Managing the Late Effects of Polio from a Life-Course Perspective

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During the past decade many investigations have been conducted concerning the etiology of post-polio syndrome (PPS). The hypothesis proposed by Wiechers and Hubbell*[1]* that post-polio people were having a degenerative fragmentation of their giant motor units as a basis for progressive muscle weakness appears now to be substantiated by many electrophysiological and pathological studies. Although several recent investigations have explored possible mechanisms that initiate motor unit deterioration, most notably immunological and hormonal dysfunction results have been inconclusive and no effective cure for PPS has been found. Therefore, clinicians must focus on effective management.

This paper reviews the implications of recent research investigations for the management of patients with PPS. It proposes that current knowledge supports the view that PPS is a secondary condition frequently occurring during the life course of people with residual motor impairment from paralytic poliomyelitis and does not support the view that PPS is a distinct pathological process which should be labeled a disease or illness.

The Institute of Medicine recently proposed a model for understanding secondary disability development (Fig 1).*[2]* In this model a secondary condition is defined as any new condition that develops in the life course of a person with a primary disabling condition. Life-style/behavioral and biological risk factors interact with the primary disability to influence the process of further disablement from the secondary condition. New pathologies may progress to greater impairment, more functional limitation or additional disability. The process of secondary disablement then interacts with quality of life. The life-course paradigm for understanding secondary disablement emphasizes developmental and aging processes that can be expected during a person's life to increase chronic impairment and evolve into more functional limitation, thus requiring frequent readaptation to avoid additional disability.*[3]*

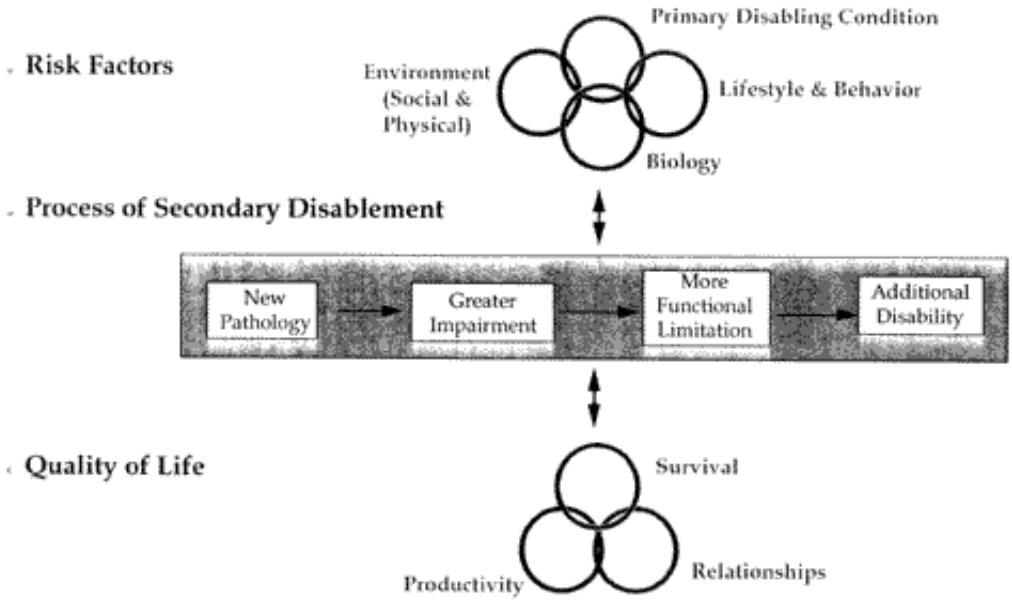

FIGURE 1. Schematic representation of a model for secondary disabilty development proposed by the Institute of Medicine.*[2]*

DIAGNOSTIC AND ETIOLOGICAL CONSIDERATIONS

If the life-course model of secondary disablement is applied to people with a past history of polio, PPS is a secondary condition that post-polio people are at risk of developing. Inasmuch as the diagnostic criteria for PPS are inherently subjective symptoms (pain, greater weakness, fatigue), PPS cannot be defined objectively, and individualized criteria must be used. A more quantifiable criterion for diagnosing PPS is the presence of a new, or greater degree of, functional limitation resulting from the level of symptoms that a person is experiencing. This definition of PPS permits a clinician to consider diagnosis and management of a person with a past history of polio holistically because many new health problems may occur during the life of a post polio person that affect the neuromusculoskeletal systems and functional abilities. Conditions that are "unrelated" as well as "related" to previous paralytic polio residuals affect function, and the consideration of what is "related" implies a subjective judgment about a causal relationship. The term "late effects of polio" may be preferable to PPS for describing specific new health conditions in which a causal relationship is likely (e.g., obstructive sleep apnea in a person who had bulbar polio and has pharyngeal muscle weakness; degenerative arthritis in a person who walked many years with a weak quadriceps muscle). Alternatively, PPS can be used when a causal relationship between current symptoms and chronic post-polio impairments is unclear, although a new functional limitation or disability has developed in association with the stereotypical symptoms of the syndrome.

