Acute flaccid paralysis (AFP)

By

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Acute flaccid paralysis (AFP) is a clinical syndrome characterized by **rapid onset of weakness**, including (less frequently) weakness of the muscles of respiration and swallowing, **progressing to maximum severity** within **several days to weeks.**

The term "**flaccid**" indicates the *absence of spasticity or other signs of disordered central nervous system motor tracts* such as *hyperreflexia, clonus, or extensor plantar responses*

AFP is a complex clinical syndrome with a broad array of potential etiologies.

 Accurate diagnosis of the cause of AFP has profound implications for therapy and prognosis.

 If untreated, AFP may not only persist but also lead to death due to failure of respiratory muscles.

Clinical Approach to AFP

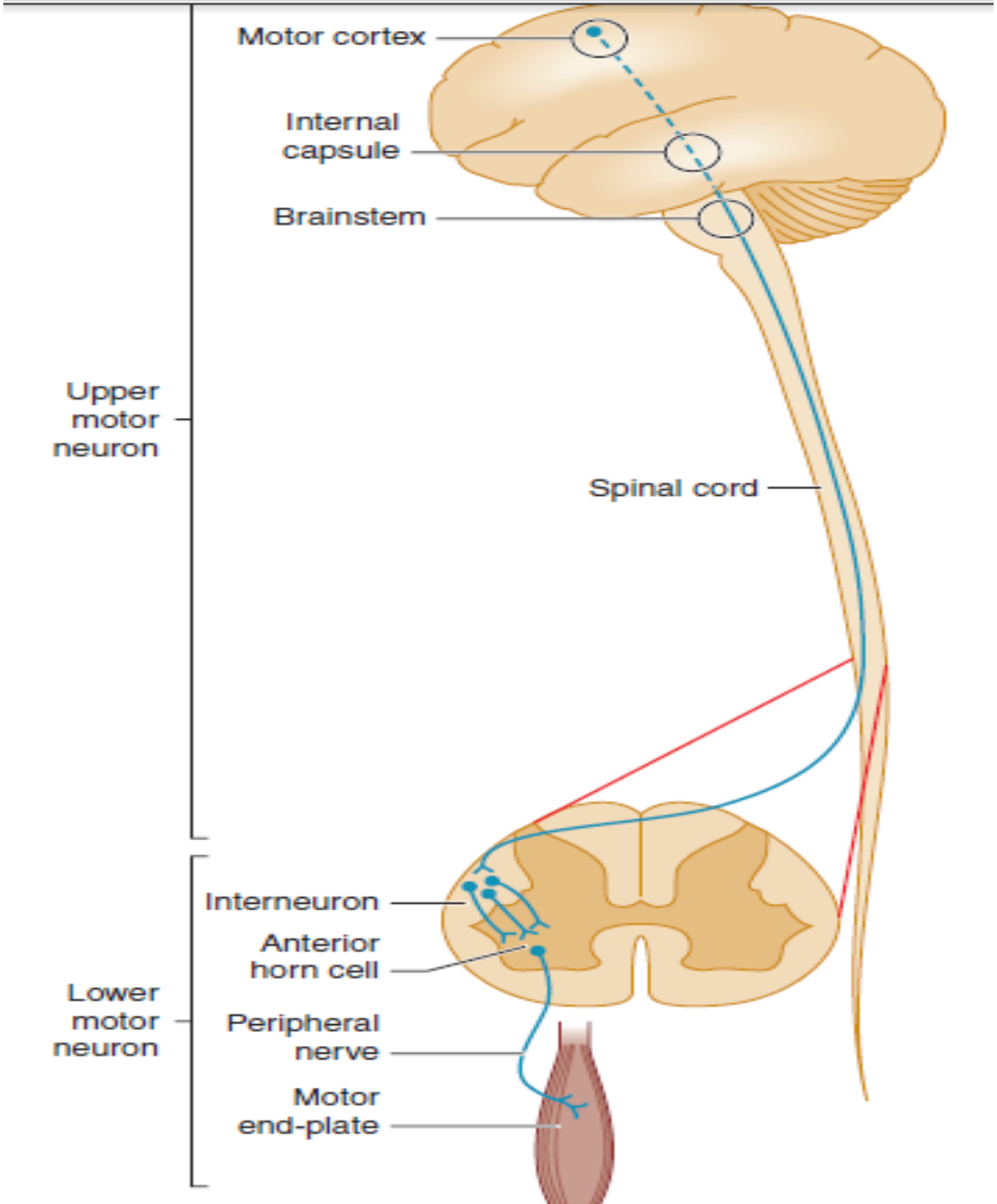
Each case of AFP is a clinical emergency and requires immediate examination.

