

JULIE K. SILVER, MD

Medical Director, Spaulding-Framingham Outpatient Center, Framingham, Mass; Assistant professor, Department of Physical Medicine and Rehabilitation, Harvard Medical School, Boston, Mass

DOROTHY D. AIELLO, PT

Senior physical therapist, Spaulding-Framingham Outpatient Center, Framingham, Mass

What internists need to know about postpolio syndrome

■ ABSTRACT

Decades after recovery from polio, many patients develop new muscle weakness and other symptoms that can lead to increased debility. Treatment is aimed at the most prominent symptoms. Medications may help, as well as physical therapy and a carefully paced exercise program. Screening for osteopenia and osteoporosis is recommended.

■ KEY POINTS

Postpolio syndrome affects an estimated 60% of "paralytic" polio survivors, plus unknown numbers of patients who had subclinical polio.

Postpolio syndrome is a diagnosis of exclusion. Symptoms are related to new muscle weakness and may include muscle atrophy, myalgias, fatigue, and problems with swallowing and breathing.

No drugs specifically address postpolio syndrome. Pyridostigmine has had mixed results for treating weakness and fatigue, as have methylphenidate and bromocriptine. Modafinil may be helpful for fatigue. Nonsteroidal anti-inflammatory drugs are used to treat pain.

Rehabilitation professionals who have expertise in treating polio survivors can be valuable resources in preserving function and preventing deconditioning.

WHEN A POLIO SURVIVOR presents with nonspecific symptoms such as weakness and fatigue, how do you determine whether they are due to postpolio syndrome or to an unrelated problem?

Postpolio syndrome is a neurologic disorder defined by a collection of symptoms occurring decades after a patient has recovered from an initial infection with the poliovirus. New muscle weakness is the hallmark, but breathing or swallowing problems, fatigue, myalgias, and cold intolerance are frequently also present.

In this review, we discuss the criteria for diagnosing postpolio syndrome, guidelines for ruling out other conditions, and treatment strategies to optimize function in postpolio patients.

■ 1 MILLION POLIO SURVIVORS

There are probably at least 1 million polio survivors in the United States, though not all have residual effects. Worldwide, there are millions more. Polio eradication is ongoing, and it is hoped that new cases will be completely eliminated over the next few years.

■ ACUTE POLIOMYELITIS MAY BE SUBCLINICAL

Historically and even recently, acute poliomyelitis has been thought of as having distinct presentations:

Abortive polio, which presents as a minor illness of fever, malaise, sore throat, anorexia, myalgias, and headache

Nonparalytic polio, which presents as aseptic meningitis

Paralytic polio, which presents as severe back, neck, and muscle pain, with the rapid or



gradual development of paralysis.¹

In fact, however, the acute viral illness is probably more of a spectrum, in which there are subclinical cases of paralysis that in the past would have been classified as nonparalytic.² This concept is important because although most patients who are at risk for postpolio syndrome had well-recognized “paralytic” polio, others who were never diagnosed with polio or were thought to have had “nonparalytic” polio may also be at risk for postpolio syndrome.

An estimated 60% of “paralytic” polio survivors are affected by postpolio syndrome.³ The prevalence in those who had subclinical illness is unknown.

■ WHAT CAUSES THE LATE SYMPTOMS?

Postpolio syndrome occurs in polio survivors who had injury to their central nervous system, generally the anterior horn cells in the spinal cord, during the initial infection. The cause of the late symptoms is not well understood but is believed to involve attrition of motor neurons during aging.⁴ Other theories abound, however, and the etiology is likely multifactorial.

When motor neurons are lost in acute polio, the surviving motor neurons sprout collateral fibers that reinnervate the denervated muscle fibers (FIGURE 1). The resulting motor units are larger than normal, and there are fewer of them than before. Therefore, the burden on each of these remaining motor neurons is higher than under normal conditions.

With age, we all gradually lose some motor neurons.⁵ Polio survivors may be more affected by this loss of motor neurons because they have fewer to begin with.

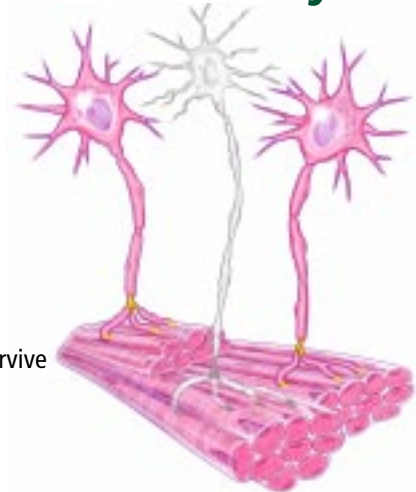
Another theory is that insufficient levels of acetylcholine are released at the neuromuscular junction, resulting in diminished muscle contraction.⁵

Maselli et al⁶ noted reduced amplitudes of miniature end plate potentials and structural abnormalities of the neuromuscular junction, such as reduced diameter of nerve terminals, but these changes were not noted in all postpolio syndrome patients.

