MYASTHENIA GRAVIS

Group B55

INTRODUCTION

- Myasthenia gravis (MG) is an autoimmune disorder characterized by weakness and fatigability of skeletal muscles.
- Muscle weakness due to dysfunction of the neuromuscular junction (myasthenia) may be an acquired disorder, and the vast majority of patients who develop generalized myasthenia in adolescence or adulthood have autoantibodies that play a pathogenically important role.

- When the symptoms of MG are isolated to the levator palpebrae superioris (LPS), orbicularis oculi, and the oculomotor muscles, it is referred to as ocular MG (OMG).
- Over half of all patients with MG initially present with isolated ptosis, diplopia, or both, and no signs or symptoms of weakness elsewhere.

EPIDEMIOLOGY

Age and sex adjusted **incidence of myasthenia gravis** was 0.68 per 100,000 person-years, with highest in the age group of 70–74 years. The **incidence** in females was 0.76 per 100,000 and 0.60 per 100,000 in males

Prevalence of the disease is increasing since 1995.

- High sensitivity & specificity of the test.
- Longer life-span due to effective treatment
- More people in the ‘at risk’ group due to increased life expectancy.

Clinical presentation

Droopy eyelids or double vision is the most common symptom at initial presentation of MG, with more than 75% of patients. These symptoms progress from mild to more severe disease over weeks to months. Difficulty in swallowing, slurred or nasal speech, difficulty chewing, and facial, neck, and extremity weakness occur. [32] On the other hand, symptoms may remain limited to the extraocular and eyelid muscles for years. Rarely, patients with severe, generalized weakness may not have associated ocular muscle weakness.

The hallmark of MG is that muscles get weaker with repeated use

PATHOGENESIS

- Myasthenia gravis is a condition that fulfills all the major criteria for a disorder mediated by autoantibodies against the acetylcholine receptor (AChR)
  - The autoantibodies are present in 80 to 90 percent of affected patients
  - The autoantibodies react with a specific antigen, the acetylcholine receptor
  - The condition can be passively transferred by the autoantibodies to an animal model, producing a similar clinical condition
  - Reduction in autoantibody levels is associated with clinical improvement

- However, the linkage between AChR antibodies and myasthenia gravis is not absolute
- Autoantibodies directed against muscle-specific receptor tyrosine kinase (MuSK) and perhaps other postsynaptic neuromuscular junction components have a pathogenic role in the development of myasthenia gravis.
- Individual patients have a mix of immunologically different antibodies to the AChR. This is partly due to the heterogeneity of the receptor.
- Some patients with myasthenia gravis who are seronegative for AChR antibodies have antibodies directed against another target on the surface of the muscle membrane,.

- Seronegative myasthenia gravis, also called antibody negative myasthenia gravis, refers to patients with myasthenia gravis who have negative standard assays for both AChR antibodies and MuSK antibodies.
- Seronegative myasthenia gravis is an autoimmune disorder with most of the same features as seropositive myasthenia gravis.
- T lymphocytes are also important in myasthenia gravis; they bind to the acetylcholine receptor and their main role is thought to be stimulation of B cell antibody production.
- The majority of patients with AChR antibody positive myasthenia gravis have thymic abnormalities (hyperplasia in 60 to 70 percent and thymoma in 10 to 12 percent.)
- The disease often improves or disappears after thymectomy.
- As a result, the thymus has been evaluated as a possible source of antigen to drive this autoimmune disease.

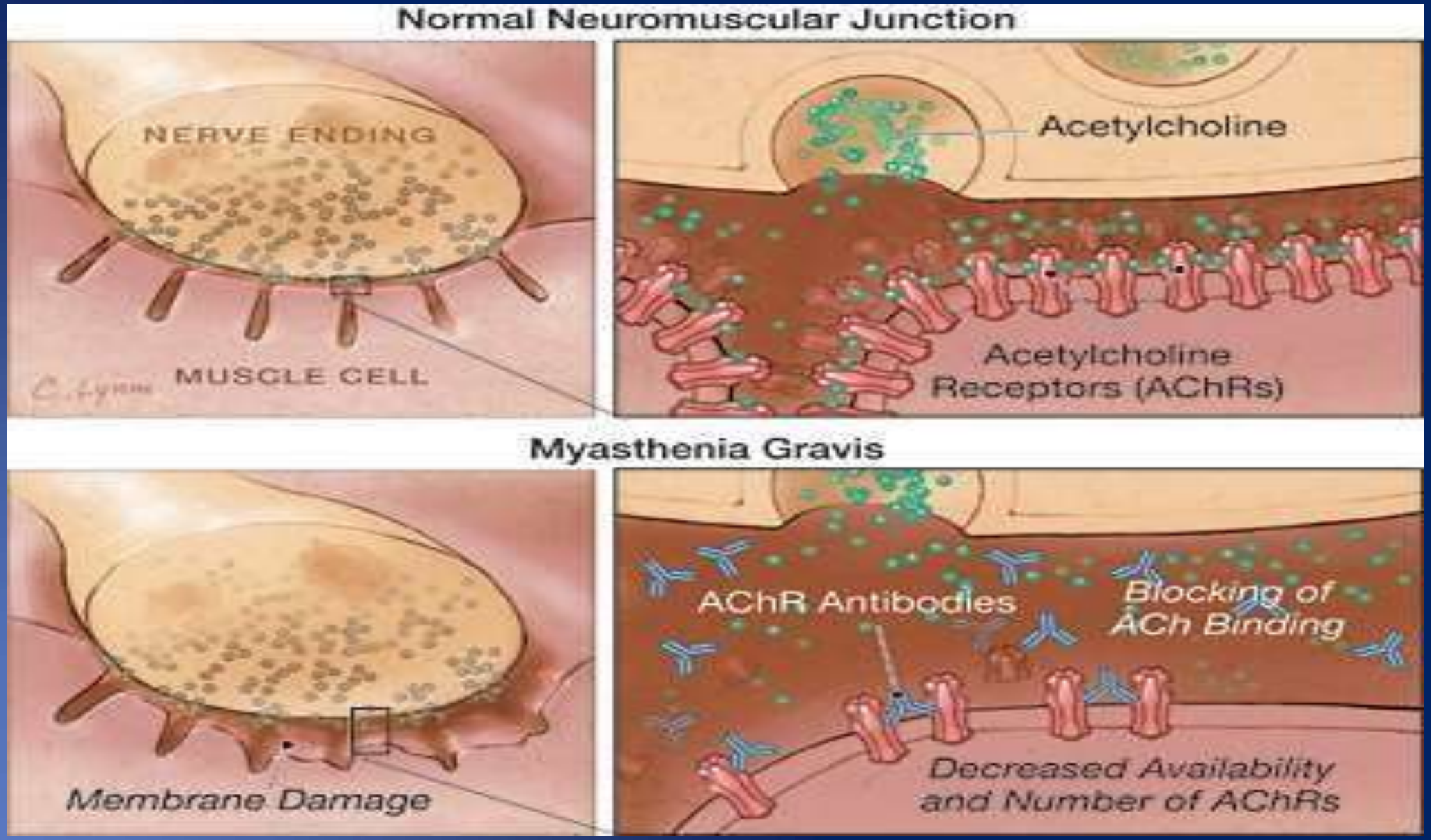
AchR antigenic peptide fragment
+ TCR (T- Cell receptor)

