Post Polio Syndrome

A guide to management for health care professionals
Contents

1 Introduction .................................................. 7
1.2 The Guideline Document ................................ 8
1.3 The Knowledge Base ..................................... 9

2 Polio and Post Polio Syndrome ... 11
2.1 Poliomyelitis ................................................. 12
2.2 Historical Treatment of Polio Survivors ............ 13
2.3 Post Polio Syndrome ....................................... 13

3 Symptoms: Presentation and Assessment ...................... 17
3.1 Overview of Presentation and Assessment ......... 18
3.2 Neuromuscular Fatigue ................................... 21
3.3 Neuromuscular Weakness and Atrophy .......... 23
3.4 General Fatigue ............................................. 25
3.5 Pain in PPS .................................................. 26
3.6 Respiratory Complications .......................... 28
3.7 Psychological Symptoms .............................. 31
3.8 Sleep .......................................................... 32
3.9 Dysphagia (swallowing) .............................. 33
3.10 Cold Intolerance Among Polio Survivors ....... 34
3.11 Bowel and Urinary Problems ...................... 36

4 Clinical Management ....................................... 37
4.1 Multidisciplinary Approach ......................... 38
4.2 Energy Management .................................... 40
4.3 Pain Management ........................................ 41
4.4 Orthotics and Footwear ............................... 43
4.5 Pharmacological Management of PPS .......... 44
4.6 Management of Respiratory Complications ... 44
4.7 Exercise ..................................................... 46
4.8 Psychological Interventions ......................... 47
4.9 Speech and Language Therapy .................... 48
4.10 Interventions That May Help ...................... 49

5 Gaps in the Evidence
Need for Research ........................................ 51

6 References .................................................... 53

APPENDICES:
7 Other Relevant Guidance and Information .......... 64
7.1 UK Clinical Information on PPS ................. 64
7.2 European Guidelines ................................. 64
7.3 International Clinical Information ............... 64
7.4 Clinical Reference Books .......................... 64
7.5 Other Relevant UK Clinical Guidelines ......... 65
7.6 Patient Information ..................................... 65
8 Glossary ...................................................... 66
Foreword

You’d be forgiven for thinking that Polio is a condition consigned to the history books; however today – in the 21st century – around 120,000 people in the UK are still living with its effects. The late effects of Polio (LEP) and in particular Post Polio Syndrome (PPS) are still real issues for thousands of people.

In the UK we are fortunate to have had a nationwide vaccination programme and since 1993 there haven’t been any new cases of Polio, but what about those people who contracted Polio as children in the 1930s, 40s and 50s? Up to 80 per cent of them are now living with PPS.

The aim of this publication is to assist GPs and other clinicians in recognising and managing PPS. After several years of stability, individuals can develop increasing weakness, fatigue and pain in previously affected or unaffected muscles, a general reduction in stamina, breathing, sleeping and/or swallowing problems and cold intolerance. These symptoms may sound familiar and misdiagnosis is common as PPS is not recognised as a factor.

Research conducted by The British Polio Fellowship in 2012 determined that only 55% of GPs were able to diagnose the symptoms and 18% of GPs did not know how to manage PPS when diagnosed. Independent research conducted in September 2015 by YouGov revealed that only 7% of the British public were aware of PPS.

There is no specific cure for PPS, but properly managed – through pacing, energy management and use of adaptive equipment, for example – it may stabilise or only progress slowly.

Further information regarding Polio, PPS and the work of The British Polio Fellowship is available on our website: www.britishpolio.org.uk
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Summary of key points

- Post Polio Syndrome (PPS) is a neurological condition that is characterised by:
  “Gradual or sudden onset of progressive and persistent new muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalised fatigue, muscle atrophy, or muscle and joint pain. (Sudden onset may follow a period of inactivity, or trauma, or surgery.) Less commonly, symptoms attributed to PPS include new problems with swallowing or breathing”

- PPS can occur in a high percentage of people with prior Polio. In the UK it is estimated there are 120,000 Polio survivors; UK surveys indicate over 60% prevalence of PPS in Polio survivors

- The impact on individuals can dramatically affect abilities, lifestyle and relationships; UK surveys of Polio survivors have reported that 31% have had to leave or change work and 60% need more help with the activities of daily life. Severe respiratory complications and dysphagia are relatively rare but can be life threatening

- Muscles that appear to have been unaffected can have significant subclinical denervation

- Commonly used assessment tools such as the MRC rating scale can give a false impression of strength as one-off movements may be strong but PPS patients often show poor endurance over repeated movements or during sustained movements

- Symptoms can be stabilised over long periods and quality of life enhanced by effective management, though there are no medications currently available that are proven to reverse muscular atrophy, improve neuromuscular strength or relieve the neuromuscular fatigue of PPS

Key recommendations:

- A multidisciplinary approach is strongly recommended, involving GPs, physiotherapists, occupational therapists, orthotists and other allied health professionals with referral to specialist consultants as required

- Effective assessment needs to review the patients’ history and status for all common PPS symptoms and should take into account their early experiences and how these may have influenced their response to new and/or increasing symptoms, their psychological well-being and the symbolic meaning of assistive devices. Regular reviews are recommended to monitor symptoms and identify any progression of the condition

- Energy management techniques can alleviate the symptoms of neuromuscular and general fatigue and reduce pain. These include:
  - Pacing activity 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

- The mainstays of pain management in this context is pacing, activity or lifestyle modification, energy conservation, activity reduction and assistive devices. Overuse pain on a daily basis is a sign that the muscles are being used maximally and activity should be reduced to a point where the muscles have some reserve

- An orthotics review by an experienced specialist can help reduce overuse and misuse and reduce energy cost of walking by optimising orthoses and footwear

- Exercise and physical activity programmes can be considered once energy management techniques have been used to control the symptoms of neuromuscular fatigue and pain. These may strengthen muscles where possible and help improve cardiovascular health leading to an overall improvement in wellbeing. However,
safe effective exercise requires an individually tailored non-fatiguing, pain-free programme and careful monitoring to avoid damage through overuse. It is recommended that a specialist physiotherapist with experience in neurological condition management provides an assessment and guidance.

- It should be recognised that respiratory complications may develop insidiously in individuals who appeared previously unaffected, but are particularly prevalent in the context of factors such as respiratory infections, periods of immobility, surgery, pregnancy or obesity.

- Patients with a history of snoring and/or day time sleepiness should be investigated for obstructive sleep apnoea (OSA) and respiratory insufficiency. If found to be in type I respiratory failure, nocturnal continuous positive pressure ventilation (CPAP) should be considered as treatment.

- Mechanical failures such as worsening kyphosis, scoliosis or respiratory muscle weakness can lead to chronic respiratory insufficiency. In the presence of type II respiratory failure Bi-level, non invasive ventilation (NIV) is the treatment of choice.

- As with other chronic diseases, pneumococcal and influenza vaccinations are prudent to offer given the respiratory issues already outlined. If a PPS patient smokes, smoking cessation advice and support can prevent worsening respiratory function and vascular complications. Dietary advice to optimise nutritional status, and consideration of weight management may also support functional status.

- Sleep disturbances need to be investigated as they may signal treatable breathing problems due to respiratory muscle weakness, sleep apnoea or abnormal sleep movements. Sleep pattern abnormalities, such as frequent wakening and decreased deep or REM sleep, found on polysomnography, may also contribute to PPS general fatigue.

- Difficulty with swallowing is a potentially serious problem for survivors of acute Polio and those with PPS; if dysphagia/swallowing problems are suspected then referral to a Speech and Language 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 due to PPS on swallowing ability. Patients should be assessed for risk of aspiration pneumonia.

- 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.

- Weight reduction may help reduce the occurrence and severity of neuromuscular fatigue and pain. An effective programme needs to take account of a probable low proportion of lean mass which may make general BMI thresholds inappropriate in determining treatment options. Energy expenditure limited by disability, fatigue and pain together with low lean mass will affect calorie requirements and activity capability. Programmes should also support adherence. Therefore referral to a dietician is recommended.
1 Introduction
1 Introduction

Post Polio syndrome (PPS) is a neurological condition that occurs in many people who have had Polio. PPS occurs at any age, in people who had either paralytic or non-paralytic Polio. After decades without any significant change in their condition, people often develop new symptoms of increasing weakness, stamina problems, fatigue and pain.

There are over 120,000 people who have had Polio in the UK, most of whom could develop PPS. The diagnosis and treatment of PPS is frequently delayed as symptoms often develop slowly and may be mistaken for other conditions: PPS is a diagnosis of exclusion.

Research has established that PPS is a distinct clinical entity, but predicting who is most likely to experience PPS is not possible. It is estimated that PPS develops in the majority of people who contracted Polio and its prevalence is expected to increase in the UK, firstly due to changing demographics from an ageing population and secondly immigration where presenting cases will be younger and have different needs, for example management during pregnancy.

Symptoms of PPS may include the following:

- general fatigue
- neuromuscular fatigue (muscle fatigue)
- muscle and joint pain
- muscle loss
- new or increasing localised weakness
- sleep disturbance
- breathing problems
- cold intolerance
- swallowing problems

A diverse range of interventions is used to manage the symptoms of PPS and there is considerable variation in service provision. Symptoms of PPS can be managed within a variety of settings ranging from the community to specialist neurological or rehabilitation services. Access to specialist services is variable and is dependent upon referral from a General Practitioner.

A comprehensive review of benefits and risks of different management strategies in both the short and long term is needed to help health professionals, patients and carers who are faced with a choice between different long-term treatments.

1.1 Other guidance

There are several published documents which give guidance on PPS (see Appendix 1 for a full list). The Map of Medicine pathway is based on a flowchart through assessment and management interventions with brief information for each stage. The European Federation of Neurological Societies published clinical guidelines based on the report of an EFNS task force. The Cochrane Collaboration published a systematic review of controlled trials for treatment of PPS.

The British Polio Fellowship produces information and factsheets for Polio survivors, with and without PPS, in the UK. Patient information is also available on NHS Choices and ‘patient.co.uk’.

This document fills a gap by providing evidence-based information and expert opinion for non-specialist primary care and allied health professionals.

1.2 The guideline document

1.2.1 Population covered by the guide

The population covered by this document is people diagnosed with PPS. No sub-groups have been identified as needing separate consideration.

1.2.2 Healthcare setting

All settings in which NHS care and publicly funded services are received.
1.2.3 Target users of the guide
- GPs
- Allied health professionals
- Clinical Commissioning Groups
- Local Authority public health professionals
- Healthcare professionals who assess benefit claimants
- People with PPS and their carers

1.2.4 Relevance to Polio survivors generally
The primary purpose of this document is to provide information on the management of the symptoms of Post Polio Syndrome. Much of this information will also be helpful for clinical management of Polio survivors who do not have a diagnosis of PPS.

1.2.5 Summary of the process
The British Polio Fellowship has asked the Expert Panel to produce clinical guidelines on the management of PPS.
In 2013, the Expert Panel developed a scoping document and, following external consultation, a final scope was agreed. This guide to management of PPS has been prepared by members of the Expert Panel with additional specialist input where required. The sources used are discussed below.

1.2.6 What is covered
The document covers the symptoms of PPS that require primary care and allied health management.

1.2.7 What is not covered
The following are outside the scope of this document
- Management of acute Poliomyelitis
- Guidance on immunisation
- Diagnosis of PPS
- Management of symptoms of PPS using orthotics
- Management of comorbidities
- Specialist management of PPS, for example surgical intervention (for a review see Sheth and Keenan[^3])

1.3 The knowledge base
The sources used in developing this guide include several literature searches, published guidelines relevant to PPS, specialist conference material, clinical Expert Panel members and the lived experience of panel members who have had Polio.

1.3.1 Sources used
The literature searches primarily utilised PubMed, further supplemented by following references in key papers and recommendations from panel members.
In addition to the published guidelines on management of PPS listed above (the Map of Medicine, EFNS guidelines and Cochrane reviews) other guidance documents have been consulted[^4-9].
Three recent international conferences included two European Post Polio conferences, Amsterdam in 2014 and Copenhagen in 2011, and Polio Health International’s conference in St. Louis in 2014.
The clinical expertise came from the Lane Fox Respiratory Unit which provides a multi-disciplinary service for people experiencing the late effects of Polio and runs a Post Polio Syndrome self management programme. The service has been running for over twenty years, seeing 600 patients each year, which equates to over 5,000 patients in total.
The personal experience of Polio survivors on the panel provided a service user perspective and input from many years of contact with Polio support groups. In addition, several had conducted academic research in the field.

1.3.2 Limitations
Interpretation of the evidence is limited by several factors. Firstly, the existence of PPS was only recently accepted[^10] and internationally agreed diagnostic criteria only published in 2000[^4]. This makes comparison between studies difficult.
Secondly, in Europe, the USA and Australia where most research so far has been done, the cohort
comes from epidemics in the 1940s and 50s. As PPS has a time-dependant onset and symptom progression, data from this cohort in 1990 compared to data in 2010 looks at different stages of the syndrome.

Thirdly, there are very few high quality controlled trials of interventions and many published studies only involve a very small number of selected participants, often those from specialist clinics who are considered fit enough for the intervention. People not seeking help or more profoundly affected are not generally included in research trials.

Finally, due to the nature of the acute Polio disease, many people could have been infected but suffered undetected non-clinical levels of neuronal loss; the consequences of such loss are largely unexplored.

1.3.3 Terminology

The terminology relating to PPS can be unclear. Several other terms have been used to describe late developing effects of Poliomyelitis such as the late effects of Polio (LEOP), Post Polio muscular atrophy (PPMA), and Post Polio sequelae.

Fatigue is a commonly reported PPS symptom but the term is poorly defined and is used to describe a wide range of effects. Many surveys and studies use the unqualified term ‘fatigue’; some distinguish subtypes of physical vs. mental or central vs. peripheral fatigue.

In this document, the term neuromuscular fatigue (or muscle fatigue) refers to the reduction in muscular force or power induced by exercise and relieved by rest - this is often localised to a muscle or muscle group. The term general fatigue is used for more widespread or centrally experienced fatigue.

Where ‘fatigue’ is used without qualification, it is because it is the term used in the reference cited.
2 Polio and Post Polio Syndrome
2 Polio and Post Polio Syndrome

Post Polio Syndrome (PPS) is a neurological condition that can occur in people who have had Poliomyelitis after a period of stability, usually in excess of 15 years. The criteria recommended for a diagnosis of PPS by the EFNS task force, which include the symptom list below\(^{11}\), are those published by the March of Dimes\(^4\) - this list is also used by NINDS (USA), Polio Australia\(^8\), and the European Polio Union\(^12\).

“Gradual or sudden onset of progressive and persistent new muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalised fatigue, muscle atrophy, or muscle and joint pain. (Sudden onset may follow a period of inactivity, or trauma, or surgery.) Less commonly, symptoms attributed to PPS include new problems with swallowing or breathing.”

It is estimated that there are 120,000 Polio survivors in the UK, similar to the estimates for people with Parkinson’s disease and more than those with multiple sclerosis\(^13\). A recent survey by The British Polio Fellowship has shown that in 85% of 378 respondents, PPS has been confirmed or is suspected (53% and 32% respectively)\(^14\). Fatigue is a very commonly reported symptom - UK based surveys report over 60% prevalence in Polio survivors and is often considered the most debilitating symptom\(^15,16\).

