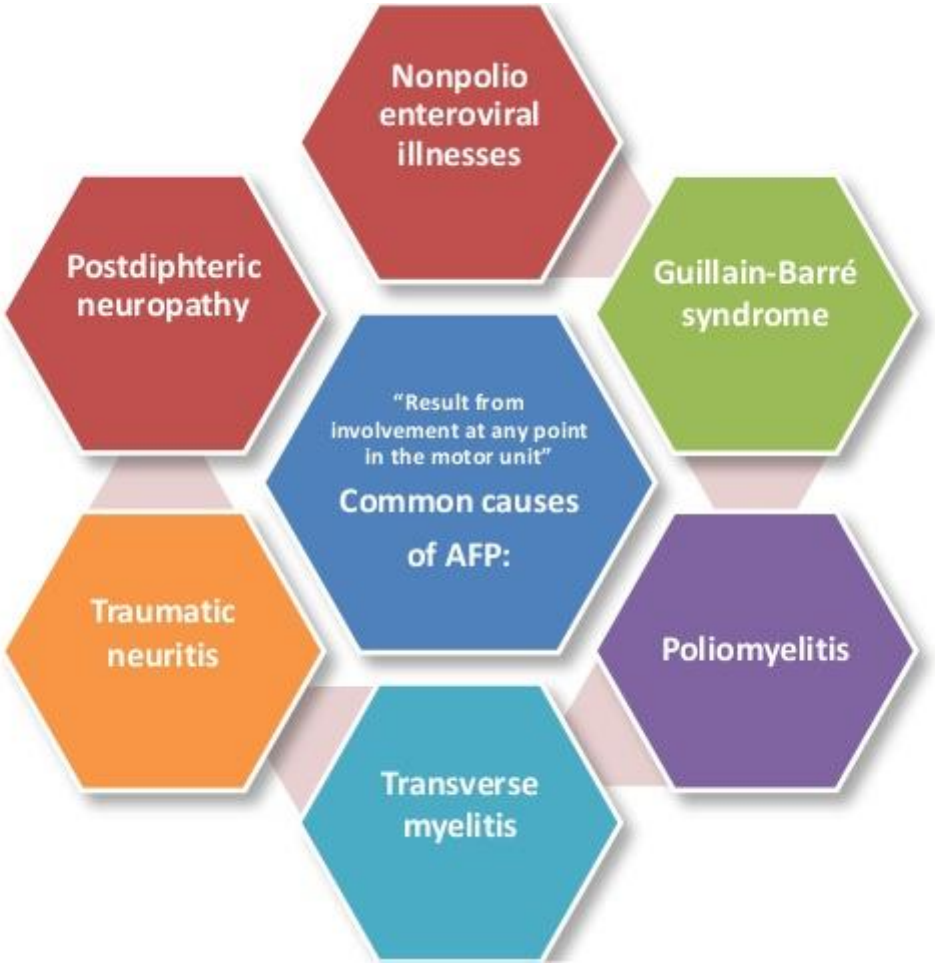
Aprroch to Flaccid paralysis

By : Reza Rikhtegar (MD)


Nonpolio enteroviral illnesses
Postdiphteric neuropathy
Guillain-Barré syndrome
"Result from involvement at any point in the motor unit"
Common causes of AFP:
Traumatic neuritis
Poliomyelitis
Transverse myelitis

Differential Diagnosis of AFP

- **SPINAL CORD**
- **Anterior horn cell disease**
- **PERIPHERAL NERVE**
- **Disorders of neuromuscular transmission**
- **MUSCLE**
- **Systemic disease**

SPINAL CORD

- ***Demyelinating diseases***
- transverse myelitis
- ***Cord compression***
- tumour
- trauma
- paraspinal abscess
- haematoma
- vascular malformation with thrombosis/bleeding
- ***Ischaemic cord damage***
- spinal cord stroke
- anterior spinal artery syndrome
- peri-operative complication

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PERIPHERAL NERVE

- ***Unilateral:***
- Enteroviral infection
- local trauma
- Focal mononeuropathy

- ***Bilateral:***
- Guillain Barré syndrome
- Acute toxic neuropathies (heavy metals, snake toxin)
- Neuropathies of infectious diseases (diphtheria)

Disorders of neuromuscular transmission

- Myasthenia gravis
- Botulism
- Insecticide (organophosphate poisoning)
- Tick bite paralysis
- Snake bite

MUSCLE

- Polymyositis
- post viral myositis
- periodic paralysis
- toxic myositis (Corticosteriods and blocking agents)
- Mitochondrial diseases (infantile type)

