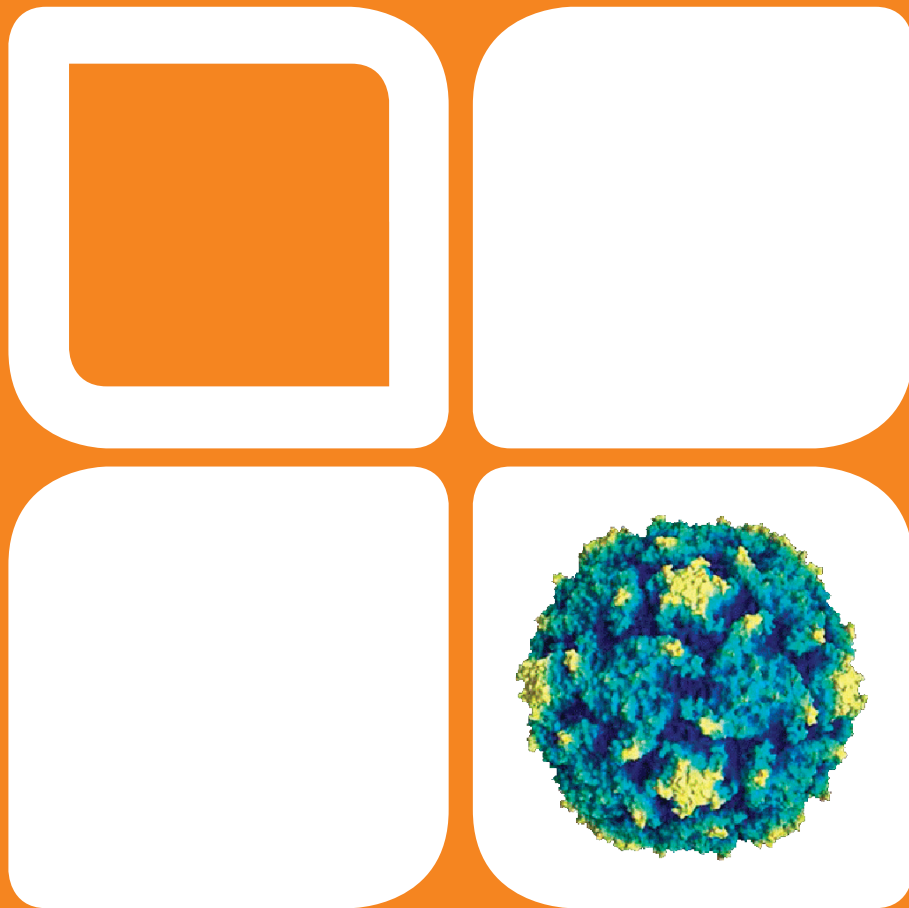


# The Late Effects of Polio:

Introduction to Clinical Practice



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## 1. Introduction: What are the Late Effects of Polio (LEoP) and Post Polio Syndrome (PPS)?

Acute poliomyelitis ('polio', 'infantile paralysis') is increasingly a disease of history as a result of the adoption in 1988 at the forty-first World Health Assembly of a resolution for the worldwide eradication of polio, and the subsequent Global Polio Eradication Initiative.

Despite this progress towards the eradication of polio, in recent years it has become increasingly apparent that new muscle weakness, atrophy and fasciculations can develop years and decades after a patient suffered an episode of acute paralytic poliomyelitis. There is a lack of consensus in the literature as to the nomenclature used to describe these new health problems being experienced by previous sufferers of acute paralytic poliomyelitis, but the terms 'The Late Effects of Polio' (LEoP), 'Post-Polio Syndrome' (PPS) and 'Post-Polio Muscular Atrophy' are frequently employed.

For the purposes of this document, the term LEoP is used to capture:

- Symptoms that are attributable to damage caused by the original acute poliomyelitis, including such aspects as residual weakness, musculoskeletal imbalance etc;
- Symptoms attributable to a failure to maintain the level of function achieved following the original acute infection - eg, new weakness and fatigue (PPS); and
- Secondary effects of chronic neuromuscular dysfunction, such as degenerative arthritis of overused joints, soft tissue diseases etc.<sup>1,2</sup>

Post-Polio Syndrome is generally considered a sub-category of LEoP, and is considered a neurologic disorder characterized by increased weakness and/or abnormal muscle fatigability occurring many years after the initial polio infection.<sup>1, 2</sup> The symptoms are thought to manifest when the compensatory neuronal processes which sprouted following nerve damage from the initial polio infection can no longer effectively innervate the muscles within their motor unit territory, resulting in an

unmasking of the neurological deficit caused by the original polio infection.<sup>3</sup> Some researchers believe there to be a neuroinflammatory component – particularly Drs Christian Borg and Henrik Gonzalez in Sweden (see their research into IV immunoglobulin therapy<sup>4</sup>).

The risk factors for the late effects of polio have not been thoroughly elucidated, although some factors thought to be more predictive of a patient's risk of LEoP have been identified. Patients who originally presented with paralytic polio appeared to have a higher risk of developing LEoP than those who had non-paralytic polio.<sup>5</sup> The degree of functional recovery during rehabilitation is also a risk factor for development of post-polio syndrome, with those achieving greater recovery during the rehabilitation phase being more predisposed to develop post-polio syndrome.<sup>6</sup> The age of the initial polio infection also appears to be a risk factor, with a younger age correlated with a higher risk of developing the LEoP.<sup>7</sup> Though, contrary to this, the reverse has also been reported, with older age when contracting polio identified as a risk factor. This has been attributed to the fact that acute polio is typically more severe in adolescents and adults than in infants and young children.<sup>8</sup> Other risk factors that have been observed in studies include the polio to post-polio interval and female gender.<sup>9, 10</sup> In addition, an inactive lifestyle can increase the risk of muscle pain and fatigue in individuals previously hospitalised with acute poliomyelitis.<sup>10</sup>

It is estimated that between 20,000 to 40,000 people were diagnosed with paralytic polio in Australia between 1930 and 1988<sup>11</sup>, with the highest rate reported in 1938 (39.1 per 100,000 population).<sup>12</sup> This figure must be increased 100-fold to obtain the estimated number of infected cases during the same period (up to 4 million people)<sup>11, 13, 14</sup>, and it does not include people who contracted polio overseas and who have since come to Australia.

Diagnosis of PPS is performed via the process of elimination and is a solely clinical assessment, with no specific tests for the diagnosis of PPS

currently available. Criteria for the diagnosis of PPS were agreed upon at the March of Dimes international conference on PPS in 2001<sup>11, 13-15</sup>. Dr Frans Nollet declared these to be validated at the European Conference on Post Polio Syndrome in Copenhagen, 2011. These criteria 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 atrophy of muscles on neurologic 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 or more) of stable neurologic function.

3. Gradual or sudden onset of progressive and persistent of new 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 breathing or swallowing.
4. Symptoms persist for at least a year.
5. Exclusion of other neurologic, medical and orthopedic problems as causes of symptoms.

The symptoms of the LEOp and strategies for their clinical management are briefly outlined in this module.

## 2. Muscle weakness and atrophy

### Key messages

- New muscle weakness can involve muscles known to have been affected by the original acute polio, as well as muscles that appeared to have been unaffected by the original acute polio. Even when muscle strength appears normal on clinical assessment, there may be significant denervation of muscle fibres.
- The management of new muscle weakness is primarily focussed on specific, appropriate physical activity, though these activities should be well paced and allow time for rest.
- Further muscle weakness and damage may be managed by use of appliances and devices (eg. walking sticks, crutches, wheelchairs, orthoses) and home modifications (eg. ramps, rails, stair lifts).

### Incidence and impact

In surveys of post-polio subjects, reports of muscular symptoms have included<sup>16</sup>

- New weakness in previously affected muscles (60% - 87%).
- New weakness in previously unaffected muscles (37% - 77%).
- New muscle atrophy (17% - 28%).
- Muscle twitching or fasciculations (39% - 41%).
- Muscle cramps (43% - 48%).

