



Illnesses presenting as Acute Flaccid Paralysis

Dr. N. Aumeer - Paediatrician

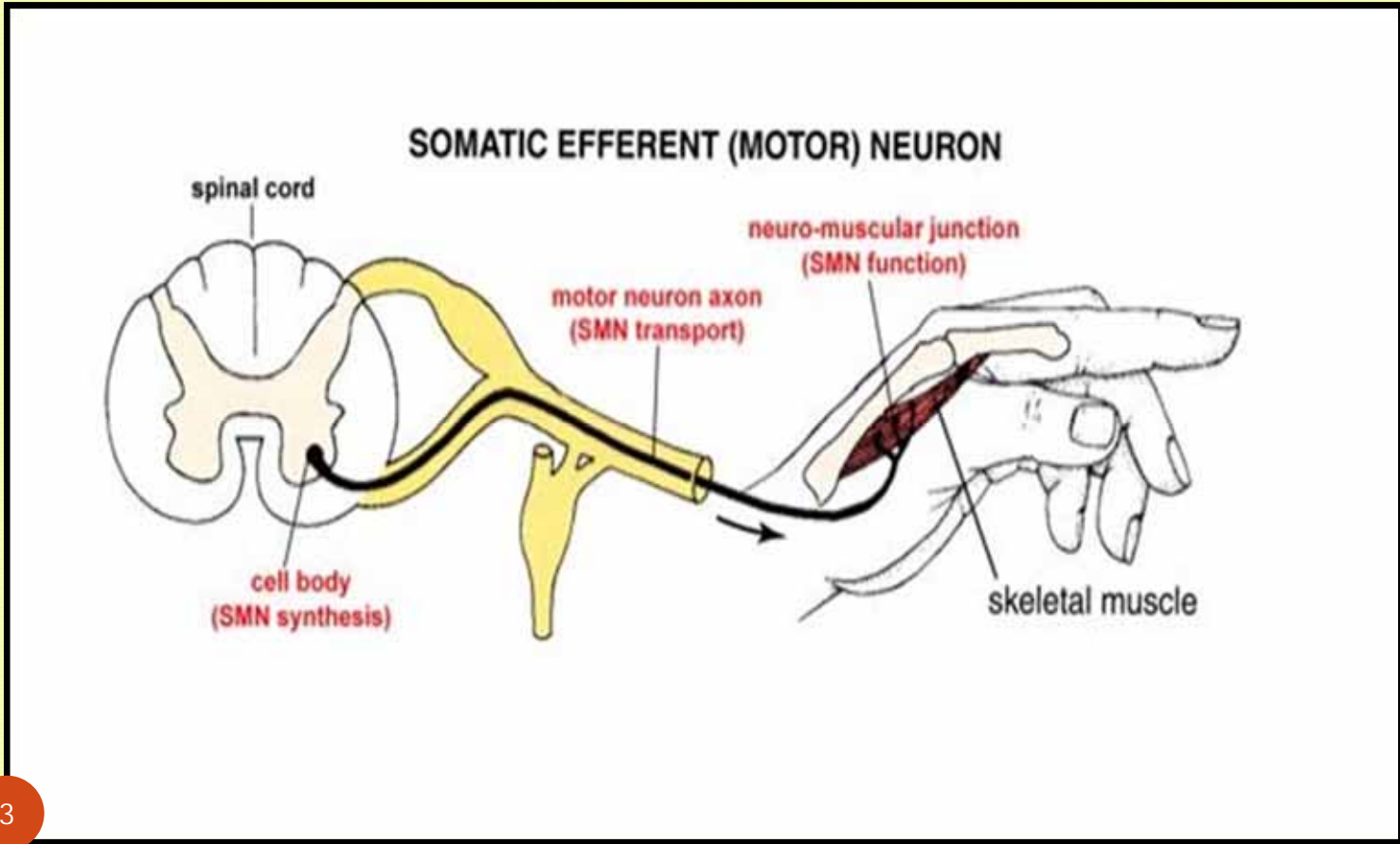
1



Acute Flaccid Paralysis (AFP)

