Management of post-polio syndrome

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Abstract

Many patients with post-poliomyelitis syndrome can benefit from a management program. When a post-polio patient presents with new symptoms, it is first essential to identify and treat other medical and neurological conditions which could produce these symptoms. New weakness can be managed with exercise (stretching, strengthening, and aerobic), avoidance of muscular overuse, weight loss, orthoses, and assistive devices. Fatigue can be managed with energy conservation techniques, lifestyle changes, pacing, regular rest periods or naps during the day, amitriptyline to improve sleep, and possibly pyridostigmine (trial in progress). The management of pain is dependent upon its cause. The treatment of post-polio muscular pain can include activity reduction, pacing (rest periods during activity), moist heat, ice, and stretching, use of assistive devices, and life style modifications. Fibromylagia can be treated with amitriptyline, cyclobenzaprine, and aerobic exercise. Joint and soft tissue abnormalities can be managed with modification of extremity use, physiotherapy, orthoses, assistive devices, non-steroidal anti-inflammatory medications, and rarely steroid injections and surgery. Superimposed neurological disorders may produce pain, and should be identified and treated. The identification and treatment of pulmonary dysfunction in a post-polio patient is an important aspect of management, and is discussed elsewhere in this issue. Dysphagia can be managed with diet changes, use of special breathing and swallowing techniques, monitoring fatigue and taking larger meals earlier and smaller meals later, and avoiding eating when fatigued. The management of psychosocial difficulties usually requires an interdisciplinary approach, and may include a post-polio support group, social worker, psychologist, and psychiatrist.

Keywords: Poliomyelitis; Rehabilitation; Exercise; Treatment

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1. Introduction

Although currently there is no specific treatment for post-poliomyelitis syndrome (PPS), patients with this condition can be managed with a resultant improvement *[1]*. Many patients can benefit from at least some of the measures described below. Because of the great variety of symptoms reported by PPS patients, an interdisciplinary approach to management is most effective. The team can include physicians such as a primary care physician, physiatrist, neurologist, pulmonary specialist, psychiatrist, orthopedist, and rheumatologist. Other health personnel who are frequently involved in the care of these patients are a physical therapist, occupational therapist, psychologist, social worker, dietician, and orthotist. Table 1 summarizes the management approach in PPS.

Because there is currently no diagnostic test for PPS, this syndrome is diagnosed by excluding other medical and neurological disorders which can produce similar symptoms. In addition, the symptom of fatigue is extremely common even in the general population, and may be caused by a variety of disorders. There are also many potential causes for pain and weakness. The differential diagnosis is discussed in a separate article in this issue. It is very important that these other causes be identified and treated first before new symptoms in a post-polio patient can be attributed to PPS.

In a post-polio patient presenting with complaints of new weakness, fatigue, and/or pain, we recommend that a thorough medical and neurological examination, with screening blood tests be performed first. Laboratory tests can include CBC, SMAC (Na, K, Cl, HCO_3, glucose, BUN, Cr, calcium, phosphorus, magnesium, total protein, albumin, total bilirubin, ALT, AST, LDH, alkaline phosphatase, creatinine kinase), TSH, ESR, and serum protein electrophoresis. Some investigators *[2]* recommend that standard electromyography (EMG)/nerve conduction studies be performed in all four extremities on virtually all patients to confirm evidence for previous motor neuron loss and to exclude peripheral nerve dysfunction, and other neuropathic and myopathic conditions. We recommend performing EMG studies in those patients who have an unclear history of previous paralytic polio, or who lack the usual findings of previous polio on physical examination. Other studies such as nerve conduction studies, F-waves, H-reflexes, can be performed when clinically indicated to rule out other neurological disorders. This is especially true for those patients who present with complaints of numbness, and/or sensory deficits on neurological examination. Radiographic studies such as plain X-rays are frequently needed to evaluate joint dysfunction. In addition, CT scans and/or MRIs may also be needed to exclude conditions such as spinal stenosis, radiculopathy, and cervical myelopathy. In general, EMG is not helpful in excluding these conditions in patients with past paralytic polio because of the changes present as a result of previous polio. Pulmonary function tests (PFTs) should be performed in virtually all patients because unexpected deficits can be present even in those patients without a history of previous or recent respiratory dysfunction. Sleep studies may be necessary in those patients with symptoms suggestive of sleep apnea (snoring, frequent waking, daytime fatigue, morning headaches, impaired cognition, irritability, anxiety, cyanosis, and depression). Swallowing studies with videofluoroscopy may also be necessary.