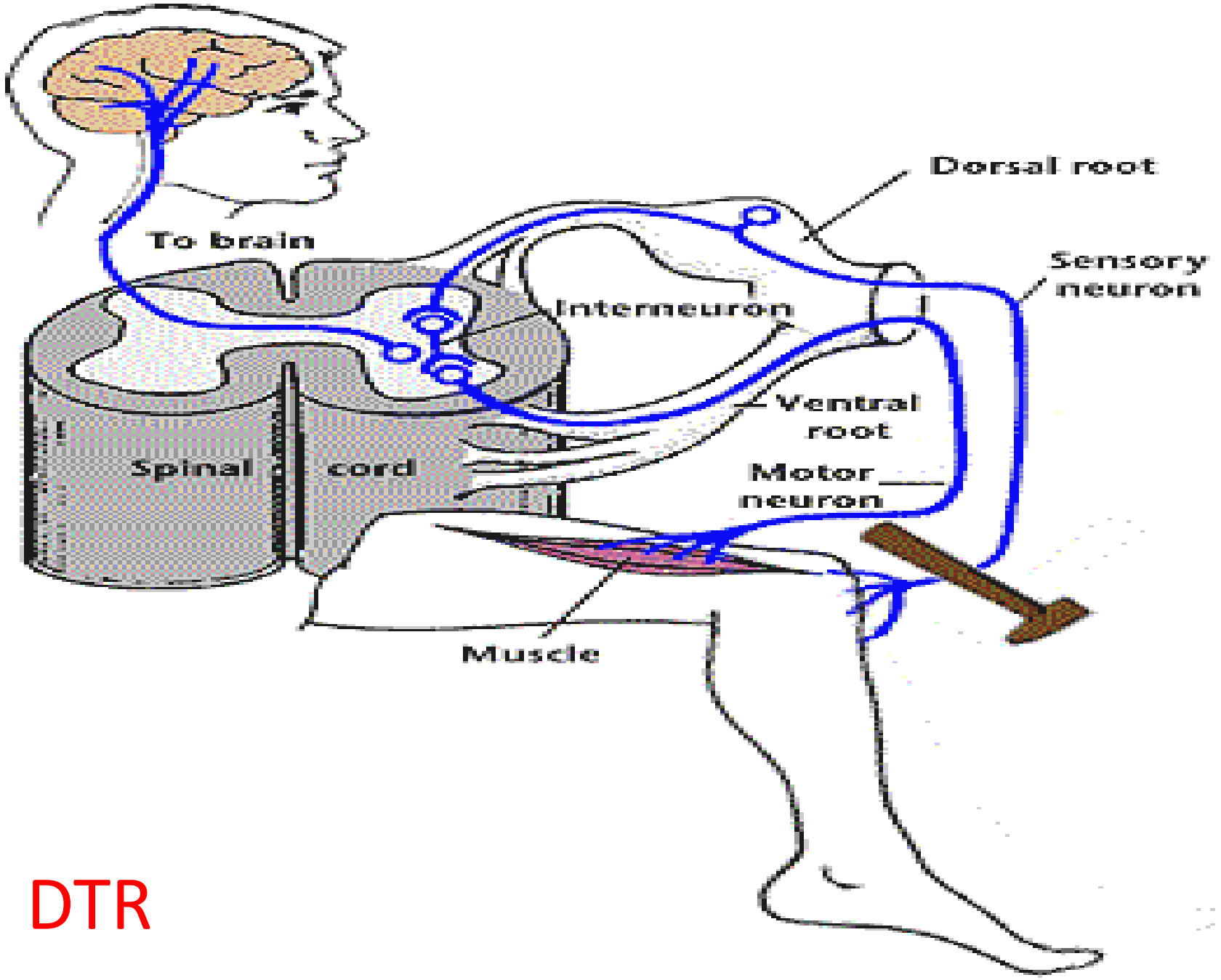
 For all cases, a **detailed clinical description** of the symptoms should be obtained, including fever, myalgia and distribution, timing, and progression of paralysis.

