

POST-POLIO SYNDROME SLIDE KIT

THE POST-POLIO TASK FORCE

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Foreword

After years of skepticism, the medical community is beginning to acknowledge the clinical entity known as Post-Polio Syndrome (PPS). This condition affects polio survivors decades after their recovery from their initial bout with polio, bringing fatigue, weakness, and pain that is often debilitating. As many as 250,000 persons may be affected by this disorder in the U.S. alone. The diagnosis of PPS is difficult – in fact, it is currently a diagnosis of exclusion – and there is no “gold standard” of treatment. However, many patients are helped with conventional management techniques such as physical rehabilitation, assistive devices, and lifestyle modifications. In addition, several promising treatments are being investigated in clinical trials, and it is hoped that ongoing research into the pathophysiology of PPS will lead to even more effective approaches.

This slide lecture kit represents a collaborative effort by the Post-Polio Task Force – a group of eight experts from the PPS healthcare and patient communities – whose mission is to raise awareness of, and disseminate information on PPS. The kit is divided into five sections: I. Epidemiology, Natural History, Definition, and Diagnosis of PPS; II. Pathophysiology of PPS; III. Evaluation and Differential Diagnosis of PPS; IV. Management of PPS; and V. Optimizing Wellness: The Role of the Patient in PPS. This kit is intended to provide the materials and information necessary to deliver a comprehensive lecture representing our current knowledge of PPS, and thus help to increase awareness of this little-known, yet potentially devastating disorder.

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I. Epidemiology, Natural History, Definition, Diagnosis of PPS

Slide 1.

Epidemiology of PPS

Epidemiologic studies of post-polio syndrome (PPS) have provided widely varying estimates of the prevalence of PPS, depending on the criteria used to define the condition – muscle atrophy (encompassing the symptoms of new weakness) or musculoskeletal stress (encompassing the symptoms of joint pain and joint dysfunction). One of the earliest epidemiologic studies¹ estimated that 17% of acute paralytic poliomyelitis (APP) survivors had new joint pain and 16% had new weakness; overall, 22% of APP survivors were reported to have experienced new problems or functional deterioration since their recovery from APP. In a later study by Speier et al,² 42% of patients who were previously hospitalized for polio reported new problems; of those 47% complained of pain or cramping and 42% complained of decreased endurance. A 1991 study by Windebank et al³ found that 64% of APP survivors experienced new manifestations and 44% had new weakness, alone or in combination with pain and/or

fatigue. And a 1992 study by Rambow et al⁴ found that 28% of survivors of paralytic polio had PPS (both new muscle weakness and new muscle pain were required for classification as PPS); 42% of the study subjects had joint pain, 38% had muscle weakness, and 38% had muscle pain. On the basis of symptoms, therefore, PPS will develop 20% to 40% of APP survivors. Better data based on prospective analyses and objective criteria are clearly needed.

1. Codd MB, Mulder DW, Kurland LT, et al. Poliomyelitis in Rochester, Minnesota, 1935-1955: Epidemiology and long-term sequelae: a preliminary report. In: Halstead LS, Wiechers DO, eds. *Late Effects of Poliomyelitis*. Miami: Symposia Foundation, 1985: 121-134.
2. Speier JL, Owen RR, Knapp M, Canine JK. Occurrence of post-polio sequelae in an epidemic population. In: Halstead LS, Wiechers DO, eds. *Research and Clinical Aspects of the Late Effects of Poliomyelitis*. White Plains, NY: March of Dimes Birth Defects Foundation, 1987:39-48.
3. Windebank AJ, Litchy WJ, Daube JR, Kurland LT, Codd MB, Iverson R. Late effects of paralytic poliomyelitis in Olmstead County, Minnesota. *Neurology*. 1991;41:501-507.
4. Ramlow J, Alexander M, LaPorte R, Kaufmann C, Kuller L. Epidemiology of the post-polio syndrome. *Am J Epidemiol*. 1992;136:769-786.

Epidemiology of PPS

- **Estimates of the prevalence of post-polio syndrome (PPS) have varied widely, depending on the criteria used to define it**
- **On the basis of symptoms, PPS will develop in about 50% of survivors of acute paralytic polio (APP)**
- **Retrospective studies using objective criteria have estimated that PPS will develop in 20% to 40% of APP survivors**
- **Better data based on prospective analyses and objective criteria are needed**

I. Epidemiology, Natural History, Definition, Diagnosis of PPS

Slide 2-5.

Natural History of PPS

The poliovirus causes APP in only a small number of cases; for most people, the infection results in nothing more than a benign viral illness that last for a few days. Those who do have APP usually recover at least some muscular function within the first few months of infection, but in some cases recovery can take several years. Subsequent to recovery is a period of neurologic stability, usually lasting 15 years or more. In up to 50% of APP survivors, this period of neurologic stability may be followed by new weakness, fatigue, and pain in the muscles and/or joints – the most common symptoms of PPS. Less common PPS symptoms include new muscle atrophy (loss of muscle bulk), respiratory insufficiency (breathing difficulties), dysarthria (speech disturbances), dysphagia (swallowing difficulties), muscle cramps, cold intolerance, fasciculation (muscle twitches), and joint deformities.

Natural History of PPS

- **Acute Anterior Poliomyelitis**
 - For vast majority: a benign viral illness for 2-3 days—fever, headaches, malaise, GI symptoms
 - <5%: CNS invasion—stiff neck, positive spinal tap, no clinical paralysis (potentially at small risk for PPS)
 - 1% to 2%: variable degree of clinical paralysis (risk of PPS varies with extent of paralysis)

Natural History of PPS (cont'd.)

- **Period of Recovery**
 - Can last from weeks to years
 - Varies with severity and age at onset of APP
 - Complicated by developmental growth (in children) and reconstructive surgery

I. Epidemiology, Natural History, Definition, Diagnosis of PPS

Natural History of PPS (cont'd.)

- **Period of Neurologic Stability (15 years)**
 - Begins with plateau of maximum neurologic and functional recovery
 - Lasts indefinitely in about 50% of persons with paralytic polio
 - For 20% to 50%, ends with onset of new weakness and other PPS symptoms

Natural History of PPS (cont'd.)

- **Onset of PPS (typically 30-50 years after polio):**
 - New weakness
 - Excessive fatigue
 - Muscle and/or joint pain
 - Muscle atrophy
 - Dysphagia
 - Breathing difficulties
 - Cold intolerance

I. Epidemiology, Natural History, Definition, Diagnosis of PPS

Slide 6.

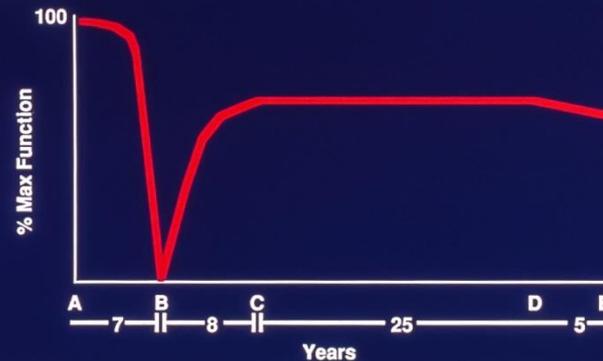
Natural History of Poliomyelitis in a Post-Polio Clinic

The accompanying graph shows changes in maximum physical function over time in 132 consecutive individuals with PPS seen at a post-polio clinic.¹ Points 'A' and 'B' indicate birth and time of acute polio, respectively. Maximum functional and neurologic recovery, designated by 'C,' occurred at a median of 8 years after acute polio. The median time of neurologic stability was 25 years (C to D), and the median time from onset of polio to onset of PPS symptoms was 33 years (B to D). Time of evaluation, indicated by (E), was a median of 5 years after onset of PPS symptoms.

1. Halstead LS, Rossi CD. Post-polio syndrome: clinical experience with 132 consecutive outpatients. In: Halstead LS, Wiechers DO, eds. *Research and Clinical Aspects of the Late Effects of Poliomyelitis*. White Plains, NY: March of Dimes Birth Defects Foundation, 1987:13-26.

Natural History of Poliomyelitis in a Post-Polio Clinic

Functional changes at key milestones for 132 persons with PPS (median times)



A=birth; B=acute polio (7 years); C=maximum neurologic and functional recovery (B to C was 8 years); D=onset of new health problems (B to D was 33 years); E=time of evaluation (D to E was 5 years).

Adapted from Halstead LS, Rossi CD. In: Halstead LS, Wiechers DO, eds. *Research and Clinical Aspects of the Late Effects of Poliomyelitis*. White Plains, NY: March of Dimes Birth Defects Foundation, 1987:13-26.

I. Epidemiology, Natural History, Definition, Diagnosis of PPS

Slide 7.

Definition of PPS

PPS is a disorder that affects polio survivors years after recovery from their initial bout with polio. An interval of 30-50 years usually elapses before the first PPS symptoms occur, but intervals as short as 8 years and as long as 71 years have been documented. New weakness, fatigue (both generalized and specific to the muscles), and pain involving the muscles and/or joints are the 3 most common symptoms of PPS, but other signs and symptoms, such as muscle atrophy, breathing and swallowing difficulties, and cold intolerance, can also occur. Progressive deterioration of motor units is the most likely cause for PPS-related weakness, abnormal muscle fatigability (ie, decreases endurance), and muscle atrophy, whereas new weakness and/or chronic musculoskeletal “wear and tear” may be the primary causes of muscle and joint pain.

Note: This definition represents a consensus statement of the Post-Polio Task Force.