Activation of T-Helper cells

B- cells converts to plasma cells

Production of antibodies

Symptoms



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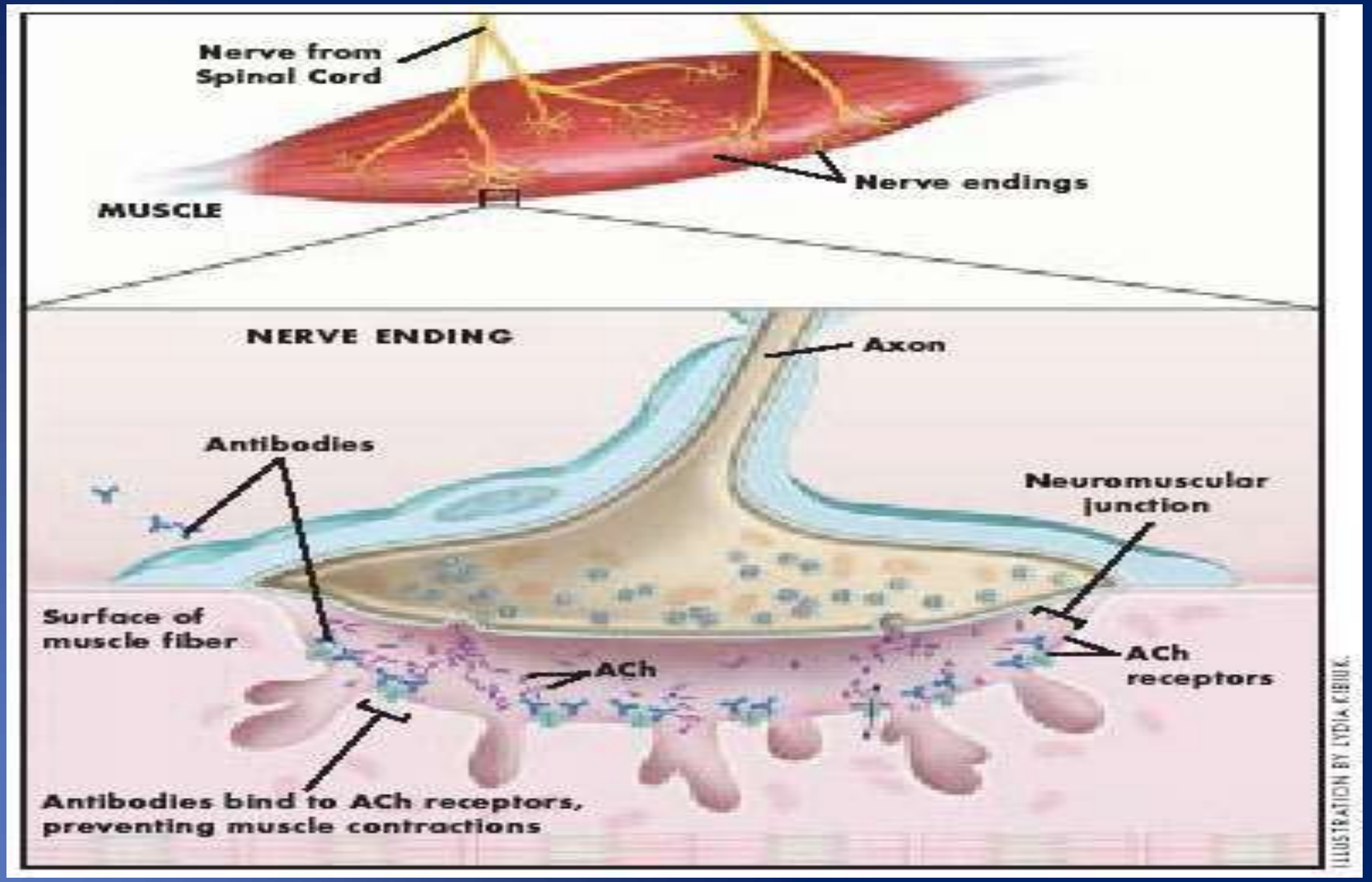

Nerve from Spinal Cord
Nerve endings
MUSCLE
NERVE ENDING
Axon
Antibodies
Neuromuscular junction
Surface of muscle fiber
ACh
ACh receptors
Antibodies bind to ACh receptors, preventing muscle contractions
ILLUSTRATION BY LYDIA KIBIUK

PATHOPHYSIOLOGY

- It is uncertain why ocular muscles are frequently involved in myasthenia and why the disease stays localized to the ocular muscles in 15 percent of cases.
- It has been proposed that subtle alterations in the function of extraocular muscles are more likely to produce symptoms than in limb muscles, and that the levator palpebrae, under constant activation during eye opening, may be more susceptible to fatigue.
- Patients with OMG are more likely to be seronegative for acetylcholine receptor antibodies than patients with GMG.
- The junctional folds of muscle endplates are sparse in the extraocular and levator muscles, perhaps producing a lower safety factor for neuromuscular transmission.
- Complement regulatory genes are expressed differently in extraocular muscles, perhaps reducing protective mechanisms to complement-mediated tissue injury.

