Features of Immune Dysregulation in Primary Immunodeficiency Patients at Soba University Hospital, Khartoum –Sudan

Goda, R.[1], Mohamed, A. S I.[1], Salim A[1], Masoud S O[1], Erwa N HH [1, 2, 3]
Soba University Hospital, University of Khartoum, Khartoum - Sudan

Online 7th ASID Congress October 1st-2nd 2021

Background

Immune dysregulation is an eminent presentation and feature of inborn errors of immunity (IEI). Patients present with autoimmune manifestations such as autoimmune cytopenias, allergy or atopy, endocrinopathies, recurrent severe infections, and more. We reviewed all patients with primary Immunodeficiency diseases (PIDs) that presented to immunology clinic at our hospital for features of immune dysregulation.

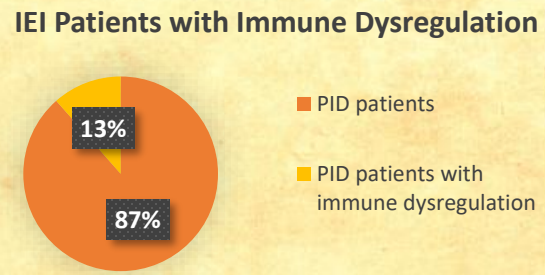
Purpose

This study describes the clinical manifestations and basic immunological findings in Sudanese IEI patients with features of immune dysregulation who presented to our clinic. Additionally, we looked into how these findings affected their management and outcome.

Patients and Methods

The records of patients with clinically diagnosed PID at Soba Immunology and Allergy Clinic in the period between May 2014 - June 2021 clinic were analyzed retrospectively for features of immune dysregulation such autoimmunity, allergy or atopy, lymphoproliferation and cytopenias.

Results



Major clinical features included: recurrent infections in all patients, autoimmunity in 75%, skin rashes in 50% and eczema in 43%. Major immunological features included hypogammaglobulinaemia in 31%, increased total IgE level, low CD4 T cells, and increased double negative T cells were, each, found in 25% of patients. FOXP3 / CD25 expression was reduced in 1 patient (not shown in Figure 2). None of the patients had a genetic/molecular diagnosis.

Figure 1: Percentage of patients with features of immune dysregulation (16 patients) among 120 PID patients at Soba immunology clinic.

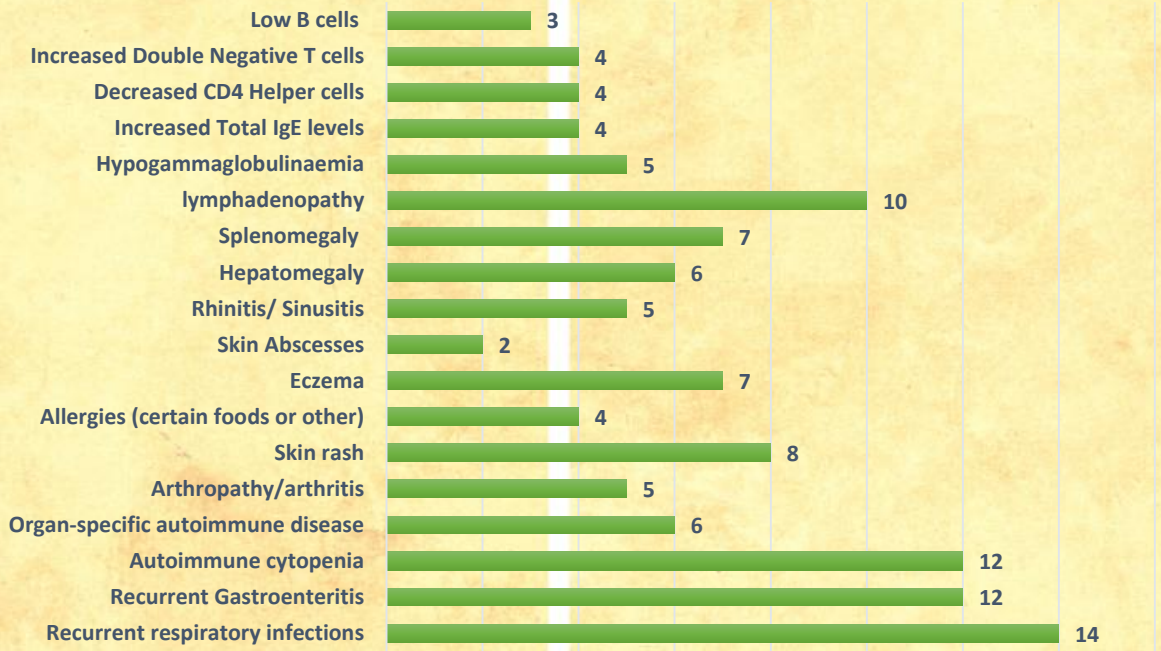


Figure 2: Frequency of immune dysregulation features among IEI patients at Soba Immunology Clinic

Conclusions

- Immune dysregulation features were overlapping and also overlapped with recurrent infections. These infections might be the result of immunosuppression in some. In the absence of genetic diagnosis disease IEI classification in this group of patients is difficult, however, the combination of clinical features favors combined immunodeficiency in the majority of this group.
- Significant management difficulties were evident with the absence of targeted therapies, refractory disease and poor follow-up compliance. Additionally, the lack of functional and molecular or genetic diagnostic facilities which all contributed to poorer clinical outcomes
- It is highly recommended that proper collaboration with pioneer centers occurs soon; to facilitate better patient diagnosis and management. Moreover, further studies and analyses are necessary to guarantee better IEI awareness and care in Sudan and Africa.

ASID
African Society for Immunodeficiencies
Société Africaine des Déficits Immunitaires

PID-IEI IN AFRICA:
WHERE AND HOW TO FIND THEM?
Online 7th ASID Congress
Khartoum, SUDAN
October 1st-2nd, 2021
African School
September 18th, 2021