

The Late Effects of Polio

Information for General Practitioners

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Introduction

Although poliomyelitis is no longer a threat in the industrialised world, the past decade has seen an increase in the interest in this disease as many survivors have acknowledged new symptoms, including fatigue, weakness and pain, which are often debilitating and having a significant impact on the individual's daily functioning and quality of life. These symptoms, which can arise in an individual years after their recovery from their initial episode of polio, are commonly referred to as the "Late Effects of Polio".

General practice is most often the first point of contact with health care for members of the community. As a result, the General Practitioner (GP) has a major role in managing the health of their patients. GPs act as gatekeepers for their patients to access the rest of the health care system.

To provide quality of care to patients experiencing symptoms related to the late effects of polio, it is essential that health professionals be aware of the diagnosis of this condition and have an understanding of the underlying aetiology and pathophysiology of the symptoms.

The primary aim of this manual is to present contemporary concepts for the assessment, diagnosis, and management of the patient with a history of polio. It endeavours to assist the GP to make decisions regarding treatment and care, and to provide suggestions for the appropriate time to seek advice from specialists and allied health professionals in the management of their patients. A multidisciplinary approach to the assessment and management of these patients is often necessary due to the complex range of symptoms that these patients may experience.

This manual is divided into several sections:

- Acute Poliomyelitis
- Late Effects of Polio
- Clinical Features of the Late Effects of Polio
- Assessment
- Management
- The Role of the General Practitioner in the Management of the Late Effects of Polio
- Resources

Although the information included in this manual has been carefully reviewed, it is only a general guide. Management decisions should only be made subject to the clinician's judgement in each individual case. No responsibility is accepted by the authors for adverse outcomes resulting from clients or clinicians acting on the recommendations contained in this manual.

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Acute Poliomyelitis

This review will begin with an overview of acute poliomyelitis and will cover epidemiology, pathogenesis, clinical features, management and prevention. The history of acute poliomyelitis in Queensland will also be considered.

Poliomyelitis (infantile paralysis), commonly referred to as “polio”, is a viral infection that was common in the Western world until the early 1960s. The name poliomyelitis derives from the Greek words “polios” meaning grey, and “myelos” meaning marrow, referring to the fact that the disease results from the destruction of neurons located in the anterior horn grey matter in the spinal cord.¹

Epidemiology

The poliovirus has been in existence for thousands of years, and has had world-wide distribution. Robbins and Daniel (1997)² reviewed the early history of polio and cite a depiction of a young man with a withered leg in a 3000-year-old Egyptian mural. Only in the 19th century did descriptions of polio epidemics begin to appear. For most of its recorded history, the poliovirus was an endemic infection in regions with poor sanitation. As a result, infants were exposed to the virus early and subsequently developed lifelong immunity. In infancy, the illness tends to be benign and only rarely produces paralysis.

During the 20th century, improvements in sanitation in developed countries led to a decrease in endemic infection. As a result, large numbers of children and young adults who were not exposed to the virus as infants, became vulnerable to sporadic and epidemic infection and consequently, the paralysis rate increased.³ Polio epidemics are seasonal, with peaks in the hot, humid, summer months.⁴

Pathology and Pathogenesis

Poliovirus is extremely infectious, but is usually benign, with 90 to 95 percent of cases being asymptomatic. In four to eight percent of cases there is a non-specific viral syndrome and only one to two percent of cases are associated with paralysis. The rate of paralysis varies with the strain of the virus and the individual's age, with the likelihood of paralysis increasing with age. In children, paralysis occurs in 1/1000 cases, while in adults, paralysis occurs in 1/75.⁵ Polio is predominantly a disease of the very young, with 70 to 90 percent of cases occurring in those younger than 3 years.⁴

The poliovirus is a positive single stranded RNA enterovirus (picornavirus) and has three antigenically distinguishable types: poliovirus numbers I (responsible for approximately 85 percent of paralytic cases), II and III. None provide cross-immunity to the others.¹ Polio type infections can also be caused by other enteroviruses, a group that consists of the polioviruses, Coxsackie A (A7, A9) and B (B2 to B4) viruses (types 1, 2, 4, 6, 7, 9, 11, 16, 18, 30) ECHO viruses and enteroviruses 70 and 71.⁶

The poliovirus accesses the body orofaecally. After passing through the stomach, the virus reaches the intestine where it establishes itself in the cells of the intestinal lining. Viruses multiply in the lymphoid tissues of the oropharynx and intestine during the one- to three-week incubation period.⁷ In most cases, the virus is present in the oropharyngeal secretions for one to two weeks and is excreted in the stools for several weeks to months. From the lymphoid tissues of the oropharynx and intestine, the virus may move into the regional lymph nodes and from there into the blood, causing a transient viremia. If the virus accesses the circulatory system and a viremia results, the central nervous system is exposed to the infection and acute paralytic polio may result.¹

Poliovirus has a predilection for the motor neurons of the anterior horn of the cervical and lumbar regions of the spinal cord, which can result in the cell death or injury of these motor neurons. Following death of the anterior horn cells, wallerian degeneration results and the muscle fibres associated with those neurons become “orphaned” (denervated), producing clinical weakness.⁸ The localisation and degree of paralysis depends on the site and the severity of neuronal lesions.⁷

Studies undertaken more recently have shown that the involvement of the central nervous system is more diffuse than simply the motor neurons.⁹⁻¹¹ Lesions are also observed in the intermediate and posterior grey columns, and occasionally in the dorsal root ganglia. In the brainstem, the reticular formation and most of the nuclei of cranial nerves can be involved. In the cerebral cortex, neuronal lesions are usually mild and restricted to the precentral gyrus, thalamus, hypothalamus and the globus pallidus.¹

Clinical Features of Polio

During the acute polio epidemics in the early 20th century, the following categories were often used to classify the extent and seriousness of the disease.¹²

Sub-Clinical Polio

The patient is unaware of infection and gains active (sometime lifelong) immunity to further infection from the strain. Many people obtained immunity this way before the development of vaccines. Sub-clinical polio usually occurred in infants and very young children.

Abortive Polio

Abortive polio is often characterised by acute respiratory infection or gastroenteritis, but is generally not dangerous. The infection is cut short by the host's defences before it can enter the central nervous system. Symptoms may include fever, headache, vomiting, diarrhoea, constipation and sore throat.

Non-Paralytic Polio

The vast majority of infected individuals remain asymptomatic or experience a self-limited illness. In non-paralytic polio, symptoms generally tend to subside after one to three days.

Symptoms may include headache, neck, back, abdominal and extremity pain, fever, vomiting, lethargy and irritability. Muscle spasm is present in the extensors of the neck and back, usually present in the hamstring muscles, and of variable presence in other muscles throughout the body. Resistance to flexion of the neck is noted after a varying range of free flexion. The patient assumes the "tripod" position on sitting up, usually by rolling to avoid flexing the back. Straight leg raising is less than 90 degrees. Spasm may be observed when the patient is at rest or may be elicited by putting each muscle through the maximum range of motion. The muscles may be tender to palpation.

Paralytic Polio

Some individuals proceed to develop more severe symptoms. If the virus crosses the blood-brain barrier, it attacks nerve cells in the brain, brainstem and spinal cord. Paralysis may occur at any time during the febrile period.

In addition to the symptoms of non-paralytic polio, tremors and muscle weakness appear. Paresthesias (due to involvement of ganglionic neurons) and urinary retention are noted occasionally. Constipation and abdominal distention (ileus) may also occur.

Clinically, paralytic polio may be divided into two forms that may coexist:

1. Spinal Polio

This condition is characterised by flaccid paralysis of muscles innervated by the motor neurons of the spinal cord and is the most common type of paralytic polio. Unless paralysis is complete, paralysis is assymetrical. Lower limbs are more commonly affected than the upper limbs and trunk.¹³

2. Bulbar Polio

This condition involves damage of neurons in the reticular formation and the nuclei of cranial nerves in the brainstem, which may lead to dysphagia, dysphonia, facial weakness, nasal voice, regurgitation of fluids through the nose, weakness of the sternocleidomastoid and trapezius muscles, difficulty in chewing, inability to swallow or expel saliva and respiratory tract secretions. The most life threatening aspect of bulbar polio is respiratory involvement due to pontile (central) involvement. Autonomic dysfunction may occur producing cardiac arrhythmia, blood pressure instability and impaired bladder and bowel function.³

Recovery from Acute Paralytic Polio

Mortality from acute paralytic polio is usually the result of respiratory or bulbar involvement.³ In those who survive the acute illness and recover, paralysis remains static for several days or weeks before a slow recovery occurs over several months to years.⁷ Muscle strength in partially denervated muscles increases to a maximum over a two year period, with 60 percent of the muscle strength recovery occurring in the first three months after onset and 80 percent in the first six months. Further improvement may continue over the next two years.⁶

Muscle strength recovery and increase in functional ability occur by several physiological processes. These include:

- Terminal sprouting;
- Myofibre hypertrophy;
- Fibre type transformation; and
- Ongoing denervation and reinnervation.

Terminal Sprouting

During the recovery process following acute paralytic polio, remaining brainstem and spinal cord motor neurons can elaborate new branches, or axonal sprouts. The physiological basis for axonal sprouting is believed to be related to the expression of neural-cell adhesion molecules by the denervated myofibres.¹⁴ These molecules seem to provide a chemotactic stimulus to the terminal axons of neighbouring, surviving motor neurons, causing these neurons to send axonal sprouts to denervated muscle fibres.¹⁵ These sprouts can reinnervate orphaned muscle fibres that have been denervated by the acute polio infection. Sprouting (or collateral innervation) can restore the capacity of voluntary muscle fibres to contract and thus improve clinical strength.⁸

There is considerable electrophysiological evidence, including single fibre and macro-electromyography (EMG) and muscle morphological data to support this concept of reinnervation.¹⁶⁻²⁰ Muscle biopsy studies have shown that the remaining motor neurons may innervate up to eight times or more the normal number of muscle fibres.²¹ Survivors of acute polio may be left with a few, significantly enlarged motor units doing the work previously performed by many units.⁵ Figure 1 provides a schematic illustration of this process.

Myofibre Hypertrophy

In addition to sprouting, the remaining innervated muscle fibres hypertrophy through exercise and activity during the rehabilitation phase after the acute illness. Muscle fibre hypertrophy contributes further to the recovery of strength after paralytic polio. Because this mechanism of neurophysiological compensation is so effective, a muscle can retain normal strength even after 50 percent of the original motor neurons have been lost. Several studies have documented that the fibre area of type I and type IIA myofibres in the quadriceps of post-polio men are, on average, twice the control value.^{22,23}

Further studies have supported these results. In a study by Borg and colleagues (1988)²⁴ marked hypertrophy of the muscle fibres, as determined by surface electromyography during gait, within the tibialis anterior muscle, was observed in post-polio subjects who excessively overused that muscle. Grimby and colleagues (1989)²² reported a significant negative correlation between muscle strength and mean fibre area in post-polio male subjects, i.e. the weakest subjects had the largest muscle fibres

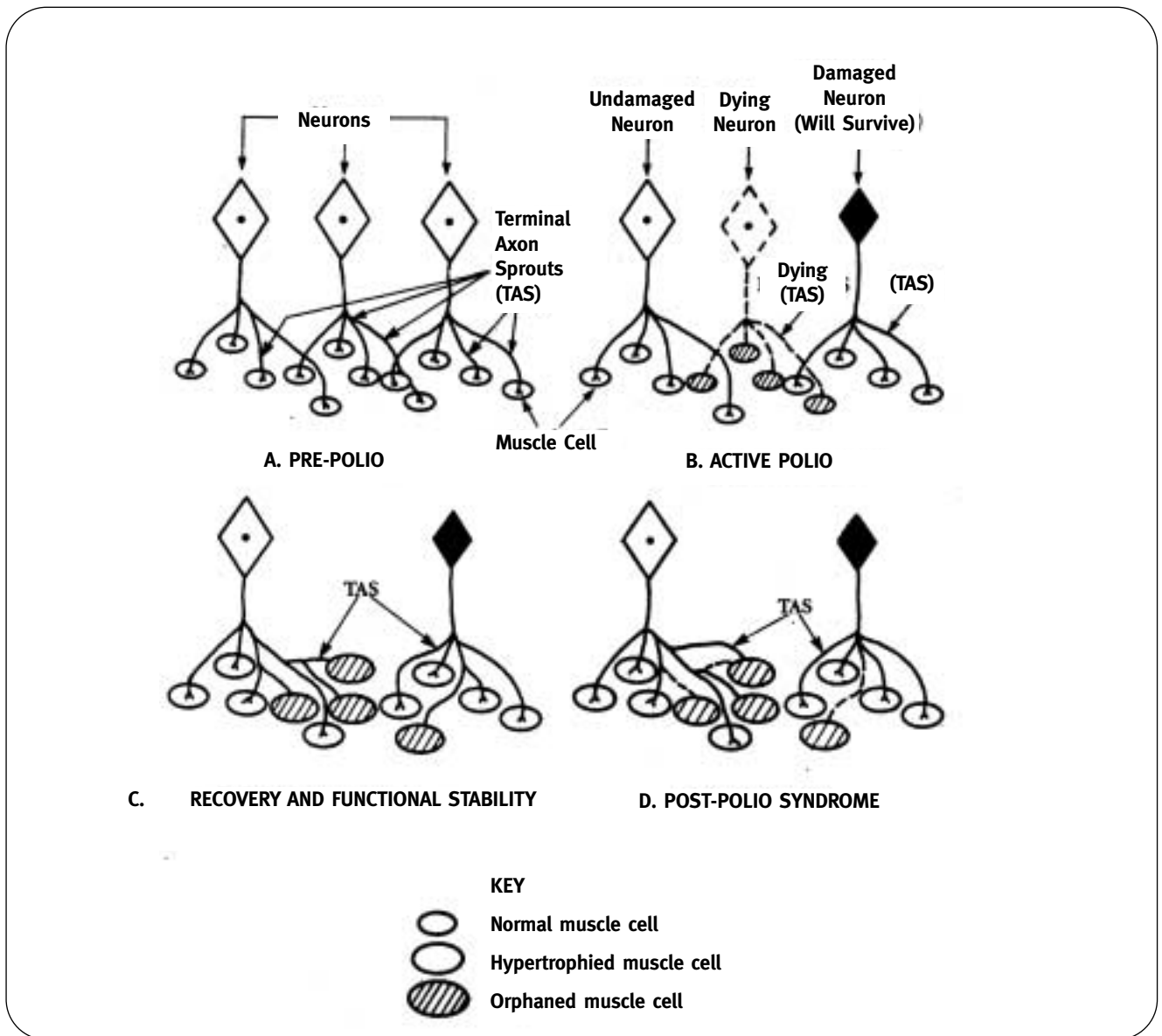


Figure 1: The Motor Neuron Unit Before and After Polio
(Adapted from Halstead, 1998)²⁵

Fibre Type Transformation

It has been speculated that fibre transformation from type II fibres (fast twitch, glycolytic) to type I (slow twitch, oxidative) fibres is another neuromuscular adaptive mechanism. A study by Grimby and colleagues (1989)²² demonstrated a significant negative correlation between the relative occurrence of type I fibres and muscle strength in the quadriceps muscles of women with a history of polio. They suggested that this may be due to transformation of type II to type I fibres in subjects with the most marked reduction in the number of muscle fibres and strength. Muscle biopsies conducted on the anterior tibialis muscle of ambulating post-polio subjects exhibited almost exclusively type I muscle fibres, while subjects who used

wheelchairs did not demonstrate type I fibre dominance. This suggested that this type I fibre dominance in some of the subjects was due to excessive use of remaining muscle fibres causing a transformation of type II to type I fibres.²⁴

Ongoing Denervation and Reinnervation

The process of denervation and reinnervation is ongoing in post-polio patients with and without complaints of new weakness. Reinnervation of recently denervated muscle fibres appears to be another adaptive neuromuscular mechanism to maintain function. Histologic²⁶ and electrophysiologic^{16,27,28} evidence supports this concept.

Such extensive compensatory physiological processes mask the profound neurological deficits caused by the disease. In addition to these physiological processes, the body possesses a number of compensatory mechanisms to maintain function in the presence of residual paralysis. These compensations include:

- Use of weak muscles at a higher level of capacity;
- Substitution of strong muscles with increased energy expenditure for the tasks; and
- Use of ligaments for stability with resulting hypermobility.