 The **symptoms** of paralysis may include gait disturbance or weakness in one or several extremities

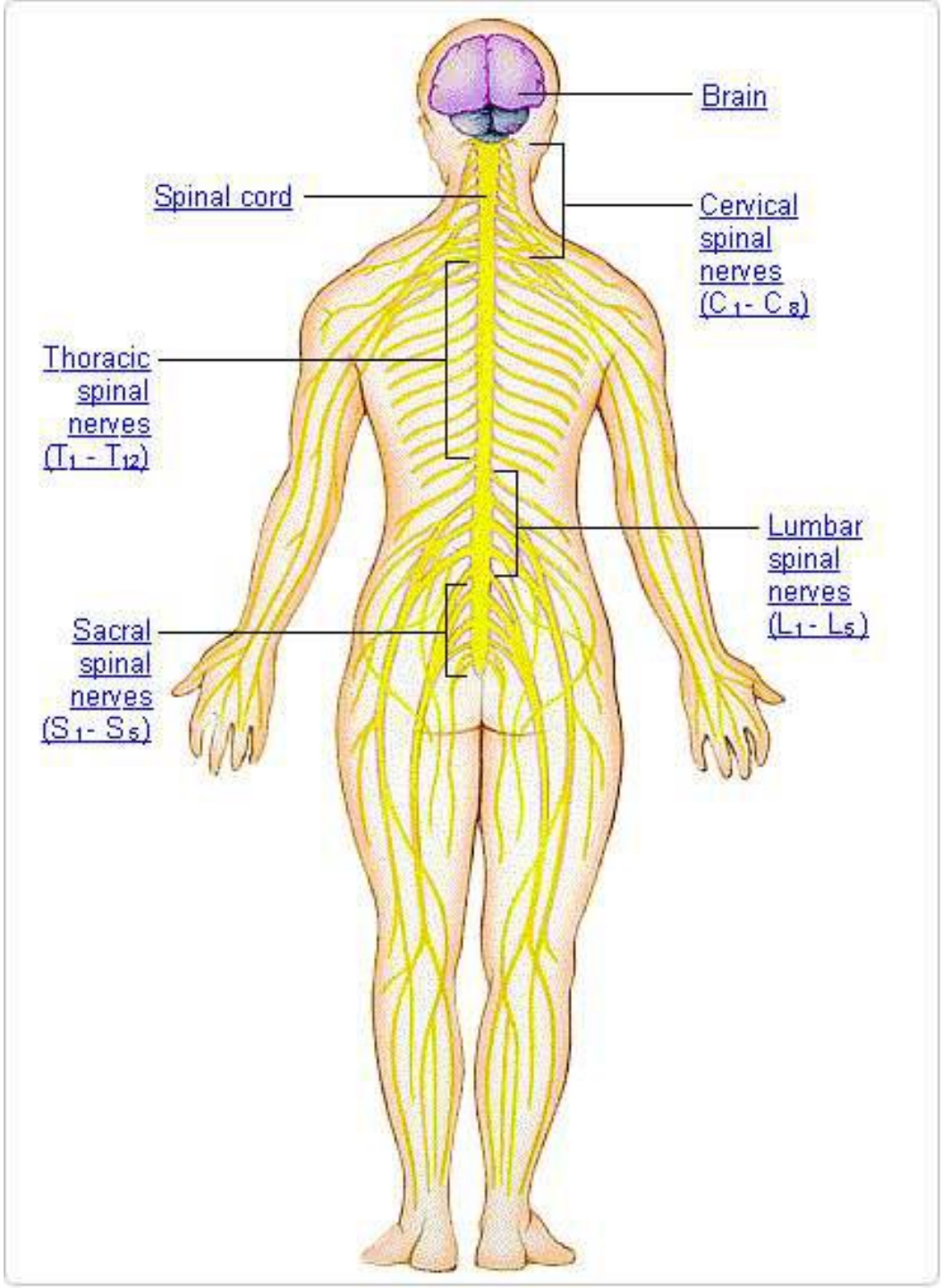
Electrophysiologic studies are very important for determining the diagnosis and prognosis of lower motor neuron disease

 Nerve conduction velocity and **electromyographic studies** are used to differentiate *demyelinating* neuropathies from *axonal* neuropathies


Motor cortex
Internal capsule
Brainstem
Upper motor neuron
Spinal cord
Interneuron
Anterior horn cell
Lower motor neuron
Peripheral nerve
Motor end-plate

