International Encyclopedia of Rehabilitation

Center for International Rehabilitation Research Information and Exchange (CIRRIE)
515 Kimball Tower
University at Buffalo, The State University of New York
Buffalo, NY 14214
E-mail: ub-cirrie@buffalo.edu
Web: http://cirrie.buffalo.edu

This publication of the Center for International Rehabilitation Research Information and Exchange is supported by funds received from the National Institute on Disability and Rehabilitation Research of the U.S. Department of Education under grant number H133A050008. The opinions contained in this publication are those of the authors and do not necessarily reflect those of CIRRIE or the Department of Education.

Post-polio Syndrome

Elisabeth Farbu, MD, PhD
Neurocenter
Stavanger University Hospital
N-4068 Stavanger
Norway

Background

The first case report on new muscle weakness several years after paralytic poliomyelitis (polio) was published in 1875 when Raymond demonstrated a 19-year old tanner who suffered from new atrophy in his shoulder more than a decade after having passed acute polio for Charcot. In the following centenary, polio was considered to be a three-phasic illness starting with acute paralysis, followed by a recovery and subsequently a stable phase with more or less residual weakness. This dogma changed as the large numbers of polio survivors in the 20th century grew older and reported new symptoms several decades after the acute illness and data were systematically recorded.

The term post-polio syndrome (PPS) was launched in 1985 by Halstead to cover all aspects of late consequences occurring several years after acute paralytic polio (Halstead and Rossi 1985). The symptoms included were new weakness, generalized fatigue, decreased muscular endurance, muscle pain, joint pain, and cold intolerance. Both Halstead and Dalakas launched suggestive criteria and definitions of PPS:

1. Confirmed history of polio
2. Partial or fairly complete neurological and functional recovery after the acute episode
3. Period of at least 15 years with neurological and functional stability
4. Two or more of the following health problems occurring after a stable period: extensive fatigue, muscle and/or joint pain, new weakness in muscles previously affected or unaffected, new muscle atrophy, functional loss, cold intolerance
5. No other medical explanation found
6. Gradual or abrupt onset of new neurogenic weakness

PPS is now reckoned to be a condition following paralytic polio in which the muscle strength and clinical function are slowly deteriorating, without any dramatic loss of muscle strength as in motor neuron diseases. Guidelines for diagnosis and management have been published the US (MoD) and Europe (EFNS) (Farbu et al. 2006, March of Dimes 2000).

It is estimated that between 20-50% of polio survivors will experience PPS, but there are diverging data about who are at greatest risk. The PPS symptoms are somewhat identical in all parts of the world, and their management should follow international guidelines. The basic care for PPS-patients is based on a proper diagnostic process in which all other possible explanations for the new deterioration have been ruled out, followed by carefully adapted rehabilitation and training programmes. Some research has supported the theory that PPS could be partially explained by increased inflammatory activity both in the nervous system and on the neuromuscular level. This pathophysiological element has given a rationale for trying immunomodulatory medical treatments. Steroids and intravenous immunoglobulin has been used in some clinical trials, but none of the results have shown any convincing improvement (Dinsmore, Damsrosia and Dalakas 1995; Elovaara et al. 2008).

Acute paralytic polio

The term poliomyelitis originates from the Greek words “polios” (gray), “myelos” (marrow), and the Latin suffix “itis”(inflammation) and refers to the specific localization of the infection in the anterior horn (gray matter) in the spinal cord. Acute paralytic polio is caused by infection of the poliovirus, which is enterovirus of the Picornaviridae family. The polio virus is an RNA-virus where the virion consists of a single-stranded RNA genome of positive polarity in a non-enveloped icosahedral protein capsid. In the majority of cases, an immunization with no clinical symptoms takes place, whereas 1% of the affected persons develop fever and signs of aseptic meningitis and flaccid paralysis. The virus attacks the motor neurons in the anterior horn of spinal cord, which can cause flaccid muscle weakness if the inflammation is severe enough. The most common location of weakness is in the lower limbs, but all levels of the spinal cord can be affected including the upper cervical cord and brain stem, causing respiratory difficulties. A random distribution of the muscle weakness is common. During the large epidemics in the USA and Europe in the 1940-50s, the mortality rate was about 7%, mainly due to respiratory weakness (Mulder 1995). There is no cure for acute polio, but vaccination is highly protective (Swennen and Levy 2001). Salks inactivated vaccine and Sabins oral vaccine and were introduced in the 1950ies, and they were quickly implemented in the industrialized world. This led to a formidable reduction of the polio epidemics and new cases of paralytic polio diminished rapidly. Europe, Australia and the Americas have now been declared polio free with no reported polio cases the last years, but despite the world wide vaccination campaign, there are still (2009) four countries in Africa and Asia were the polio virus is still endemic and new epidemics occur. The remaining countries are Afghanistan, India, Nigeria and Pakistan (WHO 2009).