Recent research on the change in muscle strength over time in the normal population over the age of 50 has shown a normal average decline of approximately one percent per year. For post-polio individuals this rate is reported to be higher, at approximately two percent per year.<sup>16</sup>

In addition, polio survivors often suffer from a significant reduction in neuromuscular innervations and as such do not have an adequate "reserve" to sustain the same level of function as normal individuals as they continue to age.<sup>17, 18</sup>

Increasing muscle weakness can have a substantial effect on the ability of a person to continue to live a normal life. In addition to creating difficulties with activities of daily living, increasing weakness can lead to reduced balance and a higher number of falls.<sup>19</sup>

Overuse of weak muscles or of other muscle groups compensating for weakened polio-affected muscles can also lead to muscle pain, also significantly impacting on a person's quality of life.<sup>20</sup>

### Clinical characteristics

New muscle weakness, with or without associated muscle atrophy, can involve both previously affected muscles which were partially or fully recovered, and muscles that appeared to be unaffected by the original polio infection. The new weakness or atrophy is usually asymmetric. Decreased muscle endurance and increased muscle fatigue is also common, and may precede the development of weakness. The first subtle sign may be that the time needed for recovery after muscle activity increases.<sup>20</sup>

### Assessment

It is important to realise that people with a history of polio may have significant denervation of muscle fibres even when muscle strength appears normal on routine clinical assessment.<sup>20</sup> Loss of motor units and a decrease in muscle strength and endurance is not necessarily associated with impaired physical function, physical activity, or social participation. This non-linear association between denervation and function is possibly because of compensatory neuromuscular mechanisms.<sup>19</sup> Diagnosis for PPS can be supported by electrophysiological examination

including electromyography (EMG), which usually reveals longstanding neurogenic signs.<sup>19</sup>

### Management

Physical activity forms the basis of management of patients with the LEOp. Most patients will benefit from appropriate physical activity and a large proportion benefit from individually chosen, specific muscle training.<sup>19</sup> It is, however, important that these activities are well-paced and allow sufficient rest time to help minimise local muscle fatigue and facilitate strength recovery.<sup>21</sup>

As the number of functioning motor fibres in polio survivors with a given muscle may be significantly less than in normal individuals, recommendations for exercise and activity must be tailored appropriately.<sup>19</sup> In addition, increasing muscle weakness may be caused by overuse, disuse or a combination of both, and thus the underlying cause of muscle weakness should be determined so that appropriate management strategies can be developed.<sup>16</sup>

The management of new muscle weakness may include<sup>16</sup>

- Strengthening exercise (isometric, isotonic, isokinetic).
- Aerobic exercise.
- Stretching exercises to decrease or prevent contractures.
- Education regarding energy conservation techniques including pacing, rest, activity reduction and the avoidance of muscular overuse.
- Weight loss; and
- Prescription of orthoses and assistive devices.

Patients should be carefully monitored to identify signs of increasing muscle weakness

and muscle pain, in order to avoid undesirable effects, especially overuse of muscles that are symptomatically unaffected but had found clinically or by use of EMG to have impaired neuromuscular function. Overuse has been proposed as a cause of increasing muscular weakness. Appropriate rest and avoidance of excessive exercise should be instituted if chronic overwork is considered to cause the weakness, but there is no evidence that overuse weakness permanently damages muscles affected by poliomyelitis; conversely, muscles affected by polio readily atrophy with disuse.<sup>19</sup>

In order to avoid potential harm, exercise in post-polio patients should be carefully monitored and activity leading to excessive fatigue, muscle pain or joint pain should be avoided.<sup>21</sup> A referral to a physiotherapist may be indicated to ensure an appropriate exercise program is developed for this population.<sup>22</sup>

Many patients will find that modifying their work situation (by going to part time work) or early retirement is necessary. These patients may need to reduce and modify their physical pursuits and will find that pacing their physical activity and including periods of rest will increase what they can do, as research has shown that polio muscles can take longer to recover from physical activity.<sup>21</sup>



### 3. Fatigue

#### Key messages

- Fatigue can be the most disabling of the LEOp symptoms.
- LEOp fatigue typically occurs every day and increases toward the evening.
- An active yet balanced lifestyle is recommended in order to minimise the impact of fatigue.

#### Incidence and impact

The most common symptoms of the LEOp include a triad of fatigue, deterioration in muscle strength, and pain.<sup>2, 19</sup> Typically, fatigue related to the LEOp will begin after a period of functional and neurological stability of at least 15 years following the initial episode of acute poliomyelitis.<sup>19</sup> In a survey of polio survivors, new or increased fatigue was reported by 91% of respondents. Respondents also reported that fatigue was interfering with self-care (25%) and with completing or performing work (41%).<sup>23</sup>

#### Clinical characteristics

Current understanding suggests the aetiology of fatigue lies in the overuse or overwork of polio damaged nerves and muscles and can be categorised in two forms.<sup>2, 16</sup>

**General Fatigue:** otherwise known as central fatigue will often occur every day and progresses during the day. General fatigue can occur following only minimal activity and is described as an overwhelming exhaustion with flu-like aching and can include hot and cold flushes and sweating in conjunction with a marked change in the level of energy, physical and mental endurance.

**Muscle Fatigue:** otherwise known as peripheral fatigue or muscle weakness, is reported as a decline in muscle strength upon exertion, which may be best described as muscle fatigability or lack of endurance. Post-polio individuals have described muscle fatigue as “a heavy sensation in the muscles”, “increased physical weakness”, and an “increased loss of strength during exercise”.<sup>24</sup> Muscle strength often returns after a period of rest.

#### Assessment

Whilst fatigue is a classical clinical symptom of the LEOp, it is non-specific with a variety of possible aetiologies. As a result, before a diagnosis of fatigue is made, it is necessary to exclude other conditions that could cause fatigue such as anaemia, cancer, chronic infections, disrupted sleep, heart failure, medications or thyroid disease. LEOp fatigue is often associated with onset later in the day and with increasing severity toward the evening.<sup>24</sup>

If fatigue is diagnosed, assessment by a multidisciplinary team, including (but not limited to) physician, physiotherapist, occupational therapist and social worker is recommended. Of particular importance to fatigue is the evaluation of the patient by a physiotherapist and/or occupational therapist prior to the prescription of any interventions such as a mobility device.

#### Management

Fatigue in patients with LEOp is a severe and persistent challenge with physical and psychological co-morbidities.<sup>25</sup> Patients with post-polio syndrome should be advised to avoid both inactivity and overuse of weak muscles.<sup>19, 20</sup> Inactive LEOp patients have a higher risk for late poliomyelitis-related symptoms compared with active patients.<sup>10</sup> Although some risk factors for fatigue (eg, age and time since the acute poliomyelitis) are non-modifiable, others (eg, stress and physical activity) are modifiable.

The management of fatigue should therefore take a multidisciplinary approach, with an emphasis on individualised physiotherapy with specific muscle training, an active but balanced lifestyle (work smarter, not harder) in combination with minimising any interrelated psychological stress.<sup>20, 25, 26</sup>

## 4. Pain

### Key messages

- Pain in muscles and joints is a major issue for people suffering with the LEOp and typically the first or second most common symptom reported to health professionals.<sup>27</sup>
- Improvement in the evaluation and treatment of pain can significantly improve comfort and restore function.
- Successful management strategies focus on improving abnormal body mechanics and postures, supporting weakened muscles with bracing, targeted exercises, and promoting lifestyle changes, (eg, weight loss, pacing, rest, assistive devices) that improve health and wellness and prevent further episodes of pain.<sup>27</sup>

### Incidence and impact

The prevalence of various symptoms associated with the LEOp differs between studies. Pain, fatigue and weakness seem to be most commonly reported (between 42% and 87%) according to descriptive surveys and clinical examination studies.<sup>11</sup>

According to experts currently working with polio survivors, what is now current thinking in pain pathophysiology may be very relevant for polio survivors. Nociception is what is occurring in the peripheries – i.e. the detection of something outside of the normal boundaries of physiological activity that has the potential to be harmful to the body. The brain then decides to produce the output of pain as a perception, depending on the circumstances.

