Post-polio syndrome

Author: Professor Frans Nollet, MD, PhD

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Department of Rehabilitation Medicine, VU University Medical Center, PO Box 7057, 1007 MB Amsterdam, Netherlands. f.nollet@vumc.nl

Abstract

Post-polio syndrome or post-polio syndrome (PPS) is the commonly accepted term to describe the neuromuscular symptoms that may develop many years after acute paralytic poliomyelitis. The prevalence estimates of late onset neuromuscular symptoms in prior polio patients vary between 25 and 74%. PPS patients are diagnosed on the basis of a confirmed history of paralytic poliomyelitis, followed by partial to fairly complete neurological recovery and functional stability for at least 15 years. After this recovery period, new or increased muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalized fatigue, muscle atrophy, or muscle and joint pain, appear usually gradually. PPS diagnosis is made by exclusion. Laboratory tests are used to show evidence of prior polio and to exclude other diseases: electromyography (EMG) displays signs of reinnervation and denervation both in symptomatic and non-symptomatic muscles. Muscle biopsy findings include type-grouping of muscle fibres as the result of reinnervation, and hypertrophy of muscle fibres as a compensation for the loss of muscle fibres. No curative treatment is available for PPS. Management is preferably multidisciplinary and aims both at reducing muscle overuse and rebalancing muscular capacities and demands. It consists of exercise, assistive devices and lifestyle changes. The etiology of PPS is still unclear. It is currently hypothesized that muscle fibres undergo denervation again due to distal degeneration of axons of enlarged motor units.

Keywords
post-polio syndrome, post-poliomyelitis syndrome, late effects of polio, poliomyelitis

Disease name and synonyms

Post-polio syndrome or Post-polio syndrome (PPS) is the commonly accepted term to describe the neuromuscular symptoms, which may develop many years after acute paralytic poliomyelitis. Several other terms are used to describe late onset symptoms ("Post-poliomyelitis progressive atrophy" (PPMA), "Post-poliomyelitis muscle dysfunction" (PPMD), "Late effects of polio" and "late onset polio sequelae") but are not synonymous (see under diagnostic criteria/definition).
Diagnostic criteria/Definition

**Post-polio syndrome**

The diagnostic criteria have been revised after an international meeting held in May 2000 by the March of Dimes Birth Defects Foundation[1] and are based on:

1) A confirmed history of paralytic poliomyelitis characterized by an acute illness with fever and a usually asymmetrically distributed, flaccid paresis of a varying number of muscle groups. Evidence of motor neuron loss on neurological examination with signs of residual weakness, atrophy, loss of tendon reflexes and intact sensation. Signs of denervation or reinnervation on electromyography (see laboratory criteria).

2) A period of partially to fairly complete neurological recovery after acute paralytic poliomyelitis followed by neurological and functional stability for at least 15 years.

3) New or increased muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalized fatigue, muscle atrophy, or muscle and joint pain.

4) Symptoms usually have a gradual, but sometimes a sudden onset and should persist for at least one year.

5) No other medical diagnosis to explain the symptoms (see under differential diagnosis).

There is insufficient basis to classify sub-types of PPS, such as "post-polio myelitis progressive atrophy".

**Post-polio myelitis progressive atrophy (PPMA)**

PPMA was intended to classify cases in which strength loss and new atrophy was objectified with serial examinations. However, the ability to detect individual changes in strength is limited[2] given the fact that the rate of decline in muscle strength is slow and has only been found in studies with a long term follow-up.[3]

**Post-polio myelitis muscle dysfunction (PPMD)**

The term was defined in a Workshop of the European Neuromuscular Center in 1994[4] PPMD refers to local signs of muscle dysfunction that include new or increased muscle weakness and/or muscle atrophy, and/or muscle pain and/or muscle fatigue. The rationale for this is that within an individual one or more muscles may be symptomatic while other muscles are not.

"Late effects of polio" and "late onset polio sequelae"

These terms are less specific and refer to the many new symptoms that patients with a history of polio may experience.[5] These symptoms may implicate new motor-unit dysfunction or may result from secondary medical conditions related to the polio residuals such as compression neuropathies after years of crutch walking or degenerative arthritis due to long-term (compensatory) overload of hypoplastic and/or deformed joints.