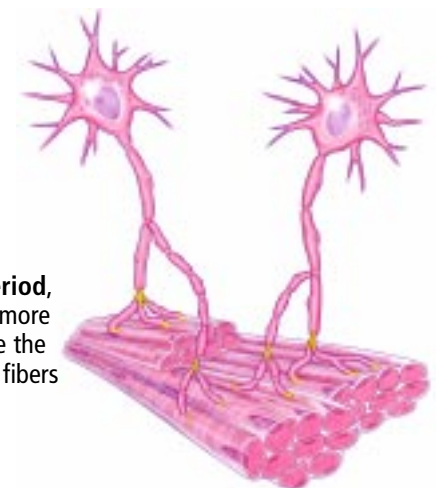
Some have found ongoing immune activation and defective viral particles in the spinal

■ What causes postpolio syndrome? One theory

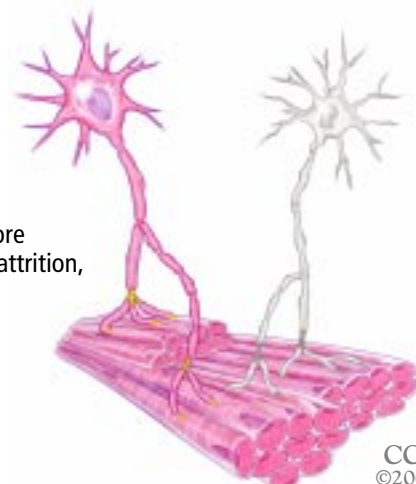
In acute polio, some neurons die, while others survive



In the stable postpolio period, the surviving neurons cover more territory than they did before the polio by sprouting collateral fibers



In postpolio syndrome, more neurons are lost by normal attrition, leading to muscle weakness



CCF
©2002

FIGURE 1

TABLE 1

Criteria for diagnosing postpolio syndrome

History of old polio, preferably with recent electrodiagnostic findings consistent with remote anterior horn cell disease

A period of at least partial recovery from the initial illness and then a long stable period (10–20 years or more)

New symptoms consistent with postpolio syndrome that are not attributable to any other medical condition; these may include weakness, myalgias, fatigue, swallowing problems, breathing problems, cold intolerance, and muscle atrophy

TABLE 2

**Not available for online publication.
See print version of the
*Cleveland Clinic Journal of Medicine***

Postpolio muscle pain classically occurs in muscles rather than joints

fluid,⁷ although the significance of these is unclear.

General fatigue may also have a central cause—an abnormality in the reticular activating system in the brain that occurred during the acute polio episode.^{5,7}

■ HALLMARK IS NEW WEAKNESS

The hallmark symptom of postpolio syndrome is new weakness, which may occur in muscles known to be previously affected or in muscles that were thought to be normal.³ The patient may report difficulty with walking or lifting items, falls, needing more assistance with transfers (eg, moving from the bed or commode to the wheelchair), and being less able to do functional tasks. The weakness characteristically worsens with increased activity and is most pronounced at the end of the day.

Symptoms may also include dyspnea on exertion due to respiratory muscle weakness, other breathing or swallowing problems, pain (myalgias), cold intolerance, and unaccustomed fatigue. New muscle atrophy may also be present.⁸

Pain can be due to factors related to the history of polio, but which are not classifiable as postpolio syndrome. For example, a patient

may present with left leg paralysis due to the initial polio and report increased limping and pain in the right hip (ie, the “good” leg). The new symptoms may be due to osteoarthritis of the hip, which is more likely to occur in a polio survivor without good muscular support around the hip and after years of additional wear and tear.

Postpolio muscle pain classically occurs in the muscles rather than in the joints. The pain is often described as aching, cramping, burning, or a “tired” feeling. It frequently occurs at night or after the person has been very active.⁸

Numbness or paresthesias are not typical symptoms of postpolio syndrome.

■ DIAGNOSING POSTPOLIO SYNDROME

Postpolio syndrome is a diagnosis of exclusion and should fit specific criteria (TABLE 1). Screening for other possible diagnoses is essential.