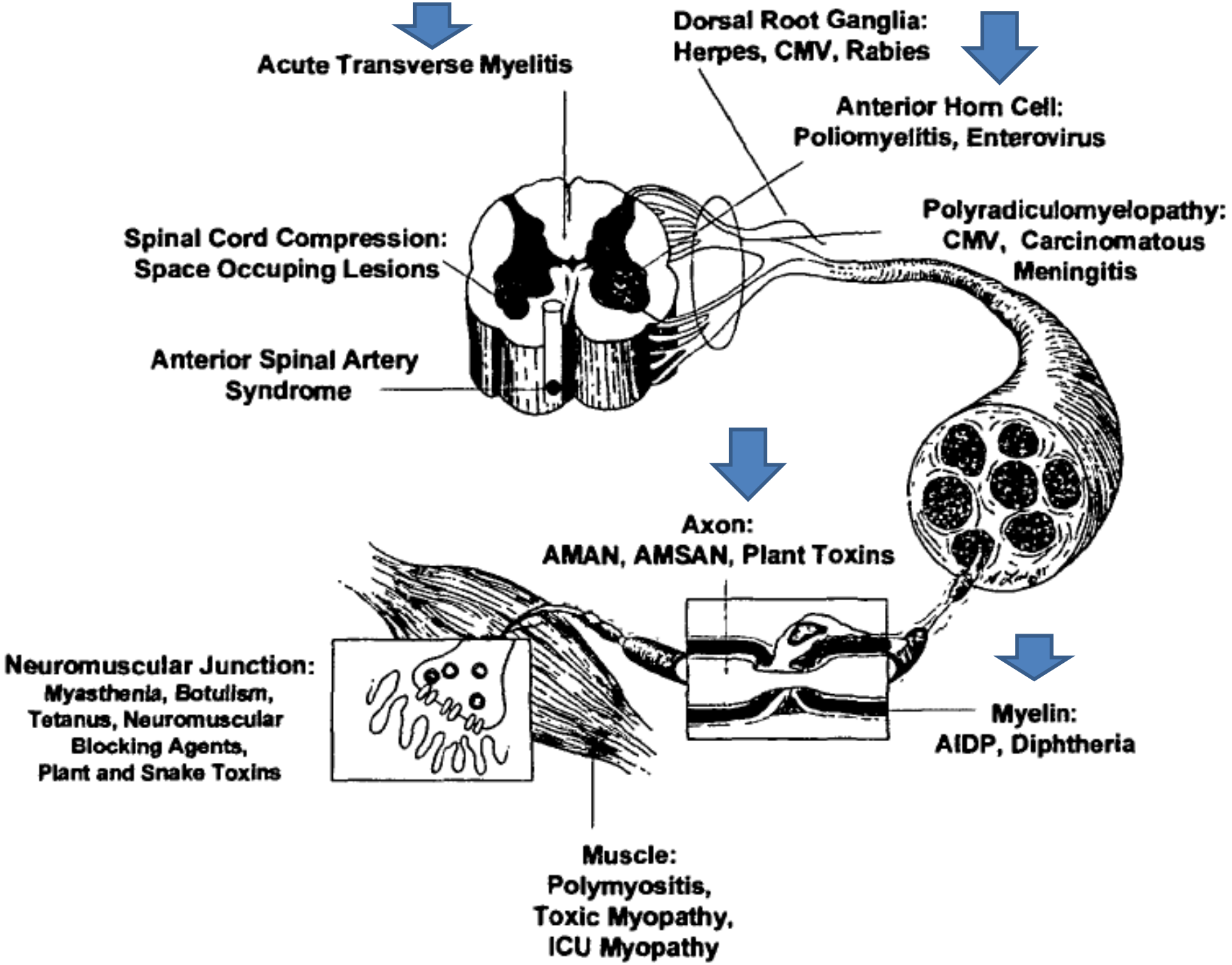

Dorsal root
To brain
Sensory neuron
Interneuron
Ventral root
Spinal cord
Motor neuron
Muscle

DTR


Brain
Spinal cord
Cervical spinal nerves (C1 - C8)
Thoracic spinal nerves (T1 - T12)
Lumbar spinal nerves (L1 - L5)
Sacral spinal nerves (S1 - S5)

Differential Diagnosis

- Guillain –Barré Syndrome
- Acute Transverse Myelitis
- Poliomyelitis
- Hypokalemic Periodic Paralysis