Residual Complications

Residual complications often result following the initial recovery process. These may include:

- Muscle paresis and paralysis, which may result in skeletal deformities, joint contractures and movement disability;
- Growth retardation of an affected limb;
- Osteoporosis and increased likelihood of fractures;
- Pain from wear and tear due to abnormal body mechanics;
- Compression neuropathy from the use of calipers or wheelchairs;
- Venous stasis due to pooling of blood in paralysed lower limbs;
- Chronic colonic distension;
- Respiratory insufficiency; and
- Intolerance to cold due to circulatory disturbances.

Management

Early management practices for paralysed muscles emphasised the need to rest the affected muscles and splint them to prevent contractures. Many paralysed patients lay in plaster body casts for months at a time. This prolonged casting often resulted in disuse and atrophy of muscles both affected and not affected by the disease.

In 1940, Sister Elizabeth Kenny, an Australian nurse, arrived in North America and challenged this approach to treatment. Having treated polio cases in rural Australia, Sister Kenny had improvised a form of therapy that aimed at relief of pain and spasm that was contrary to the opinion of much of the medical profession. Her treatment protocol involved the use of hot, moist packs to relieve muscle spasm and the prescription of early activity and exercise to maximise the strength of unaffected muscle fibres. Sister Kenny settled in Minnesota and established an institute, from which she began her world-wide crusade to advocate her system of treatment. Slowly, Sr Kenny's ideas won acceptance and by the mid 20th century had become the hallmark for the treatment of paralytic polio.²

A significant development which saved the lives of many with severe respiratory problems was the development and introduction of the mechanical tank respirator (or iron lung) during the 1930s.¹³ Other respiratory aids, such as the “rocking bed” were used in patients with less critical breathing difficulties.

Patients with residual paralysis were treated with braces and taught to compensate for lost function with the help of callipers, crutches and wheelchairs. Orthopaedic surgical procedures such as joint fixations, tendon transfers and limb lengthening and shortening, were used extensively.¹³

Prevention

The incidence of paralytic polio has reduced dramatically since the introduction of vaccination. Two forms of polio vaccine have been developed, the Salk Inactivated Polio Vaccine and Sabin Oral Polio Vaccine. Table 1 summarises the major characteristics of each of these vaccines.

Table 1: Comparison of Polio Vaccines

Salk Inactivated Polio Vaccine (IPV)	Sabin Oral Polio Vaccine (OPV)
<ul style="list-style-type: none"> • Developed in 1955 by Jonas Salk¹ • Stimulates serum IgM, IgG and IgA but not secretory U_gA – immunity induced by antibody transuding into the oropharynx⁸ • Delivered by injection • Requires trained personnel to administer injection • Confers immunity, but client can still act as a carrier • Remains indicated in pregnant, immunosuppressed and unvaccinated clients over the age of 50³¹ • No risk of vaccine associated paralysis 	<ul style="list-style-type: none"> • Developed by Albert Sabin and was approved for use in 1962¹ • Composed of attenuated or “weakened” live strains of poliovirus³⁰ • Delivered orally • Does not require extensive medical training to administer vaccine • Confers life-long immunity and prevents the client from acting as a carrier • World Health Organisation recommends the use of OPV, as it is less expensive and easier to administer to large populations of infants and children • Slight risk of vaccine associated paralysis. In a very small number of cases, vaccination is associated with paralytic polio (VAPP), which affects either individuals recently vaccinated with OPV, or non-vaccinated individuals (particularly the immunocompromised) in direct contact with healthy vaccinees.³² (Currently estimated that there is one case of VAPP per 2.5 million doses of OPV administered)¹
<ul style="list-style-type: none"> • Vaccinates only the client who receives it 	<ul style="list-style-type: none"> • May be transmitted to others for “secondary vaccination”

Eradication

In 1988, the World Health Organisation (WHO) undertook to eradicate wild poliovirus from the world, by the year 2000. Eradication is being achieved by means of:⁴

- Mass immunisation programs;
- Careful surveillance; and
- Targeting areas in which wild virus persists.

Despite the extensive, worldwide use of polio vaccines, polio has not been eradicated. As of 1 May 1999, 6277 polio cases with onset during 1998 were reported world-wide. Poliovirus transmission is now largely confined to southern Asia and parts of Africa. In the European Region, polio transmission is confined to south-eastern Turkey. The Western Pacific Region has not detected wild poliovirus since March 1997 and the Region of the Americas has been polio-free since 1991.³²

Polio in Australia

Polio was made a notifiable disease in all States and Territories of Australia in 1922. Major polio epidemics occurred during the late 1930s, early 1940s and the 1950s. It is estimated that a minimum of 20,000–40,000 individuals developed paralytic polio in Australia between 1930 and 1988, but the actual number of people infected by the virus is unknown.

Polio has now almost certainly been eradicated from Australia. The most recent case of polio, caused by wild poliovirus, was in 1978.³³ Data from the Australian Childhood Immunisation Register (1999) showed that 81 percent of one year old children in Australia had received the recommended three doses of OPV in their first year of life.³⁴

In Queensland

The annual rate of notifications in Queensland since 1917 is shown in Figure 2. The largest epidemic of polio in Queensland’s history was in 1951–1952.

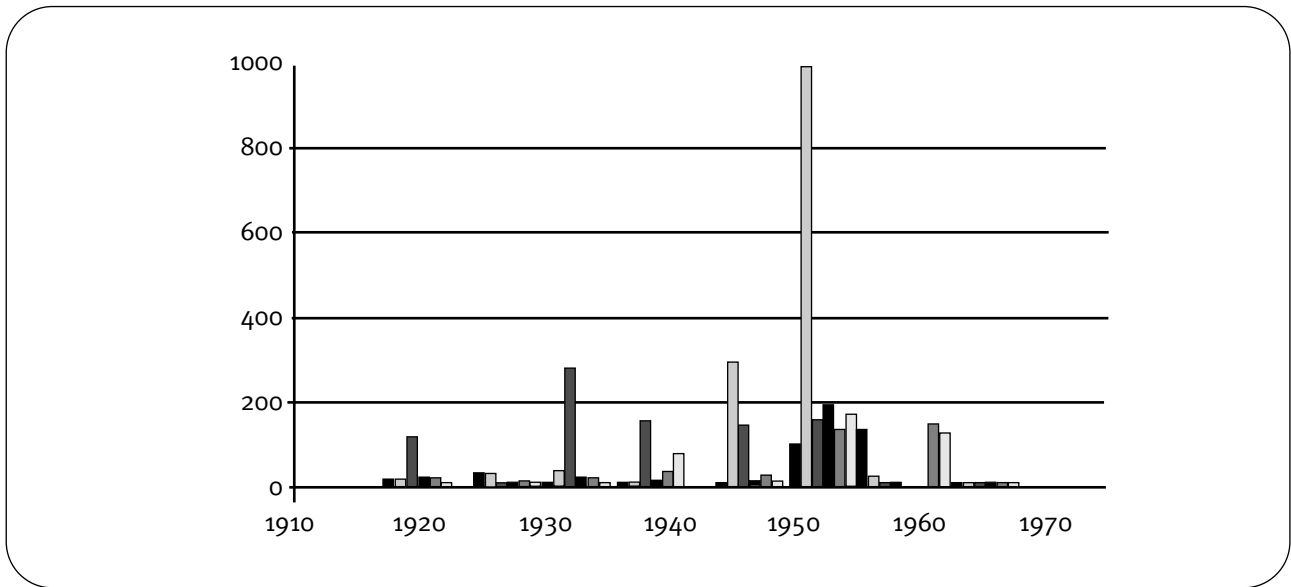


Figure 2: Annual Rate of Notifications of Acute Polio in Queensland³⁵
(Collated from the Annual Reports of Health and Medical Services Queensland 1917-1975)

In 1956, mass immunisation with the Salk vaccine commenced. For four years, very few cases of polio occurred. Then, in 1961-62, another epidemic occurred. Several cases were identified in vaccinated persons. It was found that the organism causing infection was a Type III poliovirus and that the potency of the Type III component in the vaccine being used was inadequate. After this deficiency was remedied, polio virtually disappeared from Queensland. In 1967, the oral Sabin vaccine was substituted for the injected Salk vaccine and was equally successful.³⁶ The last case of polio in Queensland was notified in 1975.³⁷

Late Effects of Polio

Polio was considered to be a chronic, yet stable disease once the acute phase was over and rehabilitation had restored a greater or lesser degree of function. Medical textbooks have until recently described polio as having three distinct stages: acute illness, period of recovery and stable disability. However, it is now known many polio survivors develop new symptoms after decades of stable functioning. In reviewing this topic, it is necessary to provide a brief historical background to the “Late Effects of Polio” (LEOP). The most appropriate diagnostic labels to use and the proposed aetiologies of the fourth stage of polio will then be considered.

Historical Background

It has been recognised for more than 100 years that new muscle weakness can occur in survivors of polio many years after their initial illness. The first descriptions were published in the French medical literature in 1875 by Jean-Martin Charcot. Since this time, other sporadic reports have appeared in the literature describing new weakness, atrophy and fasciculations occurring years after an episode of acute paralytic poliomyelitis.³⁸ It was not until the early 1980s however, that LEOP became widely acknowledged.

By 1984, a growing awareness of LEOP prompted researchers to organise an international conference at the Warm Springs Institute for Rehabilitation in the United States of America. The term “Post-Polio Syndrome” was coined around this time. The second international meeting was held at the Warm Springs Institute in 1986 and in the following years there was a dramatic increase in clinical research into the long-term effects of polio.²⁵

In 1994, the New York Academy of Sciences and the National Institute of Health co-sponsored 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 Pathogenesis and Treatment”. This conference signalled the acceptance of post-polio syndrome as a legitimate clinical entity.³⁹

Nomenclature and Definitions

There is disagreement in the literature regarding the most appropriate diagnostic labels to describe the new health problems being experienced by people with a past history of polio. The reasons for this lack of consensus are that:

- The various descriptive terms have lacked specific diagnostic criteria;
- There is currently no pathognomonic test for the condition; and
- Understanding of the underlying pathophysiology of the condition is incomplete.

The terms that have been most frequently used in the literature include “The Late Effects of Polio”, “Post-Polio Syndrome” and “Post-Polio Muscular Atrophy”.

The Late Effects of Polio (LEOP)

LEOP refers to the myriad of symptoms that individuals with a history of polio may experience. The features of LEOP can be considered in three broad categories. These include:-

1. Symptoms that can be attributed directly to damage caused by the poliovirus, including:
 - Residual weakness;
 - Musculoskeletal imbalance;
 - Growth retardation;
 - Skeletal deformities of affected limbs;
 - Respiratory insufficiency; and
 - Cold intolerance due to circulatory disturbances.

2. Symptoms thought to be related to the body’s failure to maintain the level of recovery that was achieved following the infection, such as new weakness and fatigue – The Post-Polio Syndrome.
3. Symptoms that result from secondary trauma, including:
 - Compression neuropathy, e.g. carpal tunnel syndrome after years of crutch walking;
 - Degenerative arthritis of joints that have been over-stressed due to compensatory body mechanics; and
 - Other repetitive motion problems such as tendonitis, bursitis and failing joint fusions.

Post-Polio Syndrome (PPS)

There have been many definitions of Post-Polio Syndrome (PPS) and there is still no widely agreed definition. Lauro Halstead, one of the leading researchers in this field, first proposed a definition for PPS approximately 15 years ago. He has continued to refine his definition of PPS over the years and in a more recent publication proposed the following definition:³⁹

“Post-Polio Syndrome is a neurologic disorder that produces a cluster of symptoms in individuals who had paralytic poliomyelitis many years earlier. Because these symptoms tend to occur together, they are called a syndrome. Typically, these problems occur after a period of functional and neurological stability of at least 15 years following the initial episode of poliomyelitis and include new weakness, fatigue, decreased endurance and loss of function. Some researchers also add pain as part of the syndrome, especially in muscles and joints. Less commonly, the symptoms include muscle atrophy, breathing and swallowing difficulties and cold intolerance.”

Mulder, Rosenbaum and Layton initially proposed specific criteria for the diagnosis of PPS in 1972.⁴⁰ These criteria were further refined by Halstead⁴¹ in 1991 and are outlined in Table 2.

1. A prior episode of paralytic polio confirmed by history, physical examination and typical findings on EMG.
2. Standard EMG evaluation demonstrated changes consistent with prior anterior horn cell disease.
3. Characteristic pattern of recovery – a period of neurological recovery followed by an extended interval of neurological and functional stability preceding the onset of new weakness, the interval of neurological and functional stability usually lasts 15 years or more.
4. The gradual or abrupt onset of new weakness in polio-affected muscles. This weakness may or may not be accompanied by new problems such as generalised fatigue, muscle atrophy, joint and muscle pain, decreased endurance and diminished function.
5. Exclusion of medical, orthopaedic and neurological conditions that may be causing the health problems listed in number 4 above.

The term “Post-Polio Syndrome” has developed sufficient specificity to be clinically useful and as a result, is gaining international acceptance. It has been suggested that the diagnosis of PPS should not be used indiscriminately for every person with a history of paralytic polio with a new complaint.⁵ Instead, the diagnosis of PPS should be reserved for those individuals whose symptomatology indicates motor unit dysfunction with variable musculoskeletal overuse. This opinion is further supported by Dr Pesi Katrak, Consultant in Rehabilitation Medicine, Senior Staff Specialist at Prince Henry and Prince of Wales Hospitals, Sydney. (See panel below for Dr Katrak’s comments)

Dr Pesi Katrak's comments in regard to the definition and diagnosis of PPS:

Whilst this definition of PPS is comprehensive, it includes symptoms such as muscle pain, joint pain and decreased function, which can occur as a result of mechanical factors in polio clients who have residual muscle weakness. Such musculo-skeletal problems would be expected to occur in these clients with advancing years, as indeed they would in non-polio subjects who may have a similar degree of weakness or skeletal abnormalities from any other condition. If the definitions of Mulder or Halstead are applied strictly, symptoms such as pain or declining function, which can be explained on a mechanical basis, should be considered as an "other orthopaedic condition" and hence should not be attributed to PPS.

The majority of clients present with complaints of declining function, tiredness, pain and a variety of other symptoms. In most instances, a detailed history and clinical examination, points to factors other than the PPS, which would account for the presenting symptoms. For instance:

- *Clients who have an asymmetry in limb lengths will almost inevitably suffer from degenerative or osteoarthritic changes in various major joints in the limbs and in the spine because of abnormal stresses on these structures with the passage of time. This together with the increased incidence of osteo-arthritis that occurs with increasing age results in pain and declining functional ability.*
- *It is generally accepted that physiologically there is a gradual decline in muscle strength with increasing age. Polio clients who have a moderate or severe degree of residual muscle weakness after recovery from the initial illness may find that the small amount of increase in weakness, related to age can result in a greater than anticipated decline in their ability to perform a variety of tasks. Clients will frequently perceive such a decline in function, as new weakness.*
- *Because of increased pain with physical activity or fear of falling from muscle weakness, clients tend to assume a more sedentary lifestyle and this can compound the problem of weakness due to relative disuse.*
- *Weight gain resulting in obesity and abnormal gait pattern due to focal weakness or asymmetry of limbs can often contribute to tiredness because of the increased energy requirements for carrying out day to day activities in the presence of such abnormalities.*

The diagnostic term I use for these clients, is Polio Related Problems (PRP). The term Late Effects of Polio (LEOP) would be equally acceptable.

The term PPS has been used by some clinicians to encompass many new symptoms in clients who suffered polio infection many years ago. Dr Lauro Halstead from the National Rehabilitation Hospital in Washington D.C., who has worked in this field for many years, recently noted that PPS is over diagnosed. I believe PPS is over diagnosed because of a variable interpretation of the diagnostic criteria.

My clinical impression from assessment of several hundred polio clients presenting with new symptoms is that only a very small proportion, around 15%, have PPS, i.e. progressive new weakness or excessive fatigue, which can not be explained on any other basis. I apply the diagnosis of PPS to only those clients who have a clear history of progressive new weakness or tiredness, where these symptoms cannot be explained by other factors.