Investigations

- Spinal cord MRI.
- CSF study.
- Nerve conduction study.
- OTHERS

How to differentionate among

- Polio
- Guillain-Barré syndrome
- Traumatic neuritis
- Transverse Myelitis

	Polio	Guillain-Barré syndrome	Traumatic neuritis	Transverse myelitis
Installation of paralysis	24 to 48 hours onset to full paralysis	From hours to ten days	From hours to four days	from hours to four days
Fever at onset	High, always present at onset of flaccid paralysis, gone the following day	Not common	Commonly present before, during and after flaccid paralysis	rarely present
Flaccid paralysis	Acute, usually asymmetrical, principally proximal	Generally acute, symmetrical and distal	Asymmetrical, acute and affecting only one limb	acute, lower limbs, symmetrical
Muscle tone	Reduced or absent in affected limb	Global hypotonia	Reduced or absent in affected limb	Hypotonia in lower limbs
Deep-tendon reflexes	Decreased to absent	Globally absent	Decreased to absent	Absent in lower limbs early hyperreflexia late

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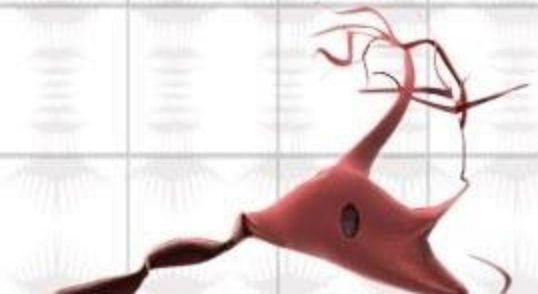
	Polio	Guillain-Barré syndrome	Traumatic neuritis	Transverse myelitis
Bladder dysfunction	Absent	Transient	Never	Present
Nerve conduction Velocity	Abnormal: anterior horn cell disease (normal during the first 2 weeks)	Abnormal: slowed conduction, decreased motor amplitudes	Abnormal: axonal damage	normal or abnormal, no diagnostic value
EMG	Abnormal	Normal	Normal	Normal
Sequel at three months and up to a year	Severe, asymmetrical atrophy, skeletal deformities developing later	Symmetrical atrophy of distal muscles	Moderate atrophy, only in affected lower limb	flaccid diplegia atrophy after years

Involvement

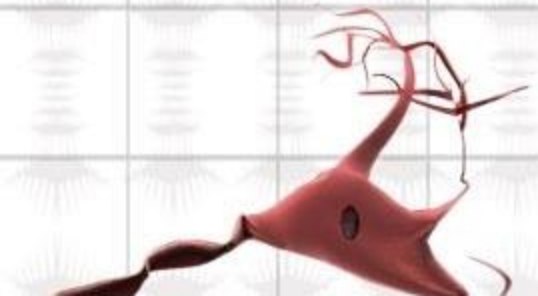
- AHC:
  1- acute poliomyelitis
  2- acute transverse myelitis

- Peripheral Nerves:
  1- roots: GBS (post-infectious)
  2- toxins: Diphteria, porphyria



Involvement

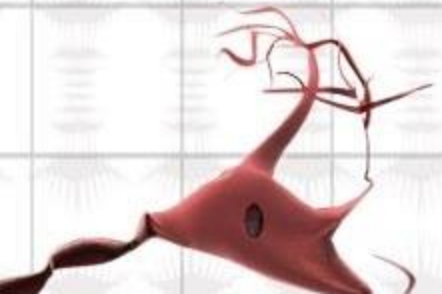
- N-M junction:
  botulinum toxin
  tick toxin
- Metabolic:
  Periodic paralysis
- Muscular:
  myositis



Guillain Barre Syndrome GBS

- The most common cause of acute flaccid paralysis (AFP) among infants.

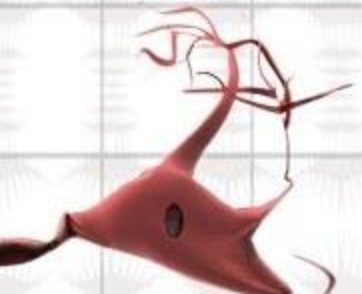
- Age : any including newborn
- Sex : any (male > female)