Table 1 Management of post-polio syndrome (PPS)

1. *Identification and treatment of other medical and neurological disorders which can mimic the*

symptoms of PPS.

2. *Management of weakness*

Strengthening exercises (isometric, isotonic, isokinetic)
Aerobic exercise
Stretching exercises to decrease or prevent contractures
Avoidance of muscular overuse
Weight loss
Orthoses
Use of assistive devices

3. *Management of fatigue*

Energy conservation techniques
Lifestyle changes
Pacing
Regular rest periods, naps during day
Amitriptyline to improve sleep
Possibly pyridostigmine (trial in progress)

4. *Management of pain*

A. Post-polio muscular pain, muscular cramps, muscular pain with activity

Activity reduction
Pacing (rest periods during activity)
Moist heat, ice, and stretching
Use of assistive devices
Life style modifications

B. Fibromyalgia

Amitriptyline
Cyclobenzaprine
Aerobic exercise

C. Joint and soft tissue abnormalities

Modification of extremity use
Physiotherapy for use of physical modalities, strengthening, stretching.
Orthoses
Assistive devices
Non-steroidal anti-inflammatory medications
Steroid injections
Surgery

D. Treatment of other superimposed neurologic disorders (e.g. carpal tunnel syndrome, radiculopathy, spinal stenosis).

5. *Pulmonary disfunction*

Preventive measures (pneumococcal vaccine, yearly influenza vaccine)
Ventilatory assistance (non-invasive methods preferred)

Identification and treatment of sleep apnea
Glossopharyngeal breathing

6. *Dysphagia*

Changing or restricting diet
Use of special breathing techniques
Use of special swallowing techniques
Monitoring fatigue, and taking larger meals earlier, and smaller meals later
Avoiding eating when fatigued

7. *Psychosocial difficulties*

Interdisciplinary approach
Post-polio support group
Evaluation and treatment by social workers, psychologists, psychiatrists.

2. Management of weakness

The management of new weakness in a postpolio patient can include a strengthening exercise program, an aerobic exercise program, stretching to decrease or prevent contractures, avoidance of muscular overuse (exercising to the point of muscle pain and fatigue), weight loss, bracing, and use of assistive devices. For more detailed information on muscular function and exercise in postpolio patients, please refer to the article on this topic in this issue.

Exercise in post-polio patients has been a controversial topic for many years, primarily because of case reports of overwork weakness in these patients *[3,4]*. These clinical findings were supported by studies in denervated muscles of rats *[5-8]*. However, more recently, several studies have found that exercise can result in an increase in strength in post-polio patients many years after the acute illness. Feldman and Feldman and Soskolne *[9,10]* were the first to report in more recent years the beneficial effects of an exercise program many years after polio. Since then several other studies have confirmed their finding of the benefical effects of an isotonic strengthening exercise program *[11,12]* as well as isokinetic and isometric strengthening exercise programs *[12-14]*. In addition, fiber density, jitter, blocking, median macro-electromyography motor unit potential amplitude, and creatinine kinase were not changed significantly with the exercise program *[12]*. In addition to strengthening exercise, aerobic exercise has also been found to be beneficial in post-polio patients. Jones et al. *[15]* found that the average level of aerobic fitness in post-polio patients was 5.6 METS, or similar to that seen in patients just after a myocardial infarction. Aerobic exercise both in the upper and lower extremities on a bicycle ergometer *[15-17]*, and on a treadmill *[18]* has been shown to be helpful. The bicycle ergometer training programs resulted in similar training effects to those observed in normals with improved maximum oxygen consumption, duration of exercise, watts attained during exercise, and maximum expired volume per unit time. Treadmill training resulted in improved economy or energy cost of walking, and thus walking duration, but no change in cardiorespiratory conditioning.

Thus, exercise has been shown to be beneficial in post-polio patients, even in muscles where new weakness has been reported. The exact exercise prescription is dependent on a number of factors. Isometric exercise is most useful in muscles with less than 3 strength (MRC scale), or in muscles over a painful joint. An isotonic exercise program is most useful in muscles with grade 3 or better strength, and without a painful joint. Isokinetic exercise could also be used when equipment is available. An aerobic

exercise program such as using a bicycle ergometer, walking, or swimming can also be useful, but preferably should be an activity which the patient enjoys to increase compliance. A warm-up and cool-down period with stretching exercises should be included *[2]*. Some patients can also benefit from more agressive stretching of contractures (if no contraindications exist), such as knee flexion contractures and ankle plantarflexion contractures to improve gait and stability. Early in the exercise program, it is best for patients to be monitored carefully, to ensure that the exercise prescription is being followed correctly and that overuse of muscles and joints is not occurring. Once the patient understands the exercise program well, and can self-monitor, less frequent follow-up such as every few months is reasonable *[19]*.

