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Letter to the Editor

Distribution and Quality of Life in Patients With Primary Immunodeficiency Diseases in a Cohort of Korean Adults

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To the editor,

Primary immunodeficiency diseases (PID) are a heterogeneous group of disorders that affect distinct components of the innate and adaptive immune system. These diseases predispose patients to various complications, including infections, autoimmune disorders, and malignancies.[1,2] PID has been considered as rare diseases; however, a recent study has shown that as many as 1% of the population may be affected with PID.[3] Most of these diseases usually present and are diagnosed in childhood.[4] However, specific subtypes of PID, such as predominantly antibody deficiencies (PAD), are commonly diagnosed in adulthood or late childhood.[5,6] Therefore, to determine the frequency, characteristics, and clinical course of PID diagnosed in adulthood, the web-based registry was constructed in 7 university hospitals in Korea. The patients were enrolled between September 2015 and April 2019, and their collected data were analyzed on diagnosis, clinical presentation, laboratory tests, treatment, and quality of life (QoL) questionnaire using the 36-Item Short-Form Health Survey questionnaire (SF-36). The diagnosis was based on the report by the International Union of Immunological Societies PID Classification Committee.[1] Statistical analyses were performed with GraphPad Prism (GraphPad Software, San Diego, CA, USA). The Kruskal-Willis and Mann-Whitney *U* tests were used to compare immunoglobulin (Ig) levels according to the PID subtypes and the Welch *t* test was used to compare the SF-36 scores between patients with PID and those with asthma. *P* values of 0.05 or less were considered statistically significant.

A total of 84 patients (male/female: 25/59) with PID were registered and their mean age was 51.4 ± 15.1 years at diagnosis. All belonged to the category of PAD; IgG subclass deficiency (IgGSD) (56 patients, 66.7%), hypogammaglobulinemia (12 patients, 14.3%), thymoma with immunodeficiency (3 patients, 3.6%), common variable immunodeficiency (CVID; 2 patients, 2.4%), IgM deficiencies (2 patients, 2.4%), IgA deficiencies (2 patients, 2.4%), X-linked agammaglobulinemia (1 patients, 1.2%), IgA deficiency with IgGSD (1 patients 1.2%), IgG deficiency with IgGSD (4 patients, 4.8%), and IgM deficiency with IgGSD (1 patient, 1.2%,

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Disclosure

There are no financial or other issues that might lead to conflict of interest.

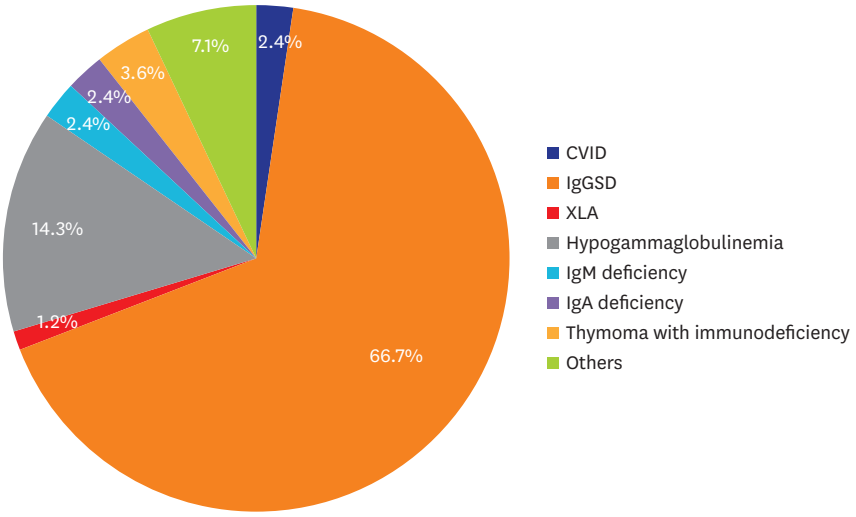

Fig. 1. Distribution of primary immunodeficiency disease in the study subjects.
CVID, common variable immunodeficiency; IgGSD, immunoglobulin G subclass deficiency; XLA, X-linked agammaglobulinemia; Ig, immunoglobulin.

