AN APPROACH TO THE PATIENT WITH SUSPECTED POST POLIO SYNDROME

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Polio survivors are at risk for the occurrence of certain physiologic changes in the nervous system which result in a characteristic set of symptoms now known as Post Polio Syndrome. In addition to these unexpected physiological changes there are anticipated complications such as arthritis, scoliosis, and entrapment syndromes that frequently accompany paralytic conditions. These anticipated complications are not the problems that distinguish PPS from other diseases of the nervous system. Post Polio Syndrome (PPS) is a major chronic illness and one which poses unique problems to its survivors and their physicians.

No Diagnostic test exists for PPS, so clinical criteria must be used to establish the diagnosis. Many Physicians lack training in the diagnosis and management of a syndrome only recently acknowledged as existing. Patients are often uncomfortable with physicians they feel do not understand their problems. They also fear increased disability, often at the same time they are coping with limitations of aging. Patients are often trapped in a "conquer the disease" mentality derived from the experience of recovering from the acute episode an average of 25 years earlier. This is incompatible with the lifestyle adjustments necessary for optimal results in PPS rehabilitation.

I. INTRODUCTION

A. DEFINITION OF POST POLIO SYNDROME

An otherwise unexplained constellation of symptoms which may include weakness, fatigue, pain, heat or cold intolerance, and swallowing, breathing, or sleep disturbance developing in a patient who had paralytic polio. Post Polio Muscle Atrophy (PPMA) has been used as the label for the above symptoms when they include progressive muscle atrophy.

B. SCOPE OF THE PROBLEM

1987 National Health Interview Survey estimated 1.63 million American polio survivors (=0.625% of population), 50% with some Post Polio Syndrome symptoms.

C. DIAGNOSTIC CRITERIA

1. PPS is a diagnosis of exclusion and should be based on a thorough history and physical exam.
2. Evidence of prior paralytic polio: via EMG, an appropriate history, or characteristic residual atrophy.
3. Period of apparent stability before any new symptoms. New symptoms may often be seen after an illness or injury.
4. Exclusion of other conditions (especially motor neuron diseases and overuse syndromes).

II. PATHOLOGY: PHYSIOLOGIC AND CLINICAL CONSEQUENCES

A. EXTENSIVE NEURONAL INVOLVEMENT IN THE ACUTE POLIO INFECTION

1. The extent of central nervous system infection by polio virus is not well appreciated. Infection is far more widespread than anterior horn cells alone. Often anterior horn cell infection is largely

subclinical due to residual capacity of uninfected and surviving neurons. Infection outside the anterior horns is likely to be largely subclinical also, but may help to explain the disabling symptoms of fatigue and pain which are subjective and controversial (because the physiologic basis is uncertain).
2. Ninety-five percent (95%) of motor neurons are infected in an average acute infection, with a 50% neuronal fatality rate.
3. There is frequent segmental involvement, accounting for the lack of symmetry of weakness.
4. In addition to the anterior horns in the spinal cord, infection involves intermediolateral horns and dorsal root ganglia.
5. Infection also involves motor cortex, hypothalamus, and globus pallidus, brainstem nuclei, reticular formation, cerebellar roof nuclei, and vermis.

B. MOTOR UNIT REMODELING IN THE POST RECOVERY PHASE

1. A normal quadriceps has, on average, 200 muscle fibers/anterior horn cell and a normal anterior horn cell can adopt as many as 1,000 orphaned muscle fibers.
2. Over 50 % of motor units may be lost without symptoms. (Normal walking uses only 15-20% of maximum muscle strength.)
3. Clinical improvement occurs acutely through recovery of mildly affected neurons, collateral sprouting, and strengthening (hypertrophy) of intact musculature.
4. Increased demand on surviving motor units results in increased firing frequency which in turn produces a change in fiber type to predominantly aerobic "slow twitch" fibers with increased vascularity.

C. DECOMPENSATION THEN PRODUCES POST POLIO SYNDROME

While a single underlying etiology for PPS has not been proven, several theories exist:

1. There is an increased metabolic burden on surviving anterior horn cells (even in asymptomatic muscles) as they direct more muscle fibers to contract, more often, to achieve the same force of contraction. This leads to anterior horn cell fatigue and can lead to premature metabolic injury, perhaps even cell loss. Fatigued neurons may be unable to continue to trophically support as many muscle fibers. The collateral sprouts to some muscle fibers will degenerate. The strength of these muscle fibers will be lost to the motor unit, and a spiral of decline may set in. This mode of decompensation augured by fatigue, may be anterior horn cell based. This appears not to be a static process and there may be dynamic denervation and reinervation.
2. Another mode of decompensation is muscle fiber based: Rapidly firing muscle fibers produce more lactic acid which may not be adequately dissipated. This is especially true with any degree of isometric contraction. Muscle fiber fatigue may, lead to muscle fiber injury, lost function, and a spiral of decline.
3. Any increase in mechanical load (such as would result from increased weight or increased physical activity) or decrease in force generating capacity (such as would result from inactivity following illness or injury) may trigger metabolic failure in motor units or in muscle fibers functioning close to their capacity.
4. The resulting relative weakness may lead to joint and muscle misuse and overuse. This may lead in turn to both arthritis and overuse syndromes.
5. In addition to anterior horn cell and muscle fiber modes of fatigue, central fatigue may also be a factor. Polio virus infection of the motor strip and the reticular activating system is well described.

