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Defining Post-Polio Problems

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The term late effects of polio is the umbrella over three subdivisions.

The cause of post-polio problems is neuromusculoskeletal failure from chronic overuse of a polio altered system. How has that system been altered? The pathology of polio involves destruction of the anterior horn cell, and the only functional residual is weakness. New polio problems are the effects of the functioning of muscle which had less than normal capability.

So, who's under the umbrella? First are the asymptomatic post-polio individuals who have a known history of polio but who are not having troubles now. Their lifestyle and their physical ability are balanced. These persons can, for example, go on running and continue what they are doing as long as they have no symptoms. If symptoms do occur, it is time to become more cautious about their activities.

The second group has symptoms which indicate the lifestyle demands now exceed their muscles' ability to meet those demands. The symptoms of pain, fatigue, and new weakness are signs of overuse with a penalty. We have a list of about 1,000 polio survivors and of that group we found 107 who thought they had full recovery. Eighty nine percent of them lived very active lives while 24% of them had been in athletics. They came to the clinic because they were having symptoms. Ninety three percent showed weakness with at least one muscle less than grade 5. (See chart below.)

When a muscle creates a force to control or create motion, it also creates a force internally and can cause

some damage. It has been demonstrated that as muscles contract they need time to repair and refuel. If the muscles are functioning almost continually, they do not have time to repair. Muscles and their ability to function are the main problem.

Another problem is the neuromuscular junction. This is evidenced by information from Gunnar Grimby, MD, PhD, Sahlgren Hospital, Göteborg, Sweden. He studied the anterior tibialis and demonstrated that a 3+ muscle, which is about 25% of normal on the Beasely scale, has marked hypertrophy (enlargement) compared to a 4+ muscle which would be about a 45% muscle. Both of them have degrees of weakness, but the 3+ is much more weak than the 4+.

Grimby also found that the 3+ tibialis anterior was not only markedly hypertrophied, but its activational rate was twice normal. One gains new or additional strength either by activating more muscle fibers or making the current muscle fibers work twice as hard. In 3+ muscles there are not very many muscle fibers, so they are worked twice as hard to meet the functional demand.

The individuals most difficult to get in balance are the ones with 3+ and 2 muscles all over. Their muscles are not strong enough to function without overtime use.

The third group, which we often overlook, is the polio survivor with joint degeneration problems. It is just plain wear and tear, either from substitutive posture used to replace inadequate muscles, or by the impact of loading a joint instead of letting it yield on the muscles. For instance, a hyper-extended knee causes bones to hit together with every step, and the joint becomes degenerative and arthritic. Joint degeneration problems do not result in muscle pain. There is pain and tenderness in the specific joint, resulting in deformity and x-ray changes.

Again, feeling that the primary cause is overuse, the principal management is lifestyle modification. No matter what else we do, we find that about 95% of people have to make lifestyle changes in order to make exercise or other devices work.

Out of 250 patients we have studied quantitatively on a scale of "better, same, worse." The majority of them were "better," meaning that they lost their symptoms of

Beasely Scale

Muscle Grade		% of True Normal
5	Normal	50%
4	Good	40%
3	Fair	20%
2	Poor	10%
1	Trace	1%

pain and fatigue by modifying their lifestyle. A small percentage did not make a change because they reject the whole idea of lifestyle modification. Some became worse and are the individuals with all muscles of grades 2 and 3. We have trouble finding a lifestyle for them which does not cause any strain.

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Today, there is no consensus, no position paper, no diagnostic test, no agreed-upon criteria about this syndrome, so there will be differences of opinion and some overlap.

From a neurologist's point of view, post-polio syndrome has to begin in the spinal cord and brain stem with the original infection and destruction of motor neurons causing paralytic polio. A common question arises: "I had non-paralytic polio. Am I susceptible to the post-polio syndrome?" A neurologist would have to say "no." There is no evidence that people who had no infection of motor neurons can develop classical post-polio syndrome. In fact, the worse the paralytic poliomyelitis, the more likely new symptoms are to develop.

Another common question is: "Am I out of the woods with post-polio syndrome after 40-years?" Burk Jubelt, MD, (SUNY, Syracuse, NY), and I reviewed every recorded case of post-polio syndrome that had been reported up to 1987 and found the mean onset was 36 years. Some people develop new symptoms 10 years after polio, and others develop symptoms 60 years after polio. Unfortunately, there is no out-of-the-woods threshold age.

There is a running controversy in the medical literature of "lumpers" and "splitters." Some authorities like to put diseases together under certain rubrics, and others like to break off certain diseases. My opinion is that it is too early to split sub-syndromes off the post-polio syndrome. My conservatism, in part, is based on work with people who had post-polio muscular atrophy. People with atrophy were just as likely to have pain and fatigue as people without new atrophy. There is not enough data to make a distinction and say this is merely musculoskeletal; that is neurogenic. They act differently and, they are treated differently.

The original meaning of the word "syndrome" comes from the Greek — "running together." These symptoms present in polio persons time after time, and the symptoms really do run together. There is something going on and, in my opinion, it is too early to dogmatize sub-groups.

