

Review

Postpolio syndrome: Epidemiology, pathogenesis and management

Abdulraheem I. S.^{1*}, Saka M. J.¹ and Saka A. O.²

¹Department of Epidemiology and Community Health, College of Medicine, University of Ilorin, Kwara State, Nigeria.

²Department of Paediatrics, College of Medicine, University of Ilorin, Kwara State, Nigeria.

Accepted 5 December, 2011

Post polio syndrome is a disorder of the nervous system that appears in many survivors of paralytic polio, usually 15 years or more after the original illness. Currently, there are more than 2 million people world wide who have been afflicted with polio as children or young adults and who now may exhibit symptoms of the post-polio syndrome. The purpose of this review is to provide the current information on the syndrome's causes and promote information exchange on the epidemiology, diagnosis and management of PPS. Post-polio syndrome is characterized by increased weakness and/or abnormal muscle fatigability, general fatigue and pain, after a period of stability in individuals who had paralytic polio many years earlier. It is suspected to be the second degeneration of the enlarged motor units formed during recovery, and another denervation of muscle fibers. The cause of this denervation is unknown, but an inflammatory process could be responsible. Diagnosis is based on the presence of a lower motor neuron disorder that is supported by neurophysiological findings, in the absence of other disorders as causes of the new symptoms. Management of patients with postpolio syndrome should be multiprofessional and multidisciplinary in approach. Individuals with postpolio syndrome should be counseled on avoidance of inactivity and overuse of weak muscles. Proper evaluation of the patients for the need of orthoses and assistive devices is mandatory. This review paper also recommends creation of State centers with comprehensive expertise in the management of all aspects of PPS. The center should also offer teaching and research services.

Key words: Post-polio syndrome, epidemiology, pathogenesis, management.

INTRODUCTION

In the late 1970s, there were reports that people who had recovered from paralytic polio many years back, probably decades earlier were developing unexpected health problems such as excessive fatigue, pain in muscles and joints and, most alarming of all, new muscle weakness. As a result of dearth of literature about delayed neurological changes in polio survivors, the initial response by many physicians was that the problems were not real. For quite some time physicians were dealing with a cluster of symptoms that had no name, and without a name there was, in essence, no disease. Having a name, even if imprecise and misleading as to

causation, at least confers an element of credibility. Due to increase in numbers, persons experiencing the late effects of polio finally started attracting the attention of the medical community, and in the early 1980s the term post-polio syndrome was finally defined. The syndrome is defined as a neurological disorder that produces a cluster of symptoms in individuals who had recovered from paralytic polio many years earlier. These symptoms typically appear from 30 to 40 years after the acute illness. The major problems are progressive muscle weakness, debilitating fatigue, loss of function and pain, especially in muscles and joints. Less common are muscle atrophy, breathing problems, swallowing difficulties and cold intolerance. Of all these health problems, however, the critical symptom of post-polio syndrome is new progressive weakness.

*Corresponding author. E-mail: ibrorahem@yahoo.com.

PPS is suspected to be the second degeneration of the enlarged motor units formed during recovery, and another denervation of muscle fibers (Trojan and Cashman, 1996). A theory is that these muscles are actually dying from years of overuse, having labored for decades to perform everyday activities. Additional theories point to a premature aging of motor units and possible persistence of the poliovirus fragments (Maynard and Headley, 1999). It is estimated that between 25 and 50% of all patients who had acute poliomyelitis will eventually demonstrate some form of post-polio syndrome (Benditt et al., 1998). This disparity is most probably caused by the use of different clinical diagnostic criteria. In this context, it is important to remember that people who have sequelae of poliomyelitis but who do not fulfil diagnostic criteria for postpolio syndrome might still have substantial loss of motor function and be in need of therapeutic interventions.

The symptoms of PPS can be divided into two groups: 1) neuromuscular and 2) musculoskeletal. The neuromuscular symptoms are believed to be caused by a progressive deterioration of motor neurons; the musculoskeletal symptoms are more likely caused by years of abnormal wear and tear. The result is muscle pain and pain from osteoarthritic joints, inflamed tendons, strained ligaments and malformed joints (Benditt et al., 1998). Faced with new and additional limitations, many polio survivors have difficulty coping and experience psychosocial challenges. They may need help in attaining a positive attitude about their health and wellness. In *Polio Network News*, Linda L. Bieniek writes (Bieniek, 2001):

"To experience wellness, we need to balance and integrate the physical, emotional, mental, social, sexual and spiritual aspects of our lives".

Patients most at risk for post-polio syndrome are those who had suffered a severe attack of paralytic polio, although some patients who seemingly had a mild attack have also developed the syndrome. The onset of these new problems often is insidious, but in many cases they appear to be precipitated by specific events such as a minor accident, a fall, a period of bed rest or surgery. Patients characteristically say that a similar event several years earlier would not have caused such a large decline in health and function. In addition to clinical examination, one of the most valuable tools one can use to evaluate and treat patients is the medical history. It does not appear to be any history form in medical practice that asks the questions:

"Have you survived polio? If yes, are you feeling any fatigue or weakness in your muscles?"

We should ask this because though not clinically evident, post-polio syndrome patients do experience symptoms that can affect their ability to obtain and maintain optimal

quality health. While increasing the number of polio patients have been reporting in medical clinics throughout the world, few physicians and relevant allied professionals have adequate knowledge of the syndrome or of best clinical practice for diagnosis and treatment, hence the purpose of this study. In addition, there is also a need for examination of current clinical practices, with the goal of developing guidelines on how best to diagnose, treat and rehabilitate persons with post-polio syndrome. This review article explicitly provides current information on the pathophysiology, clinical characteristics and management of postpolio syndrome, and suggests areas of future research improvement.

