

Post-polio syndrome – polio's legacy

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One of the first descriptions of late-onset muscle weakness following poliomyelitis was made in 1875 by Raymond, who presented a 19-year-old man to Charcot. The patient had residual paresis of his left arm and leg from infantile paralysis at the age of 6 months. He was performing heavy labour as a tanner and complained that his right arm had increasingly felt heavier and more rapidly fatigued during the last 2 years. During the previous three months, the same symptoms had developed in his right thigh. His right arm weakened, especially around the shoulder, there was moderate thenar wasting, and fibrillations were seen in many shoulder muscles. Sensation was unaffected. His right thigh also showed weakness and fasciculations to a moderate degree...Charcot suggested that this could be related to the patient's infantile paralysis.¹

It is estimated that there are currently about 120,000 people in the UK who contracted poliomyelitis and who were mostly forgotten about after the introduction of effective vaccination in the 1950s.² Some recovered fully or had mild residual muscular weakness while others had more significant persisting disability. Irrespective of the degree of recovery, up to 80% have or will develop new neurological symptoms known as post-polio syndrome (PPS), which may lead to significant new functional limitations.³

Deterioration years after acute polio, first described at Charcot's clinic in 1875, was not seriously researched until the 1980s and 1990s. The 1875 description quoted above includes the classic features of new weakness in the stronger limbs accompanied by muscle fatigue. There is now consensus that PPS symptoms usually develop after a stable period of 20 to 40 years following the initial illness, and include muscle weakness, fatigue (general, muscular or mental), muscle and joint pain, new muscle atrophy, sleep and respiratory problems, dysphagia and cold intolerance. As the cohort of polio survivors ages, there is a need for more understanding of this condition and how it can best be managed.

The cause of PPS is unknown with the leading hypothesis being that excessive metabolic stress on the remaining enlarged motor units, especially those initially damaged, results in the loss of nerve terminals and eventually of motor neurons themselves.^{4,5} Risk factors for developing PPS are greater severity of initial illness, greater degree of recovery or greater impairment, higher activity levels, older age at time of acute illness, female gender, longer interval since acute illness and neuromuscular

symptoms (cramps, fasciculations and muscle pain) during the stable period.^{6,7}

New weakness occurs both in muscles known to have been affected during the initial illness and in those thought to have been unaffected where damage was subclinical. Fatigue is the most common symptom and may include:

- a general feeling of exhaustion made worse by activity and improved by rest,
- muscle fatigue, felt often as heaviness and aching in the muscles,
- mental fatigue, experienced as difficulty processing information.

The fatigue is multifactorial and is thought to combine central and peripheral aspects.⁸ Muscle pain is usually described as an aching, burning or cramp related to physical activity. Joint pain is often due to weakness around joints, which may lead to bursitis, tendonitis and osteoarthritis.⁸ Several studies have looked at the relationship between the weakness, pain and fatigue and found that pain correlates with activity, but not weakness.⁹ A subsequent study found that pain was more common in women, and that joint and muscle pain might have different pathologies, as joint pain correlated with initial and recent increased weakness, whereas muscle pain correlated with fatigueability, activity, and younger age of acute polio.⁶

While symptoms of PPS are slowly progressive, they can usually be stabilised with appropriate management. PPS is best managed by a multidisciplinary team consisting of clinicians with a special interest, physiotherapists and occupational therapists, psychologists and orthotists.¹⁰ As PPS is a diagnosis of exclusion, an important part of the initial assessment is the identification of other neurological and musculoskeletal co-morbidities. These might include radiculomyelopathy, peripheral nerve compression syndromes (eg carpal tunnel syndrome), degenerative joint disease as well as general medical conditions associated with fatigue and musculoskeletal pain, eg hypothyroidism, polymyalgia rheumatica and other connective tissue diseases. Persistent sensory symptoms and upper motor neuron signs should alert the clinician to other pathology as polio primarily affects lower motor neurons.

After the treatment of co-morbidities and attention to musculoskeletal deformities and degeneration, the most important form of management for PPS symptoms is energy conservation (pacing physical activity), obesity management, assessment of orthotic requirements, orthopaedic intervention, if necessary, and other lifestyle changes. Non-fatiguing exercise is recommended, but the evidence is based on small highly selected studies. It is important for exercise to be monitored and for the patient to avoid overuse, which may

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lead to further deterioration. As yet no drugs have been found to be useful.⁴

Respiratory problems may arise from obstructive sleep apnoea (OSA) or from respiratory muscle weakness, often complicated by paralytic scoliosis. Symptoms of OSA include snoring, reported cessation of breathing by partners, daytime fatigue and hypersomnolence. OSA is more common in PPS than in the general public and is related to subclinical bulbar dysfunction. Symptoms of nocturnal hypoventilation and impending respiratory failure include morning headache, sleep disturbance and daytime sleepiness. This can be successfully managed with nocturnal non-invasive ventilation.⁴

PPS sleep disorders are common and usually multifactorial with pain and psychological aspects contributing to insomnia. Careful assessment and long-term monitoring are essential to ensure symptoms are treated optimally as they arise. Cold intolerance, which may be general or peripheral, rarely results in any long-term complications. Dysphagia is less common (10–20%), rarely severe, and can usually be managed with appropriate advice on swallowing techniques.⁶

The legacy of polio lives on in the cohort of polio survivors, producing new and progressive neuromuscular impairments. Expert assessment and management can minimise and stabilise symptoms. Many survivors experience suboptimal care as a result of lack of awareness of PPS. Physicians today, like Charcot 135 years ago, need to be able to spot the clinical picture and also ensure proper multidisciplinary management for this long-term neurological condition.

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