Rehabilitation for postpolio sequelae

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BACKGROUND
Postpolio sequelae (PPS) are new, late manifestations that occur many years after the initial poliomyelitis infection. Recurrence of symptoms and fear of reactivation of the polio virus is particularly distressing to polio survivors.

OBJECTIVE
This article outlines the diagnosis, pathophysiology, and management of PPS disabilities using a case vignette.

DISCUSSION
Clinical features of PPS include fatigue, joint and muscle pain, new muscular weakness and bulbar symptoms. Diagnosis can be complicated particularly in nonparalytic cases of poliomyelitis. Disabilities in PPS may not be obvious to the observer but significantly affect the quality of life of the PPS patient. Previous rehabilitation intervention focussed on physical effort and determination to overcome disability at all costs. The treatment in PPS is now modified, and aggressive physical measures that may exacerbate muscle weakness are avoided. Most disabilities in PPS can be well managed with rehabilitation interventions that address limitations in patient activities of daily living, mobility and cardiopulmonary fitness.

Postpoliomyelitis sequelae (PPS) refer to new, late manifestations that occur many years after the initial poliomyelitis infection. The time interval from acute polio to the onset of PPS is about 35 years, with a range of 8-70 years.[1] A number of patients are now presenting to their primary care physicians due to the polio epidemics of the 1940-1950s. The occurrence of PPS among patients with previous polio ranges from 28-64%.[2] The course of PPS is slow, and is reported to have an average progression based on strength measurements of 1-2% annually.[3,4]

There are an estimated 1.63 million survivors of poliomyelitis in the USA. Of these approximately 640 000 had paralytic poliomyelitis, and of these 75% now have PPS.[5] In Australia, the exact incidence of PPS is unknown. Despite large numbers of victims, only 6834 patients were on record between 1930–1940 (at the time Queensland was the only state that listed polio as a notifiable disease).[6] These poliomyelitis survivors had adapted to residual physical impairments and went on to rewarding fulfilling lives.[7] This development of new signs and symptoms of PPS is particularly distressing for these patients.

Australia is a polio free country; but is at risk of importation with immigration from polio endemic countries in southeast Asia and Africa. About 95% of all poliomyelitis infections are subclinical, 3% are abortive, 1% have nonparalytic, and 1% develop paralytic poliomyelitis. Patients may not present with specific muscle weakness or paralysis despite severe motor neuron damage (60% loss/damage to the motor neurons results in visible paralysis).[6]

What is PPS?

Poliomyelitis is considered to be a static disease. The PPS diagnosis requires new symptoms in poliomyelitis survivors after decades of stable disease. Diagnostic criteria are outlined in *Table 1* and common symptoms are detailed in *Table 2*. Exclusion of other conditions such as anaemia, thyroid disease, and cancer that can present with similar symptoms is important.

Although historically the diagnosis of PPS required a history of paralytic poliomyelitis,[9] recent studies show that similar symptoms were also reported in nonparalytic, subclinical and abortive polio cases.[10] Therefore, all polio survivors are potentially at risk of developing PPS. Particular risk factors include those:

- with paralytic poliomyelitis
- older at time of diagnosis of original polio infection
- who required hospitalisation[11]
- with greater physical activity in the intervening years.[12]

Postpolio sequelae symptoms affect the

ability to perform the activities of daily living, increase care needs and assistance, and impact on quality of life. Swedish studies have evaluated the physical functional capacity, social activities and wellbeing of PPS patients. Poor physical ability was seen in PPS patients 75–82 years of age as compared with similar aged persons without PPS. The women with PPS reported feelings of reduced emotional wellbeing compared with the general population sample.[13] Interestingly there was no difference found in 'family function' in aged matched controls and patients with PPS.[14]

Table 1. Criteria for diagnosis of PPS[18,44]

- Prior history of poliomyelitis (abortive, nonparalytic or paralytic), physical examination or laboratory studies
- Period of >15 years of neurological and functional stability after recovery from acute illness
- New symptoms: generalised fatigue, new weakness or abnormal muscle fatigue, muscle atrophy
- Exclusion of other medical diagnoses with similar manifestations (neuropathy, myopathy, spinal canal stenosis)