Even though several different types of exercise have been shown to be beneficial in the late post-polio patient, exercise should be used judiciously, and should be avoided completely in some patients. Muscle groups which are being overused may benefit from rest or supportive devices such as braces. In addition, these studies have not assessed the effect of exercise programs on patient function and general well-being, and the long term effects of exercise in these patients are still unclear.

Overuse of muscles must be avoided in postpolio patients. Muscular overuse in patients with past paralytic polio was thought to be the cause of increased weakness in several case reports *[3]*. This new weakness may be permanent. Perry et al. *[20]* performed dynamic electromyography during gait in 34 PPS patients, and found evidence for overuse (compared to normals). The usefulness of creatinine kinase levels in the monitoring of overuse in post-polio patients is unclear, however Waring and McLaurin *[21]* found a significant correlation between CK levels and distance walked during ambulatory activities in the previous 24 h, indicating that CK could be a marker of overuse. In addition, in one case report *[22]*, CK was found to be markedly elevated in a post-polio patient with symptoms of weakness, fatigue, and pain. With a reduction in exercise, serum CK levels decreased and symptoms resolved.

Waring et al. *[23]* completed a retrospective study of lower extremity orthotic management for ambulation in 104 post-polio clinic patients, and found that 78% of patients noted that appropriate orthotic prescription subjectively improved ability to walk, increased perceived walking safety, and reduced pain. Clark et al. *[24]* have described some of the more common biomechanical deficits and their orthotic management in post-polio patients.

These include inadequate dorsiflexion in swing, dorsiflexion collapse in stance, genu recurvatum (knee hyperextension), genu valgum (valgus deformity at the knee), and mediolateral ankle instability. In some patients, strengthening exercises can be tried first to control these deformities. If this is impossible, impractical, or not useful, orthoses can be used. For example, inadequate dorsiflexlon occurs secondary to·weakness of ankle dorsiflexors, and results in a foot drop, and a tendency to fall or trip. This problem can be treated with an ankle-foot-orthosis (AFO). Genu recurvatum is a common deformity in post-polio patients and is usually caused by quadriceps weakness, with a resultant tendency of the patient to 'lock' the knee to improve stability of the lower extremity. This deformity can cause pain and decreased efficiency of ambulation secondary to an increase in movement at the knee during gait. Genu recurvatum can be managed with an AFO, knee orthosis, or in patients with more severe weakness (Medical Research Council 3 or less), a knee ankle foot orthosis (KAFO) may be necessary. For further information on orthotic management, please refer to the appropriate article in this issue. In addition to orthoses, other assistive devices which can be useful for post-polio patients to improve mobility and safety are canes, crutches, manual wheelchairs, electric wheelchairs, and motorized scooters.

3. Management of fatigue

Excessive fatigue in PPS patients can be managed with the use of energy conservation techniques, life-style changes, pacing, and taking regular rest periods or naps. Tricyclic antidepressants have been reported

to reduce fatigue, possibly by improving sleep, or by other more direct effects on central neurotransmission *[2]*. The anticholinesterase pyridostigmine may be useful, and a multicentered trial of this medication in PPS is currently in progress (see below).

Energy conservation techniques can include discontinuing some unnecessary energy-consuming activities (e.g. making a bed), using a handicap license plate, seating and workstation corrections, sitting instead of standing, moving the location of certain items and supplies to make them more easily accessible (e.g. washer and drier on first floor and not in basement), and using an electric scooter for longer distances. To reduce shoulder strain, driving wheel positions can be changed and mobile arm supports can be attached to wheelchairs, desks, or tables *[25]*. Life-style changes may include discontinuing certain activities such as volunteer work, changing to a more sedentary employment, or working part-time. Regular naps during the day, especially in the early afternoon, can be beneficial in the management of general fatigue, and patients (even working patients) should be encouraged and assisted to do this.

Agre and Rodriguez *[26]* found that pacing was helpful for muscular fatigue. Seven symptomatic post-polio patients were studied on three separate occasions at least 1 week apart with three different exercise protocols. Patients, when allowed to take regular rest periods during an isometric endurance test of the quadriceps at 40% maximum voluntary contraction (MVC), were found to have less local muscle fatigue, increased work capacity, and improved recovery of strength after activity. Therefore, regular rest periods during activity can result in improved work capacity and should be encouraged.