Thus, whenever I see a symptomatic polio client, I try to decide whether the client's complaints are due to Post-Polio Syndrome or whether they are from Polio Related Problems. Sometimes this distinction is extremely difficult to make.

Progressive Post-Polio Muscular Atrophy (PPMA)

Progressive Post-Polio Muscular Atrophy is defined as:

“Progressive new weakness and atrophy in muscles with clinical or subclinical signs of chronic partial denervation/reinnervation compatible with previous acute polio.”

This term was coined by Dalakas and colleagues (1986)²⁸ to distinguish new, slowly progressive muscle weakness that is neurological in origin, from musculoskeletal problems or degeneration problems or both. PPMA is considered to refer to only a subgroup of those suffering from PPS.⁵ The term PPMA is less often used today, giving way to the term PPS.

In summary, there is currently no consistently agreed upon diagnostic name for the new health problems associated with former polio. There are currently no pathognomonic tests available and firm diagnostic criteria have not been established.

In the review of post-polio which follows, the term Late Effects of Polio (LEOP) will be used when discussing the myriad of problems that individuals with a history of polio often experience. Post-polio syndrome (PPS) will be used to describe the new neurological problems of fatigue and weakness that polio survivors are experiencing many years after their original illness.

Epidemiology of Post-Polio Syndrome

Epidemiological studies have reported varying estimates of the incidence of PPS depending on the criteria that were used to define the condition. A study by Codd and his colleagues (1985)⁴² found that 22.4 percent of patients with previous paralytic polio developed new symptoms. A further study by Windebank and colleagues (1991)⁴³ of the same group, found that 64 percent had developed new symptoms and 44 percent had reported new weakness. Further studies have yielded results ranging from 28 percent⁴⁴ to 42 percent.⁴⁵

The time period between acute polio and the onset of PPS has ranged from 8 to 71 years.⁴⁶ In various studies, the average interval has been found to be around 35 years.^{46,47}

Risk Factors

A number of surveys of new health problems in late polio have included an analysis of potential risk factors, but no firm conclusions were drawn. There are several reasons for this. Firstly, there have only been a small number of studies examining this area and their results have been conflicting. Secondly, due to methodological differences, e.g. the definition of PPS used to identify subjects, it is difficult to compare the findings of these studies. The risk factors that have been considered in surveys of polio survivors include:

Risk factors related to the acute polio episode:

- Age at onset^{43,48-50}
- Severity of paralysis^{43,44,48-52}
- Use of ventilator^{49,52}
- Hospitalisation^{49,52}
- Year of acute polio infection⁴⁹
- Gender^{44,48-50,52}

Risk factors related to the post-polio period:

- Polio to post-polio interval^{43,44,48}
- Current age^{43,48,49}
- Functional recovery and residual impairment^{45,48}
- Weight gain⁴⁸
- Presence of muscle pain associated with exercise⁴⁸

On the basis of these studies the most important risk factor appears to severity of the acute polio illness.

Progression

LEOP has only been widely recognised for a short period of time. As a result, studies that have investigated the progression of symptoms are limited in number and have tended to focus mainly on the progression of muscle weakness.

Dalakas and colleagues (1986)²⁸ followed a group of post-polio subjects over an average period of 8.2 years and examined the progression of new weakness. Using manual muscle testing (MRC scale) they found that the pace of worsening differed from subject to subject, being generally slow and variable even within the same subject. Long periods (up to 10 years) of stability were not uncommon. However, for a cumulative 10-year period, the average progression of weakness was estimated as one percent per year. They reported that the subjects' gender, age at onset of new symptoms and physical activity level preceding the development of new weakness did not appear to contribute significantly to the rate of progression. They also commented that the impact of the new weakness on the functional capabilities of the individual is variable, but appears to depend mostly on the residual deficit the person is left with. The more severe the residual polio deficit following the acute illness, the greater the functional impact of new weakness on the individual's neuromuscular function.

A more recently published study by Grimby and colleagues (1998)⁵³ investigated quadriceps muscle strength in 30 legs in 21 post-polio subjects over a period of eight years. All limbs tested had EMG evidence of previous polio. On average, there was a decrease in isometric muscle strength of nine percent, in isokinetic strength at 60°/s angle velocity of 13 percent and in isokinetic strength at 180°/s angle velocity of 15 percent. They divided the legs into those in which increased muscle weakness had been perceived during the eight-year period (unstable) (n= 20) and those with no perceived new weakness (stable) (n=10). The decrease during the eight years was 12-19 percent for the three strength measures in the unstable legs and 1-7 percent in the stable legs. This was contrasted with a reduction of 4-8 percent over an eight-year period in healthy controls.

A group of 50 subjects from the Mayo Clinic, who had previously been studied by Windebank and colleagues (1991)⁴³ were re-examined five years later by Windebank, Litchy, Daube and Iverson (1996).⁵⁴ This group of 50 paralytic polio survivors were investigated on both occasions using a structured history questionnaire, scored neurological examination, detailed electrophysiological studies, isometric muscle strength measurement, pulmonary function tests, psychological inventories and timed tests of function including gait and upper limb dexterity. All measures of neuromuscular function demonstrated stability over the five-year period in this group of subjects.

Stanghelle and Festvag (1997)⁵⁵ investigated progression of symptoms over a three to five year period in a group of 63 subjects who had all received a diagnosis of PPS based on the criteria proposed by Halstead and Rossi (1987).⁵² All subjects had previously received comprehensive multidisciplinary assessment and intervention. The subjects answered a questionnaire about their subjective symptoms, medical and social situation, and underwent spirometry and symptom-limited exercise stress testing. Seventy-five percent of subjects reported new weakness in polio-affected muscles during the follow-up period. General fatigue was an increasing problem in 77 percent of subjects and 61 percent reported increased muscle and joint pain during the follow-up period. A pronounced reduction in peak oxygen uptake (compared to normal values) was seen at the first evaluation. At the second examination, peak oxygen uptake was decreased further than predicted by increasing age.

While studies of muscle weakness in post-polio groups have shown relatively slow rates of progression, the study by Stanghelle and Festvag (1997)⁵⁵ points to a more alarming deterioration in subjective symptoms, physical disability, and cardiorespiratory fitness in post-polio individuals despite comprehensive multidisciplinary intervention. These findings were in contrast with those of Windebank and colleagues (1996)⁵⁴ who reported stability on a range of measures over a five-year period. Stanghelle and Festvag (1997)⁵⁵ acknowledged that their sample consisted of subjects who had already been diagnosed with PPS and thus the extent of progression in these subjects could not be extrapolated to the polio population in general. Obviously further research is required to establish the likely prognosis for individuals with a history of polio.

Aetiology of Post-Polio Syndrome

The pathological changes that produce the symptoms of post-polio syndrome are not well understood, but several theories have been developed over the years (Table 3).

Table 3: Proposed Aetiologies of PPS⁵

1. Motor unit dysfunction – degenerative changes within motor units
2. Muscle overuse
3. Muscle disuse
4. Loss of motor units with ageing
5. Predisposition to motor neuron degeneration because of glial, vascular, and lymphatic changes caused by acute polio
6. Chronic poliovirus infection or virus reactivation
7. An immune mediated syndrome
8. The effect of growth hormone
9. The combined effects of disuse, overuse, pain, weight gain or other illnesses

These theories will be briefly explained.

Motor Unit Dysfunction – Degenerative Changes Within Motor Units

Theory:

The new weakness and fatigue characteristic of PPS are due to degenerative changes in motor units, particularly loss of terminal axonal sprouts (peripheral disintegration).

Research:

This phenomenon, which occurs during recovery from paralytic polio (i.e. sprouting from surviving motor neurons leading to reinnervation of muscle fibres), is well documented. There is now considerable evidence that suggests that the abnormally enlarged motor units that develop after acute paralytic polio are not indefinitely stable.

Conventional electromyographic observations on post-polio muscles show that muscles which become extremely weak and atrophic have few to no motor unit action potentials (MUAPs), while other muscles, including those that are clinically normal or historically not affected, have large, polyphasic MUAPs.^{17,27} This is expected when one considers the generalised nature of the acute illness. The size of the voluntary motor unit action potentials are increased due to a very successful reinnervating process that results in giant-size motor units.⁵⁶

In a study comparing the size of MUAPs in control subjects and post-polio subjects with and without new weakness, Agre and Rodriquez (1990)¹⁷ found that the size of MUAPs was significantly larger in all post-polio subjects, and that those with new weakness had larger units than those without.

Wiechers and Hubbell (1981)¹⁶ first proposed that the abnormally enlarged motor units that develop after acute paralytic polio are not indefinitely stable, but that terminal axonal sprouts degenerate over time producing denervation of muscle fibres. It is possible that some of these denervated muscle fibres may become reinnervated by sprouts from neighbouring motor neurons, producing a continuous “remodelling” process of the post-polio motor units.

Many researchers now agree that this ongoing denervating and re-reinnervating process stresses the motor neurons, which after a number of years appear to lose their ability to maintain the metabolic demands of all their sprouts. Consequently, there is slow deterioration of some nerve terminals. As axonal sprouts die, some muscle fibres become permanently denervated and the post-polio individual experiences new weakness and other symptoms of neurological dysfunction.^{5,7,56} This process is illustrated in Figure 1 (page 4). Several electrophysiological and muscle biopsy studies have provided further evidence of neuromuscular transmission abnormalities and muscle fibre denervation in post-polio subjects.^{18,28,57}

New weakness in PPS can be explained on the basis of the reduced size of the motor unit when the reinnervation cannot keep pace with the ongoing denervation.⁵⁶ Trojan and Cashman (1997)⁹ hypothesised that if terminal axonal degeneration is the cause of new weakness in PPS, it is likely that there is a period of terminal axonal dysfunction that may precede degeneration by months to years. They believe that muscle fatigue in PPS can be attributed to neuromuscular junction transmission defects.

Muscle Overuse

Theory:

Enlarged motor units supplying post-polio muscles cannot indefinitely maintain the increased metabolic activity needed. As a result, overused motor units eventually degenerate, resulting in slowly increasing weakness and rapid fatiguing of muscles.

Research:

Increased creatinine kinase (CK) levels have been found in symptomatic subjects after polio, but not in equally weak asymptomatic subjects after polio.⁵⁴ Waring and McLaurin (1992)⁵⁸ found CK levels to be significantly correlated with distance of ambulation in post-polio subjects, suggesting that exercise is the cause of elevated CK in this population. Since elevated CK levels can be a marker for muscle injury, the increased CK levels may indicate muscle injury or overuse in post-polio individuals.

Perry, Fontaine and Mulroy (1995)⁵⁹ performed dynamic EMG studies during gait in post-polio subjects and found evidence of overuse in the biceps, gluteus maximus and quadriceps muscles.

Trojan and colleagues found in 1994 that muscle pain (especially that associated with exercise), joint pain, and a recent weight gain, were associated with PPS.⁴⁸ All of these factors can be markers of overuse, providing further evidence for the theory that overuse is a contributing factor to PPS. Numerous clinical studies have shown a correlation between the presence of PPS and a history of severe initial paralysis with a relatively good recovery of useful strength and function.^{48,52,60}

These observations support the theory that weakness and fatigue in PPS results from long-term substitutions for muscular weakness that place increased demands on muscles.

Muscle Disuse

Theory:

*Disuse produces both deconditioning and muscle weakness in individuals.*⁶¹

Research:

Post-polio subjects have been noted to have short-term increased weakness after a period of decreased activity that is secondary to illness or injury.¹⁵ The role of muscle disuse in the development of long-term weakness, however, is less clear.⁵

Loss of Motor Units with Ageing

Theory:

The natural ageing process depletes remaining anterior horn cells, leading to progressive weakness.

Research:

The normal ageing process is known to involve a gradual loss of motor neurons, but only becomes prominent after the age of 60.⁶² Several studies have failed to show a consistent correlation between the onset of new weakness and chronological age. The most consistent variable is the length of the interval between onset of polio and the appearance of new symptoms. Most subjects develop new weakness 30 to 40 years after their initial infection, and the age of onset of symptoms is variable.⁵ The superimposition of the normal ageing process on the already limited number of motor neurons present after paralytic polio may contribute to the development of PPS.⁷ The current consensus is that while chronological age may contribute to the development of new weakness, it is probably not the primary causative factor.^{5,7,56}

Less Supported Theories

Other theories that have emerged to explain the triggering mechanisms for motor unit degeneration include:

- Predisposition to motor neuron degeneration because of glial, vascular and lymphatic changes caused by acute polio – damage to the glial cells and vascular supply at the time of the acute infection can lead to secondary dysfunction of anterior horn cells.⁶³
- Chronic poliovirus infection or virus reactivation – PPS may be caused by re-exposure to live poliovirus or re-activation of persistent poliovirus in the CNS.^{28,46,64}
- An immune mediated syndrome – an ongoing inflammatory or immune response mechanism may be a precipitating factor in the development of PPS.^{5,65,66}
- The effect of growth hormone – ageing of the hypothalamus growth hormone axis may be a precipitating factor in the development of PPS.^{67,68}

While none of these theories have been completely excluded, not enough evidence exists to strongly support any of them at present.⁶⁹

The Combined Effects of Disuse, Overuse, Pain, Weight Gain or Other Illnesses

Gawne and Halstead (1995)⁵ discussed the way a number of factors may interact with each other and contribute to the development of progressive weakness and fatigue. Figure 3 is a schematic model of the possible aetiological factors for PPS and their interactions.

Chronic overuse of muscles may result in the development of new weakness, which in turn may lead to disuse. Musculoskeletal disuse leads to further weakness, atrophy, contractures, diminished endurance and weight gain. In the presence of overuse, musculoskeletal pain may occur, causing the individual to either rest, resulting in deconditioning, or compensate with improper body mechanics, leading to further overuse and possibly pain elsewhere.⁵ This complex clinical picture presents a challenge to those attempting to diagnose and manage people with the LEOP.

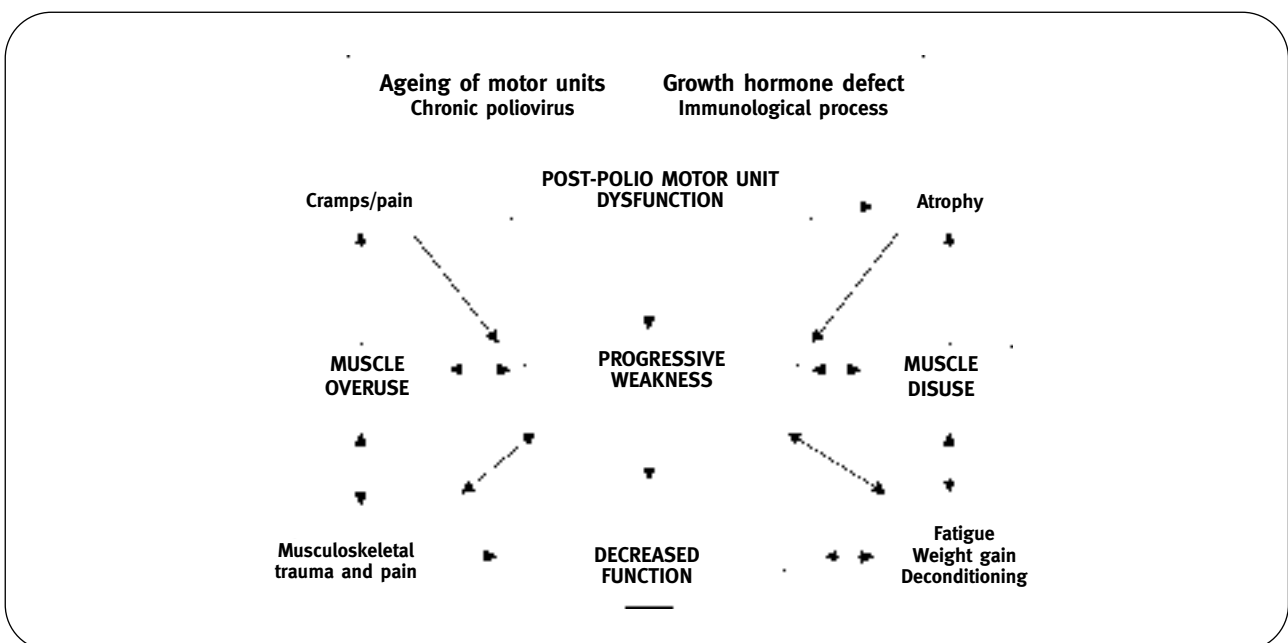


Figure 3: Schematic Model Demonstrating Possible Aetiological Factors for the Late Effects of Polio and their Interactions

(Adapted from Gawne and Halstead, 1995)⁵

Clinical Features of the Late Effects of Polio

In this section, the clinical features of LEOP will be discussed in detail. A review of some of the psychological issues facing post-polio individuals will also be presented.

