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LATE EFFECTS OF POLIOMYELITIS

PETER E. PARKER, BSc, BSc(PT), MD, FRCPC
Physiatrist, Neurorehabilitation Services,
West Park Hospital, Toronto, ON

A large number of Canadians who developed polio as children or young adults are now reaching their fifth and sixth decade. With this aging, these individuals are facing unique problems as a result of having polio many years ago, problems that have come to be known as The *Late* Effects of Poliomyelitis. The last epidemic of poliomyelitis in Canada was in 1955. As a result of the Salk and Sabin vaccines, the number of cases decreased dramatically after this year.

A ribonucleic acid virus of which there are three strains, causes poliomyelitis. The virus attacks and destroys the anterior horn cells (alpha motor neurons) of the spinal cord and brain stem. The alpha motor neuron and the many muscle fibres innervated by it is called a *'motor unit'*. Loss of these motor units results in the paralysis associated with poliomyelitis. Surviving motor units, recognizing that muscle fibres are left without innervation, send out terminal axon branches, referred to as 'collateral sprouting', thereby, re-innervating the muscle to some degree. As a result of this process, an alpha motor neuron may innervate up to seven times the normal number of muscle fibres. These large units are termed *'giant motor units,'* and account for the recovery of muscle function following the paralysis caused by the poliomyelitis virus.

Polio survivors were taught to push themselves, and regardless of the degree of neurological recovery, they have led active and productive lives. Now, due to the late effects of poliomyelitis, they must relive their experience with poliomyelitis. What was in the past can no longer remain there. The childhood memories of isolation, separation, iron lungs, bracing and pain surface as these poliomyelitis survivors age. As a result, they must deal with new problems of pain, fatigue and diminished function.

The late effects of poliomyelitis can be categorized into three distinct symptom complexes. The first, **Progressive Muscular Atrophy**, is the progressive wasting and weakening of muscle group[s] leading to progressive loss of neuromuscular function. This is relatively rare. On the other hand, the other two complexes, **Biomechanical Sequelae** of poliomyelitis and **Post-Polio Syndrome**, are common and account for a significant degree of impairment and disability in this aging population.

BIOMECHANICAL SEQUELAE

The most common biomechanical sequelae is premature osteoarthritis of both upper and lower extremity joints. Lack of normal muscle strength around lower extremity joints results in abnormal 'wear and tear' on these joints. As a result, premature degeneration develops, particularly in the knee joints. This is characterized by joint pain with weight bearing activities, joint stiffness following periods of immobility, joint effusion and joint instability. This instability develops as a result of loss of integrity of the joint capsule and ligaments in addition to muscle weakness. As a consequence, falls become increasingly frequent, leading to further injury.

Premature degeneration of the hip and ankle joints is also a common complication due to residual weakness in the lower extremities as a result of poliomyelitis. Often, these joint problems develop in the so-called 'good leg', the extremity not affected by polio. This is a result of over-work of the strong leg in order to compensate for the weaker extremity. Early degenerative osteoarthritis often develops in the upper extremities, particularly in the shoulder joints, due to prolonged use of walking-aids such as crutches or a cane. Joint pain and the concomitant loss of function has a significant impact on the quality of life.

Correspondence to: Wendy Malisani, Coordinator, Post-Polio Clinic, West Park Hospital, 82 Buttonwood Ave., Toronto, ON M6M 2J5. Tel: (416) 243-3600, ext. 2157; Fax: (416) 243-8947

POST-POLIO SYNDROME

Perhaps the most disabling and least understood late effect of poliomyelitis is Post-Polio Syndrome. This is a term, coined in 1984 following the first international Post-Polio Conference in Warm Springs, Georgia, to describe a cluster of symptoms and signs experienced by poliomyelitis survivors 25 to 40 years after their original illness. The *criteria* for the diagnosis of Post-Polio Syndrome includes **a history of paralytic poliomyelitis, a stable period of neurological status after recovery, residual neurological deficits due to the initial poliomyelitis, new subjective muscular weakness, generalized fatigue and diffuse muscle/limb pain.**

