Acute and Chronic Neurological Diseases: Quick Notes

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Neurological Conditions

- Myasthenia Gravis

Myasthenia Gravis

- A chronic autoimmune, neuromuscular disease that causes weakness in the skeletal muscles that worsens after periods of activity and improves after periods of rest.
- The skeletal muscles are responsible for functions involving breathing and moving parts of the body, including the arms and legs.
- There is no known cure,
- Available treatments can control symptoms and often allow people to have a relatively high quality of life.
- Most individuals with the condition have a normal life expectancy.

Myasthenia Gravis

The thymus gland plays an important role in the development of the immune system in early life. Its cells form a part of the body's normal immune system. The gland is somewhat large in infants, grows gradually until puberty, and then gets smaller and is replaced by fat with age.
In about 75% of adults with myasthenia gravis, the thymus gland remains large and is abnormal. It contains certain clusters of immune cells indicative of lymphoid hyperplasia—
a condition usually found only in the spleen and lymph nodes during an active immune response.
Some individuals with myasthenia gravis (10%) develop thymomas (tumors of the thymus gland). Thymomas are generally benign, but they can become malignant.

Researchers suggest that the Thymus of MG patients does not appropriately eliminate cells that produce antibodies that attack body tissues.

Epidemiology

- Affects both men and women and occurs across all racial and ethnic groups.
- It most commonly impacts young adult women (under 40) and older men (over 60), but it can occur at any age,
- Myasthenia gravis is not inherited nor is it contagious. Occasionally, the disease may occur in more than one member of the same family..
- Although myasthenia gravis is rarely seen in infants, the fetus may acquire antibodies from a mother affected with myasthenia gravis—a condition called neonatal myasthenia.
  - Neonatal myasthenia gravis is generally temporary, and the child's symptoms usually disappear within two to three months after birth.
- Rarely, children of a healthy mother may develop congenital myasthenia. This is not an autoimmune disorder but is caused by defective genes.

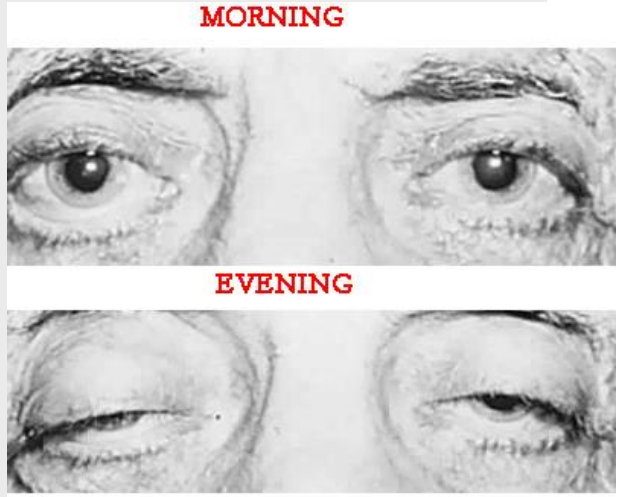
Variations

- Ocular
  - Eye and lid muscles are affected
- Generalized
  - Proximal muscles of both upper and lower extremities are involved, with ocular or bulbar involvement
- Bulbar
  - Muscles of speech, swallowing and breathing are affected
- Neonatal Transient