Definition of PPS

- **PPS is a neurologic disorder characterized by a cluster of symptoms in individuals who had paralytic polio years earlier**
- **Typically, these symptoms occur after a period of functional stability of at least 15 years after APP and include new weakness, fatigue, and pain (of muscles and/or joints)**
- **Less commonly, the symptoms include muscle atrophy, breathing and swallowing difficulties, and cold intolerance**
- **The new weakness, abnormal muscle fatigability (decreased endurance), and atrophy are most likely caused by a slowly progressive deterioration of motor units**
- **Muscle and joint pain are most likely caused by new weakness and/or chronic musculoskeletal “wear and tear”**

This definition represents a consensus statement of the Post-Polio Task Force.

I. Epidemiology, Natural History, Definition, Diagnosis of PPS

Slide 8.

Components of PPS: PPMA vs. Musculoskeletal PPS

PPS can be thought of as consisting of two distinct components: post-polio progressive muscular atrophy (PPMA), comprising the neurologic manifestations of illness, and musculoskeletal PPS, comprising the orthopedic manifestations.^{1,2} PPMA is characterized by new weakness, which is frequently accompanied by fatigue, muscle pain, and atrophy. The symptoms presumably result from degeneration of motor axonal sprouts and motor neurons. Musculoskeletal PPS is characterized by new joint pain or joint dysfunction, frequently accompanied by fatigue, joint tenderness, and joint swelling. These symptoms presumably result from chronic overstress of joints and periarticular structures (ie, tendons, ligaments, and bursae). The overstress is thought to occur because of chronic abnormal use of extremities due to residual weakness from the original poliomyelitis.

1. Wiechers DO. Late effects of poliomyelitis: case reports. In: Halstead LS, Wiechers DO, eds. *Late Effects of Poliomyelitis*. Miami: Symposia Foundation, 1985:11-31.
2. Dalakas MC. The post-polio syndrome as an evolved clinical entity: definition and clinical description. In: Dalakas MC, Bantfield H, Kurland LT, eds. *The Post-Polio Syndrome: Advances in the Pathogenesis and Treatment*. *Ann NY Acad Sci*. 1995;753:68-80.

Components of PPS: PPMA vs. Musculoskeletal PPS

- **Post-polio progressive muscular atrophy (PPMA): neurologic manifestations**
 - New weakness, often accompanied by fatigue, muscle pain, and atrophy
 - Thought to result from degeneration of motor axonal sprouts and motor neurons
- **Musculoskeletal PPS: orthopedic manifestations**
 - New joint pain/dysfunction, often accompanied by fatigue, joint tenderness, and joint swelling
 - Thought to result from chronic overstress of joints and periarticular structures because of chronic abnormal use of extremities due to residual weakness/joint instability from original poliomyelitis

I. Epidemiology, Natural History, Definition, Diagnosis of PPS

Slide 9.

Diagnostic Criteria for PPS

Diagnosis of PPS can be difficult because many of the symptoms of PPS overlap with those of other diseases, such as fibromyalgia, hypothyroidism, depression, rheumatoid arthritis, polymyalgia rheumatica, and a number of neurologic conditions. The accompanying slide presents some general criteria for making the diagnosis of PPS.

Note: These criteria represent a consensus statement of the Post-Polio Task Force. The sensitivity, specificity, and reliability of these criteria have not yet been tested in a prospective manner.

Diagnostic Criteria for PPS

- 1) A prior episode of paralytic poliomyelitis with residual motor neuron loss (which can be confirmed through a typical patient history, a neurologic examination, and, if needed, an electrodiagnostic exam)
- 2) A period of neurologic recovery followed by an interval (usually 15 years or more) of neurologic and functional stability
- 3) A gradual or abrupt onset of new weakness and/or abnormal muscle fatigability (decreased endurance), with or without generalized fatigue, muscle atrophy, and/or pain
- 4) Exclusion of medical, orthopedic, and/or neurologic conditions that may be causing the symptoms mentioned in (3)

These criteria represent a consensus statement of the Post-Polio Task Force. The sensitivity, specificity, and reliability of these criteria have not yet been tested in a prospective manner.

II. Pathophysiology of PPS

Slide 10-13.

Peripheral Disintegration Model of PPS

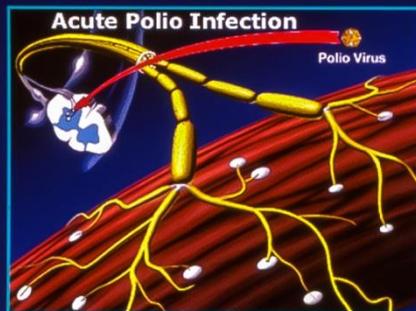
The hallmark of polio pathophysiology is the invasion and destruction of motor neurons by poliovirus, with subsequent loss of innervation in muscle fibers. The recovery phase is characterized by axonal sprouting, resulting in reinnervation of the muscle fibers. Wiechers and Hubbell proposed that PPS arises because these axonal sprouts degenerate over time, with consequent denervation of the associated muscle fibers.¹ This theory is known as the peripheral disintegration model of PPS.

The peripheral disintegration model of PPS is illustrated in the four accompanying slides. In acute polio, normal motor neurons die as a result of invasion and destruction by poliovirus (Slide 10), and the muscle fibers innervated by these neurons lose their innervation (Slide 11).

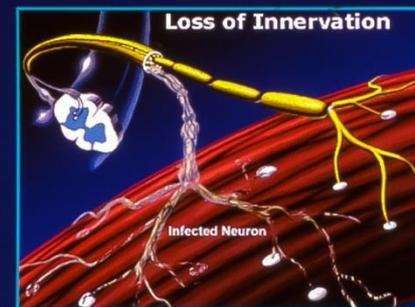
In the recovery period (Slide 12), there is axonal sprouting with reinnervation of the denervated muscle fibers. This reinnervation is not indefinitely stable however, and can decrease or degenerate with time. PPS is likely characterized by loss of innervation of the reinnervated muscle fibers (Slide 13).

1. Wiechers DO, Hubbell SL. Late changes in the motor unit after acute poliomyelitis. *Muscle Nerve*. 1981;4:524-528.

Peripheral Disintegration Model of PPS (I)

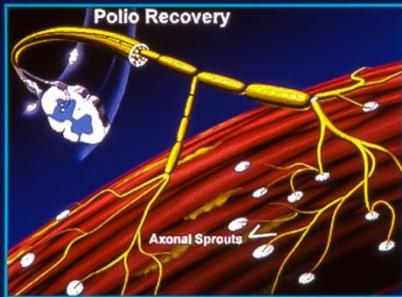


Peripheral Disintegration Model of PPS (II)



II. Pathophysiology of PPS

Peripheral Disintegration Model of PPS (III)



Peripheral Disintegration Model of PPS (IV)



II. Pathophysiology of PPS

Slide 14.

Motor Nerve Terminal Damage in PPS: Histologic Evidence

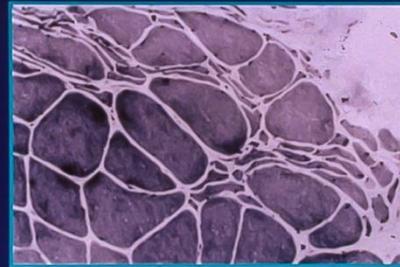
Histology studies have supported the peripheral disintegration model, demonstrating that axonal degeneration is an ongoing process in the period following acute polio. The presence of isolated angular atrophic fibers in muscle biopsy specimens indicated recent motor nerve terminal degeneration; these fibers can be clearly differentiated from fibers that lost their innervation during acute polio. The left panel of the accompanying slide demonstrates an angular atrophic muscle fiber in the rectus femoris of a PPS patient reporting 1 year of new leg weakness (hematoxylin eosin stain).

There is also compelling evidence of ongoing axonal dysfunction in PPS patients. Unlike axonal degeneration, this process may be reversible and thus may be amenable to therapeutic intervention. Investigators have identified normal-sized myofibers expressing neural-cell adhesion molecule (N-CAM), a molecule expressed by denervated muscle fibers.¹ These may be fibers that have lost innervation and will ultimately become small and angulated, or they may be only temporarily denervated and may be destined to become reinnervated. The right panel of the accompanying slide illustrates N-CAM reactivity in small- and large (normal)-diameter muscle fibers.

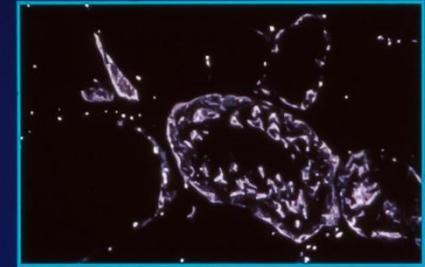
A number of hypotheses regarding the etiology of terminal axon dysfunction are being investigated. One theory is that dysfunction is an intermediate stage in degeneration. Another is that dysfunction is associated with motor unit remodeling. Motor units in a polio muscle are probably in a constant phase of dynamic remodeling, with, for example, denervation of a muscle fiber from one motor unit being followed by axonal sprouting to the fiber from another motor unit. A third possible cause of dysfunction is age-related changes in trophic support of terminal axons. For example, age-related decreases in secretion of growth hormone may result in decreases in circulating levels of insulin-like growth factor I (IGF-1), which is critical to the maintenance of terminal axons.

1. Cashman NR, Maselli K, Wollman RL, Ross R, Simon R, Antel JP. Late denervation in patients with antecedent paralytic poliomyelitis. *N Engl J Med.* 1987;317:7-12.

Motor Nerve Terminal Damage in PPS: Histologic Evidence



Angular atrophic muscle fiber (PPS patient)



N-CAM reactivity: small- and large (normal)-diameter muscle fibers

Reprinted with permission from Cashman NR et al. *NEJM.* 1987;317:7-12.

II. Pathophysiology of PPS

Slide 15.

Motor Nerve Terminal Damage in PPS: Electrophysiologic Evidence

Electrophysiologic data have also demonstrated significant motor nerve terminal dysfunction in post-polio patients. These findings include (1) a decrement in motor unit action potential in repetitive nerve stimulation studies, (2) increased single-fiber electromyography (SFEMG) jitter, and (3) abnormal stress response on stimulation SFEMG.