Recovery

As the inflammation diminishes after the acute polio phase, the temporarily disabled and infected neurons may gain function again. Other surviving motor neurons will also provide an extensive axonal sprouting and reinnervation of muscle fibers, and muscle fibers with existing innervation increase in size in a compensatory manner. These three factors contribute to the process of regaining muscle strength. The degree of residual weakness can vary from complete to partial persistent weakness. It is important to have in mind that that normal strength and muscle volume can be found despite a loss of 50% of the motor neurons within one segment (Bodian). In these cases subclinical signs of motor neuron affection with increased amplitudes on EMG-recordings can be found, confirming the impaired anterior horn function (Rekand et al. 2002). Most of the improvement is seen within the first six months, but further improvement can be found up to two years after the acute infection. The recovery phase is followed by a stable phase of neuromuscular function that can last for several decades.

PPS – definition and diagnosis

A patient with previous paralytic polio will not experience PPS until several years with a stable condition have passed. The start of the PPS may be very subtle and insidious, and the clinical course is rather modest, with no devastating progressive weakness such as in ALS. However, once the threshold for the neuromuscular compensatory mechanisms is passed, a more stepwise deterioration can be seen. The pathophysiology behind PPS is not fully understood, but overuse and metabolic stress on enlarged motor units, deterioration of the neuromuscular junction, the normal ageing process and inflammatory changes are thought to contribute to the clinical picture (Gonzalez et al. 2002; Grimby et al. 1998; McComas et al. 1997).

The symptoms reported for PPS are the same in all parts of the world. Muscle weakness, atrophy, generalised fatigue, post-exercise fatigue, muscle pain, fasciculations, cramps, cold intolerance, and joint pain dominate. These symptoms are common in the general ageing population and could be caused by a considerably amount of other conditions and illnesses, that must be ruled out as they may require other specific management. The primary goal for the physician when seeing a PPS patient for the first time will be to carefully examine and perform proper investigations to rule out other possible contributing factors.

Both Dalakas, Halstead and March of Dimes (MoD) have proposed criteria for PPS, with emphasis on the new clinical decline and muscular weakness several years after a plateauphase (Dalakas 1995, Halstead 1991, March of Dimes 2000).

The MoD-criteria are as follows

1. Prior paralytic poliomyelitis with evidence of motor neuron loss, as confirmed by history of the acute paralytic illness, signs of residual weakness, and atrophy of muscles on neurological examination, and signs of denervation on electromyography (EMG).
2. A period of partial or complete functional recovery after acute paralytic poliomyelitis, followed by an interval (usually 15 years of more) of stable neurologic function.
3. Gradual or sudden onset of progressive and persistent muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalized fatigue, muscle atrophy, or muscle and joint pain. (Sudden onset may follow a period of inactivity, or trauma, or surgery). Less commonly, symptoms attributed to PPS include new problems with swallowing or breathing.
4. Symptoms persist for at least a year.
5. Exclusion of other neurologic, medical, and orthopaedic problems as causes of symptoms.

The evidence of prior polio can sometimes be straightforward with a classical clinical picture of asymmetric and scattered muscle weakness and atrophy in a patient who became ill during one of the epidemics. If the symptoms and clinical findings are atypical such as generalised fatigue without any objective sign of motorneuron involvement, further investigation is needed. Electromyography (EMG) may show increased amplitude reflecting an enlarged motor unit, whereas nerve conduction studies should reveal normal findings for both motor and sensory nerves, except for the parameters regarding the motor units (Grimby et al. 1998). Other diagnoses such as peripheral neuropathy and myopathy can be ruled out after neurophysiological examinations. Computer tomography (CT) scans can be helpful to detect subclinical muscle atrophy (Ivanyi et al. 1998). The typical clinical pattern would be asymmetrical and often scattered weakness, involving several segments of the spinal cord, without any signs of upper motor neuron involvement, and no rapid and severe progressive deterioration. Tendon reflexes are often weakened or absent in the same scattered pattern. Fasciculations can be observed in the affected muscles, but is not generalized. Post-exercise fatigue and decreased muscular endurance during activity are frequent, and may be present before new weakness or atrophy is detected on examination. This is probably due to changes in the aerobic capacity of the muscle fibers, which are often enlarged with a reduced glycolytic activity (Borg and Henriksson 1991). The other symptoms are less specific and may be caused by several other disorders. Muscle pain may be present both in weak muscles,

but also in other healthy muscles as a result of overuse because of compensatory biomechanical use of other muscles.

Management

When the diagnosis of PPS has been established, a careful approach to the individual patient is advocated. Some patients may have very localised symptoms and findings, whereas other may need a broader follow up.

Muskuloskeletal deformities

Muskuloskeletal deformities like anisomelia, hyperextension of the knee joint, claw foot and hammer toes needs to be sorted out, and adaptation of proper devices like canes, orthoses, and individual fitted shoes can alleviate dysfunctional walking patterns, help energy economising, and relieve pain due to asymmetric work load (Brehm et al. 2007). Newer carbon orthoses is superior to heavier metal braces. Asymmetric work load may lead to degenerative changes in joints and the spine, and relevant imaging should be performed to rule out possible arthrosis or nerve root affection due to a degenerative spine. Such co-existing disorders should be managed following general guidelines for the specific disorder. The use of walking aids like stick or crutches may increase the risk for nerve entrapment in the wrist like carpal tunnel syndrome.

