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Primary Immune Deficits in children: follow-up and outcome

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Abstract

Introduction: Primary Immune Deficiencies (PID) of rare, under-determined diseases particularly in sub-Saharan Africa. Our aim was to share the results of the follow-up of these patients.

Patients and methods: We conducted a descriptive and analytical cross-sectional study at the Albert Royer National Children's Hospital in Dakar, in collaboration with other pediatric departments and the Immunology laboratory of the National Blood Transfusion Center. We included all patients received with suspected PID, from 2014 to 2021, after ruling out HIV infection. The diagnostic criteria were the recommendations of the Moroccan Society of Immunology. We did not include patients with incomplete data. A complete blood count was performed in all patients. Further explorations were carried out depending on the orientation. The data was analyzed with Excel 10.

Results: Out of 32 patients registered, 16 were included in a follow-up consultation (50%). The sex ratio was 0.6 and the mean age at diagnosis was 51.1 months. Inbreeding was observed in half of the patients (8/16). The warning signs were mainly infectious (11/16). The other symptoms were dermatological, such as eczema and warts (3/16), but also neurological, type ataxia (3/16). Anemia was observed in 12/16 children, lymphopenia in 4/16 children. Protein electrophoresis was performed in 10/16 children, immunoglobulin weight determination in 4/16 children and lymphocyte immunophenotyping in 10/16 patients. The main PIDs diagnosed were congenital neutropenia (3/16), severe combined immune deficiencies or SCID (3/16), telangiectasia ataxia (3/16), hypogammaglobulinemia (2/16), verruciform epidermodysplasia (2/16), Wiskott-Aldrich syndrome (1/16), chronic septic granulomatosis (1/16), Evans syndrome (1/16). The course was marked by relapses-remissions in 6/12 patients and discontinuation of follow-up in (5/16 patients). Bronchiectasis was observed in 2 patients, with secondary bacterial and fungal infections and digital hypocratism. 100% mortality was observed in carriers of SCID and Telangiectasia ataxia.

Conclusion: PIDs are suspected based on atypical clinical signs. Confirmation is difficult in low income countries. The development is marked by a risk of complications or death, hence the need to strengthen clinical-biological collaboration.