A motor neuron is a neuron whose cell body is located in the motor cortex, brainstem or the spinal cord, and whose axon (fiber) projects to the spinal cord or outside of the spinal cord to directly or indirectly control effector organs, mainly muscles and glands. There are two types of motor neuron – upper motor neurons and lower motor neurons. The nerves that send messages between the cerebral cortex and the spine are called _______ motor neurons, and those that relay messages from the spine to the muscles are called _______ motor neurons.

A. Upper, Lower  
B. Lower, Upper  
C. Left, Right  
D. Right, Left

Which of the following motor neuron diseases typically causes both upper and lower motor neuron signs?

A. Primary lateral sclerosis (PLS)  
B. Spinal muscle atrophy II  
C. Poliomyelitis  
D. Amyotrophic lateral sclerosis (ALS)

- **Amyotrophic lateral sclerosis (ALS):** Weakness may be due to upper or lower motor neuron loss.  
- **Upper motor neuron (UMN) signs** include weakness, spasticity, hyperreflexia, and upgoing plantar response.  
- **Lower motor neuron (LMN) signs** include weakness, atrophy, flaccidity, hyporeflexia, and fasciculations.  
- **ALS patients with UMN pathology** will often have a loss of dexterity or feeling of stiffness in their limbs. Spasticity may further exacerbate weakness and loss of function. This is due to the involvement of the vestibulospinal and reticulospinal tracts.  
- **LMN symptoms in the ALS population** include muscle weakness, with some muscle fasciculations, atrophy, and muscle cramping. Cramping of abdominal or other trunk muscles should prompt a clinician to strongly consider ALS as a possible diagnosis.  
- **Primary lateral sclerosis (PLS)** is classified as an UMN lesion. Spinal muscle atrophy II (SMA II) and poliomyelitis are classified as LMN lesions.

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**FAMOUS PEOPLE WITH THE DISEASE**

<table>
<thead>
<tr>
<th>Lou Gehrig</th>
<th>Stephen Hawking</th>
<th>Mary Valastro Pinto</th>
</tr>
</thead>
<tbody>
<tr>
<td>(June 19, 1903–June 2, 1941) Baseball player in 1920s and ’30s. Holds record for most career grand slam home runs (23). Gehrig focused national attention on ALS in a famous speech at Yankee Stadium in 1939. Gehrig at Yankee Stadium during his farewell speech.</td>
<td>(Jan. 8, 1942–present) Internationally renowned physicist who is almost completely paralyzed by ALS. Hawking’s illness has progressed more slowly than typical cases of ALS.</td>
<td>(April 17, 1948–present) Mother of “Cake Boss” reality TV show star Buddy Valastro. In the season finale last month, viewers learned Mary Valastro, a regular in her son’s bakery, had been diagnosed with ALS.</td>
</tr>
</tbody>
</table>

Stephen Hawking (Died Mar 14, 2018) / Mary Valastro Pinto (Died June 22, 2017)  
Lou Gehrig’s disease (US) / Motor Neuron Disease (UK)
Which of the following is the most common presenting form of motor neuron disease in adults?

A. Amyotrophic lateral sclerosis (ALS)
B. Poliomyelitis
C. Spinal muscular atrophy (SMA)
D. Primary lateral sclerosis (PLS)

- ALS is also called Lou Gehrig’s disease, as it was named after the New York Yankees’ first baseman who passed away from this disorder.
- This is unfortunately still the most widely known motor neuron disease and is the most common presenting form.
- The incidence of ALS is approximately 1.6 to 2.4 cases per 100,000 population.

<table>
<thead>
<tr>
<th>SLE (Systemic lupus erythematosus)</th>
<th>ALS (Amyotrophic lateral sclerosis)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lupus an autoimmune disease</td>
<td>Lou Gehrig’s disease a progressive nervous system disease that affects nerve cells in the brain and spinal cord, causing loss of muscle control. ALS is often called Lou Gehrig’s disease, after the baseball player who was diagnosed with it.</td>
</tr>
</tbody>
</table>

Which of the following is a motor neuron disease that causes the death of neurons controlling voluntary muscles?