Muscle weakness and muscle pain

Physically inactive PPS patients have more symptoms than physically active patients, and there is profound evidence that both muscle weakness and muscle pain may be helped with proper and individually fitted training programmes. However, muscle pain can also be caused by overuse of certain muscle groups (Jubelt and Agre 2000), which emphasizes the importance of balancing between activity and restitution . No serious complications or side effects have been detected after implementing training programmes, but it should be added that most of the studies have been performed on patients younger than 60 years. Training programmes may be offered both on an individual and group basis, on institutional and home basis, and they follow the main principles with aerobic muscle training, intermittent breaks, and no maximal work load. Training programmes have also shown a positive effect in a longer term perspective, with better muscle performances several months after the training programme has ended (Farbu et al. 2006). Dynamic exercises in warm water seem to be particularly useful as water training reduces the work load on weak muscles, and pain may also be diminished in warm water. The psychosocial support when performing exercises in a group with other patients could lead to an increased well-being and be of help for the individual patient (Strumse et al. 2003, Willen and Scherman 2002, Willen et al. 2001). Some smaller studies indicate that heavier muscular training with the purpose of increasing muscle strength in localised muscles can be of help, but such training should not be performed in marginally functioning muscles with very large motor units as overload could lead to an increased weakness.

Respiratory problems

PPS patients at risk for respiratory problems are mainly those who have had widespread pareses with truncal involvement and deformities of the spine with scoliosis or kyphosis (Howard et al. 1988, Kidd et al. 1997). The symptoms of respiratory weakness may be dyspnea and shortness of breath related to activity, nightly wake-ups with the sense of breathlessness, day-time tiredness or morning headache. A proper respiratory function investigation should be carried out, including both daytime function and nocturnal

registrations. If a significant muscle weakness in the neck or chest is present, assistive devices like collars or corsets may help to stabilise the most optimal anatomic positions. Non-invasive respiratory aids like Bi-level positive airway pressure (BiPAP) or continuous airway pressure (CPAP) is useful for nocturnal hypoventilation and sleep-related breathing problems. It is important to have in mind that PPS patients never have the same clinical course as patients with motor neuron diseases, and the use of respiratory aids may help the PPS patient for years, keeping the respiratory function rather stable (Bach 1995, Bergholtz et al. 1988). Overweight increases the risk of nocturnal hypoventilation, and proper advice including life style changes and regular physical for weight control is advocated to avoid overweight related respiratory problems. If an activity related dyspnea is present, proper cardiovascular fitness training like endurance training is indicated as overweight and reduced cardiovascular fitness is a common cause.

Pain and fatigue

PPS patients are at risk for experiencing different pain conditions (Conde et al. 2009). Muscular pain may be caused by overuse, and can be relieved with proper devices, reorganising of day-time activities, and physiotherapy. Tendinitis could be helped with proper physiotherapy and non-steroid anti-inflammatory drugs. When analgetics are needed, the general guidelines for pain treatment should be followed. Training in warm climate and in warm water is known to be helpful when muscular pain is present.

The background for the generalised fatigue and sense of exhaustion reported by the PPS patients is not fully understood, but probably is a more physiological than mental phenomenon (Ostlund et al. 2008). This symptom is by many patients reported to be most difficult part of their PPS to cope with. Some interventions and adjustments of daily living can alleviate the situation. Energy economising, work simplification skills and frequent periods of rest are helpful, and properly fitted aerobic training programmes in combination with lifestyle changes have shown to improve both pain and fatigue.

Other therapeutic interventions

Several medications have been tried out in PPS, but the majority of the trials have ended with the same meagre results. Amantadine and modafinil were not effective in PPS-fatigue. Steroids and coenzyme Q10 did not improve muscle strength, and pyrodostigmine did not improve muscle strength or fatigue. One open study indicated that lamotrigine improved quality of life, but this has not been investigated in placebo-controlled trials. Intravenous immunoglobulin (IvIg) has been tried in three therapeutic intervention studies, where the results are diverging when it comes to which symptoms are relieved (muscle strength, fatigue, pain, and quality of life), and can not be recommended as standard treatment up to day (Farbu et al. 2006).

Summary

In summary, PPS is a diagnosis that may lead to increasing disability in previous polio patients, but the clinical course is characterized with a modest deterioration. The diagnosis is mainly a clinical diagnosis with no specific tests, but the diagnostic process should emphasize the exclusion of the possible differential diagnoses that could lead to other specific therapies. There is no cure for PPS, but symptoms can be alleviated to a large degree by offering individually fitted training programmes, particularly in warm water, and the use of assistive devices. In the presence of respiratory insufficiency, non-invasive respiratory aids are helpful.

Life-style changes and regular activity can ameliorate pain and fatigue, and group training can give an additional positive psychosocial effect.

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