Department of Anesthesia Techniques

NEUROMUSCULAR WEAKNESS SYNDROMES

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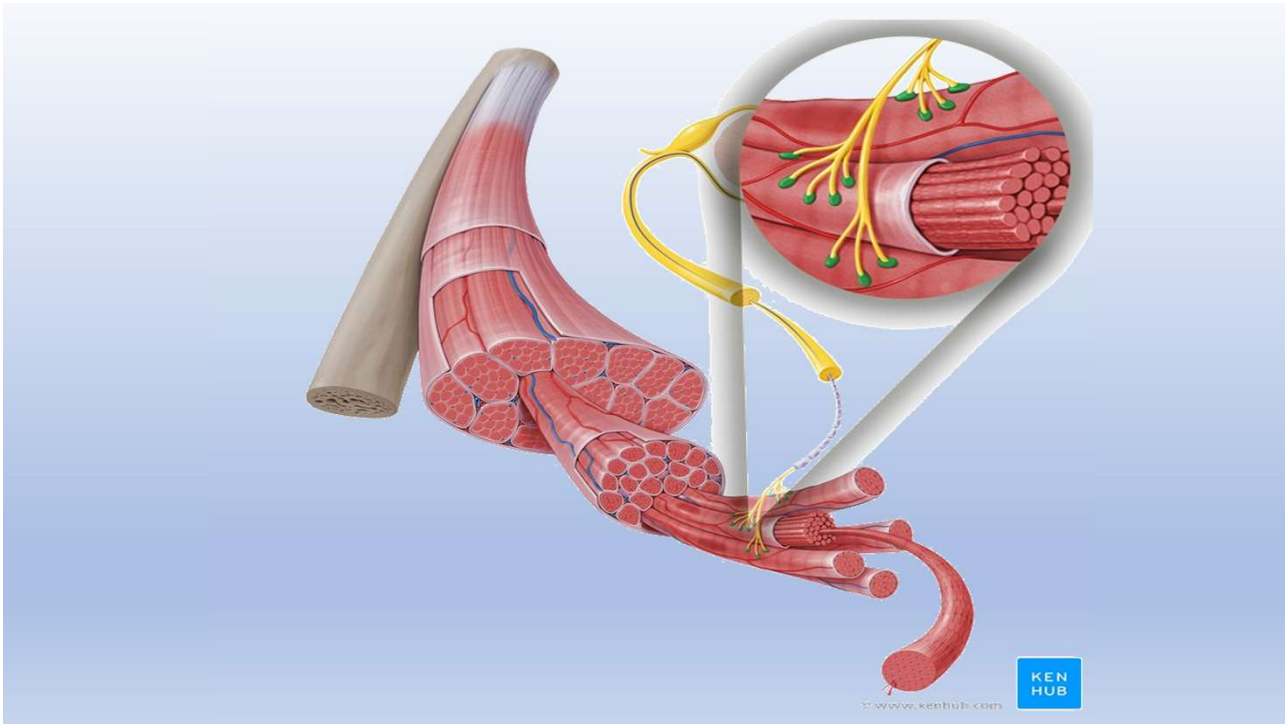
Neuromuscular weakness syndromes

