidine.<sup>3</sup>

As discussed above, OMG often converts to GMG as more muscles become involved.<sup>1,2,3,4</sup> The most serious complication of MG is referred to as myasthenic crisis, which involves extreme muscle weakness of the diaphragm and chest muscles that support breathing.<sup>1,4</sup> Patients should be educated about the symptoms of myasthenic crisis, which include difficulty breathing and swallowing, and told to seek emergent care if they occur. While many patients with MG eventually improve and may be able to cease treatment, this may take several years, and the disease may have a relapsing and remitting course.<sup>4</sup> Treatment aims to minimize patient symptoms and discomfort and to keep the patient safe until the

disease improves.<sup>4</sup>

## Discussion

Teaching instructions: Participants in this teaching case report should answer each discussion question and then read the discussion section below to enhance their knowledge of the topic. Completion of the discussion questions will allow the learning objectives listed above to be fully assessed.

*What are possible first symptoms and signs of myasthenia gravis?*

An estimated 67-85% of patients initially present with ocular symptoms, the most common being ptosis and/or diplopia.<sup>1,6</sup> These symptoms are often asymmetric and variable throughout the day and dramatically worsen with use and fatigue. Also experienced in the first few years of diagnosis is generalized weakness, specifically in the bulbar, neck and proximal limb muscles.<sup>6</sup> This can inhibit chewing, swallowing, speech, facial expression, raising arms or climbing stairs. Additionally, approximately 40% of patients with MG can develop respiratory muscle weakness that results in shortness of breath with exertion.<sup>1,6</sup> The patient discussed in this case report presented initially with ocular symptoms and later noticed labored swallowing and speech. His ptosis and ophthalmoplegia were asymmetric and worsened as the day progressed.

*What questions should we ask patients with suspected myasthenia gravis?*

When patients present with any of the ocular symptoms discussed, it is important to ask if they vary with time of day and continued effort. Other pertinent questions include whether there is difficulty with swallowing, coughing after attempting to swallow, or shortness of breath with exertion. Various clinical signs such as fatigability of speech and facial weakness can be observed by asking the patient to count out loud to 50 or perform facial expressions such as smiling or closing their eyelids.<sup>3</sup> If the patient is unable to reach 50 or speech becomes less intelligible during the test, it is a sign of enhanced dysarthria and MG should be suspected. Similarly, if a patient is unable to sustain facial expression or eyelid closure for 60 seconds, it is considered consistent with the typical nature of MG.<sup>3</sup>

*What clinical and diagnostic tests can be performed?*

If a patient presents with any of the previously mentioned symptoms, several clinical tests can be conducted in-office to see whether a diagnosis of MG is supported. A hallmark sign known as Cogan's lid twitch was first described in MG patients in 1965.<sup>7</sup> When the patient is asked to gaze downward for 15 seconds, then return to primary gaze, the affected superior lid briefly twitches upward as a result of the rapid recovery of the levator muscle. This is not specific for MG but is often noted as a characteristic sign. Ptosis is the most frequently reported initial symptom with both forms of MG. A "fatigue" test involves having the patient sustain an upward gaze for 30 seconds. If the levator becomes fatigued, a positive test results in increased ptosis after the prolonged upward gaze. The subsequent lid lag or increase in ptosis is referred to as pseudo Von Graefe's sign.<sup>1,8-10</sup> Another non-pharmacologic simple but effective test is the "ice" test. An ice pack is placed on the patient's eyes for 2-5 minutes. A positive result is an improvement of the ptosis greater than 2 millimeters. The decrease in temperature slows the breakdown of ACh, allowing an increase of its availability in the neuromuscular junction. The specificity of this test is reported as high as 98%.<sup>1,8,9,10</sup> An additional screening of the orbicularis oculi involves asking the patient to tightly squeeze their eyelids shut. Normally, the examiner should not be able to pry open a tight lid closure by finger pressure alone. A positive test for weakening of these muscles is a successful attempt at manually overcoming the blepharospasm. A longer duration test is the "sleep" test, which requires the patient to lie in a quiet room for 30 minutes while closing the eyes. The period of rest reduces the demand for and allows replenishing of ACh. A positive test result is improvement of ptosis and/or eye movements after the rest period.<sup>8,9</sup>

Diplopia is caused by a paresis or fatigability of one of the extraocular muscles and is the second most frequent symptom of both OMG and GMG.<sup>10</sup> Cover test and Parks three-step test can be used to assess the alignment and comitancy of deviations. One of the challenging aspects of diagnosing MG is the disease's ability to mimic nerve palsies or other causes of pupil-sparing ophthalmoplegias. A variable and fatigable nature of symptoms are indications for an MG diagnosis. Other historical information such as history of trauma, presence of other neurologic symptoms, systemic history and pupil involvement can help rule out differential causes of diplopia and direct appropriate referral to other services.<sup>10,11</sup>