Pathophysiology

The poliomyelitis virus damages motor neurons in the anterior horn of the spinal cord and/or brain stem resulting in lower motor neuron weakness or paralysis. The surviving motor neurons undergo axonal sprouting and hypertrophy but are unstable, constantly remodelling and more vulnerable to premature failure, and unable to maintain muscle strength over a long period of time. With the aging of these motor units,[15] the overworked neurons fail to maintain axonal sprouting and are disrupted, resulting in fatigue or/and new muscle weakness as the metabolic demand on existing motor neurons increases.[16,17]

In some patients with PPS creatine kinase levels may be elevated suggesting muscle overuse as a contributing factor.[1] Other environmental toxins and immune mediation are being investigated as aetiological agents in PPS. The combined effect of aging, overwork, weight gain, other medical comorbidities, and muscular overuse or disuse, play a role in new weakness, pain and fatigue.[18,19] It is important to note there is no reactivation of the original poliomyelitis virus or reinfection. This is often a specific anxiety in PPS patients and needs to be addressed.

Management and treatment

For many patients the rehabilitation following poliomyelitis infection focussed on physical effort and determination to overcome disability at all costs.[20] These polio survivors achieved high levels of academic, vocational and social achievement far exceeding expectations. Further, the 'type A' personality type in poliomyelitis is well recognised. The typical compulsive behaviours were thought to be in response to their earlier experiences[21] and these patients have poor compliance with treatment and are 'sensitive' to control issues. Active patient participation and planning in rehabilitation is therefore crucial to the success of the program. Regardless of their recovery, these patients express anger and are depressed about their need to redevelop techniques to cope with new PPS symptoms.[22]

Aggressive physical measures are detrimental and exacerbate muscle weakness in PPS patients.[23] The rehabilitation goal is to teach the patient to live with PPS and have an improved quality of life. Patients are assisted to maintain physical health through:

- select exercise programs
- gait aids (walking sticks, crutches, wheelchairs)
- orthotics (ankle, foot orthosis)
- adaptive equipment (long handled aids and grabbers, rails in bathroom), and
- lifestyle changes (rest breaks, part time work).[24,25]

Table 2. Common symptoms of PPS

- Generalised fatigue
- Joint and muscle pain
- New muscle and/or joint weakness
- Muscle atrophy
- Cold intolerance
- Bulbar symptoms (swallow, speech, respiratory symptoms)

Addressing specific symptoms

Fatigue

Generalised fatigue is the most common manifestation in PPS.[26] The 1995 US National Post Polio Survey found fatigue the most disabling symptom in 91% of patients; 41% and 25% reported that fatigue interfered with their ability to perform work and self care activities respectively.[27] Fatigue is described as general tiredness, lack of energy 'polio wall' with minimal activity.[28]

Fatigue in PPS may be due to damage of the basal ganglia secondary to poliovirus. Mental fatigue can also occur in severe cases due to impaired brain function rather than diffuse disintegration of motor units.[29] Fatigue is best managed with patient education and lifestyle changes including paced everyday living activities,[30] lifestyle changes,[31] work simplification, and energy conservation strategies. This is accompanied by rest breaks and daytime naps. Patients are advised on weight loss programs. The use of lower limb orthotics (ankle foot orthosis) can decrease the energy consumption and aid fatigue. Reduction in mechanical stress by using wheelchair and bracing is effective.[32] Medications for generalised fatigue (amantadine, pyridostigmine) have not been found effective.[33,34]

New weakness

The muscular weakness in PPS is slow, progressive, and asymmetrical and can occur in previously affected or clinically unaffected acute poliomyelitis. It may be accompanied by

muscle atrophy. Patients often present with poor endurance and muscle fatigue. Previously affected muscles are more likely to be involved. Clinically unaffected muscles (during acute poliomyelitis) were shown to be involved on muscle electromyography studies.[1]

Abnormal muscle fatigue manifests as increased weakness following heavy overuse and settles with several days of rest.[35] Nonfatiguing exercise programs using both maximal and submaximal strength with short repetitions may be beneficial in PPS. The emphasis is to avoid overuse, exercise for short durations with rest periods, and exercise on alternate days to allow recovery and avoid overuse. Patients with PPS and overuse may need up to 2-3 days to recover from muscle fatigue.[26]

Muscle endurance and strength is more critical in a PPS person's ability to perform routine daily activities than maximal or peak aerobic activity. Therefore training effort should focus on activities that affect performance of work tasks and forestall fatigue.[36] Hydrotherapy in PPS improves strength, flexibility and cardiorespiratory fitness.[37]

Case history – Mrs Z

Mrs Z, 70 years of age, and suffering PPS, presents to an outpatient rehabilitation clinic 2 weeks following a left total hip arthroplasty (for osteoarthritis). She had recently been discharged home but reports pain and difficulty with mobility and inability to cope with one step outside her front door. She has ongoing generalised fatigue and pain in her polio affected (nonoperated) leg. She is suffering a depressed mood and is unable to sleep at night (previously declined antidepressants). Mrs Z has home help and her husband is quite supportive. She is otherwise well but concerned that she may not be doing enough to return to her premorbid functional status.

