

# Late effects of polio

A guide to management for medical professionals



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## Background

The World Health Organisation estimates there are 20 million people worldwide currently living with some degree of disability caused by poliomyelitis (1). Not all polio survivors were diagnosed with paralytic polio. However, 99% of all polio survivors may have experienced sub-clinical damage that becomes more evident with age.

The late effects of polio is a neuromuscular condition that can occur in a high percentage of people with prior exposure to polio. In Australia, it is estimated there are tens of thousands of polio survivors. International surveys indicate there is up to 80% prevalence of late effects of polio in polio survivors.

As late polio symptoms progressively worsen, patient coping mechanisms used for many years may no longer be effective. Over time, partners and carers of polio survivors may find themselves providing more assistance to the polio survivor. As the progression may be gradual, the polio survivor and carers may not initially understand or appreciate the magnitude of the impact of the late effects of polio on their lives.

## 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 is also involved in the initial infection, particularly the reticular formation, vestibular nuclei, cerebella nuclei, hypothalamus and thalamus, and the cranial nerves. Recovery involves 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, the effects of which usually emerge as the polio survivor ages. Beyond the acute phase there is a period of stable non-progressive level of disability for several decades.

### The late effects of polio:

*a 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, trauma, or surgery. Less commonly, symptoms attributed to late effects of polio include new problems with swallowing or breathing.*

## Impact of the late effects of polio

**Pathology:** The leading causal hypothesis is that excessive metabolic stress on the 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 polio ongoing denervation and reinnervation occurs, but with time the ability to reinnervate orphaned fibres diminishes. This can occur many years later in muscles that appear normal but have subclinical denervation.

**Treatment:** There are no medications currently available to prevent or reverse muscular atrophy, improve neuromuscular strength or relieve the neuromuscular fatigue of late effects of polio.

## Presentation

Polio survivors may present with new weaknesses, pain, neuromuscular and/or general fatigue, respiratory difficulties, or sleeplessness. Although affected patients may refer to their prior episode of polio, many do not willingly disclose past exposure due to past significant stigma they may have experienced due to polio infection.

As early symptoms of the late effects of polio, such as fatigue, are commonly experienced by the general population, polio survivors may live with significant and worsening problems for a considerable time before seeking medical help.

New symptoms may also reawaken the emotional distress experienced during the initial disease, making discussion difficult for patients. This emotional distress may extend to traumatic experiences in hospitals and health care facilities, and a general wariness or mistrust of medical advice.

### Suggested recommendations for GPs include:

- › *Including a 'tick box' on the new patient registration form for your practice promoting patients about their polio history*
- › *Conduct an annual health check to review mobility, swallowing and respiratory problems of polio survivors*
- › *Check current psychological status of polio survivors.*

## Diagnosis

The diagnosis of late effects of polio relies nearly entirely on clinical information. There are no laboratory tests specific for this condition and symptoms vary greatly among individuals.

An extensive work-up including laboratory tests, imaging studies, cerebrospinal fluid sampling, detailed electrophysiological evaluation, and muscle biopsies may be required to exclude alternative diagnoses. The diagnostic criteria for Post-Polio Syndrome was first proposed by Halstead in 1991 (3) and evolved over time to the current March of Dimes diagnostic criteria (4, 5) which include:

1. Prior paralytic poliomyelitis with evidence of motor neuron loss, as confirmed by history of the acute paralytic illness, signs of residual weakness and muscle atrophy on examination, or signs of denervation on EMG.
2. A period of partial or complete functional recovery after acute paralytic poliomyelitis, followed by an interval (usually 15 years or more) of stable neuromuscular function.
3. Gradual onset (rarely abrupt) progressive and persistent new muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalised fatigue, muscle atrophy, or muscle and joint pain. Onset may at times follow trauma, surgery, or a period of inactivity. Less commonly, bulbar dysfunction or respiratory weakness occurs.
4. Symptoms that persist for at least a year.
5. Exclusion of alternative neuromuscular, medical, and orthopaedic problems as causes of symptoms.

## Diagnostic considerations:

1. Anaemia
2. Chronic infection
3. Collagen disorders
4. Deconditioning
5. Depression
6. Hypothyroidism
7. Infectious myopathy
8. Myasthenia gravis
9. Deconditioning and weakness with the ageing process
10. Weight gain



## Differential Diagnoses <sup>5</sup>

- › Amyotrophic Lateral Sclerosis in Physical Medicine and Rehabilitation
- › Hypothyroid Myopathy
- › Multiple Sclerosis

## History Checklist

Polio infection and treatment (if known)	<ul style="list-style-type: none"><li>› Age at onset, severity and progression</li><li>› Respiratory impact in acute phase</li><li>› Acute management</li><li>› Age at best recovery</li><li>› Maximum functional recovery</li></ul>
History since polio	<ul style="list-style-type: none"><li>› Change in functional ability, degree, speed and nature of change</li></ul>
Present status	<ul style="list-style-type: none"><li>› Recent changes in activity levels, employment, environment, nutritional status, general health and lifestyle</li></ul>

## Assessment

Effective assessment requires a full history and review of symptoms, including:

- › **Early experiences of polio.** Explore the physical and psychological impact on their lives. Identify areas affected in the original polio episode and extent of recovery. Document any residual long-term effects of initial polio episodes, individuals who have very severe residual weakness, individuals who had early bulbar respiratory difficulty in the acute illness, and persons who were older when they contracted acute polio. Late polio symptoms also tend to occur first in weaker muscles.
- › **Natural history of presentation.** Document and differentiate symptoms of late polio, including their pattern and progression.