- myasthenia gravis
- Guillain-Barré syndrome

A. Myasthenia Gravis (MG)

Is an autoimmune disease produced by antibody-mediated destruction of acetylcholine receptors on the postsynaptic side of neuromuscular junctions.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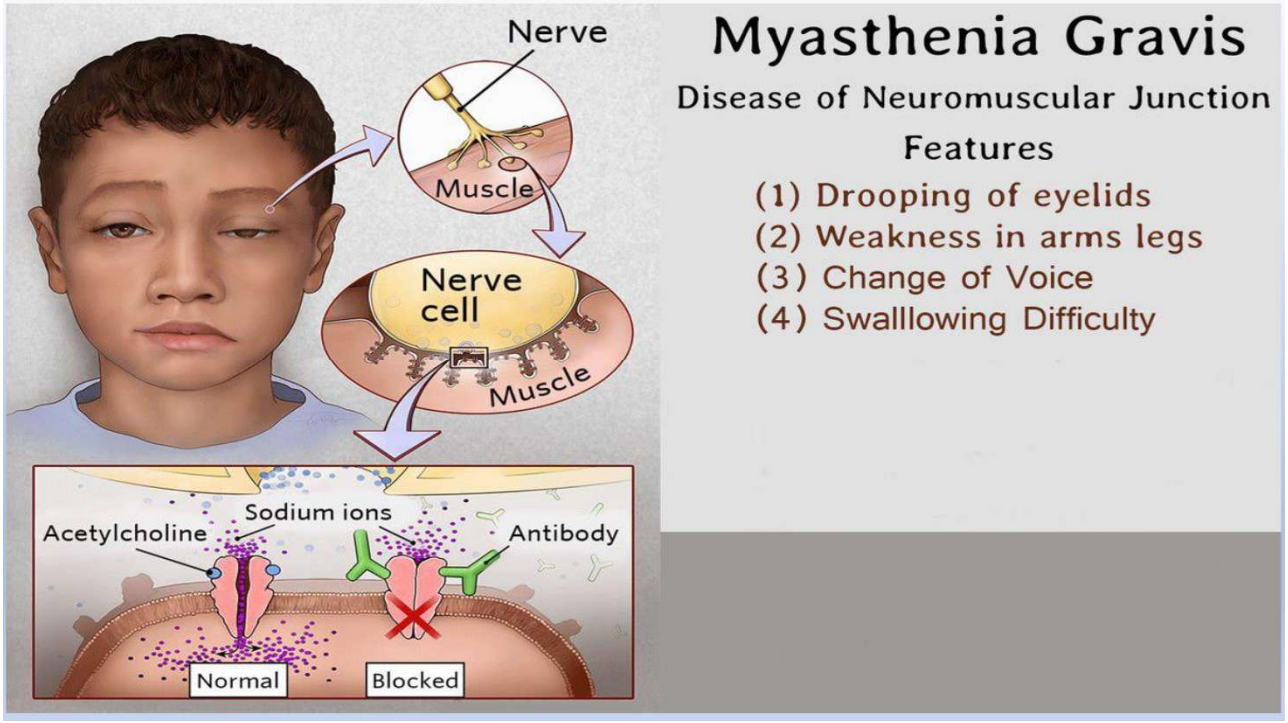

Nerve
Muscle
Nerve cell
Muscle
Sodium ions
Acetylcholine
Antibody
Normal
Blocked
Myasthenia Gravis
Disease of Neuromuscular Junction
Features
(1) Drooping of eyelids
(2) Weakness in arms legs
(3) Change of Voice
(4) Swalllowing Difficulty

Predisposing Conditions

- MG can be triggered by:
  - Major surgery
  - concurrent illness.
  - Thymic tumors are responsible for up to 20% of cases.
  - Several drugs can precipitate or aggravate MG;
    - antibiotics (e.g., aminoglycosides, ciprofloxacin)
    - cardiac drugs (e.g., beta-adrenergic blockers
    - Lidocaine
    - procainamide,
    - quinidine).

Clinical Feature

The muscle weakness in MG has the following features:

a. The weakness worsens with activity and improves with rest.

b. Weakness is first apparent in the eyelids and extraocular muscles, and limb weakness follows in 85% of cases.

c. Progressive weakness often involves the chest wall and diaphragm, and rapid progression to respiratory failure, called myasthenic crisis, occurs in 15–20% of patients.

d. The deficit is purely motor, and deep tendon reflexes are preserved.

Diagnosis

The diagnosis of MG is suggested by weakness in the eyelids or extraocular muscles that worsens with repeated use. The diagnosis is confirmed by:

A. Increased muscle strength after the administration of Edrophonium (Tensilon), an acetylcholinesterase inhibitor.

B. A positive assay for acetylcholine receptor antibodies in the blood, which are present in 85% of patients of MG.

Treatment

A. acetylcholinesterase inhibitor like pyridostigmine (Mestinon), The first line of therapy, Pyridostigmine can be given intravenously to treat myasthenic crisis: the IV dose is 1/30th of the oral dose

B. Immunotherapy is added, if needed, using either:

prednisone (1–1.5 mg/kg/day)

azathioprine (1–3 mg/kg/day)

cyclosporine (2.5 mg/kg twice daily).

C. surgical thymectomy To reduce the need for long-term immunosuppressive therapy, is often advised in patients under 60 years of age.

Advanced Cases

In advanced cases requiring mechanical ventilation, there are two treatment options:

A. Plasmapheresis to clear pathological antibodies from the bloodstream.

B. IV immunoglobulin G (0.4 to 2 gm/kg/day for 2 to 5 days) to neutralize the pathologic antibodies.

C. Both approaches are equally effective, but plasmapheresis produces a more rapid response.

Emergency Department Care

- Patients with myasthenia gravis who are in respiratory distress may be experiencing:
  - a myasthenic crisis or
  - a cholinergic crisis.
- Before these possibilities can be differentiated, ensuring adequate ventilation and oxygenation is important.
- Patients with myasthenic crisis can develop apnea very suddenly, and they must be observed closely.
- Evidence of respiratory failure may be noted through ABG determination, pulmonary function tests, or pulse oximetry.

Airway maneuvers

- Open the airway by suctioning secretions after positioning the jaw and tongue. Administer high-flow oxygen, and measure oxygen saturation by pulse oximetry.
- If respirations remain inadequate, ventilate by bag-valve mask while preparing to intubate. In the patient without an intact gag reflex, an oral airway may be placed.