Clinically the wound is well healed and Mrs Z is on PRN simple analgesics. Her right polio affected leg is 2 cm shorter and muscle strength is about 3/5. She has weakness of her left hip abductors (2+/5). Mrs Z ambulates with a walking frame with a marked Trendelenburg gait pattern. She is independent with all self care, tires easily but insists on doing all home chores and has declined help previously. She has reluctantly agreed to attend an outpatient rehabilitation clinic for 2 weeks.

Rehabilitation treatment will include:

- education regarding PPS and recent surgery, monitor wound healing, ongoing simple analgesia on a time contingent basis (especially when attending therapy)
- reinforcing routine hip precautions (no flexion past 90°, no flexion with internal rotation and no adduction of leg past midline of body)
- fatigue management strategies (pacing activity, simplify tasks, regular rest breaks)
- reinforcing wellness and lifestyle changes (optimise weight and diet, quit smoking)
- specialised exercise program, encourage weight bearing on operated leg, prescribe lower limb orthotic for the right leg and correct footwear for the leg length discrepancy. Her program will improve posture and alignment, maintain joint range of motion, improve general endurance, muscle strength and flexibility (while avoiding muscle overuse and pain), and progressive gait training and step practice with appropriate aids (elbow crutches, stick). Commence supervised hydrotherapy. Suggest scooter or wheelchair for community mobility
- retraining in everyday personal activities and practise domestic chores with pacing and adaptive equipment (long handled aids, grabers)
- a home visit to ensure safety and environmental modification (ramps, grab rails)
- provision of assistance with heavier domestic chores through the local council, meals and shopping delivered at home (short term)
- link in with local support groups and post polio society.

Pain

Increasing musculoskeletal stress due to muscle weakness and joint instability (caused by progressive muscle atrophy) results in pain, especially in the lower back and lower limbs. A recent meta-analysis[38] showed that of a review of 539 patients (mean age after polio diagnosis: 37 years) 80% reported pain in muscles and joints and 87% only had fatigue.

Joint instability with or without weakness can occur. Pain can occur with accompanying leg length discrepancy, poor posture, scoliosis, abnormal mechanics and failing tendon transfers and joint fusions.[1] The mobility levels in PPS are aggravated by disuse atrophy caused by joint and muscle pains. Again, supervised exercise programs, pacing activities, decreasing mechanical stress with bracing and the use of adaptive equipment is recommended.

Women with PPS have been reported to suffer more pain than men[39] and long term pain has a more negative effect on women than men.[40] All methods used to manage pain should include active patient participation and discussion of living habits in relation to pain.[41]

Bulbar dysfunction

Postpolio sequelae can present with new weakness in bulbar muscle dysfunction, causing dysphagia, dysarthria, aphonia and facial weakness.[1,42] Patients can develop laryngospasm resulting in malnutrition, dehydration and aspiration pneumonia. Compensatory speech strategies (sitting upright when eating, small sized bolus,

altered food consistency) can be helpful.[42]

The respiratory management in PPS requires breathing exercises, chest percussion and management of secretions. Respiratory failure can occur in postpolio patients secondary to weak respiratory muscles, reduction in lung volumes and compliance. Positive pressure ventilation at night time is useful in PPS.[43] Some patients may need a tracheostomy and permanent ventilation. Sleep apnoea can occur and be central, obstructive or mixed and needs to be addressed as in non-PPS patients. Other measures including cessation of smoking, treatment of respiratory obstructive disease and vaccination (pneumonia, influenza) are important considerations.[26]

Conclusion

For many PPS patients, the recurrence of weakness, pain and fatigue is very distressing. Patients (and their families) should be encouraged to participate in planning their care. Rehabilitation has much to offer PPS patients to optimise function and improve their participation in and quality of life.

Conflict of interest: none declared.

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