Post-Polio Sequelae

Assessment and Differential Diagnosis for Post-Polio Syndrome

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Proper assessment of post-polio patients presents both a challenge and a dilemma: a challenge because of the non-specific nature of many of the symptoms and the complex interplay between psychological and physical features; 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. Nonetheless, despite these obstacles, there is still much that can be done to ameliorate symptoms and improve function in the long-term management of these patients.

Some of the first questions clinicians ask concerning the assessment of post-polio patients are: what kinds of health and functional problems do they have; how frequently do they occur; and what are the best and most practical methods of evaluation?

In an effort to answer these questions, a review of the literature over the past decade is both helpful and misleading. Initial studies tended to summarize the results of self-report questionnaires.*[1,2]* While these were helpful in calling attention to the general types of new health problems being experienced by the postpolio population, they were misleading in several important respects: 1) they were clearly not an accurate reflection of the frequency of these problems among all polio survivors; and 2) there was no way to verify the accuracy or cause of the health problems reported.

Both of these limitations were addressed recently in a population-based study by Windebank et al*[3]* reporting on the polio-related health problems of persons in Olmsted County, Minn. Table 1 lists the new difficulties experienced by 50 subjects selected to undergo extensive examinations at the Mayo Clinic as part of a research protocol. Although these subjects were not chosen randomly, there was no systematic difference between this group by age, sex, or polio characteristics, and the rest of the Olmsted County

polio cohort. This study, therefore, is a milestone in our understanding of what health problems and functional limitations occur in the post polio population nationally. It is not, however, a reflection of the frequency or intensity of problems being seen in physicians' offices or the average post-polio clinic. For example, only seven of 50 (14%) of the Mayo Clinic subjects required new aids; only two of 50 (4%) made lifestyle changes because of their new polio problems; and none sought medical help for these problems.

Table 1

NEW DIFFICULTIES EXPERIENCED BY SUBJECTS WITH PRIOR PARALYTIC POLIO*

		Patients	
Symptom	No.	Requiring New Aids	Changing Activities
Fatigue alone	2	1	0
Pain alone	7§	0	1
Pain and fatigue	1	1	0
Weakness alone	4	0	0
Weakness and pain	9	2	0
Weakness and fatigue	2	1	0
Weakness, pain and fatigue	7	2	1
No new symptoms	18	0	0

* *Types of complaints reported by 32 of the 50 subjects with paralytic polio. Twenty-two complained of some new weakness. In seven subjects, the new symptoms necessitated the use of new aids to daily living and in two different cases, the symptoms had led to lifestyle changes.*
§ *All seven complained of nonradiating lumbar or cervical pain.*
Reprinted with permission: Windebank AJ et al. Late effects of paralytic poliomyelitis in Omsted County, Minnesota. Neurology. 1991; 41:507-507

By contrast, the experience reported by postpolio clinics is very different where, by definition, all are seeking medical help and many of the patients require new aids and are advised to make significant lifestyle changes. Table 2 lists the most common new health and functional problems of patients at two clinics.*[4,5]* The health problems included excessive fatigue, pain in muscles and/or joints, and new weakness in muscles previously affected by polio as well as muscles thought to be unaffected by the original illness. Less commonly, persons also complained of new muscle atrophy, breathing difficulties, and swallowing problems. The most common functional problems were increased difficulty with walking, climbing stairs, and dressing.

Table 2

MOST COMMON NEW HEALTH AND FUNCTIONAL PROBLEMS OF PATIENTS WITH

CONFIRMED POLIO EVALUATED IN TWO POST-POLIO CLINICS

	Texas* (N = 132)		Wisconsin§ (N = 79)	
	N	%	N	%
HEALTH PROBLEMS				
Fatigue	117	89	68	86
Muscle pain	93	71	68	86
Joint pain	93	71	61	77
Weakness:				
Previously affected muscles	91	69	63	80
Previously unaffected muscles	66	50	42	53
Combined	--	--	69	87
Atrophy	37	28	31	39
FUNCTIONAL PROBLEMS				
Walking	84	64	--	--
Climbing stairs	80	61	53	67
Dressing	23	17	13	16

