PAEDIATRICS NEUROLOGY

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ACUTE FLACCID PARALYSIS

Acute symmetric paralysis	Acute asymmetric paralysis
1. Guillain-Barre syndrome	1. poliomyelitis
2. post-diphtheric paralysis	2. non-polio viruses
3. botulism	3.pseudo-paralysis: Trauma, toxic synovitis, and osteomyelitis
4. transverse myelitis	4. post- injectional paralysis
5. spinal cord compression	

POLIOMYELITIS

It is a due to 3 antigenically distinct serotypes (1, 2, and 3).

Infants and young children (3mo-3years) are the affected age group in developing countries. **Poor sanitation and crowding** have permitted the continued transmission, despite massive global efforts to eradicate polio. Humans are the only known reservoir for the polioviruses, which are spread by the fecal-oral route for longer than 2 wk before paralysis to several weeks after the onset of symptoms.

Pathogenesis

Wild-type and vaccine strains of polioviruses gain host entry via the GIT, replicate in the mucosa of the small intestine. Regional lymph nodes are infected, and viremia occurs after 2-3 days. The virus seeds multiple sites, including the RES. Wild-type poliovirus probably accesses the CNS along peripheral nerves. Vaccine strains of polioviruses do not replicate in the CNS, a feature that accounts for the safety of the live-attenuated vaccine.

The poliovirus primarily infects motor neuron cells in the spinal cord (**the anterior horn cells**) and the medulla oblongata (the cranial nerve nuclei). Involvement of the vital centers controlling respiration and circulation may have a catastrophic outcome. Other parts of brain may be affected.

Clinical Manifestations

The incubation period 8-12 days. Poliovirus infections with wild-type virus may follow 1 of several courses:

Inapparent infection

occurs in 90-95% of cases and causes no disease and no sequelae.

Abortive Poliomyelitis

In approximately 5% of patients, a nonspecific influenza-like syndrome. The illness is short lived, lasting up to 2-3 days.

Non- paralytic Poliomyelitis

In approximately 1% of patients infected with wild-type poliovirus, signs of abortive poliomyelitis are present with more intense headache, nausea, vomiting, soreness and stiffness of the posterior muscles of the neck, trunk, and limbs, the anterior fontanel may be tense or bulging.

Paralytic Poliomyelitis

Paralytic poliomyelitis develops in approximately 0.1% of infected persons infected, causing 3 clinically recognizable syndromes that represent a continuum of infection differentiated only by the portions of the CNS most severely affected. These are (1) spinal paralytic poliomyelitis, (2) bulbar poliomyelitis, and (3) polioencephalitis.

Spinal paralytic poliomyelitis

Stages: Abortive poliomyelitis, Recovery (2-5 days), Exacerbation of the previous systemic symptoms (of non paralytic phase), then Paralytic phase: sensory and motor phenomena:

- Sensory e.g., paresthesia and hyperesthesia.
- Motor: spotty distribution, single, multiple. Asymmetric flaccid paralysis or paresis occurs. Involvement of one leg is most common, followed by involvement of one arm. The proximal areas of the extremities tend to be involved to a greater extent than the distal areas.
- Respiratory paralysis: due to inter-costal and diaphragmatic paralysis, life threatening.
- Bowel and bladder dysfunction ranging from transient incontinence to constipation and urinary retention
- Little recovery from paralysis is noted in the 1st days or weeks, but, if it is to occur, it is usually evident within 6 mo. Lack of improvement from paralysis within this period is usually evidence of permanent paralysis.

Bulbar poliomyelitis may occur as a clinical entity without apparent involvement of the spinal cord.

- Respiratory difficulty, weak cry, weak cough and nasal regurgitation.
- other are paralysis of extra ocular, facial, and masticatory muscles involvement.
- Vital centers in the medulla may be involved and manifested as irregularities in respiration; and cardiovascular alterations, including blood pressure changes

Polio-encephalitis is a rare form of the disease in which higher centers of the brain are severely involved. Seizures, coma, and spastic paralysis with increased reflexes may be observed.

