Original Article

Primary Immunodeficiency Disorder in Children of Bangladesh: A Cross-Sectional Study from a Tertiary Care Hospital

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Abstract:

Background: Primary immunodeficiency diseases (PIDDs) are a heterogeneous group of inherited disorders resulting from intrinsic defects in the immune system, leading to increased susceptibility to recurrent and severe infections. Although advances in diagnostic techniques have improved detection rates in high-income settings, early diagnosis remains a significant challenge in low- and middle-income countries. Therefore, a better understanding of the clinical and laboratory profiles of PIDD is essential to facilitate timely diagnosis and improve patient outcomes, particularly in resource-limited settings such as Bangladesh. **Objective:** To make a survey on the demographic, clinical, and laboratory characteristics of PIDD cases diagnosed among children at a tertiary care hospital in Bangladesh. **Methods:** A total of 62 clinically suspected PIDD patients from paediatric departments of Bangladesh Medical University were evaluated between March 2020 to January 2021. Quantitative immunoglobulin assays and flow cytometric immunophenotyping were performed to confirm diagnoses. **Results:** Of the 62 suspected cases, 14 (23%) were laboratory - confirmed as PIDDs. The median age at diagnosis was 21 months, and the male-to-female ratio was 1:1. Predominantly antibody deficiencies were the most frequent category (64.3%), followed by combined immunodeficiencies (35.7%). Common variable immunodeficiency (CVID) and agammaglobulinemia were the leading subtypes. Recurrent pneumonia (68%) and recurrent use of intravenous antibiotics (76%) were the most frequent warning signs. **Conclusion:** Predominantly antibody deficiencies represent the major category of PIDDs in this study, consistent with global patterns. Enhanced awareness, expanded diagnostic access, and establishment of a national PIDD registry are essential for early detection and improved patient management in Bangladesh.

Key words: Primary immunodeficiency, antibody deficiency, flow cytometry, CVID, paediatric immunology.

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Introduction:

Primary immunodeficiency disorders (PIDDs) are congenital defects in immune function[1]. They manifest as recurrent infections and autoimmune or malignant conditions[2]. Although the overall prevalence of PIDDs had been estimated to be 1 per 10,000 individuals[3,4]. Registry data show that predominantly antibody deficiencies are the most common subtype[5,6,7]. In Bangladesh, 60% of PIDD cases are antibody

deficiencies[8]. The decision of when to test or refer a child for evaluation of a suspected PIDD may be affected by several factors. History of recurrent pneumonias and/or ear, sinus and cutaneous infections or any unusual infections, a strong family history of recurrent or unusual infections herein merits immunologic evaluation. Other suspicious elements of the family history include consanguinity and early childhood mortality from infections or unexplained causes. Children with failure to thrive should also raise suspicion for underlying PIDD[9]. Diagnosis relies on immunological assessment, including serum immunoglobulins and lymphocyte subset analysis. Abnormal serum immunoglobulin levels suggest a B-cell disorder. Abnormalities on assay of the classic or alternative complement pathways suggest a complement disorder. Other important diagnostic tools include lymphocyte proliferation assays and flow cytometry which allow for the enumeration of B-cells, T-cells, and NK cells, and the evaluation of lymphocyte markers, T-cell variability, and adhesion receptors that may be associated with specific immune defects[10]. This study aimed to evaluate the demographic and laboratory profiles of children diagnosed with PIDD at a tertiary hospital.

Methods:

A total of 62 clinically suspected PIDD patients were enrolled in this cross-sectional study between March 2020 and January 2021. Patients were selected by the clinicians of the Department of Paediatrics, Bangladesh Medical University (BMU) and referred to the Department of Microbiology and Immunology, BMU, for further laboratory evaluation. Children under 18 years were included following Jeffrey Model Foundation criteria. Peripheral blood samples were collected for serum immunoglobulin measurement (IgG, IgA, IgM, IgE) via nephelometry and for flow cytometric evaluation of lymphocyte subsets (CD3, CD4, CD8, CD19, CD27, CD56, CD45RA, CD45RO). Patients with secondary immunodeficiencies were excluded.