CLINICAL FEATURES

- Characterized by ptosis and oculomotor paresis.
- Some patients also have mild orbicularis oculi weakness.
- This triad: Ptosis, Oculomotor paresis and Orbicularis oculi weakness should prompt an evaluation for MG.
- Signs and symptoms of OMG are characterized by fluctuating, fatigable weakness.
- Most patients note ocular symptoms that worsen as the day progresses or with tasks such as driving.
- Patients may report that they have mild or no symptoms upon wakening.
- The examination may elicit signs of fatigable levator and extraocular muscle weakness.

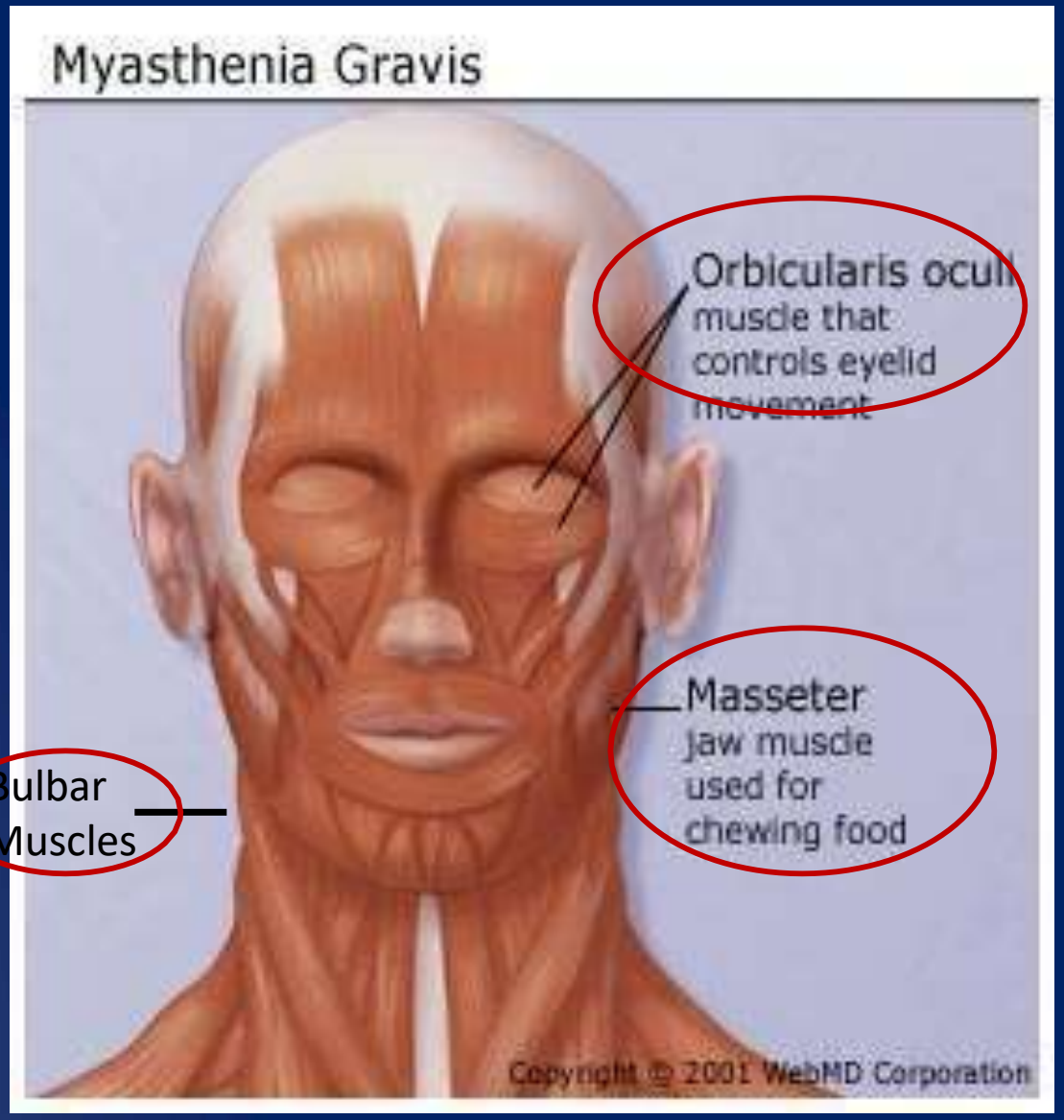
CLINICAL FEATURES

- Fluctuation weakness increasing trough the day & relieved by rest.
- Extra ocular muscle weakness
- Present in 50% of patients initially.
- Present in 90% of patients during the course of disease.
- Disease remains ocular in 16% of patients.

CLINICAL PRESENTATION

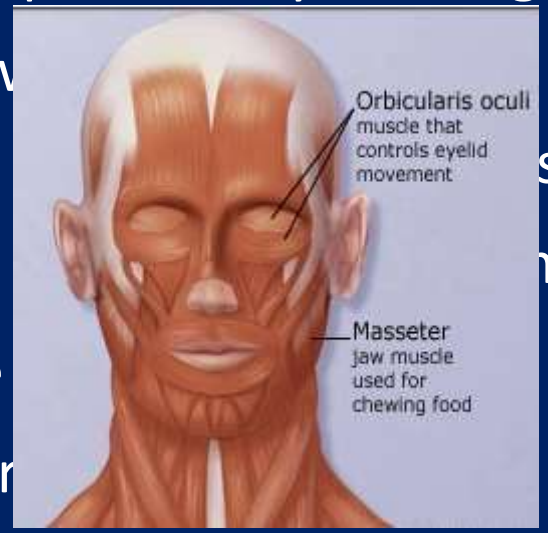
MUSCLE STRENGTH

- Ocular muscle weakness
- Facial muscle weakness
- Bulbar muscle weakness
- Limb muscle weakness
- Respiratory weakness



