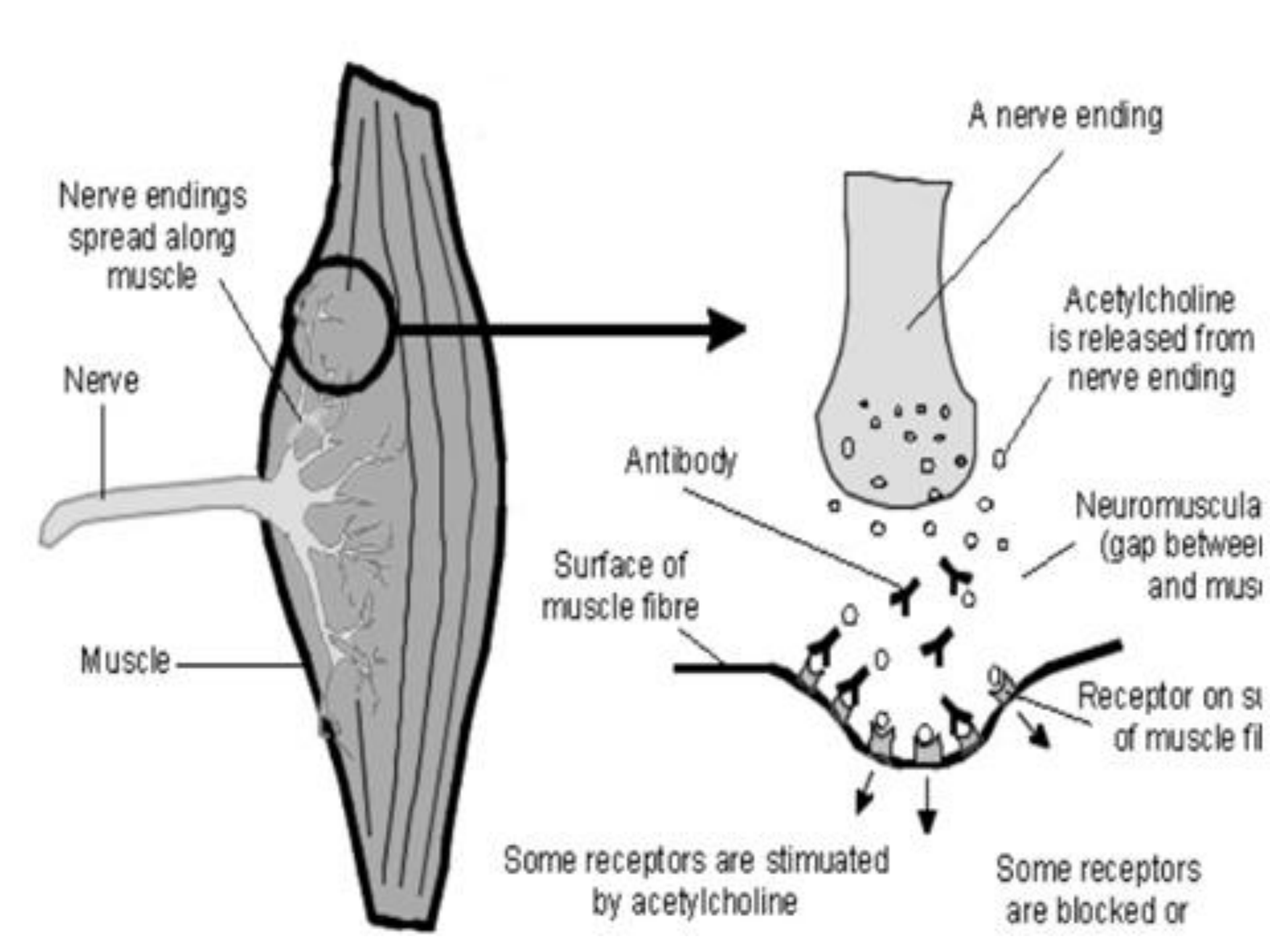
Definition

- Myasthenia gravis, an autoimmune disorder affecting the myoneural junction, is characterized by varying degrees of weakness of the voluntary muscles.
- It is an autoimmune disorder, in which weakness is caused by circulating antibodies that block acetylcholine receptors at the postsynaptic neuromuscular junction, inhibiting the excitatory effects of the neurotransmitter acetylcholine throughout neuromuscular junctions.