* *Adapted from Halstead LS. Late complications of poliomyelitis. In: Goodgold J. ed Rehabilitation Medicine. St Louis, Mo: C. V. Mosby; 1988:322.*
§ *Adapted from Agre JC, Rodriques AA, Sperling KB. Symptoms and clinical impressions of patients seen in a postpolio clinic. Arch Phys Med Rehabil. 1989; 70:367-370.*

Assessment

Assumptions and Strategies. 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 close to 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. Our assumptions include the following:

1. Polio clinic attendance may be the first health care encounter for some patients in many years. For others, because of enduring negative memories of their original treatment with polio, it may be their first health encounter ever concerning polio-related problems since the acute illness.
2. Many patients--perhaps the majority--who are seen in the clinic arrive after a considerable period of

indecision and ambivalence. They want information and help, but are not sure they want to deal with what they anticipate will be difficult choices and unpleasant news.
3. These patients have generally not been treated by health professionals who are comfortable or conversant with polio either as an acute paralytic illness or a chronic disabling condition with delayed manifestations.
4. Many patients have not had contacts with health professionals who understand the rather unique experience of some polio survivors who are confronting their limitations for the first time ("first disability") or others, who after a long period of adjustment and the onset of new symptoms, are now dealing with what they feel is a "second disability."
5. Older patients, because of their chronologic or physiologic age, are dealing simultaneously with issues of aging and disability, both separately and collectively

Based on these assumptions, we have developed an outpatient post-polio program at the National Rehabilitation Hospital which has the following elements:

1. *Comprehensive assessment*. Because of the number, diversity, and complexity of the problems presented by these patients, we have found that a comprehensive, coordinated assessment is required.
2. *Interdisciplinary Team*. The best way to provide a comprehensive, coordinated evaluation that looks at the medical, functional, and psychosocial/vocational issues of this population is to use an interdisciplinary team of persons with special expertise in disability and knowledge of polio.
3. *One stop shopping*. In an outpatient clinic, the kind of assessment described in numbers 1 and 2 above is best carried out in one setting where the patient encounters all members of the interdisciplinary team in a sequential fashion over a relatively short interval of 2 to 3 days, as opposed to a piecemeal, intermittent evaluation carried out over many days or weeks in multiple settings. This kind of structure is convenient and efficient as well as cost effective for the patient. For the staff it provides the opportunity for frequent face-to-face communication while the assessment is in progress, and on the spot, cross-discipline consultation which draws on the special interests and expertise of individual members.
4. *A dimension of time*. An assessment which extends over 2 to 3 days (rather than a single morning or afternoon) introduces a dimension of time. We find this allows the patient an opportunity to integrate the evaluation process, clarify questions, and begin to deal more realistically with some of the implications of a change in health and functional status. This, in turn, seems to make the patients more amenable to therapeutic recommendations.
5. *Diagnosis by exclusion*. Because post-polio related complications are diagnosed by *exclusion*, it is essential that every patient receive a careful history and physical exam, along with appropriate laboratory studies, radiographs, and diagnostic tests to rule out other medical, orthopedic, or neurologic conditions that might be causing or aggravating the presenting symptoms.
6. *The expectation of improvement*. We believe that everyone who comes to the clinic can be helped regardless of the underlying 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. By the same token, there is no cure, and every improvement is dependent on patient commitment and cooperation.

A typical evaluation in our clinic extends over 2½ days, with the first day reserved for evaluations by team members who include a 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 in the history to 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 and of the use and need for orthoses and other durable medical equipment. In addition, the physician determines the need for radiograph, laboratory, and electrodiagnostic studies, and initiates referrals to other rehabilitation disciplines (eg, nutritionist, vocational counselor, psychologist) as well as other medical and/or surgical specialists as needed.