A. Systemic lupus erythematosus (SLE)
B. Amyotrophic lateral sclerosis (ALS)

The Ice Bucket Challenge was an activity involving the dumping of a bucket of ice water over a person’s head, either by another person or self-administered, to promote awareness of the disease ___________ (also known as motor neuron disease) and encourage donations to research.

A. Multiple sclerosis (MS)
B. Systemic lupus erythematosus (SLE)
C. Amyotrophic lateral sclerosis (ALS)

<table>
<thead>
<tr>
<th>UMN (upper motor neuron) signs</th>
<th>LMN (lower motor neuron) signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>weakness, spasticity, hyperreflexia, upgoing plantar response</td>
<td>weakness, atrophy, flaccidity, hyporeflexia, fasciculations</td>
</tr>
</tbody>
</table>

Young children have a reflex called the __________ reflex. If you stroke the bottom of their foot, their big toe will pull back and their other toes will spread out. This reflex usually disappears after age 2. In adults, the __________ response is a sign of damage to the upper motor neuron.

A. Asymmetric tonic neck reflex (ATNR)
B. Symmetrical tonic neck reflex (STNR)
C. Sucking
D. Babinski

<table>
<thead>
<tr>
<th>Normal plantar response</th>
<th>Extensor plantar response (Babinski sign)</th>
</tr>
</thead>
</table>
Amyotrophic lateral sclerosis (ALS):

- Most common form of motor neuron disease that causes respiratory failure. Respiratory failure usually develops late in the disease and is the most common cause of death.
- Respiratory muscle weakness causes restrictive ventilation and impaired cough. If symptoms begin with limb weakness, the disorder may progress to respiratory failure in 2 to 5 years.
- The American Academy of Neurology recommends measuring FVC (forced vital capacity) at diagnosis and every three months thereafter to monitor disease progression.
- FVC below 50% of predicted suggests the risk of imminent respiratory failure and the need for ventilatory support. FVC may be falsely low with bulbar weakness due to inability to form a tight mouth seal during spirometry. Conversely, due to preserved lung volume, FVC may remain normal until there is substantial muscle weakness.
- Further testing can be done by measuring transdiaphragmatic pressure (Pdi) which is considered to be the gold standard measure of diaphragmatic strength. It is obtained by measuring gastric and esophageal pressures with gastric and esophageal balloon catheters. Pdi is the gastric pressure minus esophageal pressure.

Which of the following is considered a poor prognostic factor in patients with amyotrophic lateral sclerosis (ALS)?

A. Predominance of upper motor neuron (UMN) findings at diagnosis
B. Pulmonary dysfunction early in the clinical course
C. Long period from symptom onset to diagnosis
D. Younger age of onset

- Poor prognostic factors include predominance of lower motor neuron (LMN) findings at diagnosis, short period from symptom onset to diagnosis, and older age at time of onset.
- Bulbar and pulmonary dysfunction early in the disease course is also a poor prognostic factor.
- Women typically present with bulbar symptoms, as compared with men.
- It is important to note that electrodiagnostic indicators of poor prognostic indicators include profuse spontaneous fibrillations, positive sharp waves, and low-amplitude compound muscle action potential.

What is the prognosis for patients with amyotrophic lateral sclerosis?

A. 50% die within 3 years
B. 100% die within 3 years
C. 50% live up to 7 years
D. 50% live up to 10 years

- The overall median 50% survival rate is 2.5 years after diagnosis.
- Survival rate is largely dependent on a patient’s decision to use mechanical ventilation and/or a feeding tube, but the 5-year survival rate is between 4% and 30%. Around 10% will live for 10 years.

Which of the following pharmacologic agents is a first-line treatment for spasticity in patients with amyotrophic lateral sclerosis (ALS)?

