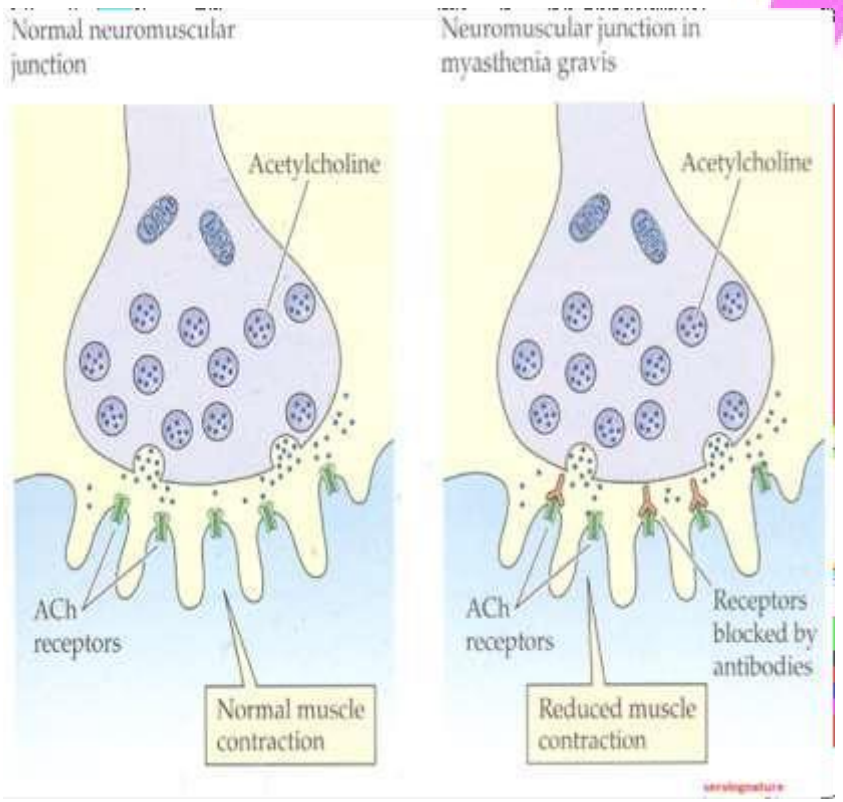
MYASTHENIA GRAVIS

INTRODUCTION

- Myasthenia gravis comes from the Greek and Latin words meaning "grave muscular weakness." The most common form of MG is a chronic autoimmune neuromuscular disorder that is characterized by fluctuating weakness of the voluntary muscle groups.