- ❑ Acute onset of weakness with reduced muscular tone in a previously normal limb.
- ❑ Usually below 15 yrs of age.
- ❑ **It is a Lower motor neurone lesion**

Lower Motor Neuron Unit





Lower motor neuron pathway

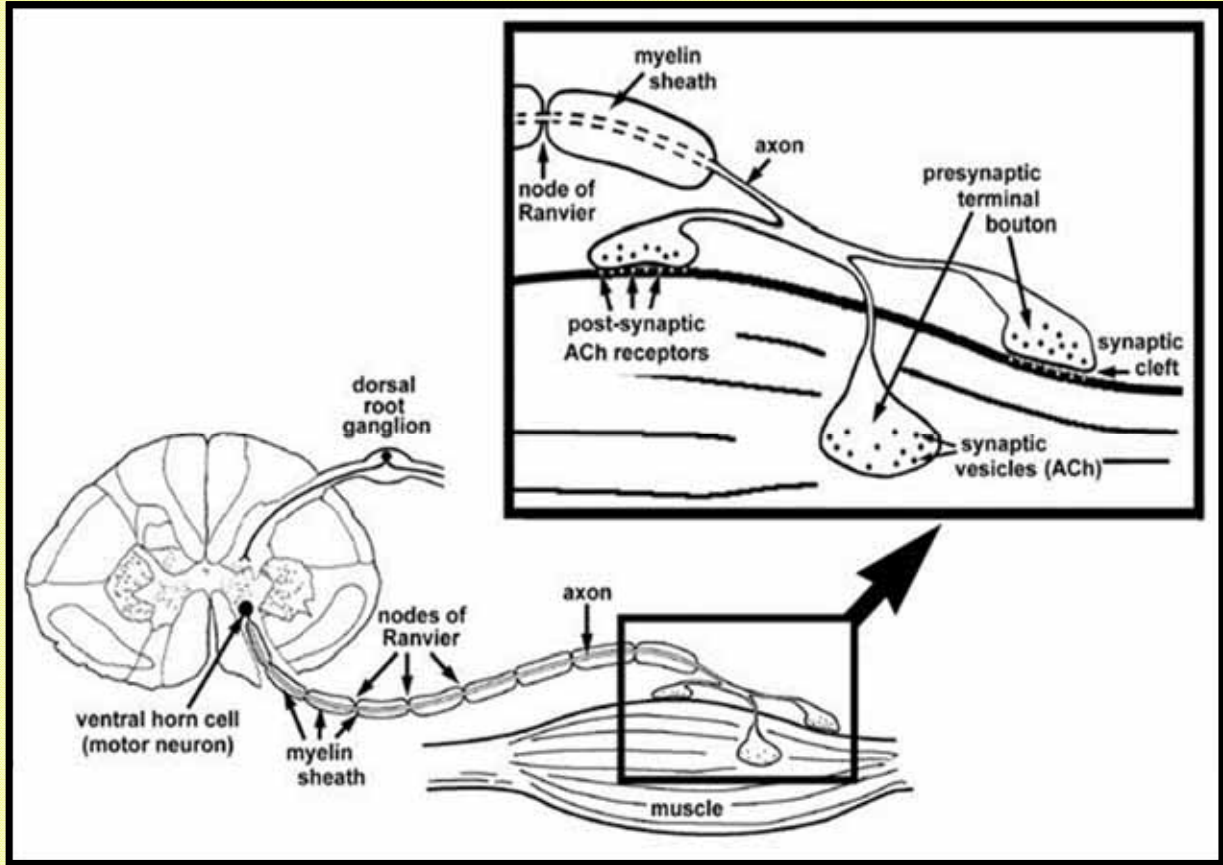
- ❑ Connect brainstem and spinal cord to muscle fibres.
- ❑ Glutamate is released from upper motor neurones
- ❑ It triggers depolarisation in **ventral horn** of lower motor neurones
- ❑ This causes action potential to propagate along axon to neuromuscular junction.
- ❑ Acetylcholine released at **neuromuscular junction** causes muscles to contract.