A. Tizanidine
B. Dantrolene
C. Baclofen
D. Benzodiazepine

- Spasticity is a condition in which muscles stiffen or tighten, preventing normal fluid movement. The muscles remain contracted and resist being stretched, thus affecting movement, speech and gait.
- Baclofen is a GABA analogue used to facilitate motor neuron inhibition at spinal levels and is the first-line treatment. Dosing can be started at 5 to 10 mg two to three times per day. It can be titrated up to 20 mg four times per day.
- Potential side effects include weakness, fatigue, and sedation. Patients must be informed that abrupt discontinuation of baclofen may cause withdrawal seizures.
- Tizanidine is an alpha-2 agonist. Benzodiazepine can be helpful, but can cause respiratory depression and somnolence. Dantrolene blocks calcium release in the sarcoplasmic reticulum and is ineffective at reducing muscle tone, but can cause generalized muscle weakness.
A 60-year-old man without any significant past medical history presents to your outpatient office with asymmetric atrophy, weakness, and fasciculations. He also complains of some difficulty swallowing his meals and complains of a strained and strangled quality in his speech. He describes normal bowel and bladder function. Which of the following is most likely his diagnosis?

- **A. Poliomyelitis**
- **B. Spinal muscle atrophy**
- **C. Primary lateral sclerosis (PLS)**
- **D. Amyotrophic lateral sclerosis (ALS)**

- **ALS** most commonly affects people in the age group of 40 to 60 years, and the mean age of onset is around 60 years.
- **Onset** is usually insidious and painless. Asymmetric weakness is the most common presentation.
- **Dysphagia** (oral, pharyngeal), dysarthria, drooling, and aspiration can occur and are signs and symptoms representing bulbar muscular weakness. **This bulbar weakness** — named for the nerves that originate from the bulblike part of the brainstem — can cause difficulty with talking (dysarthria), chewing, swallowing (dysphagia), and holding up the head.
- **Also**, strained, strangled quality of speech, reduced rate, and low pitch indicate a spastic dysarthria.
- **Bowel** and bladder function is typically spared in ALS.

Which of the following pharmacologic agents has been approved for patients with amyotrophic lateral sclerosis (ALS) to slow the progression and improve survival?

- **A. Levodopa**
- **B. Riluzole**
- **C. Baclofen**
- **D. Rebif**

- This is an antiglutamate agent that may be effective in slowing the disease, prolonging ventilator time, and may improve survival in patients with bulbar onset disease. However, side effects can include asthenia, and the medication is expensive.
- **Nonpharmacological** management of ALS includes rehabilitation, preventing contractures, submaximal exercise, tracheostomy, and respiratory therapy.
- **Rebif** is a beta interferon that is used to modify the course in multiple sclerosis patients. Baclofen is a derivative of gamma-aminobutyric acid (GABA) and is primarily used to treat spasticity. Levodopa is a medicine used to control symptoms of Parkinson’s disease. Levodopa does not slow the disease process, but it improves muscle movement and delays severe disability.

**TUESDAYS WITH MORRIE** is a memoir by American author Mitch Albom about a series of visits Albom made to his former sociology professor Morrie Schwartz, as Schwartz gradually dies of ALS. The book topped the New York Times Non-Fiction Bestsellers of 2000.

**Synopsis:** Albom is a successful sports columnist for the Detroit Free Press. After seeing his former sociology professor Morrie Schwartz appear on Nightline, Albom phones Schwartz, and is prompted to travel to Massachusetts to visit him. An ensuing newspaper strike allows Albom to visit Schwartz every week, on Tuesdays. The book recounts each of the fourteen visits Albom made to Schwartz, supplemented with Schwartz’s lectures, life experiences, and interspersed with both flashbacks and allusions to contemporary events. After being diagnosed with **Amyotrophic lateral sclerosis (ALS)**, Morrie’s final days are spent giving his former student Mitch his final lesson of life. The novel is divided into 14 different “days” that Mitch Albom spent with his professor Morrie. Throughout these days, Mitch and Morrie discuss various topics important to life and living. The novel also recounts Mitch’s memories of Morrie as a professor. - Source: Wikipedia

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**AMYOTROPHIC LATERAL SCLEROSIS (ALS/LOU GEHRIG’S DISEASE)**

| LV Blood & KD Yin deficiency | Hu Qian Wan + Zuo Gui Wan |
| SP & KD Yang deficiency | You Gui Wan + Si Jun Zi Tang |
| Qi & Yin deficiency | Zuo Gui Wan |
| Yin & Yang deficiency | Jin Gui Shen Qi Wan |
| Damp-Heat damaging the sinew | San Miao San |
| Blood stasis in network vessels | Xue Fu Zhu Yu Tang |
| Phlegm turbidity | Wen Dan Tang |

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| LV Blood & KD Yin deficiency | Hu Qian Wan + Zuo Gui Wan |
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| Yin & Yang deficiency | Jin Gui Shen Qi Wan |
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Diseases that affect the neuromuscular junction cause fluctuating muscle weakness. Certain groups of muscles are characteristically involved in each disease. The most common disorder of neuromuscular transmission is myasthenia gravis (MG). Although they are distinct from and much less common than MG, it is important for neurologists to know how to recognize and treat Lambert-Eaton myasthenic syndrome (LEMS) and botulism.