A dilemma exists in the management of fatigue because neither medical personnel nor patients have means other than symptomatology with which to measure their activity or exercise level. Because patients with past poliomyelitis can perceive exertion at the peripheral muscular level during activity in a fashion similar to non-polio control groups *[27]*, they should be taught to attend to their perception of exertion and to monitor fatigue *[28]*. The Borg Rating of Perceived Exertion (RPE) *[29]* is a useful measure which can be used to judge effort (Fig. 1). This 15 point scale with verbal anchors progressing from 6 at the lowest level, or light; to 20 at the highest, or very, very, hard level was originally developed for cardiac patients. It is an indicator of physical strain integrating peripheral muscle, joint, and cardiorespiratory signals that correlates well with heart rate (r=0.90), blood pressure *[30]*, and blood lactate levels (r=0.83) *[31]*. The RPE is routinely used as a limit of exercise capacity in other conditions *[32]*, and has been used more recently in PPS *[18,33]*. PPS individuals, after one training session, reliably (r=0.83) used the RPE in an exercise test to monitor their effort and finish the test *[33]*. This ability to use and understand the safe levels of exertion improves with intensity of activities and practise. Because the level of exertion can be reliably identified with the RPE in a PPS population, it can be used as the point at which to end any daily activity or exercise regime. Through the use of a daily/weekly diary with the RPE to identify levels of fatigue, patients can select their own appropriate level and pace their everyday activities. An RPE level 14, with the anchor word 'hard,' is the top level of exertion recommended. RPE levels with anchor words are more reproducible *[34]* and level 14 has been used in other exercise limiting cases *[18,33]*. Clear and early identification of fatigue, either at a local muscular level from exercise, or at a global body fatigue level from daily activities, by using RPE level 14 allows PPS patients to develop training strategies to prevent or reduce injury and overuse syndromes.