It has been suggested that the life course of people with a past history of paralytic polio often includes a slow decline in muscle strength that may become sufficient to affect everyday functional activities. One long-term follow-up study documented a progressive decline in strength among persons with post-polio, as measured by serial manual muscle test scores.*[4]* Although this finding has not been substantiated with serial quantitative measurements of muscle strength over one or two years, declining strength is widely reported subjectively by polio survivors.*[5]* It has also been suggested that slow decline of strength may occur during the life course of muscles with chronic partial denervation and reinnervation from other conditions than polio, for example, partially innervated/denervated muscles at the zone of partial preservation after spinal cord injury, particularly when these muscles are used repetitively at near maximal capacity during daily activities.*[6]* This phenomenon of slowly increasing weakness and fatigue may represent accelerated aging of giant neuropathic motor units and becomes clinically relevant when a functional limitation results at the point when strength is insufficient to perform a customary or important

daily activity. Cashman has characterized new postpolio weakness as a syndrome of "delayed failure of reinnervation"*[7]* Therefore any new condition affecting a person's general health will steepen the slope of age-related decline of muscle strength during the life course of a post-polio person. This view is compatible with the clinical onset of PPS after the development of non-polio-related (co-morbid) medical conditions, including severe injuries. Although PPS then results from disuse atrophy, the weakness never is completely reversed because of the reduced capacity in the later life course of persons with post-polio to again reinnervate denervated muscle fibers and/or hypertrophy sarcomeres to preinjury levels.

Several findings from an in-depth study of 120 polio survivors, with and without new symptoms and with a spectrum of residual weakness and locomotor severity, support this view of PPS.*[8]* Significant correlations were found between new functional limitations and the following individual characteristics: (1) diagnosis of non-polio-related co-morbid conditions, (2) reduced cardiovascular fitness, (3) obesity, and (4) elevated cholesterol ratio.*[9]* Musculoskeletal problems of the upper and lower limbs were also highly prevalent (Table 1), and many were significantly correlated with the presence of new functional limitations.

TABLE 1. Prevalence of Secondary Muskuloskeletal Problems among 120 Polio Survivors[1]

Secondary Condition	Percent
Sensory loss in the hand	79
Median neuropathy at wrist	58
Ulnar neuropathy at wrist	24
Carpal tunnel syndrome	31
Hand or wrist arthritis (X-ray)	48
Hand weakness	56
Impaired hand dexterity	52
Upper limb joint pain	56
Lower limb joint pain	49
Spinal pain	50
Gait abnormality	59

[1] From Maynard *et al*[8]

PSYCHOLOGICAL CONSIDERATIONS

Beyond discussions over accuracy of terminology, there are other important reasons to adopt a life-course paradigm for evaluating PPS symptoms rather than a traditional medical paradigm. In the traditional medical paradigm a physician views new symptoms as resulting from a disease and a possible cure is implied. However, based on our current understanding of the pathophysiology of post-polio motor units, a real cure seems unlikely and the goal of medical care is to minimize and control decline of strength over the life course. In my experience when symptoms are cured, it usually results from treatment of co-morbid conditions or specific secondary conditions, for example, sleep apnea. If a physician finds no specific secondary condition and communicates uncertainty over the diagnosis for symptoms, patients may become fearful. If it is suggested that symptoms have a psychoneurotic basis, patients may become angry. If symptoms are minimized by attributing them to "getting old," depression or anger can result. If a cure for pathology is sought but not found, withdrawal and depression can be expected. If physicians communicate helplessness, patients will feel hopeless. People can feel victimized by the disease and the health care system that fails them.

In the life-course paradigm for evaluating secondary disability the physician considers the development of PPS as a not-unexpected event in the life of a person with a history of polio. Physicians are reminded by this view to examine first a person's general health and well-being and to carefully consider the impact of co-morbid conditions that may have developed or need to be diagnosed. An attitude of health promotion and risk reduction for all health problems is encouraged in the patient. If current weakness has created increased functional limitation or disability, then attributing it to the expected life course and aging of a person with chronic motor impairment will promote an attitude of knowledge-seeking and coping in the patient -- just as able-bodied people want to learn more about normal aging. Post-polio patients are asked to examine their life-styles closely and to reconsider their expectations for activity performance after an objective analysis of their muscle strength and endurance. Life-style modifications, including activity pacing, planned exercise regimes, dietary change, and stress reduction are a few of the behaviors that must be nurtured.

