Ig Replacement Therapy in Primary Immunodeficiency

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Immunoglobulin (Ig) therapy is indicated as replacement treatment for patients with primary immunodeficiency (PID) characterized by absent or deficient antibody production. In 1946, Cohn developed an ethanol fractionation method to separate plasma proteins into stable fractions. In 1952, Bruton showed the effectiveness and benefit of Cohn fraction II for the treatment of agammablobulinemia. This landmark case began the modern era of Ig replacement. Ig therapy has served as lifesaving treatment in PID for over six decades. Approximately 70% of PID patients require Ig replacement to maintain their health during the disease course. Ig therapy reduces significantly the incidence of pneumonia and hospital admission, preserves organ function and improves life quality. Ig therapy should be individualized in terms of preparation, dose and frequency. The decision of Ig preparation, such as form, stabilizer, sodium and osmolarity, must be matched to specific patient needs and situations. The individualization of dosage does relate to the optimization of therapy. The target dose is that protecting the patient from significant infection. Trough level monitoring is helpful but should not be misinterpreted as benchmarks for therapy. Other factors including patient's condition, clinical course, treatment response and ongoing infection should be taken into consideration. Biologic IgG level represents the minimal serum IgG level that renders a patient as disease free as possible, which could be identified and maintained as the goal of Ig therapy. However, the current concept of Ig therapy is challenged by the findings that even the patient receives seemingly optimal treatment and shows no apparent clinical infections, silent lung airway disease progresses. Adverse reactions to Ig treatment are occasionally encountered, and classified as mild, moderate and severe. Thrombosis, anaphylaxis and hemolysis are very rare but serious complications. Use of SCIG has continued to grow in PID. In comparison to IVIG, it has similar efficacy in preventing infection, but is associated with minor local side effects and fewer systemic effects. In addition, the IgG level remains relatively consistent without the fluctuation characteristic of IVIG. According to the 2015 global network survey by Jeffrey Modell foundation, 42.9% of PID patients with an antibody deficiency received Ig replacement therapy. And the rate was even much less in Asia. Efforts are needed to improve the coverage of Ig replacement therapy in PID for the goodness of patients in Asian areas.