Guillain-Barre' Syndrome

- Post-infectious polyneuropathy; ascending polyneuropathic paralysis

- An acute, rapidly progressing and potentially fatal form of polyneuritis



Pathophysiology

Autoimmune disorder (T cell sensitization)

cause of demyelination

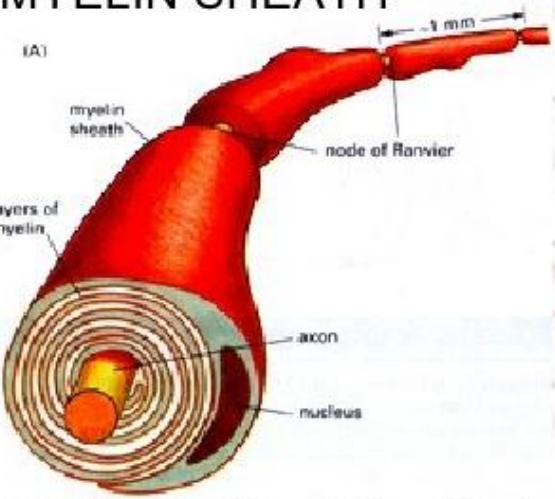
Due to attack of the myelin sheath of nerves by:

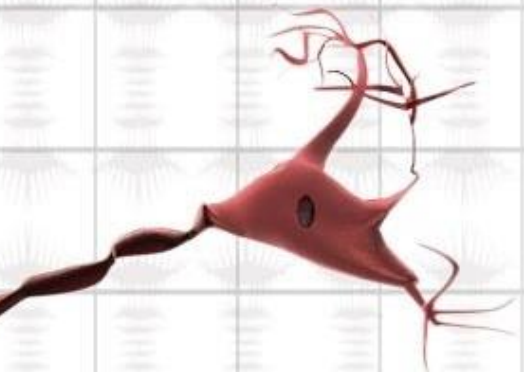
- **antibodies (Ig M, Ig G)**
- **white blood cells** (macrophages)
- **Complement activation** on the outer surface of myelinated fibers

Because (POST-)

Virus/Bacteria share antigenic sites with axons & peripheral nerve sheath or both

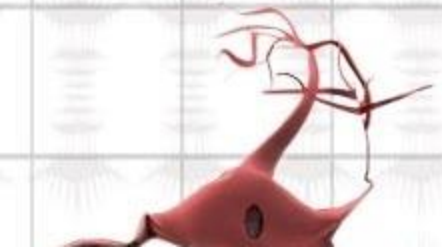
MYELIN SHEATH





pathophysiology

- inflammation causes leakage of proteins into the CSF causing raised CSF proteins without pleocytosis
- Can involve the peripheral nerves, cranial nerves,dorsal roots, dorsal root ganglia & sympathatic chain



Preceding Events : (1-3 WEEKS)

- **Respiratory infections :**

1- Viral: CMV, EBV, Varicella virus , influenza virus

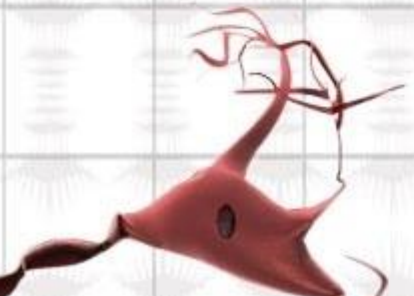
2- Bacterial: Mycoplasma pneumoniae, H influenza

- **Gastrointestinal infections :** Campylobacter-jejuni (Bloody GE)

- **Vaccinations**
- Post surgery

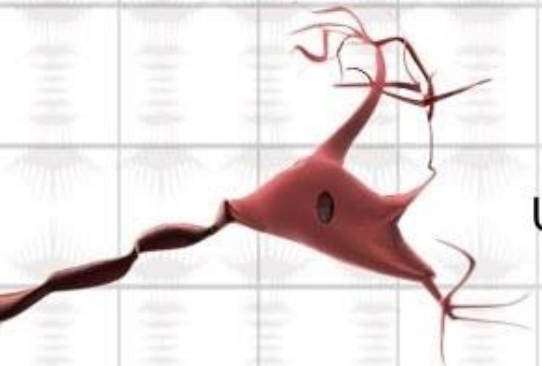
Aetiology

- Mycoplasma
  - Hepatitis B
  - CMV
  - EBV
  - Measles
  - Mumps
  - Echovirus
  - Cocksakie virus
  - Influenza virus
- Varicella virus
  - Compylobacter jejuni



classification of GBS
(Clinical, Pathological & neurophysiological)

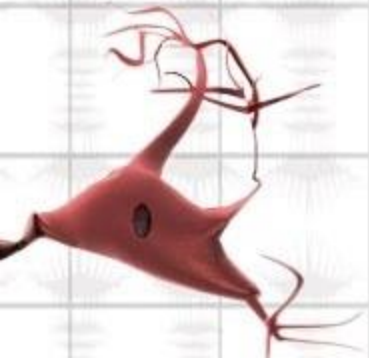
- **1-Classic type (mixed): Acute inflammatory demyelinating polyradiculo-neuropathy (AIDP)**
- **2- Pure motor GBS***
- 3- Pure sensory GBS
- 4- Pure pandysautonomia
- 5-- Miller-Fisher syndrome (hypotonia, ophthalmoplegia, ataxia)