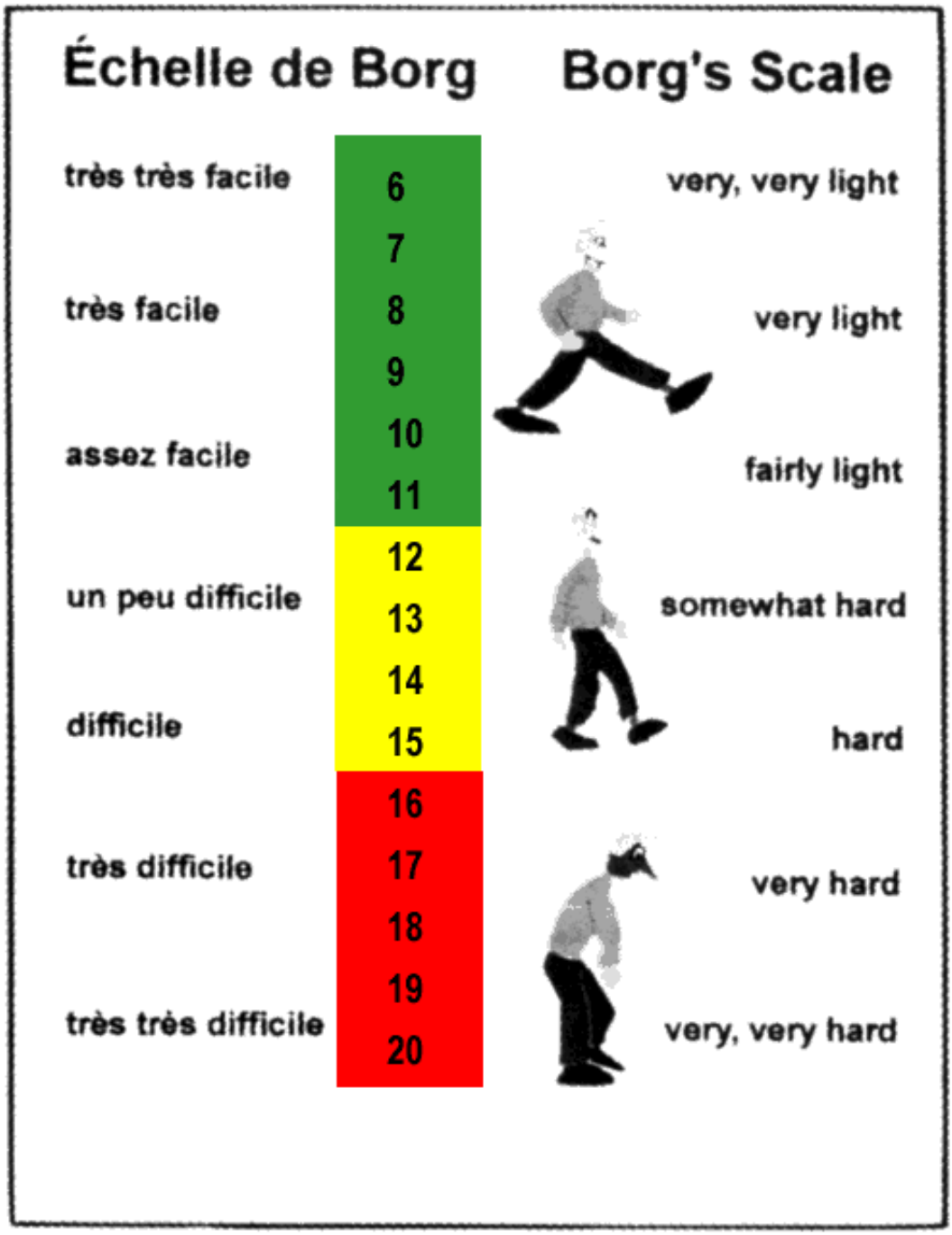

Fig. 1. The Borg scale of perceived exertion. The original Borg scale of perceived exertion has been modified. The numbers from 6 to 10 are in green, the numbers from 11 to 15 are in yellow, and the numbers from 16 to 20 are in red, similar in concept to traffic lights. A figure has been added for each colored area to depict the amount of effort represented by the numbers and anchor words. Reprinted with permission from the Scandinavian Journal of Rehabilitation Medicine *[29]. The range discrepancy between the colour scales (green and yellow) as marked in the figure and as detailed in this description, existed in the published article. Lincolnshire Post-Polio Library Administration.*

4. Management of pain

Pain in post-polio patients can occur from a variety of causes. Management of pain is thus dependent upon the etiology. Pain can be caused by muscular abnormalities, joint and soft tissue abnormalities, and other superimposed neurological abnormalities. Overuse is the most likely etiology of many of the pain

syndromes. The location of pain appears to be dependent upon the method of locomotion *[35]*. In their study, Smith and McDermott *[34]* showed that pain in the lower extremities and lower back occurred more frequently in ambulatory patients, whereas pain in the upper extremities tended to occur more frequently in those patients who used wheelchairs or crutches.

Common muscular causes for pain include a 'post-polio muscular pain,' *[2]*, muscular overuse, muscular cramps, fasciculations, and fibromylagia *[36]*. 'Post-polio muscular pain' typically occurs in muscles previously affected by polio, and is usually described as an aching sensation similar to that experienced at the time of paralytic polio. It usually occurs at the end of the day, and is aggravated by activity *[2]*. Patients can also experience a muscular pain with activity. Painful muscular cramps can occur either with activity or at the end of the day. Muscular cramps and muscular pain with activity are probably secondary to overuse, and should be avoided. Management of these types of pain include reduction of activity, taking rest periods during activity (pacing), use of moist heat and stretching, use of assistive devices, and lifestyle modifications. Fasciculations are a sign of previous motor neuron damage, and are reported by patients with previous paralytic polio. Fibromyalgia occurs commonly in patients presenting to a post-polio clinic and can be treated with amitriptyline, cyclobenzaprine, aerobic exercise, and other measures *[36]*.

Joint and soft tissue abnormalities include osteoarthritis, tendinitis, bursitis, ligamentous strain, joint deformities, and failing joint fusions. Because of residual weakness from paralytic polio, post-polio individuals may chronically use certain joints and extremities in abnormal ways which may predispose them to injury of joints, tendons, bursae, and ligaments. The resultant mechanical stresses and abnormal postures may perpetuate the pain problem. In a cross-sectional study of 61 post-polio patients, the prevalence of radiographically determined moderate to severe osteoarthritis of the hand and wrist was 13%, but the prevalence of mild osteoarthritis in the same joints was 68%. Associated factors for mild osteoarthritis were age greater than 50, lower extremity weakness, high current locomotor disability, and high current usage of assistive devices *[37]*. In addition, abnormal forces around joints may also produce joint deformities such as genu recurvatum, and genu valgum. Many of these causes for pain are treatable. Treatments can include modification of extremity use, physiotherapy for use of physical modalities [e.g. ice, superficial heat, ultrasound, transcutaneous electrical nerve stimulation (TENS)], strengthening, orthoses to control joint deformities and failing joint fusions, assistive devices, non-steroidal anti-inflammatory medications, acetominophen, and rarely steroid injections or surgery. Stretching of tight tendons and soft tissues may be helpful. However, some contractures may be biomechanically beneficial for the patient, and these should not be stretched *[38,39]*.