A landscape photograph showing a river flowing through a valley with green hills and mountains in the background under a blue sky.

Upper & Lower motor neurone lesions

- ❑ Upper motor neurone lesions cause increased muscle tone and exaggerated tendon reflexes and up going plantars.
- ❑ Lower motor neurone lesions cause decreased muscle tone and diminished tendon reflexes with down going plantars.

Lower motor neuron pathway





Case Study

- A 4-year-old boy was admitted to PICU for evaluation of acute-onset flaccid tetraparesis which started in lower limbs.
- 1 day prior to admission, he complained of severe pain in his neck.
- Few hours later, he was unable to stand & a few hrs after was unable to raise his arms.
- Mental status had not changed.
- No history of headache or blurred vision.
- 2 weeks ago he had runny nose and cough without fever.
- Previous medical & family history were unremarkable.



Clinical examination

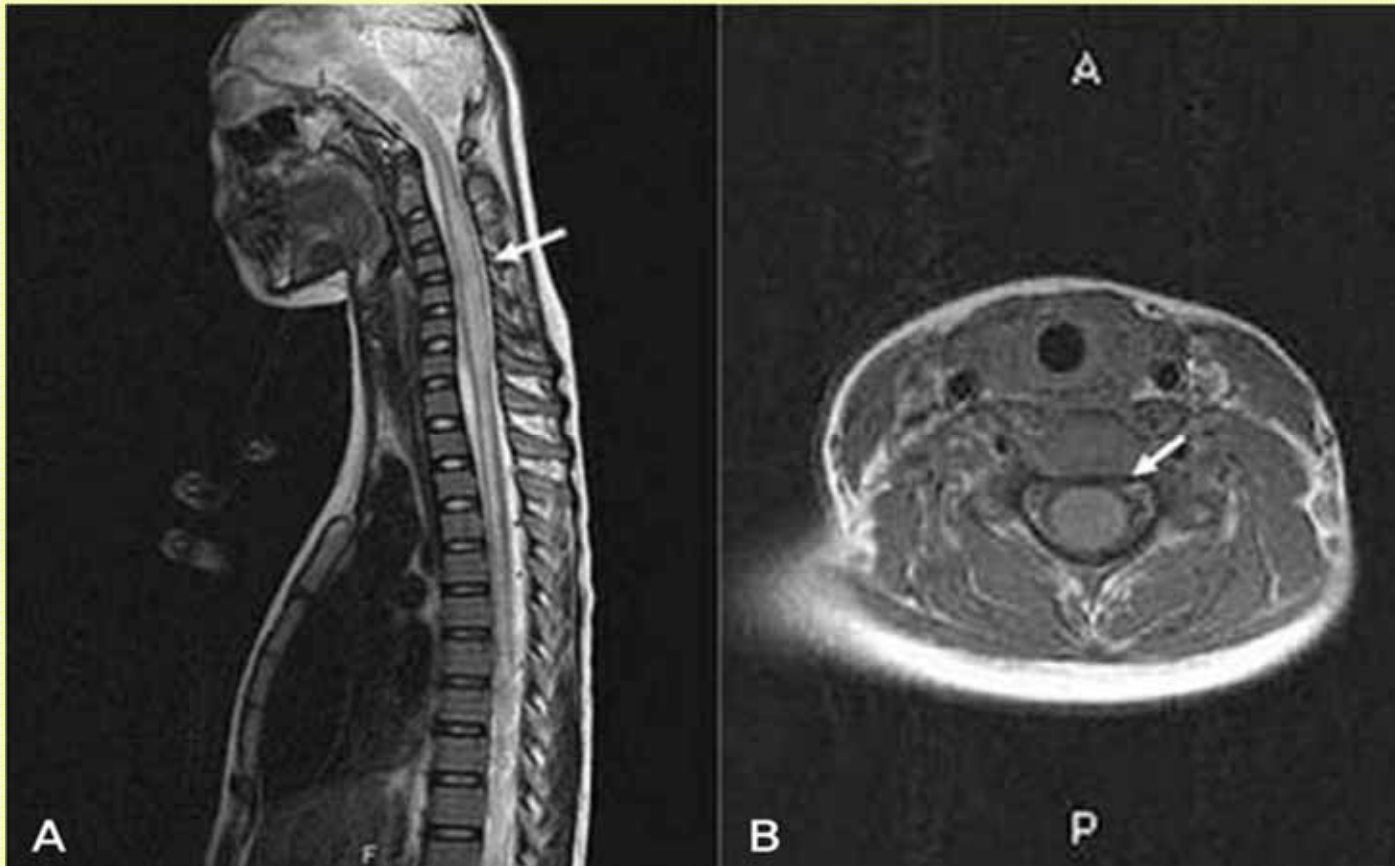
- Fully conscious & oriented . Afebrile.
- Pulse :132 / min regular. Mean BP: 64 mmHg .
- Chest** auscultation : normal. No tachypnoea
- CVS**: unremarkable.
- Abdomen**: distended. Non tender .
- Bladder palpable till umbilicus.
- CNS examination**: Unable to move any limb.
- Muscle tone decreased .Deep tendon reflexes absent upper & lower limbs.
- Cranial nerves: intact.
- Sensory level at C5.
- Anal sphinctor tone : normal.