**Neuromuscular Junction Disorders**

- **Neuromuscular Junction**
  - Electrical-chemical-electrical link b/w nerve-muscle
- **Key Disorders**
  - Myasthenia Gravis
  - Lambert-Eaton Myasthenic Syndrome
  - Presynaptic VGCC Ab
  - Botulinum Toxin
  - Presynaptic VGKC Ab
  - Neuromyotonia
- **Myasthenia Gravis**
  - **Laboratory Testing**
    - AchR Ab ~ 85% Generalized MG
    - MuSK Ab ~ 40% of Non-AchR Ab
  - **Treatment**
    - Symptomatic: Pyridostigmine
    - Immune suppression: Steroids, azathioprine, etc.
    - Rescue: Plasmapheresis and IVIG

- **Lambert-Eaton M.S.**
- **Botulinum Toxicity**
  - **Neuromuscle Junction**
    - Oculobulbar & Facial
    - Facial & EOM weakness
    - Dysarthria/Dysphagia
  - Flaccid paralysis
  - Follows oculobulbar weakness
  - Proximal > Distal
  - Respiratory arrest
  - Diaphragm failure
  - **Autoimmunity (ACh block)**
    - Constipation
    - Mydriasis
    - Hypohidrosis

**Match the Neuromuscular junction disorders to their correct etiology.**

<table>
<thead>
<tr>
<th>Botulism</th>
<th>LEMS</th>
<th>MG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clostridium botulinum toxins inhibit pre-synaptic Ach release</td>
<td>Autoimmune response against Pre-synaptic calcium channel</td>
<td>Autoimmune response against Post-synaptic Ach receptors</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Pre-synaptic</th>
<th>Post-synaptic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Botulism</td>
<td>Lambert-Eaton Myasthenic Syndrome (LEMS)</td>
</tr>
<tr>
<td>LEMS</td>
<td>Myasthenia Gravis (MG)</td>
</tr>
</tbody>
</table>

**Botulism**
- Disorder of neuromuscular transmission due to an autoimmune response against the active sites on the presynaptic membrane

**LEMS**
- Disorder of neuromuscular transmission caused by Clostridium botulinum toxins blocking exocytosis of acetylcholine from the nerve terminal

**MG**
- Disorder resulting in a decreased quantal response because of an autoimmune response against acetylcholine receptors on the postsynaptic membrane
Botulism is caused by:
A. Clostridium botulinum toxin
B. Staphylococcus aureus
C. Campylobacter jejuni
D. Autoimmune response against acetylcholine receptors on the postsynaptic membrane

- Botulism is caused by Clostridium botulinum toxin. The toxin blocks release of acetylcholine.
- The incidence of foodborne botulism is approximately 24 cases per year. The incidence of wound botulism is 3 cases per year. The incidence of infant botulism is 71 cases per year, with a mean age of 3 months.

Botulism in an infant is associated with ingestion of:
A. Mayonnaise
B. Pasta
C. Strawberries
D. Honey

- Infants should never be given honey. Honey ingestion can lead to hypotonia ("floppy baby") syndrome in an infant because of botulinum toxins in the honey.
- In infant botulism, the baby does not ingest toxin; instead, spores from the botulism bacteria produce toxin in the baby's immature digestive tract. The toxins then travel to the baby's nerve cells leading to the characteristics symptoms of weakness and the "floppy infant syndrome."