Problems arise when those who have had polio override these warning signs and potentially accelerate harm to relevant tissues.

Muscle pain is very common and is thought to be due to overuse of weak muscles or of other muscle groups which are compensating for weakened polio-affected muscles.<sup>20</sup>

Joint pain can also be significant, typically caused by joint instability, anisomelia (unequal length of limbs), disuse of muscles, abnormal biomechanical movements, and degenerative joint diseases.<sup>20</sup>

Ten percent of patients suffering with the LEOp have neuropathic pain, mainly caused by secondary disorders such as nerve compression or disc hernia.<sup>19</sup>

### Assessment & clinical characteristics

To facilitate the diagnosis and treatment of pain, a classification that divides the pain syndromes into three classes has been developed<sup>27</sup>:

- Biomechanical pain.
- Overuse pain.
- Post-polio muscle pain.

#### *Biomechanical pain*

Pain that results from poor posture is the most common type of pain reported by polio survivors. Weakness in polio-affected muscles (particularly the legs) often leads to poor muscular balance and skeletal alignment.<sup>27</sup> Years of walking on unstable joints and tissues makes polio survivors more likely to develop degenerative joint disease.<sup>27</sup> Polio survivors often experience shoulder pain due to the need for the body to compensate for weakened muscles or because of weight gain.<sup>28</sup> The majority of patients with LEOp will experience some degree of scoliosis.

Nerve compression syndromes like carpal tunnel and “pinched nerves” in the neck and back may develop from years of altered body alignment.

#### *Overuse pain*

The second most common type of pain reported by polio survivors is pain that is due to overuse of soft tissue. Muscles unaffected by polio, as well as those only mildly affected,

and tendons, bursa and ligaments are all vulnerable to overuse pain. These structures are often overused to accommodate for weakened polio muscles resulting in strains, sprains and inflammation. Tendinitis, bursitis and myofascial pain are examples of painful overuse conditions.<sup>27</sup>

### **Post-polio muscle pain**

Survivors describe post-polio muscle pain as burning, cramping or a deep ache. This type of pain is usually associated with physical activity and typically occurs at night or at the end of the day. Muscle cramps and/or fasciculations are indications of overuse of polio muscles.<sup>27</sup>

### **Management**

At this stage, there is no established specific treatment for pain associated with the LEOp.<sup>19</sup> The primary clinical management today is based on non-pharmacologic intervention<sup>20</sup>, including improving abnormal body mechanics and postures, supporting weakened muscles

with bracing, individually tailored training programs and lifestyle modification.

Adaptations recommended by a qualified health care provider, such as a physiotherapist, may include appropriate bracing, adaptive devices (walking sticks, crutches, corsets), special seating and postural modification.<sup>27</sup>

Muscle weakness and muscle pain may be helped with specific training programs - training in warm water seems to be particularly helpful.<sup>20</sup>

Lifestyle modifications can be effective in reducing overuse symptoms. Occupational interventions, weight-control programmes, group therapy (eg, water exercise) and increased use of assistive devices should be considered by the multi-professional rehabilitation team.<sup>19</sup>

Analgesics should be considered for symptomatic treatment where appropriate.<sup>19</sup>

## 5. Respiratory complications/insufficiency

### Key messages

- The most common respiratory presentation is shortness of breath.
- Other symptoms may be consistent with sleep disordered breathing (see section 6).
- Complications should be evaluated and managed by a respiratory physician specialising in neuromuscular disorders.

### Incidence and impact

Studies in Australia and overseas suggest the incidence of respiratory symptoms amongst polio survivors lies between 27% and 58%.<sup>29</sup>

<sup>30</sup> Late respiratory symptoms may be experienced by any survivor of polio, but are thought to occur more commonly in those who required artificial ventilation during the acute phase of poliomyelitis.<sup>31</sup> The frequency of respiratory symptoms has also been reported to be higher in patients who suffered acute poliomyelitis beyond the age of 10 and in those whose acute infection occurred more than 35 years ago.<sup>32, 33</sup>

Respiratory symptoms are primarily caused by a combination of respiratory muscle weakness and deformity to the chest cavity, and are characterised by a progressive loss of vital capacity and an inability to clear pulmonary secretions, particularly during infection. Complications can include ventilation/perfusion imbalance, pneumonias and pulmonary scarring.<sup>34</sup>

### Clinical characteristics

The most obvious respiratory symptoms of the late effects of polio are dyspnoea on exertion and/or at rest and difficulty clearing respiratory secretions. Other symptoms including fatigue and daytime sleepiness, impaired intellectual function (including poor concentration), morning headaches, speech difficulties, snoring and anxiety may be present and may be indicative of nocturnal hypoventilation.<sup>35</sup>

### Assessment

An evaluation of patients should begin with comprehensive patient history, in particular a history of previous ventilation requirements,

other respiratory illness and tobacco use, as well as symptoms consistent with nocturnal respiratory dysfunction such as snoring, excessive daytime sleepiness, headache etc. Other causes for respiratory symptoms should also be borne in mind as, whilst a common symptom of respiratory muscular weakness, shortness of breath may also be a manifestation of non-respiratory pathologies, such as cardiac dysfunction, primary respiratory disease, and obesity.<sup>36, 37</sup>

Investigations should include spirometry, evaluating vital capacity (both sitting and lying), inspiratory and expiratory pressure and maximum ventilatory volume. Arterial blood gas may also be indicated to evaluate lung function.<sup>36, 38</sup>

It should be noted that patients who have a history of polio may have normal respiratory function testing, but this does not take into account impaired muscle endurance<sup>19</sup>. Dyspnoea or sleep-disordered breathing may have a root in such muscle endurance impairment, and needs to be taken into account as it is not necessarily a lung related problem, but a respiratory pump problem (either muscle related or structural from a scoliosis.)

As patients suffering with the LEOp are also at risk of nocturnal hypoventilation caused by weakness of the respiratory muscles, chest wall deformity and sleep disordered breathing (eg, obstructive apnoea, central apnoea, or a mixed dyspnoea) for those presenting non-specific symptoms such as daytime somnolence, morning headache and fatigue or markedly reduced lung capacity, consideration should be given to polysomnography.<sup>19, 36</sup>

Complications such as restrictive lung disease and sleep disordered breathing should be evaluated and managed by a respiratory physician with experience in neuromuscular disease (refer to the section on *Sleep Disturbance* in this module for more information).

### Management

There have been no prospective randomised controlled trials investigating the treatment of respiratory symptoms in patients with the LEOp, but there is some evidence from other controlled trials that early recognition of respiratory impairment and introduction of non-invasive ventilator aids (eg, intermittent positive pressure ventilation (IPPV) or Bi-Level Positive Air Pressure (BIPAP) may delay further decline in respiratory function and the need for invasive ventilator intervention.<sup>1</sup> The use of ventilator aids may be appropriate in some cases but not necessarily all cases, their

use should be discussed between the patient and their respiratory specialist.<sup>39, 40</sup>

Respiratory muscle training is recommended for patients receiving ventilator treatment to assist pulmonary function.<sup>1</sup> Stopping smoking, mobilisation of secretions and cough assistance are also recommended.<sup>1</sup> In appropriate patients, weight loss and aerobic exercise, postural correction or treatment of sleep disordered breathing may be beneficial.<sup>16</sup>

If invasive ventilator aids are needed, home ventilation in tracheotomised patients is effective, though disabling.<sup>1</sup>

All persons with impaired pulmonary function and/or a history of respiratory disease should receive influenza and pneumococcal vaccines at recommended intervals according to National Health and Medical Research Council (NHMRC) guidelines.<sup>41</sup>

## 6. Sleep disturbance

### Key messages

- Sleep disturbance in patients suffering from the LEOp is usually the result of a primary sleep disorder (including obstructive sleep apnoea, central sleep apnoea and hypoventilation) or muscle twitching.
- Common clinical symptoms include tiredness and/or headache upon waking, daytime tiredness and/or sleepiness, impaired mental function and fatigue.
- Referral to a respiratory specialist for overnight oximetry and sleep studies is recommended to help identify the cause of the sleep disturbance.