2.1 Poliomyelitis

2.1.1 Epidemiology

Ancient images suggest that Poliomyelitis has existed for millennia\(^17\). It was first described clinically by Michael Underwood in 1789 as a rare and endemic ‘debility of the lower extremities in children’. The first cluster of cases of sudden paralysis in healthy children was recorded by John Badham in Worksop in 1835. Jacob von Heine first described the disease in detail in 1840, noting fever and pain suggestive of contagious illness involving the spinal cord. In 1887, Karl Oskar Medin studied 44 cases in Stockholm and first recognised Polio as an illness giving rise to epidemics. His pupil, Ivor Wickman meticulously investigated a 1905 Swedish epidemic of 1031 cases, deduced that Polio was spread through the mostly abortive or non-paralytic minor illnesses. He used the term Heine-Medin disease. In 1908 Landsteiner and Popper proved the disease was caused by a virus\(^18\). The largest epidemic of the early 1900s was in New York City in 1916 with 8991 cases, a quarter of whom died\(^19\). The epidemics increased until, in 1952, there were almost 60,000 cases of Polio in the US. Iron lungs, invented in 1928 for respiratory paralysis, were transported from one epidemic to another, and many children remember the fear of hearing the noise of the iron lungs at night or seeing that one had been placed near their bed in case it was quickly needed. In Copenhagen in 1952, there was an epidemic of 2722 patients, 316 of whom had breathing paralysis. Without enough iron lungs, 250 medical students were recruited to manually ventilate patients in eight hour shifts. In the UK, the largest epidemic was in 1950 with 7760 notified cases\(^20\).

The Poliovirus consists of three different strains of enterovirus in the picornavirus family. It is transmitted person-to-person or through contaminated water. The disease probably went from being endemic to epidemic in the late 19th century in countries where sanitation and water supplies were beginning to improve and children were no longer becoming immune at a very young age. Only about 1% of all Polio infections are paralytic\(^21\), and during the major epidemics, about 50% recovered with no obvious weakness. The death rate was around 7 to 8%, mostly those with brainstem and respiratory involvement\(^19,22\). Slightly more males than females developed Polio, but there was some evidence that the disease was more severe in females\(^23,24\). In countries where epidemics began early in the 1900s, the age distribution changed from predominantly very young children to a more even spread of ages up to adult\(^25\). In 1955 the Salk Polio vaccine was released and was gradually adopted in countries around the world. In 1961 Sabin brought out a live Polio vaccine and both vaccines are still in use\(^26\).

2.1.2 Pathology of Polio

The first description of Polio as a disease causing damage in the anterior horn cells of the spinal cord was by Duchenne de Boulogne, who invented electrodagnosis in 1855. This was confirmed through an autopsy by Charcot in 1870. It was not
until the 1940s that Bodian and Sabin carefully studied the pathology of Polio, both paralytic and non-paralytic, in live monkeys and human autopsies. Bodian was the first to discover that the brain, the motor cortex and brainstem, were affected in all Polio cases, paralytic and non-paralytic. In the spinal cord, the virus appeared to damage about 95% of motor neurons, even in mild cases, and on average 50% of neurons were destroyed, but if the damage was scattered the muscles could have normal strength. Many neurons recovered to some extent in the first month after acute Polio. After that, recovery involved compensating for the loss of motor units through reinnervation by nearby neurons until motor units could be ten times their normal size, and through muscle fibre hypertrophy. However, studies of monkeys found that in cases with mild transitory weakness there were typical lesions and neurons showing degenerative changes, which may never have been able to support enlarged motor units. In cases with low recovery there may have been many of these damaged neurons. Two studies on non-paralytic Polio patients showed that, with very careful testing and follow-up, many were found to have had paralytic Polio, although the main symptom was muscle fatigue. In recent studies of ostensibly unaffected muscles in paralytic Polio patients, up to 80% of muscles had abnormal EMGs showing enlarged motor units. Some Polio patients may have normal EMGs which could indicate damage but not loss of motor neurons.

2.2 Historical treatment of Polio survivors

The oldest survivors of Poliomyelitis in the UK are likely to have contracted the disease in the 1930s. A brief review of typical treatments for Polio at that time and subsequently will be helpful in putting the current management of PPS in context.

Until well after the end of World War Two, Poliomyelitis management was an orthopaedic domain although neurologists in the UK were coming to recognise the need for change.

The principles of management of the acute disease were prompt isolation and complete bed rest to reduce the risk of transmission and of worsening any paralysis. The greatest fear was respiratory paralysis. Prolonged immobilisation was common although the more humane and appropriate approach (advocated in the Kenny Method) was becoming part of normal practice. An important component of the Kenny Method became the psychological support of the Polio survivor during the recovery process. Acute care was usually delivered in a hospital for infectious diseases and would last several weeks. Following this, in-patient treatment in an orthopaedic hospital could last between one month and two years. Surgery was frequently recommended: Achilles tenotomy, nerve grafting, muscle re-alignment, long bone growth arrest, joint fusion and many other techniques. In addition to further hospitalisation, prolonged periods of recuperation were often necessary. Muscle spasm was relieved using hot pack application. Electrical stimulation, often to the level of pain, was used in the UK as late as the 1950s. Complex, heavy and cosmetically unsatisfactory orthotics and other equipment were provided when thought necessary.

At the completion of the treatment and rehabilitation phase, survivors were told that the disease had ended and would not return. The primary advice was to keep active, and to stay active - ‘use it or lose it’. After discharge from orthopaedic outpatient care (at age 18 if they contracted Polio as a child) there was no further Polio-related medical supervision.

The ungainly but perhaps effective orthotics were discarded and life lived to the full. However, survivors retained the emotional scars resulting from their initial and subsequent treatments: loss of family contact, isolation, loneliness, fear of treatments, especially the iron lung. When the symptoms of Post Polio Syndrome develop, the psychological impact of the apparent return of the illness can be considerable (see Section 4.8).

2.3 Post Polio Syndrome

2.3.1 Epidemiology

Polio has previously been considered a stable disease after the recovery period and no long-term follow-ups were conducted even though the first description of late onset weakness and muscle fatigue in a 19-year-old manual worker was described in 1875, followed by several dozen
more studies over the next century. New symptoms, such as weakness, fatigue, pain, sleep, respiratory and swallowing problems, years after acute Polio, were first beginning to be noticed in the late 1970s, about 30 years after the major epidemics of the 1940s and 1950s, and came to be called Post Polio Syndrome (PPS). At first there was medical disbelief, so the initial studies focused on the most severe Polio cases with obvious new weakness, which became the defining symptom of PPS. However, population studies began to show that PPS could occur in anyone with prior Polio, including those diagnosed with non-paralytic Polio or those never diagnosed at all, and a new set of criteria centring on muscle weakness and abnormal muscle fatigability was proposed and has since been widely accepted. Several population studies from the late 1990s and early 2000s in Holland, Scotland, Norway and Estonia have found a prevalence of new weakness of about 60%, and a Japanese study reported 85% with new muscle weakness, fatigue or difficulties walking. Progression of PPS has been found to be slow with estimates of the decline in strength to range from 1.5% to 2% per year. While group average change tends to be slow, individual change can be rapid. PPS patients with the largest decline in function tend to be those performing at higher levels of muscular capacity, causing more overuse symptoms, and with more widespread weakness, though not necessarily more severe weakness.

2.3.2 PPS pathology

PPS pathology derives from the Polio damage to the brainstem and motor neurons in the spinal cord. The leading causal hypothesis for PPS is that an excessive metabolic stress on remaining motor neurons, possibly due to muscle overuse over many years, results in distal degeneration of terminal axons and eventually the damaged motor neurons themselves. After acute Polio there is ongoing denervation and reinnervation, but with time the ability to reinnervate orphaned fibres diminishes, especially for motor neurons with remaining damage. This can occur in muscles that appear normal but have subclinical denervation. The symptoms of PPS mainly stem from the new muscle weakness and abnormal muscle fatigability arising from loss or damage of motor neurons and the subsequent adaptations in the muscles. Motor unit number estimation (MUNE), a measure of the number of motor neurons, has shown that motor unit numbers decrease with time in Polio survivors. In a 15 year study, Sorenson, Daube et al. found that summated MUNE declined by 44%, from 407 ±67 to 226 ±64 in the tested muscles. This study also showed a significant correlation between MUNE and strength, though the relationship was not linear, suggesting that weakness does not become apparent until a certain number of motor units have been lost. McComas, Quartly et al. also showed a reduction in MUNE of 13.4% over two years in a group of Polio survivors with 92% incidence of PPS and average age 53. This rate was twice the rate expected for healthy people over 60 years old.

The most recent electromyographic study of motor unit size in 47 people with PPS found mean motor unit action potential (MUAP) decreased by 20% over 10 years but was stable in matched controls. Baseline MUAP sizes were twice as large for people with PPS and three times as large for the most severely affected subgroup compared to controls. While some participants showed increased MUAP sizes indicating a remaining capacity for axonal sprouting, both the largest reductions and increases occurred for those with the largest initial MUAP size, with only reductions for the very largest MUAP size. The rate of decline in strength was related to the rate of decline in both MU size and number of active MUs. These results show that the motor unit size decreases with time in PPS, supporting the hypothesis of distal axonal degeneration leading to denervation of muscle fibres.

Distal axonal degeneration can also produce neuromuscular junction transmission defects and may be a factor in neuromuscular fatigue. Increased jitter on single fibre electromyography (SFEMG) has been observed in PPS with prevalence increasing with time from the acute infection.

Besides the Polio damage and loss of motor units, compensatory changes in the muscle occur, such as transition from fast to slow muscle fibre and changes in contractile properties of overused muscle fibres, which supports hypotheses for muscle strength being favoured over endurance in the muscle, and a high use of energy leading to neuromuscular fatigability and overuse muscle
The energy cost of walking in PPS patients has been found to be 40% higher than in healthy subjects\textsuperscript{58}.

During the acute Polio infection, the brainstem was also involved, particularly the reticular formation (wakefulness, respiration and cardiovascular dysfunction), vestibular nuclei, cerebellar nuclei, hypothalamus and thalamus, and the cranial nerves, for instance the vagus nerve which affects swallowing, digestion and cardiac irregularities like arrhythmias\textsuperscript{22}. Some PPS symptoms may be caused by the brainstem damage in Polio, such as central sleep apnoea\textsuperscript{22}, restless leg syndrome\textsuperscript{59}, changes in sleep architecture\textsuperscript{60}, and mental fatigue\textsuperscript{61}.

Overuse has been proposed as a contributing factor in PPS and can arise from chronic overloading of muscles during general activities of daily life (ADL)\textsuperscript{49,62}. Overloading can be due to reduced capacity of Polio-affected muscles and also to compensatory action by more healthy muscles\textsuperscript{63}. Evidence of overuse has been shown by Perry et al.\textsuperscript{62} with excessive use of two measured muscles on average during walking found in patients with symptoms of PPS. Borg, Borg et al.\textsuperscript{64} have shown that leg muscles of people with prior Polio can be maximally recruited during walking and that these muscles are markedly hypertrophic with predominantly type 1 fibres\textsuperscript{54}. There is also evidence of raised serum creatine kinase (CK) levels in Polio survivors which can be a marker for muscle damage\textsuperscript{65}. Raised levels have been found in Polio patients with new symptoms compared to asymptomatic controls. The levels correlated with voluntary elbow flexor strength, walking distance and self-reported strenuous work, but not with new or residual weakness\textsuperscript{66-69}.

PPS muscle and joint pain is most often nociceptive pain, which is caused by damage to body tissue and characterised as aching with symptoms of inflammation or a throbbing pain\textsuperscript{70}. The study of Vasiliadis, Collet et al.\textsuperscript{71} suggests that muscle pain may derive from Polio damage leading to muscle fatigue and overuse, while the joint pain, correlating with past and new weakness, is a result of joint instability from Polio weakness. One theory about the abnormal muscle fatigability and overuse pain is that they may result from changes in the adaptive mechanisms of the muscle following Polio involving fibre type and aerobic capacity\textsuperscript{55}.

Elevated activity of myeloperoxidase (MPO) has been found in the serum of PPS patients, which often indicates exercise-induced muscle damage and may initiate further damage\textsuperscript{72}. More recently, prostaglandin E2 has been found in PPS muscle, suggesting an inflammatory process relating to muscle pain, the symptom most helped by intravenous immunoglobulin\textsuperscript{73}. This inflammation may be secondary to systemic inflammation, or could suggest muscle overuse.

2.3.3 Current research

Another important hypothesis, not incompatible with the overuse theory, involves inflammation which has been found in the spinal cord and muscle in early PPS studies, and more recently in cerebrospinal fluid, serum and muscle\textsuperscript{73,74}. One study measured leptin levels, which rise with BMI and increase inflammation\textsuperscript{74}. Raised levels of myeloperoxidase (MPO) in PPS leucocytes, another sign of inflammation which increases in muscle after over activity, could be a sign in PPS that muscles are being used maximally. MPO is not found in autoimmune diseases like MS, and can cause neuron damage. Inflammation arises initially to return tissues to equilibrium after injury or disease, but may also cause further injury especially if chronic. Inflammatory markers in serum were correlated with pain\textsuperscript{74} as was elevated prostaglandin E2 found in PPS muscle\textsuperscript{73}. Another study found protein biomarkers in the cerebrospinal fluid related to neurodegeneration\textsuperscript{75}. The inflammatory response could be primary and systemic or secondary to neurodegeneration.

Intravenous immunoglobulin (IVlg) has been shown to lower cerebrospinal fluid levels of TNF-alpha and IFN-gamma, and clinically has both lowered pain and improved quality of life, especially vitality, although methods and results differed between the studies\textsuperscript{76-79}. A one-year follow-up showed a significant improvement in the SF-36 Physical Component Summary, pain score and walking ability. Not all PPS patients respond to IVlg treatment and a recent study identified responders as those with a high level of fatigue and pain, muscle atrophy in lower extremities and reduced physical function\textsuperscript{80}. The relationships between symptoms and sites of inflammation are poorly
understood. The reduction of muscle pain following IVIg may suggest that muscle overuse plays a significant role in the causation of this symptom. The efficacy and safety of IVIg in patients with PPS is currently the subject of a multicentre randomised controlled trial.

Studies to support other theories for chronic inflammation in PPS, such as an autoimmune process, Poliovirus genomic sequences and Poliovirus persistence, have had mixed results. There is some recent evidence against PPS being an autoimmune response, Poliovirus genomic sequences and Poliovirus persistence were researched in the 1990s and while several studies found viral particles in some patients, others found none. The viral particles would be incapable of destroying cells in the manner of complete viruses, but might theoretically trigger inflammatory damage. There is recent interest in reviving the Poliovirus persistence theory in the light of the IVIg trials and new anti-viral drugs.

### 2.3.4 Risk factors

Greater risk of developing PPS has been associated with greater severity of the acute disease, greater functional recovery, older age at contraction of Polio, permanent impairment after initial recovery, older age and lower disability at clinical presentation (for PPS assessment), female gender, longer time since the acute disease and possibly physical activity. Most of these factors support the view that people with the greatest motor neuron damage and greatest recovery based on motor unit enlargement are most at risk of decline.

The first PPS risk factor study was conducted on quite severely affected patients and found that those who had had the highest recovery and higher activity levels were most likely to develop PPS. However, a population study found that PPS was more common in women, and the main risk factor in mild or moderate cases was a low recovery and the use of braces or mobility aids, rather than the high recovery found in more severe cases, suggesting two routes to PPS. Another larger population study found women were more at risk of developing PPS, and although the most severe cases and those over age 15 had the highest risk, all children under 15 had a similar risk, suggesting that the youngest, thought to have been affected less, probably had more hidden damage. The higher risk for women in these studies may relate to their having had more severe and widespread Polio. In another study focusing on recovery, the most significant risk factor was having overuse symptoms (fatigue, muscle pain, twitching and cramps) during the stable period after the initial Polio.

Since 2000, PPS research has focused on understanding the complexities of the closely related overuse symptoms, fatigue and muscle pain, which are more common in women and those who were younger when they had Polio; there is no correlation with weakness, but instead with intensity of activity.
3 Symptoms: Presentation and Assessment
3 Symptoms: presentation and assessment

3.1 Overview of presentation and assessment

Effective clinical management of PPS requires a careful assessment in order to select the most appropriate intervention(s).