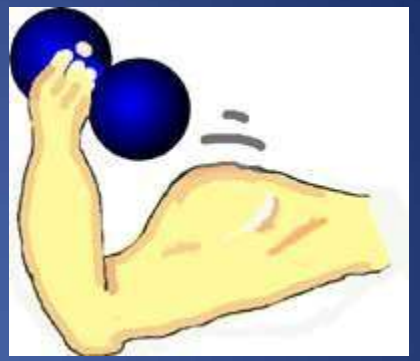
FACE AND THROAT MUSCLES

- symptoms first the gravis, myastheniawithpeopleof percent15About involve face and throat muscles, which can cause difficulties with:
- upon dependingnasal,soundor soft verybe mayspeechthe**speaking**. which muscles have been affected.
- **swallowing**. may choke very easily, which makes it difficult to eat, drink or take pills. in some cases, liquids may come out of the nose.
- out halfway through a**chewing**. the muscles used for chewing may wear meal, particularly if eating
- something hard to chev
- s.
- nay
- note "lost smile" if the
- facial expr

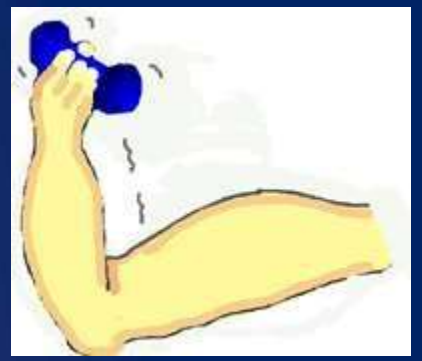


ARM AND LEG MUSCLES

- Myasthenia gravis can cause weakness in arms and legs, but this usually happens in conjunction with muscle weakness in other parts of the body – such as eyes, face or throat.
- The disorder usually affects arms more often than legs.
- If it affects legs, may waddle when walking.



Normal dumbbell



Weakness dumbbell

CAUSES

- Myasthenia gravis may be inherited as a rare, genetic disease, acquired by babies born to mothers with MG, or the disorder may develop spontaneously later in childhood.
- called chemicals,releasingby musclesthe withcommunicateNerves neurotransmitters, which fit precisely into receptor sites on the muscle cells.
- In myasthenia gravis, immune system produces antibodies that block or destroy many of the muscles' receptor sites for a neurotransmitter called acetylcholine.
- With fewer receptor sites available, muscles receive fewer nerve signals, resulting in weakness.

FACTORS WORSENING MG

- Fatigue
- Illness
- Stress
- Extreme heat
- Medications such– blockers, channelcalciumblockers,betaas quinine and some antibiotics

COMPLICATIONS

- **Myasthenic crisis**: A life-threatening condition, which occurs when the muscles that control breathing become too weak to do their jobs. Emergency treatment is needed to provide mechanical assistance with breathing. Medications and blood-filtering therapies help people recover from myasthenic crisis, so they can again breathe on their own.
- **Thymus tumors**: About 15 percent of the people who have myasthenia gravis have a tumor in their thymus, a gland under the breastbone that is involved with the immune system. Most of these tumors are noncancerous.

OTHER DISORDERS

- **Underactive or overactive thyroid**. The thyroid gland, located in the neck, secretes hormones that regulate metabolism. If thyroid is underactive, body uses energy more slowly. An overactive thyroid makes body use energy too quickly.
- **Lupus**. Disease of immune system. Common symptoms include painful or swollen joints, hair loss, extreme fatigue and a red rash on the face.
- **Rheumatoid arthritis**. Caused by problems with immune system. It is most conspicuous in the wrists and fingers, and can result in joint deformities that make it difficult to use hands.

OSSERMAN Classification

Class I	1.	Any ocular muscle weakness
Class II	.2	Mild weakness other than ocular
II a		Predominantly limb, axial, or both
II b		Predominantly orpharyngeal/respiratory
Class III	.3	Moderate weakness other than ocular
III a		Predominantly limb, axial, or both
III b		Predominantly orpharyngeal/respiratory
Class IV	.4	Severe weakness other than ocular
IV a		Predominantly limb, axial, or both
IV b		Predominantly orpharyngeal/respiratory
Class V	5.	Intubation with/without ventilation

CLINICAL FEATURES

- Prevalence: 1-7 in 10,000
- Affect all age groups
- Usual age at onset: BIMODAL PEAK
- Overall 3:2 f:m ratio
- Familial occurrence is known, but rare
- More common is a family history of one or the other autoimmune diseases, and suggests partial genetic predisposition
- Reports of the concurrence of myasthenia and MULTIPLE SCLEROSIS

PROGRESSION OF DISEASE

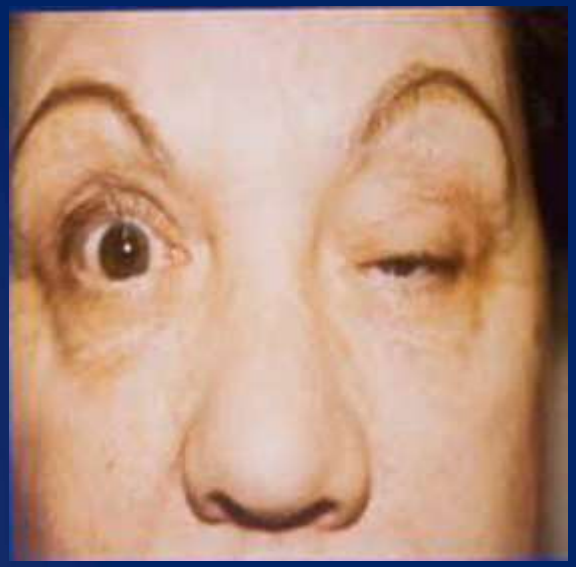
Mild to severe…..over weeks to months

- Spreads from **ocular** to **facial** to **bulbar** to **truncal and limb** muscles
- Symptoms may remain limited to EOM and eyelid muscles for years
- The disease remains ocular in 16% of patients