Etiology

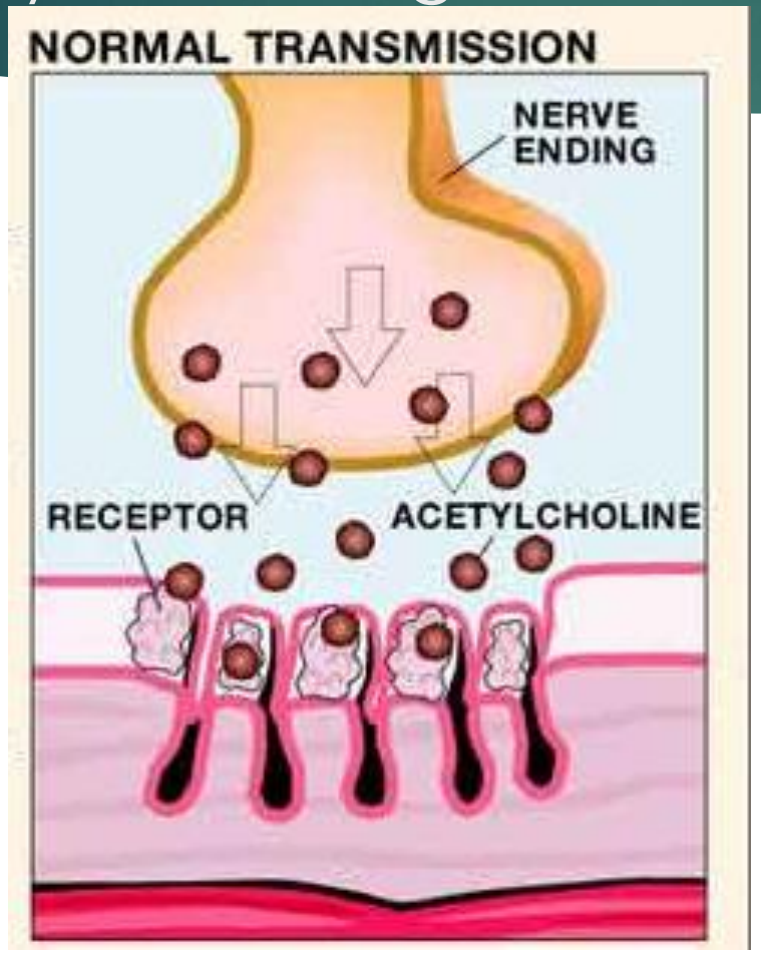
Common symptoms can include:

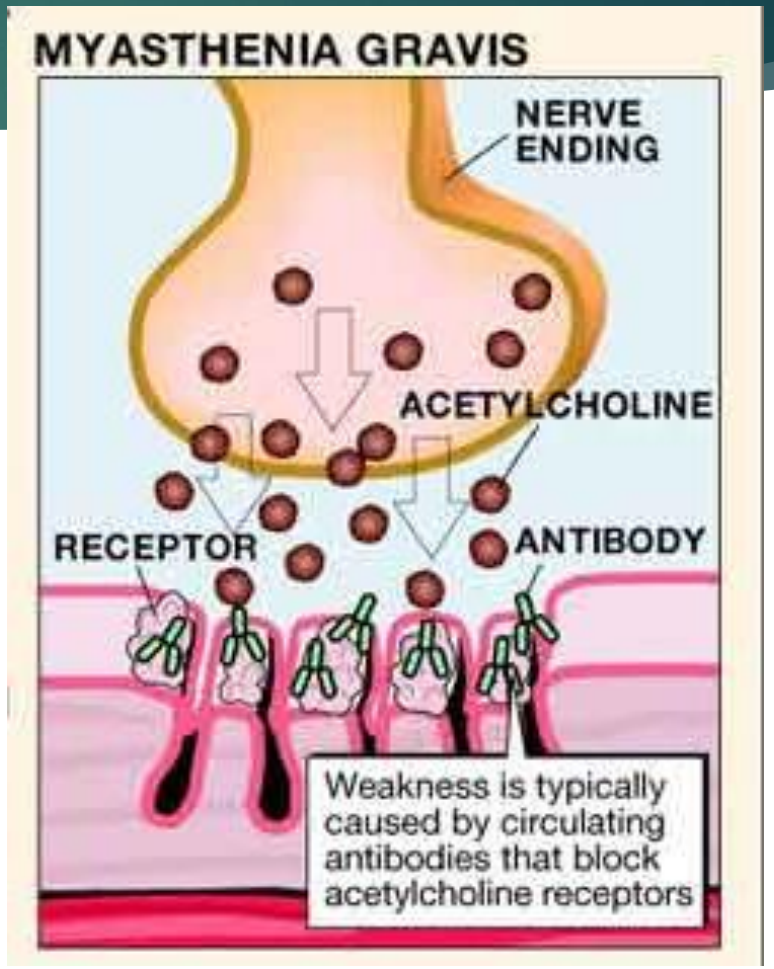
- Weakness of the eye muscles (called ocular myasthenia)
- Drooping of one or both eyelids (ptosis)
- Blurred or double vision (diplopia)
- A change in facial expression
- Difficulty swallowing
- Shortness of breath
- Impaired speech (dysarthria)
- Weakness in the arms, hands, fingers, legs, and neck.



Sometimes the severe weakness of myasthenia gravis may cause respiratory failure, which requires immediate emergency medical care.