A working definition for central fatigue is: "Increased mental effort necessary to perform a fixed amount of muscle contraction". This is very much how Post Polio Syndrome patients describe their feelings of fatigue, many report hitting a "post polio wall".

III. PATIENT PRESENTATION

A. PRIME SYMPTOMS

A common presentation is a polio survivor who previously had lower extremity involvement in a well defined polio episode. The patient may have restricted ambulation from hiking or jogging, lived a sedentary life, and did not feel disabled. After a period of relatively stability he or she may begin to notice unusual fatigue and discomfort and may further restrict activity. Denial of decreased functional capacity may lead to a crisis as the patient can no longer can meet occupational, social, and family commitments. Persistence and attempts to continue at a previous activity level may lead to a downward spiral of decreasing functional capacity with resulting depression and despair. On examination, relative obesity may be present and weakness is easily demonstrated, often in the "good" leg; limbs considered unaffected are often subclinically affected with polio and may present with "new" polio. A statistical summary of the clinical characteristics of several series of PPS patients is as follows:

1. *Fatigue, Pain, and Weakness are almost always present.* Fatigue (89%); Pain in Muscle or Joint (86%); New weakness (83%) in previously symptomatic (69%) or asymptomatic (50%) muscles.
2. New Atrophy (28%); This equates to Post Polio Muscular Atrophy (PPMA).
3. Activities of daily living difficulties (78%) = functional loss. Walking (64%); Climbing Stairs (61%); Dressing (17%).

B. ADDITIONAL PRESENTING PROBLEMS

1. **Pulmonary dysfunction:**

   Patients with Post Polio Syndrome may suffer from weakness of the breathing muscles, namely the diaphragm and ribcage. Occasionally, this can be severe enough to cause symptoms of dyspnea on exertion and even at rest, poor clearance of respiratory secretions increasing the risk of pneumonia, and elevations in the resting arterial CO2 level. Measurement of pulmonary function tests in these patients usually shows a significant restrictive pattern (small lung volumes) on the basis on neuromuscular weakness.

   If respiratory muscle weakness is severe enough mechanical ventilation may be required. Small mechanical ventilators have been developed which deliver breaths through a comfortable plastic nose mask. This is often performed while the patient is asleep at night and results in improved daytime function.

2. **Sleep Disorders:**

   Patients with Post Polio Syndrome have a high incidence of sleep disturbances with poor sleep quality and frequent awakenings which may be due to several factors. However, the most important etiology to rule out is central, obstructive and mixed sleep apneas. Nocturnal hypoxemia and hypercarbia can lead to worsening of daytime function of the breathing muscles. Nocturnal non invasive ventilation can be used in these patients to improve sleep quality and reduce symptoms of daytime sleepiness, and perhaps improve daytime respiratory muscle function.

3. **Dysphagia:**

   Many PPS patients reported some new difficulty with eating or swallowing more commonly in those with bulbar polio. Video fluoroscopy has been used for evaluation and has frequently revealed pharyngeal constrictor weakness. Laryngeal penetration and loss of the cough reflex may occur without symptoms, suggesting an underestimation of the presence and severity of dysphagia in this population. Many patients have already employed compensation such as altering diet, cutting solids into small pieces, chewing it thoroughly, taking small sips of liquids, eating slowly, and using postural maneuvers. Most patients with dysphagia had also experienced some progressive speech difficulty such as increased hoarseness, weakness, or slurring.

4. **Cold intolerance (29%):**

   Limbs may be cold and cold exposure produces weakness. This is thought to be due to intermediolateral column involvement resulting is vasoparesis, venous pooling, and excessive heat loss.

5. **Degenerative arthritis:**

   A joint that is biomechanically disadvantaged may develop degenerative arthritis.

6. **Social and psychological problems:**

   Long term disability and denial may result in social and psychological problems.

C. PAST HISTORY

1. Average age of polio onset is 7 years. Median time to maximum recovery is 8 years. Median period of stable neurologic and functional status is 25 years. Median post polio symptom duration before patient presents for evaluation is 5 years.
2. Variables associated with shorter interval to PPS: greater severity and greater age.
3. Initial symptoms are most frequent in the lower limb most affected in the acute illness. (Upper extremity weakness is easier to compensate for without overuse resulting.)
4. The onset is usually insidious but is frequently precipitated by injury, illness, bed rest, or weight gain.

