# Characteristics and Management of Postpolio Syndrome

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POSTPOLIO SYNDROME (PPS) REFERS to new, late manifestations occurring many years after acute poliomyelitis infection. Over the last 25 years, PPS has become a relatively common problem encountered by primary care physicians. A 1987 National Health Interview Survey estimated that about half of the 640 000 survivors of paralytic poliomyelitis in the United States had new late manifestations of PPS.[1] Subsequent studies in the 1990s have found the occurrence of PPS among patients with previous poliomyelitis to range from 28.5% to 64%.[2-4] The average time in various reports from the acute poliomyelitis until the onset of PPS is about 35 years, with a range from 8 to 71 years.[1] However, it is unclear if the occurrence of PPS increases with aging, which may be the case based on the most accepted etiologic hypothesis. The large number of PPS cases presently being seen is probably due to the poliomyelitis epidemics of the 1940s and 1950s.[1]

Risk factors for the development of PPS include the severity of the acute poliomyelitic paralysis, age at onset of the acute poliomyelitis (higher risk with adolescent and adult onset), the amount of recovery, and greater physical activity during the intervening years.[1,3,5] The course of PPS has been reported to have an average progression based on strength measurements of 1% to 2% per year.[6-8] Despite the fact that the course is relatively slow, PPS can present with multiple symptoms and signs.

## Clinical Manifestations

The criteria for the diagnosis of PPS used by most investigators and clinicians in the field were first described by Mulder et al[9] in 1972. These criteria include (1) a prior episode of poliomyelitis with residual motor neuron loss (can be confirmed by typical history, neurologic examination, or electromyography); (2) a period (usually ≥15 years) of neurologic and functional stability after recovery from the acute illness; (3) the gradual or rarely abrupt onset of new weakness or abnormal muscle fatigue, muscle atrophy, or generalized fatigue; and (4) exclusion of other conditions that could cause similar manifestations.[1]

The manifestations of PPS have been found to be similar around the world (TABLE 1).[1,7,10,11] Generalized fatigue is one of the most commonly reported manifestations (Table 1)[1]; it is often described as a generalized disabling exhaustion, tiredness, or a lack of energy that occurs with only minimal activity, often referred to as the "polio wall." When severe, the fatigue can affect mental as well as physical functioning, making it difficult for patients to concentrate, which has brought about a controversial hypothesis that the generalized fatigue is due to impaired brain function rather than to the diffuse disintegration of motor units and neuromuscular junctions.[12]

New weakness, sometimes accompanied by atrophy, is the most significant neurologic problem and has been referred to as postpolio progressive muscular atrophy. The new weakness is usually asymmetric and can be proximal, distal, or patchy. The weakness is usually slowly progressive and can occur in muscles previously affected (either partially or fully recovered) or clinically unaffected during the acute poliomyelitis (Table 1). Electromyographic studies indicate that many clinically unaffected muscles were involved subclinically during the acute episode of poliomyelitis.[1] Previously affected muscles are more likely than clinically unaffected muscles to later become weak (Table 1). Abnormal muscle fatigue (decreased endurance) has also been seen in patients with PPS and may be the forerunner of new weakness.[1,13] This abnormal muscle fatigue manifests as increased weakness after heavy overuse with delayed recovery after several days of rest.[14] Muscle pain (myalgias) also occurs and appears to be due to overuse of weak muscles. Muscle tenderness may be present on palpation. Fasciculations, cramps, and muscle pseudohypertrophy may be seen with or without new weakness.[1] In addition to generalized arm, leg, and body weakness, new weakness may result in respiratory insufficiency, bulbar muscle dysfunction (dysphagia, dysarthria, aphonia, facial weakness),[1,15] and sleep apnea.[1,16,17] Respiratory failure most often occurs in those with residual respiratory insufficiency and minimal reserve.[1]

Joint pain from joint instability is primarily a musculoskeletal problem and can occur without new weakness. Such pain often relates to excessive daily physical activity.[18] Residual weakness can lead to long-term overstressing of joints and periarticular soft-tissue structures (tendons, ligaments). Pain may also occur because of failing joint fusions, failing tendon transfers, uneven limb size, progressive scoliosis, poor posture, and abnormal mechanics.[1] These joint problems frequently lead to loss of mobility. Pain may also be a confounding factor for new weakness and/or reduced endurance in many patients with PPS. Patients with significant joint pain may limit their physical activity, which can lead to disuse weakness and atrophy. It is important to

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recognize this possible cause for declining muscle function in patients with PPS because appropriate treatment of the joint pain may slow down or arrest the decline.