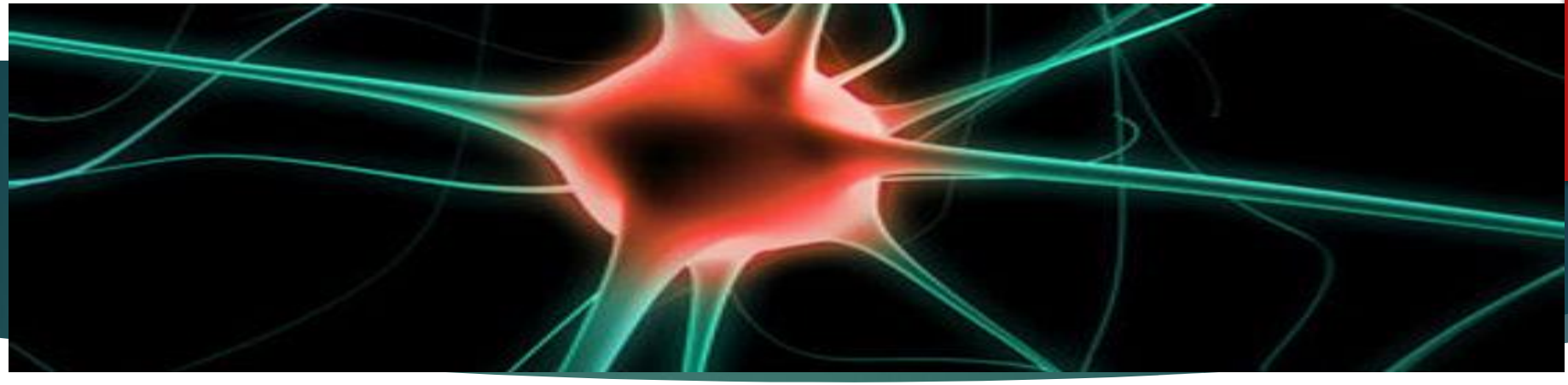
http://www.ninds.nih.gov/disorders/myasthenia_gravis/myasthenia_gravis.htm





Diagnosis

- **A physical and neurological examination**.
- **An edrophonium test**.
  - This test uses injections of edrophonium chloride to briefly relieve weakness in people with myasthenia gravis. The drug blocks the breakdown of acetylcholine and temporarily increases the levels of acetylcholine at the neuromuscular junction. It is usually used to test ocular muscle weakness.
- **A blood test**.
  - An abnormally elevated levels of acetylcholine receptor antibodies.
  - A second antibody—called the anti-MuSK antibody—has been found in about half of individuals with myasthenia gravis who do not have acetylcholine receptor antibodies.
  - However, in some individuals with myasthenia gravis, neither of these antibodies is present. These individuals are said to have seronegative (negative antibody) myasthenia.
- **Electrodiagnostics**.
  - Diagnostic tests include repetitive nerve stimulation, which repeatedly stimulates a person's nerves with small pulses of electricity to tire specific muscles.
  - . Single fiber electromyography (EMG), considered the most sensitive test for myasthenia gravis, detects impaired nerve-to-muscle transmission. EMG can be very helpful in diagnosing mild cases of myasthenia gravis when other tests fail to demonstrate abnormalities.
- **Diagnostic imaging**.
  - Computed Tomography (CT) or magnetic resonance imaging (MRI) may identify the presence of a thymoma.
- **Pulmonary function testing**.
  - Measuring breathing strength can help predict if respiration may fail and lead to a myasthenic crisis.



- **Myasthenia Crisis**
  - A myasthenic crisis is a medical emergency that occurs when the muscles that control breathing weaken to the point where individuals require a ventilator to help them breathe. It may be triggered by infection, stress, surgery, or an adverse reaction to medication. Approximately 15 to 20 percent of people with myasthenia gravis experience at least one myasthenic crisis.

MGFA Classification System

Class I: Any ocular muscle weakness; may have weakness of eye closure. All other muscle strength is normal.

Class II: Mild weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.

A. IIa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
B. IIb. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

Class III: Moderate weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.

A. IIIa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
B. IIIb. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

Class IV: Severe weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.

A. IVa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
B. IVb. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

Class V: Defined as intubation, with or without mechanical ventilation, except when employed during routine postoperative management. The use of a feeding tube without intubation places the patient in class IVb.

Management Options

- Thymectomy

  This operation to remove the thymus gland A NINDS-funded study found that thymectomy is helpful both for people with thymoma and those with no evidence of the tumors.

