Post-Polio Syndrome: Pathophysiology and Clinical Management

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ABSTRACT: Post-polio syndrome (PPS) is a progressive neuromuscular syndrome characterized by symptoms of weakness, fatigue, pain in muscles and joints, and breathing and swallowing difficulties. Survivors of poliomyelitis experience it many years after their initial infection. Although the etiology for these symptoms is unclear, it may be due to motor unit dysfunction manifested by deterioration of the peripheral axons and neuromuscular junction, probably as result of overwork. An estimated 60% of the over 640,000 paralytic polio survivors in the U.S. may suffer from the late effects of polio. Their physical and functional rehabilitation care presents a challenge for practitioners in all disciplines. To evaluate these symptoms, a comprehensive assessment must be done, as frequently PPS is a diagnosis of exclusion. Care of the patient with PPS is best carried out by an interdisciplinary team of rehabilitation specialists. This article reviews the epidemiology, pathophysiology, characteristics, assessment, and rehabilitation care of the patient with PPS.

KEY WORDS: poliomyelitis, post-polio syndrome, weakness, fatigue, exercise, pain therapy, respiratory complications.

I. INTRODUCTION.

This article is a comprehensive review of postpolio syndrome (PPS). Although poliomyelitis epidemics came to a dramatic end in most countries with the introduction of the Salk polio vaccination in 1955, acute polio remains a threat in many parts of the third world. In addition, many of those earlier survivors are now developing new problems and face a new challenge to restore strength and function.

In the past, even in the field of rehabilitation, residual motor loss from paralytic polio was generally considered a chronic, stable lesion. Following the acute illness and a period of rehabilitation, patients eventually reached a plateau of neurological and functional recovery that was believed to remain essentially static.*[1]* However, more recently, research shows that over one half of the survivors of paralytic polio experience new health problems related to their original illness.*[2]* These problems occur about 30 to 50 years after their initial polio and include new weakness, fatigue, pain, and functional loss. The cause of these new symptoms remains uncertain; however, it appears to be related to motor unit dysfunction manifested by a

deterioration of the peripheral axons and the neuromuscular junction.

This review begins with a discussion of the historical and epidemiological aspects of poliomyelitis, as well as the pathophysiology of polio. We examine possible etiologies for the development of PPS. We then review the characteristics of PPS, including physical, electrodiagnostic, and morphologic features. Finally, we discuss the assessment, differential diagnosis, and rehabilitation management of some of the problems that patients experiencing the late effects of polio may face.

II. HISTORICAL BACKGROUND.

For more than 100 years, it has been recognized that new muscle weakness occurs in polio survivors many years after their initial illness. The first descriptions appeared in 1875 when four separate case histories were reported in the French literature by Carriere,*[3]* Raymond,*[4]* and Cornil and Lepine.*[5]* All of these patients were young men who had paralytic polio in infancy. They developed new weakness not only in previously affected muscles but also in muscles believed to be uninvolved. They all had physically demanding jobs and performed repetitive activities. In a commentary on one of the cases, Jean Martin Charcot suggested that a previous disease of the spinal cord may leave an individual more susceptible to a subsequent spinal disorder and that the new weakness was secondary to overuse of the involved limbs.*[4]* In a presentation to the Royal Society of Medicine in 1962, Zilkha reviewed 11 patients with motor neuron disease who developed progressive weakness 20 to 40 years later.*[6]* He stated "It could be suggested that the subsequent development of disease of the motor neuron in those patients with a previous history of poliomyelitis, usually 25 years before, is related to the occurrence of that earlier disease".

Since these initial reports, there have been other sporadic reports of similar phenomenon. In 35 reports of almost 250 cases, authors have described new problems, including weakness and fatigue occurring up to 71 years after the acute polio episode.*[7]* These neurological changes were most commonly diagnosed as a form of progressive muscular atrophy, chronic anterior poliomyelitis, late motor denervation, and forme fruste amyotrophic lateral sclerosis.*[8,9]* However, it was not until the early 1980s, approximately 40 years after the major epidemics of the 1940s and 1950s, that PPS became widely recognized.

As the numbers of persons experiencing these new symptoms increase, this subject has been studied in depth. This review concentrates on some of the more recent research. Although the incidence of acute poliomyelitis has decreased since the introduction of the Salk vaccine in 1955 and the Sabin trivalent oral polio vaccine (TOPV) in 1961, some of the principles learned through the study of PPS can be applied to other similar neurological diseases and aging with a disability.

III. EPIDEMIOLOGY.

During the epidemics in the U.S. from 1952 to 1954, the incidence of new cases was approximately 15/100,000, as Figure 1 demonstrates.*[10]* The incidence peaked in 1952 with over 57,879 new cases reported in the U.S. After the Salk vaccine was introduced in 1955 and the Sabine vaccine in 1961 the incidence dropped to 0.04/100,000 by 1963. The last confirmed case of paralytic polio from domestic wild virus in the U.S. occurred in 1979.*[10]* Paralytic polio is now a rare complication of the current Sabin (oral) vaccination. In the U.S. between 1961 and 1964, there was an incidence of paralysis in 4.9/10 million doses compared with an incidence of 0.23/10 million in 1989, as Figure 2 shows.*[10]* The greatest risk is from the initial immunization (1/700,000).*[11]*

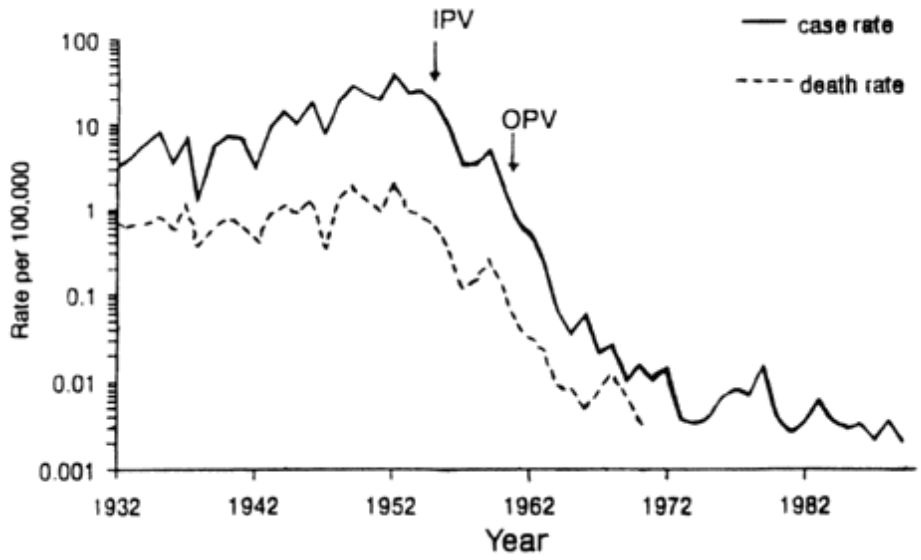

FIGURE 1. Reported rates per 100,000 persons of poliomyelitis and of death from poliomyetitis, United States, 1932 to 1989. (IPV = inactivated polio vaccine; OPV = oral polio vaccine.)

A survey by the National Center for Health Statistics in 1987 found there were more than 640,000 survivors of paralytic polio.*[12]* Thus, despite the virtual elimination of new cases, paralytic polio remains one of the most prevalent neuromuscular diseases in this country. While PPS is more common in those who have experienced moderate to severe paralysis,13 population-based surveys show that as many as 64% of polio survivors develop new symptoms.*[2]* If these samples are representative of a typical distribution, then it can be estimated that over 409,000 people may experience the late effects of polio. While some of this population has died since the original survey, there has also been an unknown and largely unexpected increase in the number of polio survivors in the U.S. resulting from the influx of affected immigrants, refugees, and illegal aliens from Southeast Asia and Latin America. In 1991, the World Health Organization (WHO) estimated that 85% of children world-wide received three doses of the TOPV vaccine, a significant increase over the percentage in 1971 (15%), largely due to the effort WHO put into their vaccination campaign.*[14]* While no cases of wild polio infection occurred in the Americas in 1993, WHO estimates that 96,500 new cases occurred in the developing world; in particular Africa, Asia, and India, due to inadequate immunization.*[15]*

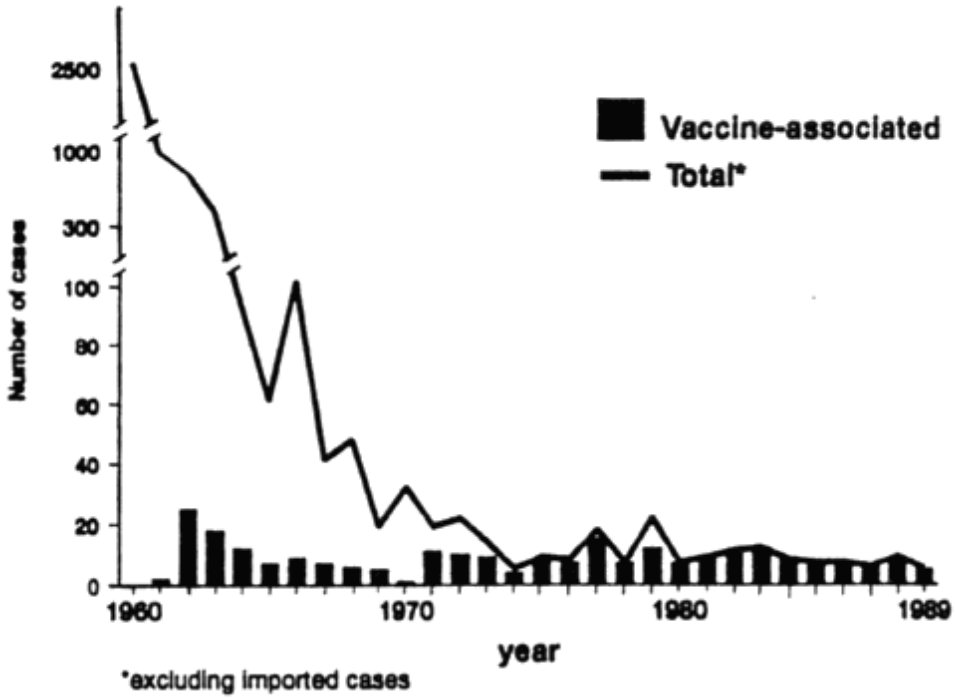

FIGURE 2. Reported cases of paralytic poliomyelitis (total and vaccine-associated per 10 million doses of Sabin vaccine), U.S., 1960 to 1989.

IV. PATHOPHYSIOLOGY IN ACUTE POLIO.

Knowledge of the pathophysiology of acute polio is necessary to understand the possible causes for PPS and to provide a rational basis for its management. The poliovirus is a positive single-stranded RNA enterovirus belonging to the picornavirus group. The virus is relatively small (30 nm in diameter) and the poliovirus genome is about 7400 nucleotides long.*[16]* It lacks a lipid coat or capsule but has a protein coat shaped like a polyhedron with 20 faces.*[17]* There are three polio viruses, numbers 1, 2, and 3, defined by the configuration of the capsid proteins. Therefore, theoretically, a person could be infected more than once.

Wild poliovirus enters the body by oral ingestion, then replicates in the lymphoid tissue of the pharynx and ileum and spreads regionally to lymphoid tissue. It is extremely infectious and usually benign. The vast majority of infected individuals (95 to 99%) remain asymptomatic or experience a self-limited illness characterized by fever, myalgia, and gastrointestinal symptoms.*[18]* However, in 1 to 5% of persons, viremia may follow, with invasion of the anterior horn cells of the central nervous system (CNS). These patients usually develop a headache, stiff neck, and back pain, similar to viral meningitis. Only 1 to 2% of all those infected develop paralysis. Finally, the rate of paralysis varies with the strain of the virus and the patient's age. In children, paralysis occurs in 1/1000 cases, while in adults 1/75 develop paralysis. Asymmetric, flaccid paralysis occurs, with legs more commonly involved than arms. Severe bulbar weakness occurs in 10 to 15% of all paralytic cases. Less frequently, there is ophthalmoplegia and bladder involvement.*[18]* The pathological findings of acute polio consist of inflammation of meninges and anterior horn cells, with loss of spinal and bulbar motor neurons.*[19]* Less prominent findings include abnormalities in the cerebellar nuclei, reticular formation, thalamus, hypothalamus, cortical neurons, and dorsal horn.*[19]*

Once the virus has invaded the CNS, neurological and functional loss occurs as anterior horn cells are lost, and thus the muscle fibers innervated by them are "orphaned". Recovery begins in weeks and reaches a plateau in 6 to 8 months. The extent of neurological and functional recovery is determined by three major factors: (1) the number of motor neurons that recover and resume their normal function, (2) the number of motor neurons that develop terminal axon sprouts to reinnervate muscle fibers left orphaned by the death of

their original motor neurons, and (3) muscle hypertrophy. The phenomenon of terminal axon sprouting makes it possible for an uninvolved or recovered motor neuron to "adopt" these orphaned muscle fibers. Stalberg has shown that a motor neuron cell can adopt five to seven additional muscle fibers commonly and occasionally, as many as 20 for every muscle cell innervated originally.*[20]* A single motor neuron that originally innervated 100 muscle fibers iriight eventually innervate 700 to 2000 fibers. As a result, the survivors of acute polio may be left with a few, significantly enlarged motor units doing the work previously performed by many units.*[21]* Figures 3, 4, 5, and 6 provide a schematic illustration of this phenomenon. Both electrophysiological evidence, including single fiber and macro-EMG and morphological data support this concept.*[22-27]*

In addition to this reinnervation, the remaining muscle fibers hypertrophy to increase the strength of the muscle group.*[25,28]* Because this mechanism of neurophysiological compensation is so effective, a muscle can retain normal strength even after 50% of the original motor neurons have been lost. Therefore, in some patients, manual muscle testing (MMT) may be normal when more than half the original anterior horn cells are destroyed.*[29]*

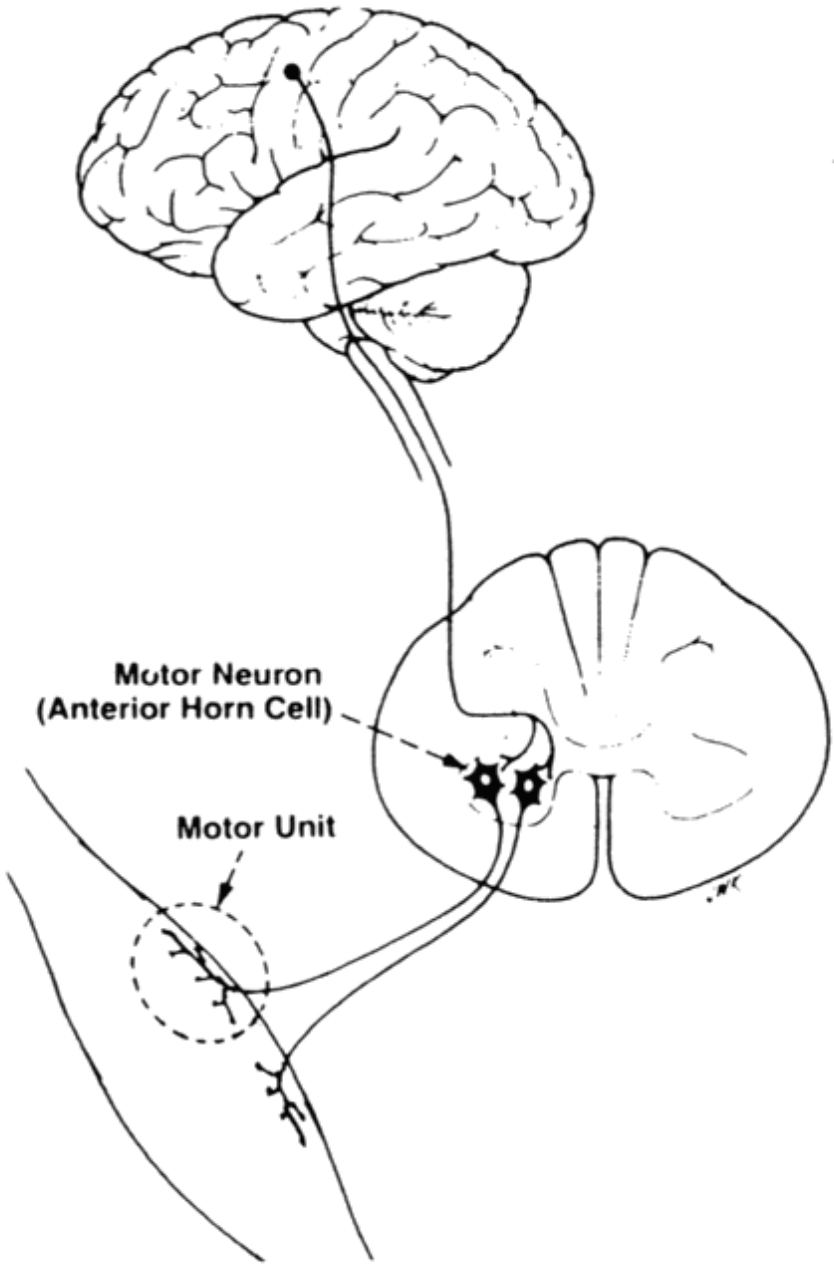

FIGURE 3. A normal motor unit containing the motor neuron and the muscle fibers it innervates.