Fig. 1). The serum IgG, IgA, and IgM levels at diagnosis were significantly different among the subtypes of PAD ($P < 0.001$ for IgG, $P = 0.024$ for IgA, and $P = 0.019$ for IgM) and the comparison of each Ig between the groups showed significant differences (**Fig. 2**), suggesting that measuring Ig is a useful tool to predict the subtype of PID before confirmation studies such as a genetic or flow cytometric method. Common infectious complications were upper respiratory tract infection (107 cases in 24 patients), followed by pneumonia (59 cases in 25 patients), and rhinosinusitis (3 cases in 3 patients). Seventy-eight patients completed the SF-36 questionnaire. As 65% of the subjects had asthma, the mean scores of SF-36 were compared to asthmatics with moderate persistent severity without PID and showed that the scores of SF-36 from PID patients were worse in all domains except physical functioning (**Fig. 3**).[7]

This cohort represents the PID distribution of the adult population in Korea with all PAD category, especially the high frequency of IgGSD. This finding is different from Western

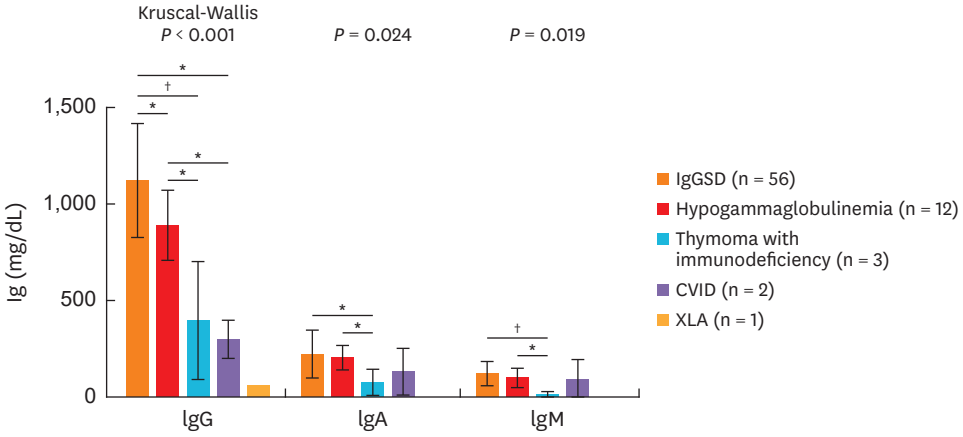

Fig. 2. Serum IgG, IgA, and IgM at diagnosis according to disease classification.
Ig, immunoglobulin; IgGSD, immunoglobulin G subclass deficiency; CVID, common variable immunodeficiency; XLA, X-linked agammaglobulinemia.
*$P < 0.05$, †$P < 0.01$ using the Kruskal-Wallis and Mann-Whitney U test.

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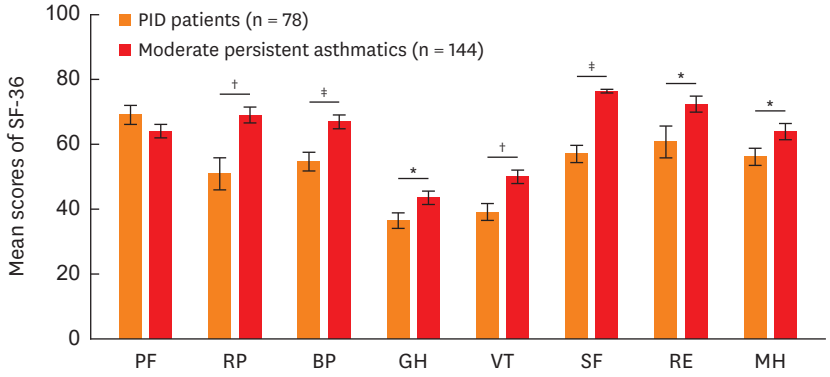

Fig. 3. Comparison of SF-36 scores between PID patients and moderate persistent asthmatics[7] in Korea.
SF-36, 36-Item Short-Form Health Survey questionnaire; PID, primary immunodeficiency diseases; PF, physical functioning; RP, role physical; BP, bodily pain; GH, general health; VT, vitality; SF, social functioning; RE, role emotional; MH, mental health.
*P < 0.05, †P < 0.01, ‡P < 0.001 using the Welch modified 2-sample t-test.

countries where CVID is the most common phenotype in the adult population, suggesting racial or genetic differences may affect the prevalence of PID. Furthermore, we found that PID significantly affects the health-related QoL of patients. This data have proven that PID is not uncommon in adults in Korea, suggesting that early diagnosis and treatment of PID are critical to minimizing morbidity and improving the QoL in this population.

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