**Laboratory criteria**

Laboratory tests ( electromyography, muscle biopsy) should show evidence of prior polio and may be indicated to exclude other disorders. Evidence of prior polio may not only be found in muscles with residual paresis and in muscles which had clinically recovered, but also in muscles that were never thought to be affected. However, with laboratory tests, PPS cannot be distinguished from stable neuromuscular functioning following polio.[6]

**Electromyography (EMG)**

EMG shows signs of reinnervation as well as signs of denervation, although to a much lesser extent, both in symptomatic and non-symptomatic muscles. No signs of polynuropathy should be found.

**Muscle biopsy**

Muscle biopsy findings include type-grouping of muscle fibres as the result of reinnervation, and hypertrophy of muscle fibres as a compensation for the loss of muscle fibres. Sometimes there is apparent type I muscle fibre predominance. Isolated small, angular muscle fibres as a sign of acute denervation may be found while group atrophy, representing the loss of complete motor neurons, is unusual.[6-8]

**Differential diagnosis**

The diagnosis of PPS is made by exclusion. Other possible causes should be thoroughly eliminated. Some neurological conditions that may be confused with PPS are:

- Progressive spinal muscular atrophy,
- Amyotrophic lateral sclerosis (Amyotrophic lateral sclerosis),
- Inclusion body myositis,
- Compression neuropathy,
- Radiculopathy and spinal stenosis.

Therefore the diagnosis of PPS should be made by a neurologist with neuromuscular expertise. Other conditions that should be excluded are orthopedic disorders such as degenerative joint diseases and general disorders such as anemia and thyroid dysfunction.

**Prevalence**

The estimates of the prevalence of late onset neuromuscular symptoms in prior polio patients vary between 25 and 74% due to differences in study populations, the definition of symptoms,
and the methods of assessment either with questionnaires or by examination.[9-14]. Three population-based studies have demonstrated that the prevalence of new symptoms is high. Windebank et al.[11] examined a sample of 50 prior polio patients. New muscle weakness was reported by 22 subjects and new neuromuscular complaints by 32 subjects. In a study with mailed questionnaires, 35% of the 551 respondents noticed new muscle weakness and 78% at least one new neuromuscular symptom at an interval of 32-39 years after the acute polio.[12] Ivanyi et al.[14] found 39 years after the 1956 Dutch polio epidemic that 58% of the 260 respondents reported new weakness and 64% increasing neuromuscular symptoms. Recently, the specificity of the symptoms of PPS has been debated since they were also found in a general population sample.[15;16] However, given the odds ratio’s (9.6 for new muscle weakness and 11.1 for fatigue in subjects with polio residuals compared with a general population sample)[16] the prevalence of late onset neuromuscular symptoms of polio has probably only been slightly overrated in questionnaire studies of population based samples.

**Risk indicators**

Several risk indicators for the development of post-polio syndrome have been identified, such as the severity of the initial polio paresis, the degree of recovery from the acute polio, the extent of residual impairments, the age at acute polio, the number of years since polio, age, and female gender.[11;12;14;17;18]

**Severity of acute polio and residual paresis**

These risk indicators are related to the number of motor neurons that were lost and the subsequent degree of adaptation resulting in increased motor unit size. The risk to develop post-polio syndrome increased when subjects had more severe residual paresis after recovery from acute polio.[12] PPS symptoms have also been found among individuals with full (subjective) recovery from the acute polio and even among cases who had suffered from non-paralytic polio.[12;14;16;19] However, the prevalence of PPS in these individuals is much lower than in individuals with polio residuals.

**Time interval and age**

The time interval since polio was found to be a risk factor for PPS.[12] The role of aging itself seems limited since most PPS patients are in their forties, an age range in which normally a loss of motor neurons is not yet supposed to occur.[20] However, muscle fibers may be lost at an earlier age, and more rapidly, due to premature metabolic exhaustion of motor neurons.[21] It is known that the severity of the acute polio paresis increases, and the capacity to recover declines, with increasing age at onset of acute polio.[22;23] Therefore, age at onset of polio may be a risk factor since individuals, who contracted polio at an older age, had recovered to a lesser extent and thus had a muscle capacity, which was more restricted ever since the acute polio.