Motor Nerve Terminal Damage in PPS: Electrophysiologic Evidence

- **Decrement in motor unit action potential in repetitive stimulation studies**
- **Increased single-fiber electromyography (SFEMG) jitter**
- **Abnormal “stress response” on stimulation SFEMG**

II. Pathophysiology of PPS

Slide 16.

Repetitive Stimulation Studies in PPS

The accompanying slide shows results of repetitive stimulation studies in PPS patients (n = 8), polio control patients (ie, those without PPS symptoms [n = 4]), and normal subjects (n = 7). Repetitive stimulation at high rates was found to produce a decrement in motor unit action potential. In measurements of the compound muscle action potential (CMAP) amplitude at 30 Hz over the 1st to 5th stimuli, PPS patients exhibited a decrease in potential greater than that seen in polio controls (P = 0.056) and significantly greater than that seen in normal subjects (P = 0.01).

Repetitive Stimulation Studies in PPS

	PPS Patients (n = 8)	Polio Controls (n = 4)	Normals (n = 7)
Percent change in CMAP amplitude at 30 Hz (1st to 5th stimuli)	-19.24 ± 9.31*	-2.25 ± 18.74†	2.73 ± 18.3
Percent change in CMAP amplitude at 30 Hz (maximum to minimum stimulus)	-50.81 ± 15.89	-60 ± 25.55	-28.01 ± 76.85
Percent change in CMAP amplitude at 3 Hz (1st to 5th stimuli)	-0.44 ± 3.39	-0.9 ± 2.86	4.29 ± 5.5

All values expressed as mean ± SD

*Difference between PPS patients and normal controls (P = 0.01)

†Difference between polio controls and PPS patients (P = 0.056)

CMAP = compound muscle action potential

II. Pathophysiology of PPS

Slide 17.

SFEMG Abnormalities in PPS Patients

Evidence of motor nerve terminal dysfunction also comes from SFEMG studies showing (1) increased “jitter” – a measure of terminal axonal dysfunction and defective neuromuscular junction transmission in polio patients – and (2) an abnormal stress response on stimulation SFEMG.

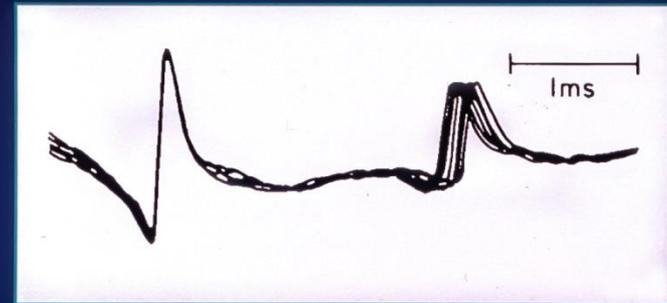
Increases jitter has been demonstrated in both polio controls and patients with PPS, and it has been found that increases in jitter is correlated with severity of original denervation in acute polio.¹ The accompanying slide represents SFEMG data from an asymptomatic, 64-year-old patient with prior paralytic polio. Shown are 10 superimposed action potentials demonstrating jitter associated with motor nerve terminal degeneration.

Electrophysiologic studies have also demonstrated an impaired “stress response” to stimulation SFEMG. In experiments in which jitter was measured in an activated neuromuscular junction over a range of rates of stimulation, it was found that patients who exhibited increased SFEMG jitter in response to increased stimulation rate were more likely to be a greater number of years removed from acute polio than were those who did not – a finding suggesting the presence of a time-dependent lesion in PPS-related terminal axonal dysfunction.²

1. Cashman NR, Maselli K, Wollman RL, Ross R, Simon R, Antel JP. Late denervation in patients with antecedent paralytic poliomyelitis. *N Engl J Med.* 1987;317:7-12.
2. Trojan DA, Gendron D, Cashman NR. Stimulation frequency-dependent neuromuscular junction transmission defects in patients with prior poliomyelitis. *J Neurol Sci.* 1993;118:150-157.

SFEMG Abnormalities in PPS Patients

SFEMG Jitter Associated With Motor Nerve Degeneration



Cashman NR et al. *NEJM.* 1987;317:2-12.

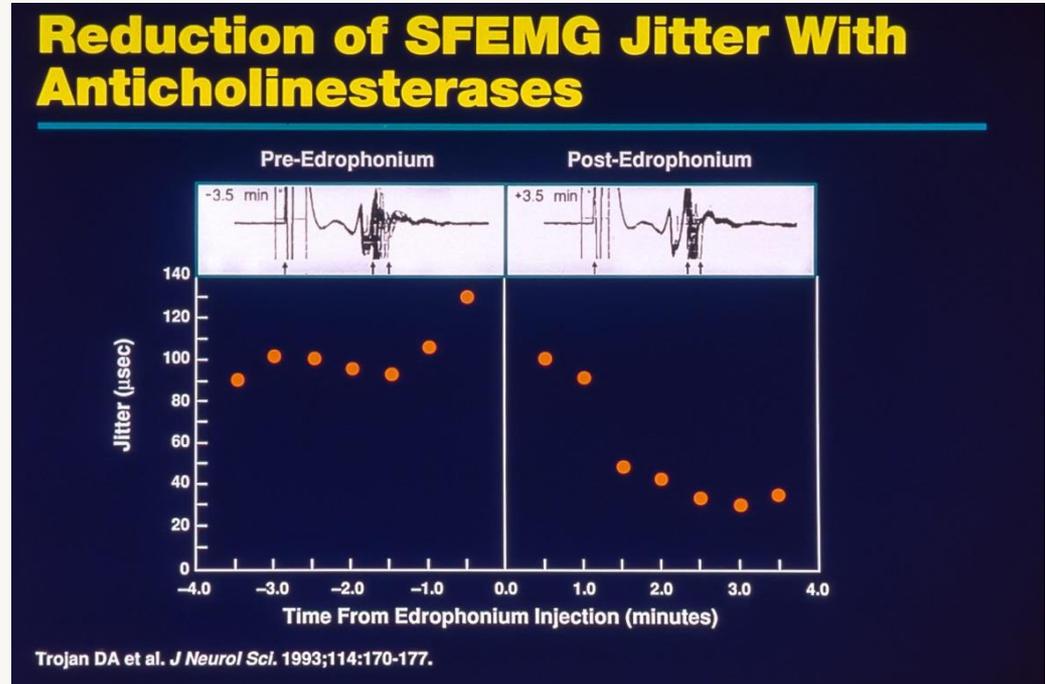
II. Pathophysiology of PPS

Slide 18.

Reduction of SFEMG Jitter With Anticholinesterases

One of the principal functions of the terminal motor axon is the release of acetylcholine to stimulate postsynaptic muscle fibers. Repetitive stimulation studies performed nearly 50 years ago showed that decrements in potentials in polio patients could be reversed with injection of the anticholinesterase neostigmine.¹

In recent studies, it has been shown that injection of the short-acting anticholinesterase edrophonium results in a marked improvement in SFEMG jitter in 50% to 60% of patient exhibiting increased jitter, suggesting the presence of a cholinergic nerve terminal defect.² The accompanying slide shows SFEMG data for one patient from a 1993 study by Trojan et al.² Jitter means were recorded every 30 seconds for 5 minutes before and 5 minutes after injection of edrophonium 10 mg. Mean jitter for the 5 minutes before edrophonium ($95.3 \pm 5.09 \mu\text{sec} \pm \text{SEM}$) significantly differed from mean jitter for the 5 minutes after edrophonium ($47.6 \pm 8.41 \mu\text{sec} \pm \text{SEM}$; $P < 0.0001$). These findings suggest the potential to treat motor nerve terminal dysfunction with anticholinesterase agents. Indeed, it has been observed that 50% to 60% of PPS patients who receive the oral anticholinesterase pyridostigmine exhibit improvement of fatigue, and that response is predictable on the basis of the electrophysiologic test response to edrophonium.²⁻⁴



1. Hodes R. Electromyographic study of defects of neuromuscular transmission in human poliomyelitis. *Arch Neurol Psych.* 1948;60:457-473.
2. Trojan DA, Gendron D, Cashman NR. Anticholinesterase-responsive neuromuscular junction transmission defects in post-poliomyelitis. *J Neurol Sci.* 1993;114:170-177.
3. Trojan DA, Cashman NR. An open trial of pyridostigmine in post-poliomyelitis syndrome. *Can J Neurol Sci.* 1995;22:223-227.
4. Seizert BP, Speier JL, Canine JK. Pyridostigmine effect on strength, endurance, and fatigue in post-polio patients. *Arch Phys Med Rehabil.* 1994;75:1049. Abstract.

III. Evaluation and Differential Diagnosis of PPS

Slide 19.

Differential Diagnosis of PPS

The diagnosis of PPS is one of exclusion; thus, differential diagnosis of this disease requires attention to a variety of conditions that could be responsible for the symptoms defining PPS. Important components in establishing the diagnosis include (1) verifying the original diagnosis of acute polio, (2) evaluating the extent and severity of acute polio residua, (3) developing a differential diagnosis for each presenting symptom complex, and (4) using appropriate diagnostic tests to exclude other potential causative conditions.

Differential Diagnosis of PPS

- **Verify the original diagnosis of acute paralytic polio (APP)**
- **Evaluate the extent and severity of APP residua**
- **Develop a differential diagnosis for each presenting symptom complex**
- **Use appropriate diagnostic tests to exclude other potential causative conditions**

III. Evaluation and Differential Diagnosis of PPS

Slide 20.

