Post-Polio Syndrome

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Decades after recovering much of their muscular strength, survivors of paralytic polio are reporting unexpected fatigue, pain and weakness. The cause appears to be degeneration of motor neurons.

In the first half of the 20th century, the scourge of paralytic poliomyelitis seemed unstoppable. A major polio epidemic hit the New York area in 1916, and in the following decades the epidemics grew in size and became more deadly. The epidemic of 1952, for instance, affected more than 50,000 Americans and had a mortality rate of about 12 percent. It is difficult to realize today the extent of the fear and panic that gripped the public. Polio haunted everyone: families stayed at home; swimming pools were closed; public events were canceled.

Children in particular were at risk. With the introduction of Jonas E. Salk's injected killed-virus polio vaccine in 1955 and Albert B. Sabin's oral live-virus vaccine six years later, the epidemics were brought to an end. By the mid-1960s the number of new polio cases dropped to an average of 20 a year. Polio had been vanquished. Or so it seemed.

For the average American, polio no longer meant a disease but a vaccine. Medically, polio came to be regarded as a static, nonprogressive neurological disorder. It was believed that after rehabilitation and reeducation most survivors could reach a plateau of neurological and functional stability that would last more or less indefinitely--and that is where our understanding of polio as a chronic disease remained until fairly recently.

Then, in the late 1970s, reports began to surface that people who had recovered from paralytic polio decades earlier were developing unexpected health problems such as excessive fatigue, pain in muscles and joints and, most alarming of all, new muscle weakness. Because there was little in modern medical literature about delayed neurological changes in polio survivors, the initial response by many physicians was that the problems were not real. For a time they were dealing with a cluster of symptoms that had no name--and without a name there was, in essence, no disease. Having a name--even if imprecise and misleading as to causation--at least confers an element of credibility.

By sheer weight of numbers, persons experiencing the late effects of polio finally started attracting the attention of the medical community, and in the early 1980s the term post-polio syndrome was coined. Today the syndrome is defined as a neurological disorder that produces a cluster of symptoms in individuals who had recovered from paralytic polio many years earlier. These symptoms typically appear from 30 to 40 years after the acute illness. The major problems are progressive muscle weakness, debilitating fatigue, loss of function and pain, especially in muscles and joints. Less common are muscle atrophy, breathing problems, swallowing difficulties and cold intolerance. Of all these health problems, however, the critical symptom of post-polio syndrome is new progressive weakness.

Patients most at risk for post-polio syndrome are those who had suffered a severe attack of paralytic polio, although some patients who seemingly had a mild attack have also developed the syndrome. The onset of these new problems often is insidious, but in many cases they appear to be precipitated by specific events such as a minor accident, a fall, a period of bed rest or surgery. Patients characteristically say that a similar event several years earlier would not have caused such a large decline in health and function.

My own experience seems to be typical of both recovery from paralytic polio and the new development of post-polio syndrome. I contracted polio during the epidemic of 1954 while traveling in Europe after my freshman year in college. I was 18 years old. My six-month journey of recovery took me from iron lung to wheelchair to foot brace and then to no assistive device at all. At times, improvement in strength seemed to happen overnight. Although my right arm remained paralyzed, the rest of my body regained most of the strength and endurance I had before my illness. As a result, I thought of myself as cured. I returned to college, learned to write with my left hand and even played intramural squash. On the morning of the third anniversary of the onset of my polio, I reached the summit of Mount Fuji in Japan after a climb of over 12,000 feet. As I watched the sun rise, I thought, "Polio is behind me. I have finally conquered it."

With the conquest of Mount Fuji fresh in my mind, I began to look for other mountains to climb. After college, I entered medical school. Internship and residency initiated yet another cycle of physically demanding years. In short, I got on with my life while polio receded ever further in my memory. Several years ago I began developing new weakness in my legs. As the weakness progressed over a period of months, I went from being a full-time walker who jogged up six flights of stairs for exercise to having to use a motorized scooter full-time at work.

Historical Background

Post-polio syndrome, it turns out, is not a new disorder after all. It was described in the French medical literature in 1875, and then, as often happens in medicine, it was forgotten. Over the next 100 years, approximately 35 reports on post-polio weakness were published in the medical literature. By 1984 a growing awareness of the delayed effects of polio prompted me and other researchers to organize an international conference at the Warm Springs Institute for Rehabilitation--the great polio treatment mecca in southern Georgia established by Franklin Delano Roosevelt, who was paralyzed by polio in 1921 at the age of 39. Roosevelt felt that swimming in the warm waters of a natural spring at a health resort strengthened his muscles, and in 1926 he purchased a hotel there and turned it into a nonprofit foundation.

