



POST POLIO MATTERS



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POST-POLIO SYNDROME This article is for medical professionals.

Post-polio syndrome (PPS) is the onset of new deterioration in function that may occur many years after partial or complete recovery from acute poliomyelitis^[1]. PPS comprises various possible symptoms such as fatigue, weakness, joint and muscle pain, reduced respiratory function or dysphagia. PPS does not necessarily imply progressive deterioration and can be helped by treatment. Management requires careful assessment and a multidisciplinary approach.

Definition/diagnosis

Accepted criteria for diagnosis of PPS are^[2]:

- History of paralytic poliomyelitis.
- Partial or complete recovery of neurological function followed by a period of stability (usually several decades).
- Persistent new muscle weakness or abnormal muscle fatigability.
- Exclusion of other causes of new symptoms.

The diagnosis is based on two or more of the following health problems occurring after the stable period: extensive fatigue, muscle and/or joint pain, new weakness in muscles previously affected or unaffected, new muscle atrophy, functional loss, cold intolerance^[3].

There is no universal definition of PPS but most sources use one similar to the above. Some experts suggest that PPS may be diagnosed in those where there is no clear history of paralytic polio but where the past history or investigations suggest that previous polio is likely.

Aetiology^[4]

There is no reactivation of the polio virus infection (patients may need reassurance about this). The exact cause of PPS is unknown:

- There may be new muscle atrophy and weakness relating to changes in motor neurons.
- Overuse or disuse of muscles plays an important role^[5].
- The cause of PPS symptoms may be multifactorial. For example, disabilities resulting from acute polio, combined with activities of daily living, can produce large stresses acting on abnormal muscles and joints. This may lead to other problems which contribute to PPS, such as:
 - Joint deformities.
 - Osteoporosis.
 - Scoliosis.
 - Cervical spondylosis.
 - Peripheral nerve entrapment (can occur with callipers, crutches, etc).

Epidemiology^[1, 5]

PPS affects 15-20 million people worldwide. It is estimated that PPS may occur in 28-75% of patients with previous polio. The time interval is usually around 35 years but may be 8-70 years after the acute polio episode.

Presentation^[5, 6]

Common symptoms include:

- Generalised fatigue.
- Joint and muscle pain.
- New muscle or joint weakness.
- Muscle atrophy.
- Cold intolerance.
- Bulbar symptoms - speech, swallowing or respiratory symptoms.
- Worsening respiratory function - may present as headaches, fatigue or sleep disorder (see below under 'Respiratory and sleep problems in post-polio syndrome').

Any of these can lead to a deterioration in day-to-day functioning. A small change in clinical terms can mean a large one in its effects on daily living.

Assessment

- The most important question to ask is not "can you do this activity?" - eg, climbing stairs - but "**how** do you do it?" This can reveal the functional change - eg, stopping to rest half way, going upstairs by shifting on their bottom^[7].
- A patient questionnaire such as "My Polio Life" can be useful^[8].
- Multidisciplinary assessment may be needed - eg, involving physiotherapist, occupational therapist, neurologist, orthopaedic/orthotic team and respiratory physician.

Bear in mind that various factors can make assessment more difficult^[9]:

- Polio survivors are used to coping and adapting. Hence the importance of asking - and observing - **how** patients carry out each activity.
- Symptoms can vary from day to day and can be affected by recent activity, overuse or rest.
- Patients may deliberately allow for a stressful hospital visit day by resting beforehand. This can give a falsely good picture. Ask what symptoms are like "on bad days".
- Results can appear normal if patients are given long rests between tests, or if only the best result is recorded.
- Respiratory and sleep problems are easily missed (see below under 'Respiratory and sleep problems in post-polio syndrome').

Anecdotally, patients with PPS have had their symptoms dismissed after apparently normal or near-normal results of tests, such as lung function or muscle power.

Investigations

These will depend on symptoms, but could include:

- Muscle tests - but be aware that simple tests of isometric muscle strength may be insensitive^[7, 10].
- Respiratory investigations (see below under 'Assessment of respiratory problems').
- Sleep studies (see below under 'Assessment of respiratory problems').
- Swallowing studies - eg, barium swallow.
- Investigations to exclude other causes.