The physical therapist's evaluation is based on a protocol outlined by Smith,*[6]* 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, sleeping, and walking. 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,*[7]* and includes an evaluation of activities that produce pain, weakness, or fatigue, when symptoms occur, and how they interfere with the person's roles. 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 of 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 or 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.*[8,9]* In addition, the social worker facilitates referrals and access to community resources and services, including the local post-polio support group.

On the second day, we obtain any necessary laboratory studies, radiographs, and diagnostic tests to help rule out other medical, orthopedic, or neurologic conditions that might be causing or aggravating the patient's presenting symptoms.

For nearly every patient, we feel a standard electromyogram/nerve conduction study (EMG/ NCS) is essential to confirm the presence of an old anterior horn cell (AHC) disease, identify major muscle groups with subclinical involvement, establish a baseline, and help exclude certain other neurologic and myopathic conditions. We do not believe more sophisticated studies with single fiber EMG (SFEMG) or macro EMG are indicated at this time in the routine clinical setting, as they have not helped separate the symptomatic from the asymptomatic patient or proved useful in guiding clinical management.*[10]* The one exception to this is the report by Trojan et al,*[11]* which describes a subgroup of patients who have a positive IV Tensilon (edrophonium chloride) test during SFEMG and respond clinically to oral Mestinon (pyridostigmine bromide). The recommended dose is gradual increments of Mestinon up to 60 mg three times a day for patients who have new neurogenic weakness and can be monitored closely for response and side effects.

The preliminary results of a more recent study by Dr. Trojan (see GINI Press Release November 30th 1997) have unexpectedly thrown some doubts on the efficacy of Pyridostigmine in PPS. "Pyridostigmine was not found to provide significant benefits with respect to quality of life, fatigue, or isometric muscle strength [of patients presenting PPS symptoms] compared with placebo, although a trend was noted towards increased strength in very weak muscles."
LincsPPN Web Administration 20th March 1998.

A standard battery of screening tests, such as an SMA 24, thyroid panel, fasting glucose, etc, used on a routine basis are generally not helpful or cost effective, with the possible exception of creatine kinase (CK). In the Mayo Clinic study,[3] 10 of 32 symptomatic subjects (31%) had mild to moderately elevated CK levels, while none of the 18 asymptomatic subjects had abnormal levels. However, whether it is useful to monitor CK levels on a regular basis to assist in determining long-term prognosis or as an aid in clinical management is still not clear. In patients who are relatively inactive, overweight, or have a family history of coronary artery disease, we recommend a plasma lipid and lipoprotein concentration study. In a preliminary report of 64 symptomatic post-polio subjects, Agre et al[12] found hyperlipidemia in 16 of 24 men (66%) and 10 of 40 women (25%). There was no evidence to suggest that this finding is related to the history of polio; more likely it is a reflection of deconditioning and the unhealthy nutritional status found in many persons with musculoskeletal problems and mobility impairments.

Patients who had respiratory involvement initially and have a history of pulmonary disease or scoliosis have a screening vital capacity measured along with their other vital signs. If the vital capacity is less than the predicted 50% or the history and clinical situation warrant, pulmonary function studies are obtained along with arterial blood games (ABGs). If the patient has significant spinal curvature, a 36-in, gravity loaded scoliosis film is obtained to provide a baseline for follow-up exams. Patients with borderline or abnormal ABGs and/or respiratory symptoms are referred to a pulmonologist for further evaluation and management.

In addition to respiratory complications, swallowing difficulties may be more common than previously realized. In a recent study by Sonies and Dalakas[13] of 32 patients, 14 (44%) had symptoms of new swallowing difficulties, and of the 18 asymptomatic subjects, all but one had some abnormality on detailed testing of oropharyngeal function. Patients who present to our clinic with any swallowing difficulties are referred to a specialist for further evaluation and management.

Finally, the morning of the third day of each patient's assessment in our clinic is used to complete any unfinished evaluations and hold a team conference with the patient and 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 are 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 a history and physical.

Differential Diagnosis

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 non-specific. 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. A description of the most common

symptoms based on the histories of patients seen in our clinic and of those described in the literature is presented along with a description of the personality characteristics commonly found in polio patients.