3.1.1 Presentation

A Polio survivor may present with new symptoms such as weakness, pain, fatigue, respiratory difficulties or sleeplessness and they may or may not refer to prior Polio. Typically, following rehabilitation from the initial Polio infection, the person will have had a stable plateau of maximum recovery lasting decades, and adapted to the residual disability with a full and active lifestyle with no medical supervision or monitoring.

To cope with the demands of daily life, many people with prior Polio have operated with little functional reserve, using muscles close to their maximum and compensating for weakened limbs, for example by excessive use of shoulders, arms and hands. Symptoms such as pain or fatigue may be the result of new or chronic overuse of Polio-affected muscles.

The development of new symptoms can be gradual, sudden or step-wise at onset, sometimes but not always linked to events such as enforced bed rest, a fall or accident. As early symptoms such as fatigue are commonly experienced, Polio survivors may live with increasing problems for a considerable time before seeking medical help.

With unanticipated new symptoms, coping mechanisms which have been used for years may no longer be effective, triggering people to seek help through primary care.

Additionally, these new symptoms may reawaken the emotional distress experienced during the initial disease and first rehabilitation, making discussion difficult for patients.

3.1.2 Assessment

Table 1 is a checklist to assist effective assessment, particularly as health practitioners may not see many Polio survivors (with or without a PPS diagnosis) and many Polio survivors will never have had a condition-specific review.

Some Polio survivors may have had an assessment and diagnosis of PPS through a specialist clinic, but most will not, and some will not even be aware of the syndrome. Assessment is complicated by several factors. The fatigue, weakness and pain symptomatic of PPS can be caused by a wide range of other conditions which need to be excluded. Also, the presentation may resemble many other conditions such as chronic fatigue syndrome amongst other diagnoses. Finally, the symptoms, may fluctuate. Given this, it is not easy to pinpoint, and even experienced clinicians may not fully understand PPS unless they have had some personal experience of it or have had neurological or rehabilitation experience.
<table>
<thead>
<tr>
<th>Table 1. Checklist for assessment: this list is an aid to assist effective assessment, mostly distilled from guidelines with additions from the authors.5,4,92,93.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Patient history and current status to include:</strong></td>
</tr>
<tr>
<td>Polio infection and treatment (if known)</td>
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<tr>
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<td></td>
</tr>
<tr>
<td>History since recovery from initial infection</td>
</tr>
<tr>
<td>Present status</td>
</tr>
<tr>
<td><strong>Full symptoms checklist - onset, duration, location, triggers</strong></td>
</tr>
<tr>
<td>Fatigue - neuromuscular and general (inc. endurance)</td>
</tr>
<tr>
<td>Weakness/atrophy</td>
</tr>
<tr>
<td>Pain - muscular, joint, soft tissue</td>
</tr>
<tr>
<td>Respiratory status</td>
</tr>
<tr>
<td>Dysphagia/swallowing status (inc. risk of aspiration pneumonia)</td>
</tr>
<tr>
<td>Sleep quality</td>
</tr>
<tr>
<td>Psychological well-being (anxiety, stress)</td>
</tr>
<tr>
<td>Mobility (inc. endurance)</td>
</tr>
<tr>
<td>Spinal or joint deformities (inc. history of change)</td>
</tr>
<tr>
<td>Bowel/urinary function</td>
</tr>
<tr>
<td>Activity level - (activity diary)</td>
</tr>
<tr>
<td>Falls history/frequency</td>
</tr>
<tr>
<td>Medication review (for possible adverse impact on symptoms)</td>
</tr>
<tr>
<td>Weight history</td>
</tr>
<tr>
<td>Smoking history</td>
</tr>
<tr>
<td>ADL function - self-care ability (including cooking, shopping)</td>
</tr>
<tr>
<td>Aids, appliances and adaptations used</td>
</tr>
<tr>
<td>Osteoporosis/osteopenia</td>
</tr>
<tr>
<td><strong>Possible tests</strong></td>
</tr>
<tr>
<td>Physical examination</td>
</tr>
<tr>
<td>Blood chemistries</td>
</tr>
<tr>
<td>Cardiac dysfunction</td>
</tr>
<tr>
<td>EMG/nerve conduction tests</td>
</tr>
<tr>
<td>Pulmonary function tests</td>
</tr>
<tr>
<td>Sleep studies</td>
</tr>
<tr>
<td>Bone density</td>
</tr>
<tr>
<td>Balance tests</td>
</tr>
<tr>
<td>Gait assessment</td>
</tr>
<tr>
<td>Nutritional assessment</td>
</tr>
</tbody>
</table>
If reference to the accepted symptoms of PPS indicates that late effects of Polio are a possible cause, then a detailed history should be taken covering what is known of the initial infection and recovery, the period of stability and the onset of new symptoms and current activity levels. Assessment will need to include a subjective discussion to establish previous levels of function and suggest to what degree all four limbs, trunk and breathing are affected by the residual symptoms of acute Polio. Physical examination must include review of all four limbs with the patient both sitting, standing and walking (when relevant) to get an idea of how new muscle weakness is affecting key functions. A quantitative history including performance in repetitive or sustained activities such as walking or standing is essential to assess endurance. PPS symptoms can exacerbate each other; for example, poor sleep due to respiratory weakness can increase general fatigue. Hence a goal for the assessment is to identify the most critical symptoms in order to select effective management interventions. Some patients may have EMGs if there is doubt about the original Polio diagnosis. This can cause confusion about which muscles were or were not affected by Polio. These tests can show the existence of enlarged motor units suggesting Polio damage, but if a muscle has a normal EMG it does not necessarily exclude Polio damage, only that the damage may be in another part of the muscle, the motor units may have been damaged and incapable of enlarging, or the abnormalities may be below the limit the test is capable of detecting. In a study of clinically uninvolved Polio limbs in patients who had one limb showing residual weakness and atrophy, although 33/34 muscles had abnormal fibre density, 14 had no EMG abnormalities94.

3.1.3 General exclusions
Many of the symptoms of PPS are also symptoms of other conditions and so other neurological, medical and orthopaedic conditions need to be excluded. Table 2 gives a non-exhaustive list of other conditions that can mimic some of the symptoms of PPS which may be useful to consider when assessing a patient with PPS.

Table 2. This is a non-exhaustive list of possible conditions to be excluded, mostly distilled from guidelines with additions from the authors5,4,92,93.

<table>
<thead>
<tr>
<th>Exclusions or co-morbidities</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypothyroidism/thyroid disease</td>
<td>Multiple sclerosis</td>
</tr>
<tr>
<td>Cancer/chemotherapy</td>
<td>Parkinson’s disease</td>
</tr>
<tr>
<td>Anaemia</td>
<td>Spinal stenosis, cervical stenosis</td>
</tr>
<tr>
<td>Cardiac conditions</td>
<td>Myasthenia gravis</td>
</tr>
<tr>
<td>Dismuse atrophy (due to injury or treatment of injury or disease)</td>
<td>Radiculopathies</td>
</tr>
<tr>
<td>Diabetes, diabetic neuropathy</td>
<td>Neuropathies (including entrapment and peripheral)</td>
</tr>
<tr>
<td>Chronic infection</td>
<td>Multifocal motor conduction block</td>
</tr>
<tr>
<td>Renal disease</td>
<td>Cauda equina syndromes</td>
</tr>
<tr>
<td>Lupus</td>
<td>Spinal tumours and infarctions</td>
</tr>
<tr>
<td>Hepatic disease</td>
<td>Joint/spine deformity, scoliosis</td>
</tr>
<tr>
<td>Depression (clinical)</td>
<td>Uraemia</td>
</tr>
<tr>
<td>Inflammatory demyelinating disease</td>
<td>Toxins (including heavy metal)</td>
</tr>
<tr>
<td>Late onset genetic dystrophies</td>
<td>Myofascial pain syndromes</td>
</tr>
<tr>
<td>Adult spinal muscular atrophy</td>
<td>Degenerative disc disease</td>
</tr>
<tr>
<td>Inflammatory myopathies</td>
<td>Spondylolisthesis</td>
</tr>
<tr>
<td>Cerebral palsy</td>
<td>Ligamentous laxity/hypermobility</td>
</tr>
<tr>
<td>Mitochondrial or metabolic myopathies</td>
<td>Degenerative arthritis/traumatic arthritis</td>
</tr>
<tr>
<td>Polymyalgia rheumatica</td>
<td>Tendonitis</td>
</tr>
<tr>
<td>Motor neuron disease (MND)</td>
<td>Bursitis</td>
</tr>
<tr>
<td>Cerebrovascular disease</td>
<td>Fibromyalgia</td>
</tr>
</tbody>
</table>
3.2 Neuromuscular fatigue

3.2.1 Presentation

The most commonly reported problem is the peripheral physical fatigue resulting from Polio-affected muscles with limited endurance - neuromuscular fatigue. The short term reduction in power is induced by physical activity and exercise, but relieved by rest. However, there is usually an additional and permanent decline in muscle strength which results in the need to work harder to perform everyday activities such as walking, lifting and carrying. Unsupportive seating may also lead to local and then general fatigue. In PPS, abnormal muscle fatigability is out of proportion to the weakness of the muscle and may exist in muscles of any strength, even those previously thought to be unaffected.

The neuromuscular fatigue experienced by Polio survivors is different from the lack of energy and tiredness reported by healthy people. Common descriptions differing from controls are of ‘heavy sensation of the muscles’, an ‘increasing loss of strength during exercise’ and a lack of endurance. This is supported by a recent study of PPS patients at a UK clinic describing physical fatigue as ‘an increasing muscle weakness and loss of strength in previously Polio-affected and non-Polio affected limbs upon any physical exertion’. A follow-up study on 273 PPS patients showed very much higher levels of fatigue in comparison to controls. It can manifest as sudden onset of exhaustion after activity and can lead to localised aching pain. The patient can feel compelled to rest and the neuromuscular fatigue can become worse in the cold. Recovery times can also be longer. Patients graphically describe neuromuscular fatigue and the overuse symptoms it leads to, transient weakness, aching pain and general fatigue, including mental fatigue (see box).

“Fatigue develops in both legs after walking a few yards and I find it increasingly difficult to maintain my balance unless the surface is completely level. I have to really concentrate on how I’m going to put that foot to the ground and how I’m going to coordinate the next move with the gait of the leg.”

“When fatigued, sometimes I cannot even think how to move. Once I fell in the supermarket and could not think how to get up again.”

“Driving the car 15 minutes, my legs felt like jelly. After walking a short distance, they began to feel like blocks of wood, with a pain like being crushed by a heavy weight. I reached a kerb and came to a halt as my legs couldn’t work out what to do next. I finally imagined diving into a swimming pool and leaned forward, my legs having no choice but to follow.”

“I walked about 50 feet from the car into Waterstones, and the name of the author I wanted kept floating in and out of my mind. After walking another 30 feet through the shop, the aching was becoming intense and my mind became a blank. I needed to rest my legs in order to remember the author I wanted, so I sat on a little footstool used for reaching high books, and waited for my memory to return.”

‘Fatigue’ is a generally used term and several approaches have been used to understand its importance in PPS. General surveys show that fatigue is one of the most common symptoms. Qualitative studies have identified both physiological and psychological factors. Much of the quantitative research has used validated self-report instruments such as the Fatigue Severity Scale or the Piper Fatigue Scale to study the relative importance of different factors and to assess the efficacy of interventions. These instruments have been used on larger study populations of several hundred and show that physical fatigue was the most significant factor.

Physiological studies of neuromuscular fatigue involve much smaller groups of patients and have been used to study peripheral and central aspects of motor fatigue. Studies using both physiological measures and patient reported outcome measures
3.2.2 Impact of neuromuscular fatigue on the individual

The impact on the individual can be life changing due to: inability to perform tasks they could do previously, reduction in the distance they are able to walk, inability to compensate for weak legs by using arms/shoulders, reduction in what they can lift, and increased tendency to fall (no longer able to correct balance). The UK survey by Field found that 31% of Polio survivors left or changed work due to their condition and that 60% needed more help for day to day tasks.

Neuromuscular fatigue can also affect self-image, restrict lifestyle, and strain relationships.

3.2.3 Assessment of neuromuscular fatigue

As fatigue in PPS is multidimensional, the nature and severity of the fatigue should be assessed to determine whether it is peripheral, central, or due to other conditions before selecting interventions. Consideration should be given to whether it is local or general, related to any particular activities, the trend during the day and the response to rest. An activity diary recording type, intensity and aids used along with energy or fatigue levels can be used to identify high energy demanding activities, and possible modifications to lifestyle to reduce fatigue and optimum methods of treatment.

Self-report questionnaires which quantify fatigue can vary in their emphasis and sensitivity. Koopman, Beelen et al. found that the Fatigue Severity Scale (FSS) and Checklist Individual Strength (CiS) did not perform well, with low sensitivity, and concluded that they could be reliably used for groups but not for detecting small changes in individuals. The Piper Fatigue Scale has been shown to have validity and reliability for this patient group. The Functional Assessment Chronic Illness Therapy (Fatigue) questionnaire (FACIT-F) is also widely used to assess fatigue.

3.2.4 Recommended management of neuromuscular fatigue

Interventions covered in this document that are helpful in managing neuromuscular fatigue include:

- energy management: energy conservation and pacing are strongly recommended to alleviate symptoms and reduce overuse; (see Section 4.2)
- assistive aids and appliances can help with ADL (see Section 4.2.3)
- orthotics: effective lightweight bracing and orthoses can help support newly weakened limbs and reduce neuromuscular fatigue (see Section 4.4)
- weight management: reduced weight may help reduce physical stress and effort required (see Section 4.10.2)
- exercise: appropriate, individually prescribed exercise may help strengthen less-affected muscles, providing it is non-fatiguing and pain-free (see Section 4.7)
3.3 Neuromuscular weakness and atrophy

New neuromuscular weakness is one of the most commonly reported symptoms of PPS; atrophy is less common. Both can occur in previously Polio-affected and clinically unaffected muscles.

A 1995 survey of 1167 Polio survivors in the UK found that 77% had deteriorated since their best recovery with 57% reporting weakness with overwhelming effect\textsuperscript{15}. In a recent health survey 38% reported muscle atrophy\textsuperscript{14}. Other surveys showed that the weakness developed not only in previously ‘affected’ muscle (54% - 88%) but also in apparently unaffected muscles (15% - 77%)\textsuperscript{5,100,110,111}. Prevalence of new atrophy ranged from 17% to 59%.

Sites of new weakness included muscles in the legs, arms, trunk, and face, along with swallowing and respiratory muscles\textsuperscript{89}.

The most widely accepted aetiology leading to progressive, persistent weakness in PPS is the distal degeneration of axons of the enlarged motor units and loss of motor neurons, though the pathophysiology is not fully understood\textsuperscript{49}. Proposed underlying causes have included normal ageing coupled with the original loss, premature ageing due to increased metabolic demand of very enlarged motor units, disuse and overuse\textsuperscript{112}. Studies of muscle strength have shown that for measured strength as low as 19% of normal controls, maximum voluntary activation is >90% for leg muscles, though lower than normal controls (96-98%), suggesting that central drive is not a primary factor\textsuperscript{113}.

Measured changes in strength have shown wide variability; some shorter term studies (< 2 years) showed no change, but longer term studies showed losses of 15% in eight years\textsuperscript{46}. The reviewers suggest that follow-up periods need to exceed four years to effectively assess decline. The onset can be slow over many years\textsuperscript{46}, suddenly over a few months or linked to periods of inactivity for example due to hospitalisation\textsuperscript{15,114}. While measured group average decline can be low, individual decline can be substantial\textsuperscript{47}.

The most recent electromyographic study in 47 people with PPS found a decrease in knee extension strength from 64.3 Nm to 54.7 Nm in ten years, compared to 120.8 Nm to 85.6 Nm for normal controls\textsuperscript{53}. For people with PPS, strength decline was not correlated with age but related to the decrease in motor unit size (20% ±44) and the number of active motor units. Those with the largest initial MUs had the largest change in MU size and decrease in strength. Normal controls had no decline in MU size and a strong correlation of strength decline with age, indicating a different mechanism consistent with ‘normal’ ageing. The authors propose that the lower proportion of strength loss in people with PPS may be related to the higher proportion of muscle used for daily activity and to the predominance of type I muscle fibres. Given the low initial strength, the functional impact of muscle loss is likely to be greater for those with PPS.

