Is Post-Polio Syndrome Progressive?

An Answer to that Question from Primary Care Physician

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Many polio survivors hear from health care professionals (and others) that Post-Polio Syndrome is *not* progressive, that it reaches a point and does not progress any further. That was the situation that prompted a question from a reader of this newsletter a month or two ago. The correct answer/response is that PPS is generally a *slowly progressive disease process.*

It is true that the original polio infection caused a certain amount of damage over the course of a few weeks, then patients had a certain amount of recovery that usually ceased after 3 - 4 years. After that they usually neither got any worse or got any better for a period of several years. That is *not* true for PPS. In *most* cases it does slowly progress. Because no one can accurately measure someone's pain or fatigue, medical professionals can only measure the muscle weakness aspect of PPS. The studies done 30 or more years ago by Agre showed an average decrease in muscle strength of 1% per year in those with PPS. I personally can attest to it getting worse over time, although for me the worsening of muscle strength that required a change in bracing has occurred about once every 10-15 years.

I would like to share the Post-Polio Syndrome Fact Sheet from the NIH.

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. Post-polio syndrome (PPS) is a non-contagious condition that can affect polio survivors usually 15 to 40 years after recovery from polio. Only a polio survivor can develop PPS, it is not contagious.

The polio vaccine has eradicated polio from the United States. However, polio still exists in some countries and cases of PPS still arise.

What are its symptoms?

Most often, polio survivors start to experience gradual new weakening in muscles that were previously affected by the polio infection. Some individuals experience only minor symptoms while others develop visible muscle weakness and atrophy. A person who was more acutely affected by polio and who attained a greater recovery may experience a more severe case of PPS.

Symptoms include:

- Slowly progressive muscle weakness
- Fatigue
- A gradual decrease in the size of muscles (muscle atrophy)
- Loss of muscle function
- Pain from joint degeneration and increasing skeletal deformities such as curvature of the spine (scoliosis)

PPS 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.

What causes PPS?

The cause of PPS is unknown. 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. 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 results in the slow deterioration of the neurons and 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 may be why PPS occurs after a delay and has periods of relative stability combined with periods of decline with progressive weakness.

How is PPS diagnosed?

There are no laboratory or diagnostic tests specific for PPS. Physicians diagnose the condition 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 muscle atrophy, and signs of motor neuron loss on electromyography (EMG).
- *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.

Diagnostic tests include:

- Magnetic resonance imaging (MRI) and computed tomography (CT) of the spinal cord
- Electrophysiological studies and other tests to investigate the course of decline in muscle strength and exclude other diseases that could be causing or contributing to the new progressive symptoms
- Muscle biopsy
- Spinal fluid analysis to exclude other, possibly treatable, conditions that mimic PPS

How is PPS treated?

There are currently no effective treatments that can stop deterioration or reverse the deficits caused by the syndrome itself, but there are recommended management strategies. Individuals with PPS should seek medical advice from a physician experienced in treating neuromuscular disorders.

A number of research studies have demonstrated that non-fatiguing exercises (those that do not cause pain or fatigue lasting more than 10 minutes) may improve muscle strength and reduce tiredness. Cardiopulmonary endurance training also is helpful. Exercise should be considered under the supervision of an experienced health professional.

Mobility aids, ventilation equipment, and revising activities of daily living activities can help to avoid rapid muscle tiring and total body exhaustion.

Counseling may help individuals and families adjust to the late effects of poliomyelitis. Support groups that encourage self-help, group participation, and positive action can be helpful.

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?

At the National Institutes of Health (NIH), the National Institute of Allergy and Infectious Diseases (NIAID) is the leading funder of PPS research. The National Institute of Neurological Disorders and Stroke (NINDS) is the leading funder of research on neuromuscular disorders. NIH is the leading supporter of biomedical research in the world.

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 learn more about its possible causes. Researchers also are developing and refining interventions to help people with chronic conditions more effectively manage fatigue and sleep disturbances.

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

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