Fatigue. 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. It is typically brought on by an accumulation of activities that previously were carried out on a daily basis without special effort or noticeable sequelae. Commonly, it occurs late in the afternoon or early evening and 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. Fatigue that occurs upon awakening may reflect sleep disturbances that are commonly due to musculoskeletal pain, but may also reflect sleep apnea. Fatigue that tends to last all day is atypical of post-polio syndrome and should make one consider other diagnoses.

The differential for fatigue is extensive, but the considerations should include such disorders as anemia, myasthenia gravis, chronic infections, hypothyroidism, collagen disorders, depression, and side effects of medications.

Pain. Post-polio pain occurs in muscles, joints, or both. The muscle pain is sometimes described as being "close to the surface" and is frequently associated with hypersensitivity and a sensation of "crawling" or "cramping," particularly at night. However it more often presents as a deep, aching pain that many patients say is similar to the muscle pain experienced during their acute illness years earlier. These muscle pains are often aggravated by physical activity, stress, and cold temperature. Joint pain is also commonly associated with specific physical activities such as weight bearing, but is only rarely accompanied by swelling and/or inflammation. The prevalence of pain by method of locomotion for 114 patients is shown in Table 3.*[14]* These pains are 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. Radiographs of painful, weight bearing joints typically show degenerative changes that are disproportionately mild compared to the amount of stress the joints have sustained, sometimes for decades. Even in seriously deformed joints, with the possible exception of the vertebral column, florid degenerative changes are uncommon.

The differential for muscle and joint pains is also extensive, but should begin with conditions commonly associated with chronic musculoskeletal wear and tear such as osteoarthritis, bursitis, tendonitis, and stress myalgia. In addition, disorders that have significant muscle and/or joint manifestations should be excluded, such as polymyalgia rheumatics, 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 polio survivors than they are in individuals with other neuromuscular diseases. If chronic overuse is the only or major underlying pathology present, then the conservative measures described in the previous paragraph can often slow or prevent further deterioration and possibly lead to a reversal of symptoms and improved function.

Table 3

PREVALENCE OF CHRONIC PAIN BY METHOD OF LOCOMOTION IN 114 POST-POLIO PATIENTS

Method of Locomotion	Number	Number (and Percent) With 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 assistant)	7	7 (100)
Total	114	102 (90)

Reprinted with permission from Smith LK, McDermott K. Addressing causes versus treating effects. In Halstead LS, Wiechers DO, eds. Research and Clinical Aspects of the Late Effects of Poliomyelitis. White Plains, NY: March of Dimes; 1987:122.

Weakness and functional loss. New weakness may appear in muscles previously affected and in muscles believed to be previously spared by polio. 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 had been marginal. One of the characteristics of many polio survivors was their ability to appear "normal" or function at an extraordinarily high level of performance on relatively few strong 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, 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 required to recover strength following activities is now increased. Persons with initial respiratory weakness may develop new difficulty with breathing, especially at night or with exertion. Patients who had swallowing difficulties with their original polio may develop new problems with swallowing or choking. Both respiratory and swallowing problems are potentially lifethreatening and should be managed as such.

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 muscle 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, eg, 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.

The workup for new neurogenic weakness should include an EMG/NCS. Although electrodiagnostic studies have their limitations even in the most skilled hands, they provide a necessary screening tool to help exclude some of the more common causes of neurogenic weakness in this population as well as other conditions that are relatively rare. 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.

In addition, every polio patient who presents with new weakness should be evaluated carefully to exclude amyotrophic lateral sclerosis (ALS). Although there are often a number of similarities, there are enough dissimilarities that the two usually can be distinguished without great difficulty. Over the years, there has been considerable speculation that an antecedent infection with polio might be associated with developing ALS later in life.*[15,16]* However, with the recent interest in the late complications of polio and the new understanding about some of the possible pathophysiologic mechanisms, it now seems likely that many of the patients who had polio and later were diagnosed as having a benign form of ALS were misdiagnosed. In a re-examination of five patients with a history of both polio and ALS, Brown and Patten*[16]* concluded that none of the patients would now be classified as having ALS, but rather as having post-polio syndrome.