Acute Transverse Myelitis
Dorsal Root Ganglia:
Herpes, CMV, Rabies
Anterior Horn Cell:
Poliomyelitis, Enterovirus
Spinal Cord Compression:
Space Occuping Lesions
Polyradiculomyelopathy:
CMV, Carcinomatous
Meningitis
Anterior Spinal Artery
Syndrome
Axon:
AMAN, AMSAN, Plant Toxins
Neuromuscular Junction:
Myasthenia, Botulism,
Tetanus, Neuromuscular
Blocking Agents,
Plant and Snake Toxins
Myelin:
AIDP, Diphtheria
Muscle:
Polymyositis,
Toxic Myopathy,
ICU Myopathy

Guillain-Barre syndrome(GBS)

the commonest cause of acute flaccid paralysis (Acute inflammatory demyelinating polyradiculoneuropathy (AIDP))

Symmetrical progressive ascending weakness, **areflexia**, variable **sensory complaints**, and **elevated CSF protein without pleocytosis .**

Pathophysiology of GBS

-Acquired, monophasic

-Immune mediated disease

-No known genetic factor

-Two thirds of cases follow a **respiratory or gastrointestinal infection**

-Campylobacter infection is the most common, but other organisms include CMV, EBV, HSV, enteroviruses, …

Clinical Features of GBS

-Two to four weeks after a **benign febrile illness**

-Common presentations are paresthesias in the fingers and toes, pain is a common presentation in children (79%), particularly *low back pain*

-Symmetrical weakness in the lower extremities, that ascends over hours to days to involve the arms, and in ***severe*** cases respiratory muscles

Cranial nerves are affected in **30%** of the cases, **most commonly** the facial nerve with bilateral facial weakness

More than 90% of patients reach the nadir of their function within **2-4 weeks**

Physical Examination in GBS

Symmetrical weakness with **diminished or absent reflexes**

Vibration and **position sensation** are affected in **40%** of cases

50% of patients have evidence of **autonomic dysfunction :**

- *Cardiac dysrythmias*
- *Orthostatic hypotension, hypertension*
- *Paralytic ileus*
- *Bladder dysfunction*

Diagnosis of GBS

Cerebrospinal fluid:

- *After the first week* of symptoms, CSF typically reveals normal pressure, normal cell count, and **elevated protein**

Electrophysiologic studies:

- **Most specific and sensitive tests** for diagnosis
- Evidence of evolving multifocal demyelination
- Normal studies after 10 days of illness make the diagnosis of GBS unlikely

GBS Management

- Expectant with **mild cases**
- Immune modulatory therapy for **rapidly progressive cases**, (most effective the first 10 days) :
  - **-Plasmapharesis**
  - **-IVIG**
  - ***Steroids** are not effective and not indicated
  - **-Critical care monitoring**
- *Most common cause of death* is **autonomic dysfunction**
- *Second most common cause of death* is **respirotory failure**

Risk factors for **respiratory failure** in GBS :

- ***Cranial nerve involvement***
- ***Short time from preceding respiratory illness***
- ***Rapid progression over less than 7 days***
- ***Elevated CSF protein in the first week***
- ***Severe weakness*** :
  - Unable to lift elbows above the bed
  - Unable to lift head above the bed
  - Unable to stand

Transverse Myelitis

Acute demyelinating disorder of the **spinal cord** that **evolves** over **days** usually but may have a **hyperacute presentation**