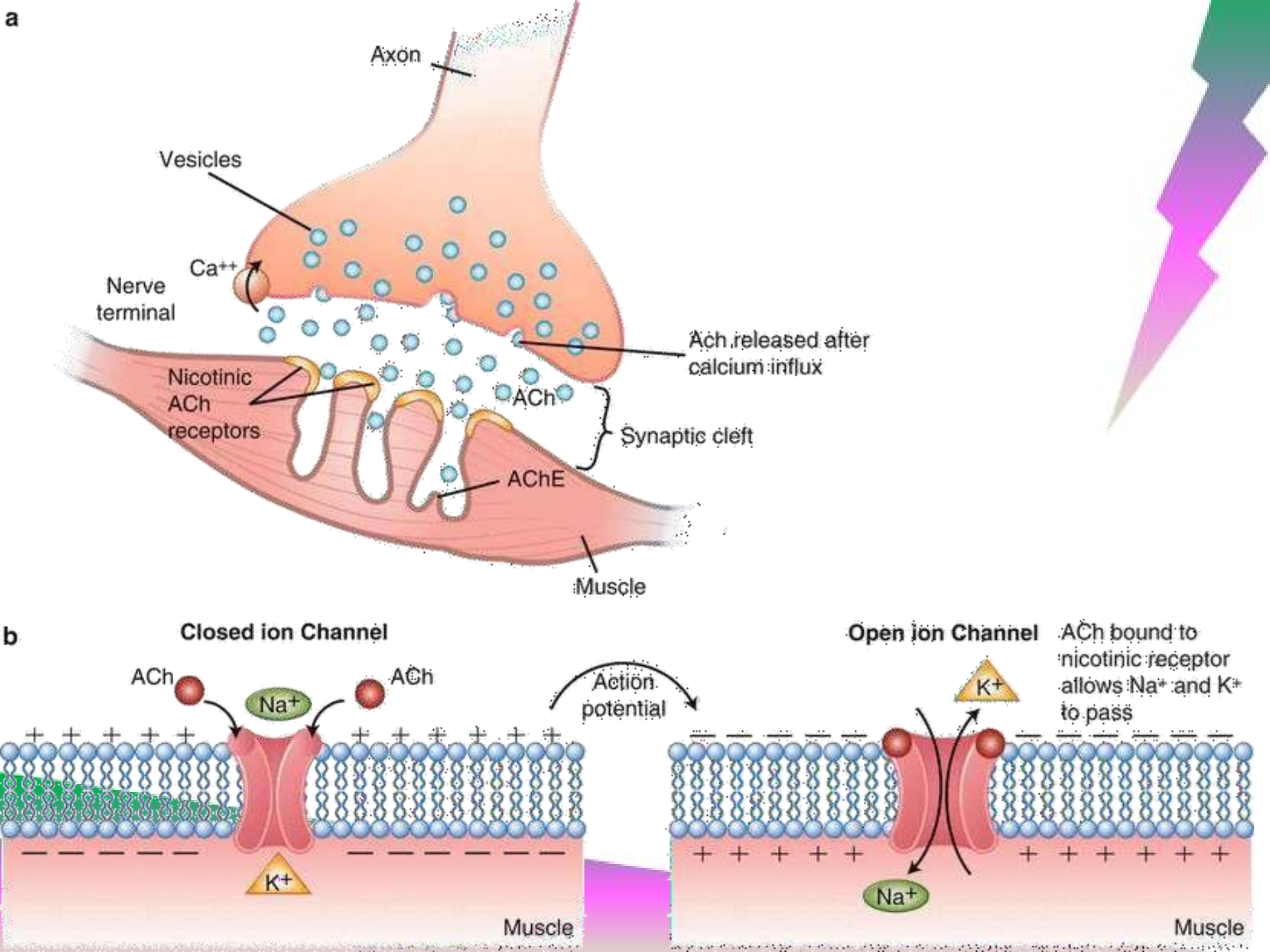


DEFINITION

- Myasthenia gravis is an autoimmune disorder affecting the myo neural junction, is characterized by varying degrees of weakness of the voluntary muscles.
- 3 times more common in women.
- Antibody attacks acetylcholine receptors.