Symptoms of botulism present how soon after spore ingestion?
A. 1 hour
B. 2 to 4 hours
C. 1 day
D. 1 month

- Symptoms such as blurred vision and diplopia can occur, along with nausea and vomiting, between 12 and 36 hours after consuming raw meat, fish, canned vegetables, or honey.
- Bulbar symptoms are noted first and include ptosis, dysphagia, or dysarthria. Gastrointestinal symptoms include nausea and vomiting, and then there may be widespread paralysis or flaccidity. If severe, there may be respiratory dysfunction.
- Lab work may reveal botulinum toxin in the stool or blood serum. Recovery occurs from collateral sprouting of nerves.
What is a requirement for the diagnosis of botulism?

A. Muscle biopsy  
B. Botulinum toxin found in stool or blood  
C. Chest x-ray  
D. All of the above

- Botulism is a rare but potentially deadly illness caused by a poison most commonly produced by a germ called Clostridium botulinum. The germ is found in soil and can survive, grow, and produce a toxin in certain conditions, such as when food is improperly canned.  
- The toxin is usually found in stool or blood.  
- Myasthenia gravis and Lambert–Eaton myasthenic syndrome would require a muscle biopsy.

Botulinum toxin (Botox) is a neurotoxic protein produced by the bacterium Clostridium botulinum and related species. It prevents the release of the neurotransmitter acetylcholine from axon endings at the neuromuscular junction and thus causes flaccid paralysis. Infection with the bacterium causes the disease botulism.

Both are acquired autoimmune disorders characterized by defective neuromuscular transmission

<table>
<thead>
<tr>
<th>LEMS</th>
<th>MG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Antibodies against voltage-gated Ca channels</td>
<td>Antibodies about acetylcholine receptors</td>
</tr>
<tr>
<td>Usually starts at extremities and moves up</td>
<td>Usually starts at eyes and moves down</td>
</tr>
<tr>
<td>Autonomic dysfunction present</td>
<td>No autonomic dysfunction</td>
</tr>
<tr>
<td>Diplopia and dysphagia uncommon</td>
<td>Diplopia and dysphagia common</td>
</tr>
<tr>
<td>Weakness improves with activity</td>
<td>Weakness worsens with activity</td>
</tr>
<tr>
<td>Associated with SCLC</td>
<td>Associated with thymoma</td>
</tr>
</tbody>
</table>

A 45-year-old man presents to your office with complaints of fatigue and weakness that are exacerbated with rest and often improved with exercise. He denies vision disturbances bilaterally. On physical exam, there is notable weakness in bilateral quadriceps. Which neuromuscular junction disorder may he have, and which location is affected?

A. Myasthenia gravis (MG); postsynaptic  
B. Lambert–Eaton myasthenic syndrome (LEMS); presynaptic  
C. Myasthenia gravis (MG); presynaptic  
D. Lambert–Eaton myasthenic syndrome (LEMS); postsynaptic

- LEMS is a disorder of neuromuscular transmission due to an autoimmune response against the active sites on the presynaptic membrane.  
- Patients complain of proximal fatigue and weakness that affects mainly the lower limbs, such as the quadriceps. Symptoms are exacerbated with rest and improved with exercise.  
- Rarely are there neck, facial, or bulbar muscle involvements. It is important to note that there is sparing of ocular muscles.  
- There is also often an association with malignancy, most commonly oat cell carcinoma of the lung.  
- Onset occurs more in males than in females (2:1) and in those older than 40 years.  
- Unlike MG, LEMS patients may experience an increase in strength after a series of muscle contractions.
Which of the following is associated with small cell (oat cell) carcinoma of the lung?

A. Lambert–Eaton myasthenic syndrome  
B. Botulism  
C. Myasthenia gravis  
D. None of the above

- **Lambert–Eaton myasthenic syndrome** is associated with small cell lung tumor. Small cell lung carcinoma: aggressive form of lung cancer most commonly occurs in smokers. It usually starts in the breathing tubes (bronchi) and grows very quickly, creating large tumors and spreading (metastasizing) throughout the body.  
- The estimated worldwide prevalence of LEMS is about 2.8 per million, making it a rare disease. There are approximately 400 known cases of LEMS in the United States. When LEMS is associated with SCLC, the patients tend to be older and are more likely to be men than women.  
- **Myasthenia gravis** is associated with thymic tumor. The prevalence of myasthenia gravis in the United States is estimated at 14 to 20 per 100,000 population, approximately 36,000 to 60,000 cases in the United States. However, myasthenia gravis remains underdiagnosed and the prevalence is probably higher.  
- **Botulism** is associated with ingestion of contaminated raw meat, canned vegetables, honey, or fish.

