POLIOMYELITIS
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Texto baseado no livro-texto Cecil (Medicina, 2007)
Definition
Poliomyelitis (acute anterior poliomyelitis, infantile paralysis) is an acute illness caused by the three strains of poliovirus. The disease selectively destroys the motor neurons of the spinal cord and brain stem, thereby resulting in flaccid asymmetrical weakness. Through the mid-20th century, poliomyelitis was one of the most feared of all human infectious diseases; however, it is now almost entirely preventable by vaccination.

Etiology
The antigenically different strains of poliovirus (types 1, 2, and 3) are classified in the genus Enterovirus within the family Picornaviridae. They are small (approximately 27 nm), roughly spherical particles with icosahedral symmetry that contain a single-stranded RNA core surrounded by a protein capsid. The majority of disease is caused by the type 1 strain. Lacking a lipid envelope, the polioviruses are resistant to lipid solvents and are stable at low pH.

Epidemiology
Since the World Health Assembly resolved in May 1988 to eradicate poliomyelitis, the estimated number of polio cases globally has declined more than 99%, and nearly 55% of the world's population is considered polio free. About 80% of the world's cases occur in India, although some African countries, particularly those engaged in ongoing warfare, continue to have low immunization rates with high rates of disease. In the United States, the number of cases of paralytic poliomyelitis has fallen to just a few cases yearly because of the widespread use of an effective vaccine. Polio is more frequent during the summer in temperate zones but has a more even distribution throughout the year in tropical areas. Poliovirus is acquired by the oral route and subsequently replicates in the oropharynx and lower gastrointestinal tract. It may be secreted for a week or two in saliva and for more prolonged periods in feces, which provide the major avenue of host-to-host transmission. Spread of polioviruses is greatly influenced by standards of hygiene, and the greatest risk occurs within families or other crowded circumstances.

Paralysis is an unusual complication of poliovirus infection. During an epidemic, only 1 to 2% of infections result in neurologic symptoms and signs; another 4 to 8% of infected persons suffer nonspecific (minor) illness. Although polio occurs most commonly in preschool children, other factors increase the incidence of paralytic disease, including advanced age, recent strenuous exercise, tonsillectomy, pregnancy, and impairment of B-lymphocyte (antibody) defenses. Immunity to each of the three types of poliovirus is lifelong, but infection with one strain does not protect against subsequent infection by another. In the United States, the incidence of poliomyelitis from live attenuated strains, though extremely rare, parallels that of wild-type virus occurring in nonimmunized subjects.

Pathobiology
Polioviruses selectively infect specific neuronal populations and induce highly stereotyped pathologic processes; in this manner they contrast with most of the viruses that cause acute encephalitis or meningitis. The unique susceptibility of anterior horn cells (motor neurons) may be due to the presence of a higher density of viral receptors or spread of the virus along motor nerve fibers.

The poliovirus invades the nervous system only after prior systemic replication. An initial alimentary phase with local replication in the intestinal mucosa and spread to the local lymphatics is followed by viremia, which seeds the nervous system. The virus may replicate in skeletal muscle and be transported via peripheral nerves to the spinal cord; this
mechanism of spread may contribute to the myalgia that precedes the onset of weakness. Convalescent poliomyelitis is characterized by loss of motor neurons and denervation atrophy of their associated skeletal muscles.

**Clinical Manifestations**

Acute poliomyelitis is separated into two distinct phases: “minor illness” and “major illness.” The minor illness coincides with viremia and consists of fever, headache, and sore throat, which resolve within 1 to 2 days. In some patients this phase is followed by the major illness, which is characterized by an abrupt onset of fever, headache, vomiting, and meningismus. Cerebrospinal fluid (CSF) pleocytosis is present at this stage. The symptoms of aseptic meningitis resolve within 5 to 10 days. Asymmetrical muscle weakness, which is the hallmark of the illness, is typically preceded by intense myalgia. The proximal muscles are more commonly involved, and legs are involved more often than arms. In mild cases, paralysis affects only parts of muscles rather than selective peripheral nerve or nerve root distributions. Sensory changes are lacking. The paralysis may render one limb useless yet entirely spare the contralateral arm or leg. Acute urinary retention develops in about 50% of patients. The trunk musculature is least commonly affected. The affected muscles are flaccid, and deep tendon reflexes may be absent. Atrophy develops rapidly, usually beginning within a week in paralyzed muscles and progressing over the ensuing weeks. The motor deficit rarely progresses for more than 3 to 5 days.

