Although polio is often considered a disease of the past, new symptoms are appearing in patients infected with the polio virus many years ago. Many patients who contracted a paralytic form of poliomyelitis 3, 4, or even 7 decades ago are now reliving their childhood symptoms in what is known as postpolio syndrome. (J Am Podiatr Med Assoc 89(4): 183-187, 1999)

Polio Today

Most physicians in the United States have never seen an active case of polio; on average, fewer than ten cases of polio a year were reported from 1975 to 1990. Worldwide, however, 6,197 cases were reported in 1995, according to the World Health Organization. However, more than half of the estimated half-million Americans who contracted a paralytic form of poliomyelitis 3, 4, or even 7 decades ago are now reliving their childhood symptoms in what is known as postpolio syndrome.

What Is Postpolio Syndrome?

One or 2 out of every 100 people infected with the polio virus developed an acute paralytic episode. In most cases, the paralysis was only temporary. Although the polio virus destroyed the nerve cells it attacked, the undamaged or partly damaged nerve cells took over the activities of the dead cells, sending sprouts to muscle fibers that had been “orphaned,” thereby restoring overall muscle strength. Years later, the nerve cells that have been carrying the workload of the destroyed nerve cells are burning out, and affected muscles are once again showing signs of fatigue and loss of innervation.

Signs and Symptoms

Postpolio syndrome is more common among those who had the worst cases of polio—those who needed hospital care, were older than age 10, had paralysis of
all four limbs, or needed mechanical assistance to breathe. In at least 1 out of 25 survivors, the postpolio symptoms are disabling. While aging and years of abnormal stress on weightbearing joints may have caused some of these complaints, they do not explain the progressive muscle weakness and atrophy. The initial symptoms of postpolio syndrome appear most frequently in the lower limb that had been most affected by the acute illness. It may be easier to compensate for weakness in the upper extremities without overuse, and therefore upper-limb weakness may be overlooked.

People with postpolio syndrome complain of many and varied symptoms, including the following:
- General fatigue. There may be unaccustomed fatigue after moderate exercise or usual activity, indicating muscle overuse.
- Joint and muscle pain. Musculoskeletal problems in most patients with postpolio syndrome include osteoarthritis of the spine and peripheral joints, scoliosis, bursitis, tendinitis, osteoporosis, myofascial pain syndrome, toe and foot deformities, carpal tunnel syndrome, and chronic pain from strain on back and neck muscles. These symptoms are generally sequelae of long-standing muscle weakness and imbalance, not merely part of the normal aging process. Chronic strain on joints used in an abnormal biomechanical fashion may occur as the body compensates for muscle weakness and may lead to premature degenerative osteoarthritis. Long-bone fractures from slight trauma due to osteoporosis are not uncommon. Generalized pain in the spine and extremities, as well as a diffuse influenza-like aching in muscles, is very common. Low-back pain becomes more common from the added stress caused by body movements made to compensate for weak hip muscles.
- Muscle weakness and loss of muscle use. Dysphagia is more common in those who have had bulbar polio. Loss of cough reflex may occur. Muscles that recovered well from the initial attack and have been used strenuously since then are the ones most commonly affected. Patients will note new weakness or pain after exercise as well as after general activity. Patients work two to three times harder to perform the activities they formerly did with relative ease. Abdominal muscles may weaken, leading to chronic back strain and injury.
- Respiratory problems and failing respiratory reserve. Insomnia, restless sleep, nightmares, morning headache or confusion, shallow breathing patterns, breathlessness (even during speaking), and sleep apnea may occur.
- Intolerance to cold. Slight changes in ambient air temperature can significantly alter muscle strength and dexterity, as well as cause pain and bluish discoloration of extremities due to blood pooling. As the feet cool excessively, the surrounding soft tissues become less elastic and subsequently become stiffer and harder to move.

Pathophysiology

Acute poliomyelitis is a viral infection, with the virus entering the body via the gastrointestinal tract. This usually causes mild gastrointestinal illness with influenza-like symptoms. However, the damage done by the virus in the central nervous system may lead to extensive paralysis or even death.