Laboratory work-up

- ❑ **Hb** 11g/dL. **Platelets** : 273,000/ mm³
- ❑ **WBC** : 9200 /mm³ (66% PNN/ 32% lymphocytes).
- ❑ **ESR** : 46 mm/1st hr.
- ❑ **CRP** : 12 g/L
- ❑ **U&E**: Normal range.
- ❑ **Coagulation** Profile : Normal.
- ❑ **Lumbar Puncture**: CSF clear. Cells: 2 WBCs, 100% lymphocytes. Protein : 25 mg/dl .Glucose: 82 mg/dl.
- ❑ **MRI** spinal cord: fusiform swelling C4-T3.

MRI Spinal cord sagittal T1-weighted images shows fusiform swelling from C4 to T3





Differential Diagnosis

- ❑ Acute Transverse Myelitis.
- ❑ Acute Disseminated Encephalomyelitis (ADEM).
- ❑ Clinically no alteration in mental status or cognitive functions & MRI findings localised.
- ❑ Guillain –Barré Syndrome: Sensory level present. CSF : no albumino-cellular dissociation.
- ❑ MRI spinal cord : normal findings in GBS.



Diagnosis & Treatment

- ❑ **Diagnosis** : Acute Transverse Myelitis .
- ❑ **Treatment**: IV methylprednisolone 30mg/Kg/day
+ IV immunoglobulin 400 mg/Kg/day both for 5 days.
- ❑ **Progress** :
 - ❖ some voluntary movements in the limbs and recovery of the deep tendon reflexes over next 15 days.
 - ❖ 6 months later: ongoing rehabilitation programs .
 - ❖ Walking with intermittent support and full control of bladder and intestine functions.

A scenic landscape photograph showing a waterfall cascading over rocks into a pool of water, surrounded by lush greenery and mountains in the background. The title 'Causes of Acute Flaccid Paralysis' is overlaid in yellow text on a dark horizontal band at the bottom of the image.

Causes of Acute Flaccid Paralysis

- Acute paralytic poliomyelitis.**
- Guillain-Barré Syndrome.**
- Transverse Myelitis.**
- Non-Polio Enteroviruses.**
- Toxic Neuropathy.**



Guillain Barre Syndrome (GBS)

- ❑ Acute inflammatory **demyelinating** polyneuropathy (AIDP)
- ❑ Autoimmune process : immunologic response against myelin component of peripheral nerves.
- ❑ Fever occurs 2 – 3 weeks **before** onset of paralysis.
- ❑ Paralysis is usually flaccid and **symmetrical** with **diminished** deep tendon reflexes .
- ❑ Paralysis is **ascending** affecting lower limbs first, followed by trunk, then upper limbs.