Our understanding about Post-Polio Syndrome is limited at the present time. We know that Post-Polio Syndrome is not a reactivation of the poliomyelitis virus. Preliminary research has demonstrated some immunological data, but currently, there is insufficient evidence to attribute this condition to an autoimmune phenomenon. There is inconclusive evidence to conclude that Post-Polio Syndrome is a result of some other infectious process. In other words the etiology of Post-Polio Syndrome is unknown.

The prevalence of Post-Polio Syndrome was originally estimated to occur in one-third of polio survivors. However, it has recently been estimated to be as high as two-thirds of all polio survivors (Personal communication, International Conference on Polio, St. Louis, Missouri, May, 1997). Perhaps, the most debilitating aspect of Post-Polio Syndrome is the fatigue associated with this condition. The fatigue varies in severity; however, many patients report hitting the proverbial 'Post-Polio Wall', referring to an inability to carry on with an activity due to fatigue. This often occurs in the early to mid-afternoon and necessitates a rest or nap. Fatigue frequently precludes socialization and other enjoyments, thereby detracting from the quality of life with advancing age. Individuals with Post-Polio Syndrome frequently report increasing muscle weakness. This new weakness is not necessarily confined to muscles or areas of the body originally affected by the poliomyelitis virus. It can occur in the so called 'normal' limbs. This weakness can also affect the trunk musculature resulting in truncal instability, impaired balance and respiratory complications.

Ten percent of motor units are lost due to degeneration with each decade after the age of 60 years due to the aging process. Poliomyelitis survivors may have fewer motor units as a result of the original poliomyelitis infection and the remaining motor units may be giant units. Therefore, a relatively small loss of motor units can have a disproportionate impact on function.

This weakness may or may not be demonstrable in a clinical assessment. This may be due to the insensitivity of manual muscle-testing or by the fact that the weakness is due more to easy fatiguability of muscle as opposed to pathophysiological weakness per se. Irrespective of the etiology of the reported weakness, it has a profound impact on the individual's level of function. Frequency of falls increase, and concerns regarding safety arise. Ambulation may become increasingly difficult, necessitating the use of braces and ambulatory aids.

Very little is understood about the pain in Post-Polio Syndrome, recently termed *'Post-Polio Pain'* (Personal communication, International Conference on Polio, St. Louis, Missouri, May, 1997). This pain does not resemble that of other more familiar conditions. It is described neither as an ache or a burning. It tends to be migratory and not necessarily localized to muscles or joints. It tends to worsen towards the evening and responds poorly to traditional analgesics.

While weakness, pain and fatigue tend to be the hallmarks of Post-Polio Syndrome, this condition can be associated with a number of other problems including decreased balance, frequent falls for no apparent reason, variable paresthesiae, daytime somnolence, non-restorative sleep, sleep apnoea, restless leg syndrome and swallowing difficulties. Furthermore, individuals may report mild cognitive problems such as impaired short-term memory and decreased concentration.

The diagnosis of Post-Polio Syndrome is one of exclusion. Other causes for a patient's symptoms should be vigorously investigated. The list of differential diagnoses is long; however, the more common conditions that Post-Polio Syndrome may mimic include fibromyalgia, hypothyroidism, early polymyositis and depression. A thorough history and physical examination along with laboratory investigations will usually lead to the underlying etiology of the patient's symptoms.

All patients should be screened for psychological factors, particularly depression. This, however, can be difficult. One must determine if the depression is a primary cause of the individual's somatic complaints or is the depression secondary to the effect that Post-Polio Syndrome has had on that individual's quality of life.

