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## Journal of Clinical Immunology

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

Rezaei, N.<sup>aj</sup>, Aghamohammadi, A.<sup>a</sup>, Moin, M.<sup>a</sup>, Pourpak, Z.<sup>a</sup>, Movahedi, M.<sup>a</sup>, Gharagozlou, M.<sup>a</sup>, Atarod, L.<sup>a</sup>, Ghazi, B.M.<sup>a</sup>, Ishaiean, A.<sup>a</sup>, Mahmoudi, M.<sup>a</sup>, Abolmaali, K.<sup>a</sup>, Mansouri, D.<sup>b</sup>, Arshi, S.<sup>c</sup>, Tarash, N.J.<sup>c</sup>, Sherkat, R.<sup>d</sup>, Akbari, H.<sup>d</sup>, Amin, R.<sup>e</sup>, Alborzi, A.<sup>e</sup>, Kashef, S.<sup>e</sup>, Farid, R.<sup>f</sup>, Mohammadzadeh, I.<sup>g</sup>, Shabestari, M.S.<sup>h</sup>, Nabavi, M.<sup>i</sup>, Farhoudi, A.<sup>a</sup>

<sup>a</sup> Department of Allergy and Clinical Immunology of Children Medical Center, Immunology, Asthma and Allergy Research Institute, Tehran University of Medical Sciences, Tehran, Iran

<sup>b</sup> Department of Infectious Diseases, Masih Daneshvari Hospital, Beheshti University of Medical Sciences, Tehran, Iran

<sup>c</sup> Department of Immunology and Allergy, Rasoul Akram Hospital, Iran University of Medical Sciences, Tehran, Iran

<sup>d</sup> Al-Zahra Hospital, Isfahan University of Medical Sciences, Isfahan, Iran

<sup>e</sup> Department of Immunology and Allergy, Namazi Hospital, Shiraz University of Medical Sciences, Shiraz, Iran

<sup>f</sup> Department of Immunology and Allergy, Mashhad University of Medical Sciences, Mashhad, Iran

<sup>g</sup> Amirkola Hospital, Babol University of Medical Sciences, Babol, Iran

<sup>h</sup> Tabriz University of Medical Sciences, Tabriz, Iran

<sup>i</sup> Semnan University of Medical Sciences, Semnan, Iran

<sup>j</sup> Department of Allergy and Clinical Immunology of Children Medical Center, Immunology, Asthma and Allergy Research Institute, Tehran University of Medical Sciences, No. 62, Dr. G. St., Keshavarz Blvd., Tehran 14194, Iran

## Abstract

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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

EMTREE medical terms: adolescent; adult; aged; article; B lymphocyte; cell count; cellular immunodeficiency; child; clinical feature; combined immunodeficiency; common variable immunodeficiency; controlled study; epidemiological data; female; granulomatosis; human; humoral immune deficiency; immune deficiency; immune dysregulation; immunoglobulin A deficiency; infant; Iran; major clinical study; male; phagocyte dysfunction; priority journal; register; T lymphocyte

MeSH: Adolescent; Adult; Age Distribution; Aged; Aged, 80 and over; Child; Child, Preschool; Consanguinity; Female; Humans; Immunologic Deficiency Syndromes; Infant; Infant, Newborn; Iran; Male; Middle Aged; Registries

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