Over the past fifteen years there have been many clinic and questionnaire based surveys of post-polio subjects. From this research it has become increasingly clear that there is a common set of symptoms and complaints that are frequently found in the post-polio population. The relative frequencies of the most common symptoms reported have been found to be similar in these various surveys.^{51, 70-74}

The most common post-polio symptoms are:

- Increased muscle weakness;
- Muscle and joint pain;
- Fatigue; and
- New difficulties with activities of daily living, particularly mobility related tasks.

Other symptoms include:

- Cold intolerance;
- Respiratory dysfunction;
- Sleep impairment;
- Dysphagia; and
- Speech difficulties.

Muscle Weakness

New muscle weakness, with or without associated muscle atrophy, can involve both previously affected muscles which were partially or fully recovered, or muscles that appeared to be unaffected by the original polio infection. New weakness is characteristically asymmetrical and is often most prominent in those muscles that were involved in the acute illness and then showed good recovery. New weakness is also commonly found in the “good” limb that was thought to be spared by the poliovirus, but which, in fact, may have had subclinical polio and has overworked over the years to compensate for the more affected limb. Other signs relating to new muscular involvement include fasciculations, muscle cramps, atrophy and elevation of muscle enzymes in the blood.⁷⁵

Surveys of post-polio subjects have reported the following frequencies of new muscular symptoms:

- | | |
|---|---------------------------------------|
| • New weakness in previously affected muscles | 60% ⁷¹ - 87% ⁷⁰ |
| • New weakness in previously unaffected muscles | 37% ⁷¹ - 77% ⁷⁰ |
| • New muscle atrophy | 17% ⁷³ - 28% ⁵⁰ |
| • Muscle twitching or fasciculations | 39% ⁵⁰ - 41% ⁷³ |
| • Muscle cramps | 43% ⁵⁰ - 48% ⁷³ |

Recent research on the course of muscle strength over time in the normal population over the age of 50 has shown a normal average decline of one percent per year, yet for post-polio individuals the rate was two percent per year.⁷⁶

It is important to realise that people with a history of paralytic polio may have significant denervation of muscle fibres even when muscle strength appears normal on clinical assessment. A study by Perry and Fleming (1985)⁷⁷ demonstrated that post-polio muscles that appeared normal (Grade 5) or good (Grade 4) clinically, as evaluated by a manual muscle assessment, may in fact have significant

denervation when tested with EMG. These results were further supported by previous research by Beasley (1961)⁷⁸ who reported that the number of functioning motor fibres in polio survivors with a given muscle test grade is significantly less than would be expected from simple percentage calculations. As a result, recommendations for exercise and activity must be correlated with this reduced number of muscle fibres.

Refer to aetiology of post-polio syndrome for possible causes of muscle weakness in the post-polio individual.

Fatigue

Fatigue is often reported to be the most common and debilitating symptom of PPS^{42,70,79,80} and has been reported by 48⁷¹ to 87⁷⁰ percent of subjects in previous surveys of LEOP. In the Queensland study by Lynch (2000)⁷⁴ 79 percent of subjects reported new symptoms of fatigue.

In two national surveys of American polio survivors, 91 percent reported new or increased fatigue, 41 percent reported fatigue significantly interfering with performing or completing their work and 25 percent reported fatigue interfering with self-care activities.^{81,82} Fatigue was reported to be triggered or exacerbated by physical overexertion in 92 percent and by emotional stress in 61 percent of study participants.

Post-polio fatigue appears to occur in two forms:

General Fatigue

General (central) fatigue has been described as an overwhelming exhaustion with flu-like aching and a marked change in the level of energy, physical and mental endurance following minimal activity. A common description of general fatigue, coined by Halstead and colleagues in 1985,⁴⁸ is that of the “polio wall”, a sudden feeling of overwhelming exhaustion. This sudden onset of symptoms includes intense fatigue, weakness, hot and cold flushes, and sweating. Commonly it occurs late in the afternoon or early evening and is typically brought on by an accumulation of activities that previously did not require special effort or cause noticeable sequelae.

Contributing causes of general fatigue may include:⁸³⁻⁸⁶

- Chronic pain;
- Respiratory compromise;
- Depression;
- Sleep disorders;
- Dysfunction of the reticular activating system; and
- Type A behaviour.

Fatigue can affect mental as well as physical function. Between 70 and 90 percent of American polio survivors with fatigue reported problems with concentration, memory, attention, word finding, maintaining wakefulness and clear thinking. In 77 percent of these individuals the cognitive difficulties were described as moderate to severe.⁹ Despite these multiple cognitive complaints, the only significant deficits that have been observed on formal neuropsychological testing in severely fatigued post-polio subjects were in the areas of attention and information processing speed.^{87,88}

Cognitive problems reported by people with a history of polio suggest that the fatigue experienced cannot be explained merely by damage to the anterior horn motor neurons.⁸⁹ Bruno and colleagues (1991)⁹ have suggested that the emergence of fatigue decades after acute polio may result from normal age related changes and the loss of brain activating system neurons that had survived the acute polio

infection, combined with an already decreased number of neurons as a result of the original poliovirus infection. During the acute illness, subjects often reported symptoms of drowsiness, lethargy, fatigue and poor attention, similar to the symptoms that they are now experiencing.¹¹ The loss of brain activating system neurons would decrease cortical activation, reduce attention and produce the symptoms of fatigue experienced by post-polio survivors.⁸⁸

Previous research has indicated that the poliovirus often damaged brain areas responsible for cortical activation and attention, including the reticular formation, posterior hypothalamus and thalamus as well as the putamen, caudate, locus ceruleus and substantia nigra.¹⁰ Reduction of neurotransmitters, in particular dopamine, due to damage to the substantia nigra, may impair the individual's ability to activate the cortex, resulting in difficulties with attention, concentration and maintaining wakefulness.

Muscle Fatigue

Muscle (peripheral) fatigue is reported as a decline in muscle strength upon exertion, which may be best described as muscle fatiguability or lack of endurance. Post-polio individuals have described muscle fatigue as “a heavy sensation in the muscles,” “increased physical weakness,” and an “increased loss of strength during exercise.”⁹⁰ Muscle strength usually returns after a period of rest.

Contributing causes for muscular fatigue may include:^{91,92}

- Overuse myopathy;
- Muscle fibre type disproportion;
- Defective muscular function; and
- Neuromuscular junction transmission defects.

Pain

Pain in muscles and joints is the first or second most common symptom in the majority of surveys of LEOP. Muscle pain has been reported by 43⁷³ to 80⁵¹ percent of subjects in post-polio surveys, and by 61 percent of subjects in the Queensland cohort.⁷⁴

Reports of joint pain in post-polio surveys have ranged from 55⁷¹ to 79⁷⁰ percent. In the Queensland study by Lynch (2000) 79 percent of subjects reported joint pain.

As observed with fatigue, the onset of new muscle and joint pain begins insidiously and often without an apparent precipitant. The subjective nature of pain often makes assessment and treatment difficult. Gawne and Halstead (1995)⁵ have proposed a classification system that divides pain experienced by post-polio patients into three categories. This classification system is used at the National Rehabilitation Hospital Post-Polio Clinic to facilitate the diagnosis and treatment of pain.

1. Post-Polio Muscle Pain (Myalgias)

- Occurs in muscles affected by polio;
- Described as either a deep muscle ache or a superficial burning pain – many individuals describe the deep muscle pain to be similar to that which they experienced during acute polio;
- Deep muscle pain is often characterised by muscle cramps or fasciculations (sensation of crawling);
- Typically occurs at night or end of day when the individual relaxes;
- Exacerbated by physical activity, stress and cold temperatures; and
- Alleviated in part by use of moist heat, slow stretching and rest.

2. Overuse Pain

- Includes injuries to soft tissues, muscle, tendons, bursa and ligaments, e.g. rotator cuff tendonitis, deltoid bursitis, myofascial pain (occurs most frequently in muscles of the upper back and shoulders and is characterised by bands of taut muscles and discrete trigger points) and fibromyalgia (commonly found in the post-polio population⁹²); and
- Occurs due to poor posture or improper body biomechanics over the years.

3. Biomechanical Pain

- Most typical form of pain reported by individuals with a history of polio;
- Presents as degenerative joint disease or pain from nerve compression syndromes (carpal tunnel syndrome, ulnar nerve impingement at the wrist or elbow, cervical or lumbosacral radiculopathies);
- Location of pain is often related to the method used by the individual to mobilise;
- Weakness in polio affected muscles and poor body mechanics places excessive and abnormal forces on joint structures, making joints more susceptible to development of degenerative joint disease;
- Commonly associated with specific activities such as weightbearing;
- Rarely accompanied by inflammation; and
- Primarily caused by long-term microtrauma from abnormal biomechanical forces as well as by injuries resulting from falls.

ADL Difficulties

Post-polio individuals often find it increasingly difficult to perform their daily activities as the result of pain, new weakness and fatigue. New or increased problems are reported in the areas of mobility related activities (such as ambulating, climbing stairs, transferring in and out of bed), performing personal hygiene and grooming tasks, lifestyle and employment. Activities that had previously caused no problems for the individual now produce new challenges.

Many people with a history of paralytic polio have functioned at extremely high levels on relatively few good muscle groups. Their bodies have managed with this loss of muscle function over the years through compensatory movements and function. With increased weakness, pain and fatigue, the compensatory mechanisms that have been used are often disrupted, affecting the patient's ability to perform their daily activities.

Postural Abnormalities

Postural deformity can lead to pain and decreased energy efficiency during various activities. Common postural malalignments found in the post-polio population include:

- Scoliosis;
- Upper thoracic kyphosis and forward head posture with compensatory lumbar hyperlordosis;
- Genu valgus and recurvatum;
- Pelvic obliquity;
- Uneven weight distribution between the limbs and through the soles of the feet; and
- Calcaneal valgus or varus.

Although many post-polio individuals may have been functioning adequately over the years with fixed deformities, the deformities can often progress and contribute to the musculoskeletal symptoms

they now experience. There are a number of reasons why this occurs. Tension in the muscles reduces tissue vascularity, leading to microdegeneration. Chronic strain causes polymer lengthening of collagen fibres.⁹³ Repeated tension results in longer fibres with increased collagen turnover and susceptibility for muscle fibres to rupture or increased curvature of an already deformed joint. Early ageing of these collagen tissues, which become stiffer and less flexible owing to extra wear and tear, along with joint cartilage degeneration, further contributes to the manifestation of musculoskeletal difficulties in these patients.⁶

Sleep Impairment

Post-polio individuals have a high incidence of sleep disturbances with poor sleep quality and frequent awakenings, which may be due to several factors.⁶ These factors include primary sleep disorders and muscle twitching.

Primary Sleep Disorders

1. Obstructive Sleep Apnoea (OSA)

OSA results when the upper airway collapses and causes repeated interruptions in airflow (apnoea). Apnoeas are terminated by arousal from sleep, which may occur many times per night, resulting in sleep disruption. This may result in either hypersomnia or insomnia. OSA is also a risk factor for hypertension, myocardial infarction, congestive heart failure and stroke. OSA is suggested by a history of loud snoring, observed interruptions in breathing and daytime sleepiness. OSA is related to pharyngeal weakness plus an increase in musculoskeletal deformities such as kyphoscoliosis or a co-existent emphysema.

2. Central Sleep Apnoea (CSA)

CSA occurs when the brains reflexes for triggering breathing during sleep are defective. This can occur due to brain diseases (such as some examples of PPS) or cardiovascular diseases, and may co-exist with other breathing problems. Many individuals with CSA have difficulty initially falling asleep because of frequent central apnoeas with arousal at the transition from wakefulness to sleep. In post-polio individuals it may be due to residual dysfunction of the surviving bulbar reticular neurons.

3. Hypoventilation

Hypoventilation results from restriction ascribed to scoliosis or respiratory muscle weakness, or both.⁹⁴

Muscle Twitching

Sleep disturbances may also result from random muscle twitching that occurs at night. A survey by Bruno and Frick (1991)⁹³ found that two-thirds of patients with a history of polio reported that their muscles twitched or jumped at night with 33 percent reporting that their sleep was disturbed by twitching. Further studies, involving monitoring PPS subjects, documented a number of abnormal movements during sleep. These included restless leg syndrome, periodic movement in sleep and generalised random myoclonus (involving contraction of muscles throughout the body), which disturbed the individual's sleep patterns.⁹⁵

Cold Sensitivity

Many individuals with a history of polio experience difficulty in tolerating cool or cold temperatures. Cold sensitivity has been reported by 46⁷¹ to 62⁷² percent of subjects in post-polio surveys. In the study by Lynch (2000)⁷⁴ on a sample of the Queensland post-polio population, 57 percent of subjects reported sensitivity to the cold.

On examination, the core body temperature of these individuals is almost always normal but limbs with significant atrophy tend to be cool to touch with a bluish discoloration and variable amounts of swelling. Individuals often report increased levels of fatigue and weakness when exposed to the cold. Bruno and colleagues (1985)⁹⁶ found that people with PPS lost 75 percent of their muscle strength when the room temperature dropped from 29 degrees to 18 degrees celsius.

Damage to the intermediolateral cell column (from acute polio infection) causes a decrease in sympathetic vasoconstrictor outflow. In paralysed muscle, the process is facilitated by an impaired muscle pump mechanism that allows the blood to pool and contributes to swelling in the limbs. The engorged capillaries of the skin release heat into the environment, decreasing skin temperature. As a result, arteriolar sphincters constrict, diminishing the blood flow and further lowering tissue temperature. This cooling produces a slowing in the nerve conduction velocity, an increase in the muscle fibre membrane refractory period, a decrease in muscle spindle firing and an increase in muscle viscosity resulting in a decreased muscle response to motor stimuli.⁹⁵

Respiratory Insufficiency

One of the most feared complications of acute paralytic polio was impaired respiratory function, which resulted in significant disability and, in many cases, death. As a result, the development of new respiratory problems in surviving post-polio individuals is of major concern to them.

New breathing difficulties have been reported by 27⁵¹ to 58⁷⁰ percent of subjects in surveys of LEOP. In the Queensland cohort, 39 percent of subjects reported new breathing difficulties.⁷⁴

Respiratory difficulties are more likely to occur in individuals who required respiratory support during the acute disease, contracted polio when older than 10 years of age,⁹⁷ or have had polio over 35 years ago.⁹⁸

Signs and symptoms of respiratory dysfunction may include:⁹⁹

- Dyspnoea on exertion and/or at rest;
- Poor clearance of respiratory secretions;
- Fatigue and daytime sleepiness;
- Impaired intellectual function (including poor concentration);
- Morning headaches;
- Speech difficulties - quite speech (with few words per breath) or difficulty speaking for extended periods;
- Snoring; and
- Anxiety.

Loss of vital capacity is directly related to respiratory muscle weakness. Post-polio individuals who develop late onset ventilatory insufficiency have been observed to lose vital capacity at a rate of 60-90 percent greater than the normal population. This vital capacity loss occurs as the result of the combination of several factors including ageing, fatigue, accelerated loss of remaining anterior horn cell collaterals, pulmonary compliance and function.⁹⁶ Other factors such as scoliosis, pulmonary disease, cardiac disease, obesity and history of smoking may also contribute to the problem.

Individuals with expiratory muscle weakness can also have difficulty clearing secretions, especially during respiratory tract infections, due to a decrease in peak cough expiratory flows (PCEF). This can lead to further complications including ventilation/perfusion imbalance, pneumonias and pulmonary scarring.⁹⁶ Other factors which may impede expiratory muscle function and further decrease PCEF include airway obstruction from laryngeal muscle incompetence, aspiration of airway secretions and food due to bulbar involvement (paralysis of lingual, laryngeal and pharyngeal structures) and vocal cord paralysis or tracheal stenosis due to previous endotracheal intubation.¹⁰⁰

Dysphagia

Dysphagia refers to any disorder of swallowing. Figures from questionnaire based surveys of LEOP have shown 6⁷¹ to 22⁷⁰ percent of post-polio individuals reporting swallowing difficulties.