NB., Pure Motor GBS*
Usually Post Campylobacter-.jejuni infection

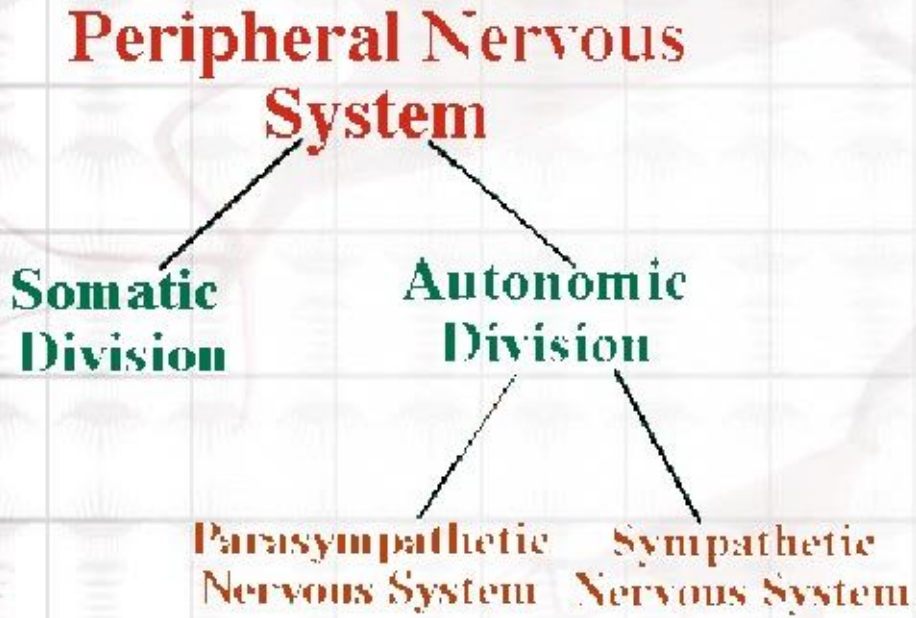
C/P:

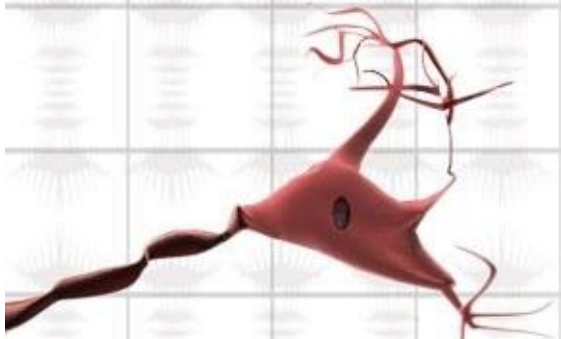
- 1- IP (afrebrile) : 1-3 weeks
- 2- CP: motor , sensory , autonomic
- 3- Serious Association



Guillain-Barre' Syndrome

- Affects the peripheral nervous system
-





Symptoms and Signs (Typical GBS)

Motor : 1- Symmetric acute progressive ascending weakness <4 wks, starting in LL

2- Areflexia or hyporeflexia

3- Atonia or hypotonia

Symptoms and Signs (Typical GBS)

Sensation : 1- C/O pain as hyperthesia or cramps
2- O/E loss of pain sensation (hypothesia) in feet/hands
3- Both C/O, and O/E

Characteristic “3A”triad:

- **ascending weakness : a- bilateral symmetrical weakness**
  b- usually start in LL, then UL
  c-then, might be affected :
  i- cranial nerves (Brain stem) :
  including glosssopharyngeal and vagus nerves (difficulty of swallowi
  even of fluid and water) and III, IV, VI cranial nerves (eye muscles in
  Miller Fisher variety), VII Facial nerve (unilateral or bilateral), and the
  respiratory muscles
  ii- respiratory muscles
  iii- phrenic nerves (diaphragm)
- **areflexia (Hallmark)**
- **atonia (hypotonia)**