Finally, older patients with new neurogenic weakness may be experiencing the effects of aging on the nervous system and the gradual loss of key giant anterior horn cells. For this reason, patients over 60 who present with a classic picture of post-polio syndrome are given a diagnosis of "new muscle weakness secondary to post-polio syndrome with an unknown contribution due to aging."

Polio personality characteristics. Many health care workers who were involved with large numbers of polio patients during the big epidemics in the 1940s and 50s have commented on the existence of a "polio personality".*[17]* Whether this was a function of social circumstances, the individual's response to the disease, or represented some kind of physiological predisposition to developing polio that is associated with certain behavioral chancteristics is unknown. However, it has been shown that there is a genetic predisposition to acquiring polio.*[18]* In addition, persons with polio 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,*[19]* they have more years of formal education on average than the general non-disabled population, and they take on marriage and family responsibilities at approximately the same rate as persons who are not disabled.*[20]* Further, many of these individuals overcame a serious and often life-changing illness.

While the behaviors that were learned in dealing with polio may have varied from individual to individual, these behaviors helped each individual survive--which is one of the reasons so many polio patients call themselves survivors. These behaviors included independence, perseverance, obstinacy, detachment, creativity, denial of limitations, etc. Because they were successful once, these same behaviors tend to emerge later in life in coping with other challenges and illness. As a group, polio survivors tend to be competent hard-driving, time-conscious, high achievers who demand high standards of themselves and others. In a questionnaire, Bruno and Frick*[20]* found that the mean Type A score in post-polio survivors

was significantly higher than that reported for a non-disabled control population. Furthermore, 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.

Diagnosis. The criteria for diagnosing postpolio syndrome are listed in Table 4. The first criterion of

diagnosing paralytic polio can almost always be confirmed with the following information: 1) a credible history of an acute, febrile illness resulting in motor loss and no sensory deficit; 2) the occurrence of a similar illness in family or neighborhood contacts; 3) the presence of focal, asymmetric weakness and/or atrophy on examination; 4) changes on EMG of chronic denervation with reinnervation compatible with prior anterior horn cell disease; and 5) examination of the original medical records whenever possible.

Table 4

CRITERIA FOR THE DIAGNOSIS OF POST-POLIO SYNDROME	
1.	A prior episode of paralytic polio confirmed by history, physical exam, and typical findings on EMG.
2.	Standard EMG evaluation demonstrates changes consistent with prior AHC disease: 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.
3.	A period of neurologic recovery followed by an extended interval of functional stability preceding the onset of new problems. The interval of neurologic and functional stability usually lasts 20 or more years.
4.	The gradual or abrupt onset of new neurogenic (non-disuse) 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.
5.	Exclusion of medical, orthopedic, and neurologic conditions that might cause the health problems listed in #4 above.

The second criterion is a characteristic pattern on EMG. The changes on routine EMG compatible with prior polio include increased amplitude and duration of motor unit action potentials, an increase in the percentage of often long duration polyphasic motor units and, in weak muscles, a decrease in the number of motor units on maximum recruitment. Occasionally, positive sharp waves and fibrillations are present and less commonly, fasciculations may be seen.

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 (Fig 1): 1) paralytic polio in childhood or later in life; 2) partial to fairly complete neurologic and functional recovery; and 3) a period of functional and neurologic stability lasting many years--usually 20 or more.