Evaluating weakness

In postpolio syndrome, weakness progresses gradually over months to years. This muscle fatigue is associated with overuse and worsens with increased activity. Rapid loss of strength



over weeks to months should suggest another diagnosis (TABLE 2).

Evaluating fatigue

Generalized fatigue has many possible causes (TABLE 3) that should be ruled out with screening tests (TABLE 4).

Since sleep disorders are common in polio survivors,^{9,10} referral to a sleep clinic should be considered. In our practice, we often see patients who feel tired but say they have no trouble sleeping. However, many of them test positive for sleep disturbances, including obstructive sleep apnea (characterized by morning fatigue, snoring, and difficulty sleeping supine) and random limb movement disorder (characterized by morning muscle pain and overall fatigue).

Depression, thyroid dysfunction, or both may be present as coexisting conditions and also contribute to fatigue.

Evaluating pain

Pain related to postpolio syndrome is due to muscle overuse or biomechanical problems, or both. Treatment of coexisting orthopedic problems is vital to alleviate pain and improve function (TABLE 5).

Evaluating respiratory problems

Difficulty breathing can be life-threatening. Conditions other than postpolio syndrome should be considered (TABLE 6).

Polio survivors with a weakened diaphragm breathe shallowly and experience dyspnea on exertion. New respiratory muscle weakness causes restrictive lung disease, which is associated with chronic alveolar hypoventilation. In obese patients, excess weight over the thoracic cage and abdominal cavity worsens the condition.

Pulmonary function tests can be used for diagnosis and to determine if supplemental oxygen is necessary. Arterial blood gas measurements and pulse oximetry can also help with the diagnosis. Referral to a pulmonologist is indicated if a patient requires a respirator.

All polio survivors who undergo surgery need special precautions beforehand to avoid potential problems with respiratory sequelae and ventilator weaning. These include pulmonary function testing and consideration of

TABLE 3

**Not available for online publication.
See print version of the
*Cleveland Clinic Journal of Medicine***

TABLE 4

**Not available for online publication.
See print version of the
*Cleveland Clinic Journal of Medicine***

TABLE 5

**Not available for online publication.
See print version of the
Cleveland Clinic
Journal of Medicine**

TABLE 6

Differential diagnosis for respiratory problems

Cardiac disease
Chronic obstructive pulmonary disease
Asthma
Anemia
Deconditioning

Even well-appearing polio survivors can have restrictive lung disease

alternatives to general anesthesia whenever possible. Even well-appearing polio survivors can have significant restrictive lung disease due to paralysis of respiratory muscles.

Evaluating swallowing difficulties

Swallowing problems can also be life-threatening. In view of the risk of choking, family members should be educated in the Heimlich maneuver. Patients presenting with difficulty swallowing may require specific testing, such as a modified barium swallow, or referral to a specialist for further evaluation and intervention.

TREATMENT RECOMMENDATIONS

Treatment of postpolio syndrome should focus on the most prominent symptoms and can include:

- Medications
- Supplemental oxygen

- Physical, occupational, and speech therapy
- An exercise program to preserve mobility and prevent deconditioning.

Medications

Drug therapy for postpolio syndrome has been generally disappointing. No medications specifically address postpolio syndrome. That said, many medications may play an important role in alleviating symptoms.

For weakness and fatigue, pyridostigmine (Mestinon), usually given as an oral dose of 60 mg three times a day, has had somewhat mixed results. One study of postpolio syndrome patients found that it improved upper extremity subjective strength and fatigue.¹¹ Another found no significant difference between patients taking pyridostigmine and placebo, except that very weak muscles (25% or less of baseline) were minimally stronger with pyridostigmine.¹²

Methylphenidate hydrochloride (Ritalin) and bromocriptine (Parlodel) have been tried for postpolio patients with chronic debilitating fatigue, also with mixed results.^{7,13}

Modafinil (Provigil) has been used to treat fatigue^{14,15} and may be useful in polio survivors. The starting dose is usually 200 mg orally in the morning and may be increased to 400 mg each morning or given in divided doses.

Side effects can be a problem with these medications. Pyridostigmine's side effects are generally dose-related and can be recalled by the acronym SLUD (increased salivation, lacrimation, urination, and defecation). Respiratory secretions may also be increased with this medication. Modafinil's side effects include headache, nausea, and nervousness, and modafinil may increase circulating levels of diazepam, phenytoin, and propranolol.