Superimposed neurological disorders which can cause pain include peripheral neuropathies, radiculopathies, and spinal stenosis. Electromyographic findings in 100 consecutive post-polio patients revealed a high prevalence of carpal tunnel syndrome (35%). Other neuropathies which were found included ulnar neuropathy at the wrist (2%), peripheral neuropathy (3%), carpal tunnel syndrome and ulnar neuropathy (3%), brachial plexopathy (1%), radiculopathy (4%), and tibial neuropathy (1%) *[40]*. Werner et al. *[41]* found that the use of assistive devices is a major risk factor for carpal tunnel syndrome in post-polio patients. Treatments for carpal tunnel syndrome can include splinting *[42]*, use of pads on canes or crutches, or use of a special grip for canes and crutches which places the wrist in a more neutral position, and increases the weight-bearing surface of the hand *[2]*. For patients with lumbo-sacral radiculopathies or low-back pain, use of lumbosacral corsets, shoe lift, back supports, or pelvic supports can be helpful. Spinal stenosis can be treated with exercise, use of a cane, TENS, and a lumbosacral orthosis. In some cases, surgery may be necessary.

Pulmonary dysfunction

Symptoms of respiratory dysfunction are common in patients with past paralytic polio *[43]*. Compromised lung function can occur in postpolio individuals irrespective of whether or not shortness of breath is present *[44]*. Respiratory compromise occurs primarily in those individuals who required ventilation at the time of acute polio, but may also occur in those who do not report previous respiratory involvement. Risk factors for respiratory compromise greater than 35 years after acute polio are the need for ventilation at acute polio and age at acute polio of greater than 10 years *[44]*. Manifest hypoventilation is rare in post-polio patients, and was reported to be present only in 2 of 40 post-polio patients who had respiratory and non-respiratory poliomyelitis at least 30 years previously *[45]*. However, many of the 87.5% of those who were initially weaned from a respirator may again require the use of a respirator *[46]*. 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 poliomyelitis *[47]*, scoliosis and kyphosis, sleep disordered breathing, obesity, other pulmonary diseases, smoking, and cardiac disease.

Sleep apnea is a common problem in post-polio patients, and may be central, obstructive, or both *[48]*. This condition is important to diagnose since if untreated, it can eventually result in acute cardiopulmonary failure *[49]*. Please refer to the article on pulmonary dysfunction in this issue for further details on this topic, sleep apnea, and the management of both conditions.

6. Dysphagia

Symptoms of swallowing dysfunction are less common in post-polio patients than the other difficulties mentioned above, however they can occur in 10-20% of selected samples of post-polio patients *[50]*. Dysphagia can occur in patients with a previous history of bulbar poliomyelitis, but can also occur in post-polio patients without previous bulbar involvement. In addition, mild to moderate abnormalities may be found on videofluoroscopy regardless of whether or not the patient is symptomatic *[51,52]*. Symptoms can include food sticking, coughing or choking during eating, and slowing of swallowing and eating *[50]*. Post-polio dysphagia is most commonly due to weakness of pharyngeal or laryngeal muscles. In patients who are complaining of swallowing or feeding dysfunction, a videofluoroscopic swallowing evaluation should be performed, preferably with a swallowing therapist. Videofluoroscopic abnormalities can include unilateral bolus transport, pooling in the valleculae or pyriform sinuses, delayed pharyngeal constriction, impaired tongue movements, and rarely aspiration *[51]*. However, it must not be assumed that all dysphagia in post-polio patients is secondary to polio, but other abnormalities such as structural lesions should also be ruled out. In one study *[53]*, 20 patients with past polio and dysphagia were evaluated with cinefluoroscopy. Other abnormalities not necessarily secondary to polio were found and included a short stricture (one patient), a Zenker diverticulum (one patient), bilateral pharyngeal pouches (five patients), and unilateral pouch (one patient). In addition, laryngeal dysfunction may contribute to dysphagia. In one study *[54]*, the laryngeal function of 9 of 21 post-polio patients with swallowing complaints was studied, and all nine patients had some degree of abnormality on laryngeal videostroboscopy. Symptoms of dysphagia and videofluoroscopic abnormalities appear to progress with time *[51,55]*. Management of dysphagia in a post-polio patient can include changing or restricting the diet to certain 'safe' substances such as purees or thickened liquids (preferably as determined by videofluoroscopy), use of special breathing techniques, use of special swallowing techniques such as turning the head to one side, monitoring fatigue and taking larger meals earlier and smaller meals later, and avoiding eating when fatigued *[2,50]*.