Pathogenesis of poliomyelitis and post-polio syndrome

Knowledge of how the poliovirus infects the body can be helpful in understanding the possible causes of post-polio syndrome. It is a small RNA virus that can enter the body when contaminated water or food is ingested, and even when contaminated hands touch the mouth. The vast majority of persons who become infected either have no symptoms or experience a self-limited illness characterized by fever and gastrointestinal upset for several days. The poliovirus, which replicates in the lymphoid tissue of the throat and small intestine, either passes harmlessly from the gut or travels in the blood to all parts of the body. In a small minority of infected persons, usually 1 to 2%, the virus invades the central nervous system and produces an unpredictable amount of paralysis. A distinctive characteristic of acute polio infection is the predilection of the poliovirus for the nerve cells that control muscles. These nerve cells, or motor neurons, consist of a cell body located in the anterior horn of the spinal cord and a long tentacle, or axon, which extends to the muscles. Near the end of each axon, tiny sprouts branch out to individual muscle cells. At the nerve-muscle interface, or synapse, the sprouts from the axon release acetylcholine, a neurotransmitter that causes the muscle fibers to contract. A motor neuron and the group of muscle cells that it activates are called a motor unit.

With uncanny precision, the poliovirus invades the motor neurons, leaving intact adjacent nerve cells that control the functions of sensation, bowel, bladder and sex. How this exquisitely targeted behavior occurs was a mystery until recently, when researchers identified poliovirus receptors at the nerve-muscle interface. These receptors apparently allow the poliovirus to enter an axon and then to migrate to the nerve cell body in the anterior horn of the spinal cord. The poliovirus typically infects more than 95 percent of the motor neurons in the spinal cord and many other cells in the brain. The infected cells either overcome the virus or die. The extent of paralysis is unpredictable. Motor neurons that survive develop new

terminal axon sprouts in response to an unknown stimulus. These new sprouts re-innervate, or reconnect, with the muscle fibers left orphaned by the death of their original motor neurons. In a sense, the growth of additional axon sprouts is the body's effort to keep as many orphaned muscle cells as possible alive and working. A single motor neuron that initially stimulated 1,000 muscle cells might eventually innervate 5,000 to 10,000 cells, creating a giant motor unit. These vastly enlarged motor units make it possible for fewer motor neurons to do the work of many. Another adaptation that leads to increased strength is the enlargement of muscle cells when they are regularly exercised. These two compensatory adaptations, increase in muscle size and axon sprouting, are so effective that up to 50% of the original number of motor neurons can be lost without the muscle losing clinically normal strength.

These adaptations are neither static nor permanent. To the contrary, after recovery from acute polio there is an ongoing process of remodeling of the motor units that consists of both denervation (losing old sprouts) and reinnervation (gaining new ones). It is this process of remodeling or constant repair that allows the motor units to achieve a steady state of muscle strength. When this steady state is disrupted, new muscle weakness occurs. The motor unit area might increase by up to 20 times, reaching a level at which further reinnervation is no longer possible. Uncompensated denervation causes atrophy of muscle fibres and subsequently loss of muscle strength (Borg, 1994; Grimby et al., 1998). There is a growing consensus among researchers that post-polio syndrome involves a slow degeneration of the terminal axon sprouts that innervate the muscle cells. David O. Wiechers and Susan L. Hubbell, then affiliated with Ohio State University, proposed this explanation in the early 1980s after diagnostic tests indicated that the functioning of motor neurons in polio survivors progressively worsens as the number of years from their recovery increases. After examining the results of muscle biopsy and electromyographic (EMG) studies in their laboratory, Trojan and Cashman (1996) postulated that there are two types of disintegration of the motor neurons: a progressive lesion and a fluctuating one. The progressive lesion, in their view, occurs when the normal regeneration of the sprouts from the axon to the muscles is interrupted and malfunctioning sprouts are not replaced. This interruption of the repair process produces irreversible, progressive muscle weakness. The fluctuating lesion, on the other hand, is thought to be caused by defective synthesis or release of the neurotransmitter acetylcholine. Trojan and Cashman (1996) and others have demonstrated that muscle weakness and fatigue can be reversed in some patients with post-polio syndrome by the drug pyridostigmine, which enhances the effectiveness of acetylcholine in triggering muscle contractions.

Post-polio syndrome (PPS) is a variable combination of new progressive muscle weakness and other symptoms

in survivor of poliomyelitis, with onset usually at least 15 years after the acute illness. Acute paralytic poliomyelitis results from polio virus invasion of the brainstem and spinal cord motor neurons and is primarily a disease of motor unit (defined as a motor neuron and all the muscle that it innervates). More diffuse nervous system involvement with encephalitis can also occur in the acute attack. Motor neuron death causes the denervation of muscle fibers with resultant weakness. Recovery of muscular force after poliomyelitis occurs by recovery of some neuron and sprouting from the remaining motor axons and by muscle fibre hypertrophy in innervated muscle. Axonal sprouting can produce the reinnervation of locally denervated muscle fibers, restoring the ability to produce muscle fiber contraction. The resulting motor units can be up to eight times normal size. The surviving motor neurons innervates many more muscle fibres than normal, and may be unable to sustain this greatly metabolic demand indefinitely. Terminal axonal sprouts may degenerate, producing denervation of muscle fibers. Some of these denervated muscle fibers may become reinnervated by neighbouring axons, causing continuous "remodeling" process, but some may become permanently denervated, and thus produced permanent increased weakness. The distal motor unit degeneration can also produce neuromuscular junction defects, which may be a cause of muscular fatigue in PPS. Possible contributing factors to the development of PPS may be normal ageing process, and overuse or disuse of muscles. The ageing process produces a progressive motor neuron loss in polio survivors, which can contribute to the onset of PPS. The length of time between acute poliomyelitis and the start of symptoms is a risk factor for postpolio syndrome (Ramlow et al., 1992), and two studies independently concluded that age was a confounding factor for the development of postpolio syndrome (Ramlow et al., 1992; Lonnberg, 1993).