Respiratory problems often improve with continuous positive airway pressure or bilevel positive airway pressure at night. Oxygen can exacerbate chronic alveolar hypoventilation and should be used with caution. A physical therapist or occupational therapist skilled in treating respiratory disorders may be helpful in teaching the patient breathing and postural techniques and help the patient conserve energy to decrease respiratory demands.

Pain can be treated with traditional non-steroidal anti-inflammatory drugs, cyclo-oxy-



genase-2 inhibitors (particularly in elderly patients or those with a history of gastrointestinal problems), and nonnarcotic analgesics. Tramadol (Ultram) may be helpful in some patients but should be avoided in those with a history of seizures.

Other medications typically used for chronic pain may also be tried, such as tricyclic antidepressants and anticonvulsants. Tricyclic antidepressants have cholinergic side effects; the most serious is the possibility of acute urinary retention in men, especially if underlying prostate problems are present.

Injections with local anesthetics or corticosteroids or both may be effective for specific conditions that are often associated with post-polio syndrome, such as myofascial pain, trochanteric bursitis, carpal tunnel syndrome, lateral epicondylitis, or rotator cuff tendonitis.

Physical and occupational therapy

From a quality-of-life perspective, perhaps the most important thing a physician can do is to help patients preserve mobility and avoid falls and resultant injuries.

Physical and occupational therapists can be extremely helpful in treating patients with musculoskeletal pain, weakness, decreased endurance, impaired balance, and difficulty walking. They can recommend appropriate adaptive equipment, such as shower grab bars, a raised toilet seat, sturdy and lightweight braces, assistive devices such as canes and crutches, and footwear modifications such as heel lifts and lateral wedges. Therapists can also advise patients on how to pace themselves, which is especially important for polio survivors. Home safety, work simplification, falls prevention, and proper exercise are also strategies that can enhance function.

Exercise

One of the most common questions polio survivors ask is, "How should I exercise?" This has been much debated. General guidelines for patients:

- Maintain an active exercise program to avoid deconditioning and cardiovascular sequelae
- Avoid overly aggressive exercise (fatiguing)

- Resist the impulse to exercise through pain.

Muscle fibers of polio survivors have very limited endurance because of the loss of aerobic enzyme activity and greater reliance on anaerobic metabolic capacity.¹⁶ Cross-training programs, such as alternating cycling with swimming and walking, are a good way to involve different muscle groups, but such programs should be consistent in terms of repetitions, resistance, and time. For most people, using daily activities as a primary way to exercise is too erratic and may lead to overuse, fatigue, and further weakness.

Is a wheelchair needed?

For patients who are having difficulty with walking or who may be at risk for falls, a motorized wheelchair or scooter can be useful, either full-time or part-time. Such vehicles can improve functional mobility, decrease risk of falls, and help conserve energy.

Manual wheelchairs have the advantage over motorized wheelchairs of being lighter and easily folded for transport. However, manual wheelchairs tend to promote overuse syndromes in the arms and are generally recommended only when another person will push the patient.

Ancillary health care

Referral to other appropriate health care providers can markedly improve the quality of life for polio survivors. For example, speech and language pathologists can be extremely helpful in teaching patients compensatory mechanisms for swallowing. Referral to a mental health counselor, with pharmacologic intervention if needed, should be considered for patients who are depressed or have other psychological sequelae.

■ AVOIDING COMPLICATIONS

Osteoporosis. Patients with significant paralysis often have associated loss of bone density. Recent studies indicate that male polio survivors are at risk for osteopenia and osteoporosis, and may be at higher risk for fracture.^{8,16,17} We recommend that all polio survivors be screened for bone density loss and be appropriately treated.

Screen all polio survivors for bone density loss



Falls. Polio survivors are also at greater risk of tripping and falling due to poor balance and weak arms or legs, and are less likely to be able to protect themselves as they fall.^{18–21} Since the complications of a fall can be serious, interventions for fall prevention are crucial. Both physical and occupa-

tional therapists typically address fall prevention.