Endotracheal intubation

- Rapid sequence intubation should be modified, because depolarizing paralytic agents (e.g., succinylcholine) have less predictable results in patients with myasthenia gravis.
- The relative lack of ACh receptors makes these patients relatively resistant to succinylcholine; therefore, higher doses must be used to induce paralysis. Once paralysis is achieved, it may be prolonged.

- A rapid-onset, nondepolarizing agent (ie, rocuronium, vecuronium) is the preferred paralytic agent for these patients.
- Although nondepolarizing agents delay the onset of paralysis, compared with succinylcholine, these medications do not result in unwanted prolonged paralysis. Following paralysis, intubation is accomplished as usual. ABG sampling guides ventilator settings.

- Preliminary studies suggest that bilevel positive airway pressure (BiPAP) can prevent intubation in patients with myasthenic crisis without overt hypercapnia and should be considered in the patient who can be closely monitored. Hypercapnia present at the time of BiPAP initiation can predict failure and the need to proceed to endotracheal intubation.

Investigation and treatment

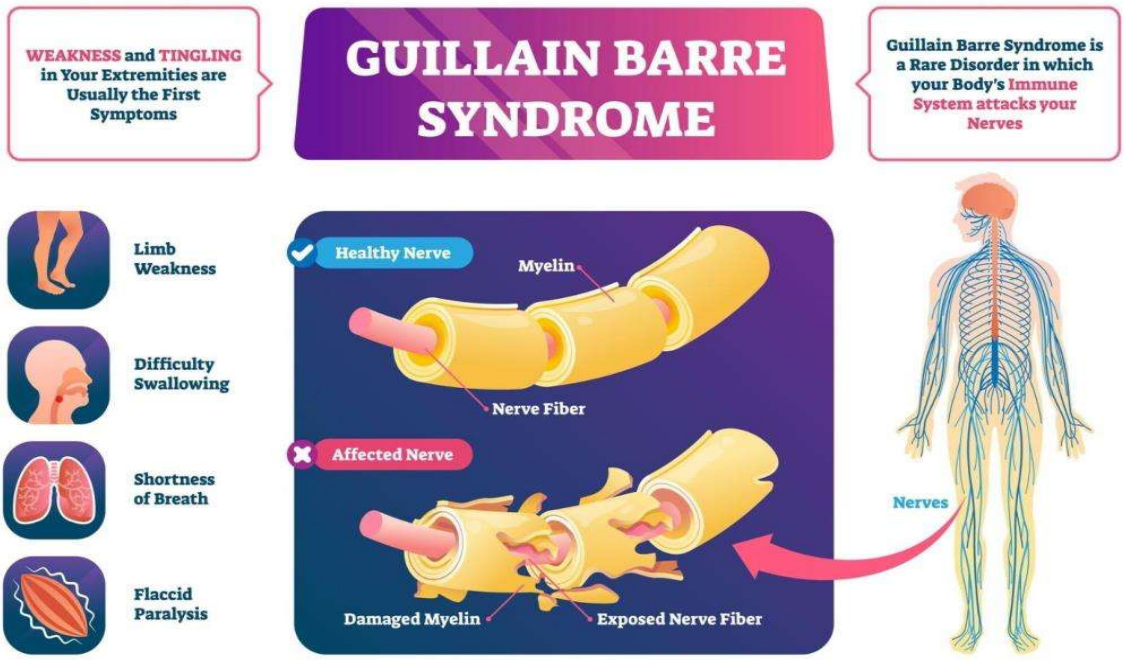
- Once the airway is secured, investigation into the cause of the exacerbation of myasthenia gravis may proceed, with the most common reason for an exacerbation being infection, followed by inadequate treatment with cholinesterase inhibitors. However, up to 30% of patients will not have an identified cause of their exacerbation.

Difference between Myasthenic Crisis and Cholinergic Crisis

Myasthenic Crisis	Cholinergic Crisis
Under medication	Overmedication
Temporary improvement of symptoms with administration of Edrophonium	Symptoms improve with administration of anticholinergics (Atropine)
Heart rate increased	Heart rate decrease
Respiratory distress	Abdominal cramps
Pupil : Mydriasis	Pupil: Miosis
Increased Blood pressure	Decreased blood pressure
Normal secretion	Increased secretion

B. Guillain-Barré Syndrome(GBS)

- is a subacute inflammatory demyelinating polyneuropathy that often follows an acute infectious illness (by 1 to 3 weeks).
- An immune etiology is suspected.