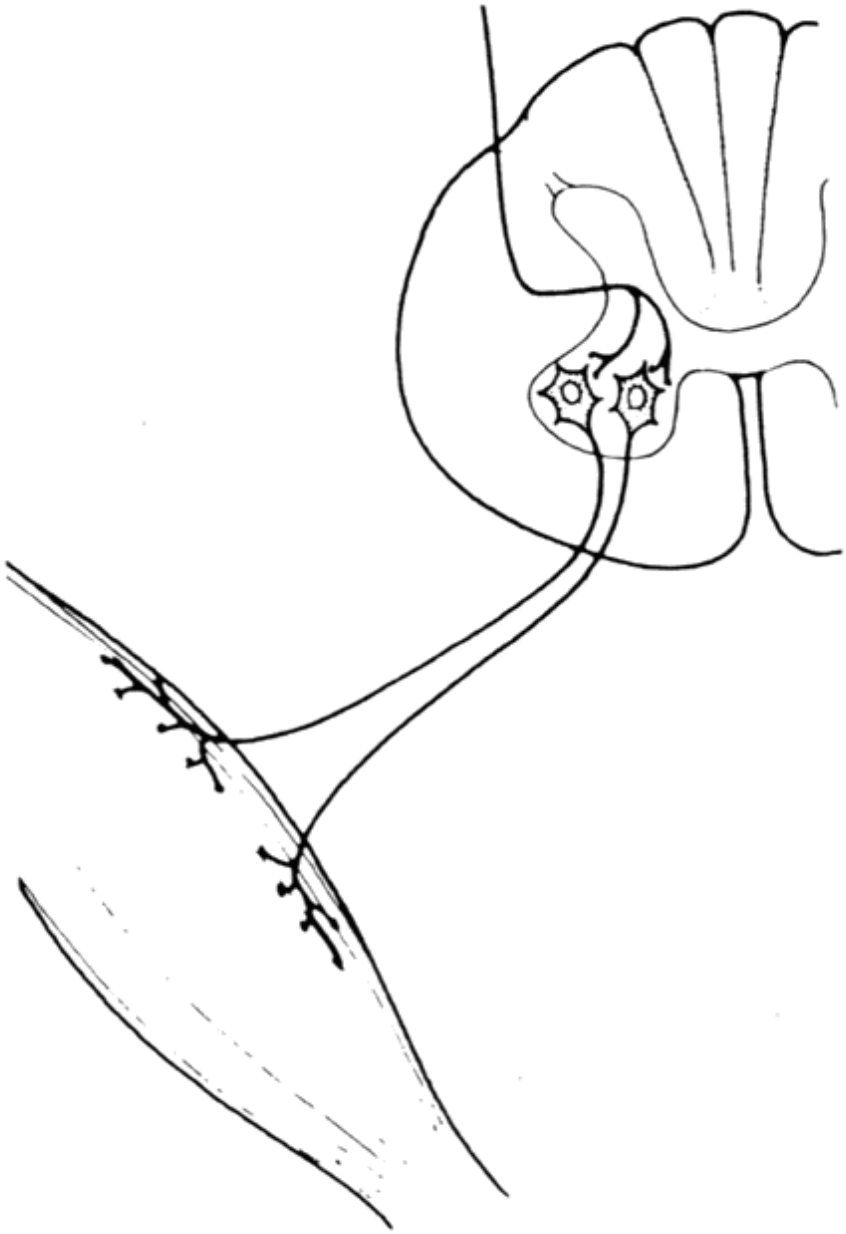

FIGURE 4. Details of the motor unit.

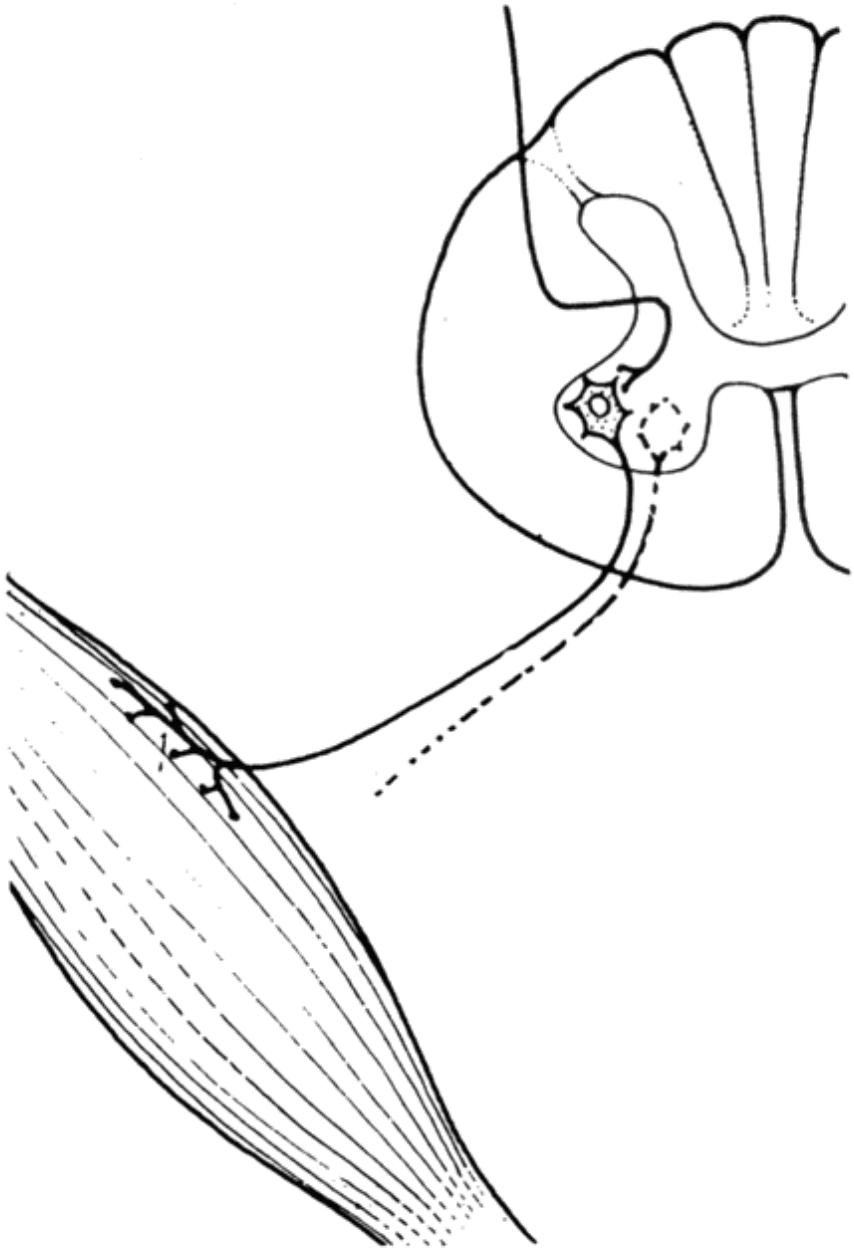

FIGURE 5. Pathophysiological changes seen with acute poliomyelitis.

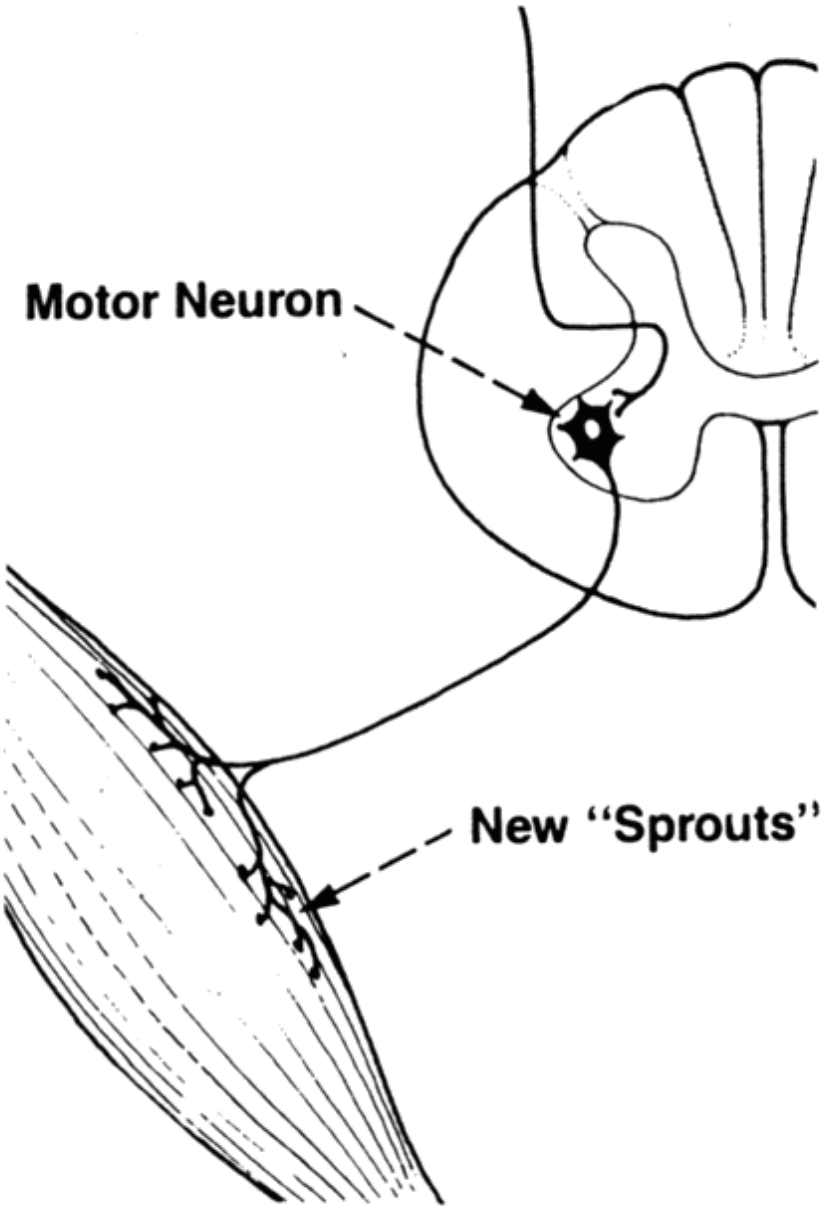

FIGURE 6. Reinnervation through collateral sprouting.

V. PATHOLOGY IN CHRONIC POLIOMYELITIS.

The pathological changes that cause the symptoms of PPS are not well understood; however, a number of possible theories have emerged in recent years.*[30]* These are summarized in Table 1. We explore each of these separately.

TABLE 1 Proposed Etiologies of Post-Polio Syndrome.

Motor unit dysfunction due to overuse or premature aging of large motor units.

Musculoskeletal overuse.

Musculoskeletal disuse.

Loss of motor units with normal aging.

Predisposition to motor neuron degeneration because of glial, vascular, and lymphatic changes caused by acute polio.

Chronic polio virus infection or virus reactivation.

An immune mediated syndrome.

The effect of growth hormone.

The combined effects of disuse, overuse, pain, weight gain, or other illnesses.

A. Motor Unit Dysfunction Due to Overwork or Premature Aging of Polio Affected Motor Units.

In 1902, Gowers proposed that progressive weakness in many degenerative neuronal diseases is due to abiotrophy; the neurons are exhausted and they simply wear out.*[31]* The original viral attack of the anterior horn cells may have left some motor neurons functional but impaired, making them more vulnerable to dysfunction as time passes. Tomlinson observed that many neurons were smaller than normal in the spinal cords of persons who survived long after the acute polio episode.*[32,33]* Consistent with Bodian's findings,*[34]* his observation led him to conclude that the protein synthetic mechanisms of any cell invaded by polio are likely to be permanently damaged.

At this time, we can only speculate that premature exhaustion may be due to abnormalities in DNA and RNA repair or protein synthesis.*[30]* If this is a major factor, it is most likely that this is a combination of damage to both neurons affected by polio and those that have increased metabolic demands because of increased motor unit territory.*[35]* According to this theory, neurological dysfunction results from this increased metabolic load after a critical number of years. This has been demonstrated in electrophysiological studies. Weichers and Hubbel*[26,27]* and Dalakas et al.*[36]* found neuromuscular transmission abnormalities suggesting that the giant motor neurons may not be able to sustain indefinitely the metabolic demands of all their sprouts. As a result, individual terminals slowly deteriorate and reinnervated muscle fibers drop off, as Figure 7 shows.*[21]*

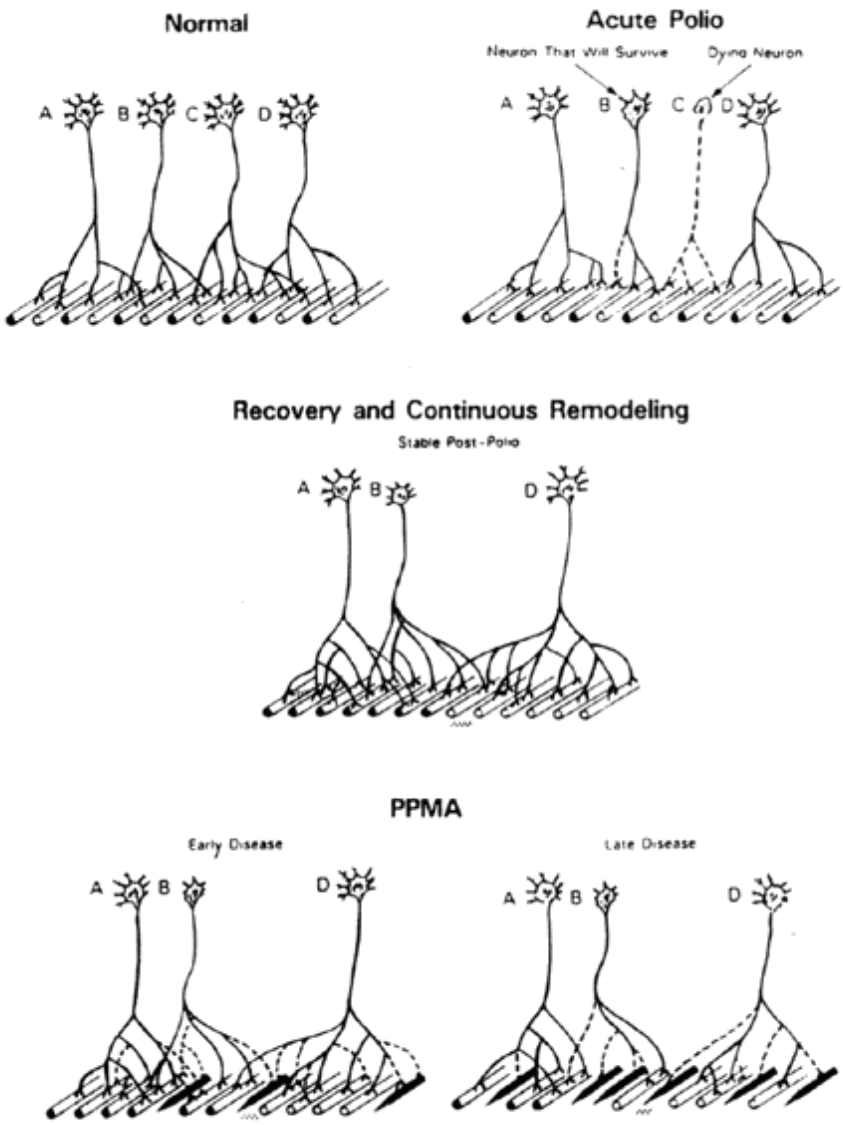

FIGURE 7. Pathophysiology in post-polio muscular atrophy (PPMA). (Adapted from ref. 21.)

Possibly then, it is prolonged overwork with increased metabolic demand of the greatly enlarged motor units that compounds injury to the motor unit sustained during acute infection. The overworked anterior horn cells' control over a greater than normal percentage of muscle function may cause them to succumb "prematurely" to the aging process, resulting in pronounced weakness beginning as early as the fourth decade and steadily worsening with advancing age. Thirty to 40 years after recovery, the giant motor units appear to have lost their ability to sustain all of the terminal sprouts supplying so many muscle fibers. Consequently, the number of muscle fibers driven by each motor neuron declines, and the polio survivor experiences new weakness and other symptoms of neurological dysfunction.

While there is no direct experimental data demonstrating that this increased metabolic demand results in premature aging of the neuron soma, pathologically it does appear that collateral reinnervation is greater, not surprisingly, in weak muscles.*[30,37]* The more muscle fibers lost, the more apparent is the slowly progressive weakness. This hypothesis is intuitively attractive and conceivably explains new weakness in some polio patients but remains unsubstantiated by muscle biopsy changes like group atrophy that would reflect the new loss of whole motor units. Also, no studies have demonstrated permanent biochemical or

physiological damage to the surviving motor neurons.*[9]*

B. Muscle Overuse.

Muscle overuse is less well understood, although studies suggest relationships between the number of motor units, muscle damage, exercise intensity, and duration.*[38-42]* However, the extent to which a primary muscle defect is weakening some polio survivors remains unknown. Overuse has a cumulative effect over time. Chronic mechanical strains on joints, ligaments, and soft tissues that have not been supported well for 30 or more years produce a self-perpetuating cycle of further complications. Recognizing overuse complications early and implementing effective interventions may avert severe post-polio disablement in middle or old age.