The presence of previous bulbar polio seems to predispose the individual to motor speech and swallowing deficits in later years. It has also been suggested that new swallowing symptoms may emerge as late effects of polio regardless of the original type of polio and whether or not the individual was aware of the symptoms of dysphagia.¹⁰¹ Laryngeal penetration and loss of the cough reflex may occur without symptoms. As a result the presence and severity of dysphagia in this population may be underestimated.

A study by Sonies and Dalakas (1991)¹⁰¹ using video fluoroscopy to evaluate post-polio subjects, revealed pharyngeal constrictor weakness, impaired tongue movements, pooling in the valleculae or pyriform sinuses and, rarely, aspiration. A further study by Silbergleit and colleagues (1990)¹⁰² found unilateral weakness of the tongue or palate in 80 percent of their subjects and laryngeal abnormalities in 57 percent.

Dysarthria

Dysarthria refers to a motor speech disorder caused by brain injury or nerve damage, which may result in paralysis, weakness, or incoordination of the muscles of the lips, tongue, soft palate, larynx and/or breathing mechanism.

Motor speech complaints in post-polio individuals include:¹⁰³

- Hypernasality;
- Intermittent aphonia;
- Reduced volume; and
- Hoarseness.

The major speech change reported by individuals with a history of polio is increased nasal resonance. This hypernasality is due to insufficient contact of the soft palate with the posterior pharyngeal wall, which allows air to escape into the nasopharynx during speech. The soft palate may be asymmetric or hemiparetic or the muscle of the hypopharynx may be weakened. Weakness of respiratory muscles and the effects of fatigue on other musculature often cause increased hoarseness, lowered pitch or volume, or loss of voice.¹⁰¹

Psychological Issues

Psychological symptoms of depression, anxiety and chronic stress have been observed in post-polio individuals. These symptoms are not only themselves causing distress but are also preventing these individuals from making lifestyle changes necessary to manage these symptoms.

It appears that a factor central to the aetiology of psychological symptoms is that polio survivors are being forced to cope with new symptoms and disability when many have not yet fully dealt with the emotional reality of having had polio years before. Polio survivors frequently report that the onset of post-polio symptoms has forced them, often for the first time, to recall and examine their acute polio experience.⁸³

Acute Polio Experience

With the onset of polio, individuals with polio realised that they had been stricken by the greatly feared disease. These fears were accentuated by the abrupt onset of the disease and by the realisation that the illness could lead to disability and possibly death.¹⁴ Many experienced long-term hospitalisation and isolation, resulting in separation from family members and friends. For many individuals, especially children, this separation was interpreted as abandonment. Due to often severe physical disabilities resulting from paralysis, many polio patients were totally dependent on hospital staff for all their basic needs.⁸³ Many patients endured sometimes painful medical (hot packs, splinting, bracing, exercise) and surgical procedures (muscle transplants, tendon lengthening and osteotomies).

On returning to the community following hospitalisation and intensive rehabilitation many individuals encountered ongoing challenges, not only from their physical disabilities but also from the attitudes of the community. For many children and adults, friends often disappeared. It was not uncommon for neighbourhood children to be prohibited from playing with “polio victims” for fear that the “crippling” disease was contagious. Due to their disabilities, many were no longer physically able to participate easily or fully in social activities and often became isolated.⁸³ Throughout childhood, adolescence and adulthood these individuals have continued to face their communities’ ignorance and intolerance with respect to disability.

Getting Normal

Patients were encouraged by health care professionals and family members to push themselves through feelings of pain, fatigue and weakness towards their goal of regaining function and mobility. Braces, crutches and wheelchairs were often considered proof that the individual had not worked hard enough to have fully recovered. Many children underwent multiple surgical procedures so that braces could be eliminated and they could look normal. Assistive devices were often quickly discarded on return to the community, regardless of the discomfort, fatigue or pain that often resulted.⁸³

Coping Styles

Many health care professionals who were involved with polio individuals have commented on the existence of a “polio personality”. It is unknown whether this was a function of social circumstances, the individual’s response to the disease, or whether it represented some kind of physiological predisposition to developing polio that is associated with certain behavioural characteristics.

Many of these individuals overcame a serious and often life-changing illness by developing coping strategies and behaviours. These behaviours included denial of their symptoms, independence, perseverance, obstinacy, detachment and creativity. Because they were successful once in overcoming their initial illnesses, these same behaviours tend to emerge later in life in coping with other challenges and illness.

People with polio tend to perform at high levels in many areas of life. It has been reported that they are employed full time at four times the rate of the general disabled population, they have more years of formal education on average than the general non-disabled population,¹⁰⁴ and they take on marriage and family responsibilities at approximately the same rate as persons who are not disabled.⁸² The post-polio population has been described as articulate, bright, successful in careers and life, independent, self sufficient and productive.

Post-polio individuals are often described in the literature as having Type A personalities (hard-driving over-achievers). In a questionnaire based study, Bruno and Frick (1987)⁸² found that the mean Type A score in post-polio survivors was significantly higher than that reported for a non-disabled

control population. They also reported that the polio group exhibited a high rate of symptoms associated with chronic stress which they felt may have initiated or exacerbated some of the new health problems.

There are a number of hypotheses why persons who had polio exhibit Type A behaviour and experience symptoms of chronic stress. It is possible that:

- Adults and even children who exhibited Type A behaviour and experienced stress were more susceptible to infection by polioviruses because of stress-induced immunosuppression; or
- In order to overcome the acute polio infection and manage in the general community, the special drive of the Type A personality was required.

Other researchers have used different models for categorising polio survivors. Maynard and Roller (1991)¹⁰⁵ proposed a model for categorising polio survivors according to the severity of muscular involvement. The model designates polio survivors as “Passers”, “Minimisers” or “Identifiers”. These labels are used to characterise the typical attitudes and behaviours that were adopted in order to cope with long-term mild, moderate or severe disability.

With this background information in mind, it is easy to understand how new physical symptoms can trigger reactive patterns of behaviour, attitudes and emotions in these individuals.

Response to the Late Effects of Polio

The new symptoms experienced by post-polio individuals may be psychologically devastating to them as they are often proud of having overcome a severe handicap. Although these individuals have often pushed themselves to the limit, this is no longer the appropriate response to the challenges that present to them in the form of the new symptoms and problems that accompany LEOP.¹⁰⁶

Their response to these new symptoms is often one of anger, fear and confusion. Individuals who experience the late effects of polio are unexpectedly faced with symptoms similar to those they experienced in their acute illness, problems that they thought they had permanently overcome. Fear is often experienced when the individual anticipates their physical condition and future capabilities – losing independence, income, physical abilities and roles. The unknown course of the disease makes it difficult to adjust as they do not know what they will be expected to adjust to. These individuals have attempted to be “normal” by successfully hiding their physical limitations over the years. Now they are experiencing new symptoms and often have to come to the realisation that they are not able to hide these limitations.

Many have sought assistance from medical professionals over the years without success. Due to the eradication of polio, few health professionals today have had training or experience in treating individuals with a history of polio. Many have limited understanding of this disease and the recurrence of symptoms.

Due to their experiences during the acute illness, many post-polio individuals fear hospitals and are wary of health professionals. As a result, faith in the medical profession has often been lost. Uncertainties of treatment and prognosis add to the stress and confusion that the post-polio individual experiences.¹⁰⁷

Previous treatment approaches used in the initial stages of the illness, i.e. ignoring pain and fatigue and to exercise as much as possible, are now being regarded as possible causes of the new symptoms. Patients are now being told to take it easy and not to exert themselves. This is a major lifestyle

change for many people and is often accompanied by resistance. Health professionals often find their patients avoiding treatment, minimising their condition and disregarding advice – in effect, not wanting to become less active and more dependent on others.¹⁰⁷

Depression

Conflicting findings have been reported on the prevalence of depression and other psychological distress among polio survivors. Frick (1985)¹⁰⁸ suggests that polio survivors may experience personal devaluation, isolation and depression as psychological responses to the onset of post-polio symptoms. A study by Conrady and colleagues (1989)¹⁰⁹ noted exceptionally high distress levels, particularly symptoms related to depression, somatisation and psychoticism in individuals with a history of polio. A study by Freidenberg and colleagues (1989)¹¹⁰ also suggests that mood disturbances are common among individuals with a history of polio with post-polio syndrome, although individuals with post-polio syndrome did not have greater levels of depression when compared to polio individuals without PPS.

Cameron (1989)¹¹¹ reported no evidence of depression in individuals diagnosed as experiencing PPS, but differences in regard to specific coping skills were observed, with patients engaging more frequently in denial as a positive coping skill. These results were further supported through a study by Tate and colleagues (1993)¹¹² who reported no major distress, depression or elevated somatic complaints when compared to the normal population, although higher levels of depression were associated with abnormally elevated levels of physical symptoms (pain).

Impact on Family

Disability is a family affair.¹¹³ New or worsening symptoms can have an impact on all members of the family, the post-polio individual, partners, children and parents. Old feelings in regard to the acute polio experience may be reawakened. Changes in the roles and responsibilities of family members may be required to assist the post-polio individual in managing some of the symptoms they are now experiencing. Increased stress experienced by the post-polio individual may have a pronounced effect on other members of the family. Intimate relationships between partners may also be affected due to the symptoms of pain, fatigue and weakness. These symptoms can also affect the individual's self-image and their sexuality, having a profound affect on relationships.

Assessment

The assessment of an individual with a history of polio who is experiencing new health problems, presents a challenge to the health professional. This is due to the number, complexity and diversity of symptoms, the absence of special diagnostic tests, the uncertainty of the underlying cause and the lack of curative medications or treatments. In this section, the diagnosis of PPS, including differential diagnosis of the primary symptoms will be reviewed.

Diagnosis of Post-Polio Syndrome

The diagnosis of PPS is one of exclusion and is clinically based. There are no serologic, enzymatic, electrodiagnostic or muscle biopsy tests that can diagnose PPS. Therefore, it is essential that each patient receives a careful history and physical examination along with appropriate laboratory, radiological and diagnostic studies to rule out other medical, neurological or orthopaedic conditions that may be producing or aggravating the symptoms the patient may be experiencing.

Halstead (1991)⁸⁶ has developed the following criteria for the diagnosis of PPS:

1. A prior episode of paralytic polio with residual motor neuron loss confirmed by history, physical examination, and typical findings on electromyography (EMG).

The diagnosis of paralytic polio usually can be confirmed by:

- Eliciting a credible history of an acute, febrile illness resulting in motor loss and not sensory deficit. Memory is often not a reliable tool when determining the location and extent of the original paralysis. Although most patients appear to remember the location of severe weakness, they may not have any recollection of extremities that incurred only mild weakness;¹¹⁴
- Noting whether other members of the patient's family or neighbours had a similar illness;
- Observing the presence of focal, asymmetric weakness and/or atrophy on examination;
- Examining whenever possible the original medical records; and
- Changes on EMG of chronic denervation with reinnervation compatible with prior anterior horn cell disease.

Non-Paralytic Polio

There is significant debate in the literature whether people with a history of non-paralytic polio (NPP) are at risk of the development of PPS. Several studies have shown that individuals with non-paralytic polio do have late onset symptoms.^{47,115} Falconer and Bollenbach (2000)¹³ have suggested a number of reasons why people with a history of NPP should not be automatically excluded from a diagnosis of PPS. These include:

- The person had non-paralytic (or abortive) polio. At the time of the acute illness there was no obvious damage to the nervous system although unobserved damage was likely. There are no established figures for the minimum amount of neuronal damage which can result in PPS symptoms. Several studies have shown that neuronal damage occurred in all non-paralytic cases of polio. For neuronal damage to be visible, at least 50-60 percent of the motor neurons must be damaged or destroyed.¹¹⁶ When fewer motor neurons are involved the patient will present with no specific muscle weakness, although neuronal damage can be present.
- The person had paralytic polio during their initial illness, but was misdiagnosed. Symptoms of paralysis and/or weakness may have been missed or the symptoms may have manifested for a short period of time. The patient recovered (apparently) fully within a matter of weeks. This type of polio often was labelled as "non-paralytic". PPS will occur in these individuals with the same frequency as in paralytic polio cases. Diagnosis of PPS on the same basis as for a patient with a history of paralytic polio is merited.
- The person did not have polio but had another disease with clinical symptoms similar or identical to polio and currently presents with PPS symptoms. Some evidence supports the hypothesis that non-polio enteroviruses can have late, post-viral effects.
- The person may have had undiagnosed polio. PPS should not be excluded and further tests may be required.

Halstead and Silver (2000)¹⁷ have suggested that the diagnostic criteria for PPS syndrome should be modified to include, “a history of remote paralytic polio or findings on history, physical examination results and laboratory studies compatible with poliovirus damage of the central nervous system earlier in life”.

2. Standard EMG evaluation demonstrates changes consistent with prior anterior horn cell disease.

The changes on conventional EMG suggestive of prior polio include:

- Increased amplitude and duration of motor unit action potentials
- Increase in the percentage of long duration polyphasic motor units
- Decrease in the number of motor units on maximum recruitment in weak muscle
- Occasionally fibrillations are present and less commonly fasciculation may be observed^{5,86}

3. Characteristic pattern of recovery – A period of neurological recovery followed by an extended interval of neurological and functional stability preceding the onset of new weakness; the interval of neurological and functional stability usually lasts 20 years or more.

The general pattern of recovery consists of (Figure 4):

- Paralytic polio in childhood or later in life
- Partial to fairly complete neurological and functional recovery
- A period of functional and neurological stability lasting many years
- Onset of new health problems

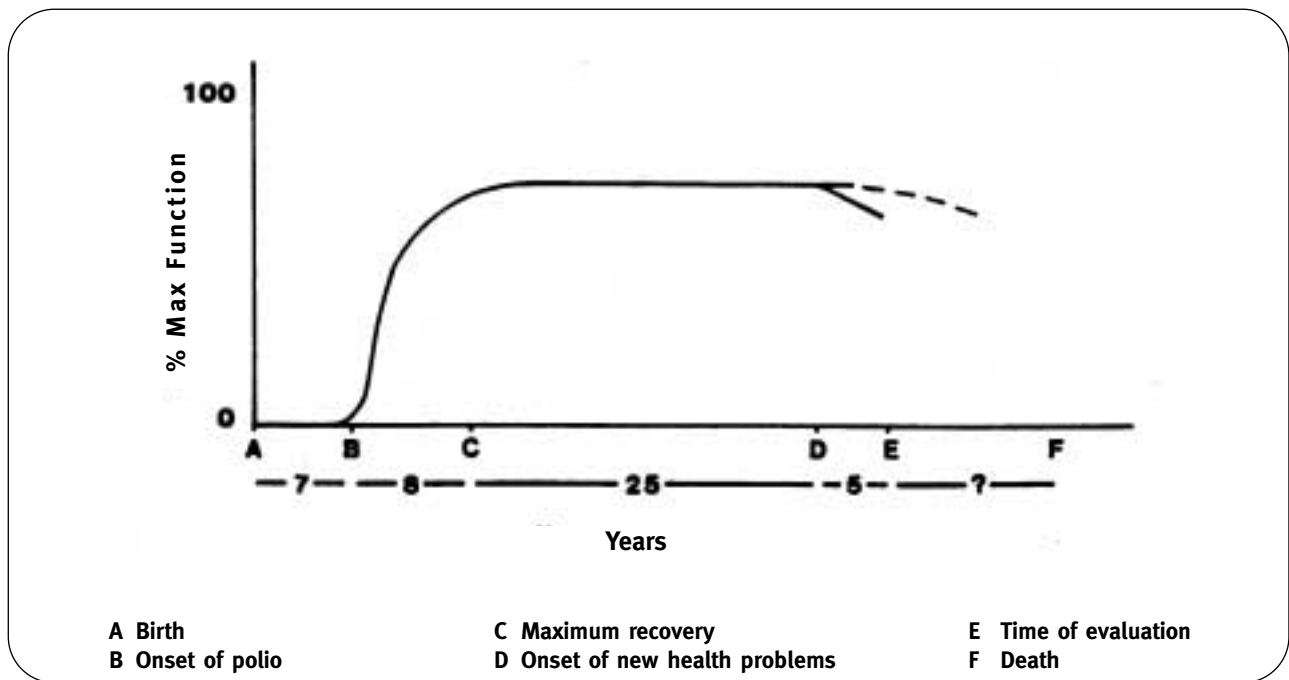


Figure 4: Diagrammatic Representation of the Recovery Stages of Polio⁹
(Natural history data from Post-Polio Clinic in Houston, Texas. Years shown are median values for the clinic group)

This pattern of recovery is very characteristic of paralytic polio. When this pattern of recovery is not reported, the diagnosis of paralytic polio should be questioned.⁸

4. The gradual or abrupt onset of new neurogenic, non-disuse weakness in previously affected and/or unaffected muscles; this may or may not be accompanied by other new health problems such as excessive fatigue, muscle pain, joint pain, decreased endurance, decreased function and atrophy.