Although researchers are still not certain what causes postpolio syndrome, most agree with Lauro Halstead, MD, director of the National Rehabilitation Hospital’s Post-polio Program in Washington, DC, and a polio survivor, who says that the most widely held theory is that the new muscle weakness is related to overuse of polio-damaged nerve cells in the spinal cord.

In polio, motor nerve cells located in the anterior horn of the spinal cord are attacked (these are the nerve cells that ultimately control muscles). Some nerve cells are permanently destroyed, while others are only temporarily damaged. After an acute paralytic attack, the body does its best to repair itself. With proper therapy and exercise, people who have had polio can usually regain some strength during the first 2 years following the initial infection.

There are three ways in which this recovery occurs:
1) Temporarily damaged nerve cells recover and strength is regained. 2) Muscle fibers innervated by surviving nerve cells can increase in size and become stronger through strengthening exercises. 3) New nerve twigs sprout from the undamaged nerve supply.

Frederick M. Maynard, MD, believes that the nerve sprouts grow to reach muscle fibers that have lost their nerve supply. Surviving nerve cells are thus connected to more muscle fibers, and each nerve cell becomes responsible for an increased number of muscle fibers. The theory is that after years of this excess load, neurons of polio survivors burn out. As the muscles lose innervation, they atrophy, leading to fatigue and weakness. Subsequently, the muscle fibers that remain innervated attempt to compensate for lost muscle function; this only exacerbates the situation. In some cases, polio survivors are beginning to revisit their past, returning to braces and wheelchairs. Some people who had bulbar polio even need the assistance of ventilators to breathe.

The process that causes other symptoms of post-polio syndrome can also cause the feet to turn blue and cold as well as become stiff and difficult to move when it is even mildly cold outside. This condition
has been termed “polio feet.” The polio virus attacks anterior horn cell motor neurons, thus affecting the muscles around blood vessels. When these atrophy, there is no motor nerve stimulating contraction. The lack of contraction causes the blood to pool, especially in the veins of the lower extremities. Heat is then lost via the superficial vasculature. The result is overheating of the body, especially the feet, where blood tends to pool anyway. Cooling of the area may then lead to difficulty in functioning of the affected musculature, which is already working at a disadvantage.

Diagnosis

Although polio is diagnosed by identifying the virus in cerebrospinal fluid, a throat culture, or feces, the diagnosis of postpolio syndrome is one of exclusion. On the basis of a thorough history and physical examination, the physician must first rule out or treat other conditions that may be causing the symptoms. Because a large percentage of muscle function (one-third to one-half) can be lost before deficits appear on a muscle-strength test done in the office, the physical examination should be followed by ancillary studies, such as electromyography studies, for evidence of prior paralytic polio. Muscle atrophy is also an important finding in postpolio syndrome. It is not uncommon for recent trauma or illness to precipitate the symptoms after years of apparent stability.

One should rule out both the congenital peripheral neuropathies, such as Charcot-Marie-Tooth disease, Roussy-Lévy syndrome, Friedreich’s ataxia, and Dejerine-Sottas atrophy, and acquired diseases, such as cerebral palsy, Duchenne’s muscular dystrophy, cerebral vascular accident, and Parkinson’s disease, in which the patient might present with peripheral symptomatology, including the characteristic cavus foot deformity.

According to Jacquelin Perry, MD, chief of pathokinesiology and polio services at Rancho Los Amigos Medical Center in Downey, California, a diagnosis of postpolio syndrome should be based on three things: 1) a history of polio, 2) a period of some recovery followed by new loss of function, and 3) a physical examination that reveals scattered muscle weakness (observed during extensive manual muscle testing from head to toe), normal sensations, and reflexes that are normal (2+) for strong muscles and diminished (0 to 1+) for weak muscles.

Treatment

Treatment of nonparalytic polio usually includes bed rest and analgesics. For paralysis patients, physical therapy prevents muscle damage while the virus is active and preserves muscle function during recovery. For respiratory paralysis, a tracheotomy and artificial respirator may be necessary.

