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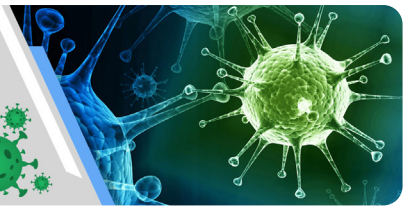

Primary Immunodeficiency Diseases in Costa Rican Adults: A Cross-Sectional Study

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Abstract

Purpose: Up to date, neither in Costa Rica nor in Central America public registries regarding the prevalence and characterization of primary immunodeficiency diseases (PID) in adult patients exist. The aim of this study was to characterize the clinical features of patients diagnosed with primary and idiopathic immune disorders treated in two specialized immunodeficiency clinics i Costa Rica.

Methods: A cross-sectional study was conducted between 2017 and 2018, including 137 adult patients diagnosed with PID at two public hospitals in Costa Rica. To estimate a robust prevalence of PID in Costa Rica, a binomial model using a hyperparameter with a Poisson distribution was implemented in WinBUGS to fit a Bayesian model to approach a posterior distribution for the prevalence estimation.

Results: Patients with PID showed a heterogeneous distribution and clinical course. Prevalence estimated resulted in a mean of 3.35 patients per 100,000 inhabitants in Costa Rica by 2018, with a 95% confidence interval of 2.98 to 4.15 patients per 100,000 inhabitants. A higher frequency of PID was observed in women and patients between 30 to 59 years of age. Humoral immunodeficiencies were predominant and most common immunodeficiency manifestations were recurrent infections and atopic syndrome. Most frequent comorbidities were dyslipidemia, acid peptic disease and hypertension.

Conclusions: In this cohort, we found different clinical manifestations in comparison to other regions, highlighting the importance of a prompt diagnose of these entities in adults. Metabolic diseases were identified as common comorbidities in this group. Appropriate prevalence estimations can address diagnostic strategies.

Keywords: primary immunodeficiency diseases, common variable immunodeficiency, adult, prevalence, Costa Rica, Bayes Theorem

Abbreviations: APS-1 (autoimmune polyglandular syndrome type I), CVID (common variable immunodeficiency), IgA (immunoglobulin A), IgE (immunoglobulin E), IgG (immunoglobulin G), IgM (immunoglobulin M), IVIG (intravenous immunoglobulin), IU/mL (International standard Unit per milliliter), LOCID (late onset combined immunodeficiency), LASID (Latin American Association for Immunodeficiencies), PID (Primary immunodeficiency diseases), SIgAD (selective IgA deficiency), SLE (systemic lupus erythematosus), SPAD (specific polysaccharide antibody deficiency), XLS (X-linked agammaglobulinemia)

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