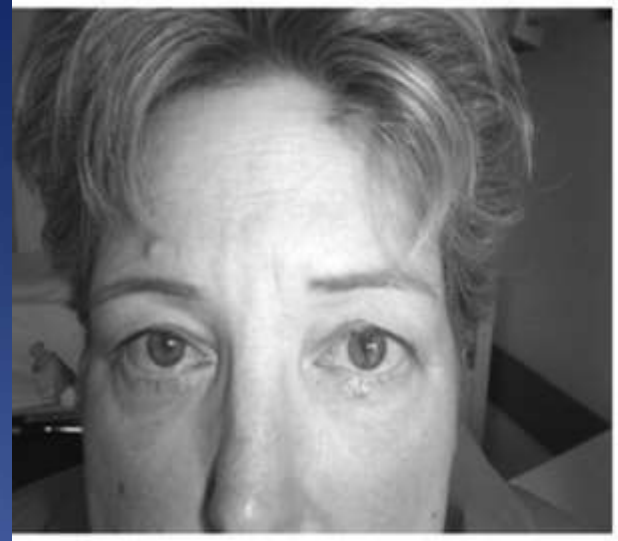
PTOSIS

Ptosis is often unilateral or asymmetric on presentation.

- A historical pattern of ptosis alternating from one side to the other is nearly always a sign of OMG.
- Old photos may help to determine if ptosis is new or longstanding.
- Measurements of the eyelid position and levator palpebrae muscle function using standard methods help to identify and quantify weakness as well as fluctuations associated with fatigue.



Myasthenia – Fatigue and Recovery Test 'Simpson plus'



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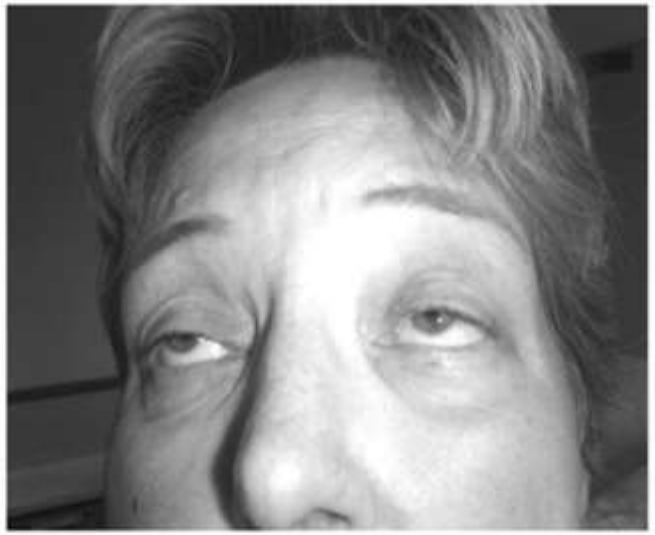
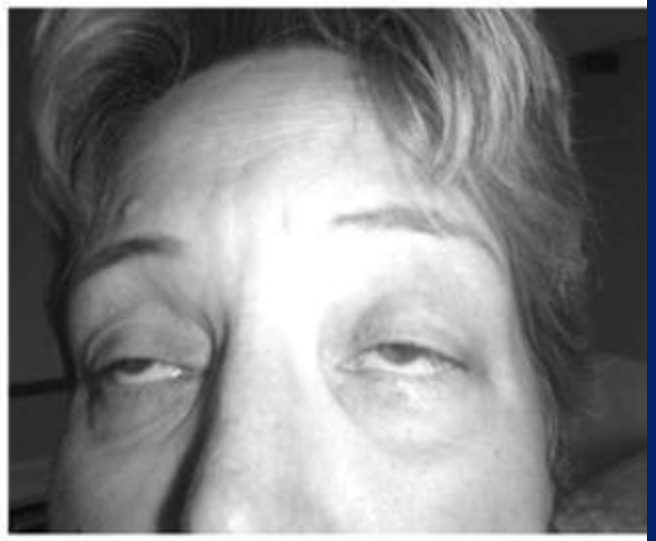
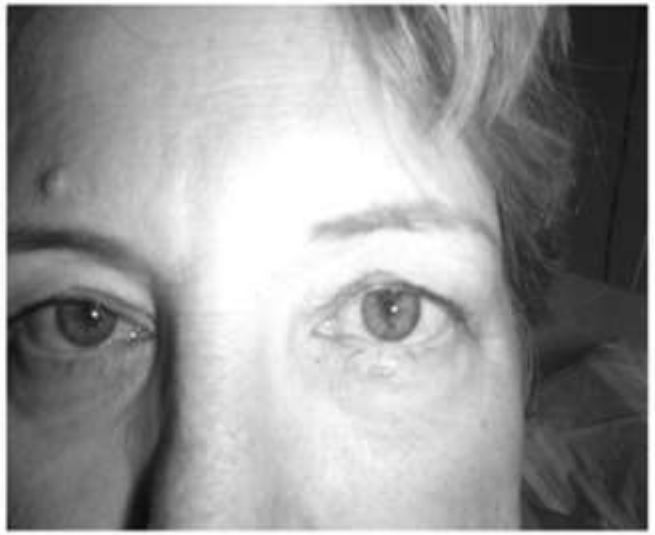


+ 10 sec upward gaze (Simpson)

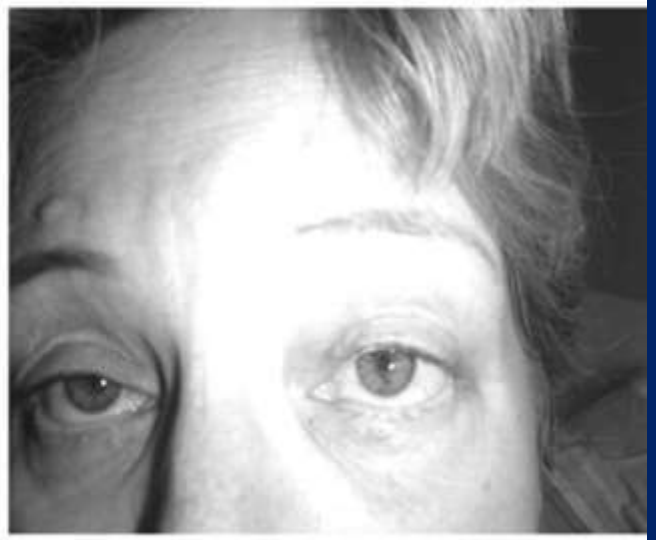


+ 30 sec upward gaze (Simpson)

Maximal lid closure 10 sec



Lid open



+ 10 sec upward gaze

EXAMINATION FOR EYELID FATIGUE

Ptosis fluctuates throughout the examination, evidence for fatigability of the levator muscle is present.