IV. EVALUATION PROCESS

A. IDENTIFY AREAS OF DYSFUNCTION

1. The history is especially useful in identifying fatigue, dysphagia, sleep disorders, and alteration in activities of daily living.
2. The Neurologic exam will identify atrophy or weakness and verify that reflexes are not increased. Pay special attention to the "good" limb as significant weakness may be present of which the patient has never been aware. With leg muscles, functional tests must be used because manual testing may not detect quadriceps weakened to 30% of normal even though this is sufficient strength for routine daily activities. Seek a mechanical advantage in manual muscle testing: Test the triceps or quadriceps with the elbow or knee flexed more than 90 degrees. Test the psoas in the supine position.

3. The general physical exam and biomechanical exam note obesity, joint deformity, overuse syndromes, and scoliosis.
4. Electromyography may be requested when needed to document previous anterior horn cell disease (especially when the previous history of polio is in doubt). EMG can also be used to rule out other neuromuscular pathologies or to identify subclinically involved muscles.
5. CK elevation may be seen in patients but may not correlate with progressive weakness.

B. FORMALIZE TREATMENT GOALS

After the diagnosis of PPS is established, a patient conference is a convenient way to formalize treatment goals and begin patient education. These areas should be addressed:

1. **Lifestyle Modifications:**

   This item is the "sine qua non" of all attempts at successful management of PPS. At the time of formal diagnosis, patients are often desperate, yet imbued with a belief in their own ability to overcome their disability through the "no pain, no gain" approach. This approach may have served them very well after their acute attack of polio many years ago but is now actually self destructive. Persistence in this approach of "overcoming" illness has led to a spiral of deteriorating function and frequently a parallel decline in self worth. Patients must understand the concept of "living with" PPS in order to lead the fullest life possible. An understanding of the need for lifestyle modification is rarely achieved at the first visit and is often best reintroduced by a knowledgeable Occupational or Physical Therapist and reinforced and monitored at subsequent physician office visits.

2. **Increase Muscle Capacity:**

   a) Muscular capacity can be increased by achieving increased strength or endurance. Strength can be increased through isometric exercise. However, muscles must be carefully selected for isometric exercises. Some muscles will already be functioning at their maximum. Exercise may actually have a deleterious effect by forcing these muscles beyond their metabolic capacity and producing injury.

   b) Endurance may be increased, susceptibility to fatigue decreased, and long term deterioration minimized through appropriate exercise supervised by a physical therapist experienced with post polio patients. Almost all patients have initial difficulties with exercise programs resulting from overdoing. They may also equate fatiguing daily activities (which challenge the weakest musculature and do not provide an effective aerobic training level) to exercise. This can be an instructive opportunity for the patient in understanding the "Lifestyle modification" and to experience its benefits. Goals in aerobic exercise are:

      1. Educate the patient to avoid potentially harmful exercise-induced fatigue. A reasonable approach would be to establish the level of peak performance by patient history. Then start at 50% of peak performance and slowly increase performance as tolerated.
      2. Select exercises which can create a training effect in the patient with weakened, atrophic musculature and overuse syndromes. Exercise intervals with intervening rests are necessary, just as is pacing of daily activities. A knowledgeable Physical Therapist can be crucial to this aspect of management.

   c) Muscle capacity can also be increased by bracing, orthotics, or other aids which extend, amplify or substitute for muscles.

d) Pharmacologic treatment of fatigue: Some medications seem to raise the threshold for fatigue. These observations are, as yet, anecdotal and await confirmation from clinical trials.

1. *Amantadine*: up to 100 mg BID as tolerated.
2. *Deprenyl*: up to 5 mg BID as tolerated.
3. *Mestinon*: up to 60 mg TID when careful monitoring is available.

Medications for the amelioration of fatigue must be understood as aids which can give a running start to the rehabilitation process. However, if they are perceived by the patient as a form of curative treatment, they will only forestall the day of reckoning.

3. **Decrease Muscle Load To Less Than Muscle Capacity:**

a) PACING of activity is the logical consequence of a successful LIFESTYLE MODIFICATION. Implementing of PACING requires that patients identify for each of the activities of daily living the length of time they may participate before experiencing fatigue. They must then break up their activities into smaller modules of time, each of which is of less duration than the time required to produce fatigue. A corollary concept to PACING is ENERGY BUDGETING which imagines that one has a fixed expenditure of energy for each day and that this sum should be "spent" on activities of the highest personal priority. (Exceeding this daily limit may be conceptualized as spending principle or acquiring debt but probably correlates to metabolic injury of the motor unit through

overuse.)

b) Other means of decreasing muscle load are diet when overweight, use of orthotics to improve mechanical efficiency, use of wheelchairs or scooters to save energy expenditure, and treatment of chronic overuse syndromes.