Both Windebank et al. at the Mayo Clinic and Maynard found that in persons with similar neurological involvement, new weakness occurred more often in the weight-bearing muscles of the legs than in the non-weight-bearing muscles of the arms.*[38,39]* And those limbs affected the most by the original disease were the most susceptible to new weakness. Perry et al. observed that patients with new lower-extremity weakness had a less efficient gait, with an increase in both the duration and intensity of the extensor muscle contraction.*[40]* Halstead and Gawne showed more patients had mild involvement or subclinical polio in their arms, while clinically unstable polio or atrophic polio was more common in the lower extremities.*[41]* Agre and co-workers found that symptomatic post-polio subjects had evidence of more severe original polio involvement by history, were weaker and capable of performing less work than asymptomatic subjects, and recovered strength less readily than controls.*[42]* Finally, evidence of anatomical damage to muscle fibers is indirectly shown by elevation of creatinine kinase levels found in patients with unstable polio.*[43,44]*

C. Muscle Disuse.

It is well known that disuse leads to both deconditioning and muscle weakness in healthy individuals.*[45]* Similarly, polio individuals have been noted to have similar short-term increased weakness when forced to remain sedentary with illness or injury.*[46]* What part this plays in the development of long-term weakness, however, is less clear.

D. Loss of Normal Motor Units with Aging.

While anatomical and electrophysiological studies have demonstrated that there is a loss of motor neurons with advancing age, this becomes prominent only after 60 years of age.*[33,35]* While some studies of post-polio survivors have failed to show a positive relationship between the onset of new weakness and chronological age,*[38,47]* a recent study by Trojan et al. demonstrates that older individuals are more likely to develop PPS.*[48]* Nevertheless, the most consistent variable is the length of the interval between onset of polio and the appearance of new symptoms, so most patients develop new weakness 30 to 40 years after their initial infection, and the age of onset of symptoms is variable. Therefore, while chronological age may contribute to the development of new weakness, it is probably not the primary causative factor. Also, it is unlikely that polio patients retain "normal" motor neurons. So, the first hypothesis (overuse of large motor units) is more likely.

E. Predisposition to Motor Neuron Degeneration Due to Glial, Vascular, and Lymphatic Damage.

Some investigators have suggested that damage to the glial cells and vascular supply at the time of infection can lead to secondary dysfunction of anterior horn cells.*[49]* While vascular damage is sometimes seen, it is believed that this is secondary to the severe inflammatory responses that can occur. Most studies show that polio affects only the neural cells and not the glial or vascular endothelial cells.*[50,51]* For these reasons, it is unlikely that these changes play a large part in the clinical deterioration seen with post-polio syndrome.

F. Virus Reactivation or Persistent Infection.

Animal studies have shown that poliovirus and other picornaviruses may persist in the CNS and produce late or chronic disease.*[52,53]* When looking for evidence of persistent infection, investigators either study the immune response, by examining oligoclonal bands in the cerebral spinal fluid (CSF), isolating the virus through histochemical or hybridization studies, or looking for evidence of viral genetic material using the polymerase chain reaction (PCR) technique and probe detection. Recently, much work has been done in this area.

Dalakas and co-workers first found evidence of oligoclonal IgG bands in the CSF in seven of 13 symptomatic post-polio patients.*[36]* However, total IgG levels, the IgG index, and IgG synthesis in the CSF were normal, and there were no antibodies to poliovirus. No oligoclonal bands were found in the CSF of six asymptomatic patients.

Evidence for possible reactivation of polio virus was demonstrated by Sharief et al. in 1991 in a study that examined the intrathecal immune response in polio survivors.*[54]* They assessed the antibody response to poliovirus and the production of interleukin-2 and soluble interleukin-2 receptors in 36 patients with PPS and 67 controls, including 13 who had a history of polio but no new weakness and 18 patients with amyotrophic lateral sclerosis. Oligoclonal IgM bands specific to poliovirus were detected in the CSF of 21 of 36 patients with PPS but in none of the controls. In quantitative studies, there was evidence of increased intrathecal synthesis of IgM antibodies to poliovirus only in those with PPS and no increased IgM to other viruses. The patients with PPS had significantly higher mean CSF levels of interleukin-2 and soluble interleukin-2 receptors, corresponding to increased levels of IgM. The presence of an intrathecal immune response to poliovirus in patients with PPS is suggestive that recrudescence of weakness may be caused by persistent or recurrent infection with some part of the poliovirus.

Following publication of Sharief's article in the *New England Journal of Medicine*, letters to the editor by Drs. Salazar-Grueso, Roos, Dalakas, Jubelt, and Cashman pointed out that there was IgM detected in 21 patients, with poliovirus specific IgG in only seven patients.*[55]* They felt it was inconsistent to have IgM but no IgG in a chronic viral infection. More studies were then done to duplicate the findings.

In 1994, Muir and Sharief examined CSF in 24 patients with PPS, 36 with stable polio, and 36 controls.*[56]* Three of 24 patients with PPS had evidence of enterovirus RNA compared with no patients in the other groups. All three of these patients had high intrathecal levels of poliovirus specific oligoclonal IgM bands, which they suggest is evidence that patients with a history of polio are susceptible to persistent enterovirus infection.

In another study, Melchers et al. examined skeletal muscle biopsies in six patients and CSF specimens in an additional 10 patients who met the criteria for PPS, examining the CSF for IgM antibodies to poliovirus and the muscle biopsy for presence of poliovirus RNA by PCR.*[57]* In none of these specimens was any evidence of poliovirus detected. Control CSF specimens in patients with acute poliovirus were positive, while controls in patients with aseptic meningitis (not polio) were negative.

On the other hand, Jubelt et al. examined sera and CSF from 19 post-polio patients and found increased anti-poliovirus (anti-PV) antibodies to type 1 and 2 in seven.*[58]* However, in another study, no anti-PV antibodies were found.*[59]* They have concluded that there was little evidence of intrathecal production of anti-PV antibody.

Leparc and colleagues examined CSF from eight patients with PPS and 10 controls.*[60]* Although no viruses were cultured, using enzymatic amplification of viral DNA, genome sequences were found in several PPS patients and in no controls. They suggest that these results are in favor of the persistence of poliovirus for several decades in PPS patients.

Finally, in 1994, Monzone and Dalakas examined serum and CSF of patients with PPS, comparing them to patients with acute polio, other neurological diseases, and normal controls.*[61]* While both IgM and IgG were highest with acute polio, moderate levels of both were seen in PPS patients compared with the controls. Poliovirus was seen in one of 18 PPS patients by PCR, and amplified genetic product was seen in four of 12 PPS patients. They concluded that high titers of IgM antiPV antibodies imply an ongoing antibody response to antigen, and the presence of viral RNA suggested possible viral persistence.

In summary, regarding the hypothesis of persistent or reactivated infection, an active controversy still exists. However, the lack of consistent findings from study to study and the failure to find conclusive findings in all patients with PPS suggests that this is not the single cause of new weakness. Most researchers feel this is an area that needs to be studied further.

G. An Immune-Mediated Syndrome.

Another hypothesis proposes immunologic involvement. A study by Pezeshkpour and Dalakas described what appeared to be evidence of an ongoing inflammatory or immune response -- active inflammatory gliosis, neuronal chromatolysis, and axonal spheroids in the spinal cords of polio patients who died many years later of other causes.*[62]* Steegman also found inflammatory infiltrates, including lymphocytes, plasma cells, and macrophages in the parenchyma and perivascular spaces in five of seven post-polio patients.*[49]* Whether these changes represent a primary lesion in the cord or a response to a lesion in the distal axon is unknown.

Ginsberg et al. described activated T cells, including significant alternations in CD4+ subsets in both symptomatic and asymptomatic post-polio subjects when compared with normal controls supporting the possibility that immunologic factors may contribute to late disease progression.*[63]* Dalakas et al. have reported preliminary evidence of a lymphocytic response in the form of anti GM_1 neuronal antibodies and IgG oligoclonal bands in the CSF of some patients with new weakness, whereas patients with no new weakness had no oligoclonal bands in their CSF.*[21,36]* He later examined muscle biopsies and found perivascular or interstitial inflammatory cells consisting of CD8+ cells, CD4+ cells, and macrophages. These findings suggest that there is a slow hut ongoing inflammatory process not only in the spinal cord but also in the muscle specimens.*[64]* More recently, immunopathological studies of a patient with PPS showed evidence of focal perivascular interparenchymal inflammatory infiltrates in the CNS. Immunoperoxide staining demonstrated that these were virtually all B lymphocytes, with rare macrophages and no T cells.*[65]* These antibodies could be directed toward neurons, nerve terminals, or postsynaptic antigens. This suggests that PPS could be an autoimmune disorder mediated by antibodies produced *in situ*, and not a cell-mediated process. It is possible that autoantigens from neurons, axons, and muscle membranes might be released during the acute phase of polio. Antibodies against neuronal elements or antiidiopathic antibodies could play a role in the pathogenesis of PPS.

H. The Effect of Growth Hormone.

There is an intriguing suggestion by Shetty, Matsson, and Rudman that the aging of the hypothalamus growth hormone (GH) axis may be a precipitating factor in the development of PPS.*[66,67]* It has been shown that GH secretion drops off dramatically in approximately one third of normal adults over the age of 40. This results in a fall in somatomedin C (SmC) or insulin growth factor (IGF-I), which plays an important role in accelerating the synthesis of DNA and skeletal muscle protein, aids in the proliferation of muscle satellite cells and the regeneration of peripheral nerve sprouting.*[68]*

In a survey of 10 men with PPS and 94 healthy men ages 35 to 63, 100% of those with PPS had SmC less than or equal to 0.40 µ/ml and 90% had values of 0.35 µ/ml or less, while in the healthy population the numbers were 40 and 27%, respectively. In the PPS group, the values did not correlate significantly with either age, functional level, body weight, or years since acute polio.*[66]* In a study of 12 stable polio patients and 10 patients with PPS, Rudman and Shetty found that SmC was markedly depressed in those with PPS and

was normal in those without PPS.*[67]*

In a subsequent study with 124 polio survivors and 261 age-matched healthy controls, Rao et al. measured IGF-I in 124 polio survivors and found that the level was significantly lower in those with a history of polio. The IGF-I levels in that group significantly correlated with age, gender, body mass index, dependency, pain, and difficulty with activities of daily living (ADLs). However, it did not correlate with subjective report of recent decline in functional status.*[69]*

Recently, this hypothesis was tested in six PPS patients with low IGF-I levels.*[70]* These patients were given low-dose human GH treatment for 3 months. Although two of five patients demonstrated improvements in strength and endurance, this was not consistent, and the overall impression was that there was no significant improvement. It was suggested that a longer trial would be necessary. Further investigation into the role of GH therapy for treatment of PPS presents a challenge for the future.

I. The Combined Effects of Overuse, Disuse, Pain, Weight Gain, or Other Illnesses.

Finally, it is hypothesized that a combination of musculoskeletal disuse, musculoskeletal overuse, or motor unit dysfunction may play a significant role in the development of progressive weakness. Furthermore, they may interact with each other in such a way to multiply the effects of any single factor, as illustrated in Figure 8.*[71]* With overuse, weakness may develop. This may lead to disuse, weight gain, and further weakness. Musculoskeletal disuse leads to atrophy, weakness, contractures, and diminished endurance, which are complications that have been studied in other groups with sedentary lifestyles or neuromuscular lesions. If there is overuse, musculoskeletal pain may occur, causing the patient to either rest, developing deconditioning, or compensate with improper body mechanics, leading to further overuse, and possibly pain elsewhere. In our experience, most patients present with some combination of these. Treatment can then be centered on minimizing the effects of one or more of these in order to allow the remaining muscles to function at a more optimal level.

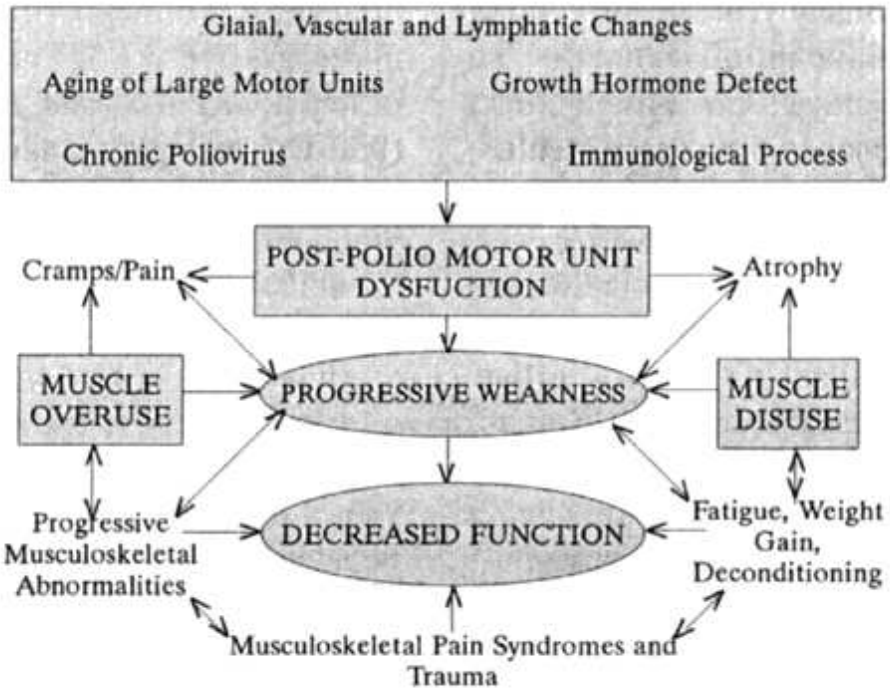

FIGURE 8. Schematic model showing possible etiological factors for the late neuromuscular and musculoskeletal complications of poliomyelitis and their interactions.

VI. CLARIFICATION OF NOMENCLATURE.

There is disagreement about the most appropriate names or diagnostic labels to describe post-polio patients with new health problems. A number of terms have been proposed, including postpolio syndrome (PPS), post-polio muscular atrophy (PPMA), late effects of polio, and postpolio sequelae. One reason for a lack of consensus is that previously these terms lacked specific diagnostic criteria. This is due to the absence of any pathognomonic tests and our incomplete understanding of the underlying pathophysiology of the presenting complaints. Another reason no single term is suitable for all individuals is that there may be one, two, or more pathologic processes present at any one time producing similar, overlapping symptoms. Separating out the origin of each symptom may not only be impractical but impossible, which gives rise to the need for a more general and less precise diagnostic term. Recently, an attempt was made to be more specific, and these terms have been defined more clearly.

PPMA refers to the clinical and pathological features seen in muscles of post-polio patients who are developing **new weakness and atrophy** in muscles both affected and apparently unaffected by the polio virus previously. Specifically, there is documented evidence of neuromuscular deterioration with muscle biopsy showing evidence of active denervation in the form of scattered angulated fibers.*[36]* Mulder first proposed the following criteria for the late progression of weakness in polio survivors in 1972: (1) a credible history of polio, (2) partial recovery of function, (3) a minimum of 10 years of stabilization, and (4) the subsequent development of progressive muscle weakness.*[9]*

In contrast to PPMA, PPS is a more heterogeneous term and therefore more practical in the typical clinical setting. However, it should not be used indiscriminately for every person with a history of paralytic polio with a new complaint. Criteria for making this diagnosis are outlined in Table 2.*[72]*

TABLE 2. Criteria for the Diagnosis of Post-Polio Syndrome.

A prior episode of paralytic polio confirmed by history, physical exam, and typical findings on EMG.
Standard EMG evaluation demonstrates changes consistent with prior AHCD: increased amplitude and duration of motor unit action potentials, an increased percentage of polyphasic potentials and, in weak muscles, a decrease in the number of motor units on maximum recruitment; fibrillations and sharp waves may or may not be present.
A period of neurologic recovery followed by an extended interval of neurological and functional stability preceding the onset of new problems; the interval of neurologic and functional stability usually lasts 20 or more years.
The gradual or abrupt onset of new neurogenic, nondisuse weakness in previously affected and/or unaffected muscles; this may or may not be accompanied by other new health problems such as excessive fatigue, muscle pain, joint pain, decreased endurance, decreased function, and atrophy.
Exclusion of medical, orthopedic, and neurologic conditions that might cause the health problems listed above.

The first criterion is a documented history of polio. The diagnosis of paralytic polio usually can be confirmed by examining, whenever possible, the original medical records; eliciting a credible history of an acute, febrile illness producing motor but no sensory loss; noting whether other members of the patient's family or neighbors had a similar illness; and by observing certain features during physical examination. One very

characteristic feature is the presence of focal, asymmetric weakness, and/or atrophy on examination.

The second criterion is a characteristic pattern on EMG. The changes on routine EMG compatible with prior polio include large polyphasic motor unit action potentials (MUAPs) and a decrease in the number of motor units on maximum recruitment in weak muscles. Occasionally, fibrillations are present. These are discussed in detail later in this article.