A second international meeting on post-polio syndrome was held at the Warm Springs Institute in 1986, and in the following years there was a dramatic increase in basic and clinical research into the long-term effects of polio. In 1994 the New York Academy of Sciences and the National Institutes of Health cosponsored another international meeting that culminated in the publication of a special issue of the *Annals of the New York Academy of Sciences:* "The Post-Polio Syndrome: Advances in the Pathogenesis and Treatment." That conference signaled the acceptance of post-polio syndrome as a legitimate clinical entity.

Surprisingly, accurate figures about the number of Americans who have had paralytic polio are not available and probably never will be. There is no national registry of persons who were diagnosed with the disease, and there is no way, after all these years, to compile accurate figures from state and local health departments. The best estimate is based on information from the federal government's National Center for Health Statistics, which collects data on health and disability issues every year from a random sample of U.S. households. Based on data from the 1987 survey, the National Center for Health Statistics calculated there were more than 640,000 survivors of paralytic polio, which would mean there are more survivors of paralytic polio than there are persons with multiple sclerosis, amyotrophic lateral sclerosis or even spinalcord injury.

Since 1987 an unknown number of polio survivors have died, but immigrants, refugees and illegal aliens have added an unknown number to the U.S. population of polio survivors. No one knows how many survivors of paralytic polio have post-polio syndrome. Some studies indicate the figure could be as high as 40 percent. If this estimate is accurate, then the total number of persons in this country currently suffering from post-polio syndrome could reach 250,000.

Knowledge of how the poliovirus infects the body can be helpful in understanding the possible causes of post-polio syndrome. It is a small RNA virus that can enter the body when contaminated water or food are ingested, and even when contaminated hands touch the mouth. The vast majority of persons who become infected either have no symptoms or experience a self-limited illness characterized by fever and gastrointestinal upset for several days. The poliovirus, which replicates in the lymphoid tissue of the throat and small intestine, either passes harmlessly from the gut or travels in the blood to all parts of the body. In a small minority of infected persons--usually 1 to 2 percent--the virus invades the central nervous system and produces an unpredictable amount of paralysis.

Mechanism of Infection

A distinctive characteristic of acute polio infection is the predilection of the poliovirus for the nerve cells that control muscles. These nerve cells, or motor neurons, consist of a cell body located in the anterior horn of the spinal cord and a long tentacle, or axon, that extends to the muscles. Near the end of each axon, tiny sprouts branch out to individual muscle cells. At the nerve-muscle interface, or synapse, the sprouts from the axon release acetylcholine, a neurotransmitter that causes the muscle fibers to contract. A motor neuron and the group of muscle cells that it activates are called a motor unit.

With uncanny precision, the poliovirus invades the motor neurons, leaving intact adjacent nerve cells that control the functions of sensation, bowel, bladder and sex. How this exquisitely targeted behavior occurs was a mystery until recently, when researchers identified poliovirus receptors at the nerve-muscle interface. These receptors apparently allow the poliovirus to enter an axon and then to migrate to the nerve cell body in the anterior horn of the spinal cord. The poliovirus typically infects more than 95 percent of the motor neurons in the spinal cord and many other cells in the brain. The infected cells either overcome the virus or die.

The extent of paralysis is unpredictable. Motor neurons that survive develop new terminal axon sprouts in response to an unknown stimulus. These new sprouts reinnervate, or reconnect, with the muscle fibers left orphaned by the death of their original motor neurons. In a sense, the growth of additional axon sprouts is the body's effort to keep as many orphaned muscle cells as possible alive and working. A single motor neuron that initially stimulated 1,000 muscle cells might eventually innervate 5,000 to 10,000 cells, creating a giant motor unit. These vastly enlarged motor units make it possible for fewer motor neurons to do the work of many.