Differential diagnosis

Other causes of fatigue or weakness - eg:

- Multiple sclerosis.
- Motor neurone disease.
- Myasthenia gravis.
- Other types of neuropathy or myopathy.
- Systemic conditions - eg, anaemia, chronic infection, hypothyroidism, collagen disorders, medication side-effects.
- Other causes of pain - eg, arthritis, bursitis, tendinopathy.
- Myalgias - polymyalgia rheumatica, fibromyalgia.

Management^[4]

EDITOR'S NOTE

August 2018 - Dr Hayley Willacy recommends the guide to management of post-polio syndrome for healthcare professionals^[11]. One of the key principles involves using energy management techniques to alleviate the symptoms of neuromuscular and general fatigue and reduce pain. These may include:

- Pacing activity, which is effective in reducing neuromuscular fatigue and pain and may improve performance for some.
- Referral to a specialist physiotherapist or occupational therapist with experience in managing neurological conditions is recommended for assessment and training in the technique.
- Energy conservation: adapting, simplifying and prioritising daily tasks can preserve energy and avoid neuromuscular fatigue and pain. Pacing can be part of this technique; occupational therapists can help optimise lifestyles, assistive devices and environments as well as providing information on how to apply pacing.

There is insufficient evidence to draw definite conclusions about the effectiveness of interventions for PPS. There is evidence that intravenous immunoglobulin, lamotrigine, muscle strengthening exercises and static magnetic fields may be beneficial but further investigation is needed to clarify whether any real and meaningful effect exists.

A multidisciplinary approach is helpful^[12]. Some important aspects of management are:

- The correct balance of rest and exercise is essential^[5]:
 - Avoid overuse, as too much exercise causes increased weakness and fatigue in damaged muscles.

- Graded exercise is beneficial - this should probably be broken up by periods of rest.
- Non-swimming exercise in warm water often helps.
- Many polio survivors are used to leading active lives and, to some extent, ignoring their disability. Adapting to PPS and the need for more rest may require lifestyle and employment changes .
- Orthopaedic and orthotic management of skeletal problems - eg:
 - Simple supports for knee, ankle and cervical spine can improve function.
 - Replace damaged aids.
- Muscle pain:
 - Physical treatments, such as warmth/cold, massage, passive stretching.
 - Transcutaneous nerve stimulation.
 - Acupuncture.
- Anaesthesia requires special considerations^[14].
- Nutrition:
 - Good nutrition and weight control.
 - Some patients find that a high-protein diet is helpful (eg, the post-polio institute 'hypoglycaemia diet') .
- Specific treatment of other problems - eg, dysphagia and respiratory and sleep problems (see below under 'Respiratory and sleep problems in post-polio syndrome').

Respiratory and sleep problems in post-polio syndrome^[15]

Importance

- Respiratory problems in PPS are an important cause of symptoms and complications, including sleep disorders.
- They may be under-diagnosed or inadequately assessed.
- Treatment can improve both quality of life and prognosis.

Aetiology

Respiratory problems in PPS may be due to one or more of:

- Respiratory muscle weakness.
- Bulbar impairment - this may affect control of the upper airway or the respiratory cycle. If the upper airway is affected, there may be obstructive sleep apnoea.
- Skeletal deformity - scoliosis or chest wall stiffness.
- Other pathology - eg, chronic obstructive pulmonary disease (COPD), asthma, obesity.
- Aspiration - if swallowing is affected.

All these are likely to worsen during sleep. The pattern of respiratory impairment may be hypoventilation, obstructive sleep apnoea, or both.

Symptoms

Respiratory failure can develop insidiously - symptoms may be subtle or unnoticed. Breathlessness may not be a symptom in patients with limited mobility. Possible symptoms are:

- Sleep disruption, eventually leading to insomnia, daytime sleepiness or fatigue.
- Morning headaches, irritability, poor concentration, anxiety or depression.
- Abnormal sleep movements, nocturnal confusion, vivid dreams.
- Breathlessness which may be positional.
- Weak cough, and chest infections.