However, the normal ageing process can also cause muscle weakness in patients with postpolio syndrome (<http://www.marchofdimess.com/files/>). Continuous presence of poliovirus fragments in polio victim may also be a cause of postpolio syndrome. Mutated poliovirus genomic sequences have been detected in the CSF of some patients (Julien et al., 1999; Dalakas, 1995), but not in the CSF of others (Melchers et al., 1992). Another study (Baj et al., 2007) reported detection of poliovirus genome fragments in all patients with postpolio syndrome. More research is needed to accept the hypothesis of a persistent viral infection in postpolio syndrome. Genetic factor has been implicated in PPS pathophysiology especially studies have reported a genetic background in postpolio syndrome. Bartholdi et al. (2000) reported that the *SMN* gene deletion, a cause of adult spinal muscular atrophy, was not present in patients with postpolio syndrome. Polymorphisms in the Fc-gamma receptor IIIA was also reported as a possible risk factor for postpolio syndrome (Rekand et al., 2002).

More research on possible gene variants that could predispose to both acute poliomyelitis and postpolio syndrome is highly needed.

Important clinical correlates

Dysphagia

The incidence of dysphagia in polio survivors has been estimated to be approximately 18% (Coelho and Ferrante, 1988). Problems with swallowing could result in residues of food remaining in areas of the pharynx and can be a risk for aspiration. Poor nutrition from a diet of soft carbohydrates or lack of eating will adversely affect the hard and soft tissues of the oral cavity. Avoidance of eating can cause weight loss and additional nutritional problems. We should consider using only high-volume evacuation for these patients during treatment. Careful review of nutrition habits should be undertaken. Suggest to your patients to chew slowly, alternate liquid and solid swallows and swallow twice for each bite of food. Other suggestions are to turn the head to one side or tilt the chin downward when swallowing (Maynard and Headley, 1999). The clinical signs of post polio syndrome are shown in Table 2.

Cardiovascular disease

There are reasons to suspect that polio survivors who are experiencing post-polio problems might be at an increased risk for cardiovascular disease. Certain features such as generalized fatigue, generalized and specific muscle weakness, and joint and/or muscle pain may result in physical inactivity, obesity and dyslipidemia, a disorder of lipoprotein metabolism. In addition, most polio survivors are at an age when cardiovascular disease becomes increasingly more likely weaker even in the absence of postpolio syndrome (Bieniek, 2001).

Sleep apnea

Many polio survivors have abnormal breathing during sleep. Obstructive sleep apnea is probably more severe in polio survivors than in people without other medical problems. If obesity is also a condition, it may be a result of inactivity (Maynard and Headley, 1999). This should be addressed when considering treatment planning. Referral for sleep studies should be made for definitive diagnosis and treatment.

Clinical presentation

Common symptoms are (Khan, 2004):

- 1) Generalised fatigue,
- 2) Joint and muscle pain,
- 3) New muscle or joint weakness,
- 4) Muscle atrophy,
- 5) Cold intolerance,
- 6) Bulbar symptoms - speech, swallowing or respiratory symptoms, and
- 7) Worsening respiratory function - may present as headaches, fatigue or sleep disorder. Any of these can lead to a deterioration in day-to-day functioning. A small change in clinical terms can mean a large one in its effects on daily living. The criteria for post-polio syndrome is shown in Box 1.

Assessment

- 1) Listen to the patient's story;
- 2) The most important question to ask is not "can you do this activity?", e.g. climbing stairs, but "how do you do it?" This can reveal the functional change, e.g. stopping to rest half way, going upstairs by shifting on their bottom (Boone, 2008);
- 3) A patient questionnaire such as "my polio life" can be useful (My Polio Life, 2007);
- 4) Multidisciplinary assessment may be needed, e.g. involving physiotherapist, occupational therapist, neurologist, orthopaedic/orthotic team and respiratory physician. Bear in mind that various factors can make assessment more difficult (Boone, 2007);
- 5) Polio survivors are used to coping and adapting. Hence, the importance of asking - and observing - how patients carry out each activity;
- 6) Symptoms can vary from day to day and can be affected by recent activity, overuse or rest;
- 7) Patients may deliberately allow for a stressful hospital visit day by resting beforehand. This can give a falsely good picture. Ask what symptoms are like "on bad days";
- 8) Results can appear normal if patients are given long rests between tests, or if only the best result is recorded;
- 9) Respiratory and sleep problems are easily missed; and
- 10) Anecdotally, patients with PPS have had their symptoms dismissed after apparently normal or near-normal results of tests, such as lung function or muscle power.

Osteoarthritis as well should be assessed using Western Ontario and McMaster Universities Osteoarthritis Index physical functioning scale (WOMAC-PF) (Bellamy et al., 1988). WOMAC-PF is also a suitable instrument to measure physical functioning in patients with late-onset sequelae of poliomyelitis. The physical functioning sub-scale (WOMAC-PF) consists of 17 items concerning daily activities that primarily involve the lower extremities, such as walking, stair-climbing, and other transfers (sitting down, reclining). The WOMAC-PF is scored on a 5-point scale, ranging from 0 (no difficulty at all) to 4 (very much

difficulty). A sub-scale sum-score is calculated, ranging from 0 to 100, with higher scores indicating higher levels of functioning or well-being. In osteoarthritis populations, the WOMAC-PF has been found to have excellent clinimetric properties (Bellamy et al., 1988). Timed up and Go test (TUG) should also be used to screen/assess PPS individuals that are prone to falls. Falls are a significant public health concern for PPS individuals; early identification of people at high risk for falling facilitates the provision of rehabilitation treatment to reduce future fall risk.

