Background

Post Polio Syndrome (PPS) is a neurological condition that can occur in a high percentage of people with prior Polio. In the UK it is estimated there are 120,000 Polio survivors; UK surveys indicate up to 80% prevalence of PPS in Polio survivors.

Definition of PPS: "Gradual or sudden onset of progressive and persistent new muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalised 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."

Impact of Polio

In the acute disease, the Polio virus attacks the motor neurons resulting in varying degrees of muscle paralysis and weakness.

The brainstem was also involved, particularly the reticular formation, vestibular nuclei, cerebellar nuclei, hypothalamus and thalamus, and the cranial nerves. Recovery involved the reinnervation of orphaned muscle fibres by nearby neurons, resulting in enlarged motor units and muscle fibre hypertrophy.

Muscles that appear unaffected can have significant subclinical denervation.

Impact of Post Polio Syndrome

Pathology: The leading causal hypothesis is that excessive metabolic stress on remaining motor neurons, possibly due to long term muscle overuse, results in distal degeneration of terminal axons and eventually loss of motor neurons. The rate of loss is about twice that of normal aging.

After acute Polio there is ongoing denervation and reinnervation, but with time the ability to reinnervate orphaned fibres diminishes. This can occur in muscles that appear normal but have subclinical denervation.

Treatment: There are no medications currently available proven to reverse muscular atrophy, improve neuromuscular strength or relieve the neuromuscular fatigue of PPS.

About the Quick Reference Guide

This guide summarises the key points contained in the British Polio Fellowship 'Post Polio Syndrome, a guide to management for health care professionals.

www.britishpolio.org.uk/pps-report

Further information:

www.mapofmedicine.com/
www.patient.info/doctor/post-polio-syndrome
www.europeanpolio.eu/healthcarepro_EFNS.php

History Checklist

Polio infection and treatment	
Polio infection and treatment (if known)	Age at onset, Severity and progression Respiratory impact in acute phase Acute management Age at best recovery Maximum functional recovery
History since Polio	Change in functional ability, degree, speed and nature of change
Present status	Recent changes in activity levels, employment, environment, nutritional status, general health and lifestyle

Symptoms Checklist

Full symptoms checklist - onset, duration, location, triggers
Fatigue - neuromuscular, mental and general (including endurance)
Weakness/atrophy
Pain - muscular, joint, soft tissue
Respiratory status
Dysphagia/swallowing status (including risk of aspiration pneumonia)
Sleep quality
Psychological well-being (anxiety, stress..)
Mobility (inc. endurance)
Spinal or joint deformities (inc. history of change)
Bowel/urinary function
Activity level - to assess overuse (activity diary)
Falls history/frequency
Medication review (for adverse impact on symptoms)
Weight history
Smoking history
Daily activities function - self-care ability (inc. cooking, shopping)
Aids, appliances and adaptations used

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Post Polio Syndrome

A guide to management for healthcare professionals

Quick Reference Guide

Presentation

A Polio survivor may present with new weakness, pain, neuromuscular and/or general fatigue, respiratory difficulties or sleeplessness and they may or may not refer to prior Polio.

As early symptoms such as fatigue are commonly experienced, Polio survivors may live with increasing problems for a considerable time before seeking medical help.

Coping mechanisms used for years may no longer be effective.

New symptoms may reawaken the emotional distress experienced during the initial disease making discussion difficult for patients.

Assessment (see History and Assessment checklists)

Effective assessment needs a full history and symptom review and should take into account their early experiences.

Chronic or new overuse of affected or unaffected areas can trigger or exacerbate PPS symptoms; activity levels in paid or unpaid work, sports/fitness or daily life need to be assessed.

Fatigue, weakness and pain can also be caused by other conditions which need to be excluded. The presentation may resemble other conditions risking misdiagnosis.

Commonly used assessment tools such as manual muscle testing can give a false impression of strength as one-off movements may be strong but PPS patients often show poor endurance over repeated movements or during sustained movements. These should be used alongside other methods such as variances from a well-documented baseline.

Respiratory insufficiency may develop insidiously, initially at night, in individuals who appeared previously unaffected, with symptoms such as unrefreshing sleep, frequent wakening, waking with headaches, daytime sleepiness, waking gasping or difficulties lying flat. Obstructive sleep apnoea is also common in PPS patients. These complications are most prevalent when there is an additional strain on the respiratory muscles such as respiratory infections, periods of immobility, surgery, pregnancy or obesity. Assessment for respiratory complications should be considered both for those with night-time symptoms or daytime effort-related shortness of breath.

