

Post-Polio Syndrome Fact Sheet

What is Post-Polio syndrome?

Polio, or poliomyelitis, is an infectious viral disease that can strike at any age and affects a person's nervous system. Between the late 1940s and early 1950s, polio crippled around 35,000 people each year in the United States alone, making it one of the most feared diseases of the twentieth century.

The polio vaccine was first introduced in 1955; its use since then has eradicated polio from the United States. The World Health Organization reports polio cases have decreased by more than 99 percent since 1988, from an estimated 350,000 cases then, to 1,352 reported cases in 2010. As a result of the global effort to eradicate the disease, only three countries (Afghanistan, Nigeria, and Pakistan) remain polio-endemic as of February 2012, down from more than 125 in 1988.

Post-Polio syndrome (PPS) is a condition that affects polio survivors' years after recovery from an initial acute attack of the poliomyelitis virus. Most often, polio survivors start to experience gradual new weakening in muscles that were previously affected by the polio infection. The most common symptoms include slowly progressive muscle weakness, fatigue (both generalized and muscular), and a gradual decrease in the size of muscles (muscle atrophy). Pain from joint degeneration and increasing skeletal deformities such as scoliosis (curvature of the spine) is common and may precede the weakness and muscle atrophy. Some individuals experience only minor symptoms while others develop visible muscle weakness and atrophy.

Post-Polio syndrome is rarely life-threatening, but the symptoms can significantly interfere with an individual's ability to function independently. Respiratory muscle weakness, for instance, can result in trouble with proper breathing, affecting daytime functions and sleep. Weakness in swallowing muscles can result in aspiration of food and liquids into the lungs and lead to pneumonia.

Who is at risk?

While polio is a contagious disease, PPS cannot be caught from others having the disorder. Only a polio survivor can develop PPS.

The severity of weakness and disability after recovery from poliomyelitis tends to predict the relative risk of developing PPS. Individuals who had minimal symptoms from the original illness are more likely to experience only mild PPS symptoms. A person who was more acutely affected by the polio virus and who attained a greater recovery may experience a more severe case of PPS, with greater loss of muscle function and more severe fatigue.

The exact incidence and prevalence of PPS is unknown. The U.S. National Health Interview Survey in 1987 contained specific questions for persons given the diagnosis of poliomyelitis with or without paralysis. No survey since then has addressed the question. Results published in 1994-1995 estimated there were about 1 million polio survivors in the U.S., with 443,000 reporting to have had paralytic polio. Accurate statistics do not exist today, as a percentage of polio survivors have died and new cases have been diagnosed. Researchers estimate that the condition affects 25 to 40 percent of polio survivors.

What causes PPS?

The cause of PPS is unknown but experts have offered several theories to explain the phenomenon—ranging from the fatigue of overworked nerve cells to possible brain damage from a viral infection to a combination of mechanisms. The new weakness of PPS appears to be related to the degeneration of individual nerve terminals in the motor units. A motor unit is formed by a nerve cell (or motor neuron) in the spinal cord or brain stem and the muscle fibers it activates. The polio virus attacks specific neurons in the brain stem and spinal cord. In an effort to compensate for the loss of these motor neurons, surviving

cells sprout new nerve-end terminals and connect with other muscle fibers. These new connections may result in recovery of movement and gradual gain in power in the affected limbs.

Years of high use of these recovered but overly extended motor units add stress to the motor neurons, which over time lose the ability to maintain the increased work demands. This results in the slow deterioration of the neurons, which leads to loss of muscle strength. Restoration of nerve function may occur in some fibers a second time, but eventually nerve terminals malfunction and permanent weakness occurs. This hypothesis explains why PPS occurs after a delay and has a slow and progressive course. Through years of studies, scientists at the National Institute of Neurological Disorders and Stroke (NINDS) and at other institutions have shown that the weakness of PPS progresses very slowly. It is marked by periods of relative stability, interspersed with periods of decline.

How is PPS diagnosed?

The diagnosis of PPS relies nearly entirely on clinical information. There are no laboratory tests specific for this condition and symptoms vary greatly among individuals. Physicians diagnose PPS after completing a comprehensive medical history and physical examination, and by excluding other disorders that could explain the symptoms.

Physicians look for the following criteria when diagnosing PPS:

- *Prior paralytic poliomyelitis with evidence of motor neuron loss.* This is confirmed by history of the acute paralytic illness, signs of residual weakness and atrophy of muscles on neuromuscular examination, and signs of motor neuron loss on electromyography (EMG). Rarely, people had subtle paralytic polio where there was no obvious deficit. In such cases, prior polio should be confirmed with an EMG study rather than a reported history of nonparalytic polio.
- *A period of partial or complete functional recovery after acute paralytic poliomyelitis,* followed by an interval (usually 15 years or more) of stable neuromuscular function.
- *Slowly progressive and persistent new muscle weakness or decreased endurance, with or without generalized fatigue, muscle atrophy, or muscle and joint pain.* Onset may at times follow trauma, surgery, or a period of inactivity, and can appear to be sudden. Less commonly, symptoms attributed to PPS include new problems with breathing or swallowing.
- *Symptoms that persist for at least a year.*
- *Exclusion of other neuromuscular, medical, and skeletal abnormalities as causes of symptoms.*

PPS may be difficult to diagnose in some people because other medical conditions can complicate the evaluation. Depression, for example, is associated with fatigue and can be misinterpreted as PPS. A number of conditions may cause problems in persons with polio that are not due to additional loss of motor neuron function. For example, shoulder osteoarthritis from walking with crutches, a chronic rotator cuff tear leading to pain and disuse weakness, or progressive scoliosis causing breathing insufficiency can occur years after polio but are not indicators of PPS.