## Pathophysiology

The origin of PPS is unknown. The leading hypothesis is that an excessive metabolic stress on remaining motor neurons over many years eventually results in the dropout of the new nerve terminals and eventually the motor neurons themselves.[8,19,20] This theory is supported by electromyographic and muscle biopsy studies.[1] Indirect evidence suggests that the excessive metabolic stress might be due to muscle overuse for many years.[5,13,14,21] Autopsy studies have revealed inflammation in the spinal cords of 16 of 17 patients with PPS.[1] This finding could be consistent with a persistent poliovirus infection, an immune-mediated (autoimmune) syndrome, or just a response to degenerating neurons. A persistent poliovirus infection seems unlikely,[22] but there are proponents to this theory.[23] The immune-mediated hypothesis has even less supportive data.[1]

## Laboratory Studies

Laboratory studies are primarily used to identify or eliminate other diseases. Routine blood test results are typically normal except for elevated creatine kinase in a minority of patients, which suggests muscle overuse.[1] Studies of the cerebrospinal fluid may reveal a nonspecific protein elevation. Electromyographic studies can identify changes consistent with previous poliomyelitis, but cannot separate those with PPS from asymptomatic patients with previous polio.[1,19] Electromyographic and nerve conduction studies are most useful for identifying and excluding other diseases (radiculopathy, neuropathy, myopathy). Imaging studies (eg, computed tomography, magnetic resonance imaging) are often needed to exclude spine disease, such as spondylosis or spinal stenosis.

## Management

Generalized fatigue is best treated with lifestyle changes consisting of energy conservation measures (pacing of physical activities combined with frequent rest periods and daytime naps), weight-loss programs, and the use of assistive devices (orthoses, canes, intermittent use of wheelchairs) (TABLE 2).[24] The use of lower extremity orthoses (ankle-foot, knee-ankle-foot) can aid in energy conservation and decrease fatigue.[24,25] Pharmacological agents (amantadine hydrochloride, pyridostigmine bromide, amitriptyline hydrochloride, fluoxetine hydrochloride, pemoline) may have a role in the amelioration of generalized fatigue, but only 2 of these agents (amantadine and pyridostigmine) have been studied in a controlled fashion, and both were found to lack benefit.[26,27]

The most important advance in the treatment of new weakness in patients with PPS is the finding in at least 8 studies that mild-to-moderate weakness can be improved with nonfatiguing exercise (Table 2).[1,28,29] All of these studies demonstrated increased muscle strength and none showed laboratory evidence of muscle overuse (eg, increase in serum creatine kinase or electromyographic or biopsy evidence of muscle damage).[1,28,29] These nonfatiguing exercise programs have used both submaximal and maximal strength combined with short-duration repetitions. The caveat to each of the above exercise studies was that overuse should be avoided. Patients should exercise for short intervals and then rest to recover between the bouts

**Table 1.** Most Common New Late Manifestations of Poliomyelitis in Patients Referred to Postpolio Clinics*

| Manifestation | No. (%) of Patients | | | |
|---|---|---|---|---|
| | Houston, Tex 1984-1985 (N = 132)† | Madison, Wis (N = 79)‡ | Syracuse, NY (N = 200)§ 1990-1993 (n = 100) | Syracuse, NY (N = 200)§ 1993-1997 (n = 100) |
| Generalized fatigue | 117 (89) | 68 (86) | 83 (83) | 86 (86) |
| Joint pain | 94 (71) | 61 (77) | 72 (72) | 73 (73) |
| Muscle pain | 94 (71) | 68 (86) | 74 (74) | 73 (73) |
| Weakness | | | | |
| Previously affected muscles | 91 (69) | 63 (80) | 88 (88) | 88 (88) |
| Previously unaffected muscles | 66 (50) | 42 (53) | 61 (61) | 59 (59) |
| Total new | NA | 69 (87) | 95 (95) | 90 (90) |
| Atrophy | 37 (28) | 31 (39) | 59 (59) | 52 (52) |
| Cold intolerance | 38 (29) | 44 (56) | 49 (49) | 53 (53) |
| Respiratory insufficiency | NA | 31 (39) | 42 (42) | 36 (36) |
| Dysphagia | NA | 24 (30) | 27 (27) | 36 (36) |

*Modified from Jubelt and Druker.[1] NA indicates data not available.
†Data from Halstead and Rossi.[10] All patients met criteria for postpolio syndrome.
‡Data from Agre et al[11]; the dates of the study were not provided. All patients had histories and examinations compatible with diagnosis of previous poliomyelitis.
§All patients had histories and examinations compatible with diagnosis of previous poliomyelitis. The exact study dates were January 1, 1990 to August 31, 1993, and September 1, 1993 to December 31, 1997.

