Post-poliomyelitis syndrome: clinical features and management

Post-poliomyelitis syndrome affects mostly women who have had paralytic polio. Most patients do very well when they are given non-fatiguing exercises in a warm environment. Patients must be correctly diagnosed with this syndrome and offered appropriate management and rehabilitation.

The term post-poliomyelitis syndrome was introduced by Halstead and Rossi in 1985. The nature of the condition remains controversial and diagnosis is essentially clinical after excluding other neurological, orthopaedic or rheumatological problems.

Post-poliomyelitis syndrome affects polio survivors, 20 or more years after recovery from an initial paralytic attack of polio virus. Weakness with generalized fatigue is the most common symptom. Other symptoms like joint pain, muscle pain, atrophy, cold intolerance, respiratory insufficiency and dysphagia may also be present (Jubelt and Drucker, 1999).

Aetiology

The factors associated with the development of post-poliomyelitis syndrome are listed in *Table 1* (Howard, 2005). Various theories have been suggested as a cause of post-poliomyelitis syndrome:

- It may be the result of viral reactivation or ongoing viral replication (Melchers et al, 1992)
- Infection of the polio survivor's motor neurons by a different enterovirus than the one responsible for the patient's polio
- Elevated levels of cytokines in spinal fluid from patients with this condition suggest ongoing inflammation in the spinal cord (Gonzalez et al, 2002)
- The effect of ageing and muscle overuse or disuse may be a contributing factor but this does not explain the post-poliomyelitis syndrome in young patients (Chang and Huang, 2001)
- There is a decompensation of a chronic denervation and reinnervation process to an extent that healthy neurons cannot maintain the new sprouts; thus denervation exceeds reinnervation
- Weakness is the result of a dysfunction of the surviving motor neurons that causes a slow disintegration of the terminals of individual nerve axons. This is not caused by a loss of whole motor neurons, as in amyotrophic lateral sclerosis (Dalakas et al, 1986)
- The new wasting and weakness is caused by distal degeneration of motor units that have been affected by poliomyelitis (Jubelt and Agre, 2000).

The most widely accepted hypothesis, proposed by Wiechers and Hubbell (1981), attributes these symptoms to a distal degeneration of axons in the greatly enlarged motor units that developed during recovery following acute paralytic poliomyelitis. The next commonly accepted theory is that distal post-poliomyelitis motor unit degeneration can produce defects of neuromuscular transmission that may cause muscular fatiguability, and a gradual denervation of muscle fibres is the likely cause of the slowly progressive weakness found in post-poliomyelitis syndrome (Trojan et al, 1991). Despite some evidence of an immune-mediated mechanism for post-poliomyelitis syndrome, this disorder is generally not considered to be immune-mediated (Melchers et al, 1992).

Table 1. Factors associated with the development of post-poliomyelitis syndrome

Onset of functional deterioration after a prolonged period of stability	
Having had acute poliomyelitis at a young age	
Severe limb, bulbar, or respiratory involvement during acute polio	
Incomplete recovery with residual disability	
Greater physical activity during the intervening years	
Development of new symptoms or impairment associated with intercurrent events	
Development of symptoms including	Pain in joints, bones, and muscles
	Fatigue
	Cramps, fasciculation
	Wasting, weakness
Deterioration in functional abilities	Activities of daily living
	Mobility
	Upper limb function, respiratory function

Adapted from Howard (2005)

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Clinical features

Post-poliomyelitis syndrome presents as a new onset of weakness, fatigue, fasciculations, and pain with addi-

tional atrophy of the muscle group involved during the initial paralytic disease 20 or more years earlier. This syndrome is more common among women and with increasing time after the acute disease. Symptoms usually appear earlier in patients who have a lot of residual weakness, early bulbar respiratory difficulty during the acute illness, and those who were older when they contracted acute polio (Cohen, 2001).

Although this predominantly affects the limbs, approximately half of the cases also have mild choking or dysphagia and weakness of the respiratory muscles which may lead to nocturnal hypoxia and hypercapnic respiratory failure. The sluggish deterioration, lack of upper motor neurone signs and previous histology distinguish post-poliomyelitis syndrome from amyotrophic lateral sclerosis (Donaghy, 2000). Patients suffer from pain in the joints, bones and muscles. They also have fatigue with muscle wasting, weakness, cramps and fasciculation. There is a severe deterioration in functional abilities including mobility and activities of daily living (Dinsmore et al, 1995). Halstead's revised criteria (Halstead, 1991) are useful in the diagnosis of post-poliomyelitis syndrome (*Table 2*).

Investigations

Investigations are focussed on the exclusion of other diagnoses. Occasionally there may be a raised serum creatinine kinase level but other blood tests are usually normal. CSF analysis, computed tomography and magnetic resonance imaging scans are helpful to rule out stroke, multiple sclerosis or tumour. Electromyography is used to distinguish between other neurological or muscular pathology (Ravits et al, 1990). It cannot distinguish between stable polio and new muscle weakness. A thorough investigation to find other co-existing neurological, rheumatological or cardiovascular disorders should be done as they need specific treatment.