Establishing the Original Diagnosis of PPS

Important elements in establishing the original diagnosis of acute paralytic polio (APP), as detailed on the accompanying slide, consist of reviewing old medical records, reviewing any history of acute illness and recovery for typical features, performing an examination for typical residua, and performing an electrodiagnostic exam in the event that diagnosis remains uncertain.

Establishing the Original Diagnosis of APP

- Obtain and review old records
- Review history of acute paralytic illness and recovery for typical features
- Examine for typical residua (asymmetric atrophy, areflexia)
- Perform electrodiagnostic exam if diagnosis is still uncertain

III. Evaluation and Differential Diagnosis of PPS

Slide 21.

Evaluating the Extent and Severity of APP Residua

Components in the evaluation of extent and severity of APP residua are shown on the accompanying slide. Such an evaluation is also important for establishing a baseline for those aspects of function that may be affected by PPS. The components of the evaluation include a comprehensive functional history, which should involve a description of physical activities that require strength and that patients may be unable to perform as well as they did in the past (eg, stair climbing, running, and walking). Other components include manual strength testing, isometric strength measurements, joint range of motion (ROM) studies, and gait evaluation, which may include laboratory gait studies. Pulmonary function testing is particularly important in any post-polio patient in whom there is any suggestion of bulbar involvement, and should be done in virtually all polio patients.

Evaluating the Extent and Severity of APP Residua

- **Comprehensive functional history**
- **Manual muscle testing**
- **Isometric strength measurement**
- **Joint ROM measurement**
- **Gait evaluation**
- **Pulmonary function testing**

III. Evaluation and Differential Diagnosis of PPS

Slide 22.

Developing a Differential Diagnosis and Plan

A suggested initial step in developing a differential diagnosis and plan in PPS patients is to define each presenting symptom complex – ie, fatigue, weakness, or pain – in terms of characteristics, onset/duration, location(s), and activities that increase or decrease symptoms. The presenting symptoms should be evaluated in the context of the patient's general health and lifestyle, including the demands of lifestyle on their strength and endurance. Elements to be considered in differential diagnosis of each symptom complex are discussed in the following slides.

Developing a Differential Diagnosis and Plan

- **Define each presenting symptom complex**
 - Characteristics
 - Onset/duration
 - Location(s)
 - Activities that increase or decrease symptoms
- **Consider symptoms in relationship to general health, APP residua, and lifestyle**
- **Develop diagnostic plan**

III. Evaluation and Differential Diagnosis of PPS

Slide 23.

Fatigue in PPS: Systemic Metabolic Disease

Three broad categories to be considered in the differential diagnosis of fatigue in a potential PPS patient are systemic metabolic disease, ventilatory dysfunction, and depressive disorders.

Systemic metabolic conditions that may be associated with fatigue include hypothyroidism; cancer or chemotherapy; anemia; heart conditions, including heart failure or coronary artery disease; diabetes; and renal or hepatic disease. For any patient presenting with potential PPS, evaluation tests should include a thyroid test, blood chemistry, complete blood count (CBC), electrocardiogram (ECG), and chest x-ray.

Fatigue in PPS: Systemic Metabolic Disease

Differential Diagnosis

Hypothyroidism

Cancer

Anemia

Heart disease

Diabetes mellitus

Kidney disease

Liver disease

Evaluation Tests

Thyroid function tests

Blood chemistry

CBC

ECG

Chest radiograph

III. Evaluation and Differential Diagnosis of PPS

Slide 24.

Fatigue in PPS: Ventilatory Dysfunction

Assessment of ventilatory dysfunction as a potential source of fatigue should include evaluation for sleep apnea, chronic alveolar hypoventilation, and hypoxemia. Evaluation tests should include a screening pulmonary function test (PFT) and arterial blood gas (ABG) analysis. If sleep apnea is suspected, overnight oximetry studies and sleep studies are appropriate.

Fatigue in PPS: Ventilatory Dysfunction

Differential Diagnosis

Sleep apnea
Chronic alveolar hypoventilation
Hypoxemia

Evaluation Tests

PFT/ABG
Overnight oximetry
Sleep studies

III. Evaluation and Differential Diagnosis of PPS

Slide 25.

Fatigue in PPS: Depression

Evaluation of depression or other mood disturbances as a potential cause of fatigue should include assessment for major depressive illness, minor depression (frequently in association with pain or functional changes), and depressive symptoms associated with sleep disturbance, fibromyalgia, or stress/post-traumatic stress syndrome. Evaluation tests should include in-depth psychiatric testing for depressive symptoms, with appropriate referral for counseling or support groups if necessary. A trial of antidepressant medication may be indicated in some cases. Functional rehabilitation may serve to alleviate physical problems that may be causing, or contributing to, mood disturbance.

Fatigue in PPS: Depression

Differential Diagnosis

Major (classic symptoms)
Minor (associated with pain and/or functional change)
Associated with sleep disturbance
Associated with fibromyalgia
Associated with stress/PTS

Evaluation Tests

Psych testing
Counseling
Support groups
Trial of medication
Functional rehab

III. Evaluation and Differential Diagnosis of PPS

Slide 26.

Weakness in PPS: New Superimposed Neurologic Conditions

Four broad categories in the differential diagnosis of weakness in potential PPS patients are new superimposed neurologic conditions, disuse atrophy, overuse or chronic strain, and symptoms associated with systemic comorbid medical conditions.

Potential superimposed neurologic conditions, as shown on the accompanying slide, include entrapment neuropathies, neuropathies, radiculopathies, spinal stenosis, and other neurologic diagnoses that may mimic PPS. Entrapment syndromes are particularly common in post-polio patients and include entrapment of the median nerve at the wrist (carpal tunnel syndrome), entrapment of the ulnar nerve of the hand (Guyon's canal) or elbow, brachial plexus palsies, entrapment of the peroneal nerve at the fibular head, lateral femoral cutaneous nerve syndromes, and tarsal tunnel syndromes.

Evaluation tests suggested to rule out the above conditions include EMG and nerve conduction studies, radiographic and other imaging studies, blood chemistries and creatine kinase (CK) analysis, and toxic metals screens.

Weakness in PPS: New Superimposed Neurologic Conditions

Differential Diagnosis

Evaluation Tests

Entrapment neuropathies

Neuropathies

Radiculopathies

Spinal stenosis

Other neurologic diagnoses

Electrodiagnosis

Radiographs

Imaging studies

Blood chemistries and CK

Toxic metals screen

III. Evaluation and Differential Diagnosis of PPS

Slide 27.

Other Neurologic Diagnoses Mimicking PPS

Numerous neurologic conditions may produce weakness that mimics that associated with PPS. These include acute inflammatory demyelinating polyneuropathy (ie, “French polio,” or Guillain-Barre syndrome), adult spinal muscular atrophy, late-onset genetic dystrophies or myopathies, inflammatory myopathies, cerebral palsy, amyotrophic lateral sclerosis, myasthenia gravis, multiple sclerosis, Parkinson’s disease, diabetic amyotrophy, spinal cord tumors or infarctions, and cauda equina syndromes.

Other Neurologic Diagnoses Mimicking PPS

- Previous acute inflammatory demyelinating polyneuropathy (“French polio”/Guillain-Barré syndrome)
- Adult spinal muscular atrophy
- Genetic muscular dystrophies/myopathies
- Inflammatory myopathies
- Cerebral palsy
- Amyotrophic lateral sclerosis
- Myasthenia gravis
- Multiple sclerosis
- Parkinson’s disease
- Diabetic amyotrophy
- Spinal cord tumors/infarctions
- Cauda equina syndromes

III. Evaluation and Differential Diagnosis of PPS

Slide 28.

Weakness in PPS: Disuse Atrophy

Components of the differential diagnosis of disuse atrophy are shown on the accompanying slide. Post-polio patients may have increased weakness because of disuse associated with injury and/or treatment of injury (eg, casting). They may also experience new weakness because of incomplete recovery from injury-induced disuse weakness. Similarly, the drastic limitations of activity that may be associated with such medical conditions as myocardial infarction (MI), acute pneumonia, or surgery may result in deficits in strength that are not recovered. Evaluation tests should include isometric strength testing and trials of strengthening exercises to determine whether patients possess strength reserves that have become dormant because of disuse.

Weakness in PPS: Disuse Atrophy

Differential Diagnosis

Evaluation Tests

Injuries (eg, fractures, strains)

Serious medical conditions (eg, MI)

Surgery (eg, hysterectomy, CABG)

Weight gain

Isometric strength testing
Trials of strengthening exercise

III. Evaluation and Differential Diagnosis of PPS

Slide 29.

Weakness in PPS: Overuse/Chronic Strain and Weakness Associated With Systemic Comorbid Medical Conditions

One element of the differential diagnosis of overuse/chronic strain as a cause of weakness in potential PPS patients is chronic repetitive use of muscles at near maximal strength during activities of daily living. Another cause of increased weakness can be greater mechanical disadvantage of muscles as a result of progressive spinal or joint deformities, including those associated with the breakdown of surgical scoliosis or joint fusions. Perceived increased weakness can also result from greater load demand on muscles caused by weight gain or by disuse atrophy associated with more sedentary lifestyles. Evaluation tests for such conditions include gait evaluation (possibly including dynamic, standing, or bending radiographs) and rehabilitation therapy to correct biomechanical conditions that may be causing weakness.

The fourth major area of differential diagnosis for weakness in potential PPS patients is weakness associated with systemic comorbid medical conditions; these conditions and the recommended evaluation tests are the same as those discussed for the differential diagnosis of fatigue (a thyroid test, blood chemistry, complete blood count [CBC], electrocardiogram [ECG], and chest radiograph).