Diagnostic methods can be utilized to help confirm the diagnosis of MG. The edrophonium (Tensilon or Enlon) method is still recognized as a gold standard diagnostic test. Edrophonium is a short-acting, reversible acetylcholinesterase inhibitor that is administered intravenously.<sup>12</sup> The onset of action is 30-60 seconds, with effects resolving within 5-10 minutes. A positive test result is an improvement in muscle function observed in the eyelids or extraocular muscles as edrophonium increases the availability of ACh at the receptor site. The edrophonium test is highly sensitive for diagnosis when measuring ptosis, as high as 86-97% in OMG and 82-100% in GMG.<sup>1,8,10</sup> Caution is advised during testing due to the rare risk of serious side effects such as bradycardia, hypotension and cardiac arrest. It is recommended to monitor blood pressure and have atropine and resuscitation equipment available during testing.<sup>12</sup> Less invasive tests are now preferred due to these risks and the cholinergic side effects that can be experienced.



Table 1. [Click to enlarge](#)

Laboratory testing for several antibodies can help support or confirm a diagnosis when MG is suspected. An elevated AChR antibody titer confirms the diagnosis, although a negative titer does not exclude the disease. This titer is most helpful in GMG, as 85-90% of GMG patients are "seropositive" and possess these antibodies, compared to only 50-70% of patients with OMG.<sup>10,12</sup> Of the AChR seronegative cases, 40-70% have antibodies to MuSK. The MuSK titers are generally used when a patient tests negative for AChR antibodies. These patients are predominantly female and less likely to have ocular features or a thymoma.<sup>1,10,12</sup> Another antibody test now available is LRP4. Anti-LRP4 antibodies have been found in approximately 9% of patients who are negative for anti-AChR and anti-MuSK antibodies.<sup>12</sup>

With seronegative patients, electrophysiologic testing such as repetitive nerve stimulation (RNS) and single-fiber electromyography (SFEMG) can be performed. With the former, patients with MG show a decrease in the action potential of the repetitively stimulated nerves. An abnormal result is found in 75% of patients with GMG and 20-50% of patients with OMG.<sup>12</sup> SFEMG is the most sensitive test for MG in adults, and up to 80% of OMG patients and 94-99% of GMG patients show an abnormal result. It is not readily available for most communities and is a specialized skill.<sup>8,9</sup> Like the edrophonium test, electrophysiologic testing is typically administered if other testing such as antibody titers are negative.

Lastly, thoracic imaging such as radiography, computed tomography (CT) or magnetic resonance imaging (MRI) can be used to detect a thymoma, which can be associated with MG. Laboratory tests for thyroid function and antibodies are also routinely obtained during initial workup as thyroid disease often coexists with MG.<sup>1,12</sup> **Table 1** summarizes the tests discussed in this section. Our patient's diagnosis of MG was confirmed with an elevated level of circulating AChR antibodies, along with unremarkable chest and neuroimaging.

*What medical treatment options for MG are available and how successful are they?*

Acetylcholinesterase inhibitors are typically the first class of medication initiated as treatment for MG symptoms. Pyridostigmine is the most used of this group and can provide relief of symptoms for up to 6

hours.<sup>8,13</sup> Typically, the dosage is 30 mg taken two to four times daily, increased over time based on efficacy and duration of symptom relief. Approximately 50% of patients with OMG have successful reduction of symptoms with pyridostigmine. Its most beneficial action has been shown to be reduction of ptosis.<sup>8</sup>

Immunosuppressive agents are the next course or additive treatment if acetylcholinesterase inhibitors fail to provide adequate therapeutic relief or cannot be tolerated due to unwanted side effects. Corticosteroids are widely used for their anti-inflammatory benefits, most frequently oral prednisone. Typically, oral prednisone is started at lower doses such as 15-20 mg daily and increased to 60 mg daily to achieve desired therapeutic effects. A reported 66-85% of MG patients benefit from this therapy though side effects must be taken into consideration when weighing treatment options.<sup>8</sup> Non-steroidal immunosuppressive therapies for those who may be poor candidates for corticosteroids can include azathioprine, cyclosporine-A and MMF, all of which have their own set of costs, risks and side effects. MMF is generally used as a second-line therapy and acts by suppressing T-cell and B-cell proliferation. Although costly, it is well-tolerated and has a good safety profile. Typical daily starting dosage is 1000-2000 mg with maintenance dosage of 2000-3000 mg. In several published studies, a significant number of patients on combined MMF and prednisone treatment were able to either decrease dosage or discontinue prednisone entirely.<sup>14,15</sup>

Intravenous immunoglobulin (IVIg) has a role for patients suffering from myasthenic crisis. Plasmapheresis has been used for GMG, but there are no current studies using it for OMG.<sup>13</sup> Monoclonal antibody drugs such as eculizumab or rituximab have been used to treat both subtypes of MG. Although costly, these drugs require less frequent dosing. Most of the latter treatments mentioned are typically used when patients have failed the mainstay drugs.

