

CLINICAL & LABORATORY PROFILE OF PRIMARY IMMUNODEFICIENCY CONDITIONS IN TERTIARY CARE CENTRE IN INDIA



B. Shenoy¹, A. Dixit², B. Bharath³

¹Head of Pediatric Infectious Diseases, Manipal Hospital, Bangalore, India.

²Pediatric Hematology, Manipal Hospital, Bangalore, India.

³Fellow in Pediatric Infectious Diseases, Manipal Hospital, Bangalore, India.

INTRODUCTION

Primary immunodeficiency disorders remain under diagnosed in developing countries. Despite several limitations and challenges, there has been significant progress in the diagnosis and management of these conditions.

METHODS

To study the clinical & laboratory profile of children with primary immunodeficiency in a tertiary care center in India. Retrospective study over a period of 5 years of children up to 16 years diagnosed with primary immunodeficiency at Manipal Hospital, Bangalore, India.

RESULTS

Out of the total 47 children with primary immunodeficiency, 22 children were less than 12 months. 25 were males and 22 were females. 18 children were diagnosed with severe combined immunodeficiency.

DIAGNOSIS	NUMBER OF CASES
Hypogammaglobulinemia	6
Familial HLH	5
Leucocyte Adhesion Defect	3
Hyper IgM syndrome	3
Mandelian Susceptibility to Mycobacterial disease	3
DiGeorge Syndrome	3
Ataxia Telangiectasia	2
Wiskott Aldrich syndrome	2
Hyper IgE syndrome	2

CLINICAL PRESENTATION	NUMBER OF CASES
Sino pulmonary infections	18
Persistent Fever	6
Failure to Thrive	9
Recurrent & Deep Skin Infections	8
BCG adenitis	3

These conditions were screened with appropriate laboratory parameters, immunoglobulin levels CD4 & CD8 counts, flow cytometry & NBT test. These cases were treated with IVIG & bone marrow transplantation. BMT was done in 12 cases & on regular follow ups with good recovery.

CONCLUSION

- Primary immunodeficiency conditions are frequently diagnosed in developing countries like India. Lack of awareness & non availability of diagnostic facilities is a major hurdle. Cost constraints to treatment like access to immunoglobulin replacements & HSCT are major barriers in the management in underdeveloped countries.

KEYWORDS

Immunodeficiency, Hypogammaglobulinemia