Neuromuscular junction

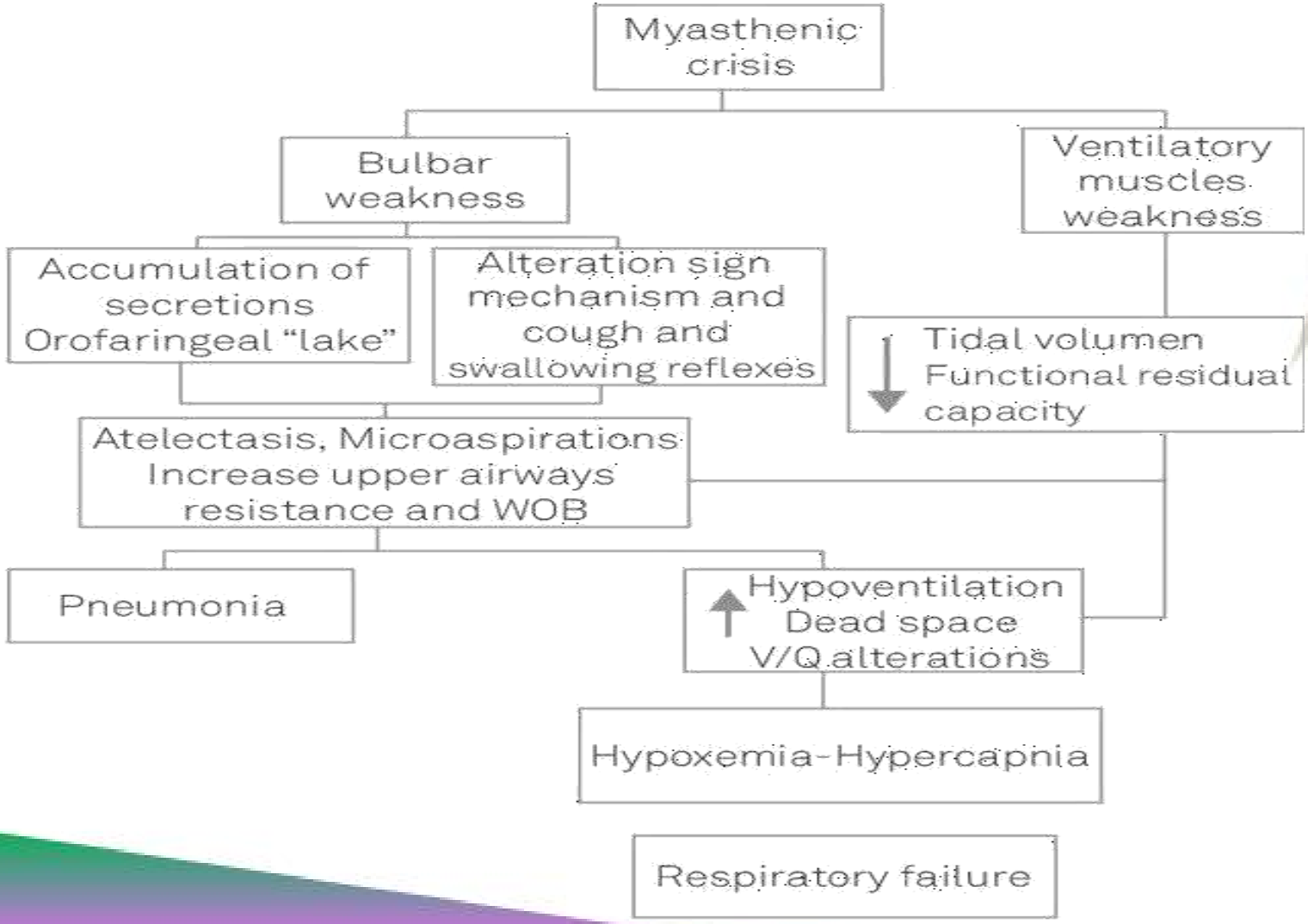
- Neuromuscular junction act as a bridge between the nervous system and the muscular system. It is a microstructure through which the process of contraction is initiated or halted in the muscles by the neurons. Any changes in the neuromuscular junction can result in impaired contractions of the skeletal muscles.


a
Axon
Vesicles
Ca++
Nerve terminal
Ach released after calcium influx
Nicotinic ACh receptors
ACh
Synaptic cleft
AChE
Muscle
b
Closed ion Channel
ACh
ACh
Na+
K+
Muscle
Action potential
Open ion Channel
ACh bound to nicotinic receptor allows Na+ and K+ to pass
K+
Na+
Muscle

Etiology

- In Myasthenia gravis, the receptors at the muscle surface are destroyed or deformed by antibodies that prevent a normal muscular reaction from occurring.
- The causative factor is unknown, but the disorder may have a genetic link.
- Autoantibodies that destroys acetylcholine receptors.
- Thymus tumors found in 15% of patients.