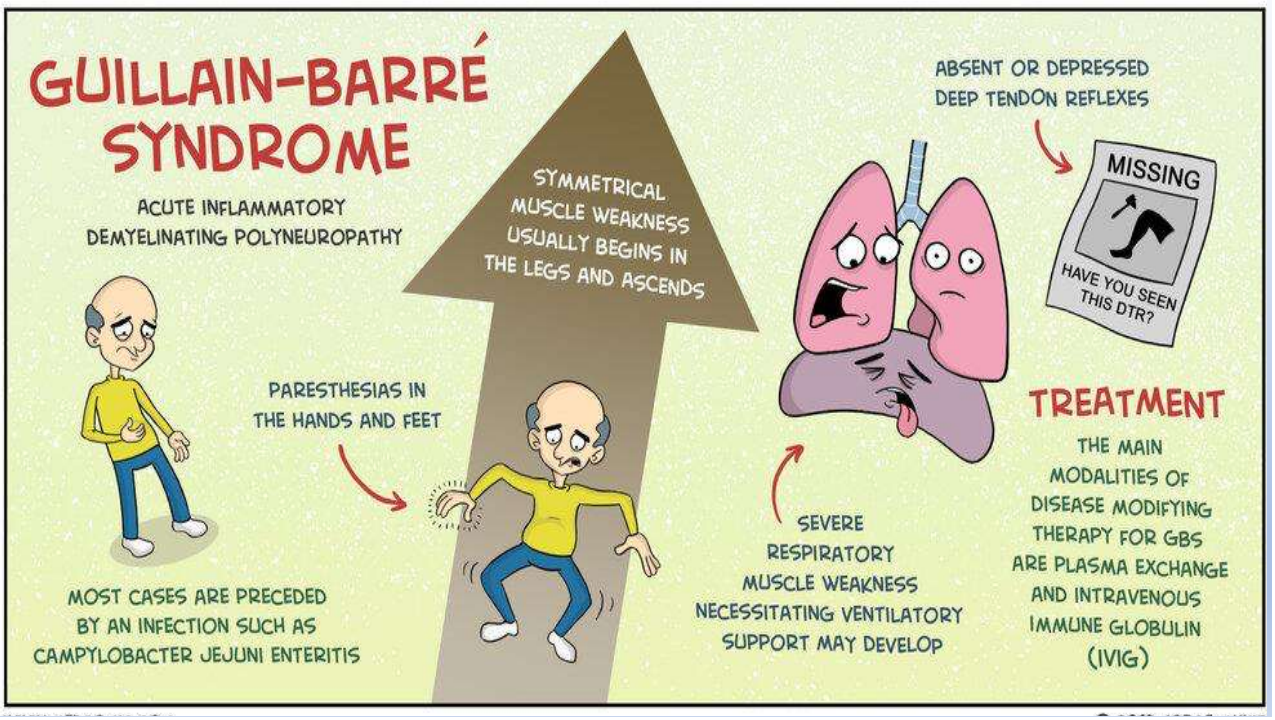


Clinical Features

A. GBS presents with distal paresthesia and symmetric limb weakness that evolves over a period of a few days to a few weeks.

B. Progression to respiratory failure occurs in 25% of cases , and autonomic instability can be a feature in advanced cases

C. The condition resolves spontaneously in about 80% of cases, but residual neurological deficits are common .



Diagnosis

- The diagnosis of GBS is based on the:
- clinical presentation (paresthesias and symmetric limb weakness)
- nerve conduction studies (slowed conduction)
- CSF analysis (elevated protein in 80% of cases).
- The features that distinguish GBS from myasthenia gravis are shown in Table below.

Table 41.3 Comparative Features of Myasthenia Gravis and Guillain-Barré Syndrome

Features	Myasthenia Gravis	Guillain-Barré Syndrome
Ocular weakness	Yes	No
Fluctuating weakness	Yes	No
Bulbar weakness	Yes	No
Deep tendon reflexes	Intact	Depressed
Autonomic instability	No	Yes
Nerve conduction	Normal	Slowed

Treatment

- Treatment is mostly supportive, but in advanced cases with respiratory failure, plasmapheresis or IV immunoglobulin G (0.4 g/kg/day for 5 days) are equally effective in producing short-term improvement. Immunoglobulin is often preferred because it is easier to implement.

MCQ TEST

1- All the following are clinical features of MG except one
   a) Ocular weakness
   b) Automatic instability
   c) Bulbar weakness
   d) Swallowing difficulty
   e) Change of voice
2- Cholinergic crisis(all true except one)
   a) Hypotension
   b) Bradycardia
   c) Increase secretion
   d) Medyriasis
   e) Abdominal crump
3- All the following are management of MG
   a) Plasmapharesisi
   b) Immunoglobulin

c) A and B
d) Non of the above
e) Edrophonium

4- Clinical features og Gullian Bare Syndrome (all true except one
a) Normal deep tendon reflexes
b) Ascending muscle weakness.
c) Hypotension
d) distal paresthesia's
e) seizures

5- patient with MG is resistant to
a) neostigmine
b) succinylcholine
c) rocuronium
d) atracuriuom
e) epinephrine