**Female gender**

In most studies of hospital populations, PPS patients were predominantly women.[24-26] Female gender was found to be a risk indicator for PPS by Ramlow et al.[12] but this finding was not confirmed in the study of Ivanyi et al.[14] Perhaps, in women the relatively smaller muscle mass and greater increase in weight with aging, a factor that has also been found to play a role in PPS,[27] leads more readily to a disturbed balance between capacity and demands than in men. It may well be that the impact of the new symptoms on functioning is greater in women than in men or, alternatively, that women are more willing to seek medical attention for their complaints.

**Clinical description**

PPS patients usually experience new or increased symptoms 30 to 40 years after the acute polio. Muscle weakness, fatigue, muscle and joint pain and loss of endurance and decreasing activity levels with regard to walking and climbing stairs were the most common complaints in the early studies describing the new neuromuscular complaints.[24-26;28] The symptoms develop gradually over time, although in some cases the onset of new symptoms may have been sudden, especially following a period of inactivity, trauma, and surgery or after vigorous activity.[1] Muscle weakness usually occurs in limbs or trunk, but may rarely also develop in swallowing or respiratory muscles.[29;30]

**Rate of strength loss**

The decline in muscle strength is usually slow. Studies with a follow up of 3 years or less failed to demonstrate strength loss.[31-33] In longer term studies the decline in strength was estimated at 1-2% per year.[3;27;34]

**Overuse and cardiorespiratory conditioning**

The symptoms of PPS such as muscle pain, increased fatigue after physical activity and delayed recovery following physical activity may signify that muscles are overused in conducting ordinary daily life activities.[35;36] Support for
such a chronic overuse of muscles in former polio subjects has been found in studies showing elevated activities of serum creatine kinase that were related to the distance walked during the previous day,[37] and in studies showing a type I fibre predominance in lower leg muscles supposedly due to fibre type transformation from chronic overload.[38;39] Also, PPS subjects have been found to recover slower from fatiguing exercise than stable polio subjects.[40;41] Another factor that is said to contribute to the symptoms is a poor cardiorespiratory condition.[42-44] However, the cardiorespiratory condition of polio subjects was not worse than that of healthy, comparably active subjects.[45]

In this study it appeared that the reduced submaximal performance capacity of the polio subjects was strongly correlated with the limited available muscle capacity and that movement economy was diminished compared with the control subjects. Lower concentrations of some oxidative enzymes in muscles of polio subjects have also been reported while other oxidative enzymes were within normal ranges.[8;39;46] The clinical significance of these findings has been debated.[47]

**Abilities and handicaps**
Growing restrictions to perform activities was mainly found for physical abilities such as walking, climbing stairs, and transfers.[11-13;24;26-28] In a recent study, physical functioning declined little over a 6-year period.[48] In agreement with the concept of overuse was the finding that the extent of paresis was the only prognostic factor for a decline in functioning. A significant increase in handicap severity for the categories mobility, occupation and social integration was found in PPS subjects over a period of 4-5 years, while in non-PPS subjects the handicap severity remained unchanged.[49]

**Management including treatment**
No curative treatment is available for PPS. Management of PPS is preferably multidisciplinary in order to restore the balance between decreasing capacities and demands.

**Pharmacological treatment**
At present no medication for PPS symptoms is available. Pyridostigmine is the only drug that has been investigated in randomised double-blinded trials.[50;51] In a multicenter study, pyridostigmine was found not to be effective.[50] In selected patients with proven neuromuscular transmission defects, pyridostigmine did not reduce fatigue, although a limited beneficial effect on physical performance was found.[51]

**Multidisciplinary management**
To reduce overuse and rebalance the capacities and demands, conservative management consists of 3 essential components: exercise, assistive devices, and life style changes. Therefore, PPS patients are best treated within a multidisciplinary, specialized rehabilitation setting. Since individuals show considerable differences in polio residua, treatment is individually adjusted and should be preceded by a thorough customised medical and functional evaluation.