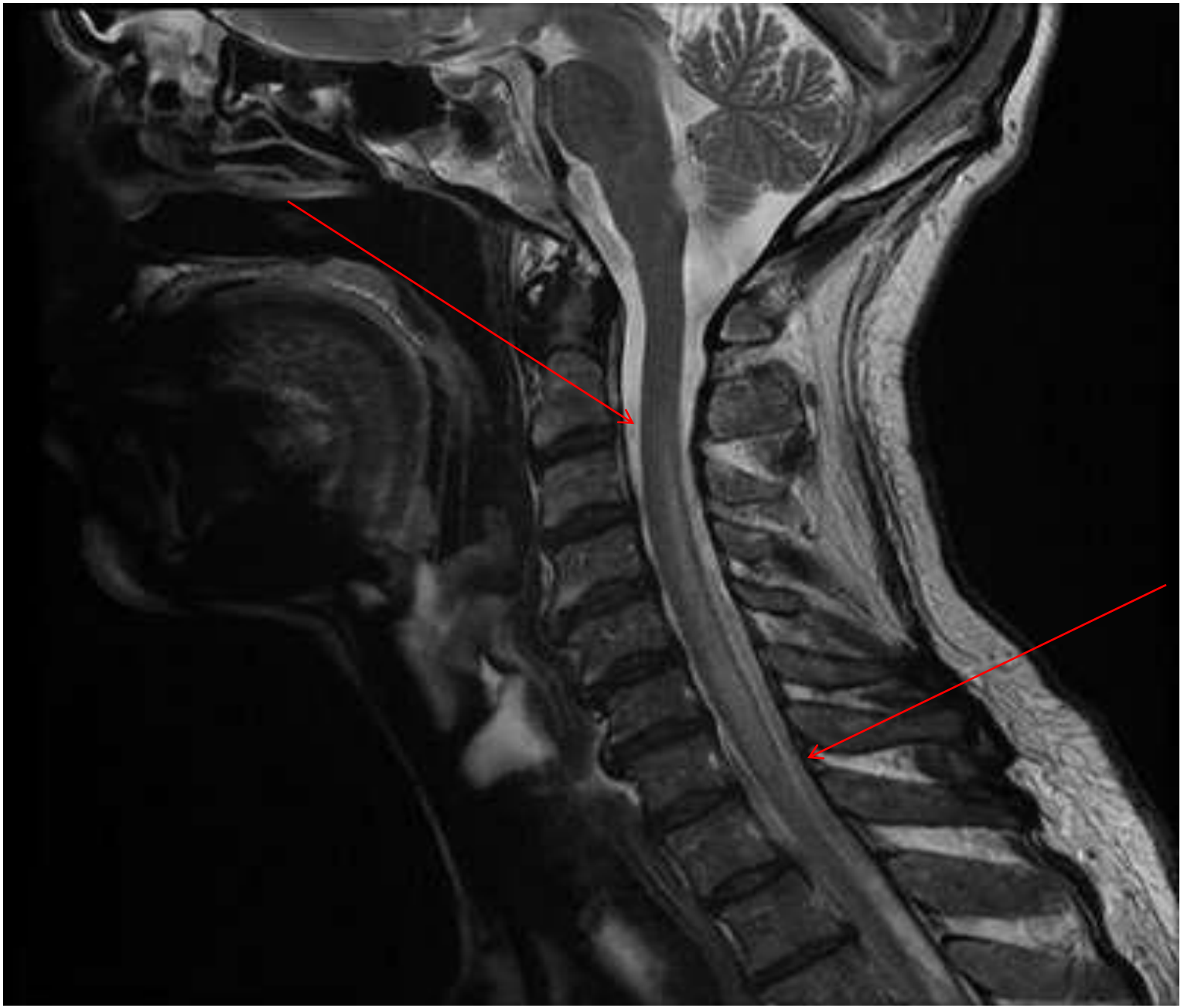
- May be associated with demyelination in other parts of the central nervous system
- **Commonly preceded by a viral infection or immunization**

- **Commonly** presents with **an ascending weakness**

- **Initially** **reflexes may be depressed or absent** because of ***spinal shock*** or involvement of the nerve roots

Symptoms **progress rapidly**, **peaking within 2 days**

- Usually level of myelitis is **thoracic**
- **Sensory level**, **asymmetrical leg weakness**, and **early bladder involvement**.
- **Back pain** is **common at the onset**
- **Tendon reflexes** may be **decreased or increased**
- **Recovery** usually **begins** after **a week of onset**



Transvers Myelitis

Treatment of TM

High doses of IV steroids (methylprednisolone) followed by **tapering** doses of prednisone

- **Prognosis**: **50%** make a **full recovery**

  40% recover incompletely

  10% do not recover

Poliomyelitis

Enteroviruses (*Poliovirus*, *cosxackievirus*, and the *echovirus group*) are **RNA viruses** that inhabit the **GI tract of humans**

- They are **neurotropic**, and produce **paralytic disease** by **destroying the motor neurons of the brainstem and spinal cord**
- **Poliovirus** causes the **most severe paralysis**, **coxsackie and echoviruses** are more likely to cause an **aseptic meningitis**

Nonpolio enteroviruses hav been associated with **polio-like paralytic disease**, frequently accompanied by **other clinical syndromes**, such as aseptic meningitis, hand-foot-and mouth disease, and acute hemorrhagic conjunctivitis.