The third criterion is a characteristic pattern of recovery. In patients with late complications of polio, the pattern of events from onset of polio to onset of new problems is so characteristic that when it is absent the diagnosis should be seriously questioned. The pattern generally consists of three stages, as shown in Figure 9: (1) paralytic polio in childhood or later in life, (2) partial to fairly complete neurologic and functional recovery, (3) a period of functional and neurologic stability lasting many years, and (4) the onset of new health problems.*[72]*

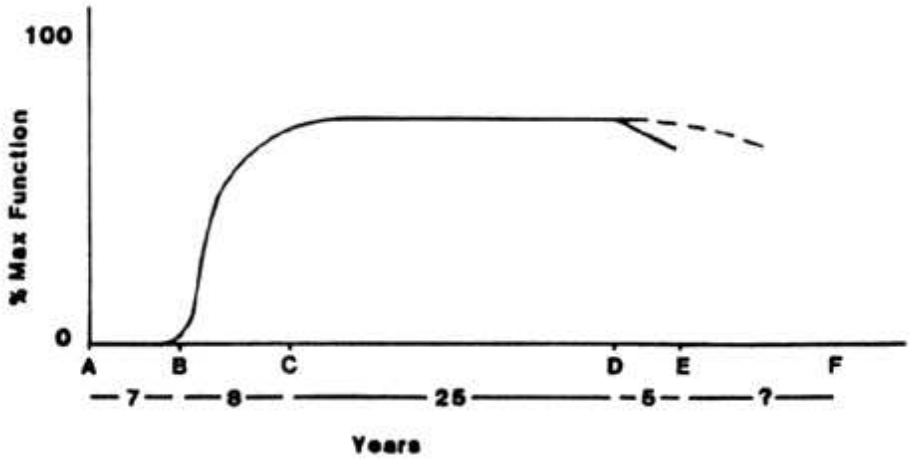

FIGURE 9. Natural history data from post-polio clinic in Houston, Texas. A = birth; B = onset of polio; C = maximum recovery; D = onset of new health problems; E = time of evaluation; F = death. (Ref. 72.)

The fourth criterion is the onset of new neurogenic, non-disuse weakness that may come on either gradually or abruptly. New neurogenic weakness is essential for making the diagnosis of PPS and presumably reflects new or continuing dysfunction of previously injured motor units. Often this new weakness is accompanied by one or more of the other new health problems listed in Table 2. Although the distinction is not always readily apparent, new neurogenic weakness, in contrast to disuse weakness, can frequently be inferred by the **onset of diminished function despite maintaining the usual level and intensity of activity**.

And, finally, the fifth criterion is the exclusion of other conditions that might cause the weakness and other health problems listed in Table 2. In addition to distinguishing between disuse and neurogenic weakness, there are several other dilemmas in making the diagnosis of PPS. First, the symptoms are frequently so general that ruling out all possible causes is not practical and can be prohibitively expensive, and, second, co-existing medical, orthopedic, and/or neurologic conditions may be present that can produce a similar set of overlapping signs and symptoms. As indicated in Figure 8, once a problem such as weakness occurs -- regardless of the underlying etiology -- it may initiate a chain reaction of other complications that makes the original problem impossible to identify.

This fifth criterion is often the most difficult to establish. A history of paralytic poliomyelitis does not exempt anyone from getting the chronic illnesses, diseases, or psychiatric disturbances that afflict the general population. When medical, orthopedic, or neurologic conditions coexist with post-polio problems, a similar

set of overlapping signs and symptoms may occur. Compression neuropathies, radiculopathies, degenerative arthritis, disc disease, obesity, anemia, diabetes, thyroid disease, and depression are some common examples. Nevertheless, because non-disuse weakness is such an important indicator of PPS, and different etiologies dictate very different management strategies, every attempt should be made to differentiate post-polio weakness from other possible causes.

The terms "the late effects of polio", or "post-polio sequelae" are less specific, and refer to the myriad of new symptoms that patients with a history of polio may experience, regardless if there is evidence of actual new motor unit dysfunction. These symptoms: (1) can be attributed directly to damage caused by the poliovirus, such as cold intolerance, or musculoskeletal imbalance; (2) are thought to be related to the body's failure to maintain the level of recovery that was achieved following the infection such as new weakness; or (3) result from a secondary trauma, such as the development of carpal tunnel syndrome (CTS) after years of crutch walking.*[73]* As new information becomes available about the underlying mechanism(s) that produce late onset complications, these criteria will undoubtedly change and new terminology be developed to fit our improved understanding.

Because diagnostic criteria remain nonspecific and pathognomonic tests unavailable, a consistent diagnostic name has not yet been established for new health problems associated with former polio. Indeed, several pathologic processes may interact at any given time to produce similar, overlapping symptoms. The use of a general rather than a precise diagnostic term takes into account the impossibility of determining a distinct origin for each new symptom. The diagnosis "PPS" should be reserved for those patients whose symptomatology indicates motor unit dysfunction with variable musculoskeletal overuse.

VII. CLINICAL FEATURES OF POSTPOLIO SYNDROME.

A variety of studies over the past 10 years demonstrate that the majority of post-polio patients develop similar symptoms, the frequency and relative rank of these are taken from a number of studies and shown in Table 3.*[13,72,73,76,77]* Of these reports, two are population based (Codd and Ramlow), one is based on a questionnaire (Halstead), and the remainder are clinically based, which accounts for the relative differences in the prevalence of symptoms. In addition to the more frequent symptoms, other problems that have been reported include increased sleep requirements, dizziness, syncope, and headaches. Major new functional problems are listed in Table 4.*[72,74,75,77]*

TABLE 3
Comparison of Most Common New Symptoms in Subjects with a History of Paralytic Polio Reported in Six Studies

Symptom	Codd*[77]* N = 28	Halstead*[72]* N = 132	Chetwynd*[76]* N = 694	Agre*[75]* N = 79	Ramlow*[13]* N = 474	Halstead*[74]* N = 539
Fatigue	59%	89%[a]	48%	86%	34%	87%[a]
Joint Pain	74%[a]	71%	60%[a]	77%	42%[a]	80%
Muscle Pain	48%	71%	52%	86%	38%	79%
New Weakness	71%	N/A	47%	69%	38%	N/A
Affected muscle	66%	69%	N/A	80%	N/A	87%
Unaffected muscle	15%	50%	N/A	53%	N/A	77%

Cold Sensitivity	46%	29%	N/A	N/A	26%	N/A

a = Most frequent symptom.
Note: N = Number of subjects reported.

TABLE 4
Most Common New Functional Problems in Persons with a History of Paralytic Polio Reported in Four Studies

Functional Problem	Codd*[77]* N = 28	Halstead*[72]* N = 132	Agre*[75]* N = 79	Halstead*[74]* N = 539
Difficulty walking	25%[a]	63%[a]	N/A	85%[a]
Difficulty climbing Stairs	N/A	61%	67%[a]	82%
Difficulty with ADLS	14%	17%	16%	62%

a = Most common new problem.
Note: N = Number of subjects reported.

In general, the patients most at risk for developing new problems are those who experienced more severe polio at onset, although some patients with typical post-polio symptoms had seemingly very mild polio with excellent clinical recovery. Table 5 summarizes the factors associated with progressive weakness.*[47,48]* Inasmuch as the age at onset of polio is a factor, persons who were older when they contracted polio appear to be at an increased risk for new neurologic symptoms. The onset of these new problems is most commonly insidious, but in many persons they are precipitated by specific events such as a minor accident, period of bed rest, or weight gain. Patients characteristically say that a similar event several years earlier would not have caused the same decline in health and function. Likewise, new problems may begin when coexisting medical problems such as diabetes develop or worsen. More specific details regarding these complaints are covered later in this article.

The time course for the development of late symptoms is fairly characteristic, as demonstrated in Figure 9. While the range between the initial episode and onset of symptoms is 8 to 71 years, the mean interval is around 35 years.*[78]*

TABLE 5 Factors Associated with the Development of Progressive Weakness.*[47,48]*

Age of initial infection more than 10 years old. [a]

History of hospitalization.

Ventilator use.

Paralytic involvement in all four limbs.

More weakness at time of acute polio.

A longer time since acute infection.

Recent weight gain.

Muscle pain associated with exercise.

Greater age at time of presentation to clinic.

a. Reports differ from study to study.

VIII. ELECTRODIAGNOSTIC FEATURES.

The EMG in acute poliomyelitis is characterized by poor MUAP recruitment at the onset of weakness, followed by the development of fibrillation potentials in widespread muscle groups after 3 to 4 weeks.*[79]* The fibrillation potentials subside as reinnervation by surviving motor units proceeds during recovery. In muscles with too few motor units to have adequate reinnervation, fibrillation potentials can persist indefinitely; however, in both partially and completely denervated muscle the fibrillation potential amplitude decays over time. Kraft found that in subjects with partial traumatic peripheral nerve lesions, initially the amplitude of the fibrillation potentials were as high as 1600 µV, with most greater than 700 µV.*[80]* One year later, all potentials were less than 100 µV.

The same is true in chronic polio. Some investigators report that there is an increase in muscle membrane instability between those patients that are clinically stable (no new weakness) and those who are clinically unstable (new weakness). Nelson found that in those patients who were seen for unrelated medical problems, three of 27 had fibrillation potentials, while in those with delayed weakness, 14 of 29 had fibrillations ($p < 0.01$).*[43]* Martinez studied 34 patients, 17 with stable polio and 17 with new weakness. He found that 41% of those with unstable polio had fibrillations, while none with stable strength did.*[37]* However, other investigators, including Cashman, Weichers, Hayward, and Seaton, examined groups of patients with and without new weakness and found evidence of ongoing enervation in the form of fibrillations in both groups.*[23,26,27,81,82]* Some electromyographers have found that muscle membrane instability is more pronounced in the more severely affected muscles.*[83,84]* The muscles that become extremely weak and atrophic have few to no MUAPs, but virtually all, including clinically normal muscles, will have large, polyphasic MUAPs and abnormal, decreased recruitment.*[79]* In a study comparing the size of MUAPs in control subjects and polio patients with and without new weakness, Agre found that the size of MUAPs was significantly larger in all polio patients, and that those with new weakness had larger units than those without. He found no significant correlation between strength and MUAP size.*[42]*

Single fiber studies have shown abnormalities, including increased jitter and blocking and increased fiber density and motor unit territory.*[85]* When comparing electrophysiological and biopsy findings, a significant correlation between the percentage of fibers exhibiting jitter and increased fiber density has been found, suggesting that muscles with the most enlarged motor units as a result of sprouting are more likely to exhibit instability later in life.*[85]*

Defects at the neuromuscular junction have been studied using repetitive stimulation techniques by Trojan et al. Single fiber EMG was performed, and jitter was measured during both low- and high-frequency stimulation in 17 PPS patients and nine normal controls. In five of 17 PPS patients and one control, jitter was significantly higher at high-frequency stimulation. The remaining patients were not significantly different from controls. Those PPS patients with abnormal jitter had a significantly longer time since their acute polio. The authors concluded that the neuromuscular junction defect in PPS patients is probably due to ineffective conduction along immature nerve sprouts and exhaustion of acetylcholine stores, and that this may be dependent on time after acute poliomyelitis.*[86]*

Findings on macro-EMG include increased macro-EMG amplitude, up to 2000% above the mean. Lange et al. found that polio-affected muscles with normal strength had large-amplitude macro-EMG signals (1000 to 4110 µV), weak but stable muscles also had increased large macro-EMG amplitude, while muscles with new weakness had smaller than expected macro-EMG amplitude (130 to 450 µV). Fiber density and jitter were increased in all three groups, but percent blocking doubled in weak muscles.*[87]*

In summary, EMG abnormalities seen in chronic polio include large polyphasic MUAPs in both conventional

and macro-EMG. Although reports differ, it is a fair consensus that there can be evidence of muscle membrane instability in the form of fibrillations in both stable and unstable muscles, although generally these fibrillation potentials are small and sparse. Findings on single fiber EMG include increased jitter, blocking, and fiber density. High-frequency stimulation accentuates this increased jitter, especially in those muscles with new weakness, reflective of defects in the neuromuscular junction. At this time, however, there is no single EMG finding that can distinguish muscles that are developing new weakness.

IX. MORPHOLOGIC FEATURES.

Dalakas et al. evaluated a population of 27 post-polio patients and documented new weakness over an 8-year period.*[24]* Biopsy findings at follow-up included changes in cell morphology, fiber type grouping, small angulated fibers, and hypertrophy of muscles that were less affected. He then separated morphologic findings according to the severity of involvement (or presumed involvement) of polio. These muscles were placed into one of four subgroups. We briefly summarize the characteristics of each of these groups.

Subgroup I included muscles originally affected, but partially recovered, with subsequent new weakness. These muscles showed a combination of myopathic features, with new and old neurogenic changes. The myopathic features included increased connective tissue, occasional necrotic/phagocytosed fibers, variations of fiber size with big and small but rounded fibers, fiber splitting and an abundance of internal nuclei. The old neurogenic changes included fiber type grouping, groups of small fibers, and small angulated fibers. New neurogenic changes seen included small scattered esterase-positive angulated fibers.

Subgroup II were those muscles originally affected but fully recovered, with new weakness. These showed evidence of extensive reinnervation with fiber type grouping consisting of very large groups of up to 170 normal size fibers and variable small scattered angulated fibers.

Subgroup III were those muscles originally clinically spared with new weakness. These muscles like those in group II showed evidence of chronic enervation/reinnervation with large fiber type grouping and occasional angulated atrophic fibers. Contrary to group II, hypertrophic fibers, internal nuclei, and fiber splitting were rare. Only minimal perivascular inflammation was noted.

Subgroup IV were muscles that were asymptomatic: either apparently unaffected or unaffected with subsequent improvement. These muscles showed fiber type grouping but no small angulated fibers or evidence of inflammation.

Grimby and Einnarsson have demonstrated that morphologic changes seen in muscles of patients with PPS weakness include an increase in fiber area with hypertrophy and an increase percentage of type I fibers. These researchers documented in some polio survivors the fiber area of type I and type IIA myofibers on the average are twice the area seen in control subjects. They also demonstrated there is a fiber type transformation from type II (fast twitch, glycolytic) fibers to type I (slow twitch, oxidative) fibers based on these findings. They suggest that excessive use of remaining muscle fibers leads to hypertrophy, and that this is most prominent in the weakest muscles.*[25,28,88]* Borg also documented an increased percentage in type I fibers. In addition, internal nuclei and fiber splitting were prominent findings.*[89]*

In summary, in severely involved muscle myopathic morphologic features include evidence of inflammation and increased connective tissue. Muscles with new enervation may show small angulated fibers, internal nuclei, and fiber splitting. In muscles with reinnervation, large fibertype grouping, fiber-type transformation, and hypertrophy may occur. All of these studies demonstrate the unpredictable nature and wide variability with which polio affects the muscles. No two persons, or even two muscles within the same person, may be affected equally. Because of this, no two people should be treated quite the same. Thus lies the challenge in treating the person with PPS.

X. EVALUATION OF THE POST-POLIO PATIENT.

Proper assessment of post-polio patients presents both a challenge and a dilemma: a challenge because of the diversity and nonspecific nature of the problems they may present with, and a dilemma because of the absence of specific diagnostic tests, the continuing uncertainty of the underlying cause or causes, and the lack of any curative therapeutic intervention. Our approach to the assessment of post-polio patients is based on a number of assumptions concerning their past health experience and present needs. These assumptions guide the format and content of our evaluation. They are based on the experience gained in assessing and managing over 1000 patients over nearly a decade and the lessons learned in organizing and running two major polio programs in two different institutional settings. Clearly, these assumptions represent a particular bias and we recognize that other professionals, with a different perspective or with different resources available to them, may want to use a modification of the approach outlined here. We believe it is essential that the evaluation includes the following elements:*[90]*

A. Comprehensive and Interdisciplinary Evaluation.

Because of the number, diversity, and complexity of the problems presented by these patients, a comprehensive, coordinated assessment is required. For many of these patients the evaluation in the post-polio clinic may be one of the first examinations done by a group of specialists that are familiar with the features of PPS. The best way to provide a comprehensive, coordinated evaluation that looks at the medical, functional, and psychosocial and vocational issues of this population is to use an interdisciplinary rehabilitation team including the physician and nurse and physical and occupational therapists. A social worker or psychologist, orthotist, and respiratory therapist may complete this team.

B. Diagnosis by Exclusion.

Because post-polio syndrome is a diagnosis by exclusion, it is essential that every patient receive a careful history and physical examination, along with appropriate laboratory, radiological, and diagnostic studies to rule out other medical, orthopedic, or neurologic conditions that might be causing or aggravating the symptoms. A psychosocial evaluation is often helpful, along with an assessment of function, gait, and orthotic needs. In addition, a baseline measure of strength and endurance in key muscle groups is essential to observe for the appearance of new weakness. Because manual muscle testing alone is not a reliable and valid measure of strength over time in post-polio patients in comparison to more quantitative measurements, we recommend objective testing of key muscle groups in the good to normal range with a myometer or isokinetic test.*[91,92,93]* For virtually every patient, we feel a standard electromyogram/nerve conduction study (EMG/NCS) of all four extremities and appropriate paraspinals is essential. We have found that this test is invaluable in confirming the presence of an old anterior horn cell disease (AHCD), identifying major muscle groups with subclinical involvement, establishing a baseline and helping exclude certain other neurologic and myopathic conditions, and in detecting additional diagnoses, including carpal tunnel syndrome (CTS), ulnar neuropathy, and radiculopathy. A summary of our experience with 100 consecutive patients is shown in Table 6.*[94]* We do not believe more sophisticated studies with single fiber EMU or macro-EMG are indicated in the routine clinical setting because they have not helped separate the symptomatic from the asymptomatic patient or proven useful in guiding clinical management.