In 10 to 15% of cases and especially in young adults, the lower brain stem motor nuclei are affected. Involvement of the ninth and tenth cranial nerve nuclei leads to paralysis of the pharyngeal and laryngeal musculature (bulbar poliomyelitis). Parts of the facial muscles may be involved, either unilaterally or bilaterally. Less often, the tongue and muscles of mastication are paralyzed. External oculomotor weakness occurs rarely, and the pupils are invariably spared. Direct involvement of the brain stem reticular formation can disrupt breathing and swallowing and produce serious disturbances in cardiovascular control, including hypertension, hypotension, and cardiac arrhythmias.

**Diagnosis**

In poliomyelitis, CSF shows increased protein content and pleocytosis with a normal glucose concentration. The virus is more commonly isolated from the nasopharynx or stool than from CSF. Molecular diagnosis with polymerase chain reaction is now the technique of choice for identifying the poliovirus serotype and for differentiating between wild and vaccine strain poliomyelitis.

**Differential Diagnosis**

Because of its rarity in the United States, poliomyelitis may present diagnostic difficulties. Its early phases must be differentiated from other acute meningitides; when paralysis ensues, a major differential diagnosis is postinfectious polyneuropathy or Guillain-Barré syndrome. Illnesses virtually identical to poliomyelitis have been reported rarely with other enteroviruses, the flaviviruses, (such as Japanese B and West Nile virus infection), tick-borne encephalitis, and human immunodeficiency virus infection. Furthermore, motor neuron disease has been reported with some herpes virus infections such as cytomegalovirus, Epstein-Barr virus, and varicella-zoster virus. Other infectious illnesses that must be considered in the differential diagnosis are rabies, Lyme disease, *Mycoplasma, Borrelia*, diphtheria, and botulism. Careful neurologic examination can distinguish other entities that might be confused with poliomyelitis because they affect different areas of the neuraxis (peripheral nerve, spinal cord, neuromuscular junction, or muscle).

**Treatment**
No specific treatment is available, but supportive care is important in reducing pain during the acute attack and in maintaining vital functions to ensure survival. Bedrest and treatment of pain are recommended during the myalgic phase. Important measures include preventing contractures, maintaining airway and cardiovascular stability, and preventing bed sores and excessive mobilization of calcium. Acute respiratory failure requiring intubation and intermittent positive pressure ventilation may develop rapidly. If prolonged ventilation is needed or coexisting bulbar weakness is present, tracheostomy may be necessary to protect the airway.

Prognosis

Death in patients with poliomyelitis is usually the result of bulbar involvement and is attributable to respiratory and cardiovascular impairment. Mortality has been considerably reduced with modern management of respiratory insufficiency. Patients who survive an episode of acute paralytic poliomyelitis usually recover considerable motor function. Generally, motor improvement begins within the first weeks after onset, and 60% of eventual recovery is achieved by 3 months.

The Postpolio Syndrome

In a number of patients with previous poliomyelitis, further motor deterioration develops later in life. In some, this worsening relates simply to musculoskeletal decompensation or other factors but does not involve new weakness. However, other persons suffer a true loss of strength, termed postpolio syndrome. This disorder is characterized by an insidiously progressive weakness typically beginning 30 or more years after an attack of poliomyelitis. Most commonly, it adds to the weakness of already affected muscles; less often, weakness develops in muscles previously thought to be normal. This weakness is often accompanied by fasciculations, and additional atrophy may develop. Muscle biopsy shows type grouping consistent with chronic denervation-reinnervation. Overall, the prognosis is good, with slow progression of further weakness, which only rarely leads to a severe increase in disability or death. The most likely pathogenesis consists of senescence of the surviving expanded motor units. In all cases it is imperative to exclude coincidental unrelated disease.

Prevention

Poliomyelitis can be prevented by live attenuated or killed polio vaccines, which are now given routinely. The practice of immunization has relaxed as the threat of development of paralytic poliomyelitis has become less conspicuous. The World Health Organization plan to eradicate polio has had a substantial impact; however, polio is still endemic in some countries and vaccine-associated polio also occurs.