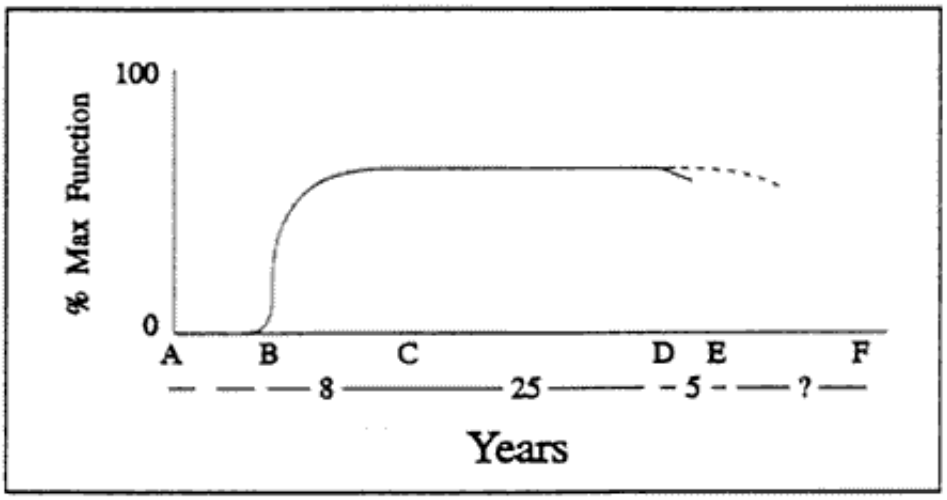

Fig 1: The natural history of polio based on data from patients evaluated in our post-polio clinic. A = birth; B = onset of polio; C = maximum recovery; D = onset of new health problems; E = time of evaluation; F = death.
Reprinted with permission of the March of Dimes Birth Defects Foundation.*[4]*

The fourth criterion is the onset of new neurologic (non-disuse) weakness which may come on either gradually or abruptly. New neurogenic weakness is essential for making the diagnosis of post-polio syndrome 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. 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.

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 post-polio syndrome. First, the symptoms are frequently so general that ruling out all possible causes is not practical and can be prohibitively expensive; and second, coexisting medical, orthopedic, and/or neurologic conditions may be present which can produce a similar set of overlapping signs and symptoms. As indicated in Figure 2, 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.