Nerve endings spread along muscle
Nerve
Muscle
A nerve ending
Acetylcholine is released from nerve ending
Antibody
Surface of muscle fibre
Some receptors are stimuated by acetylcholine
Some receptors are blocked or

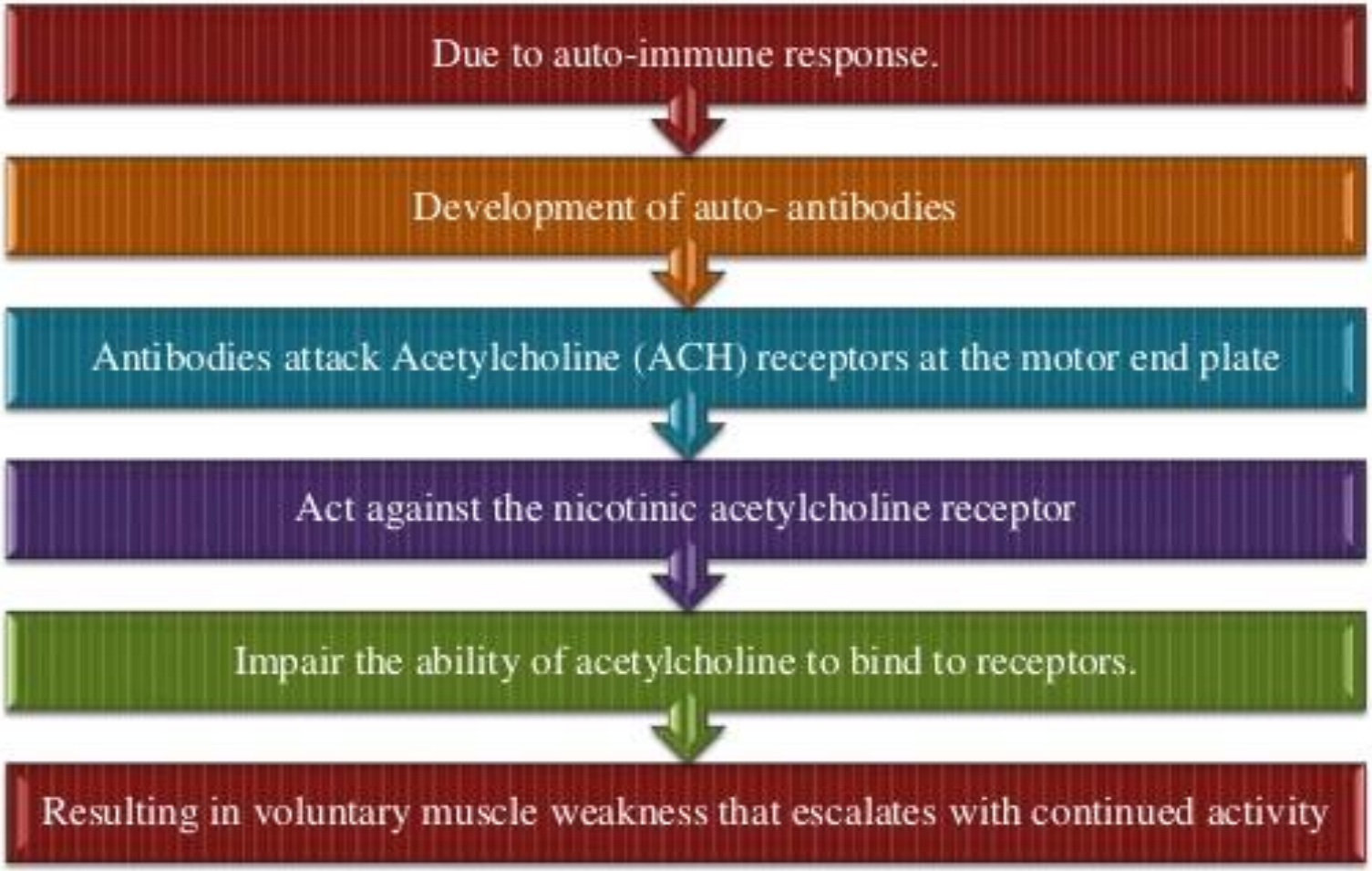
Causes

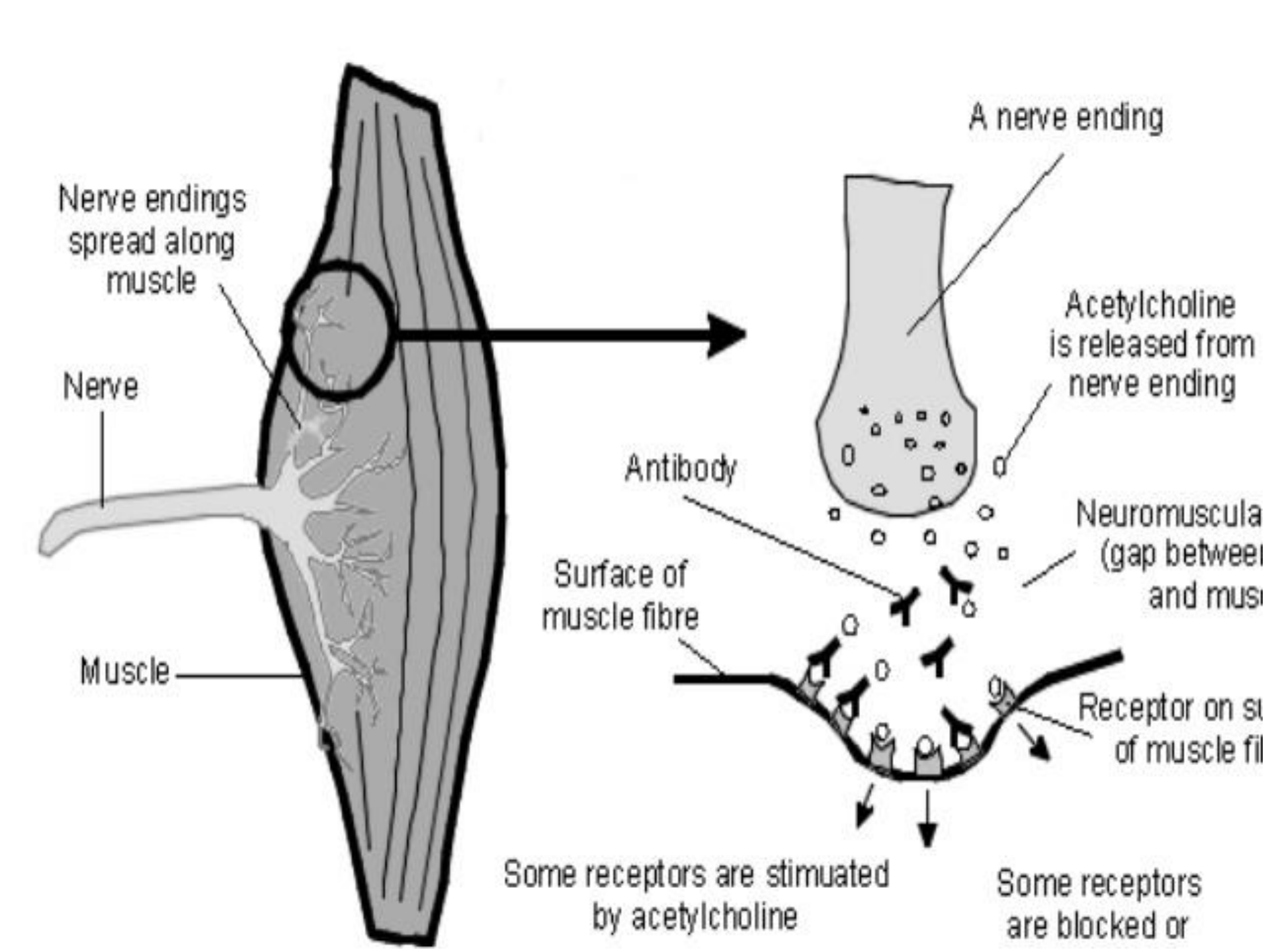
- Idiopathic.
- Autoantibodies that destroys acetylcholine receptors.
- Thymus tumors found in 15% of patients

Incidence

- MG affects 14 per 100,000 people in the United States
- Can affect any age group
- Women – peak incidence 20's to 30's
- Men – peak incidence 50's to 60's