CLINICAL VARIANTS

1–Polyneuritis cranialis

Cranial nerve involvement

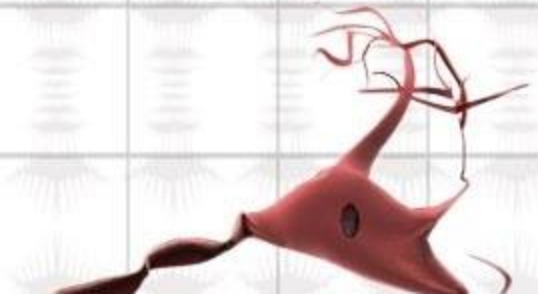
2–Miller fisher syndrome

Ophthalmoplegia, ataxia, areflexia

3–Chronic progressive GBS

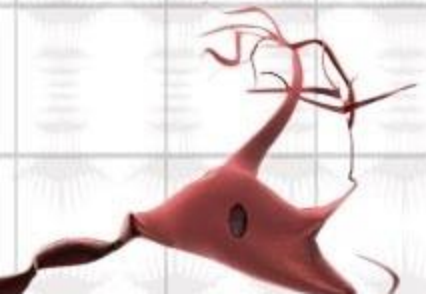
Symptoms persisting more than 6 week

4- Chronic relapsing GB



Differentiation from spinal cord syndrome

- ♦Loss of arm reflexes
- ♦Absence of sensory level
- ♦Lack of spinal tenderness
- ♦Normal bowel and bladder function



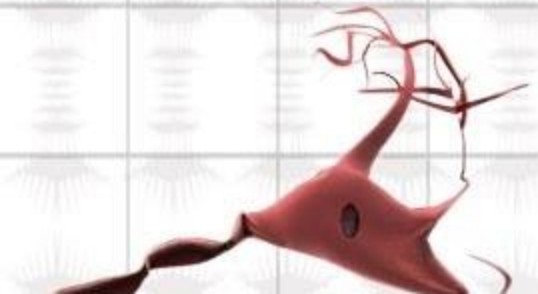
Investigations

Early

Nerve Conduction Velocity (NCV) abnormality

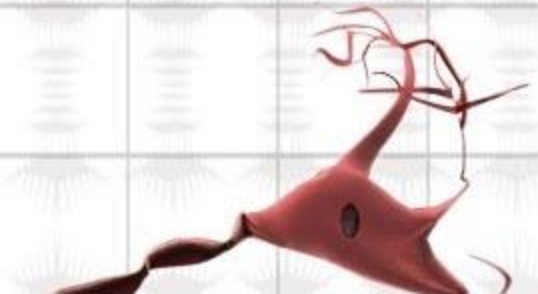
AFTER 1ST WEEK

- Late : CSF study : albuminocytogenic dissociation



1- NCV/EMG:

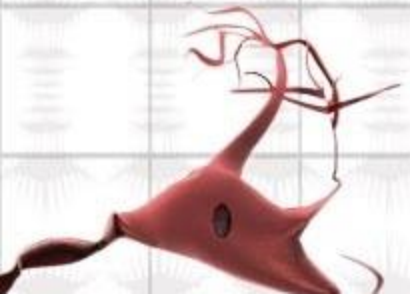
- **i- Early :**Delayed or absent F waves or H reflexes
- ii- slow or block of Nerve conduction velocity
- iii- normal EMG/ extensive fibrillation showing denervation



2- CSF: Albuminocytologic dissociation (Froin Syndrome)

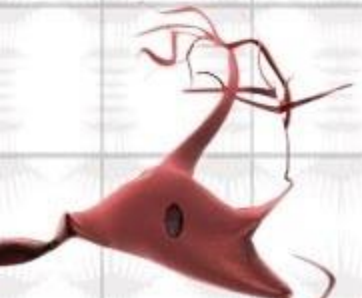
- i- Increased CSF protein with normal cells
- ii- might be normal CSF during 1st week
- ii- usually +ve after 2 weeks of onset

Differential Diagnosis of cytoalbuminous dissociation



DD

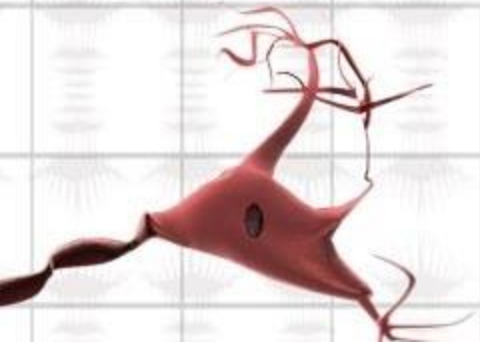
- 1- GBS
- 2- poliomyelitis
- 3- Diphteria polyneuritis
- 4- spinal cord compression
- 5- transverse myelitis
- 6- infratentorial tumor
- 7- venous sinus thrombosis
- 8- lead poison
- 9- botulism