Another adaptation that leads to increased strength is the enlargement of muscle cells when they are regularly exercised. These two compensatory adaptations--increase in muscle size and axon sprouting--are so effective that up to 50 percent of the original number of motor neurons can be lost without the muscle losing clinically normal strength. These adaptations are neither static nor permanent, however. To the contrary, after recovery from acute polio there is an ongoing process of remodeling of the motor units that consists of both denervation (losing old sprouts) and reinnervation (gaining new ones). It is this process of remodeling or constant repair that allows the motor units to achieve a steady state of muscle strength. When this steady state is disrupted, new muscle weakness occurs.

There is a growing consensus among researchers that post-polio syndrome involves a slow degeneration of the terminal axon sprouts that innervate the muscle cells. David O. Wiechers and Susan L. Hubbell, then affiliated with Ohio State University, proposed this explanation in the early 1980s after diagnostic tests indicated that the functioning of motor neurons in polio survivors progressively worsens as the number of years from their recovery increases. More recently, Daria A. Trojan and Neil R. Cashman of the Montreal Neurological Institute and Hospital, after examining the results of muscle biopsy and electromyographic (EMG) studies in their laboratory and by researchers elsewhere, postulated that there are two types of disintegration of the motor neurons: a progressive lesion and a fluctuating one.

The progressive lesion, in their view, occurs when the normal regeneration of the sprouts from the axon to the muscles is interrupted and malfunctioning sprouts are not replaced. This interruption of the repair process produces irreversible, progressive muscle weakness. The fluctuating lesion, on the other hand, is thought to be caused by defective synthesis or release of the neurotransmitter acetylcholine. Cashman, Trojan and others have demonstrated that muscle weakness and fatigue can be reversed in some patients with post-polio syndrome by the drug pyridostigmine, which enhances the effectiveness of acetylcholine

in triggering muscle contractions. Other researchers are testing another class of agents known as nerve growth factors, which stimulate both nerve and muscle cell growth.

Possible Causes

Degeneration of the axon sprouts can explain the new muscle weakness and fatigue, but what causes the degeneration in the first place remains a mystery. The most plausible hypothesis proposes that the muscle weakness is simply the result of overuse of individual motor neurons over time. This explanation assumes that after recovery from polio the surviving giant motor neurons must labor more than normal neurons just to maintain daily activities. After many years of continued overuse, these enlarged motor neurons eventually suffer from a kind of metabolic exhaustion that leads to an inability to regenerate new axon sprouts to replace degenerating ones. There is no direct way to measure metabolic fatigue in motor neurons, but evidence to support this hypothesis can be inferred from abnormalities on muscle biopsies, electrodiagnostic tests and clinical response to exercise.

In addition, some researchers report that some motor neurons in the anterior horn of the spinal cord of polio survivors appear to be smaller than normal. It is believed that these smaller motor nerve cells were somehow damaged at the time of the acute polio infection and that they are vulnerable to premature failure. Normally, significant attrition of motor neurons does not occur until a person reaches the age of 60 years or more. Because polio survivors have a greatly reduced number of motor neurons, the loss of even a few functioning motor units could result in a disproportionate loss of muscle function.

Attrition of motor neurons because of aging alone, however, may not be a significant factor in post-polio syndrome. Several studies have failed to find a positive relation between the onset of new weakness and chronological age. To the contrary, these studies suggest it is the length of the interval between onset of polio and the appearance of new symptoms that is a determining variable.

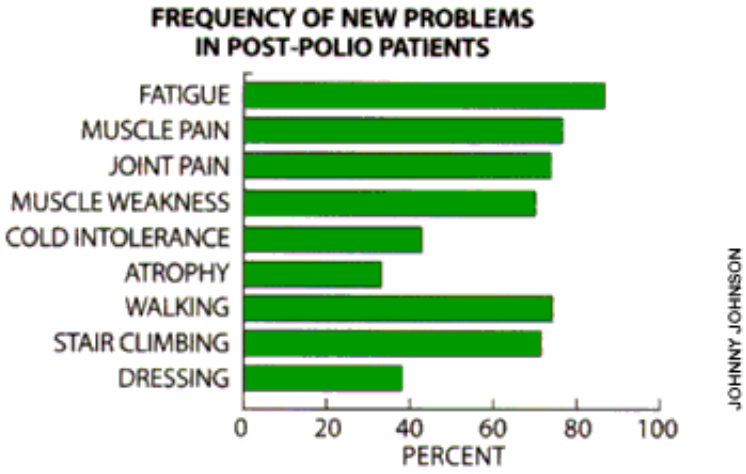
Persistence of the poliovirus in the body--or, more precisely, viral particles that have lain dormant for many years and then are reactivated by some unknown mechanism--may also play a role. Researchers at a number of centers have reported poliovirus-like RNA fragments in the spinal fluid and spinalcord tissue of some patients with postpolio syndrome but not in polio survivors who do not have the syndrome. These small RNA fragments, however, do not appear to have any infectious potential. Whether these findings are simply incidental or indicate a possible viral role is not known.