PATHOPHSIOLOGY

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CLINICAL MANIFESTATION

- Affects any of the muscles that control voluntarily, certain muscle groups are more commonly affected than others
  - Eye, face, throat, neck, limb muscles
- The hallmark of myasthenia gravis is fatigability
- Dysarthria.
- Dysphagia.
- Ptosis.
- Diplopia
- Nasal-sounding speech
- Worsening muscle weakness

Myasthenia Gravis

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1. Ptosis
2. Muscle wasting, weakness
3. Fatigue, activity induced –due to muscle weakness
4. Respiratory muscle failure causing breathing difficulties
5. Diplopia
6. Nasal regurgitation (bulbar mm weakness)
7. Food regurgitation, difficulty chewing (bulbar mm weakness)
8. Dysphagia, choking (bulbar mm weakness)

DIAGNOSTIC EVALUATION

- History:-
- Physical examination:-
- Edrophonium test (Tensilon)
- Blood analysis:-
- Repetitive nerve stimulation
- Single-fiber electromyography (EMG)
- Imaging scans:

Physical Examination

- Muscle strength and tone.
- Coordination.
- Sense of touch
- and impairment of eye movements.

Anticholinesterase test

- Edrophonium chloride (Tensilon) is injected intravenously, 2 mg at a time to a total of 10 mg. 30 seconds after injection, facial muscle weakness and ptosis should resolve for about 5 minutes.
- This immediate improvement in muscle strength after administration of this agent represents a positive test and usually confirms the diagnosis.

Blood analysis

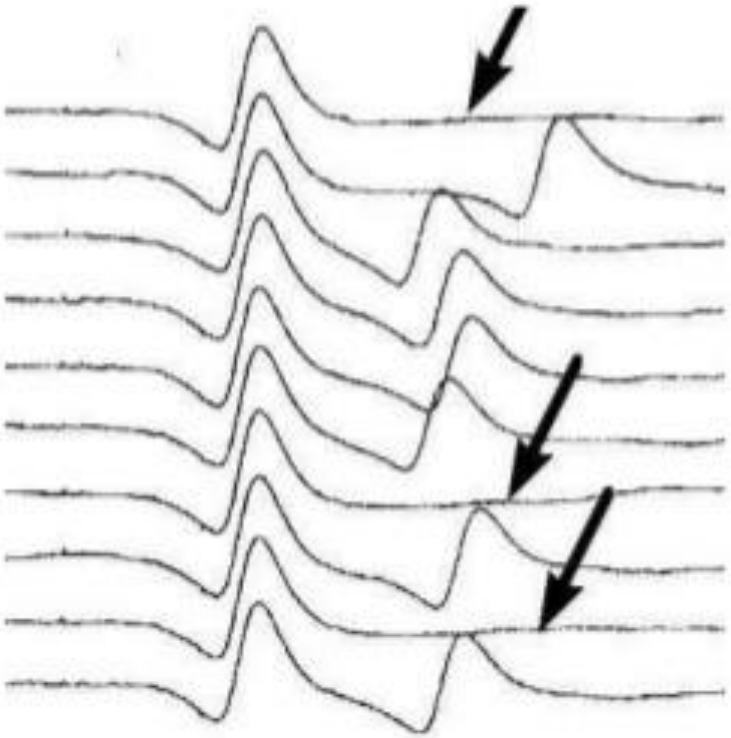
- Elevated levels of acetylcholine receptor antibodies.
- Anti MuSK(Muscle-Specific Kinase) antibody.

Repetitive Nerve stimulation

- Repetitive nerve stimulation, which repeatedly stimulates a person's nerves with small pulses of electricity to tire specific muscles.
- Muscle fibers do not respond as well to repeated electrical stimulation.

Single-fiber electromyography (EMG)

- It considered the most sensitive test for myasthenia gravis, detects impaired nerve-to-muscle transmission.