Treatment

Hospitalization

- 1- **General care**
- 2-- **Specific treatment**
- 3- complication treatment



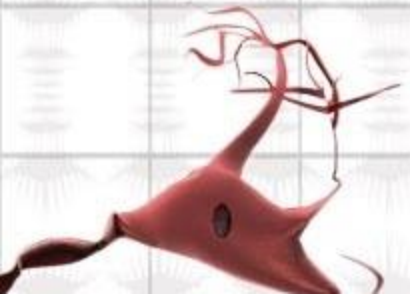
Specific treatment

- **i- IV immunoglobin**: 2 gm/kg treatment

  *at a dose of 0.4 g/kg/day for 5 consecutive days or

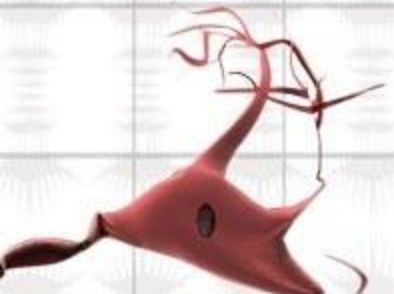
  * 1gm/kg/day for 2 days

- **ii- plasmapheresis**: 5 exchanges of 50 ml plasma/ kg on alternate days (10 days course).
- **iii- both i and ii**



Transverse Myelitis:

- ? of immunological disorder
- C/P : of AFP (acute onset of flaccid hypotonic weakness) with the following characters:
  - LL paralysis : paraplegia with areflexia
  - with sensory level of loss of sensation
  - Later : hyperreflexia

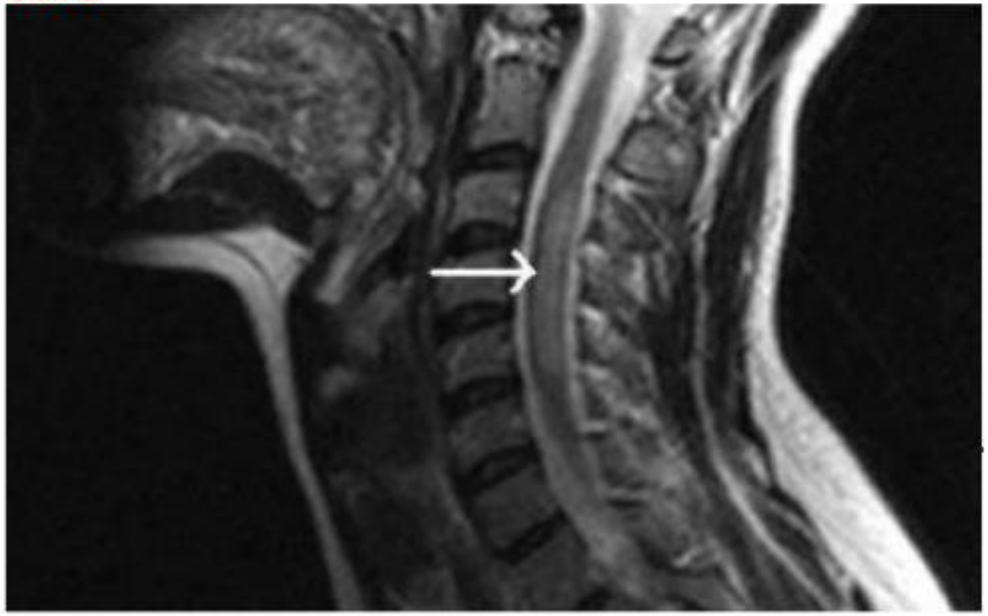


Transverse Myelitis

- Transverse myelitis is a condition characterized by rapid development of both motor and sensory deficits .