Physical mobility over ten years has shown a modest average decline (14%) but high individual variability (nine participants lost 38%). Change in walking capacity was significantly correlated to change in quadriceps strength, but could not be predicted by any baseline parameter\textsuperscript{47}. The authors conclude that the variability in progression and lack of predictability support the need for personally tailored, monitored management.

Both disuse and overuse have been associated with loss of strength\textsuperscript{119}. Immobilisation can cause a rapid loss of muscle strength which can be difficult to rehabilitate even in healthy people\textsuperscript{115}. Agre et al.\textsuperscript{114} reported that the onset of decline in 44% of 79 Post Polio patients reviewed was associated with hospitalisation, although a contemporaneous study found that 83% of 23 patients had a prolonged PPS onset, and no subsequent studies have confirmed a link between hospitalisation and PPS onset\textsuperscript{116}. They also note that both disuse and overuse can co-exist in PPS where ambulation may over-exert weak muscles and underuse others.

3.3.1 Presentation

A person with new muscle weakness and/or atrophy may report new limitations in walking, lifting, carrying, getting up from a chair, reduced mobility, increased tendency to fall and difficulty managing ADL. Weakness may present with or without other symptoms such as pain and physical fatigue. If the new weakness has destabilised a joint, then joint pain may be the presenting symptom. In trying to maintain their normal activities, patients may be...
overusing weakened or compensating muscle groups leading to peripheral fatigue and pain. Identifying the primary symptoms requires careful assessment.

3.3.2 Impact

The impact of new weakness is that affected muscles are easily exhausted and require extended rest before recovering power. A recent case study comparing motor unit behaviour between an adult who contracted acute type III spinal Poliomyelitis at three months and three healthy volunteers showed that firing rates are reduced and that the entire motor pool was utilised for each contraction, demonstrating that the Polio survivor works harder even before PPS symptoms arise. This supports other studies of the high neurological energy cost of walking for people with PPS.

Similarly to neuromuscular fatigue, the impact of new weakness and/or atrophy can be life changing, reducing the ability to work and to cope with day-to-day activities.

3.3.3 Assessment of weakness and atrophy

Assessment of weakness is complicated as muscles that appear to be unaffected can have significant denervation. Also due to compensatory mechanisms, decreased strength is not linearly related to functions such as walking ability.

Measurement of weakness using the Medical Research Council rating has been shown to be insensitive to loss of strength in Polio affected muscles. Isometric muscle strength in upper and lower limbs of 32 symptomatic, ambulant Polio survivors, eight diagnosed with PPS, was measured using the MRC rating and on a dynamometer based system. Subjects scored 14% to 63% on average for leg muscle strength but this did not register on the MRC test. The subjects reported general muscle strength on a VAS did correlate with the dynamometer measurement. The MRC scale may also give a false impression of strength as one-off movements may be strong, but PPS patients typically show poor endurance over repeated movements or during sustained movements.

New weakness can be assessed from the patient’s history of change in ability or effort needed to perform tasks. This can be made semi-quantitative by recording the numbers of stairs climbed or distance walked without difficulty, but may not take account of compensation by less-affected muscles.

It is recommended that a patient’s history of change in function be taken along with physical performance tests. The history could usefully include changes in activity level due to work, illness or change in environment. Measurements of muscle strength should be done repeatedly to assess endurance and should be performed on isolated muscle groups.

3.3.3.1 Exclusions

It is important to assess the contribution of other causes of new muscle weakness and atrophy, for example:

- nerve entrapments, radiculopathies or spinal stenosis
- degenerative diseases such as multiple sclerosis or Parkinson’s disease
- spinal tumours, infections or inflammation (ischaemia)

3.3.4 Recommended management of weakness and atrophy

Interventions covered in this document that are helpful in managing new weakness and atrophy are:

- energy management: energy conservation and pacing are strongly recommended to alleviate symptoms and reduce overuse (see Section 4.2)
- assistive devices are strongly recommended to help with ADL (see Section 4.2.3)
- orthotics: it is strongly recommended that patients with new weakness are assessed to determine any need for effective lightweight bracing and orthoses. These can help support newly weakened limbs and improve physical function (see Section 4.4)
- weight management: reduced weight may help reduce physical stress and the effort required for certain activities (see Section 4.10.2)
- exercise: appropriate, individually prescribed exercise may help strengthen less-affected muscles (see Section 4.7)
- falls prevention: may help reduce risk of falling due to reduced ability to balance (see Section 4.10.6)
3.4 General fatigue

3.4.1 Presentation

Fatigue in PPS is considered to be multidimensional in character with general, mental and emotional aspects in addition to the neuromuscular fatigue discussed in Section 3.2. Systemic or general exhaustion has been described as ‘hitting a brick wall’, a feeling of being utterly drained, brought on by activity and relieved by rest or sometimes unrelated to exertion and with a long recovery time. In one UK survey, 80% of Post Polio patients had general fatigue which was caused by activity and relieved by rest and 39% had constant fatigue. Mental or cognitive fatigue includes concentration, memory difficulties and lack of motivation. Many studies and population and clinic-based epidemiological surveys loosely use the undifferentiated term ‘fatigue’, compromising estimates of prevalence and the impact of different aspects of fatigue. Neuromuscular fatigue and weakness are easier to measure and as a result are more researched.

The aetiology of the general and mental aspects of fatigue in PPS is poorly understood and many causes have been proposed. These include the impact of other PPS symptoms such as pain, weakness, neuromuscular fatigue, respiratory and sleep problems as well as damage to the brainstem reticular activating system (in the acute phase). Poliomyelitis is considered to be mainly a disease affecting spinal cord motor neurons; however virus activity resulted in severe lesions in the brainstem (reticular formation, vestibular nuclei) with less severe lesions in the cerebellum and the cerebral cortex (precentral gyrus). Bruno, Creange et al. have proposed that this damage could play a role in general and cognitive fatigue in PPS.

Recent studies of fatigue have used the Multi-dimensional Fatigue Inventory (MFI-20) which distinguishes between general, physical and mental fatigue along with reduced activity and motivation. Trojan et al. assessed the association of a wide range of factors with general, physical and mental fatigue. Increased general fatigue was found to be significantly correlated with higher stress (r=0.63) and to a slightly lesser extent with poorer sleep, lower self efficacy, increased pain, poorer respiratory function, higher depression and lower physical activity. The multivariate model explained a major proportion (51% - 56%) of the variance in general fatigue. Increased mental fatigue was correlated with increased stress (r=0.47), depression and self-efficacy. The multivariate model only explained 22% of the variance in mental fatigue indicating that other factors are important. Ostlund et al. in a larger study showed that those with higher general fatigue had worse sleep, more pain and lower scores in mental health related QoL (SF-36 using items such as ‘nervous’, ‘down in dumps’, ‘peaceful’, ‘blue/sad’, ‘happy’).

These studies show how different factors affect general and mental fatigue and illustrate the usefulness of a multidimensional tool to investigate the different aspects of fatigue in PPS. Some of these factors are also recognised PPS symptoms. Better management of modifiable factors such as stress, pain, respiratory problems and sleep may help reduce general and mental fatigue.

3.4.2 Impact

Similarly to neuromuscular fatigue, the impact of general and mental fatigue can be life-changing due to the person’s reduced ability to work, to perform daily activities and to socialise. While there is a lack of differentiation of aspects of fatigue in population studies, qualitative studies indicate that overwhelming general fatigue can be the most distressing and difficult to treat.

3.4.3 Assessment

As recommended for neuromuscular fatigue and weakness, the nature and severity of the general fatigue should be assessed to determine whether it is peripheral or central, or due to other conditions, before selecting an intervention. Consideration should be given to whether it is related to any particular activities, the trend during the day and the response to rest. For example, fatigue in the morning may be more likely to be due to poor sleep. A diary recording activity type and level along with any aids used may be useful.

The role of other commonly reported PPS symptoms such as pain, respiratory problems and sleep quality should be assessed, together with psychological factors (stress, depression) which...
can increase general and mental fatigue (see Section 3.7).

The FSS is commonly used to assess fatigue, but does not distinguish between the different aspects. The Piper Fatigue Scale allows for qualitative responses and may be more informative.

3.4.3.1 Exclusions

Fatigue is a symptom of many conditions and it is important to assess the contribution of other causes, for example, anaemia, thyroid, cardiovascular disease, diabetes, sleep or breathing problems, stress or depression.

3.4.4 Recommended management of general fatigue

- energy management: energy conservation and pacing are strongly recommended to alleviate symptoms and reduce overuse (see Section 4.2)
- assistive aids and appliances can help with ADL (see Section 4.2.3)
- orthotics: effective lightweight bracing and orthoses may help reduce general fatigue (see Section 4.4)
- exercise: appropriate, individually prescribed exercise may help alleviate symptoms providing it is non-fatiguing and pain-free (see Section 4.7)
- psychological interventions may help reduce stress (see Section 4.8)
- weight management: reduced weight may help reduce physical stress and effort required (see Section 4.10.2)
- management of respiratory problems contributing to general fatigue may help (see Section 4.6)
- pain management may help alleviate general fatigue (see Section 4.3)

3.5 Pain in PPS

Muscle and joint pain are two of the most common symptoms of PPS. In one study, 68% of 126 patients had muscle pain and 78% had joint pain71. Muscle pain, an aching pain caused by overuse, is more common in younger women with an earlier initial Polio age and a longer period of general and muscle fatigue, while joint pain is found in younger women with higher levels of initial and new weakness, suggesting two different pathologies71. Several studies found a close correlation between pain and fatigue, so management of the two symptoms needs to proceed in tandem124-126. That pain is more common among PPS patients who are younger and have a younger initial Polio age, particularly women, has been backed up more recently by Werhagen et al.70. Muscle pain has been found not to correlate with weakness, but to relate to level of activity and perceived exertion, suggesting a problem with abnormal muscle fatigability91,127. PPS neuromuscular fatigue leads to several overuse symptoms such as aching pain, transient weakness and general fatigue, including mental fatigue. One study found that those who had overuse symptoms such as muscle pain and fatigue in the stable period were most at risk of developing PPS89. Although there is very little evidence about the long-term effects of exercise or high activity levels, in a comparison of Post Polio patients in two countries with different attitudes to disability and rehabilitation, those who were most active had the most pain128. Another study found that those with higher CK levels, indicating muscle damage, were also in the higher activity group129. Neuropathic pain occurs when there is actual nerve damage and is characterised by a sharp or burning pain. About 10% of Post Polio patients experience nerve pain, arising from nerve entrapments or other illness or injury70.

3.5.1 Presentation

In a recent study130, 91% of 63 PPS patients surveyed experienced pain, and one third experienced it constantly. Moderate pain intensity was experienced with a mean duration of 20 years. Pain was most common in the shoulders, legs and lower back. In daily activities, the pain interfered with sleep, mobility, recreational activities and work. Another study70, in which 67% of 163 patients
had pain, looked at two types of pain, nociceptive (aching with signs of inflammation) and neuropathic. Almost all the patients in this study had nociceptive pain and those with neuropathic pain were found to have another problem such as disk herniation, cervical spondylosis, polyneuropathy, or peripheral nerve injury. In a qualitative study of PPS pain, the most common locations for muscle pain were lower back, upper legs and neck, and for joint pain, knees, wrists, elbows, and neck. Most pain was described as dull (44%) followed by stabbing (22%). In this study 44% of patients experienced pain every day, 52% had the most pain in the evening or night, and 33% said that the pain depended on what they were doing during the day or had done the previous day.

Muscle pain is usually caused by overuse, and is related to activity and relieved by rest, unless it has become chronic. Overuse pain is a sign of muscle strain and muscle fatigue. It is felt as a heaviness in the limbs, stiffness, throbbing, cramps, fasciculations, and a deep aching pain that may necessitate rest. It may lead to a general physical and mental fatigue.

Joint and biomechanical pain may be caused by muscle weakness leading to instability of joints, or abnormal biomechanics due to muscle imbalance and asymmetry and is often felt as a sharper pain than muscle pain. Scoliosis is one common form of severe skeletal misalignment in Polio patients which may deteriorate over time.

### 3.5.2 Assessment and exclusion

PPS muscle and joint pain should be distinguished from overuse injuries, which are very common in Polio patients, neuropathic pain from peripheral nerve injuries, such as nerve entrapments, disk herniation, and spondylosis; and pain from osteoporosis and osteoarthritis by taking a detailed history followed by the appropriate X-rays, scans, and neurophysiological tests. Muscle fatigue, which leads to overuse pain, is assessed by repetitive muscle testing.

### 3.5.3 Recommended management of pain

The management of PPS muscle and joint pain and the most common overuse injuries as a whole is reviewed in Section 4.3.

See also:

- energy management: energy conservation and pacing are strongly recommended to reduce overuse (see Section 4.2)
- pharmacological management is strongly recommended to alleviate pain (see Section 4.5)
- assistive devices can help with ADL (see Section 4.2.3)
- orthotics: effective lightweight bracing and orthoses can help support newly weakened limbs and improve physical function (see Section 4.4)
- weight management: reduced weight may help reduce physical stress and effort required (see Section 4.10.2)
3.6 Respiratory complications

The purpose of this section is to provide UK specific guidelines for primary care health professionals, in the management of respiratory complications in patients with prior Polio and/or Post Polio Syndrome. Due to the lack of studies in an exclusively Polio population this guide includes evidence taken from a wider group of patients with neuromuscular disease or chest wall deformities.

3.6.1 Presentation of respiratory complications

Respiratory complications can present in a number of ways in this patient population. A small proportion of patients will have had on-going respiratory insufficiency since the original Polio. The recovery phase after acute Polio would have allowed the vast majority of survivors to recover their respiratory function but the amount of muscle recovery varies between individuals. Some survivors of Polio did not recover enough respiratory muscle strength to maintain respiration independently and remained on bi-level ventilation throughout the 24 hour period or nocturnally. These patients will have required ongoing medical and respiratory review throughout their lifetimes.

The majority of patients who have not required respiratory support since the original Polio may still present with new respiratory complications many years later. Each patient’s circumstances should be considered including their risk of concurrent respiratory disease from factors such as smoking.

The new respiratory issues can be categorised under the four main headings; sleep disturbed breathing, muscle weakness leading to respiratory insufficiency, restriction of the chest wall leading to reduction of respiratory volumes and secretion retention.

Symptoms that patients present with which may indicate a respiratory complication include: breathing difficulties, shortness of breath, morning headaches, daytime somnolence, difficulty sleeping, problems with swallowing, choking or changes in voice tone. Signs that may occur during respiratory impairment include: an increased respiratory rate, abdominal paradox, reduced chest expansion and use of the accessory respiratory muscles.

3.6.1.1 Sleep disturbed breathing

Obstructive sleep apnoea (OSA) presents as excessive sleepiness and unrefreshing sleep and would be evident to a bed partner through the presence of snoring and breath holds over-night. PPS patients may also present with central sleep apnoea (CSA) or a combination of both. Diagnosis is made through a sleep study or polysomnography. OSA and hypopnea syndrome are found relatively frequently in the general population. The estimated prevalence of OSA in the general population depends on the population subtype but there is a higher incidence in men and a strong association with body mass index. There is a small amount of data that suggests prevalence is higher again in a population of patients with previous Polio. OSA is characterised by repeated episodes of upper-airway collapse during periods of Rapid Eye Movement sleep (REM). During REM sleep the only muscle functioning to maintain adequate breathing is the diaphragm as the other respiratory muscle are paralysed during this sleep period. Therefore any additional upper airway resistance or diaphragm weakness will have a greater effect in REM than during other periods of sleep.