New neurogenic weakness is considered essential in the diagnosis of PPS, as it reflects new or continuing dysfunction of motor units injured during the initial illness. It is inferred by the onset of diminished function despite maintaining the usual level and intensity of activity.⁵

5. Exclusion of medical, orthopaedic, and/or neurological conditions that may cause the health problems listed above (Criteria 4) – i.e. Differential diagnosis.

Differential Diagnosis

One of the challenges facing health professionals attempting to evaluate and manage post-polio individuals with new health problems is the general nature of many of the symptoms.¹¹⁸ The differential diagnosis of PPS is complex, as there are many conditions that could be responsible for the primary symptoms of fatigue, weakness and pain that a patient may experience.

In developing a differential diagnosis, the Post-Polio Task Force (1999)¹¹⁹ suggest defining each presenting complex (i.e. fatigue, weakness and pain), in terms of:

- Characteristics;
- Onset/duration;
- Location; and
- Activities that increase or decrease symptoms.

Fatigue

Fatigue is often a non-specific complaint with a variety of possible aetiologies. As a result, before a diagnosis of PPS fatigue is made, it is necessary to exclude other conditions that can cause this symptom.

Some of the conditions that need to be considered in the differential diagnosis of fatigue include:¹¹⁹

Systemic metabolic disease:

- Hypothyroidism
- Cancer / chemotherapy
- Anaemia
- Cardiac conditions – coronary heart disease, heart failure
- Diabetes mellitus
- Chronic infection
- Renal disease
- Lupus
- Thyroid disease
- Hepatic disease

Ventilatory dysfunction:

- Sleep apnoea
- Chronic alveolar hypoventilation
- Hypoxaemia

Mood disorders:

- Depression
- Anxiety / stress

For any patient presenting with potential PPS, it is important that the characteristics, time of fatigue and the activities that produce fatigue are recorded. This information assists in determining the cause of the problem. Fatigue that is reported upon awakening usually reflects sleep disturbances that may be caused by various factors (Refer to section on sleep disturbances). Fatigue that lasts throughout the day is not typical of PPS and may indicate other conditions, e.g. chronic fatigue syndrome.

Other evaluation tests that should be considered when diagnosing the cause of fatigue include:

- Physical examination of the individual;
- Blood chemistries – complete blood count;
- Arterial blood gas;
- Thyroid function;
- Chest X-ray;
- Electrocardiogram;
- Pulmonary function test;
- Overnight oximetry and sleep studies; and
- Psychological assessment.

Weakness

Weakness is the cardinal sign of motor neuron dysfunction in PPS. Therefore, the exclusion of other causes of new weakness is essential. For many of the conditions that result in weakness, specific assessment procedures and tests are available and accurate diagnosis can usually be made. While determining the cause of weakness, it is important to remember that these conditions can occur in addition to PPS.

Some of the conditions that need to be considered in the differential diagnosis of weakness include:¹¹⁹

Superimposed neurological conditions:

- Inflammatory demyelinating disease
- Late onset genetic dystrophies
- Adult spinal muscular atrophy
- Inflammatory myopathies
- Amyotrophic lateral sclerosis
- Cerebrovascular disease
- Multiple sclerosis
- Parkinson's disease
- Spinal stenosis
- Myasthenia gravis
- Radiculopathies
- Neuropathies
- Entrapment neuropathies
- Cauda equina syndromes
- Spinal cord tumours and infarctions
- Diabetic amyotrophy

Overuse or chronic strain:

- Joint / spine deformities
- Weight gain
- Lifestyle activity patterns

Systemic comorbid medical conditions:

- Thyroid disease
- Uraemia
- Toxins

Disuse atrophy:

- Injury
- Weight gain
- Surgery

An individual with neurogenic weakness caused by PPS often experiences a pattern of diminished strength, endurance and function at a time when there have been no obvious changes in their usual level or intensity of activity. By contrast, individuals with disuse weakness often describe a clear change in the usual pace and intensity of their activity levels or in the way that individual muscles are used. This may occur due to a period of immobility due to hospitalisation, pain or illness. If disuse weakness is expected, a trial of monitored exercise should be prescribed to determine if this new weakness can be reversed.¹¹⁸

It is also important to differentiate between muscle weakness (loss of strength and endurance) and other synonymous terms the patient may be referring to, such as fatigue and lack of energy.⁵

Recommended procedures for the assessment of muscle weakness include:

- Physical examination of the individual – manual muscle assessment, range of motion, trials of strengthening exercise (as prescribed by a Physiotherapist);
- History – current and past muscle function and weakness;
- Analysis of current performance compared to performance in past;
- Blood chemistries – complete blood count, creatine kinase analysis;
- Thyroid function;
- Toxic metal screens;
- EMG / nerve conduction studies; and
- Specific neurological and medical assessments to assist in the diagnosis of other conditions which result in muscle weakness.

Pain

The number of disorders that produce pain is extensive. Assessment should commence with the identification of other conditions that could be producing pain which are commonly associated with chronic musculoskeletal wear and tear and disorders that have significant muscle and/or joint manifestations. The major areas of consideration include spinal orthopaedic conditions, fibromyalgia, muscle pain, musculotendinous conditions and limb joint conditions. Some of these conditions include:¹¹⁹

Spinal orthopaedic conditions:

- Myofascial pain syndromes
- Degenerative disc disease
- Radiculopathies
- Spondylolisthesis
- Spinal stenosis
- Scoliosis
- Chronic strain associated with leg length and gait abnormalities

Muscle pain:

- Overuse strain myalgia

Limb joint conditions:

- Internal derangements
 - Ligamentous laxity/hypermobility
 - Strain syndrome
- Degenerative arthritis
- Traumatic arthritis

Musculotendinous conditions:

- Tendonitis
- Bursitis
- Entrapment syndrome
- Repetitive strain/overuse

Fibromyalgia

Assessment of the Late Effects of Polio

Due to the complexity and nature of many of these patients' problems, it is at the health professional's discretion to select the most appropriate method of evaluation. Currently there is no definitive diagnostic test for PPS. Investigations should be problem orientated and exclude other potential causes for the patient's problems.

The most appropriate method of providing a comprehensive and coordinated evaluation that addresses the client's medical, functional, psychosocial and vocational issues is through the use of an interdisciplinary team, including physician, physiotherapist, occupational therapist and social worker. Referral to other health professionals including speech pathologists, orthotists, podiatrists, psychologists, respiratory physicians, orthopaedic specialists and neurologists is often required.

Medical Assessment

In regard to the medical assessment of the post-polio patient, Dr Pesi Katrak has written the following:

Symptoms such as fatigue and pain can be caused by a very large number of medical conditions. It is neither practicable nor cost effective to carry out investigations for all of the conditions that can cause these symptoms. One has to be guided by pointers derived from a thorough clinical assessment including a detailed history and a complete physical examination. Some clinics in North America recommend routine blood test in all patients including a complete blood count, serum electrolytes, glucose, urea, creatinine, calcium, phosphates, total protein, protein electrophoresis, albumin, magnesium, AST, ALT, LDH alkaline phosphatase, creatine kinase and thyroid function tests.

My practice is to order blood tests and other investigations only when there is some clinical indication pointing to a particular disorder which needs to be excluded. Some of the common disorders found in older adults, such as diabetes, anaemia, cardiac dysfunction, depression, sleep apnoea and fibromyalgia, should be looked for in the clinical assessment and, if necessary, excluded with additional laboratory tests or appropriate referrals. Other neurological disorders causing muscle weakness, such as spinal muscular atrophy, amyotrophic lateral sclerosis, spinal canal stenosis, diabetic/other neuropathies, entrapment neuropathy and radiculopathy, can generally be excluded on clinical assessment. Electromyography (EMG) and nerve conduction studies may sometimes be required for this purpose. Sometimes electromyography may be required to confirm a diagnosis of prior polio if the residual clinical features are unclear. EMG however does not help to differentiate between patients with new symptoms from post-polio syndrome and asymptomatic polio survivors, as similar changes of chronic denervation are seen in both groups. Sleep apnoea, which is being increasingly recognised in the general population, can lead to daytime sleepiness and fatigue, and is very similar to that seen in patients with post-polio syndrome. If the patient has a history of an irregular breathing pattern at night and tiredness or headaches on waking in the morning, I often recommend referral to a respiratory physician for consideration of sleep apnoea. Treatment of this disorder can result in significant improvement in daytime tiredness.

Management

Several reviews on the current management of LEOP and PPS have been published.^{5,118,120,121} These authors agree that many post-polio individuals can benefit from a multidisciplinary, individualised management program. This section will review the current approach to management of LEOP described in the literature. A summary of the management strategies that have been described in a number of articles that have been written on the management of this condition and several research studies that have recently emerged are outlined in Table 4

All patients with a history of polio can benefit from a thorough medical assessment and at least some of the management strategies that are outlined in this section. Although the individual may not be exhibiting late onset problems, an understanding of the disease process, a balance of activity and rest, optimal orthotic fitting and weight control can be beneficial in reducing the possible development of future problems.

Table 4: Management Strategies for the Late Effects of Polio

MANAGEMENT OF WEAKNESS

- Strengthening exercise (isometric, isotonic, isokinetic)
- Aerobic exercise
- Stretching exercise
- Avoidance of muscular overuse
- Energy conservation – pacing, rest, activity reduction
- Weight loss
- Orthoses and assistive devices

MANAGEMENT OF FATIGUE

- Energy conservation – pacing, resting, activity reduction
- Lifestyle changes
- Weight loss
- Prescription of aids
- Aerobic exercise
- Medications

MANAGEMENT OF PAIN

Post-polio muscular pain

- Stretching exercise
- Moist heat, ice and stretching
- Orthoses
- Assistive devices
- Energy conservation – pacing, activity reduction
- Medications

Overuse pain

- Medications
- Moist heat, ice, ultrasound
- Energy conservation
- Assistive devices
- Orthoses

Biomechanical pain

- Postural correction
- Strengthening exercise
- Stretching exercise
- Orthoses
- Assistive devices
- Biofeedback and muscle relaxation
- Non-steroidal anti-inflammatory medications
- Steroid injections
- Surgery

Treatment of other superimposed neurologic disorders

Includes carpal tunnel syndrome, radiculopathy, spinal stenosis

- Surgery
- Assistive devices
- Orthoses

Fibromyalgia

- Posture correction
- Medication – amitriptyline, cyclobenzaprine
- Aerobic exercise

MANAGEMENT OF RESPIRATORY DYSFUNCTION

- Preventive measures (pneumococcal vaccine, influenza vaccine)
- Ventilatory assistance
- Identification and treatment of sleep disordered breathing
- Training in glossopharyngeal breathing, assisted coughing
- Stretching of tight accessory respiratory muscles and muscle relaxation
- Postural correction – management of scoliosis
- Aerobic exercise
- Weight loss

MANAGEMENT OF DYSPHAGIA

- Referral for speech pathology assessment of swallowing
- Modification to food and fluid consistencies
- Trial and implementation of compensatory/facilitatory swallowing techniques
- Monitor fatigue

MANAGEMENT OF DYSARTHRIA

- Assessment of oromusculature and motor speech function
- Counselling
- Exercise

MANAGEMENT OF COLD INTOLERANCE

- Multiple layers of clothing
- Massage
- Localised heat

MANAGEMENT OF PSYCHOSOCIAL DIFFICULTIES

- Interdisciplinary approach
- Counselling, education, support
- Post-polio support group
- Evaluation and treatment by social workers, psychologists and psychiatrists

(Compiled from Thorsteinsson, 1997¹²⁰; Halstead, 1998¹¹⁸ and Trojan and Finch, 1997¹²¹)

Management of Weakness

New or increased muscle weakness in PPS is caused by overwork, disuse or a combination of both of these factors. It is essential that the underlying cause of muscle weakness is determined so that appropriate management strategies can be developed. The management of new weakness may include:

- Strengthening exercise (isometric, isotonic, isokinetic);
- Aerobic exercise;
- Stretching exercises to decrease or prevent contractures;
- Education regarding energy conservation techniques including pacing, rest, activity reduction and the avoidance of muscular overuse;
- Weight loss; and
- Prescription of orthoses and assistive devices.

If muscle weakness is secondary to disuse, low intensity non-fatiguing strengthening exercise or low-intensity aerobic exercise may be of benefit. Details regarding the use of exercise for post-polio individuals is outlined in the following section on exercise.

In several studies, muscular overuse in subjects with a history of paralytic polio has been thought to be the cause of increased weakness. If weakness is secondary to chronic overwork and fatigue, muscles require rest, and further overuse of muscle must be avoided. Patients should be encouraged to rest and pace activities (refer to following section on lifestyle modification). Orthoses may also be used to assist in resting isolated, weakened muscles (refer to following section on orthoses and mobility aids). Strenuous activities producing a decline in muscle strength should be avoided. However, muscles that have been affected by polio readily atrophy with disuse. It is, therefore, essential that strength changes are monitored closely following the prescription of rest, reduced activity and orthoses.

Management of Fatigue

Post-polio fatigue can be managed with the use of:

- Education regarding energy conservation techniques and lifestyle changes;
- Weight loss;
- Prescription of orthoses and assistive devices;
- Aerobic exercise; and
- Pharmacological treatment.

The most important aspect of the treatment of fatigue is educating the patient regarding energy conservation techniques. Teaching the patient the concepts of pacing activities, rest and work simplification can assist the patient in conserving their cardiopulmonary endurance and reduce the symptoms of fatigue (refer to following section on lifestyle modification). If the patient fails to adopt these basic management principles other measures, e.g. aerobic exercise, will be of little benefit.

Although some patients may be deconditioned, the presence of unrelenting fatigue may preclude the prescription of a modified aerobic training program (refer to following section on the role of exercise). In this case, the prescription of rest may have greater justification. If fatigue cannot be managed through activity reduction and rest, significant lifestyle changes may be required, e.g. changes to the work situation and home management responsibilities.

Muscle fatigue, associated with overwork of specific muscle groups, often responds well to localised rest and the use of orthoses to provide protection to the involved muscles and joints. As a result, the prescription or modification of orthoses and walking aids may be required (refer to following section on orthotics and mobility aids). Recent changes to the construction of orthoses have replaced heavier materials that were formerly used in fabrication of braces with more lightweight materials. These lighter materials significantly reduce the workload required of the individual.

Medications that have been trialed for the management of fatigue in the post-polio population include pyridostigmine, amitriptyline, selegiline and bromocriptine. (For further information regarding these trials, refer to the following section on pharmacological therapies)

Management of Pain

Pain in post-polio individuals can result from a variety of causes. Management of pain should therefore depend upon the aetiology. Pain management is based on a number of principles, which can be supplemented by specific recommendations. These basic principles include:¹¹⁸

- Improve abnormal body mechanics, such as poor posture and gait deviations;
- Relieve or support weakened muscles and joints through bracing and the use of ambulatory aids; and
- Promote lifestyle modifications that conserve energy and reduce stress.

Treatment for post-polio muscular pain may include:

- Application of therapeutic modalities including moist heat and ice;
- Stretching – to maintain extensibility of muscle and connective tissue;
- Prescription of orthoses and assistive devices;
- Education regarding lifestyle modifications, activity reduction and pacing (refer to following section on lifestyle modification); and
- Pharmacological treatment.

