

Post-Polio Syndrome – Diagnosis and Management

Post-polio syndrome is characterised by muscular weakness, pain, and fatigue several years after the acute polio. The first known clinical description dates back to 1875 when Raymond and Charcot reported a 19-year old tanner with previous infantile paralysis who presented with new paresis and atrophy in his shoulder. The subject was not investigated further during the next decade, as paralytic polio was considered to be a three-phase illness with acute pareses followed by a recovery period, and then a life-long stable phase. However, many patients with previous polio experienced a functional decline later on with new symptoms like pain, fatigue, muscle weakness, sleeping problems, and cold intolerance. Halstead introduced the term post-polio syndrome in 1986, describing the symptoms experienced by many polio survivors after decades with stable function. Halstead revised his criteria in 1991¹ (Table 1) where new muscle weakness was included as an obligatory criterion, with or without other symptoms like pain, cold intolerance, and fatigue.

The background and exact pathogenesis for the post-polio syndrome is still not known, although several theories have been explored. Ongoing viral replication or virus reactivation was suggested, but has not been confirmed.² An ongoing inflammation has been found in the spinal cord, and recent studies have shown elevated levels of inflammatory cytokines in the spinal fluid.³ The ageing process may be a contributing factor but does not explain all clinical aspects as post-polio syndrome has been found in patients before the age of 50 years.⁴ At higher age, loss of motor neurones and diminishing motor units take place at a higher rate in post-polio patients compared to patients with normal neuromuscular function.⁵ Hence, the increased muscle weakness seems to be a result of both an ongoing loss of motor neurones and a diminished ability to maintain their neurogenic supply to enlarged muscle fibres.

The prevalence of post-polio syndrome has been reported to be between 20-80% depending on the polio population being studied and the diagnostic criteria being used.

Diagnosis

Post-polio syndrome is an exclusion diagnosis (Table 1). A careful medical history and clinical examination are necessary to rule out all other conditions that may cause the same symptoms. At clinical examination, signs of lower motor neurone involvement should be present with flaccid muscle weakness or atrophy and diminished tendon reflexes. However, patients who have lost 50% of their motor neurones within one segment can still have a normal clinical picture.⁶ This means that subclinical motor neurone involvement may be present, and new muscle weakness may occur in apparently non-affected muscles. EMG can be of help to sort out other neurological and muscular illnesses, and can also establish a motor neurone involvement compatible with previous paralytic polio. However, EMG cannot distinguish between stable polio sequelae and new muscle weakness, and the major role of neurophysiology is to confirm previous polio and exclude other neuromuscular disorders.⁷ Disorders not related to the patient's previous polio may be co-existing: neurological and rheumatological disorders, cardiovascular and thyroid disorders, and depression. A thorough investigation to sort out such co-existing disorders is necessary, as these disorders need specific treatment. They should be treated in the same way as in patients without a history of previous polio.

Management

Muscular weakness and fatigue

Even though post-polio syndrome affects only between 20-50% of polio survivors, it is important to have this in mind as polio patients in general report more pain, fatigue, sleeping problems and muscle weakness than healthy controls, and they rate their health lower.⁸⁻¹⁰ This is irrespective of the presence of a post-polio syndrome or not. For all practical purposes today's management and treatment will be similar for patients with and without post-polio syndrome (PPS).

No specific medical treatment for PPS has been proven to be effective;

- Pyridostigmine and steroids have been tried in randomised studies without any positive effect with respect to muscle strength and fatigue;^{11,12}
- Muscle training at aerobic levels without maximum exercise is useful to maintain muscular function and ameliorate fatigue¹³ and muscular training in warm water seems to be particularly useful.¹⁴ Systematic training programmes in a warm climate are more effective than identical training programmes in a cold climate;¹⁵
- Reorganisation of daily life activities with short breaks (thereby conserving energy expenditure) may help to counteract fatigue. In addition, properly fitted assistive devices (i.e. intermittent use of wheel chair) can help in this respect;
- Inactivity increases the risk of obesity, diabetes, cardiovascular, and musculoskeletal problems and so polio patients who take part in physical activity have significantly less symptoms than physically inactive patients.¹⁶ All polio patients with or without post-polio syndrome should therefore be advised to take part in physical activity, but they should not be performing static muscular training at maximum effort (anaerobic level) and they should allow intermittent breaks.