- Monoclonal antibody

  This treatment targets the process by which acetylcholine antibodies injure the neuromuscular junction. In 2017, the U.S. Food and Drug Administration approved the use of eculizumab for the treatment of generalized myasthenia gravis in adults who test positive for the antiacetylcholine receptor (AchR) antibody.

- Anticholinesterase medications

  Medications to treat the disorder include anticholinesterase agents such as mestinon or pyridostigmine, which slow the breakdown of acetylcholine at the neuromuscular junction and thereby improve neuromuscular transmission and increase muscle strength.

- Immunosuppressive drugs

  These drugs improve muscle strength by suppressing the production of abnormal antibodies. They include prednisone, azathioprine, mycophenolate mofetil, and tacrolimus. The drugs can cause significant side effects and must be carefully monitored by a physician.

Medications for Myasthenia Gravis

1. Pyridostigmine
2. Azathioprine
3. Neostigmine
4. Mestinon
5. Soliris
6. Vyvgart
7. Ultomiris
8. mycophenolate mofetil
9. Rystiggo
10. Zilbrysq

ZILBRYSQ:
the first once-daily, self-administered treatment for adults with anti-AChR antibody-positive generalized myasthenia gravis (gMG).

- Results of the ZILBRYSQ clinical trial:
  - Significant improvements in the activities of daily living
  - These results were measured using the Myasthenia Gravis Activities of Daily Living scale (MG-ADL)
  - Rapid improvements in daily living
  - More than 7 in 10 participants responded to treatment.
  - Nearly 6 in 10 participants saw improvement in muscle strength

ULTOMIRIS

- Is the first and only long-acting complement C5 inhibitor, with immediate, complete, and sustained complement inhibition1-4
- The precise mechanism by which ULTOMIRIS exerts its therapeutic effect in gMG patients is not known1
- The PK/PD data derived from the 86 patients who received ravulizumab in the RCP of the CHAMPION-MG study.1,4

[Mechanism of Action | ULTOMIRIS® (ravulizumab-cwvz) | gMG]

Management Options

- Plasmapheresis and intravenous immunoglobulin

  These therapies may be options in severe cases of myasthenia gravis. Individuals can have antibodies in their plasma (a liquid component in blood) that attack the neuromuscular junction.

  These treatments remove the destructive antibodies, although their effectiveness usually only lasts for a few weeks to months.

  - Plasmapheresis is a procedure using a machine to remove harmful antibodies in plasma and replace them with good plasma or a plasma substitute.
  - Intravenous immunoglobulin is a highly concentrated injection of antibodies pooled from many healthy donors that temporarily changes the way the immune system operates. It works by binding to the antibodies that cause myasthenia gravis and removing them from circulation.

Crisis Management

- Bilevel positive airway pressure and other respiratory support systems can prevent need to for intubation or decrease recovery time
- Safe Environment
- Energy Conservation
  - Therapy times
  - Rest periods
- Nutritional support
  - Soft diet to decrease fatigue in eating
  - Caloric support if needed

Management

- Monitor for crisis events, side effects of treatments, individual tolerance and weakness
- Communicate with medical team for treatment changes based on patient response
- Assist to remain as independent as possible with ADLs
- Patient and Family Education
  - Disease Process
  - Side Effects of meds and treatments
  - Medical emergency- Myasthenic crisis or Cholinergic Crisis
  - No over the counter meds w/o consult of Dr.

Research

- Technological advances have led to more timely and accurate diagnosis,
- New and enhanced therapies have improved management of the disorder.
- There is a greater understanding about the structure and function of the neuromuscular junction, the fundamental aspects of the thymus gland and of autoimmunity, and the disorder itself.
- Researchers are seeking to learn what causes the autoimmune response in myasthenia gravis, and to better define the relationship between the thymus gland and myasthenia gravis.

Nursing Process

- Assessment
  - Full Health History
  - Assess baseline to monitor for crisis- (strongest time of day)

- Resp function
- Cardiac Function
- Bowel and Bladder function
- GI Symptoms

- Visual Acuity
- Strength and Mobility
- Swallowing
- Speech

Plan of Care

- Knowledge Deficit re: disease process and side effects of treatment
- Activity intolerance re: fatigue
- Risk for aspiration re: muscle weakness and increased secretions
- Risk for Falls
- Risk for Medical Crisis (Myasthenic Crisis)