### Incidence and impact

Sleep disturbance is common in patients suffering from the LEOp<sup>11, 42, 43</sup> and may be due to a number of factors ranging from a primary sleep disorder to disturbance from muscle twitching. The incidence of primary sleep disorders appears to be higher in post-polio patients compared to the general population<sup>43</sup>, and this may be due to chest and spinal deformities, weakened respiratory muscles and damage to the respiratory control centre following the primary poliomyelitis infection.<sup>16, 19, 20, 43</sup>

Primary sleep disorders include obstructive sleep apnoea (OSA), central sleep apnoea (CSA) and hypoventilation.<sup>16, 19, 42, 43</sup> OSA results in the interruption of airflow (apnoea) and occurs when the upper airway collapses.<sup>16</sup> Apnoeas are resolved upon waking and can occur many times per night, which in turn leads to sleep disruption.<sup>11, 16, 43</sup> One study indicated that 65% of fatigued polio survivors have sleep apnoea, with the majority of these cases (86%) classified as OSA.<sup>44</sup> OSA also increases the risk of hypertension, myocardial infarction, congestive heart failure and stroke in LEOp patients.<sup>16, 43</sup> CSA occurs when the brain reflexes signalling breathing during sleep fail. As a result, patients have trouble falling asleep as the transition from wakefulness to sleep is disrupted by frequent central apnoeas.<sup>16</sup>

Random muscle twitching at night can also disrupt sleep and can be related to conditions such as restless legs syndrome, periodic

movement in sleep and generalised random myoclonus (muscle contractions involving muscles throughout the body).<sup>16, 42</sup> A survey in LEOp patients found that two thirds of those surveyed reported muscle twitching or jumping and 33% had disrupted sleep due to twitching<sup>16</sup> (refer to the section on *Muscle Weakness and Atrophy* in this module for more information).

### Clinical characteristics

Common clinical signs of sleep disturbance are primarily the result of primary sleep disorders. They include tiredness upon waking, daytime tiredness and/or sleepiness, loud snoring (associated with OSA) headache upon waking, irritability, impaired intellectual function, poor concentration and fatigue.<sup>11, 16, 19, 42, 43</sup>

### Assessment and Management

Assessment of sleep disturbances should always involve a thorough clinical and physical examination.<sup>16, 43</sup> As sleep disturbances may be associated with a number of aetiologies, the cause of the sleep disturbance must be identified and patients should be referred for overnight oximetry and sleep studies.<sup>16</sup> Referral to a respiratory specialist should also be considered for assessment of OSA, CSA or hypoventilation<sup>11, 16, 19, 43</sup> (refer to section 5, *Respiratory complications/insufficiency* in this module for more information).

If sleep disturbance is associated with restless legs syndrome then medication to treat this condition may be indicated.<sup>19</sup>

Other considerations may include weight loss in OSA and hypoventilation related to obesity,<sup>19, 42</sup> as well as other causes of sleep disturbance including pain and stress.<sup>16</sup> It is also important to note that when assessing

patients for fatigue it is necessary to exclude sleep apnoea as a cause for this LEOp symptom.<sup>16</sup>

## 7. Swallowing difficulties (dysphagia) and speech difficulties (dysarthria)

### Key messages

- Clinical and sub-clinical damage to the bulbar nerves during acute poliomyelitis may result in the subsequent development of difficulties in swallowing and speech.
- Assessment through clinical history alone may not detect milder manifestations of disease.
- Patients with diagnosed LEOP should be referred to speech pathologists for full evaluation and, if necessary, to initiate remedial strategies to reduce the potential negative consequences of dysphagia and dysarthria.

### Incidence and impact

The incidence of new swallowing problems is between 6% and 22% of those suffering with the late effects of polio.<sup>29, 45</sup> This may be an underestimate as laryngeal penetration (passage of materials into the larynx) and loss of cough reflex can occur without obvious symptoms and in one study specific testing suggested weakness in tongue/palate and laryngeal abnormalities are present in 80% and 57% of polio survivors respectively.<sup>46</sup>

The majority of patients who experience dysphagia and/or dysarthria have a confirmed history of swallowing problems in the acute phase of polio, but even those with no history may have suffered sub-clinical damage to the bulbar nerves during the original infection.

The severity of swallowing impairment can vary substantially between patients who often develop strategies to compensate for symptoms such as tilting or turning their heads during swallowing, eating more slowly with smaller mouthfuls and avoiding foods that are difficult to swallow.<sup>47</sup> Aspiration is reported to be rare.<sup>48</sup> There is evidence of slow progression of symptoms.<sup>49</sup>

### Clinical characteristics

Intermittent or constant swallowing symptoms related to decreased pharyngeal transit, bilateral pharyngeal weakness and decreased bolus control.<sup>49</sup>

Speech problems including voice becoming easily tired and hoarse and difficulties in co-ordination of breathing and voice production (eg, during singing).<sup>47</sup>

### Assessment

Clinical history will help to determine whether patients are aware of bulbar symptoms, as well as other symptoms that may be consistent with post-polio syndrome.

An assessment by a speech and language pathologist may be very helpful to the primary care physician in the evaluation of the cause of dysphagia, as may be imaging such as video fluoroscopy. The differential diagnosis includes structural abnormalities from mouth to stomach and any disease involving muscles of swallowing (eg, motor neurone disease).

### Management

It has been suggested that modifications in swallowing position, a change in diet<sup>48</sup> and exercises to improve effectiveness of swallowing may be beneficial.<sup>16</sup>

Polio survivors with dysphagia should have their swallowing assessed at regular intervals to monitor progressive changes as well as to determine whether compensatory techniques continue to be effective.<sup>50</sup>



## 8. Impaired thermoregulation

### Key messages

- Cold intolerance and poor peripheral thermoregulation are common symptoms of the LEOp.
- Patients may not recognise that their limb is cold until they feel it is cold when touched.
- Management of cold intolerance is focused on managing symptoms.

### Incidence and impact

Cold intolerance (particularly of the extremities) is a common symptom of the LEOp which is attributed to nerve injury and muscle atrophy.<sup>51</sup> Between 46-62% of responders in post-polio surveys report cold intolerance.<sup>16</sup> Sensitivity to cold occurs when the external environment is cold, for example in winter or in air-conditioned environments.<sup>52</sup> The symptoms can present even at mild temperatures and warm indoor surroundings.<sup>51</sup>

Cold limb(s) due to the LEOp will not delay wound healing (unless the patient has other contributory factors such as diabetic vascular disease), however, it is an inconvenience for the individual.<sup>16</sup> In some cases, the symptoms can become very uncomfortable and even painful.<sup>51</sup>

### Clinical characteristics

Cold intolerance due to circulatory disturbance can be attributed directly to nerve and

muscular damage caused by the poliovirus.<sup>16</sup> Virtually all patients reporting cold intolerance will have a normal core body temperature, however limbs with significant atrophy tend to be cool to touch with a bluish discoloration and variable degrees of swelling.<sup>16</sup> Generally patients do not recognise that their limb is cold until they touch it and feel it is cold.<sup>52</sup>

### Treatment

The treatment of cold intolerance is focused on managing the symptoms. There are no medications or other types of clinical interventions to treat cold intolerance in polio survivors. Some practical approaches to dealing with cold intolerance may include<sup>16</sup>:

- Warming the patient directly, eg, wearing multiple layers of clothing or application of heat pads for short periods ( $\leq 20$  minutes).
- Warming the patient's environment eg, home insulation.