4. **Treat Specific Complications:**

a) Attention to specific complications such as dysphagia, pulmonary dysfunction and sleep disturbances may require specific referrals. The goals of these referrals can be addressed with the patient at this first conference.

b) Functional consequences also result from overuse syndromes which can lead to joint deformity. Physiatry consultation can be helpful here and orthopedic intervention is occasionally required. Evaluate need for orthotic prescriptions (i.e., splints, braces, AFO's)

c) Somatization, depression, anxiety, and self worth problems may occur as capacity decreases. Referral for counseling should be considered (MSW, psychologist) or polio support group (see reference section).

d) Evaluate and/or modify work duties through referral to occupational therapist or vocational counselor.

C. PROGNOSIS

Patients often present during a period of decompensation. Decompensation may be caused by even slight embarrassment in strength due to inactivity or injury superimposed upon aging. It may also result from slight increase in muscular work resulting from weight gain or increase in activities. In either case, a spiral

of deterioration may result from potential overuse injury to the motor unit and subsequent decrease in functional capacity can result. Patients may easily become fearful and depressed at this ominous decline in their previously stable, if compromised, neuromuscular status.

It is important to clarify for the patient the difference between deterioration in function and deterioration from disease progression. In fact, there is little evidence that any loss of function experienced by PPS patients is due to progression or recurrence of polio virus infection. If patients can understand that opposing forces of muscle strength versus muscle load are acting near a capacity threshold, they will be quicker to accept PACING concepts, to employ an appropriate exercise program, and to utilize other elements of rehabilitation. In most cases, this will allow the patient to return to or approach the previous functional baseline. It is not difficult for patients to then minimize deterioration in function over the years by:

1. Achieving an optimal balance between muscle strength and endurance (achieved and maintained by exercise) versus muscle burden (resulting from body weight, mechanical inefficiencies, and activity level).
2. Utilizing PACING and restriction of activities after the point of fatigue so that muscle work is kept within the limits of muscle capacity and decompensation does not occur.
3. Gradually decreasing total daily energy expenditure over the years much as a non PPS individual might do. This rarely results in much loss of individual activities or functions, only in the amount of each that is performed each day.

V. RESOURCES IN PATIENT MANAGEMENT

The patient with PPS is best served by having a physician who has experience evaluating post polio symptoms, formalizing treatment goals, and making the appropriate referrals such as those listed below:

A. NEUROLOGY CONSULTATION

When the Diagnosis is in question.

B. PHYSIATRY (PHYSICAL MEDICINE AND REHABILITATION)

A Physiatrist is a physician with expertise in the orchestration of the rehabilitation process. Especially when disability is severe, complex, or when biomechanical problems are prominent, physiatry consultation can help with the initial planning and selection of specific exercise programs, physical therapy, orthotics, and adaptive equipment.

C. PHYSICAL THERAPY

A Physical Therapist who is experienced regarding PPS will be of tremendous value in introducing and customizing the lifestyle modifications and in introducing the useful concepts of pacing and energy budgeting. Physical Therapists can also screen for inefficiency in movement resulting from deformity or weakness, assist in establishing your patient on a safe exercise program, and monitor for the almost inevitable initial over indulgence in that program.

D. OCCUPATIONAL THERAPY

Occupational Therapists are trained to assess the home environment and the patient's daily activities in order to restructure tasks, introduce mechanical aids like grab bars, and provide devices such as sock lifters which make possible physical activities otherwise compromised by disability. Instruction in PACING of routine daily activities and associated lifestyle modification can also be provided by an Occupational Therapist.

E. SPEECH PATHOLOGY

A speech pathologist can help in the evaluation and treatment of swallowing and speech problems.

F. PULMONOLOGY

A Pulmonologist can evaluate and manage respiratory dysfunction and sleep dysfunction.

G. PSYCHOLOGY

A psychologist or MSW can evaluate and counsel regarding reactive depression, coping strategies, pain management and life style adjustment. This is especially important to help the post-polio survivor deal with the "reemergence" of a neuromuscular disorder they thought had been previously conquered.

H. SUPPORT GROUPS

Local education/support groups meet on a monthly basis in various locales, offering education, support, and social opportunities for polio survivors and their families.

I. OTHER

Orthopedics, nutrition, and social work referrals for evaluation will occasionally be useful in specific circumstances.

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Registered Charity No. 1064177
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Document preparation: Chris Salter, Original Think-tank, Cornwall, United Kingdom.
Document Reference: <URL:http://www.zynet.co.uk/ott/polio/lincolnshire/library/easterseal/approach.html>

Created: January 1997
Last modification: 24th January 2010.