The treatment for postpolio syndrome is very different. Physical therapy and exercise of muscles that are already functioning at their maximum level could be deleterious. If, however, muscle groups are carefully selected, muscular capacity can be increased through isometric exercise. The patient must be educated to avoid potentially harmful exercise-induced fatigue. A decrease of muscle load through diet when the patient is overweight or the use of orthoses, including ankle-foot orthoses, to improve mechanical efficiency are effective treatments. Such complications as dysphagia, pulmonary dysfunction, or sleep disturbances may require specific referrals. Probably the most important aspect of treatment of postpolio syndrome is to discuss with patients the need to modify their work duties and lifestyle in order to make the most of whatever function they have left and to avoid problems from overuse.

Drug Treatment

Nonsteroidal anti-inflammatory drugs, as well as heat application to the affected area, may be used to relieve pain. Narcotic pain relievers are avoided because depressing activities of the central nervous system may adversely affect breathing.

Fatigue may be ameliorated with medications, such as amantadine hydrochloride, selegiline hydrochloride, and pyridostigmine bromide (which needs careful monitoring), that seem to raise the threshold of fatigue. These observations are still anecdotal and have yet to be confirmed by clinical trials.

A study at the National Institute of Neurological Disorders and Stroke, under the direction of Marinos Dalakas, MD, chief of the neuromuscular diseases unit, has evaluated the effects of prednisone on patients with postpolio syndrome in a double-blind study. Dalakas hypothesizes that the immune system may be involved in the syndrome, possibly in some kind of autoimmune disorder. Prednisone can suppress components of the immune system and is often helpful in relieving muscle weakness and pain in muscular inflammatory diseases, such as polymyositis.

According to Dalakas, as quoted in an article by Evelyn Zamula on the Polio Survivors’ Page on the World Wide Web,

We have found a number of irregularities in the immune system of PPS patients, which is why we’re using prednisone in this study. We don’t know if these changes are responsible for the manifestation
of muscle weakness. However, the use of prednisone as an agent that can modulate the immune system, as well as act as an anti-inflammatory agent, may prove to be effective in relieving some symptoms of PPS patients.

Research is also being done on L-carnitine and its effects on postpolio syndrome patients. L-carnitine improves the metabolism of oxygen, fat, and glucose and inhibits the use (abuse) of muscle proteins for energy production.3

Case Reports

Case 1

A 52-year-old woman presented with a chief complaint of pain and swelling in her left foot of 1 month’s duration. The patient claimed that her primary-care physician had initiated therapy to no avail. The patient related no history of known trauma coinciding with her recent left foot pain. However, she did relate that she “falls a lot.” She related not having worked for 8 years because of “difficulties in getting around.”

The patient had a medical history of poliomyelitis as an infant. She also related having lived in an area in which Lyme disease is endemic and having a history of a characteristic first-stage Lyme disease rash. The patient also related symptoms of chronic fatigue, dysphoria, and neurologic impairment with ataxia and difficulty swallowing. She also developed a generalized pain syndrome and difficulty with memory and speech. The former is consistent with a motor neuron disease, and the latter with encephalopathy as well as painful peripheral neuropathy.

The patient’s Lyme antibody test was negative; however, her erythrocyte sedimentation rate was 50 mm/h. She was treated with parenteral ceftriaxone by her primary-care physician and experienced an exacerbation of her pain, headaches, and fatigue. This exacerbation of symptoms is consistent with the initiation of antibiotics in late-stage Lyme disease. The antibiotic therapy resulted in improvements in her memory and speech. However, partial paresis of her legs and easy fatigability, which are consistent with postpolio syndrome, have persisted.

The patient presented with edema and erythema of the left foot and ankle. Upon examination, she also demonstrated an antalgic gait—she was able to walk unassisted for approximately one-quarter of a block before needing rest. Radiographs revealed a fracture of the anterior calcaneal process in the left foot; however, because there was no positive history of trauma, a bone scan was obtained. The bone scan revealed increased uptake over the anterior process of the left calcaneus, with some increased uptake noted at the talonavicular joint as well.

In light of the patient’s medical history, conservative therapy was deemed prudent. Initially, she was treated with a compressive dressing for the swelling, bracing, anti-inflammatory drugs, and contrast soaks. However, she continued to have pain. Subsequently, she was placed in a below-the-knee cast. The cast was removed at 2 weeks, when she complained of increased fatigue from “dragging the cast around.” Physical therapy was initiated but proved ineffective. She was then given multiple options, including shoe therapy (custom shoes), pads, anti-inflammatory drugs, injection therapy, and surgery. At this point, she refused further conservative care, and excision of the fracture fragment from the anterior process of the left calcaneus was performed. Postoperatively, the patient was placed in a removable cast walker. Although she experienced notable relief postoperatively, she continued to have edema and discomfort. Once again, she was referred for physical therapy and subsequently returned to normal functioning.