Treatment of overuse and biomechanical pain may include:

- Physiotherapy:
 - Electrotherapy (e.g. TENS, ultrasound)
 - Application of therapeutic modalities including heat, ice
 - Strengthening exercise
 - Joint mobilisation
 - Muscle relaxation and biofeedback
 - Stretching of tendons and soft tissues, myofascial release techniques
 - Hydrotherapy;
- Modification of extremity use, e.g. periodic rest;
- Retraining of posture and body mechanics to minimise overuse and reduce or avoid pain during activities of daily living and work;
- Pacing activities;
- Bracing to alleviate symptoms and prevent further acceleration of joint and muscle overuse, to control joint deformities and failing joint fusions (refer to following section on orthoses/bracing);
- Prescription of mobility devices (refer to following section on mobility devices);
- Pharmacological treatment – Non-steroidal anti-inflammatory medication;
- Surgery; and
- Treatments for fibromyalgia – postural correction, medication (amitriptyline, cyclobenzaprine), aerobic exercise.

If the patient experiences pain secondary to an acute injury, treatment with traditional physiotherapy modalities is helpful.

Pain can often be reduced by changing to a lifestyle that reduces physical activity and altering the biomechanics used to perform certain movements. These strategies may be difficult to accomplish because they often require the patient to develop behaviours unlike the old, familiar ones. Altering the pace and intensity of activities and learning new ways to gain more control over when and how activities are performed is essential. Unless these strategies are incorporated into the individual's life, further treatment options may become non-beneficial.

Postural Correction

Abnormal postural alignment in patients with a history of paralytic polio can frequently be attributed to muscle imbalance. Postural deformity can lead to pain and decreased energy efficiency during various activities. Addressing postural deformities can often be helpful in minimising or preventing pain and increasing endurance for sitting, standing, walking and other activities.

Management of postural deformities can be achieved through:

- Strengthening and stretching exercises;
- Education about back care and correct use of body mechanics;
- Referral to a comprehensive back care program;
- Education about appropriate posture and resting positions;
- Prescription of adaptive aids, e.g. lumbar rolls;
- Prescription or modification of orthoses/braces;
- Prescription of mobility aids; and
- Weight management – cardiovascular exercise and referral to a dietician (refer to following section on weight management).

Orthoses / Bracing

People with a history of polio often have strong and usually negative feelings about the use of orthoses. Therefore, prescription or modification of such a device becomes a challenge for the health professional. Many individuals disordered their orthoses and have relied on compensatory movements for walking. For those in which the use of an orthosis for walking was essential, the device became a part of their body image, and the patient is often reluctant to change. As a result, these individuals may continue to use the style of orthosis originally prescribed at the time of their initial illness. Although orthoses have changed considerably over the years and are now stronger and lighter, many post-polio individuals have continued to wear older style orthoses. With many of these people reporting increased pain and difficulties with ambulation, orthoses may need to be reviewed to assess their current fit and appropriateness. Individuals who may not have used orthoses in the past or those who may have disordered their orthoses may need to be reviewed for possible prescription of such a device.

There are a number of reasons why a patient may be prescribed an orthoses. These include:

- Reduce the energy requirement of walking by compensating for muscle weakness;
- Assist with balance;
- Improve positioning and stability of a joint to reduce pain;
- Equalise weight distribution – restoring weight bearing on the weaker leg and decreasing the work load of the stronger leg; and
- Reduce risk of falls and potential fractures.

Common biomechanical deficits requiring orthotic management include:

- Inadequate dorsiflexion in swing – secondary to weakness of ankle dorsiflexors. May be treated with an ankle foot orthosis (AFO);
- Dorsiflexion collapse in stance;
- Genu recurvatum (knee hyperextension) – usually caused by weakness of the quadriceps. Person often locks the knee when mobilising to improve stability of the lower extremity. Often can be managed with an AFO, knee orthosis or in patients with more severe weakness, a knee ankle foot orthosis (KAFO) may be required;
- Genu valgum (valgus deformity of the knee); and
- Mediolateral ankle instability.

Referral to an orthotist with a thorough understanding of this patient group is desirable to ensure that an appropriate, well fitting orthosis is prescribed.

Mobility Devices

For many patients with a history of polio, the extended use of crutches, the use of manual wheelchairs for many years and asymmetrical or abnormal gait patterns are the major source of weakness, pain and fatigue. Although the patient may be experiencing extreme difficulties with their mobility, changes or modifications to their current method of locomotion may be difficult for them to consider. Rather than use a mobility aid, many individuals make significant lifestyle changes to compensate for their decline in mobility, such as limiting their social and outdoor activities.

Prevention of further disability and restoration of lost function often require a decrease in the amount of walking or wheelchair propulsion and a change to methods that do not result in weakness, pain and fatigue. It is important that a thorough evaluation of the patient is conducted by a physiotherapist or occupational therapist prior to the prescription of any mobility device. Prescription of mobility aids should be based on the patient's functional status, their needs and the potential for progressive deterioration.

The objectives of prescribing a mobility aid are to:

- Ensure more stable balance and reduce risk of falls;
- Minimise fatigue and energy required for ambulation;
- Promote improved posture; and
- Alleviate load on joints.

When considering the appropriateness of a mobility aid for a post-polio patient, the following factors need to be considered:

- Balance;
- Strength and function of upper and lower limbs;
- Cardiovascular endurance and fatigue levels;
- Current or future use of orthotic devices;
- Compliance; and
- Environment.

Shoulder, elbow and wrist injuries are common in post-polio individuals who have relied on crutches or wheelchairs for ambulation. As a result, a thorough assessment of the client's upper limb function is required before the prescription of a mobility device.

Management of Respiratory Dysfunction

The conditions under which the patient experiences respiratory difficulties must be identified before development of a management program. Often the management of individuals with respiratory complications requires referral to a respiratory physician for comprehensive evaluation and management.

The following recommendations are a brief overview of some of the important factors regarding the management of respiratory dysfunction in these patients. It is essential that patients who are experiencing respiratory problems are referred for a comprehensive assessment and treatment program that a respiratory physician can provide.

General management strategies include:

- Preventive measures (pneumococcal vaccine, influenza vaccine);
- Ventilatory assistance – non invasive methods preferred;
- Identification and treatment of sleep disordered breathing;
- Training in breathing techniques and assisted coughing;
- Inspiratory muscle training;
- Stretching of tight accessory respiratory muscles and muscle relaxation;
- Postural correction – management of scoliosis;
- Aerobic exercise; and
- Weight loss.

Management of Sleep Disturbances

Sleep disturbance may be caused by pain, stress, underventilation or obstructive apnoea. Due to the complex nature of sleep disturbances, the cause needs to be identified before the development of management strategies. As a result, referral for sleep studies may be required. In addition to addressing respiratory dysfunction that may be responsible for these problems (as discussed above), modifying the patient's sleeping position and the number of pillows used, may provide some relief. Pain that the patient experiences at night may also need to be addressed.

Management of Dysphagia

Dysphagia management should be undertaken by a speech pathologist who has expertise in the assessment, diagnosis and treatment of swallowing disorders.

Speech pathology intervention of swallowing disorders may include:

- Clinical and/or radiographic assessment of swallowing;
- Provision of counselling regarding observed swallowing problems associated with PPS and strategies to facilitate safe swallowing;
- Modifying the consistency of food and/or fluids;
- Trial of compensatory/facilitatory swallowing techniques; and
- Design of exercise programs to improve range, strength, speed and coordination of muscles used in swallowing.

Management of Dysarthria

As in the case of dysphagia, patients with motor speech difficulties should be referred to a speech pathologist for management of their condition. Speech pathology management of dysarthria may include:

- Assessment of the oromusculature (e.g. lips, tongue, soft palate) and motor speech function including the parameters of respiration, phonation, resonance, articulation, prosody and speech intelligibility;
- Provision of counselling regarding identified motor speech problems associated with PPS and strategies to promote speech intelligibility. For example, using shorter sentences, reducing rate of speech, or avoiding talking over background noise; and
- Provision of exercises to improve hypernasality, voice quality and volume of voice.

Management of Cold Intolerance

The management of cold intolerance is largely symptomatic and may include:

- Multiple layers of clothing – especially when placed on the affected extremities first and then on the rest of the body;
- Massage (toward the heart); and
- Short term use of local heat (20 minutes or less) – special care must be taken when using heat by any person with diminished sensation or peripheral vascular disease.

Weight Management

Weight reduction is an important and effective way of reducing the workload on muscles. For patients experiencing pain, fatigue and muscle weakness, even relatively small increases in weight can have a significant effect. For many people with a history of polio, weight loss is a challenge due to the difficulties that they experience when exercising. Body weight norms that are used for the general population are often not appropriate for those who have had polio and have poor musculoskeletal development. The patient should ideally be in the low to middle end of the body mass index range, depending on their degree of impaired growth and development.¹²² Dietetic counselling and support is essential in ensuring that weight control is incorporated as a permanent modification to the patient's lifestyle, rather than being regarded as a short term plan.

Pharmacological Therapies

In regard to the use of pharmacological therapies in the treatment of PPS, Dr Pesí Katrak has written the following:

Anecdotally, a number of pharmacological agents have been reported to show improvement in symptoms of post-polio syndrome. All of these are case reports on a small number of patients only. There are very few randomised, controlled trials in PPS patients. The largest study was a randomised, multi-centre trial on 126 PPS patients¹²³ on the use of pyridostigmine. This was a placebo-controlled trial, which failed to show that pyridostigmine¹²⁴ produced an improvement on measures of fatigue and strength. An earlier, open trial with pyridostigmine on a small number of patients had shown improvement in fatigue and strength in patients receiving this medication. Other randomised placebo-controlled trials using Amantadin¹²⁵ on 25 patients and Prednisone¹²⁶ on 17 patients did not show a significant improvement in fatigue or strength. Small case studies with two to five patients have been reported using Selegiline,¹²⁷ Bromocriptine¹²⁸ and human growth hormone.¹²⁹ It is not possible to draw any conclusions from these case reports. Overall, there is no evidence to indicate that any pharmacological agent improves the symptoms of weakness or fatigue in post-polio syndrome patients.

Psychological Management

To overcome the combination of denial and personal history of successful coping that is often found in polio survivors, an interdisciplinary approach to psychological problems is helpful. Each member of the team brings with them specific skills and knowledge that could assist the patient address some of the issues that have resulted from the new symptoms that the patient is experiencing.

The main aims of treatment of the post-polio patient should be to:

- Increase and expand the patient's personal and external resources;
- Provide education and support; and
- Reinforce the need for the patient to have control over their lives.

Several strategies have been successfully utilised by clinicians to assist patients enhance their coping skills. These strategies have included:

- Assisting the patient in identifying their current and previous coping strategies.
- Encouraging the patient to make necessary lifestyle changes including relaxation, stress management, energy conservation and work simplification.
- Encouraging the patient to focus on past achievements and identify personal strengths rather than focusing on the new weaknesses.
- Encouraging the patient to keep the lines of communication open though:
 - Sharing literature
 - Talking with others; and
 - Attending workshops and relevant information sessions (when available)

This can be achieved through referring the patient to a local post-polio support group and by providing the patient with adequate information and resources.

- Encouraging the patient to be an active participant in managing their own needs through:
 - Setting limits
 - Reminding others of their needs; and
 - Seeking support and information.
- Encouraging the patient to be aware of their own feelings and the feelings of others (including family members) by:
 - Showing appreciation for the contribution of others; and
 - Seeking help before difficulties evolve.
- Encouraging the patient to recognise their feelings of loss and grief and provide support to them and their families through this process.
- Referring to other health professionals, including psychologists, psychiatrists and social workers, to address unresolved issues.

Lifestyle Modification

Because the aetiology of PPS is believed to be related to overwork of polio damaged nerves and muscles, it is often necessary to advise post-polio patients to make lifestyle changes to reduce the symptoms of pain, fatigue and weakness and prevent further decline in function. These lifestyle changes often present a challenge not only to the patient but also to the health professional recommending these changes.

Occupational therapists can assist the patient in making the necessary changes to their lifestyle by encouraging them to evaluate their lives, make the identified changes and to guide the individual to reframe their expectations for success by focusing on innovative ways of achieving new goals.

By educating the patient on specific energy conservation, joint protection and work simplification techniques, it is hoped that the patient develops the necessary skills required to modify or pace any activity or behaviour. These skills include the ability to:

- Identify activity related pain, fatigue and weakness and the activity that caused it;
- Analyse activities according to the potential energy use;
- Analyse body mechanics during daily activities and identify those aspects that need to be changed;
- Recognise and correct poor joint protection behaviours; and
- Analyse their posture in various situations and identify aspects that need to be changed.

To better equip the patient and their family members to make any necessary changes to their daily life, it is essential that they have a thorough understanding of the:

- Disease process;
- Reasons why they are experiencing these symptoms of fatigue, pain and muscle weakness; and
- Advantages of incorporating energy conservation techniques into their lifestyle.

The patient should be supported through this process and assured that although some of these changes may be difficult, they are necessary, and benefits will be seen. Family members should be included in all aspects of this process so that they may develop a better understanding of the new problems facing these individuals.

The lifestyle changes that may need to be incorporated into the patient's management plan and daily life include:

- Energy conservation techniques;
 - Activity analysis
 - Pacing / scheduling
 - Rest
- Joint protection; and
- Work simplification.

Exercise

Considerable controversy surrounds the prescription of exercise for post-polio patients because the pathophysiology of this condition remains unclear and because there have been case reports of overwork weakness amongst this population. It has been shown that individuals with a history of polio can increase their muscle strength and cardiovascular endurance by following a well structured, individualised training program.

Exercise prescription for the post-polio patient should only be carried out by a physiotherapist who has a thorough knowledge of the general principles of exercise prescription and an understanding of the effects of exercise on this group of patients. Even though several different forms of exercise have been shown to be beneficial in this population, exercise should be used judiciously and should be completely avoided in some patients.

Exercise must be individualised because:

- Each person has a varying degree of weakness and musculature affected;
- Exercise must be modified according to pain, fatigue, exercise tolerance and posture;
- Medical history and any previous surgical procedures must be taken into account; and
- Every person has his or her own needs, goals and interests.

Who should not be exercising?

People who push themselves beyond the point of fatigue with their daily tasks should probably not be prescribed an exercise program. These people may have weakness due to overuse of their muscles. They may notice an improvement in their strength and fatigue levels in they reduce their activity. These patients should be taught the principles of energy conservation and be assisted to modify their lifestyle to reduce fatigue and muscular overuse. Specific muscle groups that are being overused may benefit from rest or supportive devices. If these measures are effective, the person may then become a candidate for an exercise program.

Who should be exercising?

Post-polio patients who should be prescribed an exercise program include:

- Those who keep their fatigue under control by practising energy conservation and pacing themselves;
- Those who lead more sedentary lives where fatigue is no longer a problem; and
- People who are overweight.

The types of exercise that may be prescribed to a post-polio patients will be briefly explained.

Cardiovascular Exercise

The aims of aerobic exercise include:

- Improved heart function and efficiency – the average level of aerobic fitness in post-polio patients is 5 METS – similar to the fitness of patients following a myocardial infarction;
- Improved circulation;
- Improved respiratory function;
- Improved efficiency of working muscles; and
- Decreased blood pressure.

On the basis of scientific studies of the effects of aerobic exercise on post-polio subjects, the general features of aerobic exercise programs should be:

- Low intensity and non-fatiguing exercise;
- Activities the patient enjoys – to improve compliance; and
- Low resistance activities that are least likely to fatigue compromised muscles. These activities may include walking, cycling on a level surface, swimming and hydrotherapy

Strengthening Exercise

Several studies have shown that people with a history of polio can improve their muscular strength and endurance by following an individualised, carefully monitored exercise program. However, at this stage, there is no consensus in the literature regarding the best strengthening exercise protocol to follow. The key factor appears to be intensity. Most authors recommend that strengthening exercise should be non-fatiguing.

Isometric exercise is most useful in muscles with a grade of less than 3 or in muscles over a painful joint. Isometric exercise may promote circulation and help retain some joint stability in body parts with this degree of weakness. Isotonic and isokinetic exercise is most useful in muscles with grade 3 or better strength and without a painful joint.