Pathophysiology

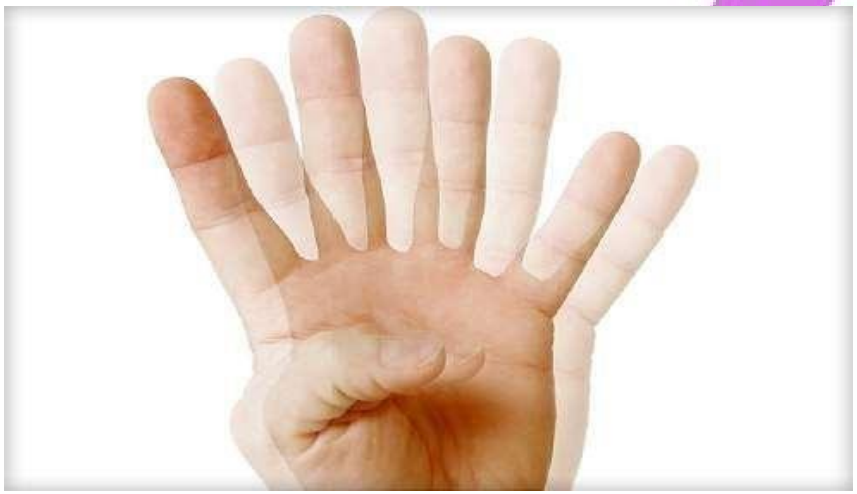


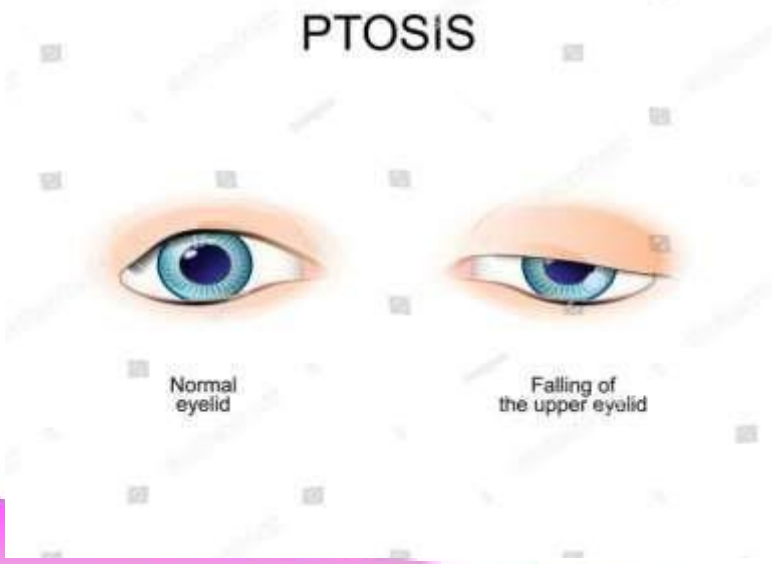
Pathophysiology

- Normally a chemical impulse precipitates the release of acetylchoiline from the vesicles on the nerve terminals at myoneural junction. These acetylcoline attach to receptor sites on the motor end plate stimulate muscle contraction.
- In myasthenia gravis autoantibodies directed at the acetylcholine receptors sites impairs the transmission of impulse at myoneural junction. Fewer receptors are avilable for stimulation.

Clinical manifestations

- Increasing muscle weakness.
- Affected muscles are ,
- Muscle used for the movement of eyes, chewing, swallowing, speaking, and breathing.
- In severe cases respiratory muscle weakness occur.
- Laryngeal involvement produces dysphonia.
- Ptosis.
- Diplopia





- Fluctuating weakness increased by exertion
  - Weakness increases during the day and improves with rest
- Extraocular muscle weakness
  - Ptosis is present initially in 50% of patients and during the course of disease in 90% of patients.
- Head extension and flexion weakness.
  - Weakness may be worse in proximal muscles