Evidence for fatigue can also be elicited:

- Prolonged activation of the levator palpebrae muscle is produced with sustained upgaze for 1 to 2 minutes. Enhancement of ptosis either during prolonged upgaze or upon return to primary gaze suggests fatigability.
- Rapid fatigue after rest is another elicitable sign. This may be observed in the eyelid as a ***Cogan's lid twitch***. The patient is asked to sustain downgaze briefly and then make a fast eye movement (or saccade) to primary gaze. An affected eyelid will quickly rise and then fall such that the lid appears to twitch.
- Eyelid "curtaining" occurs when the more ptotic eyelid is passively lifted above the iris by the examiner and the contralateral eyelid becomes more ptotic by slowly drooping or "curtaining." This phenomenon results because of equal innervation to both levator palpebrae superioris muscles (Hering's law).
- A "rest test" looks for improvement in baseline ptosis after having the patient gently close his eyes for two to five minutes.
- While each of these signs suggests fatigable levator function, none are specific for OMG.

DIPLOPIA

- Binocular diplopia is a prominent feature of OMG when ophthalmoparesis is present.
- At presentation, OMG may involve individual or multiple, bilateral ocular motor muscles.
- For patients with medial or lateral recti muscle involvement, diplopia will be binocular and horizontal.
- If the superior or inferior recti or the oblique muscles are involved, then there will be a vertical or diagonal component to the diplopia.
- Virtually every ocular motility disorder, including those associated with isolated nerve palsies and even central nervous system brainstem pathways, such as internuclear ophthalmoplegia, has been described in OMG.
- Fluctuation in either the degree of diplopia or in the direction of gaze that elicits the diplopia suggests fatigable ocular motor paresis.

OTHER FINDINGS

- Lagophthalmus, failure of the eye to fully close, is rare in OMG. However, some weakness of the orbicularis oculi is often demonstrable when the examiner attempts to open the eyes against forced eyelid closure, nearly impossible in the intact patient.
- The "peek-a-boo" sign of lagophthalmus after prolonged forced eyelid closure, suggests fatigue in this muscle.
- Pupillary involvement is generally not seen in OMG.

RESPIRATORY SYMPTOMS

- Weakness of intercostal muscles and diaphragm may lead to CO_2 retention
- Weakness of pharyngeal muscles may collapse the airway.
- O_2 saturation can be normal while CO_2 is retained. So, pulse oximetry is not reliable to detect the amount of paralysis.

OTHER SYMPTOMS

- Palatal muscle weakness
- Nasal voice
- Nasal regurgitation
- Swallowing may be difficult & regurgitation of foods can occur.
- Coughing & choking while drinking.
- Limb weakness can also be present
- Initially proximal but may follow distal muscles also.

TYPES OF MYASTHENIA GRAVIS

- **NEONATAL**: In 12% of the pregnancies with a mother with MG, she passes the antibodies to the infant through the placenta, causing neonatal myasthenia gravis. The symptoms will start in the first two days and disappear within a few weeks after birth. With the mother, it is not uncommon for the symptoms to even improve during pregnancy, but they might worsen after labor.
- **CONGENITAL**: Children of a healthy mother can, very rarely, develop myasthenic symptoms beginning at birth, congenital myasthenic syndrome or CMS. Other than myasthenia gravis, CMS is not caused by an autoimmune process, but due to synaptic malformation, which in turn is caused by genetic mutations. Thus, CMS is a hereditary disease. More than 11 different mutations have been identified, and the inheritance pattern is typically autosomal recessive.
- **JUVENILE**: myasthenia occurring in childhood, but after the peripartum period

DIFFERENTIAL DIAGNOSIS

- Conditions that cause bilateral impairment of both eyelid and oculomotor function are most likely to be considered in the differential diagnosis of OMG.
  - Thyroid ophthalmopathy
  - Chronic progressive external ophthalmoplegia
  - Muscular dystrophy
  - Brainstem and motor cranial nerve pathology

THYROID OPHTHALMOPATHY

- Grave's disease produces abnormal eye movements due to a constrictive ophthalmopathy.
- It can usually be differentiated from MG by the lack of ptosis and the presence of proptosis, lid retraction, lid lag, and periorbital edema.
- Restricted eye movement in Grave's ophthalmopathy can be confirmed by forced duction testing, and the enlarged extraocular muscles can usually be seen on CT images of the orbits.
- However, thyroid disease can coexist with MG.
- As a result, screening thyroid function studies are usually a good idea before treatment is instituted, even when the diagnosis of MG is clear.

CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA

- Chronic progressive external ophthalmoplegia (CPEO) and Kearns-Sayre syndrome (KSS) are mitochondrial disorders that produce progressive, generally symmetric, ophthalmoparesis and ptosis.
- These patients frequently have slow saccades, an early sign that may suggest CPEO rather than MG, where saccades are normal.
- CPEO generally remains isolated to the extraocular muscles, but KSS may ultimately produce generalized muscle weakness, cerebellar disease, and retinal degeneration, distinguishing this disorder from OMG.
- The majority of patients with KSS present with ptosis months to years before the onset of ophthalmoparesis.
- Patients with CPEO and KSS typically do not complain of diplopia. This is presumed to be due to the symmetric involvement of the ocular motility disturbance.