Guillain Barre Syndrome (GBS)

- ❑ Bilateral cranial nerve involvement is common, especially 7th / 9th & 10th cranial nerve .
- ❑ Pain and muscle tenderness is prominent at onset of paralysis.
- ❑ Urinary retention and constipation occur before onset of limb weakness
- ❑ **Nerve** conduction velocity is reduced in GBS.

GBS v/s paralytic poliomyelitis				
	Age	Fever	Paralysis	CSF
Paralytic Polio	<u>Below 3 yrs</u>		<ul style="list-style-type: none"> ▪ Asymmetrical ▪ Descending <ul style="list-style-type: none"> ▪ Proximal muscles involved first 	<ul style="list-style-type: none"> ▪ Normal Protein ▪ WBC 20- 300
Guillain Barre Syndrome	<u>Above 2 yrs.</u>		<ul style="list-style-type: none"> ▪ Symmetrical ▪ Ascending <ul style="list-style-type: none"> ▪ Distal small muscles involved first 	<ul style="list-style-type: none"> ▪ Increased proteins ▪ WBC < 10



- ❑ Caused by inflammation across **one** segment of the spinal cord & damage to myelin sheath of peripheral nerves.
- ❑ Damage at one segment of spinal cord affects function at that segment and segments below it.
- ❑ The segment of the spinal cord at which the damage occurs determines which parts of the body are affected.
- ❑ Demyelination usually occurs at the thoracic level causing problems with leg movement as well as bowel and bladder control.



Transverse Myelitis

- ❑ Occurs in children aged 4 years and above.
- ❑ Sudden onset of low back pain, muscle weakness or abnormal sensations in the toes and feet .
- ❑ Rapidly progresses to flaccidity of legs & paralysis followed by urinary retention and loss of bowel control.
- ❑ Paralysis is usually symmetrical in lower limbs and accompanied by profound anesthesia to all forms of sensation.
- ❑ Site of involvement is usually the thoracic cord but can be lumbar or cervical. Arms may also be partially paralyzed.



Transverse Myelitis

- ❑ Initial hypotonia gradually changes to spasticity after weeks .
- ❑ Deep Tendon reflexes are initially reduced but later become exaggerated
- ❑ Recovery is related to onset of paralysis : if onset is within hours, recovery begins after several weeks and neurological deficits remain.
- ❑ If paralysis progresses over several days recovery is complete and begins 1 to 5 days after symptoms peak.
- ❑ Most patients have only one episode of transverse myelitis.

Acute Polio v/s GBS & Transverse Myelitis

Clinical Features	ACUTE POLIOMYELITIS	GUILLAIN BARRE SYNDROME	TRANSVERSE MYELITIS
Progression of Paralysis	• takes 24 – 48 hours from onset to full paralysis.	Few hours up to 10 days	Few hours up to 4 days
Fever	•High fever at onset of paralysis. •No fever after.	Not common	Rarely Present
Onset of Flaccidity	Acute Symmetrical Proximal	•Acute Symmetrical Distal	• Acute •Lower limbs •Symmetrical





Acute onset weakness lower limbs Clinical Approach

- ❑ Demonstrable motor weakness
- ❑ DTR : absent / reduced/ normal
- ❑ Sphinctors : preserved or affected
- ❑ Sensory loss: dermatomal
- ❑ Clinical localisation : spinal cord
 - ❖ Transverse myelitis/ Tumor : spinal or extraspinal
 - ❖ Arterio -venous malformation
 - ❖ Spinal cord infarct
 - ❖ Spinal cord abscess/ Spinal TB



