

**Clinical-epidemiological pattern of primary immunodeficiencies in Malaysia 1987-2006:  
a 20 year experience in four Malaysian hospitals**

**ABSTRACT**

**Aim of study:** To determine the clinical and epidemiological characteristics of patients seen with primary immunodeficiencies referred at four Malaysian Hospitals between 1987 to 2007  
**Methods:** Patient data were retrospectively obtained from patient records and supplemented by information from a standardized questionnaires taken at the time of diagnosis from 4 participating hospitals. The completed data were transferred to document records kept by the first author. The diagnoses made were based on criteria set by WHO Scientific Committee 1986. **Results:** Fifty one (51) patients with completed records satisfied the criteria of primary immunodeficiencies based on WHO Scientific Committee 1986. Predominant Antibody deficiency (40.4%) is the commonest of the class of primary immunodeficiency (based on modified IUIS classification) followed by phagocytic defect (17.3%), combined immunodeficiencies (15.4%) and other cellular immunodeficiencies (11.5%). The commonest clinical presentation is pneumonia (54%). A positive Family history with a close family relative afflicted was a strong pointer to diagnosis for PID (52.6%) Primary immunodeficiencies are seen in all the major ethnic groups of Malaysia, predominantly among Malays. As observed in other patient registries, diagnostic delay remains the major cause of morbidity and mortality. **Conclusion:** Primary immunodeficiencies is relative rare but is an emerging disease in Malaysia. Creating awareness of the disease, may reveal more cases within the community. It is sufficient to be a health issue in Malaysia as in other developing countries in the future.

**Keyword:** Clinical patterns; Diagnostic delay; Malaysia; Primary immunodeficiency