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Guest Editorial

POST-POLIO SEQUELAE: RESEARCH AND TREATMENT IN THE SECOND DECADE

Richard L. Bruno, PhD

In 1985, 5 years after tens of thousands of polio survivors began reporting late-onset problems, ORTHOPEDICS presented the first comprehensive presentation (Vol 8, No 7) on the causes and treatment of post-polio sequelae (PPS). In 1985, there was an understanding of what PPS were not, but there was no certainty about their pathophysiology or how to treat them effectively. Today, the articles presented in this issue and subsequently in the December issue present state-of-the-art information with respect to our understanding of the etiology and treatment of PPS.

TERMINOLOGY

One problem during the first decade of post-polio sequelae was the lack of a consistent terminology. For the sake of consistency, the following terminology is used throughout these issues:

Post-polio sequelae (PPS) is a general term referring to late-onset symptoms in polio survivors that: 1) can be attributed directly to damage caused by the polio virus (eg, cold intolerance or progressive respiratory insufficiency); 2) are thought to be related to the body's failure to maintain the level of recovery that was achieved following the polio infection (eg, new fatigue or muscle weakness [see post-polio syndrome]); or 3) result from a polio-related disability (eg, carpal tunnel syndrome secondary to years of crutch walking).

Post-polio syndrome refers to new, late-onset muscle weakness with EMG evidence of previous denervation and reinnervation (see Halstead, p 1209).

Post-poliomyelitis syndrome is used by Trojan et al in the December issue to refer to the combination of new muscle weakness with other musculoskeletal symptoms, such as joint pain.

A VIRAL PATHOPHYSIOLOGY FOR PPS?

Another problem during the first decade of post-polio sequelae was the proferring of "theories" about the pathophysiology of new symptoms. Two theories that most frightened polio survivors suggested that "some kind of amyotrophic lateral sclerosis" or a reactivated poliovirus were the cause of post-polio syndrome. Sharief et al, in the September 12, 1991 issue of the *New England Journal of Medicine*, speculated that a persistent polio virus may be the cause of post-polio syndrome and provided the only preliminary empirical support for this hypothesis. They reported evidence of "intrathecal immune activation against poliovirus" in 58% of 36 individuals with post-polio syndrome and no such evidence in 13 polio survivors without post-polio syndrome.

These findings are of great interest. However, the authors' statement that it is "tempting to speculate that reactivation of latent or persistent poliovirus infection. . .may have a role in the pathogenesis of new muscle weakness in some patients with the syndrome" is frightening to polio survivors. Further, this speculation is neither consistent with the authors' data nor the current state of our knowledge about the pathophysiology and treatment of post-polio syndrome.

The authors suggest that the poliovirus could have "escaped surveillance of the immune system" for decades by "blocking" the immune

Dr Bruno is Administrative Director, Post-Polio Rehabilitation and Research Service, Kessler Institute for Rehabilitation, East Orange, NJ, and Chairperson, International Post-Polio Task Force.

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system or by being hidden in infected motor neurons. The "persistent poliovirus infection might then have a gradual but progressive cytopathic effect, which would eventually lead to either neuronal-cell lysis or alterations of specialized cellular functions." For this hypothesis to be accepted, many questions must first be answered.

If the poliovirus has been residing unnoticed outside of the motor neuron, how and when did it enter the neuron, how does it produce its "cytopathic effect," and why has this effect taken decades to become evident? Slowly progressive muscle weakness over 30 years is not pathognomonic of poliovirus infection.

If the immune response has been suppressed for decades, what has triggered the immune response, why is the immune response seen in only half of the subjects with post-polio syndrome, and why did the immune response not inactivate the poliovirus before it entered the motor neuron to cause its cytopathic effect? Notably, there is no mention as to whether subjects had been immunized against poliovirus.

If the poliovirus has been hiding from the immune system within the motor neuron and causing a cytopathic effect, what has triggered the immune response at this point? Have degenerating motor neurons finally been lysed, releasing sequestered virus? If this is the case, did the post-polio syndrome subjects with the immune response have more severe weakness, muscle atrophy, or findings consistent with motor neuron death, as opposed to EMG evidence indicating decreased motor unit territories secondary to the degeneration of axonal sprouts (see Trojan et al in the December issue)? And, if persistent poliovirus is the cause of new muscle weakness, why is an immune response seen in only *half* of the subjects with post-polio syndrome?

A viral etiology is neither necessary nor sufficient to explain post-polio syndrome and it is not consistent with the numerous clinical and research reports that reductions in physical overexertion and emotional stress are effective treatments for both new and progressive muscle weakness (see Bruno and Frick, p 1185; Peach and Olejnik, p 1199) and that some lost muscle strength can be recovered through non-fatiguing progressive resistance exercise (see Fillyaw et al, p 1253). A more parsimonious explanation of the intrathecal immune response in subjects with post-polio syndrome is that some altered form of the poliovirus has been sequestered in previously infected motor neurons and is being released as those neurons die and lyse. Neuronal failure, death, and lysis would result from persistent damage done by the poliovirus during the *acute* polio infection to the metabolic apparatus of the neuron (see Bruno et al, p 1269) and the decades of metabolic strain on metabolically vulnerable neurons that sprouted extensively to reinnervate orphaned muscle fibers (see Trojan et al in the December issue). The release of poliovirus would then be a *secondary* effect of the pathophysiology of post-polio syndrome, not its primary cause.

Finally, it must be said that including the single line "It is unclear from our data whether patients with the post-polio syndrome carry any risk of infectivity" was not appropriate since the authors' data in no way examined the issue. This statement was potentially terrifying to polio survivors and anyone with whom they come in contact. It harkens back to the days of the epidemics when polio survivors and their families were shunned for years after the acute polio infection because of unfounded fears of "infectivity" (see Bruno and Frick, p 1185).

TOWARD THE THIRD DECADE

The 1987 National Health Interview Survey estimated that there are 1.63 million American polio survivors and that nearly half of them are reporting PPS. Since the median age of polio survivors is 45, the prevalence of PPS and the need for treatment will only increase as we all move into the next century. The articles presented in these special issues make clear that PPS are psychophysiologic in nature and that a holistic approach is required for their treatment. A holistic approach would also allow PPS to be prevented. The medical and post-polio communities need to begin to consider prevention of PPS as well as their treatment, and research needs to be directed toward both areas.

Unfortunately, Congress has been unresponsive to the need for funding of PPS research. Beginning in 1985, the Post-Polio Task Force has asked Congress to hold hearings on PPS and to set funds aside for research and treatment. To date, although the Senate Appropriations Committee has twice asked the National Institutes of Health to study PPS, no hearings have been held and no funds have been allocated. Congress must be petitioned to fund PPS research and treatment.