Polio survivors with new symptoms resembling PPS should consider seeking treatment from a physician trained in neuromuscular disorders. It is important to clearly establish the origin and potential causes for declining strength and to assess progression of weakness not explained by other health problems. Magnetic resonance imaging (MRI) and computed tomography (CT) of the spinal cord, electrophysiological studies, and other tests are frequently used to investigate the course of decline in muscle strength and exclude other diseases that could be causing or contributing to the new progressive symptoms. A muscle biopsy or a spinal fluid analysis can be used to exclude other, possibly treatable, conditions that mimic PPS. Polio survivors may acquire other illnesses and should always have regular check-ups and preventive diagnostic tests. However, there is no diagnostic test for PPS, nor is there one that can identify which polio survivors are at greatest risk.

How is PPS treated?

There are currently no effective pharmaceutical treatments that can stop deterioration or reverse the deficits caused by the syndrome itself. However, a number of controlled studies have demonstrated that non-fatiguing exercises may improve muscle strength and reduce tiredness. Most of the clinical trials in PPS have focused on finding safe therapies that could reduce symptoms and improve quality of life.

Researchers at the National Institutes of Health (NIH) have tried treating persons having PPS with high doses of the steroid prednisone and demonstrated a mild improvement in their condition, but the results were not statistically significant. Also, the side effects from the treatment outweighed benefits, leading researchers to conclude that prednisone should not be used to treat PPS.

Preliminary studies indicate that intravenous immunoglobulin may reduce pain and increase quality of life in Post-Polio survivors.

A small trial to treat fatigue using lamotrigine (an anticonvulsant drug) showed modest effect but this study was limited and larger, more controlled studies with the drug were not conducted to validate the findings. Although there are no effective treatments, there are recommended management strategies. Patients should consider seeking medical advice from a physician experienced in treating neuromuscular disorders. Patients should also consider judicious use of exercise, preferably under the supervision of an experienced health professional. Physicians often advise patients on the use of mobility aids, ventilation equipment, revising activities of daily living activities to avoid rapid muscle tiring and total body exhaustion, and avoiding activities that cause pain or fatigue lasting more than 10 minutes. Most importantly, patients should avoid the temptation to attribute all signs and symptoms to prior polio, thereby missing out on important treatments for concurrent conditions.

Learning about PPS is important for polio survivors and their families. Managing PPS can involve lifestyle changes. Support groups that encourage self-help, group participation, and positive action can be helpful. Counseling may be needed to help individuals and families adjust to the late effects of poliomyelitis. Experiencing new symptoms of weakness and using assistive devices may bring back distressing memories of the original illness.

What is the role of exercise in the treatment of PPS?

Pain, weakness, and fatigue can result from the overuse of muscles and joints. These same symptoms also can result from disuse of muscles and joints. This fact has caused a misunderstanding about whether to encourage or discourage exercise for polio survivors or individuals with PPS.

Exercise is safe and effective when carefully prescribed and monitored by experienced health professionals. Exercise is more likely to benefit those muscle groups that were least affected by polio. Cardiopulmonary endurance training is usually more effective than strengthening exercises, especially when activities are paced to allow for frequent breaks and strategies are used to conserve energy. Heavy or intense resistive exercise and weight-lifting using polio-affected muscles may be counterproductive, as this can further weaken rather than strengthen these muscles.

Exercise prescriptions should include

- the specific muscle groups to be included,
- the specific muscle groups to be excluded, and
- the type of exercise, together with frequency and duration.

Exercise should be reduced or discontinued if it causes additional weakness, excessive fatigue, or unduly prolonged recovery time that is noted by either the individual with PPS or the professional monitoring the exercise. As a general rule, no muscle should be exercised to the point of causing ache, fatigue, or weakness.

Can PPS be prevented?

Polio survivors often ask if there is a way to prevent the development of PPS. Presently, no intervention has been found to stop the deterioration of surviving neurons. Physicians recommend that polio survivors get a good night's sleep, maintain a well-balanced diet, avoid unhealthy habits such as smoking and overeating, and follow a prescribed exercise program. Lifestyle changes, such as weight control, the use of assistive devices, and taking certain anti-inflammatory medications, may help with some of the symptoms of PPS.

What research is being done?

Scientists are working on a variety of investigations that may one day help individuals with PPS. Some basic researchers are studying the behavior of motor neurons many years after a polio attack. Others are looking at the mechanisms of fatigue and are trying to discover the roles played by the brain, spinal cord, peripheral nerves, neuromuscular junction (the site where a nerve cell meets the muscle cell it helps activate), and muscles.

Determining if there is an immunological link in PPS is also an area of interest. Researchers who discovered inflammation around motor neurons or muscles are trying to find out what causes this immunological response.

Where can I get more information?

BRAIN

P.O. Box 5801

Bethesda, MD 20824

800-352-9424

<http://www.ninds.nih.gov>

Post-Polio Health International

4207 Lindell Blvd.

#110

St. Louis, MO 63108-2930

info@post-polio.org

<http://www.post-polio.org>

Tel: 314-534-0475

Fax: 314-534-5070

POLIO EPIC, Inc.
Serving Arizona Polio Survivors since 1985
P.O. Box 17556 Tucson, Arizona 85731-7556
contact@polioepic.org www.polioepic.org

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