Stretching Exercise

Shortening of trunk and limb muscle is common in patients with a history of polio, particularly in those patients who are non-ambulatory. Muscle shortening may contribute to pain and decrease movement efficiency. Prescription of specific stretches for localised muscle tightness may be indicated as part of a physiotherapy program.

Gentle stretching of tight muscle in the post-polio patient may:

- Improve posture;
- Relieve muscle tension and cramping;
- Reduce the risk of musculotendinous and joint range of motion;
- Prevent irreversible contractures; and
- Increase general flexibility before strengthening exercises.

It must be noted that reduced flexibility is not necessarily detrimental. Tightness of certain structures may help to provide stability and improve function. Therefore, stretching is not always appropriate.

Hydrotherapy

Hydrotherapy is often the exercise method of choice for post-polio patients. When hydrotherapy is used in combination with other physiotherapy techniques, it has been found to improve muscle strength, range of movement, balance, coordination and endurance.

Hydrotherapy is particularly beneficial to the post-polio population because:

- The scope of water activities can benefit post-polio patients with all levels of ability;
- Exercising in water is a very pleasant and encouraging way to exercise particularly for non-mobile patients;
- Warm water promotes muscle relaxation and improved circulation;
- It allows particular movements to be either assisted, supported or resisted by the buoyancy of water; and
- It reduce weight bearing and can decrease mechanical stress on limbs, depending on the level of immersion.

It is essential that the instructor or therapist is aware of the patient's medical condition and the approach to exercise that is appropriate for these patients.

Surgical Considerations¹¹⁹

There are few studies regarding the effects of surgery or anaesthesia in post-polio individuals. However, there are certain factors that need to be considered prior to surgery. These include:

- Regional anaesthesia is preferable to general anaesthesia because it has fewer side effects. Individuals at greatest risk of complications during general anaesthesia are those with a history of ventilatory use or swallowing difficulties, those with involvement of the shoulders, arms or trunk, and individuals with a history of respiratory problems.
- In general, polio muscles tend to be more sensitive to muscle relaxants than normal muscles. As a result, a reduced dose of muscle relaxants is usually recommended.
- Close monitoring of pulmonary function is critical after general anaesthesia.
- Sleep apnoea may be aggravated following general anaesthesia.
- Healing may be delayed in paralysed limbs due to decreased blood supply.
- Polio-affected muscles may be temporarily weaker after general anaesthesia and may increase the need for ambulatory support.
- Recovery may be prolonged by two or three times beyond what is expected for the general population, depending on the individual's age, extent of paralysis and length of surgery.
- A supervised program of graded exercise may be required to reverse the effect of bed rest.

The Role of the General Practitioner in the Management of the Late Effects of Polio

General Practitioners are ideally placed to coordinate lifetime holistic care for the individual with a history of polio. In particular the GP can assist the post-polio patient by:

- Developing an assessment and management program to address the patient's specific symptoms;
- Ensuring continuity of care;
- Helping the patient and their family understand the condition (patient education); and
- Promoting a healthy lifestyle.

Developing an Evaluation and Management Program

The development of a comprehensive evaluation and management program is essential for the post-polio patient to ensure that symptoms are addressed and the progression of symptoms is reduced. This may be achieved through:

- Identifying medical, neurological and orthopaedic conditions that could be mimicking the symptoms of LEOP;
- Managing disabling symptoms – referral to other services, pharmacological treatment, etc.; and
- Monitoring patient's symptoms through ongoing evaluation.

Although the patient may not be exhibiting late onset problems, comprehensive evaluation and the development of a treatment plan that addresses optimal health and well-being may be beneficial in minimising the impact of future problems.

Continuity of Care

Continuity of care is one of the greatest contributions that the GP can provide to the patient with a chronic condition such as LEOP. Many of these patients may develop new symptoms over time and require ongoing evaluation of their medical condition to monitor their progress. The GP, who is often the patient's first contact with the health care system can provide this ongoing evaluation and refer the patient to other health professionals and services as required. GPs are uniquely placed in the health care system to advocate for their patients because of the good rapport that can be established between patient and doctor. Good rapport in the doctor/patient relationship is important in order to:

- Provide information and support;
- Develop a comprehensive treatment plan with the patient and partner/carer; and
- Refer as appropriate to health professionals and services.

Referral to Other Health Professionals

Although the GP can provide a comprehensive service to the post-polio patient, it is vital for them to recognise the importance of specialists and allied health professionals in the management of this condition. Due to the complex nature of LEOP it is often necessary to refer these patients to other health professionals who have specialist skills and knowledge that can assist the patient to manage their often disabling symptoms.

Specialists

Specialist physicians, in particular neurologists, orthopaedic surgeons and respiratory physicians, can occupy a key position in the health care of post-polio patients.

The benefits of referring to a specialist include:

- Confirmation of a diagnosis (in particular identifying other diagnoses that may be causing symptoms);
- Establishing a shared management plan;
- Access to the specialised skills and knowledge of the specialist; and
- Access to comprehensive assessment and treatment procedures.

Allied Health Professionals

Allied health professionals have specific skills that can assist the patient with a history of polio. In particular physiotherapy, occupational therapy, and social work have been utilised in post-polio clinics throughout the world and have been found to be essential in the assessment and management of all patients with a history of polio. Therapists can be accessed through both the public and private sectors, however to provide a comprehensive service to the post-polio patient, it is important that they have a thorough understanding of this condition.

A resource manual and training package has been developed by the Queensland Department of Health and distributed to Queensland Health Districts throughout the state. These resources were developed to increase the awareness of medical and allied health staff of LEOP and provide them with the necessary information that is required to provide a comprehensive service to these patients.

Physiotherapist

Through a variety of hands-on treatments, education and exercise prescription, physiotherapists can assist the post-polio patient in maximising their level of function and minimising future deterioration.

Assessments conducted by the physiotherapist may include:

- Manual muscle testing;
- Measurement of joint range of motion and leg length;
- Evaluation of posture; and
- Analysis of activities and positions that provoke or relieve muscle and joint pains.

Treatment programs for the post-polio patient may include the following:

- Prescription of an exercise program, designed for the individual;
- Correction of postural alignment;
- Pain management;
- Application of therapeutic modalities;
- Joint mobilisation;
- Referral to appropriate services;
- Hydrotherapy; and
- Prescription and training in the use of mobility aids and/or braces.

Occupational Therapist

Occupational therapists use various methods to help their patients achieve the highest level of functioning and wellness attainable in their daily lives, and to cope with the physical and emotional aspects of their disability. Occupational therapy assessment and treatment for the post-polio patient may include:

- Assessment of home and workplace environments and recommendations for necessary adaptation;
- Evaluation of activities that produce weakness, pain or fatigue;
- Recommendations and training in the use of adaptive equipment;
- Vocational counselling and rehabilitation;
- Education of the patient regarding energy conservation, joint protection and work simplification; and
- Guidance to family members and carers in safe and effective methods of caring for the patient.

Social Worker

The social worker can provide a valuable service to the post-polio patient by facilitating ongoing adjustment to physical and lifestyle changes. This may be achieved through:

- Assessing how new health problems and functional loss impact on the patient and their family, friends and colleagues;
- Assisting the patient to identify coping strategies used;
- Increasing personal and external resources – facilitate referrals to community services;
- Providing education and support; and
- Counselling.

Other Health Professionals

Referral to other health professionals for the assessment and management of some of the symptoms experienced by post-polio patients may be required including:

- *Speech Pathologists* for evaluation and management of swallowing and speech difficulties.
- *Orthotists* for prescription and fabrication of orthoses / braces.
- *Podiatrists* for assessment and treatment of foot conditions (commonly present in patients with a history of polio).
- *Psychologists* to provide counselling, education and support in regard to psychological difficulties the patient may experience.
- *Dieticians* to provide education and management strategies regarding weight management.

Patient Education

The amount of literature that is available on this condition is increasing as clinicians develop a greater understanding of the pathophysiology of this condition and the impact it has on the post-polio individual. Patients and their families can benefit from the educational resources and support groups provided by the Paraplegic and Quadriplegic Association of Queensland (refer to the section on Support Groups for further details regarding this association).

Promoting a Healthy Lifestyle

The maintenance of a healthy lifestyle is essential for post-polio patients to limit the possible development of future problems. Patients should to be encouraged to:

- Take care of their health by eating a healthy diet, maintaining a healthy weight, avoiding alcohol and ceasing smoking;
- Listen to their own bodies – change or reduce activities (including exercise) that cause pain or excessive tiredness;
- Pay attention to their lifestyle; and
- Reduce stress.

Other Issues to Consider

Employment

Due to the symptoms of pain, fatigue and weakness, the patient may have difficulties maintaining full time or part time employment. Changes in type of employment (retraining, job relocation), changes to how work is performed, modifying work schedule and ergonomic changes to the workplace may be required. Ultimately, a patient may be required to cease employment if these symptoms cannot be managed with energy conservation and work simplification techniques. This may be difficult for some patients who have expected to have several years of working life ahead.

The GP can help the post-polio patient by:

- Considering early vocational rehabilitation;
- Discussing financial implications if the patient is required to retire. Some patients and carers may be eligible for Social Security benefits; and
- Referral to other health professionals and services to assist the patient through this process, e.g. social worker, occupational therapist, vocational counsellor.

Driving

As this group of patients ages and their potential for disability increases, the GP may be required to regularly assess their patient's ability to drive. Difficulties that the patient may experience that may affect their ability to drive include:

- Reduced range of motion in arms, legs or neck;
- Reduced muscle power;
- Impaired coordination;
- Severe joint or muscle pain;
- Impaired concentration due to fatigue;
- Respiratory difficulties; and
- Sleep disorders.

The aim of medical assessment of these patients in regard to driving is to detect those drivers who may have difficulty in controlling their motor vehicle, or a specific type of motor vehicle and to identify those drivers who may benefit from specific vehicle adaptations.

Adaptive equipment may be installed in many vehicles to enable the impaired driver to operate their vehicle safely. Physical demands on drivers from certain vehicles (e.g. bus, truck) may be substantial and should be considered by the GP.

In some cases a driving assessment by a driving assessor and/or an Occupational Therapist who is specifically trained in this area, may be required.

National guidelines in regard to driving have been developed. For further information regarding a patient's fitness to drive, refer to:

Austrroads. Assessing Fitness to Drive: Austrroads Guidelines for Health Professionals and the Legal Obligations. Sydney: Austrroads Inc. 1998.

Resources

Support Services

Queensland

- **Post-Polio Support Group Network** have established support groups throughout Queensland who meet on a regular basis to share information, offer peer support and attend presentations from relevant individuals and organisations. In addition the association provides information about the late effects of polio and related issues to members, the community and health practitioners.
- For details of Post-Polio Support Groups, meeting dates and times throughout Queensland contact the Paraplegic and Quadriplegic Association of Queensland.

The Paraplegic and Quadriplegic Association of Queensland Inc.

	Brisbane	North Queensland
Address:	Cnr O'Connell St and Shafston Ave KANGAROO POINT	1 Charters Towers Road TOWNSVILLE
Postal Address:	P.O. Box 5651 WEST END 4101	P.O. Box 4132 TOWNSVILLE WEST 4810
Phone:	(07) 3391 2044 1800 810 513 (Toll Free)	(07) 4721 5800 1800 024 422 (Toll Free)
Fax:	(07) 3391 2088	(07) 4721 5802
Email:	pqaq@pqaq.com.au	pqaq@beyond.net.au

Interstate

New South Wales	Victoria	South Australia	Western Australia	Tasmania
N.S.W. Post-Polio Network P.O. Box 888 Kensington N.S.W. Phone: (02) 9663 2402 Email:polio@fastlink.com.au	Polio Network Victoria P.O. Box 1189 Collingwood VIC 3066 Phone: 1800 803 384 Email:theteam@paraquad.asn.au	Post-Polio Support Group of South Australia 23a King Willam Rd Unley S.A. 5061 Phone: (08) 8357 8909 Email:nrc@camtech.net.au	Post-Polio Network of Western Australia P.O. Box 257 Subiaco W.A. 6904 Phone: (08) 9383 9050	Post-Polio Network – Tasmania Inc. P.O. Box 194 Beaconsfield TAS 7270 Phone: (03) 6383 1690 Email:polio.tas@microtech.com.au

Internet Sites

Polio and Post-Polio Information Sites

Australian Sites

Paraplegic and Quadriplegic Association of Queensland

<http://www.pqaq.gil.com.au>

Post-Polio Network of New South Wales

<http://www.post-polionetwork.org.au>

Polio Network of Victoria

<http://www.paraquad.asn.au/services/polio/polio.html>

Post-Polio Network – Tasmania Inc.

<http://www.tased.edu.au/tasonline/polionet/>

Neurological Resource Centre – Post-Polio Support Group of S.A.

http://www.span.com.au/nrc/p_polio

Mornington Peninsula Post-Polio Support Group

<http://www.satlink.com.au/~ddlloyd>

Polio Services Victoria

<http://www.psv.svhm.org.au/>

Overseas Sites

The Lincolnshire Post-Polio Network

<http://www.zynet.co.uk/ott/polio/lincolnshire>

Post-polio information service for polio survivors and medical professionals. The site is divided into three main sections:

- Library – containing articles covering PPS, LEOP etc. All articles are the full text version;
- Networking; and
- The Directory – a catalogue of other resources both on the internet and elsewhere.

Gazette International Networking Institute (GINI)

<http://www.post-polio.org>

Consists of:

- International Polio Network – provides information to polio survivors, their families and the health care community and promotes networking among the post-polio community;
- Post-Polio Task Force – a task force of clinicians and researchers who specialise in diagnosing and treating polio survivors and symptoms of PPS, as well as patient advocates for survivors of polio; and
- International Ventilator Network – links ventilatory users with each other and with health care professionals interested in the options for mechanical ventilation and home care.

Archives of POST-POLIO-MED@MAELSTROM.STJOHNS.EDU

<http://maelstrom.stjohns.edu/archives/post-polio.html>

Questions and answers about PPS. You must be a subscriber to post to, browse or search the PPMed list.

The Polio Information Centre Online

<http://128.59.173.136/PICO/PICO.html>

Provides information about the poliovirus and links to other polio information.

Harvest Centre's Post-Polio Page

members.aol.com/harvestctr/pps/polio.html

Polio and PPS educational audio tapes, the computerised assessment program to help local doctors diagnose and treat your PPS, downloadable articles.

Post Polio Recently Published Medical Articles

<http://www.execpc.com/~epwoll/medindx.html>

This site contains a list of medical articles on PPS published from 1990 to 11/01/2000.

Post~Polio Med Web Pages

<http://angus.interspeed.net/ppmed/index.shtml>

Post~Polio Med is a forum for questions and answers to and from PPS researchers, physicians and other post-polio medical professionals, polio survivors, family, friends, students and others who are interested in PPS.

Post Polio Syndrome

[Neuro-www.mgh.harvard.edu/forum/PostPolioSynd/Menu.html](http://www.mgh.harvard.edu/forum/PostPolioSynd/Menu.html)

This is a webforum to discuss and comment on PPS.

Polio Experience Network

<http://www.polionet.org>

Large post-polio resource site including library, PPS disease and treatment information, instructional material, newsletter and vaccine information.

Polio Survivor's Page

<http://www.eskimo.com/~dempt/polio/html>

Polio and post-polio resources including articles, newsletters, an information package and related links.

MEDLINE Plus: Polio and Post Polio Syndrome

<http://www.nlm.nih.gov/medlineplus/polioandpostpoliosyndrome.html>

A service of the National Library of Medicine. Links to MEDLINE and MEDLINEplus

Post Polio Home Page and Links to same

<http://home.earthlink.net/~polioinfo/index.html>

Newsletters, chat sites, doctors reports, government help, medication, fact sheets etc.

Grace Young's Web Page

<http://www.geocities.com/HotSprings/4713>

Provides information on energy conservation

The Rollin Rat

<http://www.azstarnet.com/~rspear>

Richard Spear's home page to PPS and Disability Resources

The No Frill Edition

<http://www.azstarnet.com/~rspear/tindex.html>

Contains interesting reading on the cause and symptoms of PPS, rehabilitation and therapy, adaptations for daily living, social security information and a history of polio and PPS.

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