Weakness in PPS: Overuse/Chronic Strain; Associated With Systemic Comorbid Conditions

Differential Diagnosis

Evaluation Tests

Overuse/chronic strain

**Associated with joint/
spine deformities**

Weight gain

**Lifestyle activity patterns
(eg, work and locomotion)**

**Gait evaluation
Rehab therapy**

***Associated with systemic
comorbid medical conditions***

**Same as for fatigue
due to systemic
metabolic disease**

III. Evaluation and Differential Diagnosis of PPS

Slide 30.

Pain in PPS: Spinal Orthopedic Conditions

The major areas of consideration in the differential diagnosis of pain in potential PPS patients include spinal orthopedic conditions, limb joint conditions, limb musculotendinous conditions, fibromyalgia, and muscle pain.

Spinal orthopedic conditions that may be associated with pain are listed in the accompanying slide. Degenerative disc disease, common in the general and post-polio populations, can contribute greatly to back, buttocks, pelvic, and lower-extremity referred pain. Facet arthritis can occur independently or as an accompaniment to spinal deformity. The distal end of long spinal fusions is a primary site for spondylolisthesis or spinal stenosis resulting from spondylosis. Progression of scoliosis is especially common among postmenopausal women. Pelvic girdle somatic dysfunction may arise from asymmetrical pelvis formation during childhood polio. Localized myofascial pain is a common consequence of chronic strain of muscles, and unequal leg length can contribute to myofascial problems in the back and pelvis.

Evaluation tests should include supine and upright or other weight-bearing views to capture changes that may occur in weight bearing states. Angles radiographs may be important in scoliotic patients. Other imaging studies are computed tomography (CT) and magnetic resonance imaging (MRI), both of which may include contrast.

Pain in PPS: Spinal Orthopedic Conditions

Differential Diagnosis

- Degenerative disc disease
- Facet arthritis
- Radiculopathies
- Spondylolisthesis
- Spinal stenosis
- Progression of scoliosis
- SI joint arthritis
- Spinal/pelvic somatic dysfunction
- Myofascial pain syndromes
- Chronic strain associated with unequal leg length/gait abnormalities

Evaluation Tests

Radiographs

- upright & supine
- special views

Imaging

- CT
- MRI > ± contrast

III. Evaluation and Differential Diagnosis of PPS

Slide 31.

Pain in PPS: Limb Joint Conditions

Limb joint conditions that may be associated with pain include degenerative arthritis, frequently of the knee and less commonly of the hip; internal derangements, of which ligamentous laxity/hypermobility of the knee is the most common; and traumatic arthritis following fracture or repeated severe strains. Recommended evaluation tests are x-ray and arthroscopy.

Pain in PPS: Limb Joint Conditions

Differential Diagnosis

Evaluation Tests

Degenerative arthritis

Internal derangements
Ligamentous laxity/hypermobility
strain syndromes

Traumatic arthritis

Radiographs
Arthroscopy

III. Evaluation and Differential Diagnosis of PPS

Slide 32.

Pain in PPS: Limb Musculotendinous Conditions

Limb musculotendinous conditions that may be associated with pain include tendinitis, bursitis, enthesitis, entrapment syndrome, and repetitive strain/overuse syndromes. Anesthetic injections are useful to verify the locations of pain. Ultrasound may be useful for diagnosing such conditions as rotator cuff tear. Provocative tests may also help to localize pain. Other evaluation tests include electrodiagnostic studies and therapeutic trials of manual therapy, physical therapy, orthoses, or assistive devices.

Pain in PPS: Limb Musculotendinous Conditions

Differential Diagnosis

Tendinitis

Bursitis

Enthesitis

Entrapment syndromes

Repetitive strain/overuse syndromes

Evaluation Tests

Anesthetic injection

Ultrasound

Provocative tests

Electrodiagnostic tests

Therapeutic trials

III. Evaluation and Differential Diagnosis of PPS

Slide 33.

Pain in PPS: Limb Fibromyalgia and Muscle Pain

Fibromyalgia may be associated with tender points, chronic strain, sleep disturbance, or depression. Evaluation tests include trials of physical therapy or antidepressant therapy (eg, nighttime antidepressant treatment), changes in activities, and psychiatry/psychology referrals.

Muscle pain may be associated with both acute and chronic overuse strain myalgia. Identification of an activity that may have caused such pain should be followed by efforts to relieve the repetitive strain by altering or eliminating that activity. Muscle pain resulting from shortening or contracture of muscles may respond to stretching exercises.

Pain in PPS: Fibromyalgia and Muscle Pain

Differential Diagnosis

Fibromyalgia

Tender points

Chronic strain

Associated with sleep disturbance and/or depression

Muscle pain

Overuse strain myalgia: acute and chronic

Associated with shortening/contracture

Evaluation Tests

Therapeutic trials
Activity changes
Psychiatry/psychology referral

Therapeutic trials

IV. Management of PPS

Slide 34.

Management of Weakness in PPS

Management of weakness in patients with PPS can involve regimens of exercise as well as appropriate use of orthoses and assistive devices.^{1,2} A number of studies have shown that carefully controlled regimens of isometric, isotonic, and/or isokinetic exercises can produce an increase in strength. Similarly, aerobic exercise regimens (bicycle ergometry or treadmill) have been found to improve a number of indices, including aerobic capacity and duration of exercise.³ However, muscular overuse has been associated with apparently irreversible losses in strength; muscular pain with exercise has been found to be a predictive factor for PPS. Exercise regimens therefore need to be carefully managed and based on appropriate patient selection. In some patients, additional exercise, beyond that associated with performance of daily activities, should be avoided. With regard to other aspects of weakness management, weight gain has also been shown to be a predictive factor for PPS. Correct prescription of orthoses has been found to be associated with subjective improvements in ability to walk and walking safety, as well as with decreased pain.⁴ Assistive devices, such as canes, crutches, and wheelchairs, may be useful.

1. Gawne AC, Halstead LS. Post-polio syndrome: pathophysiology and clinical management. *Crit Rev Physical Rehabil Med.* 1995;7:147-188.
2. Trojan DA, Finch L. Management of post-polio syndrome. *NeuroRehabil.* 1997;8:93-105.
3. Agre JC, Rodriguez AA. Muscular function in late polio and the role of exercise in post-polio patients. *NeuroRehabil.* 1997;8:107-118.
4. Perry J, Clark D. Biomechanical abnormalities of post-polio patients and the implications for orthotic management. *NeuroRehabil.* 1997;8:119-138.

Management of Weakness in PPS

- **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**

Gawne AC, Halstead LS. *Crit Rev Physical Rehabil Med.* 1995;7:147-188; Trojan DA, Finch L. *NeuroRehabil.* 1997;8:93-105.

IV. Management of PPS

Slide 35.

Management of Pain in PPS: Muscular Pain, Muscular Cramps, and Muscular Pain With Activity

Post-polio Muscular Pain usually occurs in muscles previously affected by polio. Cramps typically occur at the end of the day or after a period of activity and are usually a sign of overuse or activity that should be avoided. Management of muscular pain involves pacing during activities; use of moist heat, ice, stretching, and assistive devices; and lifestyle modifications.^{1,2}

1. Gawne AC, Halstead LS. Post-polio syndrome: pathophysiology and clinical management. *Crit Rev Physical Rehabil Med.* 1995;7:147-188.
2. Trojan DA, Finch L. Management of post-polio syndrome. *NeuroRehabil.* 1997;8:93-105.

Management of Pain in PPS: Muscular Pain, Muscular Cramps, and Muscular Pain With Activity

- Pacing
- Moist heat, ice, and stretching
- Use of assistive devices
- Lifestyle modifications

Gawne AC, Halstead LS. *Crit Rev Physical Rehabil Med.* 1995;7:147-188; Trojan DA, Finch L. *NeuroRehabilitation.* 1997;8:93-105.

IV. Management of PPS

Slide 36.

Management of Pain in PPS: Fibromyalgia

Fibromyalgia, a musculoskeletal disorder characterized by generalized pain and tenderness, occurs fairly frequently in patients with previous polio. In 1 study of 105 patients presenting to a post-polio clinic, fibromyalgia was observed in 10.5% of this population, and an additional 10.5% qualified or “borderline fibromyalgia.”¹ It can be treated with amitriptyline (a tricyclic antidepressant),² cyclobenzaprine (a tricyclic amine closely related to amitriptyline and other tricyclic antidepressants),³ fluoxetine (a serotonin reuptake inhibitor),⁴ aerobic exercise,⁵ and other measures such as relaxation techniques, heat, massage, and injection of local anesthetics.⁶

1. McCain GA et al. A controlled study of the effects of a supervised cardiovascular fitness training program on the manifestations of primary fibromyalgia. *Arthritis Rheum.* 1988;31:1135-1141.
2. Goldenberg DL. Treatment of fibromyalgia syndrome. *Rheum Dis Clin North Am.* 1989;15:61-71.
3. Bennett RM et al. A comparison of cyclobenzaprine and placebo in the management of fibrositis: a doubled-blinded controlled study. *Arthritis Rheum.* 1988;31:1535-1542
4. Trojan DA, Cashman NR. Fibromyalgia is common in a post-polio clinic. *Arch Neurol.* 1995;52:620-624.
5. Goldenberg DL et al. A randomized, double-blind crossover trial of fluoxetine and amitriptyline in the treatment of fibromyalgia. *Arthritis Rheum.* 1996;39:1852-1859.
6. Goldenberg DL et al. A randomized, controlled trial of amitriptyline and naproxen in the treatment of patients with fibromyalgia. *Arthritis Rheum.* 1986;29:1371-1377.

Management of Pain in PPS: Fibromyalgia

- Pharmacologic treatments
 - Amitriptyline
 - Cyclobenzaprine
 - Fluoxetine
- Aerobic exercise
- Relaxation techniques, heat, massage, injection of local anesthetics

Bennett RM et al. *Arthritis Rheum.* 1988;31:1535-1542; Goldenberg DL et al. *Arthritis Rheum.* 1986;29:1371-1377; Goldenberg DL. *Rheum Dis Clin North Am.* 1989;15:61-71; Goldenberg D et al. *Arthritis Rheum.* 1996;39:1852-1859; McCain GA et al. *Arthritis Rheum.* 1988;31:1135-1141; Trojan DA, Cashman NR. *Arch Neurol.* 1995;52:620-624.