The perceived health status of PPS patients should be measured using Nottingham Health Profile (NHP). The NHP is a self-administered questionnaire that measures self-perceived health status, it is divided into 6 categories: 1) physical mobility, 2) energy, 3) pain, 4) social isolation, 5) emotional reactions and 6) sleep. The physical mobility category (NHP-PM) consists of 9 questions, with a score ranging from 0 (no complaints) to 100 (answered yes to all questions). To compare the NHP with the SF36 and the WOMAC, we transformed the score to 0–100, with higher scores indicating higher levels of functioning or well-being (no complaints). The

Dutch version of the NHP-PM has been found to have satisfactory clinimetric properties in patients with chronic heart failure and myocardial infarction or stroke (Erdman et al., 1993; Visser et al., 1995).

Diagnostic criteria

Clinical investigation will depend on symptoms, but could include:

- 1) Muscle tests - but be aware that simple tests of isometric muscle strength may be insensitive (Hildegunn et al., 2007);
- 2) Respiratory investigations (see under 'Assessment of respiratory problems');
- 3) Sleep studies (see under 'Assessment of respiratory problems');
- 4) Swallowing studies, e.g. barium swallow;
- 5) Investigations to exclude other causes;

The diagnostic criteria include diverse signs and symptoms. The following are important diagnostic criteria in post-polio syndrome.

1. History of paralytic poliomyelitis with evidence of motor neuron loss;
2. A period of partial or complete functional recovery after acute paralytic poliomyelitis followed by an interval (usually 15 years or more) of stable neurological function;
3. Signs of residual weakness and muscle atrophy;
4. Signs of denervation by electromyography (EMG);
5. Gradual or sudden onset of progressive and persistent muscle weakness or abnormal muscle infatigability

(decreased endurance) with or without generalized fatigue, muscle atrophy, muscle or joint pain (Sudden onset may follow a period of inactivity or trauma or surgery);

6. Persistence of symptoms for at least one year;
7. Exclusion of other orthopaedic, neurological or medical problems as causes of symptoms;
8. Less commonly, symptoms attribute to PPS include dysphagia and dyspnoea;
9. Less specific symptoms like generalized fatigue, myalgia, aches and pains, exercise and cold intolerance.

These symptoms are subjective and therefore difficult to evaluate in patients with or without prior paralytic poliomyelitis. Because symptoms are subjective, psychological problems are possible. Evaluation of patients should include medical examination for hypothyroidism, polymyalgia rheumatica, or other systemic diseases. Patients with unequivocal exercise intolerance, mitochondria or metabolic myopathy should be considered. For those with muscle pain and fatigue, fibromyalgia is a differential diagnosis. In patients with prominent fatigue, sleep and breathing disorder may be considered, therefore spirometry and sleep examinations may be requested. Fatigue can also be a consequence from serious disability of muscle weakness and atrophy but it is also a manifestation of depression. PPS patients may be more susceptible than others to neurological problems originating from the spine when there is scoliosis or paravertebral muscle weakness. Among the neurological disorders that may be confused with PPS are adult muscular atrophy, amyotrophic lateral sclerosis, caudal equinal syndrome, cervical stenosis, chronic inflammatory demyelinating polyneuropathy, diabetic neuropathy, entrapment neuropathy, heavy metal toxicity, inflammatory myopathy, multifocal motor conduction block, multiple sclerosis, myasthenia gravis, Parkinson's disease, peripheral neuropathy, radiculopathy, spinal cord tumor and spinal stenosis. Neurologic consultation is therefore important. Diagnostic problem arise if there are unusual late symptoms such as dysphagia or respiratory insufficiency, but these symptoms appear in some patients deemed to have PPS, especially if breathing and swallowing were affected in acute earlier attack. If new weakness is focal, the differential diagnosis includes radiculopathy, and appropriate imaging is helpful. For difficult diagnostic problems, neurologist or other specialist with expertise in PPS management should be consulted. Useful medical examinations in PPS include EMG, CSF examination, brain and cord MRI, as well as magnetic stimulation of motor cortex and magnetic resonance spectroscopy for evidence of upper motor neuron dysfunction. According to Thorsteinsson (1997) the differential diagnoses of PPS are classified as follows:

Other causes of fatigue or weakness include:

Table 1. Symptoms and treatment of postpolio syndrome.

Symptoms		Treatment
Weakness	Further weakness in previously weak musculature or new weakness in previously clinically unaffected muscle groups	Physical activity, muscle training, avoiding muscle overuse and disuse, and lifestyle changes
Pain	Nociceptive pain Neurogenic pain	Symptomatic medication, physiotherapy, bracing, weight reduction Decompression and pharmacological treatment for neuropathic pain
Fatigue	Most often of physical character with decreased muscular endurance. Mental fatigue can also occur, that is, brain fatigue or decreased mental endurance	Physical activity, muscle training, rest, avoiding muscle overuse and disuse, lifestyle changes, weight reduction, and, if required, medication for restless legs syndrome, depression, and sleep disturbances
Hypoventilation	Caused by weakness of respiratory muscles and chest wall deformities, obstructive apnoea, and central apnoea	Assisted ventilation and inspiratory muscle training Invasive controlled mechanical ventilation
Dysphagia and dysphonia	Caused by weakness in oesophageal and laryngeal muscles	Instruction on swallowing and voice therapy

Source: Henrik et al. (2010).

- 1) Other types of neuropathy or myopathy;
- 2) Multiple sclerosis;
- 3) Amyotrophic lateral sclerosis or other anterior horn cell diseases;
- 4) Myasthenia gravis; and
- 5) Systemic conditions, e.g. anaemia, chronic infection, hypothyroidism, collagen disorders, medication side-effects.