Upper extremity injuries. Because polio survivors tend to overuse their arms, they are also at risk for upper extremity injuries, including carpal tunnel syndrome and ulnar neuropathy.^{8,22–24}

REFERENCES

1. **Cohen JI.** Poliovirus. In: Fauci AS, Braunwald E, Isselbacher KJ, et al, editors. *Harrison's Principles of Internal Medicine*. 14th ed. New York: McGraw-Hill, 1998:1120–1121.
2. **Halstead LS, Silver JK.** Nonparalytic polio and postpolio syndrome. *Am J Phys Med Rehabil* 2000; 79:13–18.
3. **Gawne AC, Halstead LS.** Post-polio syndrome: pathophysiology and clinical management. *Crit Rev Phys Rehabil Med* 1995; 7:147–188.
4. **Dalakas MC.** Pathogenetic mechanisms of post-polio syndrome: morphological, electrophysiological, virological, and immunological correlations. *Ann NY Acad Sci* 1995; 753:167–185.
5. **Halstead LS.** *Managing Post-Polio: A Guide to Living Well with Post-polio Syndrome*. Washington, DC: NRH Press, 1998.
6. **Maselli RA, Wollmann R, Roos R.** Function and ultra-structure of the neuromuscular junction in post-polio syndrome. *Ann NY Acad Sci* 1995; 753:129–137.
7. **Dalakas MC.** Pathogenetic mechanisms of post-polio syndrome: morphological, electrophysiological, virological, and immunological correlations. *Ann NY Acad Sci* 1995; 753:167–185.
8. **Silver JK.** *Post-polio Syndrome: a Guide for Polio Survivors and their Families*. New Haven: Yale University Press, 2001.
9. **Bruno RL.** Abnormal movements in sleep as post-polio sequelae. *Am J Phys Med Rehabil* 1998; 77:339–343.
10. **Dean AC, Graham BA, Dalakas M, Sato S.** Sleep apnea in patients with postpolio syndrome. *Ann Neurol* 1998; 43:661–664.
11. **Seivert BP, Speier JL, Canine JK.** Pyridostigmine effect on strength, endurance and fatigue in post-polio patients [abstract]. *Arch Phys Med Rehabil* 1994; 75:1049.
12. **Trojan DA, Collet JP, Shapiro S, et al.** A multicenter, randomized, double-blind trial of pyridostigmine in postpolio syndrome. *Neurology* 1999; 53:1225–1233.
13. **Bruno RL, Zimmerman JR, Creange SJ, Lewis T, Molzen T, Frick NM.** Bromocriptine in the treatment of post-polio fatigue: a pilot study with implications for the pathophysiology of fatigue. *Am J Phys Med Rehabil* 1996; 75:340–347.
14. **Kingshott RN, Vennelle M, Coleman EL, Engleman HM, Mackay TW, Douglas NJ.** Randomized, double-blind, placebo-controlled crossover trial of modafinil in the treatment of residual excessive daytime sleepiness in the sleep apnea/hypopnea syndrome. *Am J Respir Crit Care Med* 2001; 163:918–923.
15. **Mitler MM, Harsh J, Hiroshkowitz M, Guilleminault C.** Long-term efficacy and safety of modafinil (PROVIGIL) for the treatment of excessive daytime sleepiness associated with narcolepsy. *Sleep Med* 2000; 1:231–243.
16. **Silver JK, Aiello DD.** Bone density and fracture risk in male polio survivors [abstract]. *Arch Phys Med Rehabil* 2001; 82:1329.
17. **Silver JK, MacNeil JR, Aiello DD.** Effect of Fosamax on bone density in a male polio survivor: a case report [abstract]. *Arch Phys Med Rehabil* 2001; 82:1329.
18. **Silver JK, Aiello DD.** Fall prevention strategies in a polio survivor: a case report [abstract]. *Arch Phys Med Rehabil* 2000; 81:1309.
19. **Silver JK, Aiello DD.** Polio survivors' attitudes regarding falls [abstract]. *Arch Phys Med Rehabil* 2000; 81:1296.
20. **Silver JK, Aiello DD.** Risk of falls in polio survivors [abstract]. *Arch Phys Med Rehabil* 2000; 81:1272.
21. **Silver JK, Aiello DD.** Polio survivors: falls and subsequent injuries. *Am J Phys Med Rehabil*. In press 2002.
22. **Veerendrakumar M, Taly AB, Nagaraja D.** Ulnar nerve palsy due to axillary crutch. *Neurol India* 2001; 49:67–70.
23. **Waring WP, Werner RA.** Clinical management of carpal tunnel syndrome in patients with long term sequelae of poliomyelitis. *J Hand Surg* 1989; 14:865–869.
24. **Slowman LS, Silver JK.** Prevalence of median and ulnar neuropathy in post-polio patients [abstract]. *Arch Phys Med Rehabil* 2001; 82:1312–1313.

ADDRESS: Julie K. Silver, MD, Spaulding-Framingham Outpatient Center, 570 Worcester Road, Framingham, MA 01702; e-mail jksilver@bics.bwh.harvard.edu.