Signs

These may be subtle - possible signs are:

- Unexplained tachypnoea.
- Use of accessory muscles.
- Abdominal paradox - this is inward movement of the abdomen on inspiration while the upper chest expands:
 - May be best seen with the patient supine during a sniff manoeuvre. When upright, it can be missed, as the diaphragm passively descends at the beginning of inspiration.
- Severe, untreated nocturnal hypoxaemia can cause pulmonary hypertension, giving signs such as raised JVP and ankle oedema.

Assessment of respiratory problems

- Listen to the patient's story and preferences.
- Assess:
 - Voice and cough.
 - Chest deformity.
 - Patients in realistic situations - eg, observe them doing repeated tests or actions, and doing everyday actions in which they may be using the necessary breathing muscles to achieve another task.
- Investigations:
 - Peak flow and cough peak flow.
 - Spirometry:
 - Both seated AND supine spirometry are needed.
 - A sensitive indicator of respiratory muscle weakness is reduction in maximal inspiratory pressure.
 - Oximetry (and possibly capnography).
 - Sleep study (polysomnogram).
 - ECG and CXR if appropriate.

Full sets of lung function tests and arterial blood gases may not be helpful in this scenario, unless intrinsic lung disease is suspected.

Management of respiratory problems

There are various options - choice will depend on the patient's individual situation and preferences.

Night-time mechanical ventilation is often used. This helps by resting the respiratory muscles at night and preventing deterioration of respiratory function during sleep. It also treats the secondary sleep disorder.

Supportive measures include:

- Not smoking.
- Avoiding sedatives and alcohol.
- Optimal weight and nutrition.
- Pneumococcal and influenza vaccination.
- Postural support if needed.
- Prompt treatment of chest infections.
- Techniques such as assisted cough or glossopharyngeal breathing ('frog breathing').
- Chest expansion exercises.

Assisted breathing options:

- Non-invasive ventilation (NIV), also called non-invasive intermittent positive pressure ventilation (NIPPV), is often useful (see below).
- Rocking bed:
 - This helps breathing by rocking a patient consecutively head up and head down. It is surprisingly effective, especially where muscle weakness is mainly diaphragmatic.
- Pneumobelt:
 - This gives intermittent abdominal pressure ventilation and is useful for daytime assistance.
- Negative pressure ventilation:
 - Examples are tank ventilators (iron lung), jacket ventilators (Tunncliffe), and cuirass ventilators. The devices are cumbersome and mainly used where NIV is not tolerated, or to provide 'respite' from NIV.
- Tracheostomy ventilation.

NIV and 'bi-levels' explained

NIV increases alveolar ventilation. It is provided by a portable ventilator and a tightly-fitting nasal or facial mask or nasal 'pillow'^[15].

NB: NIV is NOT the same as continuous positive airway pressure (CPAP). CPAP is useful for obstructive sleep apnoea because it maintains the upper airway. It is not normally indicated for hypoventilation from respiratory muscle weakness.

People with neuromuscular disease may have difficulty breathing in, so require NIV with higher inspiratory than expiratory pressures. This can be provided using a 'bi-level' ventilator:

- Bi-level ventilators were developed by modifying CPAP. The inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP) settings are adjusted

separately.

- The difference between IPAP and EPAP is called the span. For example, a patient may require an IPAP of 14 and an EPAP of 3.
- Sensitive flow triggers enable normal breathing to be supported. There may be a back-up control to provide ventilation if respiratory effort fails to trigger a breath.

Prognosis^[2]

The symptoms of post-polio syndrome are slowly progressive, with periods of stability lasting 3-10 years.

Prevention

Prevention of acute polio infection is discussed elsewhere. See the separate [Polio and Polio Vaccination](#) article.

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ARTICLE INFORMATION

- **Last Checked** 24 June 2016
- **Next Review** 23 June 2021
- **Document ID2637 (v25)**

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