Central causes of sleep apnoea can also occur in this patient group but do not have an increased prevalence compared to a population without Polio and therefore will not be addressed by this guide.

3.6.1.2 Muscle weakness leading to respiratory insufficiency

The progressive weakness and fatigability of muscles, including the inspiratory respiratory muscles, can lead to chronic respiratory insufficiency, which is often first noticed at night with difficulty sleeping, inability to lie flat, frequent wakening, nightmares, waking with a choking feeling and morning headaches. The insufficiency may only become apparent during periods of increased demand such as gaining weight to the point of obesity or respiratory illness. Respiratory muscle insufficiency may present in isolation or in combination with a restrictive chest wall deformity. The degree of respiratory insufficiency will vary, based on the pattern and amount of respiratory muscle involvement and chest wall restriction. Often the respiratory insufficiency is first apparent at night time due to the impact of body position on
muscle function, resistance to breathing when in a lying position and the impact of stages of sleep on muscle function. There are two types of respiratory failure: Type 1 being the lack of oxygen and Type II being the retention of carbon dioxide. Type I respiratory failure is more associated with nocturnal respiratory difficulties while Type II respiratory failure is less common and is more associated with muscle weakness or fatigue during the day. Both should be considered and assessed if indicated.

3.6.1.3 Reduction of respiratory volume due to a restrictive chest wall

For some patients, the Polio affected the motor neurons serving the torso. Residual weakness and an imbalance in strength between pairs of muscles in the spine can lead to kyphosis, scoliosis or a combination of both. The resulting altered spine shape reduces the ability for the rib cage to produce a ‘bucket handle’ action on inspiration. This mechanical disadvantage combined with weakness leads to poor rib excursion resulting over time in a restrictive stiff rib cage. This then leads to areas of persistent atelectasis through collapse of small airways and alveoli. The reduced excursion of the rib cage can restrict the tidal volumes of each breath and reduce the patient’s vital capacity. This can result in chronic respiratory failure.

In both, a muscle weakness leading to respiratory insufficiency and the reduction of respiratory volume due to a restrictive chest wall, patients may have adequate respiratory function under normal circumstances but display more symptoms during periods of increased demand. Survivors of Polio may not have either the strength or endurance to cope with prolonged periods of increased respiratory demand. The combination of weaknesses of the muscles and reduced range of movement of the chest muscles and chest wall means the patient is not able to produce a larger breath volume when required. Therefore additional demands lead to rapid shallow breathing\(^\text{138}\) and this can quickly lead to reduced gas exchange and acute respiratory failure. Examples of conditions that can all lead to increased respiratory demand include respiratory illness, surgery or other causes of immobility and increased weight such as pregnancy or obesity. Patients with chronic respiratory failure may also show acute chronic symptoms, a worsening of their signs and symptoms during the period of additional burden. It would be expected that nocturnal hypoventilation precedes day time hypercapnia but the nature of insidious decline may mean that patients whose breathing is challenged by their restrictive chest wall or muscular weakness are not picked up until a crisis arises in their respiratory function.

3.6.1.4 Secretion retention

Polio is not associated with an increased production of phlegm but removal of secretions during a chest infection can be an issue if the patient’s cough strength is poor. Similarly to the muscles of breathing, muscles involved in coughing may be adequate during normal circumstances but due to subclinical weakness may become ineffective when faced with the additional burden of a chest infection. An effective cough needs not only adequate volumes of inspired air but also a second compression phase against a closed glottis and a third phase to generate a large enough expiratory flow to clear secretions to the upper airways and to expectorate\(^\text{139}\). A reduction in muscle strength could affect the 1st or 3rd phase while a reduction in bulbar control the 2nd phase.

An inability to clear secretions puts people at risk of chest infections and a build-up of secretions can add to any chronic atelectasis resulting in inadequate gas exchange.

3.6.2 Assessment

3.6.2.1 Sleep disturbed breathing

Obstructive sleep apnoea (OSA) leads to short breath holds and reductions in oxygen levels. Patients rouse themselves multiple times an hour to regain adequate airway control and oxygen levels which leads to sleep fragmentation and poor sleep quality\(^\text{136}\). If sleep disturbed breathing is suspected, assessment should be carried out by a portable over-night oximetry test. Further testing via polysomnography should be carried out if the portable test indicates issues\(^\text{140}\). The polysomnography is used to measure breathing characteristics when asleep including breath holds. Treatment should be considered if the apnoea/hypopnoea index reaches moderate or severe
levels\textsuperscript{141}. An occurrence rate of 15-30 apnoea’s per hour is considered moderate and 30 or above is considered severe. The NICE guidance for the use of CPAP for treatment of OSA recommends symptom severity is also considered when deciding whether to treat.

For the Polio population, poor sleep quality can contribute to the fatigue symptoms patients may attribute to Post Polio Syndrome. Patients might describe generalised fatigue, day time sleepiness or not feeling refreshed on waking. It is clinically important to distinguish if the source of these fatigue symptoms is in fact sleep disordered breathing. The management approaches to fatigue caused by sleep disordered breathing or fatigue of other causes are very different. In the general population OSA is typically seen more prevalently in patients who snore, have more adipose tissue and larger necks. The evidence for OSA in patients with Polio suggest very similar characteristics, especially the presence of snoring which is highly associated with OSA in Polio survivors\textsuperscript{136}.

### 3.6.2.2 Muscle weakness leading to respiratory insufficiency

If there is any indication of respiratory insufficiency or a likelihood of it occurring a number of tests could be used to monitor the effect of this. Oxygen saturations via pulse oximetry, blood gases and lung function tests indicate the effectiveness of the respiratory system. Specifically, forced vital capacity (FVC) or vital capacity tests and sniff nasal inspiratory pressure or maximum inspiratory pressures (MIP) would give indications of muscle weakness effecting the respiratory system\textsuperscript{134}. Forced vital capacity should be measured in both the upright and supine (lying face-up) positions. Polysomnography may be used to make a diagnosis. Test values suggestive of respiratory insufficiency are:

- FVC $<$50\% of predicted,
- MIP $<$60 cm H$_2$O,
- PaCO$_2$ arterial blood gas $>$45 mm Hg,
- Nocturnal SpO$_2$ (oxygen saturation) $<$88\% for five continuous minutes while asleep\textsuperscript{142}

### 3.6.2.3 Reduction of respiratory volume due to a restrictive chest wall

The presence of a scoliosis, kyphosis or kyphoscoliosis does not mean a person will experience respiratory complications. A number of factors in combination have found to be predictive of abnormal lung function. These include the scoliotic and kyphotic angle, number of vertebrae involved in the curve, degree of rotation and the patient’s age\textsuperscript{143}. As there is a potential for spinal deformity to increase over time and respiratory muscle weakness to develop with increasing age, the need for respiratory mechanical support may develop in patient’s later years. Support could be required 24 hours a day but more commonly is only required nocturnally. The spinal shape should be monitored alongside lung volumes\textsuperscript{143} and blood gases. Blood gases should be used to measure the need for Non-Invasive Ventilation (NIV) for CO$_2$ retention and whether oxygen (O$_2$) is necessary either separately or with NIV. The aim of assessment is to detect if respiratory function is deteriorating so treatment can be provided in a timely manner.

### 3.6.2.4 Secretion retention

For patients who have reasons for reduced cough strength such as an admission with an acute respiratory infection or patients who report that they feel their cough is becoming weaker, the effectiveness of their cough should be reviewed. A normal strength cough is $\geq$360 litres per minute, $\leq$270 litres per minute is less effective and strategies to increase the peak cough flow should be introduced, $\leq$160 litres per minute is ineffective and secretion clearance strategies must be used\textsuperscript{144}. Peak cough flow should be a routine part of an assessment when considering secretion clearance capacities for any person over 12 year old with neuromuscular weaknesses\textsuperscript{145}.

### 3.6.3 Recommended management of respiratory complications

The recommended interventions are reviewed in Section 4.6.
3.7 Psychological symptoms

The psychological well-being of Polio survivors is impacted by their experiences of contracting and living with Polio and PPS.

Recent survey responses from 139 Polio survivors in the UK showed that 71% contracted Polio at age four or younger, with < one year old the most common age, 21% between five and ten years and the remaining 8% between 11 and 19 years old. The pain, paralysis and respiratory failure during the initial experience, frequently as a very young child, was traumatic. Treatment during the 1940s-60s often involved lengthy separation from parents and family in institutionalised environments with very restricted visits. This was before the reform of the care of children in UK hospitals.

Rehabilitation included pushing through pain and weakness; the advice at the time was to ‘use it or lose it’. For the individual, successful recovery meant at the time using the lowest level of appliances and aids and appearing as ‘normal’ as possible. Many made significant recovery from the acute infection and had demanding careers, some in quite physical jobs. Coping techniques were learned early and used effectively for decades.

The development of PPS by many Polio survivors can reawaken early fears, require a second adaptation to new disability and necessitate abandoning previously successful coping techniques. Polio survivors are often very resilient and many have had to keep adapting and changing for many years. In a qualitative study, Thoren-Jonsson traces the stages of adapting to PPS, from ignoring new symptoms and carrying on, reaching a crisis, avoiding activities to reduce symptoms, and then learning to adapt, use assistive devices, and take up the challenge of a different life.

Current studies show that Polio survivors display greater levels of emotional distress than population norms, although depression rates are higher for older people and those with chronic illnesses. The review by McNalley, Yorkston et al. of secondary health conditions in PPS reported a prevalence of depression ranging from 13% to 45% in sample sizes from 22 to 630. However, studies use different definitions and measures, so are difficult to compare. Tate, Forchheimer et al. studied Polio survivors and found low levels of depression, 15.8%, and those who were depressed had increased pain and poorer health generally. Kemp, Adams et al. showed that people diagnosed with PPS had more symptomology and a higher prevalence of depressive disorder, at 28%, than Polio survivors without PPS or nondisabled controls. Other studies have found that PPS had not affected quality of life or mental health, and depression scores were within normal limits. Clark, Dinsmore et al. found that PPS patients were within normal limits on a battery of psychological tests and that there were no significant correlations between fatigue levels and any of the psychological scores. Evaluating psychopathology in PPS patients can be particularly difficult because some measures include the physical symptoms of PPS, such as fatigue and sleep disturbance, as symptoms of depression.

From 27 people starting a UK rehabilitation course, Davidson, Auyeung et al. reported measures of anxiety (8.42, s.d. 4.44) and depression (6.62, s.d. 3.91) which are greater than normative values for the non-clinical, adult UK population (6.14, s.d. 3.76 and 3.68, s.d. 3.07). These numbers may be affected by the possibility of higher distress levels in people attending a rehabilitation programme compared to those in the community. However, the trend is consistent with other studies though the methods used and rates observed vary.

3.7.1 Recommended management of psychological symptoms

The recommended management is reviewed in Section 4.8.
3.8 Sleep

Besides the causes of insomnia common in the general population, there are four categories of sleep disturbance caused by PPS or more prevalent in the PPS population. These are obstructive and central sleep apnoea (see Section 3.6), restless legs syndrome or periodic limb movements, disturbances in sleep architecture, and sleep disturbance caused by overuse symptoms such as general fatigue and pain. These may all lead to disrupted sleep, frequent wakening and fatigue.

There has been much research into obstructive sleep apnoea (see section 3.6), and periodic limb movements and restless legs syndrome, all of which are more common in PPS patients than the general population.

The first study on abnormal movements in sleep in PPS patients was a small study of seven patients after a survey found that nearly two thirds of patients reported abnormal sleep movements. Two patients had generalised random myoclonus, two had periodic movements in sleep, two had periodic movements and restless leg syndrome (RLS), and one had contractions of the arm muscles at the beginning of sleep. Patients were treated with a low dose benzodiazepine, and, in the case of RLS, a dopaminergic agent was sometimes added. Further studies have confirmed that both periodic limb movements (PLM), which patients are often unaware of and are found during polysomnography, and restless legs syndrome, which are strange tingly feelings often in the most affected Polio leg, are significantly more common in PPS patients than in Polio patients without a diagnosis of PPS or in controls. In a retrospective study of 99 patients who had polysomnography, 16.6% had PLM (>5/hour) which is significantly higher than in the general population. Similar to sleep apnoea, PLM may disturb sleep and leads to daytime fatigue. The two conditions may occur together or separately in PPS patients and do not affect each other. In a study of 66 PPS patients, 63.6% had RLS versus 7.5% in the controls. RLS was found to have begun at the same time as PPS and is related to more fatigue and worse quality of life, although not daytime sleepiness. Both conditions are thought to be caused by deregulation of the dopaminergic system from the original Polio brainstem damage of the hypothalamus.

Two studies have looked further into PPS sleep problems, including sleep architecture. Siegel, McCutchen et al. studied 13 patients (7 bulbar) for the sequence of events leading to REM sleep and found a significantly longer latency in the bulbar group, suggesting it related to Polio damage of the reticular formation in the brainstem. A recent study of 60 relatively young PPS patients with sleep problems found them to have a pattern of abnormal sleep architecture although O₂ and CO₂ were within normal limits. The patients had lower sleep efficiency, higher arousal index, higher REM latency (length of time to first period of REM sleep) and a larger number of stage changes. Unexpectedly, up to 45% had morning headaches, often a sign of CO₂ retention. Ten patients (16.6%) used non-invasive nocturnal ventilation due to obstructive sleep apnoea (OSA). Thirteen patients (21.6%) had periodic limb movement (PLM) index >5. The sleep pattern was similar to other neuromuscular disorders with respiratory muscle weakness. Polio patients with mild respiratory muscle weakness or muscle fatigue may have altered sleep architecture with lighter sleep and less or later REM sleep, in order to prevent oxygen levels from dropping. Polio damage to the brainstem may also affect respiration, sleep arousals and REM latency. This change in sleep architecture may lead to nocturnal symptoms similar to those of respiratory insufficiency such as frequent wakening, inability to lie flat, waking with a choking feeling, feeling that their rooms are stuffy, and morning headaches.

A sleep study, or polysomnography, can be used to assess sleep disturbances. Treatment only exists for patients with OSA or PLM, while the remainder may find the resulting fatigue impacts on their quality of life and activity levels. A recent study of transcranial direct current stimulation has found improvement in sleep and fatigue in Post Polio patients.

Other possible PPS related causes of sleep disturbance are overuse pain and fatigue. Muscle overuse leading to physical and mental fatigue can cause difficulties getting to sleep or staying asleep. Reduction of activity in the evening and relaxation techniques may lessen pain, fatigue and stress, and aid sleep. Other common treatments used are drugs such as melatonin, antihistamines, valerian, amitriptyline and L-tryptophan.
3.8.1 Recommended management of sleep problems

Several other PPS symptoms can reduce sleep quality and it is recommended that these are addressed first:

- respiratory issues should be assessed (Section 3.6) and if present treated (Section 4.6)
- weight management may help if sleep apnoea is identified (see section 4.10.2)
- pain management may help (see Section 4.3)
- energy management may help (see Section 4.2)

Specialist referral may be required if sleep problems persist.

3.9 Dysphagia (swallowing)

Swallowing is the voluntary action of forming a food bolus in the mouth, transferring this to the oropharynx and then passing the bolus into the oesophagus. During this process the airway is fully protected. The centres for muscle action and coordination are in the brain and can be affected in bulbar Poliomyelitis. Shared anatomy and innervation are involved in airway protection, swallowing and voice production. The development of new weakness and fatigue in the swallowing mechanism as a result of Post Polio syndrome can place nutrition, the airway and the lungs at risk. Since Polio survivors frequently have diminished respiratory reserve, there is therefore an appreciable risk to health.