## 9. Bladder dysfunction

This section is a summary of a presentation given by Lise Kay at the Living with Polio in the 21st Century Conference, in 2009.<sup>53</sup>

### Key messages

- Symptoms of bladder dysfunction can have a significant impact on a person's quality of life.
- Potential causes of bladder dysfunction in survivors of polio include impaired detrusor muscles and nerves, oedema in the legs, restricted mobility and problematic voiding habits.
- The choice of treatment of urinary dysfunction in polio survivors varies depending upon the underlying pathology.

### Incidence and impact

During the epidemics of acute polio, bladder dysfunction was reported in approximately 20% of polio cases, with a greater prevalence among adults. The majority of patients experienced urinary retention, although incontinence, urinary stasis and urinary stones were also reported. Symptoms of bladder dysfunction in acute polio generally lasted one week, however approximately 15% of cases resulted in permanent damage and ongoing sequelae.

Urinary incontinence can have a significant impact on a person's quality of life. Patients can feel embarrassed about their symptoms and consequently restrict their involvement in social activities outside of their home.<sup>54</sup>

### Clinical characteristics

There are a number of potential causes of bladder dysfunction in survivors of polio, including impaired detrusor muscles and nerves, oedema in the legs, restricted mobility and problematic voiding habits.

A weak detrusor muscle may cause incomplete voiding and consequently voiding becomes more frequent and overflow incontinence may result. Polio survivors may also have a weak sphincter/pelvic floor leading to stress incontinence (leakage of urine), or an imbalance in the autonomic nervous system giving rise to urge incontinence (difficulty inhibiting the desire to void) or difficulties in initiation of voiding.

Polio survivors with restricted mobility due to weak leg and arm muscles may experience functional incontinence because they are unable to make it to the toilet in time to avoid leakage of urine. Polio survivors with paralysed legs may experience urinary incontinence as a result of oedema in the legs - fluid which accumulates in the legs during the day becomes mobilised when these patients lay down in bed, resulting in a larger urine production at night.

Another factor contributing to bladder dysfunction in polio survivors is problematic voiding habits, in particular suppressing the need to void leading to an overstretched detrusor muscle.

### Assessment

Generally, three simple tests and a screen for other diseases are required for a diagnosis of bladder dysfunction:

- A drinking/voiding chart for 3 days to be completed by the patient.
- Measurement of velocity of urine flow via a flowmeter.
- Measurement of residual urine via ultrasound.
- Screening for other diseases by urinary stick, vaginal-rectal examination and ultrasound.

If further work-up is required to reach a diagnosis, the patient should be referred to a urologist for full urodynamic investigation.

## Treatment

The choice of treatment of urinary dysfunction in polio survivors varies depending upon the underlying cause of symptoms.

Stress incontinence may be treated by strengthening the sphincter/pelvic floor with specific exercises, however urge incontinence may require pharmacotherapy.

Patients who experience incomplete voiding (residual urine > 100ml) may find it beneficial to trial double voiding (i.e. void once again at the same toilet visit). If this is unsuccessful,

clean intermittent self-catheterisation should be considered, with permanent catheterisation reserved as a final treatment option.

If urinary flow is less than 15ml per second or if the patient is unable to obtain voiding volumes greater than 100ml, they should be referred for a full urodynamic investigation.

In patients presenting with oedema of the legs and a large urine production at night, daytime fluid retention should be prevented by elevation of the legs and/or elastic stockings, supplemented with a mild diuretic in the evening (about 5pm) when necessary.

## 10. Surgical considerations

### Key messages

- When a polio survivor presents for surgery, special precautions are necessary as these patients may suffer complications during and post-surgery as a consequence of the LEOp.
- The choice of anaesthesia requires special consideration and lower doses are generally recommended for general anaesthesia but higher doses (eg, twice the dose) are required to control pain for local anaesthesia in dental surgery.
- Intensive monitoring may be required in the post-operative period and recovery may be prolonged.

### Pre-operative assessment

Patients with the LEOp should have the following assessments prior to surgery:

- Assessment for contractures or spinal deformities to establish a baseline condition and predict positioning requirements during surgery.<sup>55</sup>
- A detailed respiratory evaluation should be conducted, regardless of whether the patient experiences respiratory symptoms related to the LEOp or not. Any patient with symptoms suggestive of a decreased respiratory reserve should be referred for a baseline chest radiograph and spirometry.<sup>55</sup>
- Assessment for a history of sleep apnoea or hypoventilation syndrome as these patients are at a higher risk of cardiac dysfunction.<sup>55</sup>

### Peri-operative considerations

The following are important factors for health care providers to take into account when a patient suffering the LEOp undergoes surgery:

- Regional anaesthesia is preferable to general anaesthesia in LEOp patients as it is associated with fewer side effects.<sup>16</sup>
- The choice of general anaesthetics requires special consideration. Generally, selection of shorter-acting agents with titration to desired effects is preferred in patients with the LEOp.<sup>55</sup>

- Patients with the LEOp may have an increased sensitivity to the effects of induction drugs, maintenance drugs, muscle relaxants and opioids. Consequently, a lower dose of any such medication is generally recommended in this patient population.<sup>55</sup>
- Baseline twitch response to peripheral nerve stimulation should be measured before administration of neuromuscular blocking agent, as this response may be abnormally small in some muscles in post-polio patients.<sup>55</sup>
- Patients should be comfortably positioned with consideration given to limbs with contractures.<sup>55</sup>
- Blankets or warming devices may be needed during surgery for LEOp patients with cold intolerance.<sup>55</sup>
- Prophylactic anti-emetic medication may be required as some LEOp patients have bulbar dysfunction and an increased risk of aspiration. It is also crucial to carefully suction the laryngopharynx prior to emergence from anaesthesia.<sup>55</sup>

### Post-operative management

Important considerations for health care providers during the convalescent period include<sup>16</sup>:

- Recovery from surgery in LEOp patients may be prolonged by 2 or 3 times beyond the expected duration for the general population.

- It should be recognised that being outside of one's comfort zone may lead to the need for more assistance (a need which may not be identified by staff unaware of post-polio limitations).
- Some patients may require intensive monitoring in the post-operative period, particularly in order to monitor their pulmonary function.
- LEOp patients with sleep apnoea may experience a worsening of their symptoms following general anaesthesia.
- Polio affected muscles may be temporarily weaker after general anaesthesia and patients may require mobility aids.
- Paralysed limbs may have delayed wound healing due to decreased blood supply.

### Dental surgery

Although polio survivors with LEOp are more sensitive to general anaesthesia, they seem to require about twice the typical dose of local anaesthetic for dental surgery because of their increased sensitivity to pain. A problem is that the increased dosage may cause paralysis of facial, tongue and pharyngeal muscles and impair the ability to swallow saliva or breathe.<sup>56</sup>

Swallowing and breathing may be further compromised in those who had bulbar polio or paralysis of the respiratory muscles simply by reclining in a dental chair. A safe and

comfortable reclined position should be identified before any dental procedure begins.<sup>56</sup>

For these reasons, polio survivors need to communicate their polio histories and any requirement for physical assistance in transferring to and from the dental chair in the pre-operative period. Their medical history should include swallowing difficulties and the need to use ventilatory support. They should also inform the dentist of all medications taken since high doses of painkillers such as aspirin can cause excessive bleeding of the gums and anti-cholesterol drugs can further weaken muscles.<sup>56</sup>

Polio survivors with osteoporosis (refer to "Falls" section) should inform their dentist if they are being treated with bisphosphonate (Fosamax) because of the risk, albeit no more than 1/10,000, of developing osteonecrosis of the jaw, especially if the bisphosphonate therapy has been continued for more than 3 years.<sup>57</sup>

Recovery from dental surgery with local anaesthesia in LEOp patients may also be prolonged by 2 or 3 times beyond the expected duration for the general population. This is a major reason why same-day oral surgery for complicated dental procedures in polio survivors with LEOp is not advisable. Further, sedation-impaired coordination after dental surgery makes falling more likely and the need for physical assistance or mobility aids much greater.<sup>56</sup>

## 11. Falls

### Key messages

- Tailored exercise programs that include exercises to challenge balance should be implemented for polio survivors with LEO/PPS to reduce their high fall rate.
- Osteoporosis is common in the hips of polio survivors, especially those with polio-affected lower limbs, so bone mineral density of both hips should be determined in males and females in this population and treatment instituted, if indicated.
- The removal or modification of environmental hazards in the home and the avoidance of risky behaviour and external hazards can prevent falls.
- The use of braces and walking aids contributes to the prevention of falls.