Muscle strength

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

- Facial muscle weakness is almost always present
  - Ptosis and bilateral facial muscle weakness
  - Sclera below limbus may be exposed due to weak lower lids
- Bulbar muscle weakness
  - Palatal muscles
    - “Nasal voice”, nasal regurgitation
    - Chewing may become difficult
    - Severe jaw weakness may cause jaw to hang open
    - Swallowing may be difficult and aspiration may occur with fluids—coughing and choking while drinking
- Neck muscles Neck flexors affected more than extensors

- Limb muscle weakness
  - Upper limbs more common than lower limbs
- Respiratory muscle weakness
  - Weakness of the intercostal muscles and the diaghram may result in CO_2 retention due to hypoventilation
  - Weakness of pharyngeal muscles may collapse the upper airway
    - Monitor negative inspiratory force, vital capacity and tidal volume
    - Arterial blood oxygenation may be normal while CO2 is retained

- Occular muscle weakness
  - Asymmetric
    - Usually affects more than one extraocular muscle and is not limited to muscles innervated by one cranial nerve
    - Weakness of lateral and medial recti may produce a pseudointernuclear opthalmoplegia
  - Ptosis caused by eyelid weakness
  - Diplopia is very common.

Diagnosis

- History and physical examination
- Blood analysis Elevated levels of acetylcholine receptor antibodies.
- Anti MuSK(Muscle- Specific Kinase) antibody.
- Tensilon test(Edraphonium test)-

Tensilon test

- A medicine called Tensilon (also called edrophonium) (It inhibit Ach breakdown. Therefore more Ach are available) is given, IV. 2 mg at a time to a total of 10 mg. 30 seconds after injection, the patient is observed for improvement in muscle strength.
- If the muscle strength increases in response to Tensilon, then it is likely that myasthenia gravis
- Iv atropine is get ready if any allergic reaction occurs.

Single-fiber electromyography (EMG)

- It is considered as the most sensitive test for myasthenia gravis, detects impaired nerve-to-muscle transmission.
- Imaging scans CT Or MRI: Its shows thymus enlargement.

Pharmacological Management

- Immunosuppressive Therapy
- Prednisone
- Azathioprine
- Acetylcholinesterase Inhibitors
- First line of therapy
  - Neostigmine bromide (Pyridostigmine)
  - Edrophonium chloride (Tensilon)
- Immunoglobulin Therapy

- **Corticosteroids –** to suppress the immune system. To decreases production of autoantibody.
- **Plasmapheresis-** process by which plasma is seperated from formed elements of blood , plasma is discarded and RBC electrolyte are returned to the clients.

Complications

Myasthenic Crisis

- Under medication,
- Increased HR/BP/RR Bowel and bladder incontinence
- Decreased urine output
- Absent cough and swallow reflex May need mechanical ventilation
- Temporary improvement of symptoms with administration of Tensilon

Cholinergic Crisis

- Overmedication,
- Decreased BP Abd cramps N/V, Diarrhea
- Blurred vision
- Pallor Facial muscle twitching
- Constriction of pupils
- Tensilon has no effect Symptoms improve with administration of anticholinergics (Atropine)

Nursing Interventions

- Maintain patent airway
- Assess swallowing to prevent aspiration
- Keep appropriate equipment available at the bedside: Oxygen, suction, Ambu bag, endotracheal intubation
- Promote energy conservation measures
- Consult Physical Therapy.
- Consult OT for assistive devices to facilitate ADLs
- Consult with speech and language therapist if weakening facial muscles impact communication
- Monitor I/O, serum albumin levels, and daily weights Know the signs and symptoms of both Myasthenic Crisis.
- Administer Medications as per order.

Patient teaching

- Teach patient/family disease process, complications, and treatments
- Teach patient about their medications uses dosage etc.
- Teach medications to use with caution that is muscle exacerbation
- Beta blockers, calcium channel blockers, quinine, quinidine.

Nursing diagnosis

- Ineffective breathing pattern related to intercosta muscle weakness.
- Ineffective airway clearance related to muscle weakness
- Impaired verbal communication related to weakening of larynx
- Disturbed sensory perception related to ptosis.

THANK YOU