Transverse Myelitis

- disorder caused by **inflammation of the spinal cord**

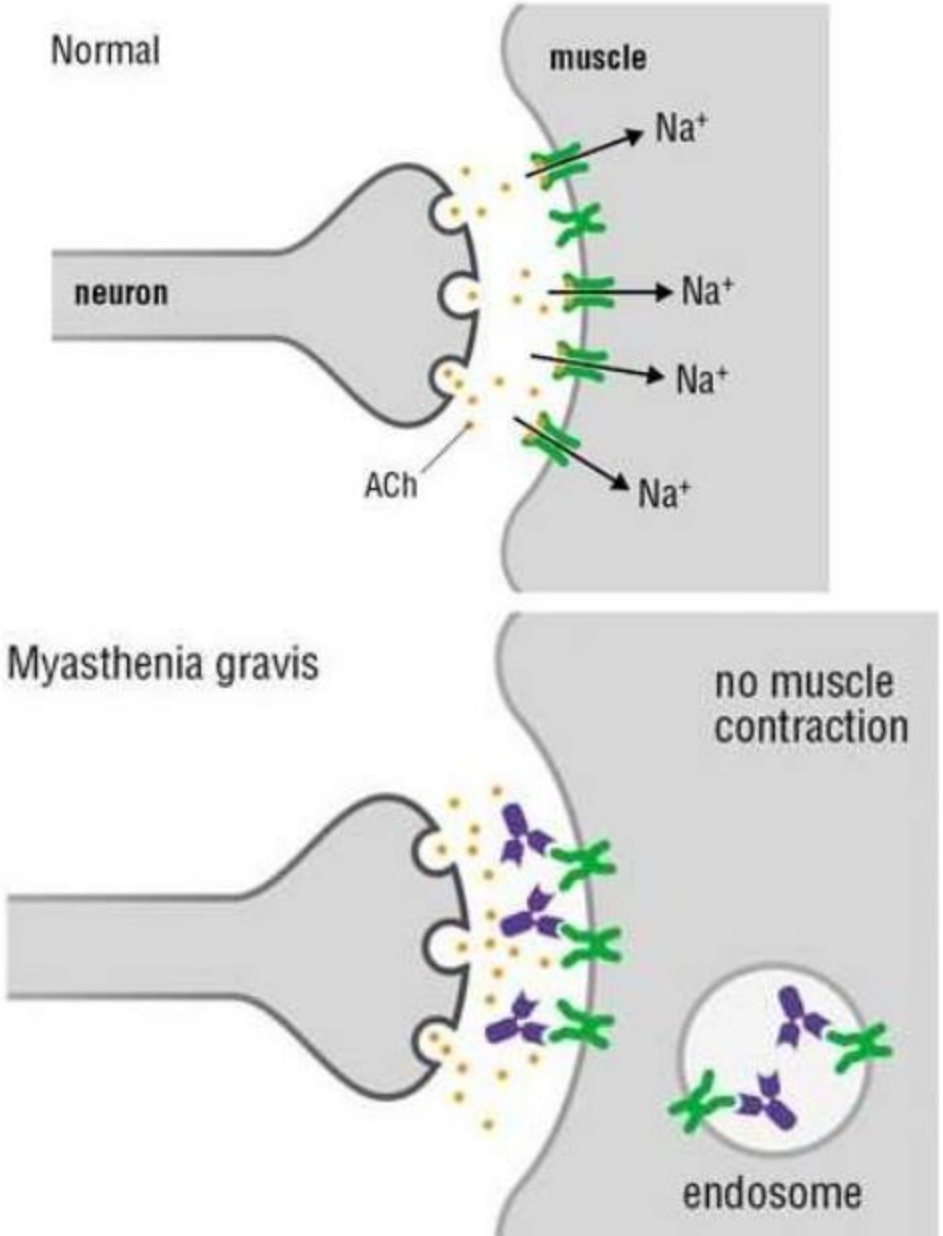


MYASTHENIA GRAVIS

- An autoimmune of neuromuscular junction.
- Weakness of skeletal muscles.
- Fatigability on exertion.

MYASTHENIA GRAVIS

- PATHOPHYSIOLOGY;



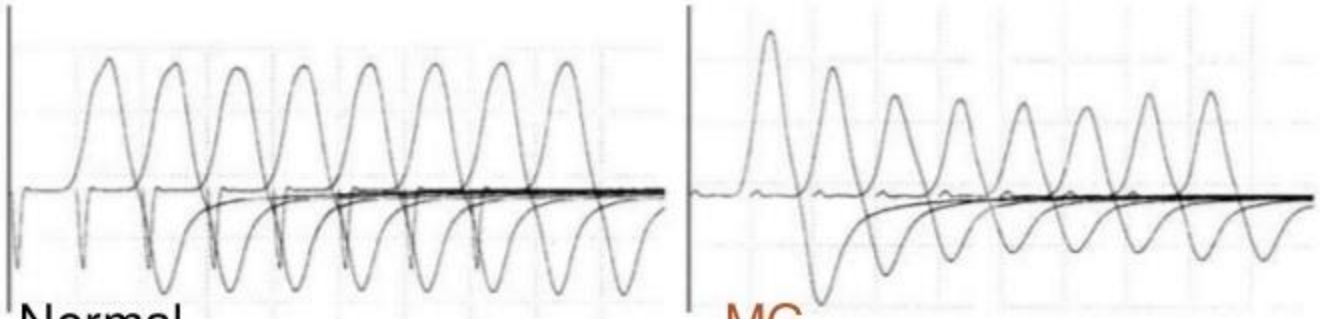
MYASTHENIA GRAVIS

- Myasthenia Gravis is often associated with:
  - Hashimoto thyroiditis.
  - Some collagen vascular diseases.
  - Thymoma (*mostly with adults; rarely in children*).