Difficulty with swallowing is a commonly reported problem for survivors of acute Polio (10%-15%) and those with PPS (10%-22%) and as many as 20%-60% of those who had acute bulbar Polio. A survey of Polio survivors in the USA showed that 27% of the sample population complained of swallowing difficulties. A subsample of twenty one individuals underwent video-fluoroscopy; twenty (95%) showed swallowing abnormalities. Evidence of bulbar Poliomyelitis was found in 75% of Polio survivors with swallowing difficulties. This study further showed that, in a randomly selected group of Polio survivors, 31 of 32 showed evidence of disordered swallowing with two subjects having some evidence of aspiration. In a more recent investigation, Söderholm, Lehtinen et al. found a prevalence of daily difficulties in swallowing or voice production of 29%. Disordered swallowing occurs in both paralytic and non-paralytic Polio survivors; Ramlow et al. found such problems in 6% of a non-paralytic sample and in 7% of a larger paralytic sample. A UK survey reported 18% with swallowing difficulties.

There is evidence of a slowly progressive deterioration similar to that of PPS and also some evidence that new symptoms may not correspond to the original symptoms and that people with mild symptoms may not report them.

Complications associated with difficulty swallowing, such as aspiration pneumonia, can be life-threatening and it is important that patients with PPS are assessed for possible risks.

3.9.1 Presentation:

The following symptoms and signs should raise concerns about dysphagia:

- prolonged mealtimes especially when fatigued
- choking or coughing especially with liquids
- occasional inability to complete a swallow
- voice change after eating (“gurgling”)
- unintended weight loss
- episodes of airway obstruction
- chest infections, including aspiration pneumonia

3.9.2 Assessment

A self-assessment questionnaire is a first step in determining the level and impact of swallowing difficulty (an example is given in Sonies). Effects of medication and other medical conditions or surgical procedures need to be excluded. Any tests which include observing muscle action need to involve several repetitions to assess the impact of abnormal neuromuscular fatigue due to PPS on swallowing ability.
3.9.3 Recommended management of dysphagia

If dysphagia is suspected then referral, which should be classed as urgent if there is a significant risk of aspiration, to a Speech and Language Therapist is recommended as investigation is a specialist undertaking.

A careful history and assessment of the respiratory system, phonation and swallowing function should be followed, when indicated, by a barium swallow and fibre-optic endoscopic examination of the swallowing process.

The referral should advise that any tests which involving observing muscle action need to involve several repetitions to assess the impact of abnormal neuromuscular fatigue due to PPS on swallowing ability.

3.9.3.1 Precautions

The patient history with Polio and the possibility of sub-clinical damage needs to be taken into account when assessing and prescribing tests or treatments.

3.9.3.2 Monitoring

Because weakness in PPS can progress, regular review of any Polio survivor who complains of swallowing difficulty should be undertaken with specialist referral and investigation as above as necessary. Lung function should be assessed during each review.

3.10 Cold intolerance among Polio survivors

Cold intolerance is defined as an abnormal sensitivity to a cold environment or low ambient temperature. It is a commonly reported new symptom among people with PPS, with prevalence rates in some studies between 29% and 56%, consistent with a survey in Lothian reporting 70% with cold intolerance, over half of long standing. Reported effects of cold exposure are increased weakness (62%), muscle pain (60%) and fatigue (39%). The challenge of a cold environment seems to increase with advancing age.

3.10.1 Pathophysiology

Normal thermoregulation when faced with the challenge of a cold temperature depends on healthy muscle activity (vigorous movement and shivering, fluid return from the extremities), normal functioning of the sympathetic nervous system (vasoconstriction to reduce heat loss) and intact control of body temperature (hypothalamic function). Post-mortem investigations have shown that the Polio virus attacks all of these components of the thermoregulatory mechanism.

There are very few studies of the cause of cold intolerance in Polio survivors. A study of motor and sensory functioning of hands in Post Polio patients at low temperature (dropping from 30°C to 20°C) found lower mean blood flow in comparison with controls indicating damaged sympathetic vasoconstriction neurons. Hand function performance also worsened. This is supported by a recent study which measured lower sympathetic skin response in comparison to controls.

The ‘perfect storm’ of weakened muscles, poor sympathetic activity and hypothalamic dysfunction can mean that Polio survivors’ ability to restrict warm blood flow to the skin with lower temperature is damaged leading to cold extremities (especially the feet - the “Polio foot”), profound discomfort and increased pain.
3.10.2 Presentation and impact

Although commonly experienced, cold intolerance can be underreported in clinics due to more pressing problems. The symptom affects comfort and function as mentioned above. It can have an impact on quality of life through the discomfort and pain of feeling cold. The increased peripheral fatigue can restrict activities especially in the winter. The financial cost of keeping warm can be significant.

Exacerbating factors include poor self care (for example nutrition, inadequate clothing) and environmental deficiencies (low ambient temperature, inappropriate heat sources, poor insulation, drafts).

3.10.3 Assessment

Assessment is based on the patient’s history and exclusion of other conditions.

3.10.3.1 Exclusions

Cold intolerance can be affected by other conditions or medications such as:

- Anaemia
- Anorexia nervosa
- Hypothyroidism
- Hypothalamic disorder
- Chronic ill health (e.g. SLE, chronic liver, lung or cerebrovascular)
- Depression
- Medication: e.g. antihypertensive drugs (e.g. blockers), sedatives (e.g. diazepam), antipsychotics (chlorpromazine), alcohol, opiates
- Peripheral vascular disease
- Raynaud’s disease
- Scleroderma

3.10.4 Recommended management of cold intolerance

Management is mostly based on self-care, though some interventions in this document may be helpful.

- Nutrition - (see Section 4.10.3)
- Self-management training (see Section 4.10.1)
- Pacing (see Section 4.2.2)
- Exercise (see Section 4.7)
- Some pharmaceuticals can help but are not licensed for this indication. These include:
  - Nifedipine, an antihypertensive, may be helpful, however Nifedipine and calcium channel blockers may also lead to side effects such as ankle swelling which may affect mobility
  - Naltrexone in low doses may help. This is a recommendation in the USA literature
  - Hormone replacement therapy has been used in appropriate clinical situations
  - Pregabalin has been used when cold related pain is predominant
  - Referral to a dietician to assess nutritional adequacy may help. An occupational therapist may be able to advise on lifestyle and environment changes
3.11 Bowel and urinary problems

While not a primary symptom of PPS, many Polio survivors experience bowel and urinary problems. These can arise from impaired muscles and nerves and difficulty in accessing toilets due to limited mobility\textsuperscript{177}. A recent survey of Polio survivors found that 76.5\% had ‘bothersome’ bladder symptoms and that the prevalence was about double that of the normal population\textsuperscript{178}. Johnson, Hubbard et al.\textsuperscript{179} also found that bladder disorders were common among people with PPS, but noted no voiding dysfunction. One survey of gastrointestinal problems in 754 PPS patients found symptoms suggesting extensive gastrointestinal dysmotility from the oropharynx to the colon\textsuperscript{180}.

3.11.1 Assessment

After screening to exclude other diseases, assessment should include a patient history, mobility assessment, a voiding and drinking diary and a pelvic floor assessment\textsuperscript{181}.

3.11.2 Recommended management

Incontinence may be addressed by changes in toilet habits, fluid habits, bathroom access and equipment, pelvic floor training and sometimes laxatives. Diet and particularly fibre may help with constipation.

Continuing problems may need specialist referral.

3.11.2.1 Precautions

The possibility of weak or sub-clinical Polio-damaged pelvic floor muscles must be borne in mind when prescribing muscle training.
4 Clinical Management
4 Clinical management

4.1 Multidisciplinary approach

Because of the wide range of symptoms and the psychosocial impact of early trauma and life-long disability, a multidisciplinary management approach is required. The core multidisciplinary team can include GPs, physiotherapists, occupational therapists, dieticians and psychotherapists with referral to rehabilitation consultants, neurologists and orthotists among others. These disciplines form the teams in rehabilitation programmes developed in specialist Post Polio clinics worldwide. The long term holistic care needed should be reflected in an individualised management programme for the patient’s unique set of symptoms (these can vary widely) while ensuring continuity of care and helping improve patient understanding and self-management skills.

The principal management interventions are energy and pain management together with optimising orthotics, aids and appliances to improve neuromuscular fatigue, manage weakness and pain and to reduce overuse. Table 3 summarises the recommended interventions for PPS symptoms.

Management also needs to include treatment of other conditions which occur commonly in Polio survivors such as osteoporosis and peripheral neuropathies. A recent Irish study of 50 Post Polio patients found that 56% had osteoporosis and 40% had osteopenia and that 38% had fractured bones in the preceding five years. In a study of electrodiagnostic findings in 100 Post Polio patients, 49% had additional peripheral neurologic disorders. Bladder and bowel symptoms may also need to be addressed (see Section 3.11).

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

In many localities, there are Proactive Care Multidisciplinary Teams who identify patients with complex needs and develop personalised care and support plans. Some patients with PPS may benefit from referral to such teams.
Table 3. Symptoms and recommended interventions - the table summarises recommended interventions for PPS symptoms and the appropriate health professions. This is only a guide as interventions need to be individually prescribed; see the text for precautions when prescribing exercise, physical activity and weight management.

<table>
<thead>
<tr>
<th>Intervention</th>
<th>Principal health professional for delivery of the intervention</th>
<th>Neurmuscular fatigue</th>
<th>Neuromuscular weakness, atrophy</th>
<th>General fatigue</th>
<th>Pain</th>
<th>Respiratory</th>
<th>Psychological symptoms</th>
<th>Sleep problems</th>
<th>Dysphagia (Swallowing)</th>
<th>Cold intolerance</th>
<th>Bowel and urinary problems</th>
</tr>
</thead>
<tbody>
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<td>Pacing</td>
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**KEY:** ☀ ☀ ☀ - strongly recommended; ☀ ☀ - recommended for some; ☀ ☀ ! - recommended for some with precautions (see text); ✳ - may help; ✳ ✳ - may help with precautions (see text);
4.2 Energy management

Several energy management approaches are used to alleviate the symptoms of PPS such as neuromuscular fatigue, weakness and pain.

4.2.1 Energy conservation

Energy conservation consists of strategies for using energy more efficiently by adapting or reducing activities and this requires activity or lifestyle modifications such as:

- simplifying tasks at work and home (cutting out unnecessary steps, planning simpler meals, reduced ironing, ordering food online, using a clothes dryer rather than hanging clothes out)
- reorganising the timing of tasks (doing chores less often, spreading them out across the day with rests between, or spreading them across the week)
- modifying tasks (sitting rather than standing)
- delegating tasks to someone else (friend, family, carer, hired help)
- reducing or changing activities (doing a certain activity less often or for less time, or changing from a more physically challenging activity to a physically easier one) e.g. having a shower rather than a bath)
- deciding some tasks are unnecessary
- mix and match activities (instead of having two physical activities at one go; e.g. shower and then cleaning house, mix and match physical and mental activities - sorting out mails and then cleaning the house)

In the case of muscle weakness, modifying tasks or the environment are the first priority in order to maintain other desired activities. For neuromuscular fatigue, reducing the time spent on tasks or introducing frequent rests and special seating, would be the first step, followed by altering and reducing tasks if necessary.

Conservation of energy in performing the activities of daily life can reduce the level of neuromuscular fatigue and pain experienced. Occupational therapists can help people with PPS to examine their daily activities and related fatigue/energy level, and advise on pacing, through the process of prioritising, adapting and simplifying tasks and the use of assistive devices both in the home and at work.187,188

Assistive devices are another means of conserving energy and may help in the management of fatigue and pain (see Section 4.2.3).

4.2.2 Pacing

Training to pace activities has been successfully used in multi-disciplinary PPS rehabilitation programmes in the UK and elsewhere.7,111,162,183 The primary aim is to prevent ‘boom and bust’ cycles of over activity or under-activity leading to periods of acute symptoms.

In some instances, pacing may enable people to achieve more by stopping activity before fatigue occurs and allowing the system to rest before resuming. In a group of seven Post Polio subjects with grade 4 quadriceps muscle strength, Agre and Rodriguez189 have shown that activity interspersed with rest breaks reduced muscle fatigue and increased performance capacity.

Referral to a specialist physiotherapist with experience in neurological condition management or occupational therapist with experience in fatigue management is recommended for assessment and effective training in strategies including pacing, adaptation of tasks and prioritisation. This will help individuals maintain activities of value and importance while avoiding over-use and excessive fatigue.

4.2.3 Assistive aids and appliances

Assistive devices include any equipment or technology that helps in energy conservation, management of neuromuscular fatigue, pain relief, lifestyle modification, improving function and reducing risks such as falls.

Patients can be assessed by hospital or community occupational therapists. Referral can be made by patient themselves, via GP or by the acute hospital team if patient is under their care. Some smaller items such as kitchen aids or functional chairs or stair lifts can be privately purchased.

PPS patients are often reluctant to use assistive devices and mobility aids, which may eliminate symptoms and stabilise their deterioration, but...
remind them of the original Polio. However, a new brace, wheelchair or scooter will often bring independence and a return to activities that had to be given up.

Frequently used aids for people with PPS include:

- Mobility aids – orthotics, crutches, walking sticks, mobility scooters, walkers, wheelchairs (manual or power): referral made to the local wheelchair team
- Manual handling equipment – hoist or standing and transfer aids (e.g. bed lever, bath board, toilet frame)
- Personal care aids – single-arm use dressing technique or long handled aids
- Joint protection aids – e.g. large handled tap turner, key turner or kettle tipper
- Hand weakness and carpal tunnel syndrome – wrist splint. Referral to the local hand therapy for further treatment plan. Advice on reducing activities related to wrist extension
- Ergonomics – to have optimised furniture height at home or work (e.g. chair/settee raisers, raised toilets)
- Household adaptations
- Computer adaptations and interfaces such as voice recognition

4.3 Pain management

The mainstays of pain management are pacing, activity or lifestyle modification, energy conservation, activity reduction and assistive devices. Overuse pain on a daily basis is a sign that the muscles are being used maximally and activity should be reduced to a point where the muscles have some reserve.

Pacing involves recognising activity time limits before symptoms set in, and taking regular rests between bouts of activity.

Energy conservation involves strategies of using energy efficiently by simplifying tasks breaking up tasks, changing activities from more to less physically taxing ones, or doing some tasks less often or not at all. Whereas, in the case of weakness, energy conservation involves altering activities that may have become very difficult or impossible, sometimes neuromuscular fatigue and overuse pain may be alleviated by limiting the time of the activities.

Activity or lifestyle modification is similar to energy conservation but focuses on simplifying or reducing different aspects of daily activity such as housework, social life, walking and travel, outdoor activity and exercise, diet, or rest. Assistive devices used for managing pain may include equipment or technology such as mobility aids, household adaptations or computer adaptations such as voice recognition. Orthoses have been shown to improve walking ability and reduce pain. Sometimes a wheelchair or scooter, at first possibly only used outside, is the only way to control overuse pain and fatigue, and can open up new opportunities.

In one study that looked at strength, activity and PPS pain, it was not strength or activity that was associated with pain but higher strain, activity intensity and perceived exertion. Activities were being performed at a level close to maximal capacity and the authors concluded that daily activity needs to be modified to reduce muscle strain. The Post Polio group also had higher activity levels than the controls. It is important for clinicians to understand that PPS patients who recovered well with little or no residual weakness may still have significant problems with Polio-related neuromuscular fatigue and pain without marked new weakness.

In a study by Stoelb et al., patients were asked about the efficacy of a wide range of other treatments, although not the most commonly advised – pacing, energy conservation and rest. The treatments that were most effective and used by the most people were massage, physical therapy, heat, ice, range of motion exercises, narcotics, paracetamol and aspirin. Less used, but very effective, were nerve blocks, marijuana, diazepam and carbamazepine (Tegretol). Chiropractic was also relatively effective. Gabapentin is often prescribed for pain in PPS but was not found to be very effective in this sample. In the study by Widar et al., 78% of the sample found pain was alleviated by rest, 48% by heat, and 44% by medication. Other helpful therapies mentioned were physical therapy, massage...
and relaxation. Massage has been found to reduce inflammation and promote new growth of mitochondria in muscles damaged through exercise. Activity is encouraged in all people for general health, fitness and mobility, but exercises are not prescribed for muscles experiencing overuse pain or a feeling of increased weakness. Those with the most widespread neuromuscular fatigue and overuse pain (in many parts of the body), who may have very few strong muscles able to compensate for weaker ones, need to be most careful about exercise. Nollet et al. found that PPS patients with the most symptoms, indicating the most overuse, and most widespread weakness, rather than the most severe weakness, had more decline in physical functioning. If overuse pain can be controlled through energy conservation so that muscles have some reserve, then a non-fatiguing exercise programme can be slowly implemented as long as pain does not develop. In the only unselected trial of non-fatiguing, strengthening exercise in 23 PPS patients without pain, only half had enough reserve to benefit. The others had no gain or their symptoms increased.