Acute onset weakness in lower limbs Clinical Approach

- ❑ Demonstrable motor weakness
- ❑ Deep Tendon Reflexes : reduced or normal
- ❑ Sphincter function preserved
- ❑ No Sensory loss
- ❑ Clinical localisation : muscle
 - ❖ Post viral myositis
 - ❖ Toxic myositis
 - ❖ Periodic paralysis



Acute onset weakness in lower limbs Clinical Approach

- ❑ Demonstrable motor weakness
- ❑ Sphincter function : preserved
- ❑ Sensory loss : glove and stocking
- ❑ Deep Tendon Reflexes: absent
- ❑ Clinical localisation : Peripheral nerve
- ❖ If unilateral : Enterovirus infection or Local trauma
- ❖ If Bilateral : Guillain Barre or Toxic neuropathy



Acute onset weakness lower limbs

Diagnostic work-up

- ❑ **Accurate Clinical history is essential**
- ❑ **AFP work up required in all cases.**
- ❑ **Muscle:** Creatine kinase/ U&E/ Urine Myoglobin
- ❑ **Peripheral Nerve :**
 - ❖ If Unilateral :
 - Nerve conduction velocity
 - MRI lumbosacral plexus
 - ❖ If Bilateral :
 - CSF proteins & cells
 - Nerve conduction velocity/ Forced vital Capacity
- ❑ **Spinal Cord Lesion:**
 - Urgent MRI spinal cord
 - CSF : cells/ protein/ glucose/ culture / TB isolation/
 - PCR for Cryptococcal antigen / oligoclonal bands
 - FBC / ESR/ C3/ C4 / Antinuclear Factor





Non-Polio Enterovirus (NPEV)

- Coxsackie A & Coxsackie B viruses
- Echoviruses
- Enterovirus types 70 & 71
- Mumps virus (paramyxo virus)
- Associated with both mild and severe neurolytic disease.
- Most cases show complete recovery.
- In some cases sequel may mimic paralysis caused by wild poliovirus.
- Healthy children excrete non-polio enteroviruses.
- Hence their isolation from patients with AFP may not be proof of causal relationship.



AFP: Peripheral Neuropathy

- ❑ Metabolic disorder : Acute diabetic mononeuropathy
- ❑ Toxic Neuropathy : Pharmacological products : pyridoxin / Cisplatin / Amiodarone / Vincristine
- ❑ Hereditary disease : Charcot –Marie –Tooth : peroneal muscular atrophy. Autosomal dominant genetic disorder with degeneration of peripheral nerves (axons / myelin sheath). Onset of symptoms is often in adolescence or early adulthood.
- ❑ Food poisoning : Diphtheria toxin and Botulism Toxin
- ❑ **Myasthenia gravis** : autoimmune disorder of peripheral nerves
Antibodies form against acetylcholine nicotinic postsynaptic receptors at the neuromuscular junction. A reduction in the number of acetylcholine receptors results in a characteristic pattern of progressively reduced muscle strength with repeated use of the muscle and recovery of muscle strength following a period of rest. Bulbar muscles are most commonly affected.



AFP : Peripheral Neuropathy

- ❑ **LYME Disease** caused by spirochete *Borrelia burgdorferi*, transmitted by *Ixodid* tick species. Cranioneuropathies, especially peripheral 7th nerve, are common.
- ❑ **Tumors** may cause progressive flaccid paralysis that is asymmetrical without associated fever. Distribution of nerve involvement is dependent upon the anatomic location of the space-occupying lesion.
- ❑ **HIV-associated polyneuropathy** : more common in adults. Associated with low CD4 counts. Either due to CMV/ H. Zoster or lymphoma of the spinal cord.
- ❑ **Hypokalemia** may present with generalized acute flaccid paralysis of all 4 limbs and neck flop. Weakness is noticed first in limb muscles, followed by trunk and respiratory muscles. Areflexia, paralysis, death from respiratory muscle failure and cardiac arrest can occur.



