Diagnostic spectrum and clinical profile of primary immunodeficiencies in children at an Algerian department of general pediatrics.

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Background

Primary immunodeficiencies (PIDs) are heterogeneous disorders, characterized by variable clinical immunological features and an increased susceptibility to infections autoimmune disorders and malignancies. Early diagnosis and appropriate treatment are critical for reducing morbidity and mortality.

Purpose:

The aim of this study was to estimate the disease burden of PIDs in a department of general pediatrics in Algiers and to appreciate the trends.

Methods:

Retrospective single center study conducted All cases of PIDs seen in our department between January 1st 2003 and december 31th 2020 were enrolled.

PIDs were classified according to the International Union of Immunological Societies expert committee for Primary Immunodeficiency.

Results:

87 PIDs patients were identified with 49 boys. Mean age at diagnosis was 32 months. Parental consanguinity was found in 34% of cases. Main clinical manifestations were recurrent respiratory infections (51%), growth failure (40%), chronic diarrhea (30%) and eczema (21)

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Based on the updated IUIS classification, PID distribution in our hospital showed that predominantly combined T and B cell immunodeficiency account for the majority of cases (39%), followed by combined immunodeficiencies with associated or syndromic features (23%) predominantly antibody deficiencies (17%), congenital defects of phagocyte (6,8%), diseases of immune dysregulation (6,7%) and autoinflammatory diseases (5%).

Among combined ID category, SCID was the most common condition (26%) followed by CMH II deficiency (8%). Use of intravenous immunoglobulin (IVIG), antibacterial and antifungal prophylaxis varied according to PID category. IVIG was used in 34 patients. A genetic study was carried out in 31 patients including 8 in Algeria.Bone marrow transplantation (BMT) was performed, mainly abroad, in 7 patients (3 Wiskott-Aldrich syndrome, 2 CMH II deficiency, 1 CGD, Chediak Higachi) at a mean age of 58 months. The global mortality of our series was 26 %.

Conclusion:

Combined T and B cell immunodeficiency followed by combined immunodeficiencies with associated or syndromic features are the commonest primary immune deficiencies (PIDs) identified in our center experience. . Long lag time in diagnosis and high mortality in our cohort emphasizes the need for early diagnosis and early referral.

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