TABLE 6
Additional Electrodiagnostic Findings in 100 Consecutive Post-Polio Patients (Ref. 94.)

Finding	N	%
Carpal Tunnel Syndrome (CTS)	35	35
Ulnar Neuropathy at the Wrist	2	2

CTS and Ulnar Neuropathy	3	3
Peripheral Neuropathy	3	3
Brachial Plexopathy	1	1
Tibial Neuropathy	1	1
Radiculopathy	4	4
Total	49	49
Subclinical Polio	49	49

A standard battery of screening tests such as an electrolyte panel, fasting glucose, etc. used on a routine basis are generally not helpful; however, when indicated we may obtain a hematocrit, blood glucose, creatine phosphokinase (CPK), or thyroid function tests. Whether it is useful to monitor CPK levels on a regular basis to assist in determining long-term prognosis or as an aid in clinical management is still not clear.

Patients who had respiratory involvement initially and have a history of pulmonary disease or scoliosis should have a screening vital capacity (VC) and functional expiratory volume (FEV1) measured along with their other vital signs. If the vital capacity is less than 50% of predicted or the history and clinical situation warrant, further pulmonary function tests (PETs) are obtained. If the patient has significant spinal curvature, a 36-in gravity loaded scoliosis film is obtained to provide a baseline for follow up exams. If degenerative joint disease (DJD) or other skeletal abnormalities are suspected, appropriate radiographs are obtained.

C. Expectation of Improvement.

We believe that everyone who comes to the clinic can be helped, regardless of the etiology or severity of disability. As a result, our goal is that everyone, even if they can implement only some of the recommendations and interventions, will feel better physically and emotionally and achieve an improved level of function.

D. Convenience and Efficiency.

Because of the comprehensive, interdisciplinary nature of our evaluation, and frequently the decreased stamina these patients may have, we attempt to complete the evaluation in 1 or 2 days, having team members come to the patient in a central location rather than having the patient come for a series of single service outpatient evaluations. All laboratory, radiological, and electrodiagnostic procedures are performed on location the day of the evaluations.

A typical evaluation in the NRH Post-polio Clinic extends over 2 days, with the first day reserved for evaluations by team members, including a rehabilitation nurse, physician, physical therapist, occupational therapist, social worker, and, if needed, an orthotist.

The patient is seen initially by the nurse, who makes a brief assessment of the past and current health status, clarifies the patient's goals for the clinic visit, coordinates the evaluations by the team members, schedules diagnostic tests, and assists with patient and family education. The medical evaluation consists of a comprehensive history and physical exam, with special attention to the history of the details of the initial illness with acute polio and its management and a special focus during the physical exam on neurologic and musculoskeletal findings. There is also an analysis of station and gait to determine the need for orthoses and other durable medical equipment. In addition, the physician determines the need for X-ray, laboratory and electrodiagnostic studies, and initiates referrals to other rehabilitation disciplines, such as a nutritionist, vocational counselor, speech language pathologist, or psychologist, as well as other medical and/or surgical

specialists if needed.

The physical therapist's evaluation is based on a protocol outlined by Smith and includes a baseline manual test of major muscle groups, measurement of major joint range of motion and leg length, and an evaluation of habitual postures during standing, sitting, lying down, and walking.*[95]* It also includes an analysis of activities and positions that provoke or relieve muscle and joint pains. The occupational therapist's assessment is based on a format described by Young and includes an evaluation of activities that produce pain, weakness, or fatigue; when they occur; and how these problems interfere with the person's activities of daily living (ADL).*[96]* Special attention is paid to the frequency and intensity of activities in the home, at work, in the community and during travel, and to the use or need for adaptive aids.

The social work evaluation focuses on how new health problems and functional loss impact on the patient, the family, significant others, and colleagues at work, school, or elsewhere outside the home. There is also an effort to identify coping strategies used by, and available to, the individual and assess the emotional impact of the original polio experience and relate it to current feelings of having a second disability.*[97,98]* In addition, the social worker facilitates referrals and access to community resources and services, including the local post-polio support group. In the afternoon, we obtain any necessary diagnostic tests to help rule out other medical, orthopedic, or neurologic conditions that might be causing or aggravating the patient’s presenting symptoms.

Finally, the morning of the second day is used to complete any unfinished evaluations and to hold a team conference with the patient and his/her family. This conference is used to review the results of diagnostic tests and discuss our impressions and recommendations for interventions. Patients are given a written copy of recommendations and then seen in follow-up 6 to 8 weeks later to evaluate the effectiveness of the interventions and make any modifications or additional suggestions for management. Thereafter, patients are seen as needed and at annual intervals for a repeat functional evaluation and manual muscle test as well as an interim history and a physical.

In summary, the assessments provided by special diagnostic testing are generally more fruitful in excluding certain conditions than in assisting either with the diagnosis or management of post-polio syndrome. Despite the growing body of evidence that suggests that the major pathologic process is motor unit dysfunction, there is still no objective method to predict who might develop new weakness in the future or to monitor the progress of the underlying pathology in the subject who has already becoming weaker. Specifically, no serologic, enzymatic, electrodiagnostic, or muscle biopsy test can diagnose PPS. In general, we rely on a careful patient history to distinguish between those patients who have no new weakness (clinically stable) and those who are experiencing new weakness (clinically unstable).

XI. DIFFERENTIAL DIAGNOSIS AND REHABILITATION MANAGEMENT.

In general, we have found that patients most at risk for developing new problems are those who experienced more severe polio at onset, although it is not unusual to see patients with typical post-polio symptoms who had seemingly very mild polio with excellent clinical recovery. Most commonly, the onset of these new problems is insidious, but in many persons they may be precipitated by specific events such as a minor accident, fall, period of bed rest, or weight gain. Characteristically, patients state that a similar event experienced several years earlier would not have caused the same decline in health and function. Likewise, new problems may begin when coexisting medical problems, such as diabetes, develop or worsen.

The symptoms experienced by polio survivors, unfortunately, are fairly common and nonspecific. The lack of specificity and a characteristic cluster of symptoms have led some observers to question both the validity of the symptoms and the existence of a diagnosis of post-polio syndrome. Until a pathognomic test is found, this dilemma will undoubtedly persist. However, health professionals who have become experienced in this field in recent years generally agree there are definite qualitative features of these symptoms that are reasonably

characteristic. The differential diagnosis of symptoms experienced by persons with a history of polio is complex, and they must be evaluated in a symptom by symptom manner. We review these symptoms, their differential diagnosis, and outline an approach to the evaluation and rehabilitation management that we have found to be successful.

A. Weakness and Functional Loss.

1. Differential Diagnosis.

Because of the importance of weakness as a cardinal symptom of motor neuron dysfunction and post-polio changes in general, it should be addressed early and one should make the differentiation between post-polio syndrome as a cause of new weakness or other causes of new weakness. New weakness may appear in muscles previously affected and/or muscles believed to be previously spared. The weakness is usually most prominent, however, in the muscles most severely involved in the initial illness. Diminished functional capacity tends to parallel the muscle weakness and can be quite dramatic if functional reserve was marginal. One of the characteristics of many polios was their ability to appear "normal" or function at an extraordinarily high level of performance on relatively few good muscle groups. This was possible because of the random, scattered nature of the motor deficits and the body's uncanny ability to compensate with unconventional muscle and joint function. In this situation then, late onset weakness of a critical muscle often leads to disruption of a delicate balance that has been maintained for years, leading to a disproportionate amount of functional loss. Persons with involvement of one or both legs may have increased difficulty in walking, standing, climbing stairs, or other endurance activities. individuals with presumably normal upper extremities who have been "walking" on their arms with crutches for years may find that ambulating, transfers, driving a car, or even dressing are more taxing and the time to recover takes longer than it used to.

When patients present with complaints of "new weakness", the first task the practitioner needs to do is differentiate between true weakness (a loss in muscle strength) and other synonymous terms the patient may be referring to, such as lassitude, fatigue, lack of energy, and languor. Whereas the polio patient may also experience these symptoms, they are complaints of fatigue, not progressive muscle weakness. If possible, objective muscle testing with a myometer or isokinetic system should be done to establish a reliable baseline. *[91-93]* This baseline is essential for monitoring changes in strength in the future. If there is a history of new weakness (often revealed as decreased endurance with diminished function) combined with objective changes, the major differential diagnoses include focal neurological disease such as a radiculopathy, focal compressive neuropathy, or spinal cord lesion and medical causes of neuropathy such as diabetes, thyroid disease, uremia, alcohol, toxins, and, uncommonly, hereditary neuromuscular disease. In most cases, history and a physical alone can make this distinction; however, when necessary, laboratory or electrodiagnostic data may be needed to clarify the appropriate diagnosis. Once other causes are ruled out, an attempt should be made to distinguish the etiology of this weakness (disuse vs. overuse), so that appropriate recommendations regarding exercise and activity can be made.

For patients with new weakness (with or without atrophy), the major differential confronting the clinician is to distinguish between neurogenic weakness due to polio and disuse weakness caused by diminished activity. Although the distinction is not always readily apparent in the clinical setting, when new neurogenic weakness is present a careful history can usually elicit a pattern of decreased strength, endurance, and function despite attempts to maintain the usual level and intensity of activity. Routine, daily activities that require repetition or sustained contractions, such as walking, climbing stairs, standing, or pushing a wheelchair can sometimes provide a semi-quantitative picture of new weakness when current performance is compared with similar activities in the past, for example, number of stairs climbed without difficulty 1, 3, or 5 years ago vs. now. When the presence of new neurogenic weakness is in doubt, a trial of carefully monitored exercise is indicated to exclude the possibility of disuse weakness.

As indicated earlier, we feel the workup for new neurogenic weakness should include an EMG/NCS.

Although electrodiagnostic studies even in the most skilled hands have their limitations, they provide a necessary screening tool to help exclude some of the more common causes of neurogenic weakness in this population. Specifically, EMG studies can indicate the presence of radiculopathies from disc disease or other causes and help differentiate old polio from other neuromuscular disorders such as adult onset spinal muscular atrophy and myopathies. Nerve conduction studies can identify the presence of localized compression neuropathies as well as generalized peripheral neuropathies. Follow-up laboratory and imaging studies can help clarify the underlying etiology suggested by the electrodiagnostic exam and reveal other causes for weakness such as occult tumors, toxic metal exposure, and endocrine disorders. New weakness may appear in muscles previously affected and muscles believed to be previously spared. The weakness is usually most prominent, however, in the muscles most severely involved initially. Diminished functional capacity tends to parallel muscle weakness and can be quite dramatic if functional reserve was marginal. This occurs because of the functional compensations that are made early within the recovery period.

2. Management.

There has been a controversy about the management of new weakness with exercise in the post-polio patient because the pathophysiology of PPS remains unclear. Traditional therapy such as exercise may cause further weakness, so it must be used cautiously. As there is now a considerable body of literature on this subject -- some of it contradictory -- we will provide a quick review of what is known before proposing specific recommendations.

One of the first formal isotonic strengthening programs for patients with a history of polio was described in 1948 by Delorme and Watkins, who applied the principle of progressive resistive exercises (PREs) in 19 post-polio subjects.*[92]* In the end, 17 of 27 muscle groups demonstrated gross gains in strength measured with MMT. Muscle power in 15 of 27 quadriceps muscles doubled or more than doubled as measured by a spring scale. All except three muscles showed an increase in work capacity. In 1950, Gureswitsch evaluated 13 subjects who were in the initial phases of recovery from their polio.*[99]* They exercised with a modification of Delorme's protocol. After training, both muscle strength and endurance increased 50%.

In the early 1980s, specific exercise studies were carried out for survivors with PPS. In 1984, Feldman and Soskolne developed an exercise protocol they described as "non-fatiguing strengthening exercises" for a population of six subjects with post-polio symptoms.*[100]* This was performed three times weekly for a period of 3 to 6 months. They found that by using this routine for at least 24 weeks, 14 muscles (46%) got stronger, 17 (53%) showed no change, and 1 muscle got weaker, as measured by myometry. There was no relationship between the initial weakness and improvement in strength.

Grimby and Einarsson reported an isokinetic exercise program for 12 post-polio subjects, including nine with symptoms of PPS, in 1987.*[101]* These exercises were performed for a total of 6 weeks. Only one leg was trained and the other served as a control. There were significant strength gains in the trained leg. The investigators concluded that increases in strength might be explained through both muscular as well as neural adaptations. In 1991, Einarsson used a similar protocol for muscle conditioning of knee extensors in 30 post-polio subjects.*[102]* This was performed three times a week for 6 weeks. There was a significant improvement in knee extension strength, with no change in the strength of the knee flexors. These changes were also associated with subjective improvements in functional tasks and general well-being.

Also in 1991, Fillyaw and colleagues used Delorme's training program with 17 PPS subjects.*[103]* The quadriceps or biceps were exercised, with the contralateral extremity used as a control. Three sets of 10 repetitions with a 5-min rest between sets were performed every other day. Sixteen of 17 subjects demonstrated significant strength gains, but there was no evidence of increased endurance. They cautioned patients should undergo periodic quantitative muscle testing under the supervision of a physical therapist to avoid overwork weakness.

Agre and Rodriguez have described an exercise technique known as "pacing", mixing periods of exercise with periods of rest, in post-polio subjects.*[104-106]* They found when subjects paced themselves they had less evidence of muscle fatigue, increased capacity to perform work and increased ability to recover strength after activity.

In addition to strengthening programs, studies were also done to determine the cardiovascular fitness of post-polio survivors and to assess their response to a cardiovascular training program. In 1985, Owen and Jones evaluated the cardiovascular endurance of 21 subjects in an EKG-monitored, symptom-limited graded exercise test.*[107]* They found that the subjects had an average maximum fitness level of 5.6 metabolic equivalents (METS), indicative of severe deconditioning. They proposed an exercise program as follows: intensity of 65 to 80% of reserve HR (maximum HR-resting HR), a duration of 15 to 30 min, and a frequency of three to five times per week on alternate days.

In 1987, Alba and co-workers evaluated the work capacity of 35 subjects, including 33 that were complaining of new symptoms.*[108]* Parameters evaluated included muscle strength using MMT, body weight, maximum METS, maximum heart rate (Hrmax), maximum oxygen uptake (VO_2 max), and vital capacity (VC). Significant findings included a decreased VC, especially in those who smoked or had a history of respiratory involvement. There was also a decreased Hrmax, decreased maximal cardiac output, and decreased work capacity. She recommended that post-polio survivors partake in any repetitive activity that appealed to them, if it was considered "safe" and recommended stopping the activity if it caused "undue pain, muscle fatigue or a sense of weakness" that required more than the usual time to recover.

Dean and Ross examined the effects of a modified exercise program in three post-polio subjects in 1988.*[109]* Each subject met the diagnostic criteria for PPS and was ambulatory without assistive devices. Using one subject as an untrained control, the other two walked on a treadmill three times a week at a submaximal rate for a total of 8 weeks, advancing their walking duration from 22 to 31 min. The rating of perceived exertion (RPE) was monitored using a 1 to 10 scale,*[110]* and pain was monitored using a 1 to 4 scale. An attempt was made to keep the RPE below 2 (light). After training, on submaximal testing, both the trained subjects demonstrated reductions in VO_2, HR, blood pressure (BP), RPE, and energy cost at similar work loads compared with the pretest, while the untrained subject showed no change. There was no apparent effect of training on pulmonary function. The mechanism for the change was felt to be both cardiovascular conditioning as well as muscle adaptation.

In 1989, Jones and colleagues evaluated the cardiorespiratory responses to aerobic training in 16 post-polio subjects who participated in a 16-week exercise program.*[111]* Baseline tests, including resting HR and BP, HRmax, BPmax, VO_2, and expiratory volume (Ve), were performed on a bicycle ergometer. Subjects were divided into exercise and control groups. The subjects exercised three times a week for a period of 15 to 30 minutes using a predetermined target HR. Significant results on post-test included an improvement in the total exercise time, total work per time, VO_2, and Vemax within the exercise group. In 1992, in a similar study Kriz and colleagues examined the effects of an upper-extremity arm ergometry program in 29 post-polio subjects.*[112]* The exercise group participated three times a week in a 16-week training program with a heart rate of 70 to 75% HR reserve, with 1 d of rest between exercise sessions. After the 16-week program, the exercise group showed significant improvement in VO_2 max, Vemax, power, and exercise time. None of the subjects in the exercise group developed problems with pain or overuse.