Other causes of pain, for example:

- 1) Arthritis, bursitis, tendinitis,
- 2) Myalgias - polymyalgia rheumatica, fibromyalgia, and
- 3) Neuropathies.

Management

There is no specific management for PPS. An interdisciplinary approach to management, with a team of physicians and allied health personnel, is appropriate for many PPS patients because they often have a variety of problems. The team may include Primary health Care Physician, Neurologist, Psychiatrist, physical and occupational therapist. Consultants may include pulmonologist, speech pathologist, psychiatrist or psychologist, orthopaedist, rheumatologist, dietician, orthotist, nurse and respiratory therapist. Management should be symptom specific. Management should also address conditions such as osteoporosis or pressure palsies that may occur more commonly in people with previous

paralytic poliomyelitis than in the general population. The specific symptoms and treatment are shown in Table 1

Fatigue

Management of fatigue in PPS include energy conservation techniques, lifestyle changes, pacing of activities, regular rest periods during the day and improvement of sleep by relaxation techniques and medications. These include use of antihistamine, valerian, melatonin, amitriptyline, L-tryptophan and gabapentin. Non invasive ventilation at night may alleviate fatigue in patients with under-ventilation. Energy conservation techniques include discontinuing unnecessary consuming activities such as making a bed, sitting instead of standing. Lifestyle changes include changing to more sedentary job or working few hours and discontinuing selected activities. Regular naps or rest periods during the day especially in the early afternoon, can help in managing general fatigue and should be encouraged for patients especially with significant fatigue. Pacing (regular rest periods during activity is useful in managing focal (not general) muscle fatigue in PPS. Use of pedometer is important so as to know what the patient is doing most especially in terms of walking distance.

Pain

Pain in PPS is due to diverse causes, including joint and

Table 2. Clinical signs of post-polio syndrome.

Parameter	Clinical Signs	Investigations
Muscle function clinical examination Neurophysiology (electromyography) MRI	Weak or no muscle strength. Lower motor neuron lesion with fl accid paralysis	Clinical examination Neurophysiology (electromyography) MRI
Tendon reflexes	Weak or no tendon reflexes in affected muscle groups	Clinical examination
Sensory function Clinical examination	No sensory loss, cold intolerance might be present	Neurophysiology (nerve conduction velocity)
Cranial nerves Clinical examination	Most often normal but might be impaired (bulbar poliomyelitis)	Investigation on oesophageal and laryngeal muscle function
Pulmonary function and sleep-disordered breathing	Weak respiratory muscles, chest wall and spinal deformities Daytime sleepiness	Pulmonary investigation, including complete Spirometry Polysomnography

Source: Henrik et al. (2010)

- 1 Prior paralytic poliomyelitis with evidence of motor neuron loss, as confirmed by history of the acute paralytic illness, signs of residual weakness and atrophy of muscles on neurological examination, and signs of denervation on electromyography.
- 2 A period of partial or complete functional recovery after acute poliomyelitis, followed by an interval (usually 15 years or more) of stable neurological function.
- 3 Gradual or sudden onset of progressive and persistent new muscle weakness or abnormal 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 postpolio syndrome include new problems with breathing or swallowing.
- 4 Symptoms persist for at least a year.
- 5 Exclusion of other neurological, medical, and orthopaedic problems as causes of symptoms

Box 1. Criteria for postpolio syndrome 1. Source: March of Dimes. Post-polio syndrome: identifying best practice in diagnosis and care. http://www.marchofdimes.com/files/PPS_report.pdf (accessed November 27, 2011).

soft tissue abnormalities, pressure injuries of nerves in the arms and legs, including crutch injuries to the brachial plexus and carpal tunnel syndrome. Treatment depends upon the cause of pain. Management include life style changes, reduction of activity, pacing, stretching and use of ice or moist heat and assistive devices. Analgesics can be given but benzodiazepines and opiates should be avoided. Many causes of pain are treatable with

modification of extremity use, physical therapy, strengthening (when possible), orthoses to control joint deformities and difficulties, assistive devices, non-steroidal anti-inflammatory drugs, acetaminophen, and rarely steroid injection (because of the side effects) or surgery. These decisions warrant the attention of a specialist. Treatment for carpal tunnel syndrome include splinting, use of pads on canes or crutches to place the

wrist in more neutral position and increase the weight bearing surface of the hand, steroid and capal tunnel release. For patients with low back pain, lumbosacral orthoses, shoe lifts, back or pelvic support, non-steroidal anti-inflammatory drugs and physical therapy are options, before surgery is contemplated. Spinal stenosis can be treated with exercise, use of a cane or other walking device, trans-cutaneous electrical nerve stimulation (TENS), lumbosacra orthoses, epidural steroids, and in some cases surgery.

Respiratory problem

Respiratory muscle weakness is the main cause of respiratory insufficiency in post-polio patients. However, other contributing or causal factors include central hypoventilation due to previous damage from bulbar polio, scoliosis, kyphosis, sleep disordered breathing, obesity, other lung diseases, smoking and heart disease. Any contributing factors to respiratory problems that may be reversed should be eliminated or treated. All patients with respiratory dysfunction should receive pneumococcal vaccine and yearly influenza vaccinations. Ventilatory assistance may help those with hypoventilation or sleep-disordered breathing. Supplemental oxygen should usually be used only for acutely ill hypoxemic patients with intrinsic lung disease, after alveolar ventilation and airway secretions have been optimally managed with respiratory muscle acids. Glossopharyngeal breathing can be taught to ventilator dependent patients to allow periods of free ventilator breathing.