- **Muscular dystrophy** — Myotonic dystrophy and oculopharyngeal dystrophy may produce ptosis and ophthalmoparesis.

- ***Myotonic dystrophy*** is an autosomal dominant disorder characterized by variable ptosis and weakness of the face, jaw, and neck as well as weakness of the extremities.

- Associated abnormalities include cataracts, cardiac conduction defects, characteristic faces (long face with atrophy of temporalis and masseter), frontal balding, and variable intellectual impairment.

- Patients may also develop dysphagia and ophthalmoparesis in some cases.

- ***Oculopharyngeal dystrophy*** is muscular dystrophy characterized by slowly progressive ptosis and dysphagia with onset in the fourth or fifth decade of life.

- Weakness of the proximal muscles is commonly present upon initial presentation, but ophthalmoparesis typically develops after the onset of ptosis and dysphagia.

BRAINSTEM AND MOTOR CRANIAL NERVE PATHOLOGY

- Structural disease of the brainstem can cause isolated ocular symptoms. As examples, parasellar tumors and aneurysms can impair function of the third, fourth, and sixth cranial nerves, leading to symptoms similar to ocular myasthenia.
- The presence of trigeminal dysfunction and/or pupillary abnormalities is inconsistent with OMG and may point to a structural lesion. Brain MRI to exclude these disorders is warranted in unconfirmed cases of OMG.
- Multiple motor cranial neuropathies, such as those produced by carcinomatous or lymphomatous meningitis, may also produce eye movement abnormalities that may be confused with MG.
- If the diagnosis of MG is not firmly established in cases of possible multiple cranial nerve abnormalities, examination of the cerebrospinal fluid for abnormal cells and cytology is usually necessary.

DIAGNOSTIC TESTS

- The diagnosis of ocular myasthenia gravis (OMG) can often be made on a clinical basis when the history and examination findings are classic.
- However, confirmation by diagnostic testing is usually desired. The sensitivity and specificity of tests are different for generalized versus OMG

ICE PACK TEST

- The ice pack test can be used in patients with ptosis, particularly those in whom the Tensilon test is considered too risky.
- It is not helpful for those with extraocular muscle weakness. It is based on the physiologic principle of improved neuromuscular transmission at lower muscle temperatures.
- In the ice pack test, a bag is filled with ice and placed on the closed lid for one minute.
- The ice is then removed and the extent of ptosis is assessed immediately; the duration of improvement is short (less than one minute). The sensitivity appears to be about 80 percent in those with prominent ptosis.

TENSILON TEST

- Edrophonium chloride (Tensilon) inhibits acetyl cholinesterase and can transiently reverse signs of weakness due to OMG, such as ptosis and extraocular muscle paresis.
- The test is positive if there is significant improvement in ptosis or ophthalmoparesis.
- The sensitivity of the Tensilon test for OMG is similar to that for GMG, 85 to 95 percent, but it is associated with false-negative and false-positive results.

SERUM ANTIBODY STUDIES

- While more than 85 percent of patients with generalized myasthenia gravis (GMG) have serum antibodies against the acetylcholine receptor, the sensitivity of AChR-Ab testing in OMG may be as low as 45 to 60 percent.
- This is the most specific test for MG; no false positives have been reported.
- Some patients with GMG who are seronegative for AChR-Ab have antibodies against the muscle-specific tyrosine kinase (MuSK). Once thought to be unassociated with OMG, MuSK antibodies have been detected in patients with OMG in a few case reports.
- A number of reports have linked LRP4 antibodies with GMG.
- In three reported cases of OMG, patients who were found to be seronegative for both AchR-ab and MuSK, were found to have positive LRP4 antibodies.

ELECTROPHYSIOLOGY

- Electrodiagnostic studies are an important supplement to the immunologic studies and may also provide confirmation of the diagnosis of myasthenia.
- **Repetitive nerve stimulation** — Repetitive nerve stimulation (RNS) studies demonstrate ***decrement in the amplitude*** of the compound muscle action potential after repetitive stimulation of the motor nerve to that muscle.
- The ***orbicularis oculi*** may be studied in patients with OMG.
- Among patients with GMG, the sensitivity of RNS is greater than 70 percent. In contrast, among patients with OMG, the sensitivity has been as low as 15 percent in some series and only slightly higher. The specificity is 89 percent; both false negatives and false positives do occur.

SINGLE-FIBER EMG

- Single-fiber electromyography (SFEMG) identifies abnormal neuromuscular transmission by measuring temporal variability in the firing of adjacent motor nerve fibers from a single motor unit, a phenomenon called "jitter.“
- SF-EMG is more sensitive than repetitive nerve stimulation and may identify electrophysiologic abnormalities in clinically strong muscles.
- Depending on which and how many muscles are examined, the sensitivity of SFEMG for OMG is between 63 to 100 percent.
- Evaluation of the orbicularis oculi and the superior rectus levator palpebrae complex increase the sensitivity in patients with OMG to over 95 percent.
- However, this testing, particularly with examination of the superior rectus muscle, is available only at specialized centers with neuromuscular expertise.

DIAGNOSTIC EVALUATION

- The diagnosis of ocular myasthenia gravis (OMG) can usually be made in a patient with a typical history and examination findings and a positive result on ice or Tensilon test.
- Further confirmation should be obtained by a positive acetylcholine receptor antibody titer (AChR-Ab).
- If this is negative (as it will be in about half of patients with OMG), then EMG with repetitive nerve stimulation (RNS) or single-fiber electromyography (SFEMG) should be performed.
- If the titer is positive, then electrophysiologic testing is generally unnecessary.
- If the diagnosis remains unclear after EMG, then a brain MRI and lumbar puncture (LP) with spinal fluid examination may be needed to exclude treatable inflammatory or structural disease of the brainstem or cranial nerve roots.
- Patients with suspected OMG should also have a chest CT scan to rule out thymoma and thyroid function tests to rule out associated thyroid dysfunction.

