Chronic inflammatory demyelinating polyradiculoneuropathy

Assessment of long-term disabilities and response to treatment

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This PhD dissertation was accepted by the Faculty of Health Sciences of the University of Aarhus, and defended on October 3, 2008.

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Dan Med Bull 2009;56:72

ABSTRACT

The PhD dissertation is based on clinical studies on 24 patients with chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) carried out at the Department of Neurology, Aarhus University Hospital, during 2004 to 2008. The studies are presented as three individual papers.

CIDP is an autoimmune disease of the peripheral nervous system characterized with slowly progressive or relapsing symmetrical weakness and sensory loss. Responsiveness to immune modulating therapy is an important feature of CIDP and intravenous immunoglobulin (IVIG) is effective.

Although CIDP is a primary demyelinating polyneuropathy concomitant axonal loss is common as well. This finding appears to be clinical important because it is possible that long-term prognosis depends on the magnitude of axonal loss rather than on degree of demyelination.

The aims of my thesis were in long-term CIDP 1) to describe the short-term responsiveness to IVIG treatment 2) to determine the severity and distribution of assessed muscle weakness and to relate muscle performance to measures of function and quality of life and 3) to determine whether secondary axonal loss and/or segmental demyelination underlie permanent weakness and disability.

Important outcome measures were muscle strength assessed with isokinetic dynamometry, electrophysiological studies, and health related quality of life.

The acute motor response after a single course of IVIG was described in a prospective study of eight CIDP patients in IVIG maintenance therapy. Improvement of strength occurred after 5 days with a steady progress until day ten when a 12% increase was established without further increase for 10-15 days.

In a cross-sectional study of CIDP patients with a disease duration of 8.7 (3.3-11.5) years, an extensive health profile was provided. The average loss of muscle strength was 19.4% compared to matched healthy controls. Isokinetic muscle strength was associated with walking performance, functional health, and health-related quality of life.

Finally, a decreased distal compound muscle action potential and distal motor latency suggesting axonal loss were correlated to length dependent muscle weakness and functional disability whereas demyelinative features were not. Suggesting that permanent muscle weakness and disability in CIDP are closely related to axonal loss but not to signs of demyelination.