4.3.1 Evidence-based treatment for pain
Orthotics: The prescription of new or improved orthotics has been shown to improve walking ability and reduce pain.

Lamotrigine: One small, randomised, controlled study (15 patients, 15 controls) of Lamotrigine (50-100 mg) showed significant improvements for the patient group in fatigue, pain and health-related quality of life at two and four weeks and was well tolerated. Although Lamotrigine has been found to be effective for neuropathic pain, in these patients it was also effective for overuse pain which increased during and after physical activity.

Intravenous immunoglobulin: As an increased level of cytokines has been found in the cerebrospinal fluid, peripheral blood and muscle of PPS patients which has been reduced by intravenous immunoglobulin (IVIg) treatment, several studies have been conducted to test the effect of IVIg treatment. Some improvements were seen in pain and quality of life. One study which re-evaluated patients after one year found significant improvements in physical functioning, quality of life, pain reduction and walking ability, but the results of a smaller study did not reach significance after four months. As the results have been mixed, recent studies looked at the possibility of ‘responder’ subgroups. In a study of IVIg and pain, Werhagen et al. found that patients who had Polio before the age of ten and more pronounced weakness responded more to treatment. Another study of responders found that those who were below 65 years of age, had weakness in the lower limbs, had higher levels of pain and no other comorbidities were most likely to improve significantly in pain and quality of life. However, the effect of IVIg treatment on motor neuron loss is unknown. The inflammatory response could be primary and systemic or secondary to neurodegeneration and muscle overuse.

4.3.2 Types of pain and their management
In this section, the recommended management of muscle pain, joint pain and overuse soft tissue injuries, which are extremely common in weak and fatigued muscles, will be discussed.

Studies of pain in PPS give a good picture of the types of pain, their aetiology, and their effect on patients but the studies on management are more difficult to interpret as there is no differentiation between types of pain and the results are inconsistent. There is, however, some consensus about how the different types of pain in PPS patients should be managed. Reduction of overuse pain through modifying activity and using assistive devices can help in stabilising PPS deterioration, but this may be challenging as many Polio patients were brought up to lead normal lives and find it difficult to change their very active ways. Other common modalities for all kinds of pain are heat (heating pads, hot baths), relaxation, hydrotherapy, acupuncture, osteopathy and massage.

4.3.2.1 Muscle pain
The mainstays of management are reduction of activity, pacing, energy conservation and lifestyle modification, heat, cold, stretching, and assistive devices including orthotics. If pain cannot be managed through lifestyle changes or assistive devices, various categories of pain
medications are used; these must be prescribed as appropriate for the individual but could include anti-inflammatory drugs, antidepressants, anti-seizure medications and narcotics.

Exercise is not appropriate for muscles with overuse pain but it has been shown in Post Polio patients, although not necessarily patients with PPS, that strengthening pain-free leg muscles may be helpful in relieving the strain on painful arm muscles and thereby diminishing pain63. If overuse pain develops in a muscle while exercising, then the exercise should be stopped. After the muscle has been given time to rest and heal, a reduced exercise session, at 50% or 25% of the earlier session, could be tried197. Exercise recommendations, especially appropriate stretching and exercise aimed at increasing endurance of activity before muscle pain is induced, should be guided by a physiotherapist.

Myalgia may be drug-induced through statins, therefore a medication review and test of serum creatine kinase levels may be warranted together with withdrawal of the statin if appropriate.

4.3.2.3 Soft tissue injuries

Soft tissue injuries, due to muscle overuse, weakness, or asymmetry, such as tendinitis, bursitis, ligament injuries, are very common and should initially be managed conservatively with pacing (Section 4.2), lifestyle modifications, physiotherapy and assistive devices (Section 4.2.3). If conservative treatment is unsuccessful, further options may include cortisone injections and topical counter-stimulants.

4.3.2.4 Entrapment syndromes

The assessing clinician should remain vigilant to the possibility of peripheral nerve entrapment syndromes (for example ulnar neuropathy or carpal tunnel syndrome) and radiculopathy which may co-exist with PPS and may be difficult to identify in the presence of pre-existing muscle wasting. A vital clue suggesting further investigation is required is the presence of new sensory symptoms – numbness or tingling, since Polio is primarily a disorder of motor neurons.

4.3.2.2 Joint pain

Joint pain from instability of joints or abnormal biomechanics may be managed by modifying the use of joints and improving posture and body mechanics through physiotherapy, orthotics (Section 4.4) or other assistive devices (Section 4.2.3).

Careful assessment of the underlying causes of joint pain is recommended, particularly when the shoulders are involved. Imaging of the shoulder joints with ultrasound is often helpful in identifying the nature of pathology in the shoulder joints. For example sub-acromial bursitis may be identified and the extent of rotator cuff tears, if present, can be quantified.

Hip pain may also benefit from further investigation initially with plain X-rays and sometimes MRI scanning. Specific treatment may be recommended in the light of the results of such investigations.

4.4 Orthotics and footwear

People with PPS often are using orthotics that were prescribed many years before and may no longer be optimal following an increase in neuromuscular weakness and atrophy.

Correctly fitting orthotics and footwear used to support weak limbs, reduce pain and improve mobility can help reduce fatigue and overuse symptoms. Recent advances in lightweight orthoses have been shown to improve energy expenditure while walking198,199.

An assessment by an orthotist or approved healthcare professional is essential to ensure that the most effective devices are being used and no damage is being caused. This should be followed by regular review to ensure orthotics remain in good working order and are updated as needs change.
4.5 Pharmacological management of PPS

There are no medications currently available proven to reverse muscular atrophy, improve strength or relieve the neuromuscular fatigue of PPS although research is ongoing.

A recent Cochrane review looked at ten studies of pharmacological treatments (modafinil, intravenous immunoglobulin (IVig), pyridostigmine, lamotrigine, amantadine, prednisone) and found inconsistent evidence on the effectiveness on muscle strength\textsuperscript{108}. Of these, the only positive effect (though with low quality evidence) was shown by lamotrigine on activity limitation with inconsistent results for fatigue. The authors conclude that the lack of high quality data makes drawing definite conclusions impossible.

The observation of increased levels of cytokines in people with PPS indicating an inflammatory process has led to current trials of intravenous immunoglobulin to determine effectiveness\textsuperscript{200} (see Section 2.3.3).

Anabolic steroids are not recommended; they improve muscle bulk and power but the side effects, such as risk of prostate cancer in men and masculinisation in women, greatly outweigh the potential benefits.

Anti-oxidant medications and supplements have never been formally tested to verify any effect on the rate of progression of Post Polio Syndrome.

Metabolic stimulants such as L-carnitine and co-enzyme Q10, which are thought to improve the ability of muscles to make energy (and possibly reduce fatigue and improve strength) have also been studied but not proven to be effective\textsuperscript{201}. A clinical trial on the effect of coenzyme Q10 found that it did not reduce the fatigue of PPS\textsuperscript{202}.

Other medications are used to alleviate the individual symptoms of PPS such as pain (Section 4.3.2.1), sleep (Section 3.8) and cold intolerance (Section 3.10).

4.6 Management of respiratory complications

4.6.1 Evidence based interventions

4.6.1.1 Sleep disturbed breathing

The recommended treatment for patients diagnosed with OSA is continuous positive airway pressure (CPAP). It is also recommended that if patients are unable to adhere to CPAP or suffer adverse effects to the CPAP that a mandibular advancement device is considered\textsuperscript{203}.

4.6.1.2 Muscle weakness leading to respiratory insufficiency

Home mechanical ventilation, positive pressure ventilation, has mostly replaced negative pressure ventilation as the preferred ventilation method in the home.

NIV is the recommended treatment for established respiratory failure due to respiratory muscle weakness. Constant pressure or Bi-level non-invasive ventilation will be more appropriate depending on whether the patient presents in type I or type II respiratory failure. In the absence of other comorbidities causing interstitial lung disease, oxygen alone is not indicated but can be considered if clinically indicated and given with NIV support\textsuperscript{204}.

4.6.1.3 Reduction of respiratory volume due to restrictive chest wall

NIV is the recommended treatment for established respiratory failure due to a restrictive chest wall.

4.6.1.4 Secretion retention

It has been shown that, in a mixed group of patients with neuromuscular pathologies, mechanical devices such as the mechanical insufflator/ exsufflator (MI:E) produce a higher peak cough flow than manual techniques alone\textsuperscript{205}. If patients are admitted to hospital due to chest infections and have secretion retention, physiotherapy to aid secretion removal is indicated. There is some evidence that in a population of neuromuscular patients with acute chest infections, physiotherapy techniques combined with the MI:E reduces...
secretion removal treatment time compared to physiotherapy techniques without the addition of the Mi:E206.

However, a Cochrane review207 was unable to make a recommendation on the use of the Mi:E in the neuromuscular disease group due to the lack of high quality evidence. The recommendation of this guide is that the strength and effectiveness of the cough should be considered when a patient with old Polio or Post Polio Syndrome is admitted to hospital with an acute chest infection. Other techniques such as active cycle of breathing, positioning and huffing should be used first to explore the patient’s ability to expectorate, but if these simple techniques are not working therapists should quickly move to other techniques because of the poor respiratory muscle reserve and likelihood of tiring. The Mi:E device should be considered as a treatment option, especially in cases of poor cough strength.

4.6.2 Assessments and Interventions that may help

4.6.2.1 Reduction of respiratory volume due to restrictive chest wall

In this patient group, to gain adequate resolution of gas exchange, higher NIV pressures may be required. This may not be comfortable for the patient. In this situation, and also as it may prove to be more effective, NIV utilising volume control rather than pressure control could be trialled to overcome resistance to the delivery of the inspiratory breath.

It is recommended in both impairment of respiratory function due to restrictive chest wall and muscle weakness, that if the patient has an acute deterioration requiring admission to hospital, that NIV is started if the respiratory rate is above 20 and the vital capacity is less than one litre. This is even before the occurrence of hypercapnia. This is due to the reduced reserve of the respiratory system in these patient groups.

4.6.2.2 Secretion removal

For patients who have chronic retention of secretions, techniques that assist cough augmentation have been in use prior to the Mi:E device being available. Techniques include hyperinflation via breath stacking to aid inspiration and manual assisted coughing to aid expiration. They can be useful, and if effective, free the patient from the need to rely on equipment. However, a manual assisted cough (MAC) normally requires assistance from another person and can be uncomfortable as it requires pressure to be applied to the abdomen to produce the cough. The low level of evidence for these treatments suggests they can be used to produce a cough improvement with combined breath stacking and MAC being most effective139,144. A further combination of Mi:E and MAC can be used if neither technique is effective on its own.

It is also suggested if the patient has a vital capacity of less than 2000ml or 50% of predicted capacity, that breath stacking, practiced daily, can be helpful as part of prophylactic airway clearance144. For patients with very thick secretions, mucolytics could be considered, however this will need to be monitored to assess if this truly makes expectoration easier. There is no reason for patients with Polio and without other concurrent disease to have particularly viscous saliva or excess secretion production. In the situation of acute chest infection, mucolytics and other secretion removal techniques will have more relevance.
4.6.2.3 Bulbar dysfunction

Bulbar dysfunction, which has not been addressed in this guide, is an additional condition that may occur with the onset of Post Polio Syndrome. The prevalence amongst this patient group is unknown. It has been more frequently reviewed in other conditions such as motor neuron disease (MND), where bulbar dysfunction is a common progression of the disease. Within the MND group, bulbar dysfunction, alongside peak cough flow and peak cough flow/peak velocity time have been found to be reliable indicators of a cough that will become inadequate during an acute chest infection\textsuperscript{208}. This may be applicable to Post Polio Syndrome group as well but this would require further research to evaluate.

4.6.2.4 Vaccination and health promotion

As with other chronic diseases, pneumococcal and influenza vaccinations are prudent to offer given the respiratory issues already outlined\textsuperscript{209-211}. If a PPS patient smokes, smoking cessation advice and support can prevent worsening of their respiratory function and vascular complications. Dietary advice to optimise nutritional status and consideration of weight management may also support functional status.

4.7 Exercise

Once energy management techniques have been effectively learned, and pain controlled as far as possible, carefully designed and monitored exercise programmes can be considered. Physical activity and exercise are key components of PPS management interventions as they are important for cardiovascular health and may be able to strengthen unaffected muscles or less affected muscles that are not already being used maximally. Exercise may also promote relaxation; relaxation and promotion of sleep quality can be part of a planned approach to management of energy.

In the UK, the majority of people who contracted Polio did so in the 1940s and 50s and are now predominantly 50 to 75 years old. This is an age range where sarcopenia is accelerating; exercise is a key factor to treat this additional muscle loss\textsuperscript{212}. Studies of aerobic exercise in people with PPS have shown improvement in cardiovascular health\textsuperscript{213-217}. Likewise, resistance training has been shown to improve muscle strength\textsuperscript{201,218-220}.

Strengthening exercise is sometimes recommended for less affected muscles; however a systematic review of the evidence shows a lack of high quality studies and inconsistent results\textsuperscript{221}. These studies have shown improvement in objectively measured strength with no evidence of damage but tend to have small sample sizes and to be of short duration so give no information on long term effects\textsuperscript{201,218}.

However, all these studies involve carefully prescribed and supervised interventions. They have involved small groups of patients, generally under 60 years old, and selected by their ability to do exercise without increasing their symptoms; few involve longer term follow-up. One issue with assessing the exercise capability of an individual with PPS, is that muscles that appear clinically unaffected may have lost more than 50% of their motor neurons. Concern over damaging weak muscles through overwork has been a barrier to recommending exercise which can lead to a deconditioning cycle\textsuperscript{222}. Also, people with PPS are already using a high degree of energy in daily life functioning which should be factored into any calculation of their fitness. About 70% have long-standing overuse pain\textsuperscript{70} and 80% have moderate to severe fatigue\textsuperscript{126}, so it is recommended that patients try to minimise overuse pain and fatigue through lifestyle changes before beginning an exercise programme. A recent study found that severely fatigued patients did not improve in strength or cardiovascular fitness after a four-month exercise programme; the authors hypothesise this may be because their reduced muscle had already adapted optimally\textsuperscript{223}.

The recommendations for safe, effective exercise are\textsuperscript{224}:

- the capability of the individual is carefully assessed
- the exercise programme is individually tailored, started gradually, with periods of rest and take into account the longer recovery times required
- it should be pain-free and non-fatiguing
- people are monitored closely to identify any increases in muscle weakness or pain and
- progression to higher intensity is slow
Initial aerobic intensity levels at a rating of perceived exertion (RPE) of 11 on the Borg scale\textsuperscript{225} or 50-75\% of 3 repetition maximum (3RM) are advised\textsuperscript{226}. Aquatic exercise in warm water has also been shown to be beneficial\textsuperscript{216}. Warm water can provide an environment which reduces concerns such as falling, and the buoyancy can offer more movement to limbs which are constrained by the load of gravity and mechanisms of the limb on land. Anecdotally the main barrier to exercising in water is difficulty in accessing a pool. If this can be overcome, most patients find this an enjoyable and feasible way to exercise.

Strengthening exercise is not recommended for people with very weak or fatigued muscles who are already using all their strength for activities of daily life\textsuperscript{224}. As a basic guide, if the patient struggles to move their limb against gravity (and this is not due to a period of disuse), or has chronic injuries in the limb, then they will already be using the available muscle maximally during daily living.

