

## **LONG-TERM SURVIVAL ANALYSIS OF PRIMARY IMMUNODEFICIENCY DISORDERS, INSIGHTS FROM A LOW-MIDDLE INCOME COUNTRY.**

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We conducted a retrospective analysis of 34 patients diagnosed with PID between January 2011 and December 2023. Among the patients, 28 underwent hematopoietic stem cell transplantation (HSCT) using donors who were either fully matched (18) or haploidentical (10) from their siblings or parents.

**RESULTS:** The overall survival for primary immunodeficiency disorders (PID) was 65.9%. However, for patients who received early diagnosis followed by HSCT, the overall survival rate significantly improved to 78.57%. Notably, a vast majority 27 (96.4%) of patients presented with infections prior to transplantation, with 19 (67.8%) of them having persistent infection during the transplant procedure. Consanguinity was notable in 79% of cases. Two successful high-risk haploidentical transplants were performed for Chediak-Higashi syndrome with acute HLH and Reticular dysgenesis.

**DISCUSSION:** Majority of these patients remain undiagnosed. Only a handful cases are identified each year. Initially, good supportive care to counter infections followed by HSCT is the sole curative treatment for most PID. Given that a significant proportion of marriages occur within extended families in our country, it becomes crucial to offer counseling to family members, so that the birth of affected children can be prevented or facilitate early diagnosis followed by HSCT.