PROGNOSIS

- Patients presenting with ocular myasthenia gravis (OMG), two-thirds will go on to develop signs and symptoms of extremity weakness and other bulbar muscle weakness, while one-third will have pure OMG.
- Most (78 percent) of those who will develop generalized MG (GMG) do so within the first year, and virtually all (94 percent) will do so by three years.
- Neither age nor sex has been consistently shown to alter the course of disease.

TREATMENT

- Treatment considerations in the management of ocular myasthenia gravis (OMG) include
  - symptomatic and immunomodulatory treatment of myasthenia,
  - thymectomy,
  - and corrective treatments of ptosis and strabismus.

SYMPTOMATIC MANAGEMENT

- Assistive devices (ptosis crutches and lid adhesive devices) can aid in the treatment of ptosis pending a response to acetyl cholinesterase inhibitors or immunosuppressive therapy.
- Use of lubricating drops and periods of abstinence from device use are critical to prevent corneal dryness and exposure keratopathy.
- An eye patch, opaque contact lens, or occlusion of an eyeglass lens are simple ways to eliminate diplopia.
- A patch that is concave (to avoid injury to the cornea) can be worn over either eye; it is not necessary to alternate the patch between eyes in adults.
- These interventions have the disadvantage of eliminating depth perception, which usually requires unoccluded vision in two eyes.
- Some patients require stereoscopic vision in their occupation.
- Other activities of daily living, such as driving and watching television, are also impaired with a loss of depth perception.

- **Prism lenses** offer another non pharmacologic alternative for patients with diplopia.
- If the ophthalmoparesis is stable over several weeks or months, prism lenses may minimize diplopia.
- However, fatigability and significant variability of eye movements will limit the benefit of prisms in patients with OMG until stabilization occurs.
- A disadvantage is that they can distort vision when the amount of prism needed is significant.

ANTICHOLINESTERASE AGENTS

- Pyridostigmine (Mestinon) is the most commonly used anticholinesterase agent for symptomatic treatment of myasthenia.
- Unfortunately pyridostigmine treatment alone rarely results in resolution of ocular symptoms, particularly diplopia.
- It is best used for very mild cases of OMG or as adjunctive symptomatic treatment for moderate or severe OMG.
- The dosing regimen and side-effect profile for pyridostigmine is the same for patients with GMG and OMG.

IMMUNOSUPPRESSIVE AGENTS

- Some clinicians prefer to reserve immunosuppressive treatment for patients with more severe disease than that manifested by pure OMG.
- However, in order to achieve resolution of ptosis or ophthalmoparesis, patients with OMG usually require immunosuppression. Many patients are not satisfied with eye patching, prism lenses, or non pharmacologic therapy despite the known risks.

Plasmapheresis and intravenous immune globulin are used for the short-term management of severe GMG and have no role in patients with OMG.

STEROID SPARING AGENTS

are second-line immune Azathioprine, mycophenolate, mofetil, and cyclosporine suppressants for patients who do not respond to or tolerate prednisone. These agents can also be used with prednisone in order to taper or wean off of prednisone. The side effects of these medications often limit their use for patients with OMG, but they may be necessary to control symptoms and may be preferred by patients if they do not experience major side effects.

BEHAVIORAL MODIFICATIONS

- Diet
  - Thickened liquids are preferred, when dysphagia arises to counteract the fear of aspiration.
  - Asparagus should be taken as it contains steroid-like substance.
- Activity
  - Patients should be as active as possible but should take rest in between.
  - Yoga exercises to stretch the weakened muscles should be done.
  - This not only strengthens the muscles but also provides oxygen & removes carbon dioxide from them.

SURGERY FOR PTOSIS AND DIPLOPIA

- Surgical correction can be performed on patients with stable ptosis.
- Recurrence of ptosis for patients who do not demonstrate stability prior to surgery is common due to the nature of myasthenia.
- The optimal duration of stability prior to surgery is unknown, although there are some indications that three to four years of stability prior to ptosis repair is appropriate.
- Extraocular muscle resection and recession can be performed on patients with stable ophthalmoparesis.
- Similar to the situation for ptosis, recurrence of diplopia for patients who do not demonstrate stability prior to surgery is common.
- The optimal duration of stability prior to surgery is unknown; shorter durations of stability (five to six months) prior to strabismus surgery are advocated compared with ptosis surgery.

SUMMARY AND RECOMMENDATIONS

- Ocular myasthenia gravis (OMG) refers to MG in which the signs and symptoms are confined to the ocular muscles.
- The diagnosis of OMG is strongly suggested by the triad of ophthalmoparesis, ptosis, and orbicularis oculi weakness.
- However, isolated ptosis and ophthalmoparesis frequently occur.
- The demonstration of fatigue in the ocular muscles is a helpful but not infallible diagnostic sign for OMG.
- Clinicians should consider thyroid disease, muscle disease, and other brainstem and cranial nerve lesions in the differential diagnosis of OMG.
- The diagnosis of OMG can usually be made in a patient with a typical history and examination findings and a positive result on ice or Tensilon test.
- If this is negative, then an electrophysiologic test should be performed.
- If the diagnosis remains unclear, then a brain MRI and lumbar puncture (LP) should be performed to rule out other diagnoses.
- Patients with suspected diagnosis of OMG should have thyroid function tests and a chest CT to exclude thymoma.

- Two-thirds of patients with OMG will go on to develop signs and symptoms of extremity weakness and other bulbar muscle weakness.
- Most of those who will develop generalized MG (GMG) do so within the first year, and virtually all will do so by three years.
- Use anticholinesterase agents as a first treatment in OMG. Most patients have some benefit from anticholinesterase agents, although many will not fully respond.
- For patients who continue to have disabling symptoms despite treatment with an anticholinesterase agent, immunosuppressive therapy is suggested.
- Prednisone is most commonly used. In deciding to use prednisone, clinicians and patients must balance the severity of symptoms and the efficacy of nonpharmacologic measures, with the difficulty in weaning prednisone in some patients and side effects.
- Patients with stable diplopia or ptosis despite maximal therapy may benefit from surgical interventions. A period of stability of several months to a few years is suggested prior to surgical intervention.

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THANK YOU

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