Poliomyelitis Clinical presentation

Epidemics usually occur in the **spring and summer**

- Usually a **brief illness** characterized by **fever, malaise** and **GI symptoms** precedes the **paralytic illness**
- **After the febrile illness**, there is a **brief period of apparent well being**, **after which** the **fever recurs**, with **headache**, **vomiting** and **meningeal irritation**

- **Pain** in the ***limbs and spine*** is followed rapidly by **limb weakness**

- **Pattern of limb weakness** is variable, but is **generally asymmetric**

- **Weakness**, **diminished reflexes** and **muscle atrophy** are seen

- **Paralysis**

- **Bulbar polio** may occur with or without **spinal polio** and is life threatening

- Affected children have **prolonged periods of apnea** and require mechanical ventilation

- **Extraoccular muscles are spared**

- **Paralytic polio** is **rarely** seen after the introduction of the **polio vaccine**

Diagnosis:

- **Clinical suspicion**
- **CSF leukocytosis** is seen the acute phase,**elevated protein** may also be seen
- **CBC** shows **leukocytosis**
- **Virus** recovery from **stool** is **essential**
- Obtain ***stool, blood and throat samples*** for **viral serology,** demonstrating a **four fold rise in IgG** is *helpful* but *not always* easy.
- **Positive IgM antibodies is diagnostic**

Treatment: *mainly supportive*

- **Mechanical ventilation** may be needed in **bulbar** involvement
- **Pain management** for paresthesias
- **Physical therapy**

Polio Vaccine

A **single dose** of Sabin's **oral polio vaccine** produces immunity to **all three poliovirus serotypes** in approximately **50%** of recipients. **Three doses** of live-attenuated OPV produce protective antibody to **all three poliovirus** types in **more than 95%** of recipients.

After **two doses** of **IPV** (given by injection), **90% or more** of individuals develop protective antibody to **all three serotypes** of poliovirus, and **at least 99%** are immune to poliovirus following **three doses**.

Acute Periodic Paralysis

CALCIUM CHANNEL DISEASE

This disease has become clinically apparent **after adolescence** and has been **much more severe in males**.

The usual pattern of inheritance is **autosomal dominant with reduced penetrance in women** (**male-to-female ratio** of **3** or **4:1**)

The **typical attack** comes on **during the second half of the night or the early morning hours**, *after* **a day of unusually strenuous exercise; a meal rich in carbohydrates** favors its development.

Excessive hunger or thirst, *dry mouth*, *palpitation*, *sweating*, *diarrhea*, *nervousness*, and a *sense of fatigue* are mentioned as **prodromata** but *do not necessarily* precede an attack.

the **weakness lasts a few hours** if mild or **several days** if severe.

Limbs are affected **earlier and often more severely than trunk muscles**, and **proximal muscles are possibly more susceptible than distal ones.**

The muscles most likely to **escape** are those of the eyes, face, tongue , pharynx, larynx, diaphragm, and sphincters,

As the attack subsides, **strength** generally returns **first** to the **muscles** that were **last** to be affected.

Laboratory Findings

-**Reduction in serum K levels**, as low as 1.8 mEq/L (The serum K levels return to normal during recovery)

-The fall in serum K is associated with **little or no increase in urinary K excretion** (large quantities of K enter the muscle fibers during an attack)

Diagnosis at a time **when the patient is normal** may be facilitated by **provocative tests** with the carefully monitored, including the use of ECG, the oral administration of 50 to 100 g of glucose or loading with 2 g of NaCl every hour for 7 doses, followed by vigorous exercise, brings on an attack, which then can be terminated by 2 to 4 g of oral KCl

Treatment

A **low-sodium diet** (*less than 160 mEq/d*), **avoidance of large meals** and **of exposure to cold**, and **acetazolamide 250 mg tid** may be helpful in **preventing attacks.**

For an **acute attack**, **0.25 mEq KCl/kg** should be given **orally** or, if this is *not tolerated*, **some other K salt** may be tried. This **dose may be insufficient** and if there is no improvement in 1 or 2 h, **KCl** may have to be given **intravenously**: **0.05 to 0.1 mEq/kg** initially in a **bolus** at a **safe rate**, followed by **20 to 40 mEq KCl in 5 percent mannitol,** avoiding glucose or NaCl as the carrier solution


END