INTRAVENOUS IMMUNOGLOBULIN (IVIG) IS THE FIRST-LINE THERAPY IN CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIDP)

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CIDP is the most common acquired autoimmune neuropathy and the most gratifying PNS disorder because it responds to immunotherapies It is typically characterized by slow or subacute onset (>8 weeks from outset) of sensorimotor involvement with proximal and distal weakness, progressive or relapsing-remitting course, areflexia, increased CSF protein, demyelination on EMG/NCV studies and no other known cause. The immunopathogenesis of CIDP is not well studied. Macrophages, antibodies and T cells play a role. In the patients' nerves there are deposits of immunoglobulins and complement and prominent upregulation of cytokines, chemokines, adhesion molecules and co-stimulatory molecules.

CIDP has been the prototypic steroid-responsive neuropathy. Because the disease is slowly progressive, it requires chronic therapy. Based on controlled trials, the treatment options for CIDP include Prednisone, IVIG, Plasmapheresis and Immunosupressants. Controlled studies have shown almost equal efficacy of Prednisone, IVIG and Plasmapheresis but opinions differ on how treatment should begin. The controversy is not based on the different degree of efficacy among these therapies but mostly on cost and long-term side effects.

I will argue that the treatment of choice in CIDP is with IVIg, followed by steroids. Immunosuppressants (Azathioprine, Mycophenolate, Methotrexate, Cyclophosphamide or Cyclosporin) are used as steroid-sparing agents. Plasmapheresis and combination therapy is applied in those bone fide patients who do not respond to the initial treatment of IVIg and steroids. Rituximab is emerging as a potentially useful therapy in some difficult cases. Optimal protocols, safety concerns and uncertainties in the administration of IVIg will be discussed.