Dysphagia

The most common cause of dysphagia in PPS is weakness of pharyngeal or laryngeal muscles. Management include changing or restricting the diet to "safe" substances such as purees and thickened fluids (usually when the need is determined by video fluoroscopy, use of special swallowing techniques (such as turning the head to one side while swallowing, and avoiding eating while fatigued or taking larger meals earlier in the day and smaller meals later.

Psychosocial problems

Treatment of psychosocial problem may be accomplished with an interdisciplinary approach that include psychology, social worker and psychiatry as necessary.

Weakness

Management of weakness in PPS includes judicious

exercise, avoiding muscle over (exertion to the point muscle pain and fatigue), weight reduction, bracing weakened muscles and assistive devices such as crutches, manual wheelchairs, electric wheelchairs, and motorized scooters.

Exercise

To be safe and effective, exercise programs in PPS should be cautiously customized to each person's needs, residual strength of individuals muscles and symptoms patterns. For many individuals, the level of exercise is muscle stretching while for others it is inform of aerobic training. However additional exercise should be completely avoided in patients who are too weak and fatigued, and are already spending most of their energy simply performing activities of daily living. Patients should be monitored when starting the exercise, in order to ensure that exercise is done correctly, and that there are no adverse effects. Patients should be monitored, until they fully understood the exercise program properly.

Aquatic/training

Training in warm water reduces the stress on joints and muscles, and warm water may have an analgesic effect. Dynamic water exercises, not involving swimming, are a good alternative for patients with PPS, with positive effects on pain relief, improved cardiovascular conditioning, and increased subjective well-being (Willen et al., 2001). A study by Willen et al. (2001) has found that exercising in water, or aquatic therapy, helped post-polio syndrome patients relieve pain and assisted their muscle function. Although physical therapy and exercises help post-polio syndrome patients, health care providers must balance keeping such patients fit against the fact that over-exercise can exhaust weakened muscles and subsequently cause long periods of inactivity.

Orthotic management

Orthopaedic braces can restrict unwanted movements—supporting joints and muscles and reducing the impact of bodyweight, particularly in legs and feet. The use of braces should improve mobility, reduce pain, and reduce overuse of the still well functioning parts of the muscles, joints, tendons, and ligaments (Henrik et al., 2010). Use of lightweight knee–ankle–foot braces can have a beneficial effect (measured as decreased oxygen consumption during walking) and save energy (Brehm et al., 2007; Hachisuka et al., 2007). Patients with postpolio syndrome should be supplied with appropriate orthoses and braces on the basis of individual needs (Steinfeldt et al., 2003). In order to improve the energy efficiency of

these braces, carbon-composite material is preferred (Brehm et al., 2007; Hachisuka et al., 2007; Steinfeldt et al., 2003). In addition to orthoses, assistive devices like crutches, manual and electrical wheelchairs, and motorised scooters, should also be used in order to increase the functional activity (<http://www.marchofdimes.com/files/>).

Surgery

Indications for surgery include joint deformities, arthrosis, and limb-length inequality may require surgery. Increased function can be achieved by arthrodesis, tendon transfers, and muscle transplantation (Adnan, 2006). For other secondary disorders, such as spinal stenosis and disc hernia, surgical treatment can help. However, motor neurons of patients with postpolio syndrome might be more sensitive to effects of anaesthetic drugs and thus patients should be carefully assessed before and monitored both during an operation and postoperatively (Lambert et al., 2005).

Respiratory and sleep problems in post-polio syndrome

According to study reports (Howard and Davidson, 2003; Lincolnshire Post-Polio Information Newsletter, 2006; Bach and Tilton, 1997) the importance of respiratory problems in PPS are:

- 1) Respiratory problems in PPS are an important cause of symptoms and complications, including sleep disorders;
- 2) They may be under-diagnosed or inadequately assessed; and
- 3) Treatment can improve both quality of life and prognosis.

Aetiology

A study (Lincolnshire Post-Polio Information Newsletter, 2006) have shown various causes of respiratory problems in PPS. These include:

- 1) Respiratory muscle weakness;
- 2) Bulbar impairment - this may affect control of the upper airway or the respiratory cycle. If the upper airway is affected, there may be obstructive sleep apnoea;
- 3) Skeletal deformity - scoliosis or chest wall stiffness.
- 4) Other pathology, e.g. COPD, asthma, obesity;
- 5) Aspiration - if swallowing affected.

All these are likely to worsen during sleep. The pattern of respiratory impairment may be hypoventilation, obstructive sleep apnoea, or both (Hsu and Staats, 1998).

Symptoms

Respiratory failure can develop insidiously - symptoms may be subtle or unnoticed. Breathlessness may not be a symptom in patients with limited mobility. Possible symptoms are:

- 1) Sleep disruption, eventually leading to insomnia, daytime sleepiness or fatigue;
- 2) Morning headaches, irritability, poor concentration, anxiety or depression;
- 3) Abnormal sleep movements, nocturnal confusion, vivid dreams;
- 4) Breathlessness may be positional; and
- 5) Weak cough, chest infections.

Signs

May be subtle and possible signs are:

- 1) Unexplained tachypnoea;
- 2) Use of accessory muscles;
- 3) Abdominal paradox - This is inward movement of the abdomen on inspiration while the upper chest expands. It may best be seen with patient supine during a sniff manoeuvre. When upright, can be missed as the diaphragm passively descends at the beginning of inspiration; and
- 4) Severe, untreated nocturnal hypoxaemia can cause pulmonary hypertension, giving signs such as raised JVP and ankle oedema.

Assessment of respiratory problems

Listen to the patient's story and preferences, and assess:

- 1) Voice and cough;
- 2) Chest deformity;
- 3) Observe patients in realistic situations, e.g. doing repeated tests or actions, and doing everyday actions in which they may be using the necessary breathing muscles to achieve another task.