**Exercise**
Exercise can optimise cardiorespiratory fitness and may add to the patient’s sense of well-being.[52-54] Exercise should be non-fatiguing and performed at submaximal levels to avoid overloading of the limited muscle capacity. Exercise can improve muscle strength especially in case of disuse and muscle groups that are only moderately affected.[55] Intensive strengthening exercises are not generally recommended, although they may occasionally be indicated. Functional training may also be useful to improve the efficiency of ambulation.

**Orthoses and assistive devices**
Braces may be helpful to support weak muscles and to stabilize (painful) joints. The condition of existing, often old braces should be carefully examined and judged whether they are still adequate, based on biochemical evaluation of walking abnormalities.[56;57] Assistive devices comprise crutches, the use of a wheelchair, motorized scooters and home adaptations such as elevators, seating devices in the kitchen or shower. All of these devices should be individually indicated.

**Life style changes**
Pacing of activities and taking rest intervals are of major importance to relieve symptoms. It has for instance been shown that upper extremity complaints often result from overuse of shoulder and arm muscles.[58] Usually PPS patients have successfully been learned to deny their symptoms from child on to achieve a normal life.[59] Therefore, PPS patients may have great difficulty with adapting their life style to their decreasing abilities and psychological support may be indicated.

**Etiology**
The etiology of PPS is still unclear. It is currently hypothesized that muscle fibres undergo denervation again due to distal degeneration of axons of enlarged motor units.[60] Probably, the large motor units never really stabilize following the recovery from the acute poliomyelitis. There
is evidence for an initially balanced, but ongoing denervation and reinnervation of muscle fibers.[61;62] With time the balance cannot be maintained and denervation gets the upper hand supposedly from premature aging of metabolically exhausted motor neurons.[60] A reduction in size of enlarged motor units was also found in prospective follow-up studies with macro-EMG.[63;64] McComas et al. [21] performed motor unit number estimation in prior poliomyelitis patients with a 2-year interval. They found a decreasing size of motor units after 2 years and also that the rate of motor unit loss was twice that occurring in healthy subjects aged above 60 years. Other factors that may contribute to the new muscle weakness are neuromuscular transmission defects [65] and an impaired ability to activate the muscles.[66;67]

**Diagnostic methods**

Careful history taking and physical examination are essential for the diagnosis of PPS. In addition to investigations indicated to exclude other medical conditions, muscle imaging (CT or MRI) and pulmonary functions tests may provide useful information:

**Muscle imaging**

The measurement range of manual muscle testing has an upper limit. Therefore, a considerable loss of muscle strength can be missed during physical examination. Beasley showed that polio subjects with only 55% of the muscle strength values of healthy subjects had manual muscle testing results of normal strength. [68] Fatty infiltration of muscles and atrophy can be visualized with computed tomography or magnetic resonance imaging, even in muscles that were considered to be unaffected during the acute polio, and thus provide additional information about the available muscle capacity.[67;69]

**Pulmonary function tests**

Sleep complaints, morning headache, severe daytime fatigue or drowsiness may point to respiratory insufficiency. These problems are mostly seen if the respiratory muscles were affected at the time of acute polio, which often resulted in severe residual paresis of trunk muscles with kyphoscoliosis. A sleep apnea syndrome may also be present. Therefore, these complaints require a careful evaluation of respiratory function. Sleep registrations may be indicated as well as repeated over-night examinations of blood gasses to identify an increase in arterial carbon dioxide tension.[70] Some PPS patients may require (non-invasive) nightly ventilatory support.[71]

**Unresolved questions**

A better understanding of the cause of the degeneration of motor units is needed and may provide new clues for pharmacological interventions.

Multidisciplinary rehabilitation programs need to be developed further and evaluated; the role of exercise in maintaining the available muscle capacity should be further investigated.

**References**

12. Ramlow J, Alexander M, LaPorte R, Kaufmann C, Kuller L. Epidemiology of the post-
41. Agre JC, Rodriguez AA, Franke TM. Subjective recovery time after exhausting

http://www.orpha.net/data/patho/GB/uk-PP.pdf


