

IL 10-2004-018

In Reply Refer To: 111

December 7, 2004

UNDER SECRETARY FOR HEALTH'S INFORMATION LETTER

POST-POLIO SYNDROME (LATE SEQUELAE)

1. Background

a. While "late post-polio sequelae," also known as post-polio syndrome or post-poliomyelitis syndrome (PPS), is not a common entity, it does occur in veterans, and recognition of the scope and consequences are important. This information letter serves to generally familiarize practitioners with PPS and emphasize the need for care that may require expertise related to this uncommon symptomatology.

b. PPS represents an aggregate of often unexpected and sometimes disabling symptoms of muscle weakness, muscle and joint pain, abnormal sleep, heightened sensitivity to anesthesia, cold and pain intolerance, as well as difficulty with swallowing and/or breathing. These symptoms occur approximately 20-40 years after the acute poliovirus infection in 20-30 percent of previously paralyzed patients and perhaps some non-paralyzed polio survivors. There are about two million North American polio survivors and twenty million polio survivors worldwide. PPS may occur in veterans, including those who become ill with polio during or near time of service, or who may have served despite an earlier episode of paralytic anterior horn cell disease with recovery prior to military induction.

2. The Disease. While the specific cause(s) of PPS has (have) not been elucidated, it is suspected that this entity may be due to loss of neurons during the initial infection. After initial polio infection, some speculate that the remaining neurons compensate by increasing their motor unit territories in order to maximize muscle use. Some authorities attribute the late progression of muscle weakness to be the result of physiologic attrition of motor units innervating muscles and muscle groups already less innervated from the earlier acute infection. The true mechanism, however, is unknown.

3. Diagnosis. PPS is a diagnosis of exclusion. There are no specific diagnostic tests for PPS. While an electromyogram (EMG) can show that an individual has had anterior horn cell disease, it cannot define the extent of the PPS. It is necessary, therefore, to exclude all other causes forearm and leg twitching, excessive fatigue and muscle weakness, decrease in pulmonary function related to diaphragm and other muscle strength, abnormal breathing, particularly at night, and other neuromuscular symptoms or signs. Special note needs to be paid to changes in pulmonary function particularly related to anesthesia or concomitant conditions such as chronic obstructive pulmonary disease (COPD) since weakening of the respiratory muscles may lead to an inordinate decrease in respiration that can be life threatening.

A reasonable definition of PPS has been suggested with the following criteria:

- a. A confirmed history of paralytic polio;
- b. Partial-to-complete neurologic and functional recovery;
- c. A period of neurologic and functional stability of at least 15 years duration;
- d. The onset of two or more of the following health problems since achieving a period of stability: unaccustomed fatigue, muscle and/or joint pain, new weakness in muscles previously affected and/or unaffected, functional loss, cold intolerance, new atrophy; and
- e. No other medical diagnosis to explain these health problems.

4. Treatment. The primary component of treatment is recognition of the entity. Once recognized, this allows effective coordination of numerous clinical specialty areas to devise an appropriate plan of care designed at maximizing activities of daily living and functioning, as well as avoiding potential adverse outcomes that may occur when PPS patients interact with the medical community (e.g., anesthesia sensitivity). It is clear that many specialties need to be aware of PPS, including Primary Care, Neurology, Pulmonary and Critical Care, Anesthesia, Surgery, and, most importantly, Physical Medicine and Rehabilitation. Interventions such as assistive devices, changes in activity levels, and attention to other general health issues are important to the current and future well-being of persons who have had problems in the past.

5. Access to Care. Since PPS is not a common entity, it is likely that persons most familiar with this set of signs and symptoms will be those in Physical Medicine and Rehabilitation, and Neurology. In most inpatient Department of Veterans Affairs (VA) facilities, these specialties are readily available and need to be provided, as needed, to patients with known or suspected PPS. In settings where such consultation is not available, referral to other VA facilities where such expertise resides needs to be considered. If this is not feasible because of location, patient illness, or other contingencies, following appropriate legal and regulatory authority, outside VA referrals need to be considered based on patient need and local provider expertise.

6. References

- a. Dalakas M.C., Sever J.L., Madden D.L., et al. "Late poliomyelitis muscular atrophy: Clinical, virologic, and immunologic studies," *Reviews of Infectious Diseases*. 6:S562-S567; 1984.
- b. Dalakas M.C., Elder G., Hallet M., et al. "A long-term follow-up study of patients with post-poliomyelitis neuromuscular symptoms," *New England Journal of Medicine*. 314:959-963, 1986.
- c. Ramlow J., Alexander M., LaPorte R., et al. "Epidemiology of the post-polio syndrome." *American Journal of Epidemiology*. 136:769-786; 1992.
- d. Johnson R.T. "Late progression of poliomyelitis paralysis: Discussion of pathogenesis." *Reviews of Infectious Diseases*. 6:S568-S569; 1989.

- e. Modlin J.F. "Poliovirus," published in Mandell, Douglas and Bennett's Principles and Practices of Infectious Diseases, 5th ed. Vol. 2, Chapter 159, pgs. 1895-1903. Churchill Livingstone, Philadelphia, PA, 2000.
- f. Agre J.C., Mathews D.J. "Rehabilitation Concepts in Motor Neuron Diseases," published in Physical Medicine and Rehabilitation, ed. by R.L. Braddon. Chapter 5, pgs. 955-971, 1996. W.B. Saunders Company, Philadelphia, PA, 1996.
- g. Agre J.C., Rodriguez A.A. "Poliomyelitis and Postpolio Syndrome," published in Physical Medicine and Rehabilitation: the Complete Approach, ed. by M. Grabois, et al. Chapter 87, pgs. 1591-1610. Blackwell Science, Inc. Malden, MA, 2000
- h. Jubelt B. and Agre J.C. "Characteristics and Management of Post-Polio- Syndrome," Journal of the American Medical Association (JAMA). 284:412-412, 2000.
- i. Jubelt B. "Post-Polio Syndrome," Current Treatment Options in Neurology. 6:87-93, 2004.

7. Inquiries. Specific clinical questions may be referred to the Infectious Diseases Program Office at (513) 475-6398 or the Neurology Service at (203) 932-5711, ext. 3544, or the Physical Medicine and Rehabilitation Office at (612) 725-2044.

Jonathan B. Perlin, MD, PhD, MSHA, FACP
Acting Under Secretary for Health

DISTRIBUTION: CO:

E-mailed 12/9/04

FLD: VISN, MA, DO, OC, OCRO, and 200 – E-mailed 12/9/04