Investigations

- 1) Peak flow and cough peak flow,
- 2) Spirometry-Both seated and supine spirometry are needed. A sensitive indicator of respiratory muscle weakness is reduction in maximal inspiratory pressure
- 3) Oximetry (and possibly capnography).
- 4) Sleep study (polysomnogram)
- 5) ECG and chest X-ray if appropriate

Full sets of lung function tests and arterial blood gases may not be helpful in this scenario, unless intrinsic lung disease is suspected.

Management of respiratory problems in PPS

There are various options - choice will depend on the patient's individual situation and preferences. Night-time mechanical ventilation is often used. This helps by resting the respiratory muscles at night, and preventing deterioration of respiratory function during sleep. It also treats the secondary sleep disorder. Supportive measures include:

- 1) Not smoking;
- 2) Avoiding sedatives and alcohol;
- 3) Optimal weight and nutrition;
- 4) Pneumococcal and influenza vaccination;
- 5) Postural support if needed;
- 6) Prompt treatment of chest infections;
- 7) Techniques such as assisted cough or glossopharyngeal breathing ('frog breathing');
- 8) Chest expansion exercises.

Assisted breathing options are:

- 1) Non-invasive intermittent positive pressure ventilation (NIV or NPPV) is often useful (Box 1).
- 2) Rocking bed: This helps breathing by rocking a patient consecutively head up and head down. It is surprisingly effective, especially where muscle weakness is mainly diaphragmatic.
- 3) Pneumobelt: This gives intermittent abdominal pressure ventilation and is useful for daytime assistance
- 4) Negative pressure ventilation: Examples are tank ventilators (iron lung), jacket ventilators (Tunncliffe), and cuirass ventilators. The devices are cumbersome, and mainly used where NIV is not tolerated, or to provide 'respite' from NIV.
- 5) Tracheostomy ventilation.

Prognosis

In PPS functional deterioration does not necessarily progress. Fatigue and reduction in mobility may stabilise or progress very slowly.

Prevention

Given the various known contributing factors, it seems possible that PPS problems might be reduced by:

- 1) Careful management of exercise and daily living activities to optimise muscle and joint use, and prevent overuse or disuse.
- 2) Correct maintenance of aids and prostheses.
- 3) Monitoring and early treatment of associated/contributing problems such as:

- i) Osteoporosis.
- ii) Obesity.

iii) Respiratory problems.

Postpolio management center

Considering the number of polio cases since the inception of PEI activities in Nigeria, Postpolio management center should be established in Nigeria and headed by a neurologist or a specialist in rehabilitative medicine. All necessary specialists should be represented. The center should make provision for consultation in orthopedics, respiratory medicine, psychiatry and pain management. Staff at the center should include allied professionals who would provide expertise in physical and occupational therapy, orthotics, psychology, speech pathology, nursing, respiratory therapy, nutrition, social work, swallowing and assisted technologies (wheel chair and seating clinic). The center should have diagnostic facilities for EMG, pulmonary function testing, swallowing and sleeping studies, blood laboratory, radiological capabilities for magnetic resonance imaging, blood laboratory, computerized tomography and X-ray. The center should be responsible for training physicians and other health care workers in the management of PPS. The center should also be actively involved in research.

CONCLUSION

The impact of PPS on an individual and society is largely un-measured. This fact hinders the assessment of unmet needs of PPS patients. PPS-specific, responsive, reliable, and valid measures should be developed to assess the disease progression and effectiveness of interventions. When these measures are validated, they should be widely disseminated. Even though pharmacological and rehabilitation interventions seem effective in patients with postpolio syndrome, a combinations of different therapeutic strategies are important in achieving the best outcomes.

Detailed clinical analysis is a cornerstone in differentiating between postpolio syndrome and poliomyelitis-related secondary disorders. Researches are needed to establish, if there are clinical markers or risk factors that might predict the development of postpolio syndrome. When found, strategies and means for preventing postpolio syndrome should be started at an early stage. Study of subgroups of patients with postpolio syndrome (e.g, assessing rate of progression of muscle weakness and different forms of fatigue) and identification of measures to distinguish these subgroups early and thus tailor treatment accordingly are very important.

REFERENCES

- Adnan A (2006). Poliomyelitis: orthopaedic management. *Curr. Orthop.*, 20: 41-46.