Finally, thymectomy is performed for those patients with thymoma. It has also been shown to benefit non-thymomatous patients who are early in the course of their disease or younger than age 60. A benefit to thymectomy is the potential for remission of symptoms and discontinuation of medical therapy.<sup>8,13-14</sup>

A controversial topic is whether immunosuppressive therapies, such as corticosteroids or azathioprine, may delay or prevent progression from OMG to GMG as has been suggested in several retrospective studies.<sup>16</sup> Others point out the limitations of these studies, and instead suggest that the effects of chronic immunosuppression may mask the signs and symptoms of conversion to GMG.<sup>3</sup> Further studies are needed for better a understanding of the true effects of immunosuppressive treatment.

#### *What side effects can patients experience from treatment?*

As a mainstay drug for the symptomatic treatment of MG, pyridostigmine is generally considered to have a good safety profile.<sup>14</sup> Its effects can be noticed within 30 minutes of the dose being taken and it has a short half-life. Common adverse events include gastrointestinal disorders such as stomach cramping, nausea, vomiting and diarrhea. Concurrent use of glycopyrrolate or atropine can help eliminate some of the unwanted side effects. Higher doses of acetylcholinesterase inhibitors can cause cholinergic crisis, which is rare but has a mortality rate of 3-25%, most often due to respiratory failure.<sup>17</sup> Symptoms include increased muscle weakness and fasciculations and subsequent exacerbation of cholinergic symptoms such as excessive secretions, gastrointestinal distress and pupil miosis. In cholinergic crisis, acetylcholinesterase inhibitor treatment should be immediately ceased. Due to the many overlapping clinical symptoms, it is imperative this condition is quickly differentiated from myasthenic crisis with examination and edrophonium test, as the treatments are independent.<sup>17</sup> **Table 2** highlights the differences between these conditions.



Table 2. [Click to enlarge](#)

Corticosteroids generally start to improve MG symptoms within 2 weeks and are a low-cost treatment option. Side effects from long-term use of corticosteroids are well-known and include osteoporosis, cataracts, diabetes or worsening of diabetes and hypertension, and weight gain.<sup>13,14</sup> Long-term use of steroids can lead to ocular complications such as cataract progression and intraocular pressure increase that should be monitored routinely by the patient's eyecare provider.

Among the non-steroidal immunosuppressive agents, azathioprine and MMF are widely used. Azathioprine generally has a delayed effect with improvement of symptoms in 6-15 months. Most of the literature indicates it has a safe profile, but patients should be monitored for hepatotoxicity and leukopenia.<sup>8</sup> The therapeutic benefits of MMF are typically observed after 2 months of use.<sup>8,13</sup> While serious side effects are rare, patients more commonly experience gastrointestinal intolerance such as nausea and diarrhea.<sup>14,15</sup>

The patient in our case was started on a conventional dose of pyridostigmine and initially showed improvement of symptoms. After only a short period, he unfortunately experienced the aforementioned condition of cholinergic crisis. This case demonstrates the significance of frequent monitoring with any MG medication, as well as the crucial liaison role of an optometrist to other specialties when new side effects are reported. Thus far, our patient has shown stable symptoms and has tolerated MMF.

#### *What ophthalmic treatment options are available to manage ocular manifestations?*

After diagnosis and appropriate neurology referral, our role as optometrists is focused on relief of persistent ocular symptoms. Occlusion therapy is often an effective treatment for the variable diplopia synonymous with MG. Patching or the use of a frosted spectacle lens can be of great symptomatic relief when dealing with fluctuating diplopia throughout the day. In the rarer cases of stable diplopia, optometrists can utilize application of temporary Fresnel or ground-in prism glasses. In the setting of severe or longstanding ptosis, non-surgical treatments can include botulinum toxin injections or use of a ptosis crutch attached to a spectacle frame.<sup>9</sup> Surgical treatment options include ptosis repair surgery, blepharoplasty, frontalis suspension, external levator advancement and tarsomyectomy.<sup>10</sup> Exposure keratopathy can result from orbicularis oculi weakness, incomplete or reduced blinking with ptosis crutch or botulinum toxin injections and can be managed with topical lubrication.

In addition to taking medication, the patient presented in this case report ultimately benefitted from the use of several optical therapies including Fresnel prism, frosted spectacle lenses and ground-in prism glasses. Optometrists can play a vital role in the management of MG patients to relieve ocular symptoms and provide appropriate surgical referral when indicated.

#### **Conclusion**

Myasthenia gravis is often called "the great masquerader" because the initial presentation of symptoms is widely variable and can mimic other conditions such as cranial nerve palsies. This case provides an example of how challenging diagnosis can be when only extraocular muscles are initially involved. Optometrists should be aware of the potential manifestations of this condition in order to facilitate appropriate and timely testing and diagnosis. Moreover, optometrists should be familiar with treatment options, as well as their potential side effects, so they can effectively co-manage these patients with their neurologists, primary care providers and other specialists. Myasthenia gravis can be a debilitating disease, but optometrists can play a crucial role in minimizing patients' symptoms and suffering.

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