Imaging scans

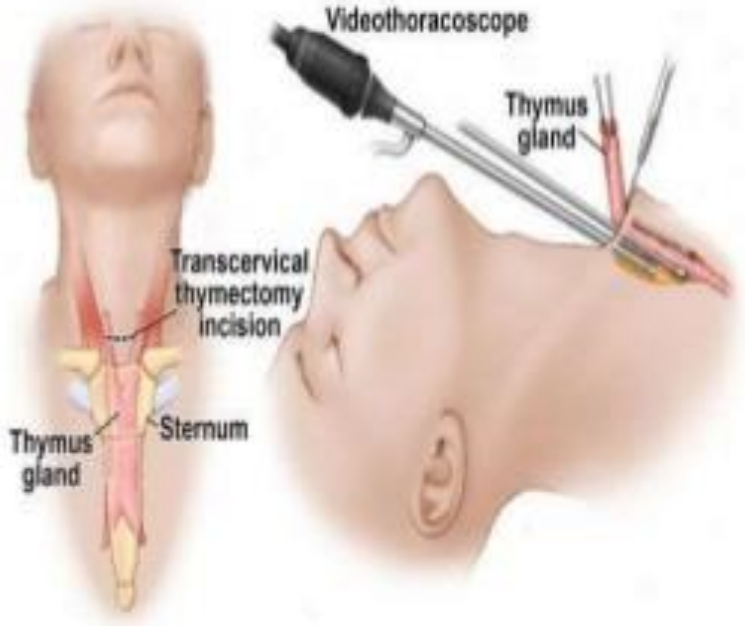
- CT Or MRI: Its shows thymus enlargement.

Pharmacological Management

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

Surgical management

- Thymectomy: (surgical removal of the thymus gland) can produce antigen-specific immunosuppression and result in clinical improvement.



COMPLICATION

⌗ Myasthenic Crisis:-

A myasthenic crisis is an exacerbation of the disease process characterized by severe generalized muscle weakness and respiratory and bulbar weakness that may result in respiratory failure.

Myasthenic Crisis VS. Cholinergic Crisis

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

- 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, procainamide, some antibiotics, neuromuscular blocking agents

Prognosis

- Chronic disease with periods of exacerbation and sometimes remissions .
- Disease course is highly variable.
- Symptoms respond well to treatment and in most cases the patient can live a normal or nearly normal life.
- Ocular Myasthenia has the best prognosis.

- -y71ear-old male complains of intermittent weakness and muscle fatigue progressively worsening over the past month.
- A previous long-distance runner, he now has difficulty getting his mail.
- His symptoms of profound leg weakness and fatigue are attributed to age and his underlying history of CAD atrial fibrillation.
- Over the past few months, he also reports having noted “eye strain” when working at the computer or reading for long periods of time.
- He has developed intermittent double vision that seems to be worse when reading at bedtime

• Meds: warfarin, atorvastatin, and amiodarone• No changes in weight; no fever, chills, SOB, CP

• Vitals: WNL
• Cognition, sensation, and cerebellar function intact
• Absence of Lhermitte's sign
• Weakness of right EOM with repetition; positive nystagmus; positive ice test; PERRLA
• Symmetric upper extremity weakness with fasciculation
• Decreased repetitive SLR and rising from seated position
• No muscular atrophy

- IgG antibodies attack AChRs or MuSK
- Antibody-antigen complex and inflammation inhibit neuromuscular transmission
- Breakdown of immune tolerance involves thymus
- Afffects 140-200/1,000,000 population
- 36,000-60,000 Americans
- Women > men before age 50
- Peak onset 20-40 years
- Men more commonly diagnosed ages 60-80

Clinical Features of MG

- Painless, striated muscle weakness
- Worse with activity; improved with rest
- Exacerbating factors include emotional stress, rapid changes in body temperature, infection, trauma, and multiple medications
- Ocular manifestations common: diplopia and ptosis
- Ocular MG 10-40% of all cases
- Generalized MG symptoms vary based on muscle groups: oropharyngeal, skeletal, or respiratory
- UE weakness more common than LE weakness

Differential Diagnoses of MG

- Amyotrophic Lateral Sclerosis
- Botulism
- Cranial Nerve Palsies
- Guillain-Barré Syndrome
- Lambert-Eaton Myasthenic Syndrome
- Multiple Sclerosis
- Polymyositis
- Transient Ischemic Attack and Stroke
- Tumors

Treatment of MG •

- Symptomatic therapy
  - AChE inhibitors
- Immune-directed therapy
  - Corticosteroids
  - Non-steroid immunomodulators
    - Azathioprine
    - Mycophenolate mofetil
    - Cyclosporine
    - Cyclophosphamide
    - Tacrolimus
    - Rituximab
    - Etanercept

Treatment of MG •

- Acute management therapies
  - Plasmapheresis
  - IV IG Infusion
- Surgical intervention