In summary, although it is true that individuals with a history of polio may develop new weakness many years after the original episode, it has been shown that many can improve both muscle strength and cardiovascular endurance from a well-planned training program. There appears to be positive benefits regardless of whether the muscle group is with or without new weakness. The type of exercise program needs to be selected according to both the needs of the individual and the resources available. An isometric program is of benefit in those who have less than antigravity strength, a painful joint, or a joint immobilized in a cast due to surgery

or a fracture. An isotonic program is more appropriate for a home exercise program for a non-painful joint with greater than antigravity strength. An isokinetic program can be used when the equipment is available and the muscles have greater than antigravity strength. While the long-term safety of a strenuous strengthening program on muscle that has been severely affected is not known, the positive effects of exercise on other systems, including cardiovascular and respiratory systems are clear. Therefore, it can be concluded that a carefully monitored program is beneficial for most individuals with a history of polio. It is also possible that many of the secondary symptoms such as generalized fatigue can be reduced as patients become conditioned and are able to perform similar amounts of work with less expenditure of energy. An ideal cardiovascular program should exercise the muscles least affected by polio in order to get maximum cardiovascular benefits, while avoiding overuse or secondary degenerative effects on the more affected extremities. For instance, if the legs are the more involved limbs, then the arms can be used in a more strenuous program. The exercise program should be initially supervised by a physical therapist in order to teach proper techniques, including the monitoring of HR and RPE. As with all exercise programs, a warmup followed by gentle stretching should be done to improve flexibility and reduce the possibility of injury. Both aerobic and strengthening exercises can be done. Finally, after exercising, a cooldown period should take place. The type of activity should be one that the participant enjoys to minimize lack of interest and potential for drop out.

In order to prescribe the safest and most appropriate exercise program for post-polio patients in our clinic, we have developed a new limb-specific muscle classification system called the National Rehabilitation Hospital (NRH) Post-Polio Limb Classification.*[41,113]* A standardized taxonomy provides a means of establishing a common language that can be used by clinicians and researchers alike. This makes it possible to establish an objective baseline for each muscle and limb before initiating treatment and then apply a systematic, rational protocol appropriate for the degree of polio involvement. When results of various protocols and clinical trials are compared, we have a better basis for understanding their efficacy and their applicability to other patients or limbs with similar involvement. It can be used to give recommendations regarding activity, prognosis, and need for assistive equipment.

To classify a limb, a combination of history, physical exam, and EMG is used. Key elements of the history include both remote and recent history of weakness. Physical examination focuses on strength, sensation, and reflexes. On electrodiagnosis in addition to evaluating for specific clinical conditions, a screening examination is performed. It includes nerve conduction studies (NCS) of bilateral median and sensory nerves, as well as needle examination of at least three muscles in each extremity.*[94,114]* Following the EMG, each muscle is classified separately and the limb is classified according to the most severely involved muscle. Table 7 summarizes this classification.

TABLE 7
The National Rehabilitation Hospital (NRH) Post-Polio Limb Classification

NRH Class I	No Clinical Polio
NRH Class II	Subclinical Polio
NRH Class III	Clinically Stable Polio
NRH Class IV	Clinically Unstable Polio
NRH Class V	Severely Atrophic Polio

NRH Class I muscles (no clinical polio) have no history of past or new weakness. Strength ranges from good to normal, and there is no atrophy, sensory, or reflex changes. On monopolar EMG there is no evidence of muscle membrane instability such as fibrillation potentials (fibs) or positive sharp waves (PSW). The MUAPs are normal in size and configuration with normal recruitment. Single fiber EMG or muscle biopsy may show evidence of old anterior horn cell disease if extensive testing is carried out; however, in a clinical setting, this is not feasible. The objective of the exercise program for Class I muscles or limbs is to increase muscle

strength and cardiovascular endurance. Exercise recommendations consist of a strengthening program with PREs as described by DeLorme. These muscles can be used selectively in an aerobic program to improve cardiovascular conditioning following the American College of Sports Medicine (ACSM) recommendations for frequency and duration.*[114]* A typical program might include the following: exercise three to four times a week for a period of 15 to 30 min at a heart rate of 60 to 80% of HRmax or an equivalent of 8 to 9 METS (Table 8).*[115]*

TABLE 8
Table of Metabolic Equivalents (METS) (Ref. 113.)

1 - 2 METS
Doing seated ADLs (eating, performing facial hygiene, resting)
Doing seated recreation (sewing, playing cards, painting)
Doing seated occupational activities (writing, typing, doing clerical work)

2 - 3 METS
Standing ADLs (dressing, showering, shaving, doing light housework)
Standing occupation (mechanic, bartender, auto repair)
Standing recreation (fishing, playing billiards, shuffleboard)
Walking 2.5 mph
Bike riding 5-6 mph

4 - 5 METS
Doing heavy housework (scrubbing floor, hanging wash)
Canoeing, golfing, playing softball, tennis (doubles)
Social dancing, cross country hiking
Swimming 20 yards/min
Walking 4 mph (level), 3 mph (5% grade)
Bike riding 10 mph

6 - 7 METS
Heavy gardening (digging dirt, lawn mowing, hoeing)
Skating, water skiing, playing tennis (singles)
Stair climbing (<27 ft/min)
Swimming 25 yards/min
Walking 5 mph (level), 3.5 mph (5% grade)

8 - 9 METS
Active occupation (sawing wood, digging ditches, shoveling snow)
Active recreation (downhill skiing, playing ice hockey, paddleball)
Bike riding 12 to 14 mph, stair climbing (27 ft/min)
Swimming 35 yards/min
Walking 9 mph, 3 mph (15% grade)

NRH Class II muscles (subclinical polio) have no history of past or new weakness, or if they were affected they have fully recovered. Strength is good to normal, sensation and reflexes are normal. EMG is consistent with anterior horn cell disease (AHCD). The goal of the exercise program for Class II extremities is to increase strength in those muscles in the good range and maintain normal strength in the remainder. If other extremities are more severely affected, Class II extremities can be used to improve cardiovascular endurance using ACSM guidelines for frequency and HR. However, the session should be paced, with exercise periods of 4 to 5 min and rest breaks of 1 min. The frequency of exercise should also be paced, alternating exercise and rest days. As a training effect develops and the individual is able to perform a similar workload with less fatigue, both the amount of resistance used and the frequency and duration of the exercise can be altered to

meet the changing increases in strength.

NRH Class III muscles (clinically stable polio) have a remote history of weakness with some improvement and no complaints of new weakness. On physical examination, the strength ranges from fair to good. Sensation is normal and reflexes are normal or decreased proportional to the muscle strength. EMG shows evidence of old AHCD. These muscles and limbs along with those in Class I and II represent those previously described as "asymptomatic".*[42]* The goal for Class III extremities is to at least maintain strength and if possible gain strength in those muscles that are deconditioned. Recommendations include strengthening exercises, similar to Class II, with pacing. Strength should be carefully monitored and the program modified if weakness develops. Aerobic activities for Class III extremities with strength greater than or equal to antigravity are similar to those for Class II extremities. For those extremities with less than antigravity strength, cardiovascular exercises in the 4 to 5 MET range are appropriate, pacing 2 to 3 min of activity with 1 min rest, three times a week, on alternating days. For limbs with degenerative joint disease (DJD), non-weight-bearing exercises such as a pool program are preferred.

NRH Class IV muscles (clinically unstable polio) are those that are developing new weakness and sometimes atrophy. Sometimes these limbs or muscles were described in the literature as "symptomatic".*[42]* They are usually weaker, with less dynamic and isometric strength than Class III. Sensation is normal, old atrophy usually is present, and reflexes are decreased. EMG findings are largely similar to Class III, as indicated previously. The goal in this class is to prevent further weakness, so we recommend first decreasing activity if overuse is suspected. If disuse is suspected, or rest does not help, then an exercise program is begun. A nonfatiguing exercise program would be appropriate for strengthening. Because many muscles may have less than antigravity strength, exercises should be done in a gravity-eliminated position or in a pool. Muscle strength is carefully monitored. This program should be done no more than three times a week and modified if symptoms of pain, new weakness, or fatigue develop. For cardiovascular conditioning we generally recommend that they be used in ADLs only. As we have found that many of our patients have less affected upper extremities, we frequently advise our patients to do a cardiovascular exercise using only their arms, such as swimming or arm ergometry. For those limbs with severe weakness, we recommend avoiding weight bearing by using an assistive device, wheelchair, or motorized scooter. Bracing is frequently necessary.

NRH Class V muscles (severely atrophic polio) are those that are originally affected with severe weakness and little improvement. New weakness may be present. On physical examination they are extremely weak (trace to poor) with marked atrophy, no sensory changes, and are areflexic. On EMG there is decreased insertional activity, few fibs or PSW, little to no MUAPs, with variable amplitude, increased polyphasics, and markedly decreased recruitment. These limbs cannot be exercised; however, a passive range of motion program is suggested.

Table 9 shows the result of classification of 400 limbs in 100 consecutive post-polio patients evaluated in our clinic.*[41]* The distribution of our findings in both the upper and lower extremities are detailed in Figures 10 and 11. Note the higher percentage of upper extremities in Classes I and II with a higher percentage of lower extremities in Classes III, IV, and V.

It has been demonstrated that people with a history of polio can improve muscular strength and endurance as well as cardiovascular conditioning in individualized, carefully monitored exercise programs. With this in mind, in conjunction with an appropriately trained professional, a prescription for a rational exercise program can be developed using this limb-specific classification system with the goal that virtually all post-polio individuals can more safely enjoy many of the additional positive benefits that regular exercise can bring.

TABLE 9
NRH Limb Classification Of 100 Consecutive Post-Polio Outpatients N = 400[41]

NRH Class	Upper Extremity	Lower Extremity	Total

I	84	10	94
II	42	46	88
III	42	53	95
IV	25	50	75
V	7	41	48
Total	200	200	400

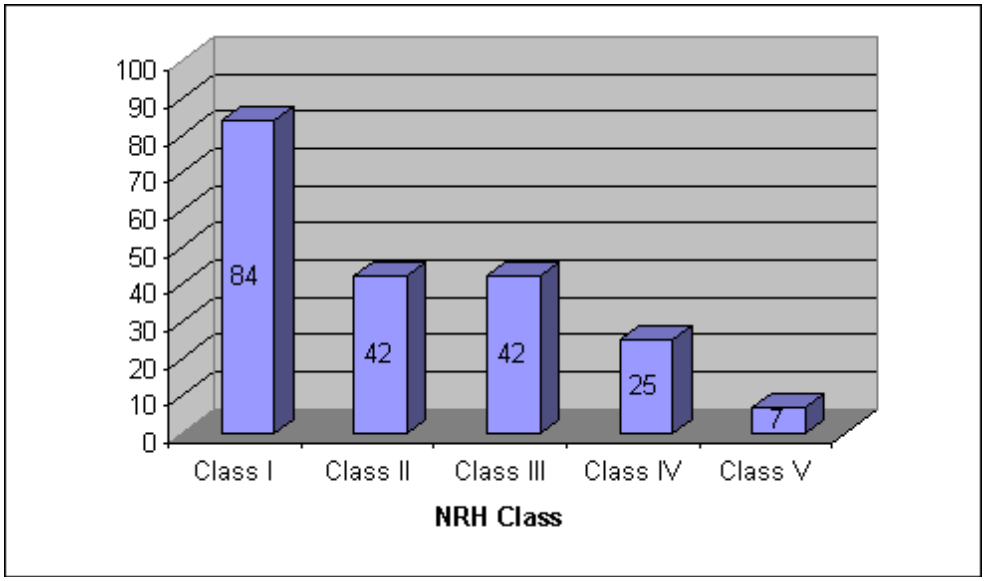

FIGURE 10. Distribution of upper extremity limbs by NRH Limb Classification in 100 consecutive post-polio patients (N = 200). (Ref. 41.)

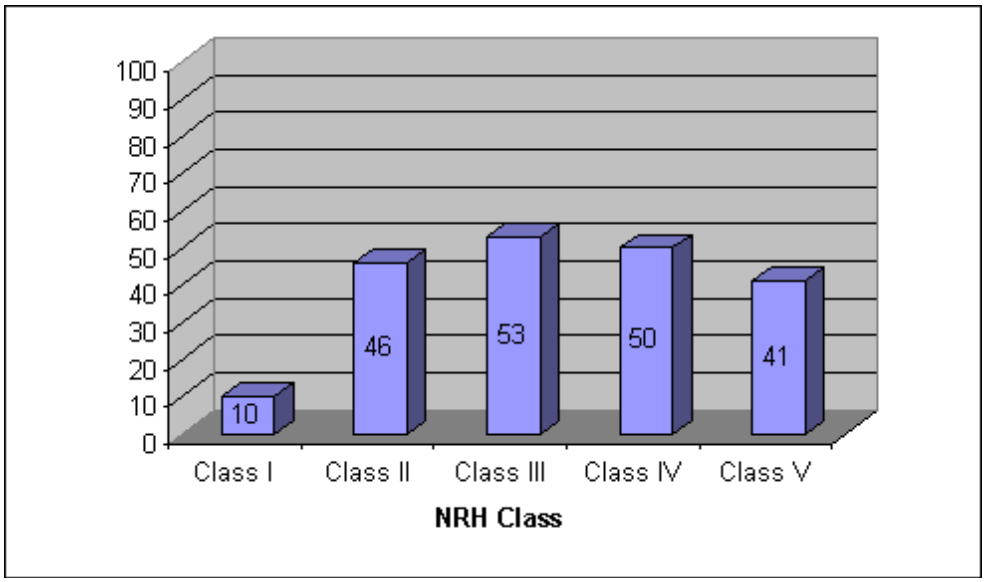

FIGURE 11. Distribution of lower extremity limbs by NRH Limb Classification in 100 consecutive post-polio patients (N = 200). (Ref. 41.)

B. Fatigue.

1. Differential Diagnosis.

Fatigue is often a nonspecific complaint, with a variety of possible etiologies. The fatigue is sometimes focal but more typically generalized and is usually described as overwhelming exhaustion or flu-like aching accompanied by a marked change in the level of energy, endurance, and sometimes mental alertness. Frequently, patients will complain of fatigue when the problem is actually new weakness and vice versa. In a study of polio survivors and healthy controls, Berly found that both groups described fatigue as "tiredness and lack of energy", while polio survivors were significantly different from controls in describing their fatigue as "increasing physical weakness", "increasing loss of strength during exercise", and "heavy sensation of the muscles".*[116]* The fatigue usually occurs on a daily basis and progresses during the day. It is typically brought on by an accumulation of activities that had been carried out previously on a daily basis without special effort or noticeable sequelae. Late in the afternoon or early evening it peaks, and it is described by some people like "hitting a wall". When this occurs, it becomes necessary for individuals to stop what they are doing, rest, and, if possible, take a short nap. Of all the new health problems, this is often the most distressing because it imposes limits on people's lives without obvious objective changes that others can easily identify.

Fatigue that occurs after awakening may reflect sleep disturbances that are commonly due to musculoskeletal pain but may also reflect nocturnal pulmonary abnormalities. Fatigue that tends to last all day is atypical of PPS and should make one consider other diagnoses. The differential for fatigue is extensive but the considerations should include such disorders as other medical diseases, including anemia, chronic infections, collagen disorders, thyroid disease, diabetes, or cancer. Depression can also cause symptoms of fatigue. Fatigue can result from a number of medications, both prescription medications such as beta blockers or sedatives, or over-the-counter medications such as antihistamines. Deconditioning and obesity can also contribute to feelings of fatigue. Once these have been ruled out, then a careful evaluation of lifestyle is needed. Many patients are both active and competitive, awakening early, then working one job and sometimes two before coming home to work in the home and getting to sleep late. No matter what their vocation or avocation, many patients sustain a level of activity more strenuous than their strength and endurance allows. Because of disproportionate levels of activity, we have found it useful to have patients keep a diary both of their sleep and daytime activities, as well as the response to their schedule.

Bruno and colleagues have suggested that post-polio fatigue is caused by poliovirus-induced damage to the reticular activating system (RAS).*[117-119]* Acutely, drowsiness, lethargy, disorientation, and apathy were noted in poliovirus patients.*[120,121]* The reports of drowsiness, lethargy, fatigue, and fleeting attention in patients with acute polio virus are similar to the symptoms of decreased attention and fatigue that post-polio patients are now experiencing.*[119]* Such damage would impair the ability of the RAS to activate the cortex and would generate problems with attention and concentration that polio survivors describe as "brain fatigue". For this hypothesis to be supported, this fatigue should be associated with impaired function on neuropsychological tests that measure these functions. In a study of six patients, three with reports of severe fatigue, and three with no complaints of fatigue, Bruno demonstrated that those with severe fatigue showed significant deficits in their performance on measures of attention (double cancellation and trails making tests) and deficits in their ability to process complex information and sustain attention.*[118]*

Post-mortem histological studies of poliomyelitis victims have demonstrated the presence of poliovirus lesions in areas of the brainstem, including the midbrain reticular formation.*[122]* Bruno et al. performed MRI scans on 22 post-polio patients and in 8 of the subjects found areas of hyperintense signal in the periventricular and deep white matter, putamen, rostral reticular formation, and centrum semiovale.*[117]* All of these patients had complaints of severe fatigue. This is significantly different than what was found in aged-matched controls. Seven other patients with severe fatigue had normal MRIs and none of the patients without complaints of fatigue had abnormalities.