IV. Management of PPS

Slide 38.

Management of Pain in PPS: Treatment of Superimposed Neurologic Disorders (Some Examples)

Superimposed neurologic disorders that can cause pain include peripheral neuropathies (such as carpal tunnel syndrome), radiculopathies, and spinal stenosis.¹⁻⁴ Carpal tunnel syndrome can be treated with splints,³ pads on canes and crutches, special grips on canes and crutches that place the wrist in a more natural position and increase the weight-bearing surface of the hand, and carpal tunnel releases.^{2,5} Lumbosacral radiculopathies or low-back pain can be treated with anti-inflammatory medications, lumbosacral corsets, shoe lifts, back or pelvic supports, physical therapy, and surgery.^{2,5} Spinal stenosis can be treated with exercise, use of a cane or lumbosacral orthoses, transcutaneous electrical nerve stimulation (TENS), and surgery.^{2,5}

1. Gawne AC et al. Electrodiagnostic findings in 108 consecutive patients referred to a post-polio clinic. *Ann NY Acad Sci.* 1995;753:383-385.
2. Gawne AC, Halstead LS. Post-polio syndrome: pathophysiology and clinical management. *Crit Rev Physical Rehabil Med.* 1995;7:147-188.
3. Kruger VL et al. Carpal tunnel syndrome: objective measures and splint use. *Arch Phys Med Rehabil.* 1991;72:517-520.
4. Werner RA et al. Risk factors for median mononeuropathy of the wrist in post-poliomyelitis patients. *Arch Phys Med Rehabil.* 1989;70:464-467.
5. Trojan DA, Finch L. Management of post-polio syndrome. *NeuroRehabil.* 1997;8:93-105.

Management of Pain in PPS: Treatment of Superimposed Neurologic Disorders (Some Examples)

- **Carpal tunnel syndrome** (splints, pads, and special hand grips on canes or crutches, carpal tunnel release)
- **Radiculopathy** (anti-inflammatory medications, lumbosacral corsets, shoe lifts, back or pelvic supports, physical therapy, surgery)
- **Spinal stenosis** (exercise, cane, lumbosacral orthosis, TENS, surgery)

Gawne AC et al. *Ann NY Acad Sci.* 1995;753:383-385; Gawne AC, Halstead LS. *Crit Rev Physical Rehabil Med.* 1995;7:147-188; Kruger VL et al. *Arch Phys Med Rehabil.* 1991;72:517-520; Trojan DA, Finch L. *NeuroRehabil.* 1997;8:93-105; Werner RA et al. *Arch Phys Med Rehabil.* 1989;70:464-467.

IV. Management of PPS

Slide 39.

Management of Pulmonary Dysfunction in PPS

In the management of pulmonary dysfunction, it is important to first treat any reversible contributing causes. All patients with respiratory dysfunction should receive a pneumococcal vaccine once and should receive influenza vaccinations yearly.

Ventilatory assistance may be necessary for those with sleep-disordered breathing or hypoventilation; noninvasive methods are preferred because of better tolerance by patients and caregivers and lower complication rates.^{1,2} In addition, it is important to identify and treat sleep apnea because it is a potentially life-threatening condition.¹⁻³ Continuous positive airway pressure (CPAP) is frequently used for treatment.

Finally, glossopharyngeal breathing – a method of projecting a bolus of air into the lungs by using the tongue and pharyngeal muscles – can be taught to patients. This technique can allow ventilator-dependent patients to breathe for a few hours without a ventilator.^{1,2}

1. Bach JR, Tilton M. Pulmonary dysfunction and its management in post-polio patients. *NeuroRehabil.* 1997;8:139-153.
2. Gawne AC, Halstead LS. Post-polio syndrome: pathophysiology and clinical management. *Crit Rev Physical Rehabil Med.* 1995;7:147-188.
3. Steljes DG et al. Sleep in post-polio syndrome. *Chest.* 1990;98:133-140.

Management of Pulmonary Dysfunction in PPS

- Preventive measures (pneumococcal vaccine, yearly influenza vaccine)
- Ventilatory assistance (noninvasive methods preferred)
- Identification and treatment of sleep apnea
- Glossopharyngeal breathing

Bach JR, Tilton M. *NeuroRehabil.* 1997;8:139-153; Gawne AC, Halstead LS. *Crit Rev Phys Rehabil Med.* 1995;7:147-188; Steljes DG et al. *Chest.* 1990;98:133-140.

IV. Management of PPS

Slide 40.

Management of Dysphagia in PPS

Swallowing dysfunction has been observed in 10% to 20% of selected samples of post-polio patients. It can occur in patients with or without bulbar polio.¹ Some patients without clinical symptoms have been found to exhibit videofluoroscopic abnormalities, which may progress with time. Management of dysphagia may include change or restriction of diet, use of special breathing and/or swallowing techniques, monitoring of fatigue by taking larger meals earlier and smaller meals later in the day, and avoidance of eating when fatigued.^{2,3}

1. Bucholz DW, Jones B. Post-polio dysphagia: alarm or caution? *Orthopedics*. 1991;14:1303-1305.
2. Gawne AC, Halstead LS. Post-polio syndrome: pathophysiology and clinical management. *Crit Rev Physical Rehabil Med*. 1995;7:147-188.
3. Trojan DA, Finch L. Management of post-polio syndrome. *NeuroRehabil*. 1997;8:93-105.

Management of Dysphagia in PPS

- Change or restrict diet
- Use special breathing techniques
- Use special swallowing techniques
- Monitor fatigue; take larger meals earlier, smaller meals later
- Avoid eating when fatigued

Gawne AC, Halstead LS. *Crit Rev Physical Rehabil Med*. 1995;7:147-188; Trojan DA, Finch L. *NeuroRehabil*. 1997;8:93-105.

IV. Management of PPS

Slide 41.

Symptomatic Fatigue in PPS

Fatigue in patients with PPS may be associated with a low energy state attributable to poor sleep or mood disturbance, mental fatigue, reduced muscular endurance, or delayed recovery after exercise.¹ Factors contributing to fatigue may include depressive illness or symptoms, deconditioning, overuse of muscles or muscle groups, disturbed sleep patterns, and muscle fiber transformation. With regard to the latter, it has been suggested that muscles fibers in patients with PPS may undergo changes that render them increasingly fatigable.²

1. Sharma KR et al. Excessive muscular fatigue in the post-polio syndrome. *Neurology*. 1994;44:642-646.
2. Borg K et al. Motor neuron firing and isomyosin type of muscle fibers in prior polio. *J Neurol Neurosurg Psychiatry*. 1989;52:1141-1148.

Symptomatic Fatigue in PPS

- Low energy state/mood disturbance
- Mental fatigue
- Reduced muscular endurance
- Delayed recovery after exercise
- Contributing factors:
 - Depression
 - Deconditioning or overuse
 - Disturbed sleep
 - Muscle fiber transformation

IV. Management of PPS

Slide 42.

Management of Fatigue in PPS

Management of fatigue in patients with PPS may involve lifestyle changes relating to energy conservation and pacing, use of antidepressant medications, normalization of sleep patterns, and reconditioning exercises.¹ In addition, a number of pharmacologic approaches for treatment are under investigation.²

1. Bach JR, Alba AS. Pulmonary dysfunction and sleep disordered breathing as post-polio sequelae: evaluation and management. *Orthopedics*. 1991;14:1329-1337.
2. Trojan DA, Cashman NR. *Current Trends in Post-Poliomyelitis Syndrome*. New York, NY: Milestone Medical Communications; 1996.

Management of Fatigue in PPS

- Energy conservation/pacing
- Antidepressants
- Normalization of sleep patterns
- Reconditioning exercises

IV. Management of PPS

Slide 43.

Disturbed Sleep in PPS

It has become increasingly apparent that many patients with PPS have disturbed sleep patterns, with symptoms of daytime sleepiness, snoring/frequent waking, morning headache, and cognitive impairment during the day.¹ Management of such disturbances should include evaluation by nocturnal oximetry and polysomnogram. Initial treatment may involve a trial of amitriptyline. Those with documented major sleep disturbances can benefit greatly from continuous or bilevel positive airway treatment (CPAP, Bi-PAP).²

1. Steljes DG et al. Sleep in post-polio syndrome. *Chest*. 1990;98:133-140.
2. Bach JR, Alba AS. Pulmonary dysfunction and sleep disordered breathing as post-polio sequelae: evaluation and management. *Orthopedics*. 1991;14:1329-1337.

Disturbed Sleep in PPS

- Daytime sleepiness
- Snoring, frequent waking
- Morning headache
- Cognitive impairment
- Management strategies:
 - Nocturnal oximetry
 - Polysomnogram
 - Amitriptyline
 - CPAP, Bi-PAP

IV. Management of PPS

Slide 44 and 45.

Prescribing Exercise in PPS

Prescription of exercise in PPS patients should be individualized on the basis of a number of factors. History and grading of strength should be used to determine when, and for which muscle groups deconditioning (as opposed to overuse) is responsible for symptoms. Deconditioned muscles or muscle groups are likely to benefit from exercise regimens.¹⁻⁵ Caution is needed in prescribing exercise for muscles with strength of antigravity, or less, because such muscles may be at greater risk for overuse. Aerobic exercise can help to prevent weight gain but should be prescribed with the same considerations used when prescribing strengthening exercises. Symptoms of joint/soft tissue irritation should be considered in prescribing any therapeutic exercise program.

