

## 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:<sup>39</sup>

“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.<sup>40</sup> These criteria were further refined by Halstead<sup>41</sup> in 1991 and are outlined in Table 2.

<b>Table 2: Criteria for the Diagnosis of PPS</b>
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.<sup>5</sup> 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)<sup>28</sup> 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.<sup>5</sup> 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.