  - **Post-infectious myasthenia:**
    - Affects children.
    - Follows infection with varicella zoster.
    - Transient.

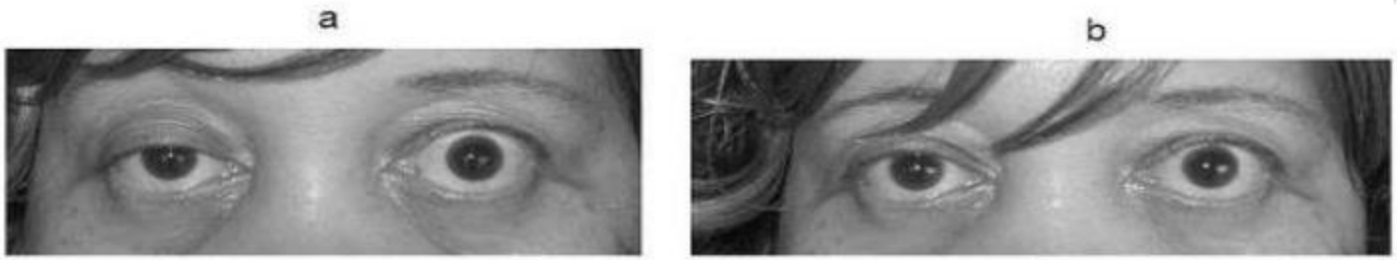
MYASTHENIA GRAVIS
DIAGNOSIS

- DIAGNOSTIC STUDIES;
- EMG.
  - *More diagnostic than Bx.*



Normal. MG.

- Anti-Ach Abs.



- Tensilon test (Edrophonium Test)
  - *Ptosis and ophthalmoplegia improve within a few seconds, and fatigability of other muscles decreases.*

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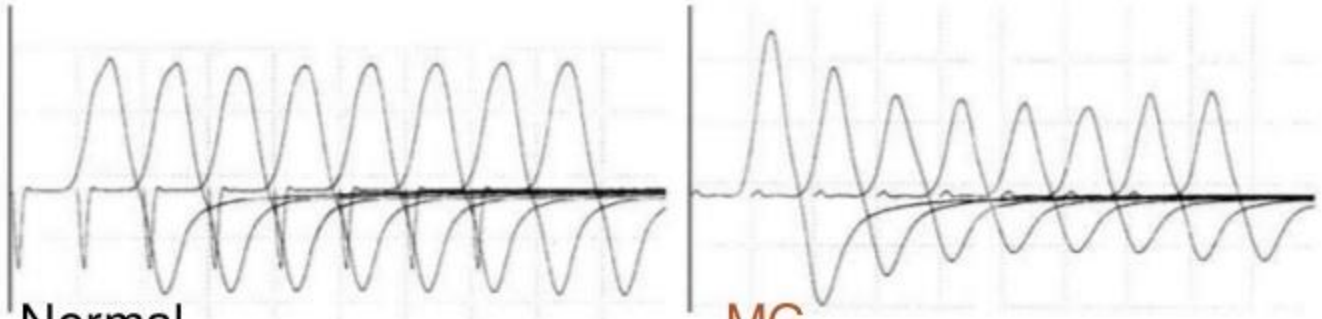

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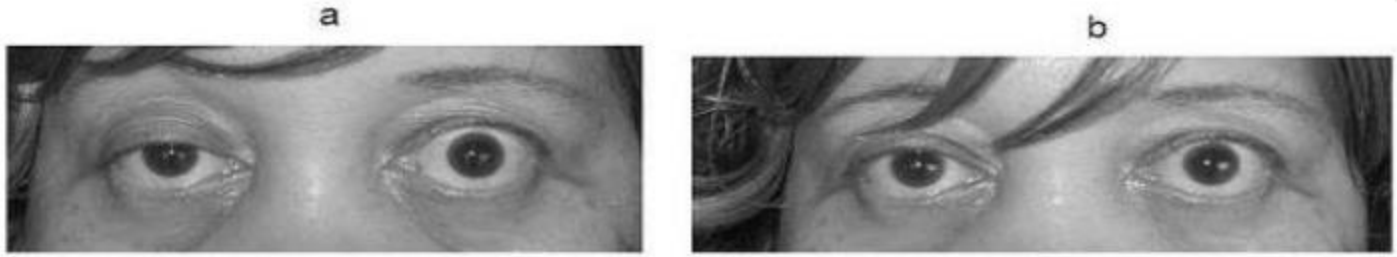

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Hypokalemic Periodic Paralysis