Management

Managing patients with post-poliomyelitis syndrome will need a multidisciplinary approach.

- Medications like steroids and pyridostigmine, most of which address fatigue, have been used with only partial success (Dinsmore et al, 1995; Horemans et al, 2003)
- Antivirals have no significant benefit compared to placebo medicines
- As post-poliomyelitis syndrome may result from the degeneration of nerve sprouts, growth factors can target these and help to regenerate new ones. Unfortunately, one small study (Rao et al, 1993) showed that insulin-like growth factor (IGF-1), which can enhance the ability of motor neurons to sprout new branches and maintain existing branches, was not helpful
- The patient may present with various joint deformities, which may require orthoses, casts or splints. If severe the patient should be considered for hip or knee replacement (Kidd et al, 1997)
- Appropriate exercise programmes should be introduced to reduce fatigue and maintain good muscle functioning (Willen et al, 2001)
- Strengthening exercises should be non-fatiguing like swimming or water aerobics. It has been reported that physical training in a warm environment has beneficial effects on muscles and joints, and study participants report less pain than at home (Strumse et al, 2003). It is not clear why this benefit occurs – whether the warm climate allows better warming up before the exercise, or whether this is partly a psychosocial effect
- Prolonged immobility and paralysis may be associated with loss of bone density and osteoporosis. Such patients should be screened for osteoporosis
- Electrical stimulation helps to strengthen the weakened muscle, decrease the pain and re-educate the muscles
- Heat, trigger point injections, bio-feedback, muscle relaxation exercises or static magnetic fields help with myofascial pain. Patients who undertake physical activity have significantly fewer symptoms than those who are physically inactive (Rekand et al, 2004)

Table 2. Criteria for the diagnosis of post-poliomyelitis syndrome

A prior episode of paralytic polio confirmed by history, physical examination and typical findings on electromyogram	
Standard electromyogram evaluation demonstrates changes consistent with prior anterior horn cell disease:	Increased amplitude and duration of motor unit action potentials, increased percentage of polyphasic potentials and, in weak muscles, a decrease in the number of motor units on maximum recruitment. Fibrillations and sharp waves may or may not be present
A period of neurological recovery followed by an extended interval of functional stability preceding the onset of new problems. The interval of neurological and functional stability usually lasts for 20 or more years	
The gradual or abrupt onset of new neurogenic (non-disuse) weakness in previously affected and/or unaffected muscles. This may or may not be accompanied by other new health problems such as excessive fatigue, muscle pain, joint pain, decreased endurance, decreased function and atrophy	
Exclusion of medical, orthopaedic, and neurological condition that might cause health problems listed above	

From Halstead (1991)

- Post-poliomyelitis syndrome patients will need adaptive techniques and equipment to perform daily activities with good education and encouragement
- Swallowing evaluation with dynamic imaging including cine- and video-fluoroscopy should be performed in post-poliomyelitis syndrome patients with dysphagia (Jones et al, 1992). Laryngeal pathology may be present in patients with dysphagia, and voice assessments should be considered (Driscoll et al, 1995)
- Sleep disorders could be related to the pain intensity and its frequency, therefore the sleep duration and quality will improve by managing the pain (Farbu et al, 2003)
- Respiratory muscle training can improve pulmonary function. Recognition of respiratory impairment and early introduction of non-invasive ventilatory aids prevent or delay further respiratory decline and the need for invasive respiratory aid (Farbu et al, 2006)
- Sleep evaluation can help with recognition of sleep apnoea and nocturnal hypoventilation. Some patients may need mechanical respiratory support because of respiratory insufficiency but most do very well.

Conclusions

After a period of prolonged stability, many patients with residual impairments following paralytic poliomyelitis develop new disabilities. Improved health and hygiene in western society has eradicated polio, but nearly one third of the people who suffered from paralytic polio has post-poliomyelitis syndrome. This syndrome should be identified earlier to reduce severe disability and appropriate treatment should be offered to all the eligible patients. Post-poliomyelitis syndrome has not been seen to necessarily progress after its onset. However, prognostic factors need to be evaluated by further studies. **BJHM**

KEY POINTS

- Post-poliomyelitis syndrome is a neurological disorder that occurs in individuals who have recovered from paralytic poliomyelitis.
- Major clinical features are new weakness, muscular fatiguability, general fatigue and pain.
- The diagnosis of post-poliomyelitis syndrome requires a history of paralytic poliomyelitis, partial or complete recovery of neurological function followed by a period of stability, persistent new muscle weakness or abnormal muscle fatiguability, and the exclusion of other causes of new symptoms.
- Contributing factors to post-poliomyelitis syndrome may be ageing with motor neuron loss, overuse and disuse.
- An interdisciplinary management programme can be useful in controlling symptoms, even though there is no specific treatment for post-poliomyelitis syndrome.

Conflict of interest: none.

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