### Incidence and Impact

The latest Cochrane review of the medical literature on falls in people over 65 years of age and living in the community confirms that approximately 30% fall each year.<sup>58</sup>

In contrast, the frequency of falls in polio survivors is significantly higher. A 2010 publication from The Netherlands reported that 74% of 305 polio survivors sustained at least one fall in the previous year with 60% reporting more than one fall.<sup>59</sup> An earlier report from the United States recorded a fall rate of 64% among 233 polio survivors in the previous year with 61% of the falls requiring medical attention, including 35% who had at least one bone fracture.<sup>60</sup> A report published on 50 post-polio patients in Ireland in 2009 recorded a fall rate of 64% over the previous six months; 19 of those 50 had fractured a bone as the result of a fall over the previous five years.<sup>61</sup>

### Assessment

Osteoporosis is common in the hips of post-polio populations, especially in the hip that is associated with a polio-affected lower limb.<sup>62</sup> Bone mineral density should be measured in both hips in males and females in such populations and appropriate treatment instituted, if indicated. Twenty eight of the 50 Irish post-polio patients were diagnosed with osteoporosis and 20 with osteopaenia, yet only eight of the 48 were receiving treatment for their low bone mineral density to reduce the risk of hip fractures.<sup>61</sup>

Reduced muscle strength, relatively rapid muscle fatigability and the impaired balance and gait associated with LEO/PPS are risk factors for falling. Other risk factors to be assessed in this population are visual impairment, dizziness on standing (eg, due to low blood pressure), and the taking of certain medications (eg, psychotropic drugs).<sup>58</sup>

Environmental hazards in the home and risky behaviour by the occupants should be assessed, preferably by an occupational therapist, where frequent falls have occurred in the home.<sup>58, 63</sup>

### Intervention and Prevention

Reviews of the medical literature provide strong evidence that exercise programs can reduce fall rates in older populations but the exercises that are more likely to be effective are those that challenge balance.<sup>58, 64</sup> Such exercises involve standing with feet close together or on one leg while practising controlled movements that strengthen the core trunk muscles.<sup>64</sup> The particular exercise program will need to be tailored to the capabilities of the individual polio survivor with LEO/PPS under the direction of a physiotherapist who has been trained or is experienced in devising appropriate exercise programs for polio survivors.

The removal or modification of environmental hazards in the home under the direction of an occupational therapist has been shown to prevent falls among older people who are at

increased risk of falling. In addition, the home visits may also lead to changes in behaviour that enable older people to move more safely in the home and in the external environment.<sup>63</sup> Rough terrain, sloped surfaces, wind and crowds in the external environment increase the risk of falling for those with LEO/PPS and are best avoided, if possible.

Bracing that prevents foot drop and stabilizes joints also contributes to the prevention of falls,

as does the use of a wheeled walker instead of a walking stick as muscles become weaker.

Polio survivors who live alone should be encouraged to wear personal alarms since they may not be able to get up from a fall, even in the absence of a bone fracture. A coded key safe box containing the house keys should be installed on the outside of the house near the front door to ensure prompt access when help arrives.

## 12. Psychological considerations

### Key messages

- Psychological and emotional factors may be significant contributors to the severity of symptoms associated with the LEOp.
- Conversely the severity of symptoms may in itself have an impact on psychological and emotional well being.
- Psychological state is inextricably linked to physical wellbeing and recovery.

### Incidence and impact

Psychological symptoms in polio survivors vary greatly along with the incidence and severity of the LEOp symptoms. The intertwined triad of LEOp (fatigue, pain and muscle weakness) often results in social isolation and strained interpersonal relationships which puts marriages and friendships at risk.<sup>11, 65</sup> Central to the aetiology of psychological symptoms may be polio survivors reliving many of the realities, emotional and physical, of their acute paralytic poliomyelitis. Polio survivors frequently report that the onset of post-polio symptoms have forced them, often for the first time, to recall and examine their acute polio experience.<sup>66</sup>

In surveys that have reviewed LEOp, depressed, anxious or stressed patients report a more severe physical deterioration, more pain with a higher rate of somatic complaints, poorer coping mechanisms, a lesser quality of life and more social exclusion.<sup>66</sup> Interestingly, most studies do not report an increased incidence of psychosocial or depressive events amongst LEOp patients.<sup>67</sup> This may in part be due to reports of the Type-A personalities (hard driving over-achievers) of some polio survivors, who frequently demand perfection of themselves in all aspects of their lives, and are confronted with new, progressive disability<sup>23</sup>. These unique circumstances surrounding the development of an unexpected second disability are thought to result in particular psychosocial difficulties.<sup>11</sup>

### Clinical characteristics

As the physical causes and optimum treatment regimes for post-polio symptoms are being clarified, psychological symptoms including chronic stress, anxiety, depression, and compulsive behaviour are becoming evident in polio survivors.<sup>23</sup> It is important for clinicians to consider the psychological impact of a chronic illness like polio, as these symptoms are commonly missed.<sup>65</sup> Importantly, these symptoms are not only exacerbating the LEOp but often prevent patients from making the lifestyle changes necessary to achieve a benefit from treatment programs.<sup>15, 66</sup>

Individuals who lose abilities which they previously re-gained through strenuous rehabilitation, may experience a deep feeling of bereavement resulting in social withdrawal, isolation, relationship hardships and a change in self perception. Polio survivors will often respond to these new symptoms with anger, fear and confusion.<sup>66</sup> Due to their experiences during the acute illness, many post-polio individuals fear hospitals and are wary of health professionals. As a result, faith in the medical profession has often been lost.<sup>11, 16, 67</sup>

### Assessment

It can often be difficult to separate the symptoms of the LEOp due to the interwoven connectivity of physical and emotional states. In order to provide the best therapeutic advice, it is essential to have a good knowledge of the symptoms but also take the time and listen to



patients in order to differentiate the underlying physical and psychological components.<sup>67</sup> The most appropriate method of providing a comprehensive and coordinated evaluation that addresses the polio survivor's medical, functional, psychosocial and vocational needs is through the use of an interdisciplinary team, including physician, physiotherapist, occupational therapist, psychologist and social worker.<sup>11, 16</sup>

### Management

The main aims of addressing the psychological needs of the post-polio patient should be to:<sup>11, 16</sup>

- increase and expand the patient's personal and external resources;
- provide education and support (to both the patient and family); and
- reinforce the need for the patient to have control over their lives.

Psychologists can provide counselling, education and support with regards to emotional difficulties the individual and close family members may experience. Intimate relationships between partners may also be affected due to the symptoms of pain, fatigue and weakness which affect the individual's self image and their sexuality.<sup>11, 16</sup> Psychological support can also assist the patient in their evaluation of therapeutic choices (rehabilitation, orthotics, mobility devices, medications, speech pathology or surgery), as well as the choice between treatment or an abstention of treatment.<sup>67</sup> In particular, the concept of relying on mobility devices such as walking sticks and wheelchairs can be extremely traumatic for polio survivors, so it is suggested that patients first test out these devices to help enhance their enjoyment of an activity (eg, visiting an art gallery or at an airport).

