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Frequency and clinical manifestations of patients with primary immunodeficiency disorders in Iran: Update from the Iranian primary immunodeficiency registry

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Abstract

Primary immunodeficiency disorders (PID) are a heterogeneous group of diseases, characterized by an increased susceptibility to infections. A total of 930 patients (573 males and 357 females) are registered in Iranian PID Registry (IPIDR) during three decades. Predominantly antibody deficiencies were the most common (38.4%), followed by congenital defects of phagocyte number and/or function (28.3%), other well-defined immunodeficiency syndromes (17.7%), combined T- and B-cell immunodeficiencies (11.0%), complement deficiencies (2.4%), and diseases of immune dysregulation (2.3%). Common variable immunodeficiency was the most frequent disorder (20.8%), followed by chronic granulomatous disease, ataxia-telangiectasia, btk deficiency, selective IgA deficiency, and T-B-severe combined immunodeficiency. The frequency of other PID disorders was less than 50 in number (<5%). There is an increasing trend in recognition of more PID in the recent years. Construction of such registry is not only important for its epidemiological aspect but also for its role in increasing the physician's knowledge about such disorders. © 2006 Springer Science+Business Media, LLC.

Author keywords

Epidemiology; Immunological deficiency syndromes; Infection; Iran

Indexed Keywords

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