Other causes of post-polio syndrome, such as an immune-mediated response, hormone deficiencies and environmental toxins, have also been proposed. Although some of these hypotheses, and others, seem plausible--and none has been completely excluded--at the present time there is not enough evidence to justify strong support for them.

No definitive tests exist for post-polio syndrome. Diagnostic tests of the blood, muscle biopsies and EMG recordings of muscle activity often show essentially the same abnormalities in polio survivors with post-polio syndrome as in those without it. Therefore, diagnosis relies primarily on systematically ruling out other causes of a patient's symptoms.

Diagnosis and Treatment

A set of criteria for diagnosing post-polio syndrome has been developed by the Post-Polio Task Force, a group of researchers, clinicians and polio survivors. The onset of new muscular weakness after many years of stable functioning is perhaps the most characteristic symptom. Yet many of the symptoms of post-polio syndrome are so general that ruling out all possible causes is often impractical. For instance, new weakness may result simply from the lack of use of the muscles. Muscle weakness from disuse can mimic post-polio syndrome and complicate it. Regardless of the underlying cause, once the weakness begins it may initiate a cascade of other complaints that makes the original symptom impossible to identify.



MOST COMMON new health problems reported by post-polio patients in several clinical studies are fatigue, muscle pain, joint pain and weakness. Functional problems include walking and stair climbing.

As with many other chronic conditions, the essence of good medical care is to relieve symptoms, improve muscular function and enhance the patient's sense of well-being. This management strategy is frequently referred to as bracing and pacing. Effective intervention can be as simple as prescribing a cane or a wheelchair or as complicated as putting someone in an iron lung or providing some other form of assisted ventilation. For both physical and psychological reasons, patients are encouraged to remain as active as possible. The new lifestyle, however, should incorporate regular rest breaks. An exercise program is desirable for virtually all patients. For some, this may be nothing more strenuous than gentle stretching or various types of yoga. For others, it may be considerably more vigorous and even include aerobic workouts.

With certain types of exercise in carefully monitored settings, some patients have been able to regain and maintain muscle strength. Although there are no magic bullets, medications can occasionally be helpful: for example, low doses of a tricyclic antidepressant may relieve muscle pain in some patients, and pyridostigmine may reduce fatigue and improve muscle strength.

As a general rule, the progression of symptoms is fairly slow and the overall prognosis is good, unless there are severe breathing or swallowing difficulties. Yet when reserves of strength and stamina are low, a minor change in the stability of the motor neurons can result in a disproportionately large loss of muscular function, which often is psychologically devastating.

Personal Legacies of Polio

Most polio survivors tell a story of struggle and triumph: the sudden, random onset of paralysis, the gradual restoration of strength seemingly as a result of individual willpower and, finally, for many, resumption of an active, productive life, which leads them to believe they have put polio behind them.

The story, for most, is made possible by denying their disability and the reality of what was lost and the life that might have been. Perhaps the most famous example of disability denial is provided by our 32nd president, Franklin D. Roosevelt. Although he regained normal use of his arms fairly quickly after his polio illness, he spent many years in a heroic, but largely unsuccessful, effort to relearn to walk. Even after he became president, Roosevelt went to extraordinary lengths to disguise the extent of his disability. In the waning months of his life, he experienced increasing muscular weakness that appears to be compatible with post-polio syndrome.

Virtually every polio survivor I have met has displayed self-deception or denial. Until recently, most of us tended to avoid other polio survivors and polio help groups. We knew we weren't physically normal, but if we thought about it at all, we considered ourselves as inconvenienced, not disabled. By retraining the muscles that remained, we felt we could do just about anything, even become president, like Roosevelt.