Thus, it appears that in some cases subjective complaints of fatigue can be correlated to both poorer function on certain aspects of neuropsychological testing and there may be anatomical correlations with areas of the

brain that have been affected by the polio virus. Although preliminary, these findings may have some practical clinical implications. While many of the lifestyle changes discussed later can be helpful to some, there may still be a group of people refractory to these energy conservation techniques. For this group, this hypothesis may be a way to gain a better understanding of their fatigue and in turn may lead to the development of more effective pharmacological treatment.

2. Treatment.

Many times a few lifestyle adjustments or education regarding energy conservation techniques will reduce this fatigue. These energy conservation techniques include the use of handicap license plates, planning to make as few as trips as possible, balancing activity and rest, sitting instead of standing, moving the location of supplies, and the use of a scooter or similar motorized vehicle when traveling a distance.*[96,123,124]* The importance of expecting reasonable demands and asking for help when needed is emphasized.

When these techniques do not help, a few medications have been introduced recently to help some of these symptoms. Cashman and Trojan have found success with the anticholinesterase inhibiter Mestinon. In a clinical trial with 17 PPS patients experiencing fatigue using jitter as a measure of electromyographic stability, they measured the response to 10 mg IV edronphonium and found seven patients (41%) had decreased jitter. It was unchanged in eight and increased in two. On the Hare fatigue scale, with a dose of 180 mg mestinon per day, nine patients (53%) noted improvement.*[125]* Clinical trials using the antiviral agent Amantadine were performed by Stein et al. using a placebo-controlled trial in 25 patients for 6 weeks.*[126]* These investigators used the visual analog and fatigue severity scale to measure the response. Their findings suggested that amantadine was not significantly better than placebo in controlling fatigue. However, three patients desired to continue the medication, and the investigators suggest that their scales may not be sensitive enough to detect a real change. Although not directly related to the treatment of fatigue, high-dose prednisone was tested to monitor the strength response in a group of 17 PPS patients.*[127]* Strength was measured on Medical Research Council (MRC) scale and by the Tufts Quantitative Neuromuscular Exam. There were no statistically significant changes, and three subjects withdrew from the study. All of these studies were well controlled, using objective, quantitative measurements. Unfortunately, fatigue is frequently subjective.

Recently, we have had success treating fatigue using low levels of tricyclic antidepressants (TCAs). Whereas these reports are only anecdotal so far, clinical trials will begin soon. The mechanism for this is not clear. Many patients report that they are sleeping better due to the pain relief and possibly the sedative effect of the TCA; however, there is possibly a direct effect on the serotinergic or norepinephrine neurotransmitters. This research demonstrates that pharmacological agents may play a part in the treatment of PPS patients. Further well-controlled studies using objective scales are needed on the use of medications in treating PPS symptoms, especially fatigue.

C. Pain.

1. Differential Diagnosis.

Chronic pain is the first or second most prevalent symptom in post-polio patients in most studies.*[72,74-77]* The differential diagnosis is extensive but should begin with conditions commonly associated with chronic musculoskeletal wear and tear such as osteoarthritis, bursitis, tendinitis, and myofascial pain. In addition, disorders that have significant muscle and/or joint manifestations should be excluded, such as polymyalgia rheumatica, fibromyalgia, polymyositis, and rheumatoid arthritis. Many of the problems that appear to be related to "overuse" of weak muscles, along with abnormal joint and limb biomechanics, may simply represent the inevitable consequences of chronic disability and be no more common in post-polios than they are in individuals with other neuromuscular diseases. In order to facilitate the diagnosis and treatment of pain, we have developed a classification that divides the pain syndromes into three categories.*[128]*

Type I pain or "post-polio muscle pain" (PPMP) occurs only in muscles affected by polio. It is either a deep or superficial aching pain that many patients say is similar to the muscle pain experienced during their acute illness years earlier. It is characterized by muscle cramps, fasciculations, or a crawling sensation. Typically, it occurs at night or the end of the day when the patient tries to relax. It is exacerbated by physical activity, stress, and cold temperature and is alleviated in part by the use of moist heat and slow stretching.

Type II pain or overuse pain includes injuries to the soft tissue, muscle, tendons, bursa, and ligaments. Common examples are rotator cuff tendinitis, deltoid bursitis, and myofascial pain (especially in muscle of the upper back and shoulders). Myofascial pain in post-polio patients is similar to that in other patients and is characterized by bands of taut muscles and discrete "trigger points" that elicit a "jump" response when palpated. These occur due to poor posture or improper body biomechanics. Fibromyalgia, with its associated symptoms, is another cause of muscle pain.

Type III pain or biomechanical pain presents as degenerative joint disease (DJD), low-back pain, and pain from nerve compression syndromes. Weakness induced by polio muscles as well as poor body mechanics makes the joints, especially in the lower extremities, more susceptible to the development of DJD, as years of ambulating on unstable joints and supporting tissue increases the energy expenditure to perform a given task. These costs accumulate silently over many years until they cross a critical threshold. In a study of 111 post-polio clients, Smith found that 100% demonstrated abnormal gait deviations, 40% demonstrated an uneven pelvic base, and 33% showed major trunk deviations.*[124]* In addition, the joints of the upper extremity, especially the wrist and shoulders, are prone to DJD when they assume a weight-bearing role, as occurs with the use of assistive devices (canes, crutches, walkers, or wheelchairs) over an extended length of time.*[129]* The joint pains are only rarely accompanied by swelling and/or inflammation, tenderness, and abnormal range of motion. X-rays of painful, weight-bearing joints may show degenerative changes that are proportional with the amount of stress the joints have sustained; however, even in seriously deformed joints, with the possible exception of the vertebral column, florid degenerative changes are uncommon. Autoimmune syndromes such as rheumatoid arthritis are uncommon; however, the prevalence in post-polio patients is similar to the general population, so if symptoms consistent with these arise, joint arthrocentesis or appropriate laboratory tests should be obtained.

Nerve compression syndromes, including CTS, ulnar mononeuropathy at the wrist or elbow, and cervical or lumbosacral radiculopathy, are syndromes that can cause pain as well as neurological deficits in the post-polio patient.*[94,130-132]* Risk factors for the development of focal neuropathies of the median and ulnar nerves at the wrist include use of an assistive device such as a cane, crutch, or wheelchair and the length of time that assistive device has been used.*[130,132]* These neuropathies can be detected on EMG/NCS even before the patient has symptoms characteristic of CTS.*[130]* In addition to these more common neuropathies, a number of less common ones have been described, including a tibial neuropathy at the ankle (due to compression of a poorly fitting brace), tarsal tunnel syndrome, and brachial plexopathy (associated with the use of axillary crutches).*[94]* While some of these patients are initially asymptomatic, most go on to develop pain and associated disabilities.

In order to study this further, we retrospectively reviewed the records of 40 consecutive patients seen in our post-polio clinic, gathering information about the type of pain experienced and its location.*[128]* In this group of 40 patients, 38/40 (95%) had pain complaints. Of the 38, seven (18%) had type I pain, 18 (47%) had type II pain, and 30 (77%) patients had type III pain. The location and type of pain for these patients is shown in Table 10. In another study by Smith and McDermott, the location of pain by method of locomotion for 114 patients was investigated. The results of this study are shown in Table 11.*[95]*

In general, patients who are ambulators develop degenerative joint disease in the lower extremities, while those who use wheelchairs or assistive devices for ambulation are prone to DJD or overuse syndromes in their upper extremities.

TABLE 10
Distribution of Complaints in Upper and Lower Extremities by Type of Pain in Forty Consecutive Post-Polio Patients (Ref. 128)

Extremity	PPMP Type I	Overuse Type II	Biomechanical Type III	Total (Percent) a
Upper	3	13	5	21 (53%)
Lower	5	2	24	31 (78%)

a Percents are greater than 100 since some patients have multiple pain complaints.

TABLE 11
Prevalence of Chronic Pain by Method of Locomotion in 114 Post-polio Patients (Ref. 95)

Method of locomotion	Population #	Pain #	Pain %
Ambulatory no brace (independent)	67	56	(84)
Ambulatory with brace (independent)	12	11	(92)
Ambulatory with crutches (independent)	21	21	(100)
Wheelchair locomotion (independent)	7	7	(100)
Wheelchair locomotion (need personal assistance)	7	7	(100)
Total	114	102	(90)

2. Treatment.

Pain management in post-polio patients is based on a few basic principles, which can be supplemented by class specific recommendations. These basic principles include: (1) improve abnormal body mechanics, (2) correct and minimize postural and gait deviations mechanically, (3) relieve or support weakened muscles and joints, (4) promote lifestyle modifications, and (5) decrease the abnormally high workload of muscles relative to their limited capacity.*[71,124]* Treatment for type I pain (PPMP) includes periodic rest, stretching, and heat. Stretching may have a role in maintaining the extensibility of muscle and connective tissue; however, it must be performed judiciously, as there are situations in which the patient may derive greater functional benefit and be safer with tighter tendons and reduced joint range of motion.*[135]* A variety of medications have been used to treat PPMP, but most common ones such as aspirin and other NSAIDS, acetaminophen, and narcotics are of little use. We have had success with the use of muscle relaxants in the benzodiazepam class, such as diazepam or TCAs, especially amitriptyline, a medication used commonly for the treatment of chronic pain.*[136]*

Treatment for type II pain (overuse pain) includes modification in extremity use, followed by modalities such

as ice, heat or ultrasound, TENS, and occasionally NSAID medications. Treatment for myofascial pain consists of myofascial release techniques, including spray and stretch and trigger point injections. Many times rest is not possible because many patients rely on their upper extremities for both locomotion and self-care. In rare cases, steroid injection or surgery may be needed.

Treatment for type III pain (biomechanical pain) includes posture and back care education and decreased weight-bearing or stress through use of assistive devices such as braces, crutches, wheelchairs, and scooters. Abnormal biomechanics can often be modified with fairly simple and practical interventions such as cervical pillows, lumbar rolls, gluteal pads, dorsal-lumbar corsets, and heel lifts. This pain is usually improved by conservative measures aimed at reducing mechanical stress, pacing activities, supporting weakened muscles, stabilizing abnormal joint movements, and improving biomechanics of the body during common daily activities. We use antiinflammatory agents sparingly, and then only in low doses to supplement conservative measures. In particular, efforts should be directed at improving routine activities that occur on a daily basis such as sitting, standing, walking, and sleeping, as well as any repetitious activities at work. Weight bearing with the wrist hyperextended and radially deviated should be avoided.*[133]*

For patients with carpal tunnel syndrome who must use a cane or crutch, we prescribe an Ortho-ease or "pistol" grip to place the wrist in a more neutral position and increase the weightbearing surface of the palm. Providing adequate support for weakened muscles and unstable joints can often be a difficult challenge; however, the basic orthotic principles are similar to those used in the management of other neuromuscular diseases. For patients with low back pain, lumbosacral corsets, a shoe lift and pelvic lift can help improve biomechanics.*[134]* For genu recurvatum or genu valgus due to quadriceps weakness or ligamentous instability, a KAFO with a free ankle and an extension stop at the knee is used. Those with dorsiflexor weakness or ankle instability can benefit from an athletic ankle splint, high-top shoes, or an AFO.*[95]* Many patients need an orthosis that combines strength and lightness. The new plastics and lightweight metals can often be used alone or in combination, but, unfortunately, the most functional solution may not be cosmetically acceptable. Frequently, patients prefer to repair and use their old braces than to start over again with new ones. Others may resist using any kind of brace for cosmetic and psychological reasons. In a study of 104 postpolio patients, Waring et al., prescribed lower extremity orthoses for 36 patients.*[134]* Orthotics were recommended for the following: (1) improve safety by reducing the risk of falls, (2) reduce pain, (3) decrease fatigue by improving gait speed and symmetry. Subjects who used orthoses reported significant improvements in pain relief, especially at the knee.

Reductions in stress, activity, and weight are lifestyle changes that have the most impact on reducing pain. These strategies may be the most difficult to accomplish, however, because they often require developing behaviors very unlike the old, familiar ways of coping. Essential is altering the pace and intensity of discretionary activities and learning new ways to gain more control over when and how activities are performed. Functional restoration as well as relief of pain is accomplished with the use of an interdisciplinary team, including physical therapists, occupational therapists, psychologists, rehabilitation engineers, and physicians.

D. Cold Intolerance.

1. Differential Diagnosis.

In addition to cold intolerance, patients may also develop color changes ranging from cyanosis to violet color and blanching of the affected extremity, flushing, and hot and cold flashes.*[137,138]* This can be accompanied by hyperesthesia, burning pain, and decreased manual dexterity.*[139]* These symptoms may be due to reduced blood flow through areas of atrophic muscle but may also be related to sympathetic intermediolateral column damage to vasoconstrictor neurons at the time of the acute poliovirus infection. This allows passive dilation of cutaneous venous capacitance beds, producing venous congestion and decreased arterial inflow to the skin. Loss of musculature due to atrophy and the dependent position further hinders

return of venous blood to the heart, causing edema. Cutaneous venous pooling combined with a decrease in warm blood flowing to subcutaneous tissues causes heat loss and cooling of the extremity. The limb then becomes resistant to warming as the sensitivity of the alpha adrenoceptors increase, accentuating vasoconstriction. Because the extremity is cold, the rate of nerve conduction is slower, although the amplitude of the sensory-evoked potential may be large. Bruno found that polio subjects were more sensitive to electrical stimuli in both affected and apparently unaffected extremities when compared with controls.*[138]*

2. Treatment.

At the present time, treatment for cold extremities is largely symptomatic, with the use of multiple layers of clothes, especially in the extremities. Patients report relief with the use of nylon panty hose and woolen long underwear, even in warmer weather.

E. Respiratory Complications

1. Differential Diagnosis.

During the acute phase of polio the most feared complication was impaired respiratory function. The prevalence of patients who developed respiratory compromise requiring the use of an iron lung was estimated to be 15%.*[140]* Now, 30 to 40 years later, patients with initial respiratory weakness may develop new difficulty with breathing, especially at night or with exertion. Several preliminary studies suggest that as many as 18 to 38% of polio survivors who were successfully weaned off a respirator after acute illness now require ventilatory assistance full or part time.*[141]*

In a national study of the late effects of polio, Halstead reported that 42% of respondents reported new problems with breathing.*[74]* Even with improvement in respiratory care, the difference between the prevalence of acute problems (15%) and late effects (42%) suggests either subclinical involvement initially or the combined effects of other cardiopulmonary disease and deconditioning.

More than half of patients interviewed 21 to 30 years after acute polio reported that the respiratory treatment needed had not changed since one year after polio onset, 27% believed impairment had worsened, and 17% felt it had improved; only four needed more daily respiratory support than formerly.*[141]* The persons at greatest risk for serious late onset pulmonary complications had moderate to severe respiratory involvement initially and usually required ventilatory assistance. The other group at risk are those with severe spine deformities: scoliosis and kyphoscoliosis. While the long-term prognosis for those who were never weaned completely from a ventilator appears favorable, both respiratory problems and dysphagia are potentially life-threatening and should be managed as such.

Pulmonary problems include both obstructive and restrictive lung disease, with symptoms of exertional dyspnea, sleep apnea, and reduced pulmonary endurance. These are due to weakness of respiratory musculature, chronic alveolar hypoventilation, increased scoliosis, decreased pulmonary compliance, and effects of smoking or other diseases, such as asthma.

In a study looking at the prevalence of symptoms suggestive of sleep-disordered breathing, Fischer found that in a group of 155 polio patients, 59% woke frequently, 39% snored, and 41% reported daytime fatigue, significantly different from his 90 controls, where the figures were 8, 8 and 6%, respectively.*[142]*

The pathophysiology of late-onset pulmonary dysfunction frequently results from complications of chronic alveolar hypoventilation (CAH) and sleep-disordered breathing.*[143]* CAH is a restrictive disorder caused as a result of inspiratory respiratory muscle weakness, scoliosis, and, often, obesity. There is a loss of vital capacity and ventilatory insufficiency, with subsequent hypercapnia and hypoxia. Patients with expiratory muscle weakness can have difficulty clearing secretions, especially during upper-respiratory infections. This

can lead to mucous plugging, ventilation/perfusion imbalance, atelectasis, pneumonia, and pulmonary scarring. Mucous plugs can also cause sudden hypoxia and respiratory failure. Proper evaluation for those with complaints of respiratory difficulties begins with a comprehensive history and physical. This includes remote history of both the need for respiratory support and length of time it was needed, information on the use of tobacco, past medical history regarding pulmonary diseases, including upper respiratory infections, asthma, or chronic obstructive pulmonary disease (COPD). In addition, a review of symptoms should include questions regarding snoring or nighttime awakening, and daytime sleepiness, headaches, muscle weakness, dyspnea, fatigue, changes in body weight, impaired cognition, cyanosis, irritability, anxiety, depression, and decreased cognition, all which are possible characteristics of CAH. On physical examination, the auscultation of lungs in both inspiration and forced expiration should be done, noting any wheezes or evidence of obstructive lung disease. The degree of scoliosis and obesity in the abdomen can help determine the possible lung volume.