Toxic Neuropathy : Organophosphates

- Mechanism of action: **irreversible inhibition of acetylcholinesterase** which is found in red blood cells and in nicotinic and muscarinic receptors in nerve, muscle, and gray matter of the brain.
- ❑ Accumulation of acetylcholine at the neuromuscular junction causes persistent depolarization of skeletal muscle, resulting in weakness and fasciculations.
- ❑ Symptoms occurs within hours of exposure. The acronym SLUDGE is used to describe the muscarinic manifestations of salivation, lacrimation, urination, defecation, GI distress, and emesis.
- ❑ Delayed effects take at least 10 days to develop following a single acute exposure. Cramping, tingling, ataxia, and weakness in the lower extremities, progressing to generalized weakness may be seen in severe cases.





Botulism Neurotoxicity

- ❑ Neurotoxins secreted by Gram-positive anaerobic *Clostridium botulinum* bind pre-synaptically and prevent release of acetylcholine.
- ❑ The organism or its heat-resistant spore proliferates in anaerobic conditions in the colon/ in wounds /in food.
- ❑ Bilateral symmetrical & descending flaccid paralysis occurs after 12-36 hr.
- ❑ Dry mouth, ptosis, diplopia ,dilated pupils , constipation, ileus / urinary retention & hypotension develop .
- ❑ Bulbar paresis (dysarthria /dysphagia) may result in aspiration pneumonia & respiratory paralysis . Facial weakness with absent sucking & gag reflex occur in infants.
- ❑ There is No fever. Consciousness is not impaired
- ❑ Deep Tendon reflexes are absent. There is No sensory loss.
- ❑ CSF examination is normal.
- ❑ Complete recovery takes weeks to months.



Diphtheria Neurotoxicity

- ❑ Caused by toxins secreted by aerobic *Corynebacterium diphtheriae*.
- ❑ Incubation period 2 - 5 days.
- ❑ Nerve damage often occurs first in the throat, palate and ocular muscles and may subsequently become generalised.
- ❑ Throat infection : pseudomembranes, dysphagia, airways obstruction, markedly enlarged cervical lymph nodes and leukocytosis ; followed by myocarditis around the 2nd week and sometimes peripheral paralysis around the 3rd to 6th week.
- ❑ Untreated patients remain infectious for 2 to 3 weeks.
- ❑ Treatment consists of antibiotics : macrolides or penicillins and diphtheria antitoxin.



Traumatic Neuritis

- ❑ Traumatic neuritis is caused by injections
- ❑ It may lead to AFP of the lower extremity.
- ❑ Onset of AFP in the affected lower limb occurs from 1 hour to 5 days after injection in the gluteal region.
- ❑ Fever is usually present before the onset of paralysis when injection is given for a pre-existing febrile illness.
- ❑ AFP is usually accompanied by pain in the gluteal region or along the affected leg.
- ❑ Atrophy may appear 40 to 60 days later. However, atrophy caused by a traumatic injection never reaches the degree seen in polio. Differences in calf circumference usually do not exceed 0.5 to 1.5 cm
- ❑ Knee jerk is present. Ankle jerk is absent or diminished.
- ❑ Hip and knee strength is normal.
- ❑ Child walks with a foot drop.
- ❑ Gradual recovery with physiotherapy usually occurs within 3 to 9 months.



Traumatic Neuritis v/s Polio

- ❑ In polio : paralysis involves large proximal muscle groups although any group of muscles may be affected. Tendon reflexes are diminished.
- ❑ In Traumatic Neuritis : only one leg is involved below the knee. knee jerk is normal while the ankle jerk is diminished.
- ❖ **Age:** Polio occurs mainly below 3 years of age, while in TN there is no specific age distribution.
- ❖ **The paralytic sequel of polio is more severe and is permanent.**
- ❖ **Atrophy of muscles and shortening of one lower limb may be present.**
- ❖ **AFP due to causes other than polio tends to resolve or improve within 60 days of onset.**

Thank you