Later in life, when polio survivors begin to experience the new weakness of post-polio syndrome, the denial usually is still intact, which makes understanding and accepting the new changes all the more difficult. As they begin to accept the fact that they are disabled, they may be overcome by feelings of anger, bitterness and despair. Fortunately, a post-polio support movement has sprung up, and there are now more than 300 support groups throughout the country. In my own case, it took me several years after developing post-polio syndrome before I joined a support group and began talking with other polio survivors about my new weakness and pain. Only then did I start to grieve for the body I lost 35 years earlier.

Many post-polio survivors exhibit an extraordinary commitment to exercise, a legacy from their recovery from polio. When a physical therapist prescribed 10 repetitions twice a day to strengthen a certain muscle, patients typically would do 20 or more repetitions three times a day. For many, exercise became a daily obsession, for others, almost a religious devotion. Thus, survivors of polio developed a special relation to their bodies unknown to able-bodied persons. They experienced a new mastery over their muscles and movements, an element of control that had not existed before polio. It was a visceral lesson that carried over into other aspects of their lives and probably accounts for why so many polio survivors have excelled at school and at work.

Individuals who have recovered from paralytic polio have, on average, more years of formal education than the general population, and they take on marriage and family responsibilities at approximately the same rate as persons who are not disabled. Also, the rate of employment of polio survivors is reported to be about four times the rate of other disabled persons.

Over the past few decades much of the leadership for the disability movement has come from polio survivors. Their efforts have led to the founding of the Independent Living movement and to the passage of legislation such as the Architectural Barriers Act and the Americans with Disabilities Act.

What is not widely known is that many of these leaders were among the most disabled: walking with braces and crutches, riding motorized wheelchairs and attached to portable ventilators. In his novel about the plague, Albert Camus wrote, "It helps men to rise above themselves." He could have been describing polio.

Social Legacies

Polio has had a far-reaching effect on medicine. The successful development of a safe vaccine after years of effort was a triumph of enormous propertions. It involved the eager participation of millions of ordinary Americans, initially through door-to-door fund-raising and later in volunteering their children to participate in the 1954 field test of the Salk vaccine. The massive research effort to develop a polio vaccine led to many discoveries that have since revolutionized the fields of virology and immunology.

The polio epidemics accelerated the development of rehabilitation medicine. In contrast to traditional medicine with its focus on the curing and repairing of diseased organs, rehabilitation medicine emphasizes the rebuilding of body functions and the teaching of the skills necessary for independent living. The principles developed to treat polio patients decades ago are essentially the ones used today to rehabilitate persons with head and spinal injuries, strokes and degenerative disorders.

The intensive care unit that is now an integral part of every modern hospital is another legacy from the era of polio epidemics. When large numbers of polio patients requiring iron-lung ventilators started to overwhelm the staff in rehabilitation centers, the iron lungs were clustered together so they could be readily monitored by only a handful of nurses. In addition, regional respiratory centers were created to treat the most severely affected polio patients. Some of these centers have survived and continue to provide care for persons with spinal-cord injuries.

The money that originally funded these respiratory centers was raised by the National Foundation for Infantile Paralysis, which later became known as the March of Dimes. It was Eddie Canter, the famous singer and entertainer, who suggested that Americans could be asked to mail dimes to support the fight against polio. For a while, the fight against polio brought out many of the best qualities of American society: neighbors carried food to families quarantined at home, teen clubs raised money to help hospitalized classmates, and women's groups adopted local polio wards for the year.

Then the vaccines were developed, and not only did polio disappear in America but the war against polio seemed to be forgotten--along with the survivors. Once held up as examples of heroic human fortitude, thousands of polio survivors who continued to need medical and financial help were largely ignored by the public. As veterans of other wars would continue to discover, the public does not like to be reminded of the wounded and the dead after the war is over. Also, as with other conflicts, the polio war left more wounded and uncounted survivors than are generally recognized.

Even the miracle vaccines have their problems. Most scientists agree that the Sabin oral vaccine is superior to Salk's injected one; however; the oral polio vaccine is not free of risk. In fact, it causes the very disease it is designed to prevent in 10 to 15 people every year because of either a mutation in the virus or an immune deficiency in the recipient that allows the weakened virus to take hold and produce paralysis. The federal government has changed its vaccination policy and is now recommending two initial immunizations with the injected killed-virus vaccine followed some time later by two doses of oral live-virus vaccine. In theory, this combination provides the advantages of both vaccines and, we hope, will end forever this nation's rendezvous with polio.

The Author

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Further Reading

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