There is no 'test' for Post-Polio Syndrome. Electromyography (EMG) can demonstrate features of the original poliomyelitis infection, i.e. giant motor units with no ongoing denervation. EMG cannot, however, diagnose Post-Polio Syndrome, and the EMG may, in fact, be normal in spite of the history of poliomyelitis.

Therefore, the diagnosis of Post-Polio Syndrome should only be made when the criteria outlined above have been met and all other potential etiologies have been excluded.

TREATMENT

Unfortunately, there is no 'cure' for Post-Polio Syndrome. As noted above, traditional analgesics tend to work poorly in this patient population. Non-traditional therapies such as tricylic agents may, in some cases, be effective. Traditional physiotherapy tends to be of limited value, whereas, acupuncture and Shiatsu massage tend to be more effective for pain relief.

Various pharmacological agents have been used to treat the fatigue of Post-Polio Syndrome with little success. Methylphenidate (Ritalin) has been used, with anecdotal reporting of improvement in fatigue. No controlled studies are available to evaluate the efficacy of this drug. Pyridostigmine (Mestinon) has also been tried. This drug has recently been evaluated in a clinical study by Doctors Neil Cashman and Daria Trojan of the Montreal Neurological Institute. Unfortunately, there was no evidence of improvement in reported severity of fatigue in individuals using this drug.[1] The authors do not recommend the use of pyridostigmine for the fatigue of Post-Polio Syndrome. Therefore, the primary management of fatigue in this condition is through energy conservation techniques and pacing strategies.

Appropriate exercise is extremely important in poliomyelitis survivors. In the early days, individuals recovering from poliomyelitis were instructed to exercise. They were led to believe that the harder they worked at exercise, the better their recovery would be. Now, however, it is believed that overwork of giant motor units may cause premature degeneration of these units due to 'burn out', leading to muscle weakness and loss of function. On the other hand, an individual should avoid muscle weakness that results from lack of use. Therefore, these individuals should perform regular exercise that is of low resistance and non-fatiguing in nature. Given the many orthopaedic problems associated with the late effects of poliomyelitis, water exercise is usually the most effective and safe method of maximizing and maintaining muscle strength.

The goal of treatment is to enhance the quality of life in those individuals with the late effects of poliomyelitis. This is achieved through life-style management, consisting of energy conservation and appropriate exercise. Various assistive devices are available to enhance the individual's level of function. When indicated, pulmonary function tests and a swallowing evaluation will help to direct appropriate treatment of these problems.

Psychological counseling may be required, particularly when a reactive depression exists. Psychological intervention can help these individuals deal with their physical limitations and functional losses. Some require assistance to accept the assistive devices that are now necessary for day to day activities. Others need help to deal with repressed childhood memories that can no longer remain buried.

Although there is no cure for Post-Polio Syndrome, the prognosis can still be positive. With the appropriate interventions and a willingness on the part of the poliomyelitis survivor to adopt the life-style changes required, these individuals can maintain a high level of quality of living as they age.

The Post-Polio Clinic, West Park Hospital, Toronto, is the only one in Ontario and one of only a few such clinics in North America. The clinic provides a comprehensive multidisciplinary evaluation of poliomyelitis survivors. The core team consists of a clinic coordinator, physiatrist, occupational therapist, physical therapist and social worker. Support services include psychological, othotics, dietary, respiratory, sleep and swallowing programs. Following their assessment in the Post-Polio Clinic, problems related to the late-effects of poliomyelitis are identified and management recommendations are offered to the client and their referring physician. The objective of the Post-Polio program is to address the needs of this very unique aging population of poliomyelitis survivors. The Post-Polio Clinic can be accessed by any physician wishing to refer one of their patients for assessment.

More information regarding the services offered through the Post-Polio Clinic, West Park Hospital, can be obtained by telephone, (416) 243-3600, ext. 2260, or by fax, (416) 243-8947.

REFERENCE

1. Polio Network News, International Polio Network, Saint Louis, Missouri, Winter 1998, Vol. 14, No. 1.