7. Psychosocial difficulties

Post-polio patients who are now faced with PPS may have great difficulty adjusting to this second and

unexpected disability. Many of these patients have had to come to terms with the residua of a severe childhood illness, and now they are being asked to once again deal with polio-related difficulties. Because polio is now considered to be a 'conquered disease,' patient problems are compounded by the general lack of knowledge among the medical profession about both acute polio and PPS *[56]*.

Some investigators have described a 'polio personality,' and have suggested that this may actually contribute to post-polio sequelae *[57,58]*. Polio survivors tend to be well-educated, competent, hard-driving individuals who demand perfection both from themselves and others around them *[2,59]*. In a survey conducted by Bruno and Frick *[57]*, 676 polio survivors were recruited by sending out 1200 questionnaires to post-polio clinics and support groups in the US. The mean type A score was found to be significantly higher in this post-polio group than in a previously reported non-disabled control group. In addition, type A score was significantly higher in those with muscle pain and fatigue. Despite their difficulties, polio survivors have more years of formal education and a larger proportion are married than the general disabled and non-disabled populations. In addition, they have higher levels of employment than the general disabled population *[59]*.

Frick *[56]* suggested that individuals with PPS may experience personal devaluation, isolation, and depression as psychological responses. In one study *[60]*, the Symptom Check List-90 (SCL90R), Psychosocial Adjustment to Illness ScaleSelf Report (PAIS-SR), and questionnaire about polio histories was administered to 93 individuals with a past history of polio (71 from a post-polio clinic and 22 recruited from a post-polio support group). Results indicated psychologic distress. Elevated SCL-90R scores in men occurred in somatization, depression, anxiety, hostility, and phobia. In women, elevated scores occurred in somatization, depression, anxiety, and psychoticism. In another study *[61]*, in which 116 polio survivors were recruited from a polio registry, the prevalence of depression and distress was found to be 15.8%. Psychological distress/depression was correlated with physical symptoms. The emotional responses to a new disability may pose difficulties for the treatment of these patients. Patients may resist making the necessary changes in their life to effectively manage their new difficulties.

Treatment of psychosocial difficulties related to PPS is best managed with an interdisciplinary approach. It may include obtaining help from a post-polio support group, and evaluation and treatment by health professionals such as psychologists, social workers, and psychiatrists.

8. Drug trials and pharmacotherapy

Several small clinical trials of pharmacological treatments have been completed in PPS patients. Pyridostigmine (Mestinon) 180 mg per day has been evaluated in an open trial using an objective measure of neuromuscular junction transmission (jitter as measured by stimulation single fiber electromyography) in 17 patients by Trojan et al. *[62]*. A significant relationship between subjective fatigue response to pyridostigmine and improvement in neuromuscular junction transmission with edrophonium (a short-acting anti-cholinesterase similar to pyridostigmine) was found. This suggests that fatigue in PPS is due to anticholinesterase-responsive neuromuscular junction transmission deficits in at least a proportion of patients (Fig. 2). In addition, Seizert et al. *[63]* have found improvements in some objective measures of strength and subjective measures of fatigue with pyridostigmine 180 mg per day in a double-blinded, placebo-controlled, crossover trial in 27 PPS patients. Because of these encouraging preliminary results, a multicentered trial of this medication is currently underway.

Amantadine and high dose prednisone have also been assessed in PPS. Stein et al *[64]* randomly assigned 25 PPS patients with fatigue to amantadine 100 mg twice a day or to placebo for 6 weeks. Fatigue level was assessed with the Fatigue Severity Scale, and Visual Analog Scale for Fatigue. No association was found between amantadine and clinical response to fatigue, although 54% of amantadine patients, and

43% of placebo patients reported improvement in fatigue. Dinsmore et al *[65]* studied the effect of high dose prednisone in 17 PPS patients (with new muscle weakness). Patients were randomly assigned either to prednisone (80 mg per day for 28 days, followed by a gradual reduction) or placebo. Outcome assessments included measures of isometric strength by a modified Tufts Quantitative Neuromuscular Examination (TQNE) *[66]*, manual muscle testing, and subjective fatigue assessment. There were no significant differences between groups in terms of muscular strength and subjective fatigue. Another medication which may warrant further study is deprenyl (selegiline), a neuroprotective agent. In a report of two cases *[67]*, deprenyl was found to produce an improvement in PPS symptoms, and this improvement ceased with discontinuation of the drug. Thus, several potential pharmacological treatments have been evaluated in PPS, and of these pyridostigmine appears to be most promising.