<table>
<thead>
<tr>
<th>LEMS</th>
<th>MG</th>
</tr>
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<tbody>
<tr>
<td>exacerbated with rest</td>
<td>exacerbated with exercise</td>
</tr>
</tbody>
</table>

Which of the following disorders is exacerbated with rest?

A. Botulism  
B. Myasthenia gravis (MG)  
C. Lambert–Eaton myasthenic syndrome (LEMS)  
D. None of the above

- The symptoms of LEMS are the result of an insufficient release of neurotransmitter by nerve cells.  
- Continued use of the muscles may lead to a buildup of the neurotransmitter to normal levels, so symptoms of LEMS can often be lessened or alleviated.  
- Symptoms of myasthenia gravis do not improve with continued muscle use.

What is the etiology of Lambert–Eaton myasthenic syndrome (LEMS)?

A. Disorder of neuromuscular transmission caused by Clostridium botulinum toxins blocking exocytosis of acetylcholine from the nerve terminal  
B. Disorder of neuromuscular transmission caused by C. botulinum toxins blocking endocytosis of acetylcholine from the nerve terminal  
C. Disorder resulting in a decreased quantal response because of an autoimmune response against acetylcholine receptors on the postsynaptic membrane  
D. Disorder of neuromuscular transmission due to an autoimmune response against the active sites on the presynaptic membrane

- This decreases calcium entry into the cell, causing a decreased release of acetylcholine into the synaptic cleft.  
- There is a strong association with malignancy, such as small cell (oat cell) carcinoma of the lung.  
- Answer choice A describes botulism, and answer choice C describes myasthenia gravis.
Which of the following is a postsynaptic neuromuscular junction disorder?

A. Myasthenia gravis  
B. Botulism  
C. Lambert–Eaton myasthenic syndrome  
D. None of the above

* Myasthenia gravis is a postsynaptic disorder. The other disorders are presynaptic.

In myasthenia gravis, there is an autoimmune response against:

A. Calcium channels  
B. Acetylcholine receptors  
C. Sodium channels  
D. Sodium potassium pump

* There is an autoimmune response to acetylcholine receptors in myasthenia gravis.

In myasthenia gravis (MG), early monitoring of which of the following can help prognosticate?

A. Exertion related fatigue  
B. Onset of diplopia  
C. Response to edrophonium  
D. Spirometry

* It is vital to do pulmonary function tests (PFTs) in patients diagnosed with MG. The forced vital capacity (FVC) is probably the most important PFT, as it correlates to the function of the pulmonary muscles, including the diaphragm. Severe myasthenia may cause respiratory failure due to exhaustion of the respiratory muscles.
Edrophonium or (Tensilon) test is used to help diagnose:

A. Botulism
B. Myasthenia gravis
C. Lambert–Eaton myasthenic syndrome
D. None of the above

- Edrophonium is a reversible acetylcholinesterase inhibitor.
- The Edrophonium (Tensilon) test can be used to confirm the diagnosis of MG. Facial weakness is provoked by repeated facial movements. Edrophonium chloride, a short-acting anticholinesterase, is then given by slow IV injection. In MG, the facial weakness is rapidly relieved by this test. Objective testing of muscular power elsewhere in the body will reveal similar responses. This test should be always undertaken in hospital where there are resuscitation facilities and with a drawn up syringe of atropine present.

<table>
<thead>
<tr>
<th>Edrophonium</th>
<th>Pyridostigmine</th>
</tr>
</thead>
<tbody>
<tr>
<td>for diagnosis</td>
<td>to get rid of symptoms</td>
</tr>
</tbody>
</table>

A 49-year-old female presents to your clinic with complaints of fluctuating double vision and droopy eyelids. The patient is sent for a test where edrophonium chloride is injected, and there is brief improvement in her symptoms. Which of the following treatments is most appropriate?