In instituting exercise regimens, it is important to observe the maxim of starting low and going slow. Graduated exercise appears to be much better tolerated and is less likely to result in overwork weakness. Rests between brief exercise periods and separation of exercise days by rest days are advisable. Exercise that produces fatigue is by definition too strenuous, as is that which produces muscular pain. For these reasons, it is important to monitor exercise regimens, at least in the initial stages.

A sample protocol for exercise in patients with PPS includes a warm-up period and a stretching period to avoid muscle pain, followed by strengthening exercises, gentle aerobic exercises, and a stretching and cool-down period.¹⁻⁵

1. Agre JC, Rodriguez M. Intermittent isometric activity: its effect on muscle fatigue in post-polio subjects. *Arch Phys Med Rehab.* 1991;72:971-975.
2. Dean E, Ross J. Effect of modified aerobic training on movement energetics in polio survivors. *Orthopedics.* 1991;14:1243-1246.
3. Jones DR et al. Cardiorespiratory responses to aerobic training by patients with postpoliomyelitis sequelae. *JAMA.* 1989;261:3255-3258.
4. Kriz JL et al. Cardiorespiratory responses to upper extremity aerobic training by post-polio subjects. *Arch Phys Med Rehab.* 1992;73:49-54
5. Owen RR, Jones DR. Polio residuals clinic: conditioning exercise program. *Orthopedics.* 1985;8:882-883.

Prescribing Exercise in PPS

- Reverse deconditioning
- Avoid overuse
- Muscles >MRC 3
- Prevent weight gain
- Avoid joint/soft tissue irritation
- Start low, go slow
- Take rest breaks (mins/days)

Prescribing Exercise in PPS (cont'd.)

- Avoid excessive fatigue and muscular pain
- Coach, monitor
- Warm up
- Stretch
- Strengthening
- Gentle aerobics
- Stretch, cool down

IV. Management of PPS

Slide 46 and 47.

Therapeutic Trials in PPS

Several clinical trials of pharmacological treatments for PPS have been completed.¹ Amantadine, an antiviral agent used to treat influenza and fatigue associated with neurologic illnesses, has been evaluated for PPS-related fatigue. In one study, 25 PPS patients with fatigue were randomly selected to receive 100 mg amantadine or placebo for 6 weeks; no association was found between amantadine and clinical response to fatigue.²

The effects of high-dose prednisone – a powerful anti-inflammatory corticosteroid used to treat rheumatoid arthritis and other inflammatory disorders – were assessed in a study of 17 PPS patients with new muscle weakness. No significant differences were found between groups in terms of muscular strength or subjective fatigue.³

Human growth hormone (Humatrope) has been found to increase the production of a hormone in the body – insulin-like growth factor 1 (IGF-1) – that stimulates nerve cells to sprout additional extensions (axons). This drug, which is already approved for other uses, was tested in 6 patients with PPS and produced little or no improvement in their muscle strength, endurance, or recovery after fatigue.⁴

A placebo-controlled trial of recombinant human IGF-1 (rhIGF-1) in 22 patients with PPS has recently been completed. Subcutaneous rhIGF-1 enhanced recovery after fatiguing exercises in patients with PPS but did not alleviate excessive exercise-induced fatigability or weakness.⁵

Selegiline (Deprenyl), a neuroprotective agent primarily used for the treatment of Parkinson’s disease, may warrant further study on the basis of initial results from case reports. Two cases showed that selegiline produced an improvement in PPS symptoms; this improvement ended after the drug was discontinued.⁶

Bromocriptine mesylate (Parlodel), a dopamine receptor agonist used to treat conditions such as Parkinson’s disease, was evaluated in 5 patients with PPS and produced improvements in fatigue symptoms in 3 of the participants.⁷

Therapeutic Trials in PPS

Drug	Category	Type of Trial	N	Results
Amantadine	Antiviral	Randomized, placebo-controlled trial (Stein et al)	25	No significant improvement in fatigue
Prednisone (high-dose)	Steroid, anti-inflammatory	Randomized, placebo-controlled trial (Dinsmore et al)	17	No significant improvement in strength or fatigue
Human growth hormone	Hormone	Open trial (Gupta et al)	6	Little or no improvement in muscle strength
Insulin-like growth factor-1 (IGF-1)	Growth factor	Randomized, placebo-controlled trial (Miller et al)	22	Improvement in recovery after exercise; no change in strength, fatigability

Stein DP et al. *Ann NY Acad Sci.* 1995;753:296-302; Dinsmore S et al. *Ann NY Acad Sci.* 1995;753:303-313; Gupta KL et al. *Arch Phys Med Rehabil.* 1994;75:889-894; Miller RG et al. *Neurology.* 1997;48:A217.

IV. Management of PPS

Slide 46 and 47 (cont'd).

Therapeutic Trials in PPS

The anticholinesterase pyridostigmine (Mestinon), which is already approved for treatment of myasthenia gravis, has demonstrated some promise in relieving the symptoms of weakness and fatigue in patients with PPS. In an open trial of 17 patients taking a daily dose of 180 mg pyridostigmine, a significant relationship was found between subjective fatigue response to pyridostigmine and improvement in neuromuscular junction transmission after administration of edrophonium (a short-acting anticholinesterase similar to pyridostigmine).⁸ In another open trial with 27 patients, 59% who received pyridostigmine reported an improvement in fatigue with the medication and requested continuation of treatment.⁹ In addition, a double-blind, placebo-controlled, crossover trial demonstrated improvements in some objective measures of strength and subjective measures of fatigue when pyridostigmine, 180 mg/day, was given to 27 patients.¹⁰

Adverse events, including loose stool and intestinal cramps, blurred vision, increased urinary frequency, muscle cramps, and fasciculations or muscle twitches, were seen in approximately 50% of patients participating in these 3 trials.

Therapeutic Trials in PPS (cont'd.)

Drug	Category	Type of Trial	N	Results
Selegiline	Neuroprotective agent	Case studies (Bamford et al)	2	Improvement in PPS symptoms
Bromocriptine	Dopamine receptor agonist	Placebo-controlled, crossover trial (Bruno et al)	5	Improvement in fatigue symptoms in 3 patients
Pyridostigmine	Anticholinesterase	Open trials (Trojan et al [1993,1995])	17, 27	Improvement in fatigue
		Placebo-controlled, crossover trial (Seizert et al)	27	Improvement in fatigue, strength
		Multicenter, randomized, placebo-controlled trial (Trojan et al, 1997)	126	No significant improvement in health-related quality of life, fatigue, isometric strength, but trend to increased strength in very weak muscles

Bamford CR et al. *Int J Neurosci.* 1993;71:183-188; Bruno RL et al. *Am J Phys Med Rehabil.* 1996;75:340-347; Trojan DA et al. *J Neurol Sci.* 1993;114:170-177; Trojan DA, Cashman NR. *Can J Neurol Sci.* 1995;22:223-227; Seizert BP et al. *Arch Phys Med Rehabil.* 1994;75:1049; Trojan DA et al. *Arch Phys Med Rehabil.* 1997;78:1027.

The ongoing North American Postpoliomyelitis Pyridostigmine Study (NAPPS) is a multicenter, randomized, double-blind, placebo-controlled phase II trial involving 126 patients with PPS. The study is designed to determine the efficacy of pyridostigmine, 180 mg/day, in relieving PPS symptoms using measures of fatigue, muscle strength, and health-related quality of life. NAPPS was also designed to determine the incidence of severity of any side effects associated with pyridostigmine. The study showed no difference between patients who received placebo with regard to health-related quality of life, fatigue, and most measures of isolated strength. However, a trend to increased strength in very weak muscles was seen at 6 months with pyridostigmine. The result of no significant difference in outcome measures between patient groups did not correlate with the investigators' clinical impressions that the medication appeared to be beneficial in a proportion of patients with PPS.¹¹

1. Trojan DA, Finch L. Management of post-polio syndrome. *NeuroRehabil.* 1997;8:93-105.
2. Stein DP, Dambrosia J, Dalakas MC. A double-blind, placebo-controlled trial of amantadine for the treatment of fatigue in patients with post-polio syndrome. *Ann NY Acad Sci.* 1995;753:296-302.

IV. Management of PPS

Slide 46 and 47 (cont'd). Therapeutic Trials in PPS

3. Dinsmore S, Dambrosia J, Dalakas MC. A double-blind, placebo-controlled trial of high-dose prednisone for the treatment of post-polio syndrome. *Ann NY Acad Sci.* 1995;753:303-313.
4. Gupta KL et al. Human growth hormone effect on serum IGF-1 and muscle function in poliomyelitis survivors. *Arch Phys Med Rehabil.* 1994;75:889-894.
5. Miller RG, Gelinus DF, Kent-Braun J. The effect of recombinant insulin-like growth factor 1 (rhIGF-1) upon exercised-induced fatigue and recovery in patients with post-polio syndrome. *Neurology.* 1997;48:A217. Abstract S29.005.
6. Bamford CR, Montgomery EB Jr, Munoz JE, Stumpf C, Pry S. Post-polio syndrome: response to deprenyl (selegiline). *Int J Neurosci.* 1993;71:183-188.
7. Bruno RL et al. Bromocriptine in the treatment of post-polio fatigue. *Am J Phys Med Rehabil.* 1996;75:340-347.
8. Trojan DA, Gendron D, Cashman NR. Anti-cholinesterase-responsive neuromuscular junction transmission defects in post-poliomyelitis fatigue. *J Neurol Sci.* 1993;114:170-177.
9. Trojan DA, Cashman NR. An open trial of pyridostigmine in post-poliomyelitis syndrome. *Can J Neurol Sci.* 1995;22:223-227.
10. Seizert BP, Speier JL, Canine JK. Pyridostigmine effect on strength endurance, and fatigue in post-polio patients. *Arch Phys Med Rehabil.* 1994;75:1049. Abstract.
11. Trojan DA et al. The response of post-poliomyelitis syndrome symptoms to pyridostigmine (Mestinon): a multi-centered, placebo-controlled trial. *Arch Phys Med Rehabil.* 1997;78:1027. Abstract.