Importantly, previous approaches to polio treatment have been to ignore pain and fatigue and to exercise as much as possible; these therapeutic strategies are now being regarded as possible contributors to the post-polio

symptoms.<sup>11, 16</sup> So post-polio patients and their families are now being challenged and are being told to dramatically change their approach to managing their symptoms. This is a major obstacle for many individuals and often results in higher levels of non-compliance in treatment programs.<sup>66</sup>

Unfortunately, the support of family and friends is often not sought by polio survivors and, even when it is, may not prove to be adequate to the patient. Newly diagnosed patients frequently experience problems in communicating effectively about LEOp with their families and friends, and in obtaining help from them. It has been reported that only 39% of polio survivors requested help from their family and of these only 52% found it "very helpful" to do so. While 75% had talked with their family about LEOp only 40% rated this as a "very helpful" experience. In addition, this study also found that 74% had talked with friends about LEOp but only 23% of these found this "very helpful". Personal coping strategies (such as becoming more involved in interests they can still pursue, developing their philosophy of life, reading more about LEOp) were more frequently reported as being adopted than were interpersonal coping strategies and were more frequently rated as "very helpful". The length of time required to get a diagnosis results in many survivors' symptoms being discounted by their families and often health practitioners.<sup>30, 68</sup> As with the patient's assessment, the most successful approach to managing the complex psychological components of the LEOp will involve a comprehensive interdisciplinary treatment program comprising physicians, physiotherapists, social workers and psychologists.<sup>11, 16, 66</sup> Each team member brings with them specific skills and knowledge that could assist the patient to address some of the aspects of their condition. In addition, patients should be encouraged to connect with post-polio support groups to facilitate communication and awareness about their illness, and to allow access to an additional support network.<sup>16</sup> Together they can address the LEOp as a whole.

## 13. Pharmacological considerations (summary of Cochrane Review)

In 2011 the Cochrane Collaboration undertook a review of the treatments for symptoms associated with the LEOp.<sup>15, 69</sup> This section briefly summarises that review.

### Key messages

- Due to the lack of both good quality data and randomised controlled studies, definite conclusions on the efficacy of various treatment options for the LEOp could not be drawn.
- Results showed that intravenous immunoglobulin, lamotrigine, muscle strength training and static magnetic fields may be beneficial for the management of the LEOp, however require further investigation.

### Background and objective

The late effects of polio can affect 15% to 80% of those who survive paralytic poliomyelitis. The efficacy and optimum use of pharmacological and rehabilitation treatments to manage the LEOp is yet to be definitively established. The objective of the Cochrane review was to systematically review the efficacy of a range of potential LEOp treatments, both pharmacological and non-pharmacological, compared to placebo, usual care or no treatment.

### Pharmacological treatments

The pharmacological treatments included in the Cochrane review were:

- Amantadine, bromocriptine and modafinil: these drugs act on various areas of the brain and are used in an attempt to address general fatigue.
- Insulin-like growth factor (IGF-I) and human growth hormone: these agents may improve muscle strength by promoting regeneration of peripheral nerves.
- High-dose prednisone and intravenous immunoglobulin (IVIG): muscle strength, fatigue and pain may be improved by the immunosuppressive and immunomodulating properties of these drugs.
- Pyridostigmine: inhibits the breakdown of acetylcholine in the neuromuscular

synapse and may have a positive effect on fatigue and other LEOp symptoms.

- Lamotrigine: its postulated neuroprotective effects may reduce fatigue and pain.
- Coenzyme Q10 and selegiline: may assist with general symptoms of the LEOp via their effects on muscle metabolism and muscle strength.

### Rehabilitation (non-pharmacological) treatments

The three non-pharmacological treatments listed below were included in this review:

- Muscle strengthening: may improve functional capacities however literature suggests that muscle overuse and intensive training may worsen muscle weakness and fatigue resulting in further loss of muscle strength. Physically active LEOp patients, however, were found to have fewer symptoms and greater functional capacity.
- Rehabilitation in warm and cold climates: may have positive effects on physical, psychological and social aspects of health.
- Static magnetic fields.

### Summary of main results

Commentary was provided for the following treatments:

- Amantadine: 200mg/day for six weeks did not reduce fatigue compared to placebo.

These results were based on a small population and therefore it was concluded that there is very low quality of evidence on the benefits or otherwise of amantidine.

- Modafinil: 400mg/day did not reduce activity limitations, fatigue or pain compared to placebo. The studies investigating the use of modafinil were considered to be of high quality and hence the authors concluded there are no beneficial effects of modafinil.
- IVG: two infusions of 90g or one of 2g/kg body weight did not show improvements in activity limitations or fatigue. The effects of IVG on muscle strength and pain were inconsistent. The evidence is considered of moderate quality and further investigation is required.
- Prednisone: 80mg/day for four weeks followed by a 20 week tapering scheme showed no beneficial effects on fatigue. The authors concluded that the evidence is of very low quality based on the sample size of the study.
- Pyridostigmine: there is moderate quality evidence that 180mg or 240mg/day showed no effect on activity limitations, muscle function, fatigue or pain. Daily doses of 540mg to 720mg are indicated for treatment of myasthenia gravis and plasma concentrations vary significantly between individuals. Based on this, the authors concluded further investigation is warranted.
- Lamotrigine: 50mg to 100mg/day for four weeks was shown to improve activity limitations and pain. However, the evidence is of very low quality and further investigation is required to establish the efficacy of lamotrigine.
- Muscle strengthening: progressive resistance training of thumb muscles affected by polio showed an improvement in muscle strength;

however the evidence is of low quality. The authors commented on the value in assessing the effects of strength training of larger muscle groups such as the lower limbs, which are more frequently affected by the LEOp.

- Rehabilitation in warm and cold climates: there is low quality evidence that rehabilitation in warm or cold climates is of no benefit three months after treatment.
- Static magnetic fields: a reduction in pain was observed immediately after application of static magnetic fields over pain trigger points. The evidence is of moderate quality. Long term effects and effects on activity limitation were not studied and require further investigation.

#### Authors' conclusion

It was concluded that it was impossible to draw definite conclusions on the efficacy of the various treatment options for the LEOp due to the lack of both good quality data and randomised controlled studies. Results showed that IVG, lamotrigine, muscle strength training and static magnetic fields may be beneficial however all require further investigation.

#### Addendum: Medical Alert

The following therapeutic drugs may worsen the symptoms of LEOp/PPS and should be avoided or used with caution. The decision whether or not to take any drug always has to weigh up the benefits and the possible side effects:

- Cholesterol-reducing drugs such as statins.
- Beta-blockers.
- Benzodiazepines.
- Central nervous system depressants.
- Muscle relaxants.
- Local anaesthetics.

## 14. Comorbidity considerations

This section provides a summary of a presentation given by Irene Tersteeg at the European Conference on Post polio Syndrome in 2011.<sup>66, 70</sup>

### Key messages

- A higher level of comorbidity has been shown to be associated with a lower level of physical functioning.
- Polio survivors have more cardiac disease, respiratory disease, endocrine and metabolic disease, and more bone disease than the non-polio-affected population.
- Screening tests are important and should be done for diabetes, hyperlipidemia, hypothyroidism, osteoporosis, and lung function.

### Incidence and Impact

Polio survivors have paresis, are less able to lead an active lifestyle and are therefore more prone to certain types of comorbidity. A higher level of comorbidity has been shown to be associated with a lower level of physical functioning and a faster decline in physical functioning in polio survivors. Lifestyle-related factors, physical inactivity and excess weight are also associated with a lower level of functioning in polio survivors.

### Clinical Characteristics

Compared with the non-polio-affected population, polio survivors have more disease of the heart and blood vessels, such as heart attacks, hypertension and cardiac arrhythmias.

They also have more respiratory disease such as chronic pulmonary disease and asthma. When polio survivors age they may encounter late onset respiratory failure due to weakening of respiratory muscles and deformities of the chest.

The incidence of endocrine and metabolic disease such as diabetes, hyperlipidaemia, and hypothyroidism are also higher in polio survivors.

Due to an imbalance in muscle load, polio survivors have more disease of the bones,

muscles and tendons such as arthrosis and osteoporosis. Increased muscle weakness can lead to reduced balance and combined with osteoporosis can increase the risk of fractures after a fall.

### Assessment & Management

Many of the comorbidities associated with the LEOp are asymptomatic in their early stages, highlighting the importance of regular screening tests. While fatigue is a common symptom associated with the LEOp, it is important to rule out fatigue from other causes such as hypothyroidism. Screening should be done for diabetes, hyperlipidemia, hypothyroidism, osteoporosis, and lung function.