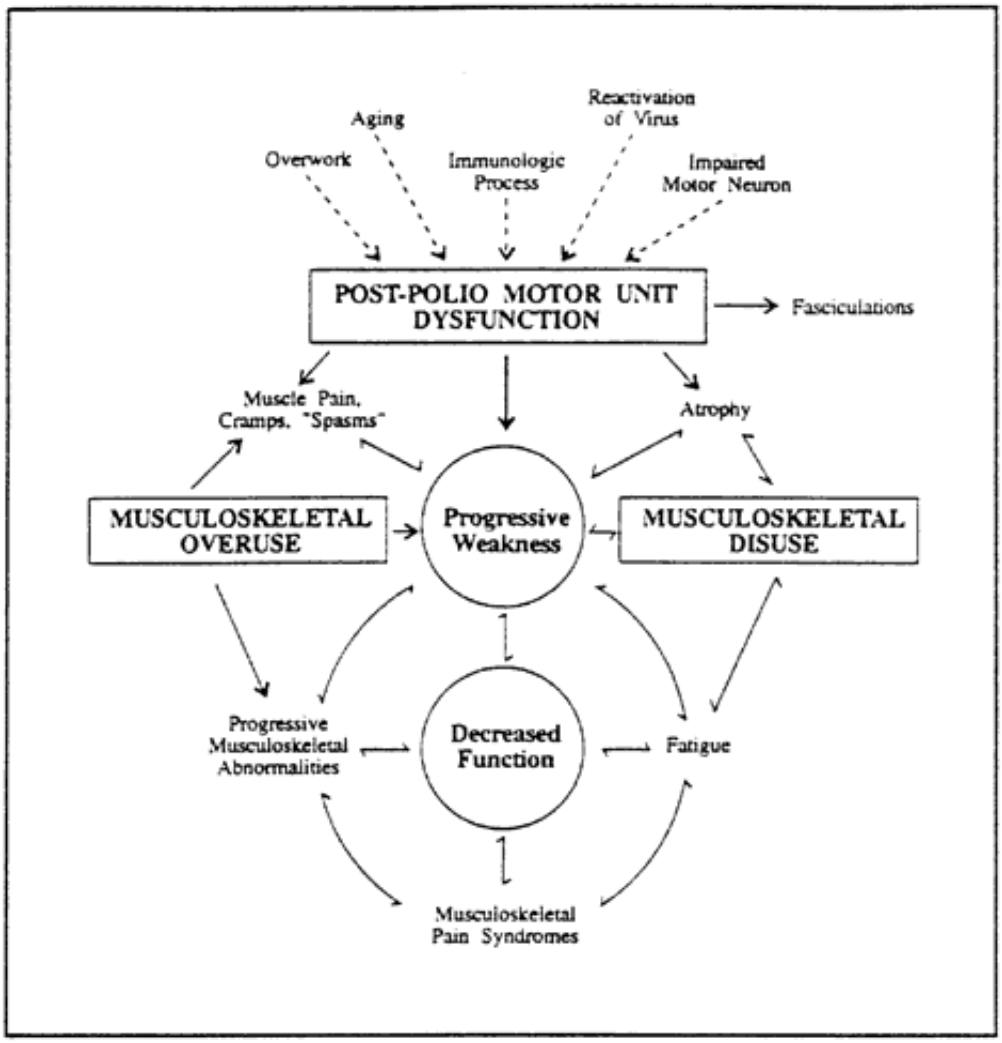

Fig 2: Schematic model showing three possible causes for the late neuromuscular and muskuloskeletal complications of polio and their interactions.

Nomenclature. There is no clear consensus about the most appropriate name or diagnostic label to use in post-polio patients with new health problems. A number of terms have been proposed, including post-polio syndrome, postpolio progressive muscular atrophy, and the late effects of polio. One of the reasons that none of these terms is suitable for all individuals is the lack of specificity in diagnostic criteria. This, in turn, is related 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 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.

At present, post-polio progressive muscular atrophy (PPMA) has the most specific criteria. The criteria for making the diagnosis of PPMA include the five items listed in Table 3 in addition to the requirement of new muscle atrophy. Changes on muscle biopsy in these patients show evidence of active denervation in the form of scattered angulated fibers.*[21]* An alternative term to PPMA is post-polio motor neuron disease (PPMND), which has the same meaning but has the advantage of being more generic and less

restrictive.

Post-polio syndrome 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 based on the assumption that the major pathologic process involves new motor unit dysfunction with a variable contribution from musculoskeletal overuse. For this reason, the presence of neurogenic weakness is considered a necessary finding to make this diagnosis. As new information becomes available about the underlying mechanism(s) which produce late onset complications, these criteria will undoubtedly change and new terminology be developed to fit our improved understanding.

Future Considerations

Until a definite test is available, post-polio syndrome will remain a diagnostic challenge that probably will become more difficult in coming years. It is true that the natural history and symptoms of post-polio syndrome are already widely known. However, as interest and knowledge about post-polio syndrome continue to grow and expand, polio survivors (and those who mistakenly believe they had polio) will become more sophisticated in matching their symptoms --either consciously or unconsciously-- with those described in the medical literature. If this becomes a widespread phenomenon, it could decrease the credibility of the diagnosis of post-polio syndrome with several possible unfortunate consequences: 1) it may make it more difficult to obtain disability retirement using post-polio syndrome as the primary diagnosis; and 2) it may cause third party payers to be less inclined to cover medical expenses associated with the diagnosis of post-polio syndrome. For these reasons, it is crucial that all patients continue to have a thorough evaluation and that the criteria for diagnosis outlined in Table 4 be followed.

Another problem concerns the confounding of the symptoms of post-polio syndrome with the effects of aging. The mean age of persons attending post-polio clinics around the country is in the mid to late 40s. As the polio population ages, there will be an increasing number of co-morbidities, and sorting out the cause of each symptom will be increasingly complicated. In the final analysis, our major hope lies with finding a diagnostic test that reflects the underlying pathology of post-polio syndrome which will provide a means of monitoring its natural history as well as the effect of interventions.

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