## Pattern and impact of physical activity

Chronic overuse of affected or unaffected muscles can trigger or exacerbate symptoms. Health professionals need to make enquiries into activity levels in paid or unpaid work, sports/fitness training or daily living.

Enquiries need to include any changes to levels or intensity of physical activity.

**Fatigue, weakness and pain.** Exclude other possible causes of fatigue. The presentation of fatigue in a polio survivor patient may resemble other conditions and risk potential misdiagnosis.

Commonly used assessment tools such as manual muscle testing can give a false impression of strength as one-off movements may be strong. However, polio survivors often show poor endurance over repeated movements or during sustained activity.

**Respiratory insufficiency** may develop insidiously, initially at night, in individuals who appeared previously unaffected, although respiratory disorders are most prevalent in patients with residual muscle weakness. Symptoms may include unrefreshing sleep, frequent waking, waking with headaches, daytime sleepiness, waking gasping or difficulties lying flat. Obstructive sleep apnoea is also common in polio survivors.

These complications are most prevalent when there is an additional strain on the respiratory muscles such as respiratory infections, periods of immobility, surgery 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 people living with late effects of polio. Common symptoms include increased swallowing time and food catching in the throat. Weakness in pharyngeal constrictor muscles are common in late polio, even in the absence of symptoms. Patients should also be assessed for risk of aspiration pneumonia.

If swallowing difficulties are suspected, referral to a speech therapist is strongly recommended. The referral should advise that any tests involving observing muscle action need to involve several repetitions to assess the impact of abnormal neuromuscular fatigue.

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

### **Walking and postural disturbances.**

Difficulty in walking can be due to progressive weaknesses and joint instability. Walking problems are more common in people who have used mobility assistance devices in the past and have discarded them. Postural problems can be related to spinal malformations and weaknesses in truncal muscles.



## Recommended management interventions

**A multidisciplinary approach:** ◆◆◆◆ The ideal approach involves coordinated multidisciplinary treatment often involving rehabilitation specialist doctors, GPs, physiotherapists, occupational therapists, orthotists, other medical specialists and allied health professionals as required (see referral criteria).

**Energy management techniques:** Successful energy management techniques, best taught through referral to a specialist physiotherapist or occupational therapist, can help alleviate the symptoms of neuromuscular and general fatigue and reduce pain. Techniques include:

- ▶ **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. Late effects of polio 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 respiratory insufficiency, confirmed by a respiratory specialist, 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 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 late effects of polio requires an individually tailored non-fatiguing, pain-free program and careful monitoring to avoid overuse.*

**Pharmacological management:** ◆ As there is no medication proven to reverse the progress of late effects of polio, 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 late effects of polio. Some medications can alleviate pain and are used after energy management techniques have been tried.

**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.

The strength of the evidence is indicated:

◆◆◆◆ very strong. ◆◆◆ strong. ◆◆ for some. ◆ may help, caution advised!

Side effects of some medications can worsen symptoms of late effects of polio including increased weakness, fatigue, respiratory depression or muscle pain/cramps:

- › **Anabolic steroids:** 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 Coenzyme Q10 have been studied and used in some parts of Australia but have not been proven to be effective.
- › **Intravenous immunoglobulin:** The efficacy and safety of intravenous immunoglobulin is currently the subject of a multi-centre randomised controlled trial.
- › **Statins:** Many polio survivors have read that some medication classes are bad for them, including statins. While some statins may lead to muscle fatigue and pain, health professionals are best placed to provide advice to patients with late effects of polio and to monitor any potential side effects of statins.
- › **Anaesthetics:** Anaesthetics can cause gastro-oesophageal reflux, tachyarrhythmia and, sometimes, difficulty maintaining blood pressure.
- › **Sedatives:** Patients with the late effects of polio are nearly always very sensitive to sedative medications, and emergence can be prolonged. This is probably due to central neuronal changes, especially in the Reticular Activating System, from the original disease.
- › **Analgesics:** People with late effects of polio may experience breathing problems. As opioid based analgesics can depress breathing, precaution should be taken when prescribing them in a patient with the late effects of polio.
- › **Vaccination:** Pneumococcal and influenza vaccinations are recommended.

**Nutrition and weight management:** ♦♦♦ Dietary advice to optimise nutritional status may also support functional status of the person with the late effects of polio. Weight loss may help reduce neuromuscular fatigue and pain, taking account of probable low proportion of lean mass and low mobility. A person being underweight may be due to poor diet, swallowing issues or difficulty shopping/cooking and may impact late effects of polio symptoms.

*Caution: if advising exercise, ensure you recommend a safe and effective program.*

**Other conditions:** Management also needs to include treatment of other conditions which occur more commonly in polio survivors such as osteoporosis and peripheral neuropathies. Smoking cessation advice and support may help prevent worsening respiratory function and vascular complications in a person with the late effects of polio.

**Regular monitoring:** As the polio survivor with features of the late effects of poliomyelitis will potentially decline functionally with time, regular annual review helps identify changing and progressive symptoms and early opportunities to intervene and adapt the individual's management program accordingly.

## Criteria for referral to secondary care

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

## Resources and References

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This Post Polio booklet has been officially recognised as an Accepted Clinical Resource by The Royal Australian College of General Practitioners.



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