The safest term, to date, is post-polio syndrome, and the safest definition of the post-polio syndrome is a new clinical syndrome of pain, weakness, and fatigue in individuals who recovered from acute paralytic poliomyelitis.

**Lauro S. Halstead, MD, Physical Medicine and
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Washington, D.C.**

From the point of view of a clinician who sees polio individuals in a clinic, "What diagnosis do I use?" often makes a great deal of difference, because of the implications for insurance and disability payments. We do not have enough information, insight, or wisdom to separate specific definitions except for clinical use.

The three major diagnostic terms — the late effects of polio, post-polio syndrome, and post-polio muscular atrophy — can be described by imagining three concentric circles. A large outer circle labeled the late effects of polio represents a grab bag. Anyone who has serious involvement with a neuromuscular disease, and it does not have to be polio, will experience a variety of problems, if they live long enough.

The late effects of polio refers to a group of symptoms and signs which people who had polio many years ago now experience and include weakness, fatigue, muscle pain, joint pain, decreased endurance, and new atrophy. They also include increased weight gain, osteoporosis, increased risk of fractures, scoliosis, increase in pulmonary problems, sleep difficulties, and psychological problems.

People can experience a whole list of problems that are in this big circle as a result of having had polio, but they do not have post-polio syndrome.

Inside the big circle is a smaller circle labeled post-polio syndrome. More narrowly defined, a diagnosis of post-polio syndrome requires the presence of new neurogenic weakness. This new weakness is caused by some dysfunction of the motor neuron that was originally affected. Post-polio syndrome is a neurologic disease which occurs as a result of having had an invasion of the polio virus to part of the nervous system many years earlier.

**There are five elements to make the
diagnosis of post-polio syndrome:**

1. a history of paralytic polio;
2. a fair to good recovery (something had to recover to have loss later on);
3. a period of neurologic stability;
4. onset of new weakness; and
5. exclusion of other causes for the new weakness.

Currently, making a diagnosis is difficult because there is no diagnostic test; unfortunately at this stage, diagnosis is still primarily by history and physical examination.

The smallest and most inner circle is post-polio muscular atrophy and is reasonably straightforward. It is post-polio syndrome in someone with new muscle atrophy.

Another better term that might be used for these last two diagnoses is post-polio motor neuron disease which focuses on the motor neuron as the primary source of pathology.

Frederick Maynard, MD, Physical Medicine and Rehabilitation, MetroHealth Center, Cleveland, Ohio

In the broadest terms, there are many new health problems that persons with a previous history of polio may experience. These health problems can include those that are clearly unrelated to the history of polio, such as glaucoma or gall stones.

For other health problems, such as coronary artery disease, it may be unclear whether there is a relationship to previous paralytic because a sedentary lifestyle in polio survivors may cause an increased risk of developing coronary artery disease.

Systemic health problems, such as heart disease or diabetes, can impact and produce symptoms that overlap and mimic those of post-polio syndrome (fatigue, weakness, and pain). This issue has led to confusion, misunderstanding, and lack of consensus about what is meant by post-polio syndrome.

The term "late effects of polio" implies that new health problems are related to the original polio impairments, such as muscle weakness and related joint deformities. The term "post-polio syndrome" is a looser term which does not specify etiology, or cause of symptoms. Post-polio syndrome commonly develops in people with polio residuals and mild symptoms are almost inevitable in people who are growing older with polio.

As a "syndrome," post-polio is a common symptom cluster — classically, pain, fatigue, and new weakness — that is seen over and over again in post-polio people.

The bottom line for having post-polio syndrome is new loss of function. Individuals with post-polio syndrome have new disability.

Post-polio muscular atrophy (PPMA) refers specifically to people who definitely had old polio weakness and are now having new weakness and atrophy from no other identifiable cause. PPMA is new or additional anterior horn cell disease that appears to be unexplainable.

A slightly different concept has recently been proposed by the Institute of Medicine (see below) to describe new disabilities in people with long-standing disability.

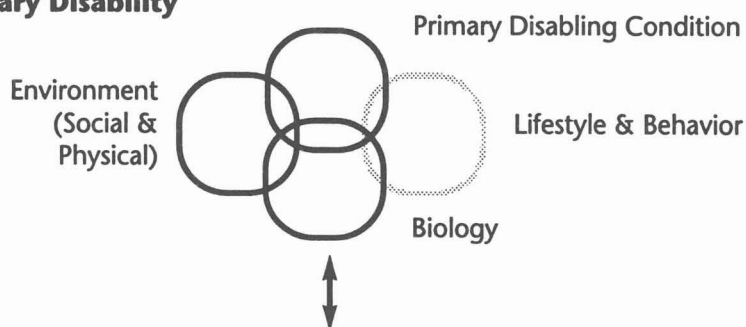
This has also been referred to as the development of secondary disability in a person who has always had a primary disability. For example, people with post-polio residuals, after full recovery and rehabilitation, were left with a primary disabling condition. Other health problems may then cause the residuals (weakness, deformity) to become worse and result in additional new functional limitations.