- Bach JR, Tilton M (1997). Pulmonary dysfunction and its management in post-polio patients. *NeuroRehabilitation*, In Lincolnshire Post-Polio Library, 8: 139-153.
- Baj A, Monaco S, Zanusso G, Dall'ora E, Bertolasi L, Toniolo A (2007). Virology of the post-polio syndrome. *Future Virol.*, 2: 183-92.
- Bartholdi D, Gonzalez H, Borg K, Melki J (2000). Absence of SMN gene deletion in post-polio syndrome. *Neuromuscul. Disord.*, 10: 99.
- Bellamy N, Buchanan WW, Goldsmith CH, Campbell J, Stitt LW (1988). Validation study of WOMAC: a health status instrument for measuring clinically important patient relevant outcomes to antirheumatic drug therapy in patients with osteoarthritis of the hip or knee. *J Rheumatol.*, 15: 1833-1840.
- Benditt J, Cantini E, Chamblin D (1998). An approach to the patient with suspected post polio syndrome. Washington, DC: Easter Seal Society of Washington, pp. 1-3.
- Bieniek LL (2001). Emotional bridges to wellness. *International Polio Network Polio Network News*, 17(4).
- Boone H (2007). When test results do not match important symptoms. Conference presentation, Post Polio Association of South Florida, Miami., Lincolnshire Post-Polio Network, UK, p. 8.
- Boone H (2008). Problems experienced by polio survivors and suggested solutions. Presentation for occupational therapists, given at the Neurological Occupation Therapy Conference, October., Lincolnshire Post-Polio Network, UK.
- Borg K (1996). Post-polio muscle dysfunction 29th ENMC workshop 14–16 October 1994, Naarden, the Netherlands. *Neuromuscul. Disord.*, 6: 75-80.
- Brehm MA, Beelen A, Doorenbosch CA, Harlaar J, Nollet F (2007). Effect of carbon-composite knee-ankle-foot orthoses on walking efficiency and gait in former polio patients. *J. Rehabil. Med.*, 39: 651-57.
- Coelho CA, Ferrante R (1988). Dysphasia in post-polio sequelae: report of three cases. *Arch. Phys. Med. Rehabil.*, 69(8): 634-636.
- Dalakas MC (1995). Pathogenetic mechanisms of post-polio syndrome: morphological, electrophysiological, virological, and immunological correlations. *Ann. NY Acad. Sci.*, 753: 167-185.
- Erdman RA, Psschier J, Kooijman M, Stronks DL (1993). The Dutch version of the Nottingham Health Profile: investigations of psychometric aspects. *Psychol Rep.*, 72: 1027-1035.
- Grimby G, Stalberg E, Sandberg A, Sunnerhagen KS (1998). An 8-year longitudinal study of muscle strength, muscle fiber size, and dynamic electromyogram in individuals with late polio. *Muscle Nerve.*, 21: 1428-1437
- Hachisuka K, Makino K, Wada F, Saeki S, Yoshimoto N (2007). Oxygen consumption, oxygen cost and physiological cost index in polio survivors: A comparison of walking without orthosis, with an ordinary or a carbon-fiber reinforced plastic knee-ankle-foot orthosis. *J Rehabil Med.*, 39: 646-650.
- Henrik G, Tomas O, Kristian B (2010). Management of postpolio syndrome. *Lancet Neurol.*, 9: 634-642
- Hildegunn L, Jones K, Grenstad T, Dreyer V, Farbu E, Rekand T (2007). Perceived disability, fatigue, pain and measured isometric muscle strength in patients with post-polio symptoms. *Physiother. Res. Int.*, 12(1): 39-49.
- Howard RS, Davidson C (2003). Long term ventilation in neurogenic respiratory failure. *J. Neurol. Neurosurg. Psychiatr.*, 74(3): 24-30.
- Hsu AA, Staats BA (1998). "Postpolio" sequelae and sleep-related disordered breathing. *Mayo Clin. Proc.*, 73(3): 216-224
- Julien J, Leparac-Goff art I, Lina B, Fuchs F, Foray S, Janatova I (1999). Postpolio syndrome: poliovirus persistence is involved in the pathogenesis. *J. Neurol.*, 246: 472-476.
- Khan F (2004). Rehabilitation for postpolio sequelae. *Aust. Fam. Phys.*, 33(8): 621-624
- Lambert DA, Giannouli E, Schmidt BJ (2005). Postpolio syndrome and anesthesia. *Anesthesiology*, 103: 638-644.
- Lincolnshire Post-Polio Information Newsletter (2006). Issue covering respiratory problems, 5: 9
- Lonnberg F(1993). Late onset polio sequelae in Denmark: results of a nationwide survey of 3,607 polio survivors. *Scand J. Rehabil. Med. Suppl.*, 28: 1-32.
- March of Dimes (2011). Post-polio syndrome: identifying best practice in diagnosis and care. <http://www.marchofdimes.com/files/PPSreport.pdf> (accessed 20th November, 2011).
- March of Dimes (2011). Post-polio syndrome: identifying best practice in diagnosis and care. <http://www.marchofdimes.com/files/PPSreport.pdf> (accessed November 27, 2011).
- Melchers W, de Visser M, Jongen P, Van Loon A, Nibbeling R, Oostvogel.(1992). The postpolio syndrome: no evidence for poliovirus persistence. *Ann. Neurol.*, 32: 728-732.
- My Polio Life (2007). A patient questionnaire providing baseline information and comparison charts, for use by polio survivors and their health professionals. Lincolnshire Post-Polio Network.
- Ramlow J, Alexander M, LaPorte R, Kaufmann C, Kuller L (1992). Epidemiology of the post-polio syndrome. *Am. J. Epidemiol.*, 136: 769-786.
- Rekand T, Langeland N, Aarli JA, Vedeler CA (2002). Cgamma receptor IIIA polymorphism as a risk factor for acute poliomyelitis. *J. Infect. Dis.*, 186: 1840-1843.
- Steinfeldt F, Seifert W, Gunther KP (2003). [Modern carbon fiber orthoses in the management of polio patients—A critical evaluation of the functional aspects]. *Z Orthop Ihre Grenzgeb.*, 141: 357-361 (in German).
- Thorsteinsson G (1997). Management of postpolio syndrome. *Mayo Clin. Proc.*, 72(7): 627-638.
- Trojan D, Cashman N (1996). Current trends in post-poliomyelitis syndrome. New York: Milestone Medical Communications.
- Maynard FM, Headley JL (1999). Eds. Handbook on the late effects of poliomyelitis for physicians and survivors. St. Louis: Gazette International Networking Institute., pp. 412-414.
- Visser MC, Koudstaal PJ, Erdman RA, Deckers JW, Passchier J, van Gijn J, Grobbee DE (1995). Measuring quality of life in patients with myocardial infarction or stroke: A feasibility study of four questionnaires in the Netherlands. *J. Epidemiol. Community Health*, 49: 513-157.
- Willen C, Sunnerhagen KS, Grimby G (2001). Dynamic water exercise in individuals with late poliomyelitis. *Arch. Phys. Med. Rehabil.*, 82(1): 66-72.