Recent research in psychology suggests that among people with spinal cord injury a person's attribution for the cause of their injury was highly predictive of successful coping.*[10]* In a similar way, attribution theory may influence the way a person with post-polio copes with new disability. For example, a person with long-standing moderate leg weakness from paralytic polio who has walked with two crutches for 30 years develops painful arthritis or tendonitis of the shoulders which limits walking and leads to greater arm weakness and more shoulder strain. These strain-induced shoulder problems could be considered to not unexpectedly result from prolonged, high-frequency heavy loading of tissues which show normative age-related reduced capacity for heavy loading without strain. Alternatively, one could attribute these shoulder strain problems to the onset of progressive weakening of muscles that is unique to people with a previous history of polio, or PPS. In the first view, symptoms are considered to result from age-related normative changes in organs (shoulders) that can be expected to have characteristic manifestations (degenerative arthritis/tendonitis) and functional implications (greater difficulty walking) among people with chronic motor impairments (walkers with crutches). In other words, problems are attributed to not unexpected life course changes. In the second explanation for the cause of shoulder problems, symptoms are attributed to condition-specific (post-polio) rates and types of changes in specific organ systems (nerve and muscle) that result in new functional limitations (PPS). The latter explanation can lead to a "self-fulfilling prophesy," or nocebo effect, that predicts worsening because no attribution is made to a person`s present or past activity that is under their own control. Psychological distress may be greater with the latter explanation than the former because it implies that PPS is a new disease with an unpredictable likelihood of rapid progression. Some people, particularly those who are relatively young (e.g., in their

early forties) may be more distressed by an explanation that attributes early aging, rather than a new disease, because the notion of growing old threatens their self-image of vitality and implies acquiring other disability characteristics of older people. For other people, attributing the problem to overuse strain may evoke guilt because of their own role in causing it or anger at health professionals who did not warn them about its future likelihood. Although the true cause of these problems may include both explanations, what may be most important for successful coping is an individuals attribution and/or understanding of the cause. The role of the physician in helping patients to understand their condition may strongly influence their attribution and subsequent coping. This role can be very challenging if done in a way that promotes successful coping among post-polio patients.

In a recent study that compared distressed/depressed to nondistressed/nondepressed polio survivors, the former were found more likely to be living alone, to be experiencing new health problems, to seek professional help, to view their health as poor, to report greater pain, to be less satisfied with their occupational status and life in general, and to exhibit poorer coping skills in relation to their disability. Coping factors associated with nondistressed/nondepressed subjects included positive self-acceptance, information seeking/sharing about disability, and social activism.*[11]* Therefore, conceptualization of PPS as an event in the life course of people with chronic post-polio motor impairments, rather than as an acute illness, may help promote successful coping. Writing in a Hastings Center report on ethical challenges in chronic illness, Jennings states that within the acute illness model, illness is viewed as an "alien threat to the self and the goal is to defend and restore the self by curing or compensating for the illness."*[12]* According to the acute illness model, the provider and patient enter into a contract wherein the goal is to defeat the enemy, that is, the illness. Restoration of functioning to a premorbid status is the aim, via destruction of the enemy within, to eventuate a cure. Inherent within the acute medical model is the traditional doctor-patient relationship, wherein the role of the patient is typically passive in nature. Care for people with chronic disabling conditions should be directed toward the minimization of the impact of any further disablement. Diplomacy becomes a better metaphor than warfare for appropriate medical care. If successful living with a chronic disability is a process of negotiation, the role of medicine is to facilitate that process.

CLINICAL EVALUATION AND MANAGEMENT

What then should a physician's approach be in treating patients with post-polio syndrome? It should begin with a comprehensive and detailed history that allows them to gain a life-course perspective on the person's functional abilities and activity patterns and on the severity of any residual polio impairments and functional limitations. This will enable a practitioner to consider all current symptoms in the appropriate context and to evaluate them in relation to current muscle strength, joint deformities, functional abilities, and other general health indicators. On the basis of the initial comprehensive assessment a further in-depth review of systems may be needed to consider the impact of all other health issues on musculoskeletal functioning and PPS symptomatology. The practitioner must adopt a primary-care-physician approach to the health of the person with postpolio and decide if a diagnostic workup is needed to identify other conditions that may be treatable and that may impact on the general health and functional capacity of the person. Some of the most common general health issues that must be considered include cardiopulmonary diseases that affect vitality and endurance; endocrine diseases such as hypothyroidism and diabetes; immunological abnormalities including allergic disorders and states of chronic stress; nocturnal hyperventilation and sleep apnea; dysphagia; neuromusculoskeletal problems including spinal disorders associated with spinal cord or nerve root impingement; peripheral nerve compression syndromes; myofascial pain syndromes; arthritis; and psychoemotional disorders such as depression and posttraumatic stress syndromes.

After all secondary conditions have been fully evaluated and optimally treated or managed, persons with post-polio must build a partnership with their physician based on effective communication and trust. Patients should receive all rehabilitative therapies that can assist them to improve functional limitations, reestablish mental and physical equilibrium in their lives, and adopt an attitude of negotiation with any residual new (secondary) disability. They should be informed about and offered adjunctive treatment options, such as exercise, drugs for fatigue or new assistive devices. Consultations with occupational therapists, social workers or psychologists, as well as participation in support groups, can be of additional assistance in considering beneficial changes in life-style.

Lastly, the physician needs to facilitate the patient's adoption of a health promotion and risk prevention attitude toward their health and life-style. After reviewing their vulnerability to progressive functional loss over the life course, patients should be encouraged to participate in wellness programs that can provide them with information and guide them in training behavioral changes that focus on nutrition, exercise, functional activity, posture, stress reduction, and coping. In summary, new scientific knowledge and advances about the late effects of polio will be used optimally in the lives of persons with post-polio only if applied with advanced skill in the art of medicine.

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