**Table 2.** Evidence-Based Treatment for Patients With Postpolio Syndrome*

- Generalized fatigue
  - Institute lifestyle changes including energy conservation and weight-loss programs[24]
  - Prescribe appropriate lower extremity orthoses[24,25]
- Muscle weakness and fatigue
  - Prescribe nonfatiguing strengthening exercise program[1,28-30]
  - Institute physical activity pacing with rest periods[31]
  - Avoid overuse of weakened muscles[14]
- Bulbar muscle weakness
  - Respiratory failure
    - Noninvasive positive-pressure ventilation at night and as needed[32]
    - Tracheostomy and permanent ventilation
  - Dysphagia
    - Instruction on swallowing techniques[15]
- Musculoskeletal pain and joint instability
  - Decrease mechanical stress on joints and muscles with lifestyle changes[24] such as weight loss, decrease activities causing overwork, return to using assistive devices (including orthoses,[25] wheelchairs, and adaptive equipment)
  - Prescribe anti-inflammatory medications, heat, massage
- Cardiopulmonary conditioning
  - Cycle or arm ergometer exercise[28,33]
  - Aquatic exercise training[34,35]

*All of the interventions are evidence-based except for tracheostomy and permanent ventilation; and prescribing anti-inflammatory medications, heat, and massage.

of exercise. Exercise was also usually performed on alternate days to allow for full recovery and to avoid overuse. A physical therapist is needed to instruct the patient with PPS on appropriate exercise techniques and to monitor the patient so that he/she does not overexert himself/herself. Patients with PPS who were able to exercise at a level that avoided overuse (excessive muscle fatigue or increasing muscle or joint pain) have experienced positive results.[30] Muscle fatigue, similar to the treatment of generalized fatigue, can be improved by interspersing bouts of activity with rest breaks (pacing) to avoid excessive fatigue.[31] This simple procedure significantly improved strength recovery after activity.[31] Patients with PPS who exert their weak muscles to the point of exhaustion (overuse) may require 2 to 3 days to recover from the resulting muscle fatigue. Pharmacological intervention to increase muscle strength (controlled trials with pyridostigmine and prednisone) has not been found to be beneficial.[26,27]

Bulbar muscle weakness may lead to respiratory failure, dysphagia, and sleep disorders. Respiratory insufficiency in patients with PPS can often be managed with nighttime noninvasive positive-pressure ventilation.[32] Only a small minority of patients eventually require treacheostomy and permanent ventilation. Incentive spirometry might be useful, but it has not been studied as a treatment for patients with PPS. Pneumonia and influenza vaccines are probably indicated. Smoking should be eliminated and obstructive disease treated. Dysphagia can be improved with instruction on swallowing techniques.[15] Sleep disorders occur frequently in patients with PPS and it is important to determine if these are due to respiratory insufficiency or sleep apnea. Sleep apnea may be central, obstructive, or mixed and is treated similarly to sleep apnea seen in patients without PPS.[1]

Musculoskeletal pain (joint or muscle pain) and joint instability can be treated by pacing activities,[18] making lifestyle changes,[24] and decreasing mechanical stress with bracing and wheelchairs.[25] The judicious use of anti-inflammatory medications, heat, and massage appear to be somewhat beneficial, but have not been studied objectively.

Part of the management of PPS includes the exclusion of medical problems (eg, anemia, thyroid disease, cancer) that may produce symptoms similar to those seen in PPS (fatigue, pain, and weakness). Related problems may also include respiratory insufficiency and cardiac failure. Cardiac conditioning is important for all patients, and 3 trials using aerobic or general conditioning exercise with lower-limb cycle or arm ergometers have shown significant improvement in cardiorespiratory fitness in patients with PPS but without deleterious adverse effects.[28,30,33] Two studies have demonstrated that flexibility, strength, and cardiorespiratory fitness can be improved in patients with PPS through aquatic exercise programs.[34,35] Again, similar to individual muscle-strengthening programs, muscle overuse must be avoided. Finally, it is important to remember that psychological symptoms related to the reemergence of a supposedly resolved problem and to the stresses of the required major lifestyle changes can be overwhelming at times.[1]

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