Difficulty with swallowing is a potentially serious problem for Polio survivors and those with PPS; if suspected referral to a Speech and Language Therapist is strongly recommended. This should advise that any tests involving observing muscle action need to involve several repetitions to assess the impact of abnormal neuromuscular fatigue. Patients should be assessed for risk of aspiration pneumonia.

Cold intolerance is common but underreported and can affect motor function and comfort.

Management interventions

A multidisciplinary approach: Strongly recommended, involving GPs, physiotherapists, occupational therapists, orthotists and other allied health professionals with referral to specialist consultants as required (see referral critiera).

Energy management techniques: Can alleviate the symptoms of neuromuscular and general fatigue and reduce pain. Referral to a specialist physiotherapist or occupational therapist with experience in managing neurological conditions is recommended for assessment and training in these techniques:

- **Pacing activity** ♦♦♦ Effective in reducing neuromuscular fatigue and pain and may improve performance for some.
- **Energy conservation** ♦♦♦ Adapting, simplifying and prioritising daily tasks can preserve energy and avoid neuromuscular fatigue and pain.
- **Aids and appliances** ♦♦♦ These can help in energy management. PPS patients may need encouragement to use aids, which may remind them of the original Polio.

Orthotics: ♦♦♦ An orthotics review by an experienced specialist can help reduce overuse and misuse and reduce energy cost of walking by optimising orthoses and footwear. Optimised orthotics may help reduce falls.

Respiratory management: ♦♦♦ For confirmed respiratory insufficiency, due to weak respiratory muscles or scoliosis, respiratory support in the form of non-invasive ventilation, normally bilevel positive pressure (BiPAP), usually only at night, is the recommended treatment. Continuous positive airway pressure (CPAP) is used for obstructive sleep apnoea. If secretion retention is an issue, a respiratory physiotherapist can advise on positions of treatment and also devices such as cough assist that might be useful in the case of a persistently weak cough.

Exercise and physical activity: ♦♦! Once good energy management is established, these can be considered under the guidance of a specialist physiotherapist with experience in management of neurological conditions, to strengthen muscles where possible and help improve cardiovascular health.
Caution: safe effective exercise for people with PPS requires an individually tailored non-fatiguing, pain-free programme and careful monitoring to avoid overuse.

Pharmacological management: ♦ As there is no medication proven to reverse the progress of PPS, pharmaceutical interventions are aimed at alleviating symptoms such as fatigue, pain and poor sleep. To date none have been proven to reduce the fatigue and neurological weakness of PPS. Some medications can alleviate pain and are used after energy management techniques have been tried.

Side effects of some medications can worsen PPS symptoms such as increased weakness, fatigue, respiratory depression or muscle pain/cramps.

Anabolic steroids are not recommended to improve muscle bulk as the risks due to side effects greatly outweigh the potential benefits. Metabolic stimulants such as L-carnitine and co-enzyme Q10 have been studied but not been proven to be effective.

The efficacy and safety of intravenous immunoglobulin is currently the subject of a multi-centre randomised controlled trial.

Vaccination: As with other chronic diseases, pneumococcal and influenza vaccinations are prudent to offer given the respiratory issues already outlined.

Psychological therapies: ♦♦ May be helpful in treating symptoms such as depression and anxiety. They may also enhance the efficacy of physical interventions by promoting behaviour change and improving the ability to cope with physical symptoms.

Nutrition and weight management: ♦♦! Dietary advice to optimise nutritional status may also support functional status. Weight loss may help reduce neuromuscular fatigue and pain, taking account of probable low proportion of lean mass and low mobility. Underweight may be due to poor diet, swallowing issues or difficulty shopping/cooking and may impact PPS symptoms. **Caution:** if advising exercise, see the advice on a safe effective programme.

Other conditions: Management also needs to include treatment of other conditions which occur more commonly in Polio survivors such as osteoporosis and peripheral neuropathies. If a PPS patient smokes, smoking cessation advice and support can prevent worsening respiratory function and vascular complications.

Review: As PPS is a progressive condition, regular review, ideally once a year, is essential to identify increasing muscle weakness, fatigue and/or pain and to adapt the individual's management programme accordingly.

The strength of the evidence is indicated: ♦♦♦ strong, ♦♦ for some, ♦ may help; ! caution advised

Criteria for referral to secondary care

- Development of new neurological symptoms
- Progression or deterioration of longstanding neurological symptoms
- Uncertainty regarding the diagnosis of PPS
- Advice about symptom management in PPS, especially where respiratory complications or dysphagia are suspected
- Need for advice on failure of treatment that was previously effective in PPS
- Need for specialist advice on orthotics, biomechanics and orthopaedic problems