It is recommended that referral to a specialist physiotherapist with experience in neurological condition management is essential for assessment, prescription and monitoring of a customised exercise or physical activity programme.

4.7.1 Precautions

**Overuse**

The cause of the ongoing denervation leading to PPS has not been proven but the stress or overuse of the neuromuscular system has been proposed as a possible factor\textsuperscript{219}. Hence it is important that any exercise programme is non-fatiguing and does not cause pain or increased weakness. For people with severe weakness and/or neuromuscular fatigue, exercise may not be appropriate as they may be using their muscles maximally in everyday life.

4.8 Psychological interventions

Psychological interventions may help people with PPS make the challenging behaviour and lifestyle changes necessary for the management approaches recommended in this document and also improve adherence. There are few studies on the efficacy of psychotherapy specific to Polio survivors either with or without a diagnosis of PPS.

A recent computer-based positive psychology intervention study included people with PPS and found positive results for pain control and depression\textsuperscript{227}. Koopman, Beelen et al.\textsuperscript{228} are conducting a trial of CBT in the management of PPS; however, early results have shown no significant effect when compared to usual care\textsuperscript{229}. This is not consistent with proven efficacy for a wide range of long-term conditions (see below) and it is possible that the study population from rehabilitation centres is being supported in behaviour change as part of this usual care and thus the result is not relevant for use of CBT in a non-specialist setting or for those seeking help with the symptoms of PPS for the first time.

The multi-disciplinary rehabilitation programme at St. Thomas’ Hospital has shown significant positive outcomes; this incorporated education on behaviour change, group discussions and goal setting\textsuperscript{162}.

For more general long term conditions, the King’s Fund has reviewed the links with psychological health, and reported that the overall evidence supports the increased incidence of depression and anxiety in this population group. It concludes that outcomes can be improved by integrating psychological support into the primary care of chronic physical conditions\textsuperscript{230}. This is further supported by the NICE Guideline CG\textsuperscript{91}, ‘Depression in adults with a chronic physical health problem’.

The use of psychological therapies for depression and anxiety is embedded in the NHS IAPT programme, ‘Improving Access to Psychological Therapies’\textsuperscript{231}. The related four year plan of action\textsuperscript{232} reported that people with long term physical conditions are between three and four times more likely to experience depression and anxiety than the general population while also being less likely to be referred for psychotherapy.
The plan includes an IAPT training module on psychological therapies for people with LTCs and development of a collaborative care approach. Recent evidence on the benefits of the IAPT programme for a range of long term physical conditions has shown increased adherence to treatment and proposes that improving psychological health may improve the ability to cope with physical symptoms\textsuperscript{233}. 

Taken together, the evidence for increased psychological symptoms amongst people with PPS and the benefits of psychological therapies for people with long term physical conditions, these show that such therapies have a role to play in the management of Post Polio Syndrome, both directly and by supporting behaviour change in other interventions.

**4.8.1 Recommendations**

The management of people with PPS should take into account their early experiences and how these may have influenced their psychological wellbeing, their response to new and/or increasing symptoms and the symbolic meaning of assistive devices.

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.

**4.9 Speech and language therapy**

Referral to a speech and language therapist with experience in neurological conditions is essential in order to correctly assess and devise a management strategy for the swallowing (and sometimes speech) difficulties experienced by Polio survivors with PPS\textsuperscript{168}.

**4.9.1 Precautions**

The patient history with Polio and PPS, and the possibility of sub clinical damage, needs to be taken into account when assessing and prescribing tests or treatments. Any tests involving observing muscle action need to involve several repetitions to assess the impact of abnormal neuromuscular fatigue due to PPS on swallowing ability.

PPS patients with signs of dysphagia should be reviewed periodically to monitor any symptom progression. It has been advised that even if symptoms are minimal, people with PPS often accommodate changes and can risk exacerbation of their swallowing difficulties\textsuperscript{172}.
4.10 Interventions that may help

4.10.1 Self-management training

Most of the interventions discussed here require significant behavioural changes as do other lifestyle changes which people with PPS find helpful\(^{101,234}\). All of these require effective self management skills. Enabling and supporting self management is a key service component in primary and allied health care\(^{235}\).

Referral to structured education programmes may be helpful; these range from symptom-specific (pain management) to generic training such as the Expert Patient Programme Chronic Disease Self Management Course (CDSMC)\(^{236,237}\). In a qualitative 12 month follow up study, three of nine people on a CCDSMC were diagnosed with PPS - all reported using the learned techniques at 12 month follow up\(^{238}\).

4.10.2 Weight management

A qualitative study of Polio survivors in the UK has shown that people can detect changes in performance with a weight difference of only a few pounds\(^{146}\). Excess weight requires more effort and weight management is recommended in PPS rehabilitation programmes\(^{6,239}\). It may help reduce the occurrence and severity of neuromuscular fatigue and pain, and to maintain function when faced with new weakness.

There are several issues in assessing whether or not a Polio survivor would benefit from weight loss and how best to prescribe a weight management programme (WMP). Studies have shown that their body composition is different from age matched controls with a lower proportion of lean mass indicating that the standard BMI categories may not be relevant\(^{240}\). For physically disabled people in general the standard energy expenditure equations are not as useful\(^{241,242}\).

Exercise as a component of a WMP may be less helpful due to mobility limitations on physical activity level (PAL)\(^{242}\) or in the ability to find and access suitable facilities such as warm swimming pools and accessible venues. Audits of WMP do not clearly indicate how well physically disabled people are accommodated.

Barriers to achieving a healthy weight may include the importance of food as a social activity, long-term comfort eating, a belief in the inability to exercise and feelings of fatigue and cold on a reduced calorie intake. Additionally, many have difficulty in shopping and cooking.

4.10.2.1 Precautions

The reduction in calorie intake necessary for weight loss in people with PPS, limited mobility and reduced muscle mass may be low enough to risk inadequate nutrition.

It is recommended that people are individually assessed, that supervision by a dietician is essential and that any WMP is undertaken gradually.

4.10.3 Nutrition

It is possible that good nutritional behaviour, both in ensuring adequate levels of nutrition and frequent regular meals, may help reduce the severity of general fatigue, improve bone health and bowel and urinary function. Referral to a dietician may prove helpful.

4.10.4 Hydrotherapy

Swimming and exercise in warm water has proved both enjoyable and beneficial for many Polio survivors. With new problems from PPS, a course of hydrotherapy may help relieve symptoms and also be useful in discovering how to safely exercise in water (see section 4.7).

4.10.5 Relaxation

As part of introducing regular rests, relaxation techniques could be helpful if patients are struggling to pace their activities. They are also useful to refresh mind and body in order to cope with daily stress or as aids to fall asleep.

There are various relaxation techniques available such as
- Jacobson’s muscle tense relaxation
- Using relaxed breathing
- Guided imaginary relaxation
- Jane Madders relaxation technique
Ideally an effective relaxation session should be 20-30 minutes long.

4.10.6 Falls prevention training

A Falls Prevention course may be helpful in minimising falls risks due to new weakness by advising on balance exercise and home adaptation.

There is some evidence that fall mechanisms in Polio survivors are different from those for elderly people. In a study by Bickerstaffe et al.105, it was found that Polio survivors fell mostly in the afternoon as opposed to morning or night in community-dwelling elderly people. Another difference was that more falls occurred in the home than just outside with the opposite true for older adults. Likewise, medication use and increasing age were not associated with falls in the Polio survivor population. The authors propose that muscle fatigue is a contributor and found that quadriceps weakness in the weakest leg and problems maintaining balance were factors.

Bickerstaffe et al.105 recommend tailor-made interventions that focus on safety in walking and reducing domestic hazards and also increasing muscle strength or stability where possible.

Any training could also bear in mind that Polio survivors have been coping with a high risk of falling for decades and will already be employing falls avoidance strategies.

4.10.6.1 Precautions

The existence of weak, fatigued and overused muscles, including the possibility of sub-clinical damage, must be borne in mind when prescribing any muscle training.
5 Gaps in the evidence - need for research
5 Gaps in the evidence - need for research

Many areas relating to the progression and management of PPS are not well researched and would benefit from further study. The paragraphs below give a few examples.

Longitudinal studies following the developmental course of PPS symptoms along with individual behaviours such as physical activity are needed to clarify the role of overuse as a risk factor and at what level and for which individuals exercise is beneficial.

The characteristics of fatigue in PPS, and especially the factors affecting general fatigue, are not well understood. It would be helpful to have a widely agreed definition of the different types of fatigue (neuromuscular, general, mental). Studies on patients who specifically have abnormal neuromuscular fatigue and overuse symptoms are needed, as is research into the exacerbating factors and efficacy of management interventions.

A major gap in the evidence on PPS pain are studies that clarify patients’ long-term experience of managing PPS and especially their experience of abnormal neuromuscular fatigue, showing the link between neuromuscular fatigue and overuse pain. Studies on the management of pain should divide patients into those with overuse pain worsened by activity and those with other pain.

Researchers have found that there are probably subgroups of patients who have cerebrospinal inflammation, and benefit from IVlg treatment. More in-depth research is needed into the specific symptoms or clusters of symptoms that patients with inflammation have and that are affected by IVlg, and whether, for instance, they are brought on by activity.

The specific area of Polio, Post Polio Syndrome and respiratory dysfunction is not well researched and much evidence is extrapolated from other neuromuscular groups or from studies with mixed populations. Therefore most areas would benefit from further research.

The British Thoracic society guideline for the ventilator management of acute hypercapnic respiratory failure in adults is due out 2015/16 and should provide more guidance on the management of this patient group in the event of an acute event.

Respiratory muscle training is not commonly used in clinical practice with this patient group. The evidence in the chronic obstructive pulmonary disease group has not shown significant benefits to objective measurements of lung volumes and muscle strength in combination to an improved perception of impact by the patient. There is not enough clinical evidence in this patient group and the evidence in other patient groups is not compelling enough to suggest it should be initiated as a treatment option.

Cold intolerance is not well understood; the area would benefit from studies of the prevalence, impact and management.
6 References
6 References


7 Appendices
7 Other relevant guidance and information

7.1 UK clinical information on PPS

7.1.1 Map of Medicine
The Map of Medicine is a central NHS repository of evidence-based pathways. Two entries relate to PPS:
- Map of Medicine: Post Polio Syndrome (PPS) - suspected
- Map of Medicine: Post Polio Syndrome (PPS) - rehabilitation and management

7.1.2 The Scottish Medical and Scientific Advisory Committee
This committee set up a working group and produced a broad report on PPS

7.1.3 Patient.co.uk and NHS Choices
Short summaries are available at -

7.2 European guidelines

7.2.1 European Federation of Neurological Societies (EFNS)
An EFNS task force reviewed the evidence for therapeutic interventions for PPS and produced clinical guidelines in 2006. An update was published in the European Handbook of Neurological Management in 2011

7.3 International clinical information

7.3.1 Cochrane Collaboration
The evidence for treatment of PPS was reviewed in 2011 and updated in 2015

7.3.2 Ireland

7.3.3 Australia
- MANAGING MUSCLES AND MOBILITY. Clinical Practice For Health Professionals. Polio Australia Incorporated. (2014)

7.3.4 USA
- Post Polio Health International has online resources for health professionals at http://www.post-Polio.org/edu/hpros/index.html

7.4 Clinical reference books
  - Written by international experts, this book covers the features, diagnosis and management of PPS with evidence-based reviews of therapies
7.5 Other relevant UK clinical guidelines

- NICE (2007) CG53: Chronic fatigue syndrome/myalgic encephalomyelitis (or encephalopathy)
- NICE. (2012). Clinical Guideline 146: Osteoporosis: assessing the risk of fragility fracture
- NICE. (2013). CG161: Falls - Assessment and prevention of falls in older people

7.6 Patient information

7.6.1 Lane Fox Respiratory Unit

The specialist clinic at St. Thomas’ Hospital London has produced a patient advice booklet

- Lane Fox Unit. (2015). Post Polio Syndrome - advice on the management of your symptoms. Guy’s and St Thomas’ NHS Foundation Trust

7.6.2 Relevant British Polio Fellowship documents

The British Polio Fellowship produces information and factsheets for Polio survivors in the UK

- Post Polio Syndrome: A practical guide to understanding and living with the condition, SS11, 10/09
- Pacing for Activity and Exercise, SS07, 11/07
- Pain and How to Manage it: A Guide for People with Polio and Post Polio Syndrome (PPS)
- Swallowing problems: A guide for people with Polio or Post Polio Syndrome (PPS), SS14 03/13
- PPS factsheets covering healthy eating, cold intolerance, dental care and a hospital pack

7.6.3 Books for Polio survivors

### 8 Glossary

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition: meaning within this document</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neuromuscular weakness</td>
<td>Inability to produce expected muscular force/power even on first trying from a rested state</td>
</tr>
<tr>
<td>Neuromuscular fatigue</td>
<td>Reduction in muscular force/power induced by exercise and relieved by rest, resulting in the need to work harder to produce the desired force/power</td>
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<tr>
<td>Exercise</td>
<td>A structured programme of physical activity with the aim of improving one or more aspects of physical performance - can include aerobic, flexibility and strengthening activities</td>
</tr>
<tr>
<td>Obstructive sleep apnoea (OSA)</td>
<td>Obstruction of the airway that only occurs during sleeping as the tone of the soft tissues reduces. Characterised by repetitive breath holds and snoring due to the effort needed to overcome the resistance of the soft tissues</td>
</tr>
<tr>
<td>Continuous positive airway pressure ventilation (CPAP)</td>
<td>Mechanical delivery of an airway pressure, usually via a face mask, throughout the breathing cycle. The continuous pressure maintains the airways open to allow more effective gas exchange and reduces the effort of the inspiratory breath</td>
</tr>
<tr>
<td>Negative pressure ventilation</td>
<td>Negative pressure is created outside the body in a vacuum which causes expansion of the thorax and lungs thus pulling air in. The breath out is achieved passively by the negative pressure being released. This type of ventilation is the original form of mechanical ventilation</td>
</tr>
<tr>
<td>Iron lung or tank respirator</td>
<td>A form of negative pressure ventilation that was large enough for the patient to lie in with just their head external to the device. This device prevented many patients dying from respiratory failure during the acute epidemics of Polio</td>
</tr>
<tr>
<td>Cuirass</td>
<td>An interface for delivery of negative pressure that is still in use. The shell covers just the patient’s thorax and abdomen</td>
</tr>
<tr>
<td>Positive pressure ventilation</td>
<td>Conversely to negative pressure ventilation, the air is pushed in by an external devise. There are a number of forms of positive pressure ventilation which address various issues with gas exchange or of matching the patients breathing pattern</td>
</tr>
<tr>
<td>Non-invasive ventilation (NIV)</td>
<td>The umbrella term for ventilation that provides respiratory support external to the body and without the need for a tracheostomy or endotracheal tube</td>
</tr>
<tr>
<td>Abdominal paradox</td>
<td>Under normal circumstances, the breath in and contraction of the diaphragm causes a flattening movement in a caudal direction. The abdominal contents are pushed down and the abdominal wall out. The Abdominal paradox is the drawing in of the abdomen on the breath in. This is a sign of the diaphragm being dysfunctional or weak. The breath in is created by the accessory muscles of the rib cage and neck contracting but the diaphragm moves in a cephalad direction causing the hollowing of the abdominal wall</td>
</tr>
<tr>
<td>Hypercapnia</td>
<td>An abnormally high level of carbon dioxide in the arterial blood, above the normal range of 4.6-6Kpa</td>
</tr>
<tr>
<td>Mechanical insufflation/exsufflation</td>
<td>A mechanical device that augments the effectiveness of a cough for secretion clearance. The insufflation is a positive pressure to deliver a larger lung inhalation. A sudden switch to negative pressure simulates the airflow changes that occur during a cough</td>
</tr>
</tbody>
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