A. Trivalent ABE antitoxin
B. Guanidine
C. Treat the malignancy
D. Pyridostigmine (Mestinon)

- This patient has myasthenia gravis (MG). Pyridostigmine (Mestinon) reversibly binds to and inactivates acetylcholinesterase, which is a cholinesterase inhibitor. Mestinon 60 to 120 mg orally every 3 to 8 hours can be used to treat MG. Other treatment options for MG include thymectomy, corticosteroids, immunosuppressive agents, and plasmapheresis.
- Answer choice A is used for patients with botulism. Answer choice C is correct in Lambert–Eaton myasthenic syndrome (LEMS) patients with malignancy. Initial treatment should be aimed at the neoplasm because weakness frequently improves with effective cancer therapy. No further LEMS treatment may be necessary in some patients.

Of the following, which is a disorder of neuromuscular transmission due to an autoimmune response against acetylcholine receptors on the postsynaptic membrane?

A. Botulism  
B. Lambert–Eaton myasthenic syndrome (LEMS)  
C. Myasthenia gravis (MG)  
D. Amyotrophic lateral sclerosis (ALS)

- Neuromuscular junction disorders can be classified into two categories: presynaptic or postsynaptic. LEMS and botulism are presynaptic disorders, and myasthenia gravis is a postsynaptic disorder.
- Myasthenia gravis is a disorder resulting in a decreased quantal response because of an autoimmune response against acetylcholine receptors on the postsynaptic membrane.
- LEMS is a disorder of neuromuscular transmission due to an autoimmune response against the active sites on the presynaptic membrane.
- Botulism is a disorder of neuromuscular transmission caused by Clostridium botulinum toxins blocking exocytosis of acetylcholine from the nerve terminal.
- ALS is a motor neuron disease that presents with both upper and lower motor neuron lesions that is caused by degeneration of the anterior horn cell.

A 49-year-old woman presents to your clinic with complaints of fluctuating double vision and droopy eyelids. The patient is sent for a test where edrophonium chloride is injected, and there is brief improvement in her symptoms. Which of the following does she most likely have?

A. Myasthenia gravis (MG)  
B. Lambert–Eaton myasthenic syndrome (LEMS)  
C. Botulism  
D. Amyotrophic lateral sclerosis (ALS)

- Myasthenia gravis patients may complain of proximal muscle fatigue and weakness exacerbated with exercise, heat, or time of day (evening). It is important to note that there can be facial or bulbar symptoms, including ptosis, diplopia, dysphagia, or dysarthria.
- This patient presents with ocular myasthenia gravis. Ocular involvement is an extremely common initial presentation in MG patients. Drooping of eyelids and intermittent diplopia result from levator palpebrae (extraocular muscles) involvement, which is seen in 90% of MG cases.
- The test here describes the edrophonium (Tensilon) test.
  - An intravenous solution of edrophonium chloride is injected into a patient.
  - A total of 10 mg of the cholinergic drug is prepared, and a 2 mg dose is injected.
  - If there is no reaction in 30 seconds, the remaining 8 mg is administered.
  - A brief improvement in muscle activity is regarded as a positive result.
- Edrophonium chloride is also used to distinguish between myasthenia gravis and a cholinergic crisis. Because edrophonium chloride can precipitate respiratory depression, the test should not be performed unless an anticholinergic antidote, such as atropine, and respiratory resuscitation equipment are available.

<table>
<thead>
<tr>
<th>4 needle technique</th>
<th>Tonify _______</th>
<th>Sedate _______</th>
</tr>
</thead>
<tbody>
<tr>
<td>___ tonification</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

| MYASTHENIA GRAVIS    | Qi deficiency falling downward | Bu Zhong Yi Qi Tang | DU24, DU20, DU16, DU14, ST44, ST41 | • ST36, RN6, UB20  
|                     | Qi & Yin dual deficiency       | Liu Wei Di Huang T. + Sheng Mai San |                                   | • ST36, SP6, UB20  
|                     | SP & KD Yang deficiency        | You Gui Yin + Li Zhong Tang |                                   | • SP3, KD7, UB20, UB23  
|                     | Damp-Phlegm                    | Dao Tan Tang |                                   | • ST40, SP9, ST36  
|                     | Blood stasis                   | Xiong Long Tang |                                   | • LI4, SP6, RN17  

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