A screening PET, including vital capacity (VC), tidal volume (TV), and forced expiratory volume (FEV1) in both a sitting and supine position should be performed as part of the evaluation, as these tests may reveal unsuspected problems only in the supine position. If the initial VC is less than 50% of predicted or less than 1500 ml, more complete testing should be done. This may include repeat PFTs, including a measurement of the maximum volume of air that can be held with a closed glottis. This is a function of pulmonary compliance and the strength of bulbar musculature. For those with evidence of obstructive disease such as COPD, PETs with a bronchodilator can be done. At times it is necessary to obtain an arterial blood gas (ABG) to measure O_2, CO_2 saturation, and Ph.*[144]* Sleep-disordered breathing, including central or obstructive apnea, occur in the post-polio population to a greater degree than non-polios. Sleep-disordered breathing can result in hypoxia, right ventricular strain, and, finally, cardiopulmonary failure. When not corrected, insidiously progressive hypercapnia leads to a compensatory metabolic alkalosis. The resulting central nervous system bicarbonate levels can lead to depression of the ventilatory response to the hypercapnia and hypoxia and worsening of the CAH.*[144]*

When a nocturnal sleep disorder is suspected, overnight oxyhemoglobin saturation monitoring using a pulse or ear oximeter should be performed.*[145]* In some cases, polysomnography may be needed.*[146,147]* Transcutaneous CO_2 sleep studies can be performed with CO_2-sensing electrodes. A CO_2 greater than 50 mmHg or $\%O_2$ less than 95% for 1 h or more in a patient with a VC less than 50% predicted is diagnostic of CAH. For symptomatic patients with VC greater than 50% predicted, and inconclusive sleep studies, sleep-disordered breathing and inspiratory muscle weakness may be responsible for symptoms.

2. Treatment.

Both sleep-disordered breathing and CAH can be reversed and symptoms improved with the initiation of ventilatory assistance. Inspiratory positive pressure ventilation (IPPV) can be delivered in many ways. Continuous positive airway pressure (CPAP) or bilevel positive pressure airway pressure (Bi-pap), which independently varies the inspiratory (IPAP) and expiratory (EPAP) pressures, are both suitable alternatives. These can be delivered via an oral, nasal, or oral-nasal ventilator hose.*[148,149]* Because of the development of newer noninvasive methods of delivering positive pressure, a tracheostomy is seldom needed and should be avoided if at all possible, due to the high rate of complications, including vocal cord paralysis, endobronchial intubation, and laryngeal or tracheal stenosis.*[150]* Negative pressure body ventilator (NPBVs) such as the iron lung, the Porta-lung, and the chest shell currais are still options for some patients. Drawbacks to these include sleep interference, poor portability, and a high occurrence of apnea, hypoxia, and hypercapnia. Ventilators that work directly on the body are the intermittent abdominal pressure ventilator and the rocking bed. These have some of the difficulties of NPBVs and are generally less effective. Finally, frog breathing or glossopharyngeal breathing (GPB), a method of using tongue and pharyngeal muscles to project a bolus of air past the vocal cords into the lungs can be used. Immediate vocal cord closure traps the bolus in the bronchial system. Bach et al. studied 49 patients trained in GPB.*[151]* Of those 49 patients who were taught, 26 used it

while speaking to keep a more consistent volume and duration despite a mechanical ventilator. Ten used it when changing mechanical breathing aids and 13 did not practice or use the technique. Using this method, people can take up to 200 cc of air per bolus, for up to almost 60 boluses, for a total of up to 3 liters per "breath". Oxygen therapy alone, without maintaining adequate insufflation, can worsen hypoventilation and hypercapnia. This increases pulmonary compliance and, ultimately, respiratory arrest.*[144]*

Additional considerations include the use of assisted coughing to help clear airway secretions in those who lack sufficient expiratory musculature to do so.*[152]* Mechanically assisted coughing is more efficient and less labor-intensive, and should be used when manual techniques are not able to generate 5 l/s of peak cough expiratory flow (PCEF). Either manual or mechanical chest percussion can help with atelectasis or mobilization of secretions. All patients with impaired pulmonary function or a history of recurrent respiratory infections should receive influenza vaccines on a regular basis and Pneumovax at least once. Obstructive pulmonary diseases can be treated with combinations of aerosol bronchodilators, theophylline, and periodic corticosteroids.

F. Psychosocial.

1. Differential Diagnosis.

To understand some of the psychosocial responses of many post-polio survivors, it is useful to explore the feelings they may have experienced during the initial polio experience. With the onset of paralysis, many individuals believed they were stricken by "the feared disease".*[153]* They were placed in isolation, and, if paralysis was severe, they came to count on others for basic needs, losing a sense of autonomy, and in many cases their identity. Many polio victims learned to deal with loss of control, pain, and fear by submitting to those in control, complying fully with external expectations and denying personal needs, physical and emotional pain, and even their own individuality. Bruno describes some of the behaviors they were expected to show, including "listen to the doctors, obey the nurses, do not be bad, and be good in school.*[153]* Some people believe that many of these survivors may have continued to follow these "good rules for behavior" after returning to the community.

Many health-care workers have commented on the existence of a "polio personality".*[154]* Whether this was a function of social circumstances as described above, the individual's response to the disease, or represented some kind of natural selection also associated with certain behavioral characteristics is unknown. Whereas the behaviors learned in dealing with this illness may have varied from individual to individual, they were behaviors that helped each individual survive -- which is one of the reasons so many polio patients call themselves "survivors". There does appear to be certain coping mechanisms many survivors share. These include independence, patience, perseverance, creativity, industrious behavior, detachment, and denial of limitations.*[71]* One of the hardest issues most post-polio survivors face is the fact that they are now faced with a second disability. Years after they felt they beat polio, recovering strength and function, they must now deal with recurrence of new symptoms, with the knowledge that the same strategies they used before may be detrimental. Most of all they must deal with the uncertainty regarding the etiology of their symptoms and their prognosis. As a group, polio survivors tend to be competent, hard-driving, time-conscious high achievers who demand high standards of themselves and others and tend to perform at high levels in many areas. For example, it has been reported that they are employed full-time at four times the rate of the general disabled population,*[77]* they have more years of formal education on average than the general able-bodied population,*[153]* and they take on marriage and family responsibilities at approximately the same rate as persons who are not disabled.*[155]* Further, many of these individuals overcame a serious and often life-changing illness. Because they were successful once, these same behaviors tend to emerge later in life in coping with other challenges and illness.

It is believed by some that post-polio survivors frequently exhibit "type A" behavior. In a study done by Bruno and Frick of 676 post-polio subjects, they concluded "type A" behavior was more prevalent than in a

nondisabled control population, and the "type A" score was higher in respondents reporting muscle pain and fatigue when compared with subjects without these complaints.*[155]* They found the polio group exhibited a high rate of symptoms associated with chronic stress, which they felt may have initiated or exacerbated some of the new health problems. To further evaluate psychological traits of post-polio survivors, Conrady et al. tested individuals using a self-report inventory, The Symptom Check List 90-revised (SCL-90R).*[156]* Peak scores for men in both groups occurred at somatization, depression, anxiety, and phobia. Peak scores for women were at somatization, depression, anxiety, and psychoticism. In general, the tests documented significant psychological distress, especially depression and anxiety (60 to 65%), supporting the hypothesis that devaluation, depression, and isolation may occur with a second disability. However, the researchers are uncertain that the degree of distress experienced can be solely attributed to progressive decline in function, as their data did not show any correlation between psychological distress and physical decrement.

In another study done by Tate et al. using The Brief Symptom inventory (BSI), only 15% had elevated scores for depression.*[157]* These patients also reported their health was poor or very poor, with more complaints of pain and fatigue. They reported less satisfaction with their employment status and also demonstrated poorer coping strategies than nondepressed polio survivors. Studies of controls (non-polio patients) in the community show a prevalence of depression from 15 to 30%, and those with other disabilities or health problems have BSI scores similar to those with polio. The main differences between these studies is that Conrady recruited subjects from either a clinic population or a post-polio support group, while Tate sent out questionnaires to individuals registered in a statewide polio registry. Therefore, one might conclude that while the post-polio population as a whole demonstrate psychological profiles similar to others in the general population, especially those with health problems, the group of patients who seek out help from either professionals or through peer support groups have higher levels of psychological distress.

Emotional responses to experiencing new medical problems related to polio can be as traumatic and disabling as the physical problems. Although people may experience any combination of denial, anger, frustration, and hopelessness, post-polio patients generally exhibit one of three distinct categories of psychological responses: (1) those who do not regard themselves as handicapped, regardless of the extent of involvement and presence of obvious deformities; (2) those who feel disabled now, but who never did in the past, even during the acute illness; and (3) those who feel that, because they are experiencing polio for the second time, they are "twice cursed".

Polio survivors may resist making lifestyle changes to accommodate weakness, fatigue, and other post-polio symptoms because they have worked so hard to overcome the initial paralysis and achieved a high level of functional performance and personal fulfillment. They may no longer perceive themselves as handicapped and believe, even if some disability remains, that they have conquered polio; the long struggle with polio is over. Instead, new limitations unexpectedly and abruptly developed 25 to 50 years later. However, patients still expect to regain lost function and feel better by persevering and working harder when better advice may be to slow down. Compliance is a large problem with this population. Waring et al. reported that only 41% of patients sporadically used a prescribed brace, while 70% refused to use a recommended crutch or cane because "they didn't want to".*[134]* In Peach's study on compliance, reasons stated for not following through with recommendations included: (1) refusals to change job or lifestyle; (2) refusal to lose weight; (3) refusal to wear orthotic equipment; and (4) inability to change job or purchase equipment due to financial reasons.*[158]*

2. Treatment.

To overcome this combination of denial and a personal history of successful coping, an interdisciplinary approach is helpful. Bruno and colleagues describe an initial evaluation that includes a psychosocial evaluation and psychological studies, with administration of the Reinforcement Motivation Study (RMS) and the Beck Depression Inventory.*[153]* The treatment of PPS requires a team approach from a psychophysiologic perspective using techniques of multimodal therapy.

The use of a treatment log where patients document physical and emotional symptoms along with activities can help the treatment team design a plan to decrease behaviors that may cause physical symptoms. These include incorporating techniques for time and stress management, energy conservation, and using relaxation techniques. The team can help the patient modify old coping mechanisms that are familiar. For example, have the old brace repaired after the patient's first visit rather than prescribing a new one. Beginning with a minor, acceptable intervention may prepare the patient to make necessary major changes later. A patient who has been ambulatory for 35 years may reject buying a wheelchair but agree to use a cane or restrict wheelchair use to the airport. The wheelchair may become more acceptable, however, as the patient learns that the cane is helpful but insufficient to relieve symptoms. Polio survivors may also reject anything that publicly advertises their handicapped status. Changes that enable them to retain some sense of control, such as displaying a handicap placard on the dashboard when desired instead of getting handicapped license plates, may enhance compliance. Getting help through peer support groups may also be a way of coping with new disabilities, as well as being a form through which patients can get state-of-the-art information from professionals who treat patients as well as do research in the area of post-polio. It is also a way they can share advice and practical tips on how to improve independence and deal with the common inconveniences they face on a daily basis. Networking through groups such as the Polio Society and Gazette International Networking Institute (G-I-N-I) is also a way to help meet these needs.

G. Dysphagia.

1. Differential Diagnosis.

Difficulty with swallowing has been reported to be a significant complaint in 10 to 15% of all patients with acute poliomyelitis*[159]* and between 10 to 22% of all patients with PPS.*[160]* In some cases bulbar involvement may be mild, with complaints of "food sticking"; however, in more severe cases it can lead to aspiration pneumonia and asphyxia. Dysphagia results from bulbar involvement, more specifically in the nuclei of the IX, X and XII cranial nerves. Control of swallowing can be divided into three phases. In the early oral stage food is positioned in the mouth, masticated as needed, and formed into a bolus. The late oral phase involves moving the bolus posteriorly into the region of the anterior faucial pillars, which move medially around the base of the tongue. Phase 2 is the pharyngeal phase, where the swallow response is triggered and the bolus moves through the pharynx. The final stage, the esophageal phase, is when the bolus moves past the crico-pharyngeal membrane, where the epiglottis tilts backward as the vocal cords approximate, preventing aspiration. The bolus then proceeds down the esophagus through peristalsis.*[161]*

In a recent study by Sonies and Dalakas of 32 post-polio patients, 14 (44%) had symptoms of new swallowing difficulties.*[162]* Twelve of these patients (38%) had a history of prior bulbar involvement. New swallowing difficulties were noted by nine of the 12 patients with bulbar involvement and five of the remaining 20 with nonbulbar involvement. Detailed evaluation of these 32 patients, including ultrasonography and video fluoroscopy, revealed abnormalities in 31 patients, regardless of whether they were symptomatic or had a history of previous bulbar involvement. Signs of abnormalities frequently included tongue pumping, delayed esophageal motility, pooling in the valleculae, unilateral bolus transport through pharynx, uncontrolled bolus flow into pharynx, difficulty swallowing fluids, delayed pharyngeal constriction, esophageal reflux, and delayed initiation of swallowing reflux. Less common abnormalities included impaired tongue activity, pooling in the cricopharyngeal area, and nasal reflux. Trace aspiration was noted in only two subjects.

In a study of 109 post-polio patients, Coelho evaluated 21 who had complaints of difficulty swallowing.*[163]* Of those patients, 12 complained that foods got stuck, five had difficulty swallowing pills or dry foods, two complained of frequent choking, and two had complaints of coughing or tightness in the throat. These patients were evaluated using a modified barium swallow (MBS), which used video fluoroscopy to examine swallowing of liquids, paste, and crackers coated with barium. Twenty of these 21 patients had abnormal studies, including decreased pharyngeal transit in 81% of patients and decreased bolus control due to velar and lingual weakness. Twelve patients (57%) had mild involvement, six (29%) had moderate involvement,

and two (10%) were severely involved. Only one patient had a normal study. In addition, pulmonary function tests demonstrated abnormalities, including decreased peak expiratory flow (PEF) and maximum expiratory pressure (Pemax) in all but three subjects. Therefore, it was concluded that this combination made the patient at risk for aspiration. In another study of three symptomatic patients, abnormalities on videoflouroscopy included mild to moderate dysfunction in the pharyngeal phase, such as decreased peristalsis, pooling of liquids in the pyriform sinuses, and a delayed swallowing reflex.*[164]*

Buckholtz found additional abnormalities not necessarily related to a history of polio in 25 patients that he evaluated.*[165]* These included hiatal hernia in 11, pharyngeal pouches in eight, other neuromuscular disease in four, lateral diverticulum in two, and a Zenkers diverticulum and stricture in one patient. He reports common findings on physical examination may include asymmetric palate or pharynx, dysarthria or hoarseness, limb abnormalities scoliosis, and facial asymmetry.*[160]*

Finally, in a follow-up study, Sonies reevaluated 11 of her 32 original subjects 3 to 4 years later using similar clinical measures. All patients continued to show mild to moderate findings. Eight had no significant change and three got worse. In addition, one developed pneumonia. These observations suggest a variable progression of symptoms, regardless of patient awareness.*[166]* It is notable that abnormalities on videoflouroscopy can be seen in both symptomatic and asymptomatic patients, and in both those with and without a history of bulbar involvement, suggesting initial subclinical involvement.

2. Treatment.

For patients who present with symptoms of dysphagia, evaluation and management includes a thorough neurological exam, with particular attention to the evaluation of the cranial nerves, a screening pulmonary function test, and videoflouroscopy supervised by a speech language pathologist. If abnormalities are noted that place the patient at risk for aspiration, compensatory techniques include: (1) change the consistency of the food or liquid; (2) turn the head to one side; (3) tuck the chin; (4) alternate food and liquid; and (5) avoid eating when fatigued.

XII. PROGNOSIS.

Because all evidence suggests that the pathological processes involved are benign, post-polio syndrome is not life-threatening unless there is severe pulmonary involvement or a swallowing disorder. Dalakas et al. (1986) found an average loss of strength of 1% per year in 27 persons followed for a mean of 8.2 years.*[36]* This represents a rate of natural progression, as no one in the group studied was being treated to combat or modify the weakness. Although the long-term effects of such interventions remain to be studied, many clinicians have reported that patients who conscientiously adjust their lifestyles do improve, often increasing strength and stabilizing function. If weakness is, in part, due to overwork of the motor unit combined with musculoskeletal overuse, then interventions designed to reduce the metabolic demand on the overextended motor unit could conceivably alter this rate of decline. Munin, on the other hand, studied seven patients with quadriceps strength less than 5/5, new weakness and fatigue on one side and a contralateral leg without new weakness.*[167]* They tested strength objectively with an isokinetic, isometric protocol. They saw no decline on either side over a 4-year period. However, EMGs were not done to confirm AHCD, and the level activity was not monitored. Therefore, before conclusive statements can be made about prognosis, well-controlled, long-term studies need to be performed. In the meantime, we must use our best clinical judgement.

XIII. CONCLUSION.

We can only speculate about how these late complications will affect polio survivors; relatively few who survived the large epidemics of the 1940s and 1950s have reached their sixties and seventies, and post-polio problems have been widely recognized for study only since the early 1980s. Similarly, no group has been followed long enough to estimate the mean survival time. Future study of post-polio patients promises not

only to shed light on the many unanswered questions about pathological mechanisms, best forms of treatment, the role of exercise, and long-term prognosis, but also to reveal much about the aging process in other disabled groups. Furthermore, health-care workers and policy makers should keep in mind that thousands of people who survived paralytic polio will continue to survive well into the twenty-first century.

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