Disorders related to previous polio

Patients with previous polio are more prone to developing overuse symptoms and disorders like soft tissue inflammation, arthrosis, spinal degenerative disorders, and nerve entrapment due to asymmetric weight bearing. Carefully fitted orthoses, casts, splints, and other assisting devices may prevent or delay these symptoms.^{10,17} If severe arthrosis is present, surgical treatment with hip or knee replacement should be considered in a similar fashion as to that done for patients without previous polio, but the post-operative rehabilitation period



Elisabeth Farbu is a consultant in the Department of Neurology, Stavanger University Hospital, Norway. She has a PhD in clinical neurology from the University of Bergen. Her clinical and scientific interests are in neuromuscular disorders and post-polio syndrome.

Correspondence to:

Elisabeth Farbu,
Department of Neurology,
Stavanger University Hospital,
Norway.
Email: elfa@sir.no



Table 1: Criteria for the diagnosis of post-polio syndrome¹

1. A prior episode of paralytic polio confirmed by history, physical exam, and typical findings on EMG
2. Standard EMG evaluation demonstrates changes consistent with prior anterior horn cell disease: increased amplitude and duration of motor unit action potentials, increased percentage of polyphasic potentials and, in weak muscles, a decrease in the number of motor units on maximum recruitment. Fibrillations and sharp waves may or may not be present.
3. A period of neurologic recovery followed by an extended interval of functional stability preceding the onset of new problems. The interval of neurologic and functional stability usually lasts for 20 or more years.
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.
5. Exclusion of medical, orthopaedic, and neurologic condition that might cause health problems listed above.

may be prolonged and require particular training programmes. Severe scoliosis or degenerative spine changes should be considered for surgery if the neurological and/or respiratory function is threatened. Sleeping problems can be related to the frequency and intensity of pain, and proper pain management may improve sleep.¹⁰ Sleeping problems can also be a part of nocturnal hypoventilation and patients with chest wall deformities and respiratory muscle weakness are at risk of developing respiratory insufficiency.¹⁷ Co-existing obesity increases the risk. Symptoms on respiratory insufficiency can manifest as daytime sleepiness, morning headache, sleeping problems, fatigue, dyspnea, recurrent respiratory tract infections, and if not treated properly, secondary right sided heart failure (cor pulmonale). If the respiratory insufficiency is due to a mechanical deficit only (i.e. muscle weakness) with intact lung tissue, the effect and prognosis for using artificial ventilatory aids is excellent. In

most cases, artificial ventilation is only needed during nighttime, and non-invasive ventilators are the first choice. Biphasic positive-pressure ventilators (BIPAP) and nasal intermittent positive-pressure ventilators (NIPPV) are often used in polio-related respiratory insufficiency with good results. General precautions like stopping smoking, a reduction in weight if obese, the use of influenza- and pneumococcal vaccines, and aggressive treatment of respiratory tract infections are particularly important for these patients.

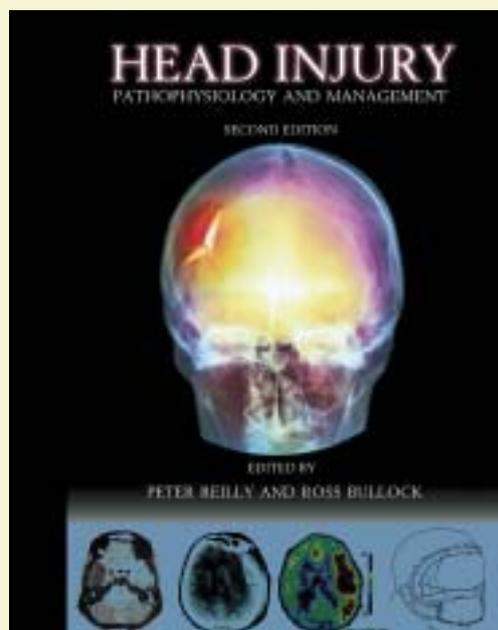
In conclusion, PPS is an exclusion diagnosis based on a thorough investigation in a patient with a previous history of polio. Proper treatment of other disorders, both polio-related and non-polio-related, is important with the management of PPS being primarily based on physical therapy and muscular training along with intermittent ventilatory support if necessary.

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