Case 2

A 230-pound, muscular 56-year-old man with a history of polio presented complaining of periods when his feet were cold and purple. The patient related a history of an arthroplasty 6 years previously due to a 20-year history of pain and stiffness in the second metatarsophalangeal joint of the left foot. Social history revealed that the patient performed physical labor for 25 years as a meat cutter, was a nonsmoker, and consumed no alcohol.

The patient had been hospitalized when he was 5 years old, in 1946, for “an entire summer” with bulbar polio. He recalled experiencing paralysis of the lower extremities with extreme weakness in his neck as well as being on a waiting list for an iron lung because of breathing difficulties. He claimed to have regained full strength approximately 2 years after his initial infection with the polio virus. He had led a seemingly normal life, free of any recognizable neuromuscular problems, until his forties.

The patient related an abnormal decline in his physical ability to perform work-related tasks. In 1993, he presented to a general practitioner complaining of abnormal fatigue, as well as some joint and muscle pain and weakness following routine physical exercise as well as normal everyday activities. The patient also complained of a 5-year history of muscle fatigue, spasms, and “electrical-like” sensations in the musculature of his back. He was referred to a neurologist, who performed electromyographic
studies of the dorsal paraspinal muscles. The results of these were inconclusive.

The patient also complained of intolerance to cold (especially in his feet) as well as of hot spots on his skin. He related periodic presentation of spots about the size of a silver dollar with a burning sensation. He said that he would quickly look to see what was burning him and find nothing there. He claimed that rubbing the hot spots made them quickly dissipate. He also related an approximately 4- to 5-year history of a "numb itch, like when the anesthesia wears off at the dentist," on his scalp and forehead above the left eye.

It is also pertinent that while in his thirties, the patient was running 15 miles a day, but was forced to discontinue this because of asthmatic symptoms—despite a negative diagnosis for asthma. In his forties, the patient experienced a severe continual headache. At that time, a computed tomographic scan was done to rule out a brain tumor. Later the same year, the patient presented with sleep apnea (he reportedly slept only a few hours a night) and morning headaches. He was diagnosed as having depression and treated with amitriptyline hydrochloride. The periodic headaches and sleeping problems continued.

The patient was advised to wear cotton socks over acrylic ones to wick the moisture away from his feet and keep them dry and warm. He was also advised that thermal socks were another alternative and that he might need to change socks several times throughout the day. Furthermore, he was educated about the effects of postpolio syndrome and the importance of lifestyle modifications in order to avoid fatigue.

**Discussion**

Postpolio syndrome is difficult to diagnose. In fact, there are many varied opinions concerning this syndrome. Some physicians believe that the diagnostic criteria should be fairly strict. Because the symptoms of postpolio syndrome may present idiopathically, this is difficult. Even though a patient may have years without symptoms, a positive history of polio (especially of an acute, paralytic nature) should lead to a high index of suspicion for postpolio syndrome. The road to diagnosis of postpolio syndrome is a long one, but every effort should be made to speed the process. Once postpolio syndrome is diagnosed, the goal should be to bring about the required lifestyle modifications and keep the patient functioning comfortably. As stated earlier, specific systemic complications should be addressed individually. Care should also be taken to maximize the functioning capacity the patient has left and avoid potential problems from overuse. Orthoses are an effective tool for optimizing mechanical efficacy and thus reducing the risk of overuse. The longer a postpolio syndrome patient remains undiagnosed, the more the patient's everyday activities may damage motor neurons that are already being taxed.


**References**

1. **BRUNO RL:** *Post-Polio: You Can Live with It*, Easter Seal Society of Washington/Polio Outreach Advisory Council, Seattle, WA.
4. **MAYNARD FM:** The *Post-Polio Syndrome and Rehabilitation*, Easter Seal Society of Washington/Polio Outreach Advisory Council, Seattle, WA.