Walking aids may be needed in patients with increased muscle weakness in order to reduce the risk of falls and related bone fractures. Advice should also be offered regarding ways to manage diet and weight. Daily physical activity should be encouraged, with activities prioritised to lowering symptoms of fatigue and pain and to minimising the decline in physical functioning. Training in warm water is recommended, as it reduces the stress on joints and muscles and warm water may have an analgesic effect. Dynamic water exercises, not involving swimming, are a good alternative to weight-bearing exercises.

## 15. The Post-Polio Health Team

Due to the numerous and varied considerations for the diagnosis and management of patients suffering the LEOp, a broad team of healthcare professionals may be required to make up the “Post-polio Health Team”. A list of experts that could make up this team is provided below.

- **Rehabilitation specialist:** To support the management of pain and energy levels.
- **Neurologist:** To provide definitive diagnoses and comprehensive treatment plans for the neurological sequelae in polio survivors.
- **General Practitioner:** To develop a management program to address the post-polio patient’s specific symptoms, ensure continuity of care, educate the patient and promote a healthy lifestyle.
- **Physiotherapist:** To support polio survivors in improving/maintaining mobility, function and provide relief from pain.
- **Occupational therapist:** To assist polio survivors to employ strategies to help maintain overall health and encourage the highest level of lifestyle independence.
- **Orthotist:** To fit appropriate and well-fitting orthoses for patients who require them.
- **Podiatrist:** for assessment and treatment of foot conditions (commonly present in patients with a history of polio).
- **Speech Pathologist:** To support the management of patients with speech or swallowing disorders.
- **Respiratory Specialist / Sleep Specialist:** For management of respiratory dysfunction in patients with LEOp including preventative measures, ventilator assistance and treatment of sleep apnoea.
- **Psychologist/Social worker:** To provide counselling, education and support.
- **Dietitian/Nutritionist:** To provide education and management strategies regarding weight management.
- **Massage Therapist:** Can assist in the management of cold intolerance and pain control.
- **Osteopath:** To provide support for the management of postural / biomechanical issues in patients with LEOp.

## 16. Summary

- The late effects of polio (LEoP) is a set of sequelae in polio survivors characterised by the chronic impairments caused by the original polio infection, the secondary traumatic effects of those impairments, and from Post-Polio Syndrome (PPS), a neurologic disorder characterized by increased weakness and/or abnormal muscle fatigability and pain occurring many years after the initial polio infection.
- New muscle weakness can involve previously affected muscles, as well as muscles that appeared to be originally unaffected. Management of this new muscle weakness requires appropriately paced physical activity.
- Frequent fatigue is one of the most common and disabling symptoms of the LEoP, and a balanced, active lifestyle is recommended to minimise this symptom.
- Muscle and joint pain is a major and common issue for people suffering from the LEoP. Successful management strategies focus on improving abnormal body mechanics and posture, supporting weakened muscles with bracing and mobility devices, targeted exercises, and promoting lifestyle changes such as weight loss.
- The most common presentation of respiratory complications is shortness of breath and other symptoms may be consistent with sleep disordered breathing. These complications should be evaluated by a respiratory physician.
- Swallowing and speech difficulties are caused by damage to the Bulbar nerves during the acute phase of polio and may not be detected by clinical history assessment alone. Referral to a speech pathologist for full evaluation and management is recommended.
- A common symptom associated with the LEoP is cold intolerance, which is managed by managing the symptoms of impaired thermoregulation, rather than the cause.
- Disturbance to sleep is common in patients suffering from the LEoP and may be due to chest and spinal deformities and weakened respiratory muscles following primary polio infection. Assessment should always involve a thorough clinical and physical examination.
- Bladder dysfunction, potentially caused by impaired detrusor muscles and nerves, leg oedema and restricted mobility, can have a significant impact on the sufferer's quality of life. Treatments vary depending upon the underlying cause of symptoms.
- Special precautions are necessary when a polio survivor presents for surgery (including dental surgery) due to the higher risk of complications as a result of the LEoP, and a potentially longer recovery period. The choice and dose of anaesthesia requires special consideration and more intensive monitoring during the post operative period may be required.
- The modification of environmental hazards and tailored exercise programs to challenge balance should be implemented to help prevent falls. If necessary, the use of mobility aids can also be of help.
- Psychological and emotional factors may be significant contributors to the severity of symptoms associated with the LEoP. Physical wellbeing and management of symptoms can be linked to psychological state.
- The efficacy of pharmacological and rehabilitation treatments to manage the LEoP are yet to be firmly established. Definite conclusions cannot be drawn on the efficacy of various treatments due to the lack of good quality data and randomised controlled studies.

## 17. Useful resources for patients

The following is a list of useful resources that may be useful for post-polio patients:

### Resources for Polio Survivors

Polio Australia / Resources / Resources for Polio Survivors

[http://www.polioaustralia.org.au/?page\\_id=45](http://www.polioaustralia.org.au/?page_id=45)

### State Polio Networks

Polio Australia / About Us / State Polio Networks

[http://www.polioaustralia.org.au/?page\\_id=22](http://www.polioaustralia.org.au/?page_id=22)

### Fact Sheets for Polio Survivors

Polio Services Victoria / Resources

<http://www.svhm.org.au/services/polioservicesvictoria/Pages/Resources.aspx>

### Post-polio health care considerations for families and friends

Post-Polio Health International

<http://www.post-polio.org/edu/healthcare/index.html>

### Save Our Shoulders

A Guide for Polio Survivors

<http://www.polioaustralia.org.au/wp-content/uploads/2010/09/Save-Our-Shoulders.pdf>

### PolioPlace

A Service of Post-Polio International

<http://www.polioplace.org/>

### Frequently Asked Questions

Post-Polio Health International

<http://post-polio.org/faq.html>

### PolioToday

Salk Institute for Biological Studies

<http://poliotoday.org/>

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## 19. List of Abbreviations

<b>BIPAP</b>	Bi-Level Positive Airway Pressure
<b>CSA</b>	Central Sleep Apnoea
<b>EMG</b>	Electromyography
<b>IPPV</b>	Intermittent Positive Pressure Ventilation
<b>LEoP</b>	Late Effects of Polio
<b>NHRMC</b>	National Health and Medical Research Council
<b>OSA</b>	Obstructive Sleep Apnoea
<b>PPS</b>	Post-Polio Syndrome

## 20. Credits

### *Commissioned by:*

Polio Australia Incorporated

### *Written by:*

Members of the GlaxoSmithKline Australia Vaccines Medical Department as part of a volunteering program: Iris Depaz, Claire Borg, David Crump, Christopher Clarke, Pooi Eu Nham, Chris Vearing, Gabrielle Formosa, Helen Ho and Jutta Richter.

Dr Margaret Peel, Committee of Management Member (Qld), Polio Australia

Dr Mary Westbrook, Clinical Advisory Group Member

### *Reviewed by:*

Polio Australia's Clinical Advisory Group Members: Ann Duncan (Respiratory Nurse), Gnanalechumy Jegasothy (Physiotherapist), Natasha Layton (Occupational Therapist), and Dr Mary Westbrook (Psychologist)

Blaise Doran, Physiotherapist and Coordinator, Polio Services Victoria, St Vincent's Hospital

Tessa Jupp, CEO/Clinic Nurse, Post Polio Network of WA

Mary-ann Liethof, National Program Manager, Polio Australia

Elizabeth Telford, President, Post Polio Victoria

Gillian Thomas, President, Polio Australia

### *About Polio Australia*

Polio Australia is a community based, not-for-profit, national peak consumer body committed to standardising quality polio information and service provision across Australia for polio survivors.

Polio Australia's vision is that all polio survivors in Australia have access to appropriate health care and the support required to maintain independence and make informed lifestyle choices.

For more information: [www.polioaustralia.org.au](http://www.polioaustralia.org.au)