Other potential treatments which could be evaluated in PPS in the future include other neuroprotective agents such as vitamin E, and neurotrophic factors such as insulin-like growth factor (IGF-1), and brain-derived neurotrophic factor (BDNF).

FIGURE 2 Graph

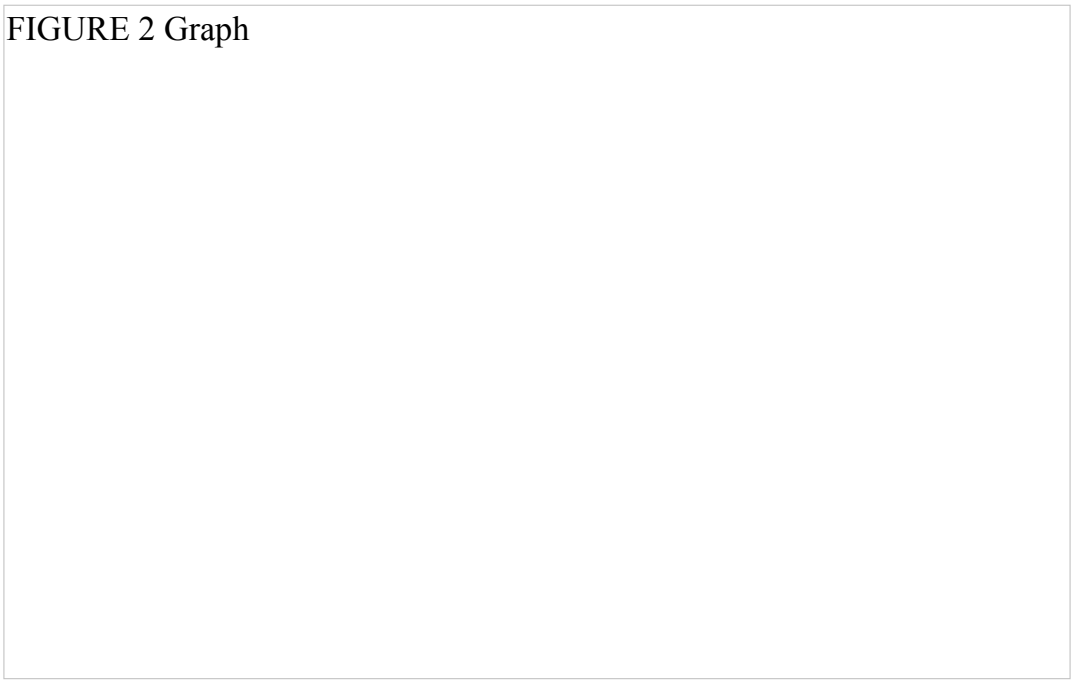

Fig 2. Reduction of fatigue with pyridostigmine is associated with reduction of jitter (on stimulation single fiber electromyography) with edrophonium injection. Jitter is a measure of neoromuscular junction transmission adequacy, and is abnormally increased in post-polio patients. Pyridostigmine-responders (open circles) are those who report a reduction in fatigue with pral pyridostigmine 180 mg per day. Pyridostigmine non-responders are illustrated with closed circles. Jitter plots are normalized to 100% of pre-edrophonium mean jitter. A significant difference in jitter means is observed with comparison of pyridostigmine-responders (n=9) before and after edrophonium injection (Bonferroni corrected, $P < 0.0001$). A significant difference is also observed between pyridostigmine-responding and non-responding patients (n=8), after edrophonium injection ($P < 0.001$). There is no significant difference in jitter means of pyridostigmine-non-responders before and after edrophonium injection ($P > 0.8$). Error bars=SEM. Reprinted from the Journal of the Neurological Sciences *[62]*, with kind permission of Elsiver Science -- NL, Sara Burgerhartstraat 25, 1055 KV Amsterdam, The Netherlands.

9. Summary

Other medical and neurological causes of new symptoms in a post-polio patient should be identified and treated before attributing new difficulties to PPS. Many patients with PPS can benefit from an interdisciplinary, individualized, management program which is dependent upon the specific symptoms. This program can include pacing with regular rest periods and naps, avoidance of overuse, judicious exercise, orthotics, assistive devices, energy conservation techniques, and life style changes.

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