Another new concept is the notion of "life course" of people with a history of polio. As people with paralytic polio histories become older, they can be expected to experience a level of functional decline when they are 70 or 80, similar to what non-polio people experience. However, their expected life course of slowly progressive weakness can be drastically accelerated by other life events, such as injury or the onset of other medical conditions, for example diabetes, heart disease, and arthritis. □

Institute of Medicine's Model of Secondary Disability

Figure 1.

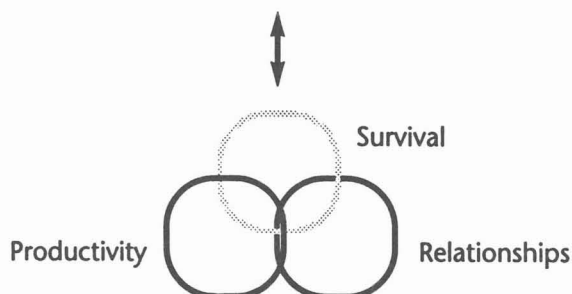
◆ Risk Factors



◆ Process of Secondary Disablement



◆ Quality of Life

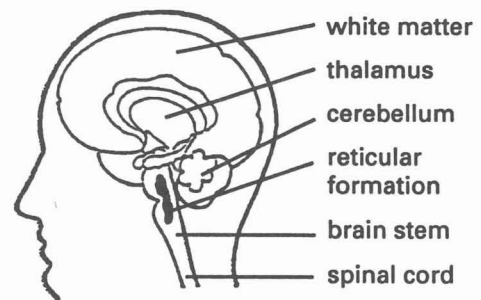
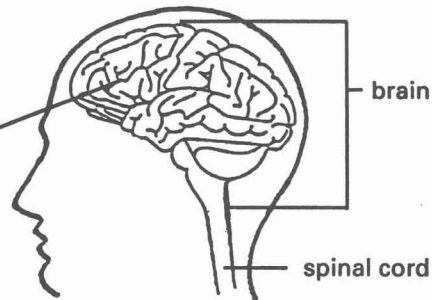


NOTE: Descriptive sentences in Figures B and C were inadvertently shortened. This is the completed version.

DIAGRAMS AND DEFINITIONS FOR POST-POLIO SYNDROME 101

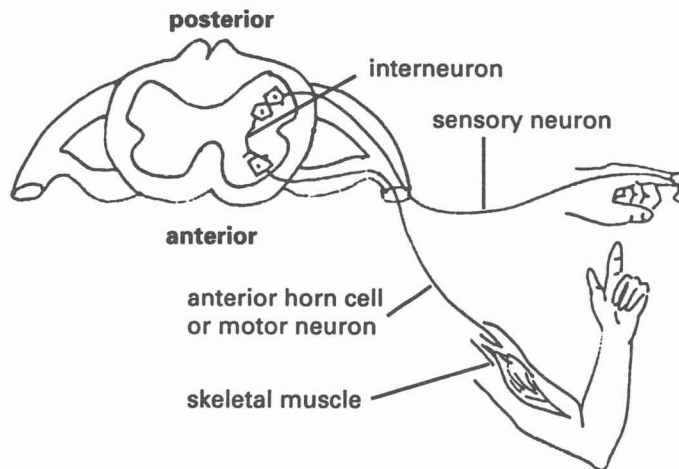
The central nervous system is composed of the brain and the spinal cord.

cerebral cortex (gray matter)



The polio virus infected the anterior horn cells, thus affecting the skeletal muscles causing partial or complete paralysis.

Cross section of spinal cord



A motor unit is composed of a nerve cell and all the muscle fibers it innervates. The neuromuscular junction is the junction between the nerve cell and the muscle fiber.

Figure A

During infection

Two of the five nerve cells have been infected by the polio virus. The middle one has temporarily stopped functioning.

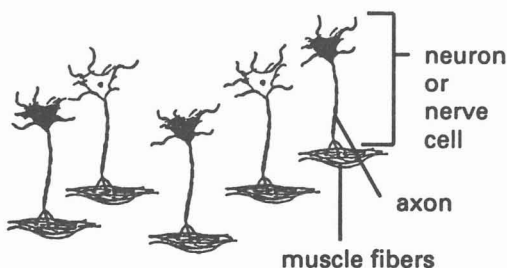


Figure B

A few weeks later

The middle nerve cell has recovered. Two nerve cells have died leaving the muscle fibers denervated or orphaned.

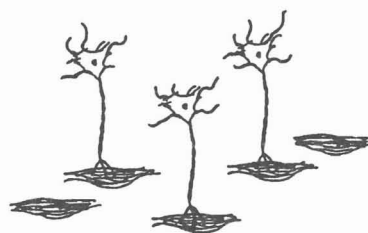


Figure C

Months, even years later

The surviving nerve cells "sprouted" to innervate the orphaned muscle fibers.