V. Optimizing Wellness: The Role of the Patient in PPS

Slide 48.

Patient Assessments of 15 Treatments for PPS

Supporting the notion of self-care can help promote wellness in patients with PPS. It is considered to be of paramount importance that physicians form a partnership with patients in assessing PPS symptoms and in making decisions on how to manage these symptoms.

An Australian study¹ of 176 people with PPS has provided some indication of how patients respond to PPS symptoms. The survey concluded that a coping style that focuses on symptoms and attempts to maintain previous activity levels was associated with increased feelings of helplessness, depression, and anger. The accompanying slide shows the top 6 tried, and top 6 rated as very helpful among 15 prescribed or self-selected treatments for PPS symptoms.

1. Westbrook M, McIlwain D. Living with the late effects of disability: a five year follow-up survey of coping among post-polio survivors. *Aust Occup Ther J.* 1996;43:60-71.

Patient Assessments of 15 Treatments for PPS

Top 6 Tried

Drugs for pain relief (45%)
Swimming (31%)
Exercise program (31%)
Massage (29%)
Vitamins (29%)
Hot baths (28%)

Top 6 Rated as Very Helpful

Massage (67%)
Hot baths (59%)
Special shoes (55%)
Swimming (47%)
Hydrotherapy (43%)
Counseling (36%)

Westbrook M, McIlwain D. *Aust Occup Ther J.* 1996;43:60-71.

V. Optimizing Wellness: The Role of the Patient in PPS

Slide 49.

When Medical Options Are Not Enough

Conventional therapy (surgery, pharmaceuticals, assistive devices) is not always adequate for reducing symptoms and/or increasing quality of life. According to a 1990 survey in the *New England Journal of Medicine*,¹ an estimated 61 million Americans made 425 million visits to alternative medicine practitioners in 1990 for treatment of symptoms such as stress, pain, fatigue, and psychological distress. This can be compared with 388 million visits to conventional primary-care physicians. The type of alternative therapy reported in this survey were, in alphabetical order, acupuncture, biofeedback, chiropractic services, commercial weight loss programs, energy healing, herbal medicine, homeopathy, hypnosis, imagery, lifestyle diets including macrobiotics, massage, megavitamin therapy, self-help groups, spiritual healing, and relaxation therapies.

1. Eisenberg DM, Kessler RC, Foster C, Norlock FE, Calkins DR, Delbanco TL. Unconventional medicine in the United States: prevalence, costs, and patterns of use. *N Engl J Med.* 1993;328:246-252.

When Medical Options Are Not Enough

- **61 million Americans made 425 million visits to alternative medicine practitioners in 1990 (compared with 388 million visits to conventional primary-care physicians)**
- **Symptoms have included stress, pain, fatigue, and psychological distress**
- **72% did not tell their physicians about these visits**

Eisenberg DM et al. *N Engl J Med.* 1993;328:246-252.

V. Optimizing Wellness: The Role of the Patient in PPS

Slide 50.

Complementary Therapies

Elliott Dacher, a practicing physician, has noted that the emerging public interest in health promotion, self-care, alternative healing practices, and mind/body medicine has developed in response to the limitations of conventional medical practices.¹ According to the expanded medical model proposed by Dacher, the relationship of the health practitioner to the patient can become more of a partnership if the treatment plan has a long-term focus and includes health promotion practices whose orientation is more internal, rather than external. This expanded medical model also extends the physician's vision to include the mind/body healing system.

Although the terms "alternative" and "complementary" are often used interchangeably in the context of medical treatments, the latter term more accurately characterizes the value of many nontraditional therapies. When a patient is said to have chosen an alternative treatment, this implies that another treatment has been excluded; however, when a patient is said to have chosen a complementary treatment, this indicated that multiple therapies – such as mind/body healing in conjunction with a conventional pharmacologic treatment – are being used to obtain more complete, or "holistic," medical care.

1. Dacher E. A systems theory approach to an expanded medical model: a challenge for biomedicine. *J Alternative Complementary Med.* 1995;1:187-196.

Complementary Therapies

- **The relationship between physician and patient becomes more of a partnership when the treatment plan includes the mind/body healing system and health promotion practices**
- **"Alternative" implies that one option has been chosen over another, whereas "complementary" indicates that an option has been supplemented to make it more complete**
- **Complementary therapies can be used to augment medical practices**

Dacher E. *J Altern Complementary Med.* 1995;1:187-196.

V. Optimizing Wellness: The Role of the Patient in PPS

Slide 51.

Relaxation Studies

Elicitation of a relaxation response is a self-care technique with numerous benefits. Relaxation techniques have been shown not only to decrease oxygen consumption (by 10% to 17%), respiration rate, heart rate, blood pressure, and muscle tension, but also to stabilize blood flow to arm and leg muscles.¹ In addition, research relevant to polio survivors has shown that patients with chronic pain can reduce the severity of pain, anxiety, depression, and anger through the use of relaxation techniques. These patients also exhibited increased levels of activity and paid 36% fewer visits to managed care facilities 2 years after beginning the program.²

1. Benson H, Stark M. *Timeless Healing: The Power and Biology of Belief*. New York: Scribner, 1996.
2. Caudill MA, Schnable R, Zuttermeister P, Benson H, Friedman R. Decreased clinic use by chronic pain patients: response to behavioral medicine intervention. *Clin J Pain*. 1991;17:305-310.

Relaxation Studies

Benefits of the relaxation response:

- **Decreased oxygen consumption, respiration rate, heart rate, blood pressure, muscle tension**
- **Stabilized blood flow to arm and leg muscles**

Chronic pain program teaching relaxation techniques resulted in:

- **Decreased severity of pain, anxiety, depression, anger; increased activity**
- **36% fewer visits to managed care facilities (2 years after program)**

Benson H, Stark M. *Timeless Healing: The Power and Biology of Belief*. New York: Scribner, 1996;
Caudill MA et al. *Clin J Pain*. 1991;17:305-310.

V. Optimizing Wellness: The Role of the Patient in PPS

Slide 52.

Spirituality Related to Health

According to Dacher, a spiritual perspective can have a profound influence on personal attitudes, values, and behaviors, which in turn can substantially affect biochemical and physiological factors.¹ Moreover, a 1991 study by Woods Smith² found that polio survivors manifested a greater level of spirituality than people who had not experienced polio or a similar life event – 91% of polio survivors reported that they had a religious affiliation, 82% reported that they prayed regularly, and 18% reported meditating at least once a week.

1. Dacher E. A systems theory approach to an expanded medical model: a challenge for biomedicine. *J Alternative Complementary Med.* 1995;1:187-196.
2. Woods Smith D. Power and spirituality in polio survivors. *Nurs Sci Q.* 1995;8:133-139.

Spirituality Related to Health

- **“Spirituality has profound effect on personal attitudes, values, and behaviors, and...on biochemistry and physiology” (Dacher, 1995)**
- **91% of polio survivors in a national study reported religious affiliations; 82% said that they prayed regularly (Woods Smith, 1995)**

Dacher E. *J Altern Complementary Med.* 1995;1:187–196; Woods Smith D. *Nurs Sci Q.* 1995;8:133–139.

V. Optimizing Wellness: The Role of the Patient in PPS

Slide 53.

Interpersonal Strategies for PPS Management

The accompanying slide, adapted from a 1993 study by Westbrook et al,¹ shows the top 6 interpersonal strategies tried for relief of PPS symptoms and the top 6 rated as very helpful. Some lifestyle changes were rated as being more helpful than treatment or interpersonal strategies. Employing household help, buying special equipment and furniture, and modifying the home were rated as being very helpful by at least 70% of respondents. Personal strategies of becoming more involved in interests that one is still able to pursue, developing a philosophy of life, and reading more about post-polio were rated as very helpful by at least 65%.

1. Westbrook M, McIlwain D. Living with the late effects of disability: a five year follow-up survey of coping among post-polio survivors. *Aust Occup Ther J.* 1996;43:60-71.

Interpersonal Strategies for PPS Management

Top 6 Tried

Talked with family (75%)

Talked with friends (74%)

Talked with other polio survivors (68%)

Joined a post-polio support group (65%)

Asked family for help (39%)

Asked friends for help (19%)

Top 6 Rated as Very Helpful

Asked friends for help (55%)

Asked family for help (52%)

Joined a post-polio support group (44%)

Talked with other polio survivors (43%)

Talked with family (40%)

Talked with friends (23%)

Westbrook M, McIlwain D. *Aust Occup Ther J.* 1996;43:60-71.

V. Optimizing Wellness: The Role of the Patient in PPS

Slides 54 and 55.

Interpersonal Strategies for PPS Management

To help post-polio patients avoid feelings of powerlessness, hopelessness, and dependence, it is important for physicians to be aware of, and sensitive to, the special needs of people who have had polio and to help those with new symptom onset to become aware of PPS.

Management options should be explored with patients, and decisions should reflect both patient and physician input. Patient involvement in management is crucial to the outcome; thus, the goals of disease management should be highly individualized.

Physician/Patient Partnerships: Physician Awareness

It is important for physicians to be aware of, and sensitive to, the special needs of people who had polio and to help polio survivors become aware of PPS.

Physician/Patient Partnerships: Physician Awareness

- It is important for physicians to make decisions with —not for— polio survivors
- “Life’s best survivors share a key characteristic: they take an active role in responding to personal trauma” (Siebert, 1994)

Siebert A. *The Survivor Personality*. Portland, Ore: Practical Psychology Press, 1994.