Diagnosis

Poliomyelitis should be considered in any unimmunized or incompletely immunized child with paralytic disease. WHO recommends that the laboratory diagnosis of poliomyelitis be confirmed by isolation and identification of poliovirus in the stool, with specific identification of wild-type and vaccine-type strains. In suspected cases of acute flaccid paralysis, 2 stool specimens should be collected 24-48 hr apart as soon as possible after the diagnosis of poliomyelitis is suspected. Poliovirus concentrations are high in the stool in the 1st wk after the onset of paralysis, which is the optimal time for collection of stool specimens.

Management of poliomyelitis;

1. isolation; especially from susceptible infants.
2. observation; especially muscles of respiration, observation for degree of involvement is important, blood gas analysis and in case of hypoxemia and CO2 retention, mechanical ventilation is life saving.
3. Rest of the paralysed limb; should be kept in neutral position, All intramuscular injections and surgical procedures are contraindicated during the acute phase of the illness, especially in the 1st wk of illness, because they might result in progression of disease, passive movement of the limb is gradually allowed during the second week.

There is no specific antiviral treatment for poliomyelitis

GUILLAIN –BARRE SYNDROME
POST-INFECTIOUS POLYNEURITIS

It is an autoimmune disorder involving mainly motor but also sensory and sometimes autonomic nerves. It is primarily axonal degeneration. It is the most common cause of acute paralysis in children, although infants can be affected too, most cases are seen in children more than 3years old. The paralysis usually follows a nonspecific GIT or Resp. infection by approximately 10 days. The original infection might have caused only gastrointestinal *(*especially *C. jejuni,* but also *H. pylori*) or respiratory tract (especially *M. pneumoniae*) symptoms.

Clinical manifestations

Acute symmetric paralysis: Initial symptoms include numbness and paresthesia, followed by weakness which is symmetrically in the lower extremities and progressively involves the trunk, the upper limbs, and, finally, the bulbar muscles, a pattern known as **Landry ascending paralysis.** Asymmetry is found in 9% of patients. The onset is gradual and progresses over days or weeks; the process plateaus in 1-28 days. Tenderness on palpation and pain in muscles are common in the initial stages. Affected children are irritable. Weakness can progress to inability or refusal to walk and later to flaccid tetraplegia with loss of tendon

reflexes, respiratory paralysis may occur in some cases and necessitates hospitalization and mechanical ventilation for few weeks.

Bulbar involvement occurs in about 50% of cases. Respiratory insufficiency can result. Dysphagia and facial weakness are often impending signs of respiratory failure with increased the risk of aspiration. Some young patients exhibit symptoms of viral meningitis or meningoencephalitis.

Miller-Fisher syndrome (MFS) consists of acute external and occasionally internal ophthalmoplegia, ataxia, and areflexia.

The **autonomic nervous system** is also involved in some cases. Liability of BP and HR and occasional asystole may occur. Cardiovascular monitoring is important.

Chronic inflammatory relapsing polyneuritis or **Chronic unremitting polyradiculoneuropathy** are chronic varieties of Guillain-Barré syndrome

The course is benign in most cases and gradual complete recovery usually occur over few weeks or few months.

Laboratory findings and diagnosis

- CSF studies are essential for diagnosis: The CSF protein is elevated to more than twice the upper limit of normal, the glucose level is normal, and there is no pleocytosis. This called protein cellular dissociation, which develop at 2nd week.
- MRI findings include thickening of the cauda equina and intrathecal nerve roots with gadolinium enhancement.
- Motor nerve conduction velocities are greatly reduced.
- Electromyography shows evidence of acute denervation of muscle.
- Serum creatine kinase level is normal.

Treatment

1. No isolation is required

2. Observation: at hospital, for involvement of respiratory and or bulbar muscle, intubation and mechanical ventilation for several weeks may be necessary and in bulbar paralysis N/G tube feeding is required.

3. In rapidly progressive and sever paralysis:

- Immunoglobulin IV: A commonly recommended protocol is IVIG 0.4 g/kg/day for 3- 5 consecutive days

- Plasmapheresis and/or immunosuppressive drugs if IVIG is ineffective.

- Steroids are not effective.

4. Supportive treatment: treatment of secondary infections and prevention of contractures by physiotherapy.