Results:

A total of 62 suspected Primary Immunodeficiency Disease (PIDD) patients were enrolled in this study as cases. Among them 14 (23%) were diagnosed with PIDDs based on the laboratory test results evidence. Median ages of the patients ranged from 2 to 180 months. Median age of confirmed PIDD cases was 21 months and non-PIDD cases was 11 months. Median age of onset of symptom was 5 months in the suspected PIDD cases. The male to female (M: F) ratio was 1:1 in the confirmed PIDD cases (Table I).

Table-I: Demographic characteristics of study population

Variable	PIDD (n=14)	Non-PIDD (n=48)	Total suspected PID cases (n=62)	P-value
Gender Male Female	7 (50%) 7 (50%)	28 (58.3%) 20 (41.7%)	35 (56.5%) 27 (43.5%)	0.760[a]
Age (month) Median(range)	21.0 (4-120)	11.0 (2-180)	11.5 (2-180)	0.284[b]
Age of onset (month) Median (range)	12 (2-72)	4.5 (1-109)	5 (1-109)	0.060[b]

a= Chi-square test, b= Mann-Whitney U-test

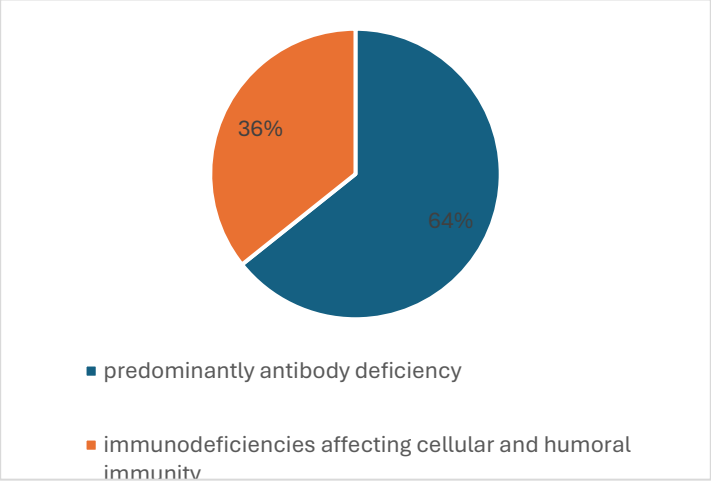

Figure 1: Pie diagram shows distribution of PIDD cases (n=14)

Figure 1 shows distributions of different categories of diagnosed PIDD. Out of 14 laboratory-confirmed PIDD cases, 9 (64.28%) patients had predominantly antibody deficiency and 5 (35.71%) patients had immunodeficiencies affecting cellular and humoral immunity. On subcategorizing cases based on symptom complex and available investigations: common variable immune deficiency, agammaglobulinemia and combined immunodeficiencies other than severe combined immune deficiency (SCID) consisted of 3 cases in each clinical type. One in each case was diagnosed with transient hypogammaglobulinemia of

infancy, selective IgA deficiency, selective IgM deficiency (Table II).

Table II: Distribution of PIDD cases according to types (n=14)

PIDD types	No. of cases (%)
Predominantly antibody deficiency	9(64.28%)
Agammaglobulinemia	3(21.43%)
Common variable immunodeficiency (CVID)	3(21.43%)
Transient hypogammaglobulinemia of infancy	1(7.14%)
Selective IgA deficiency	1(7.14%)
Selective IgM deficiency	1(7.14%)
Immunodeficiencies affecting cellular and humoral immunity	5(35.71%)
Severe combined immunodeficiencies (SCID)	2(14.28%)
Combined immunodeficiencies other than SCID	3(21.43%)

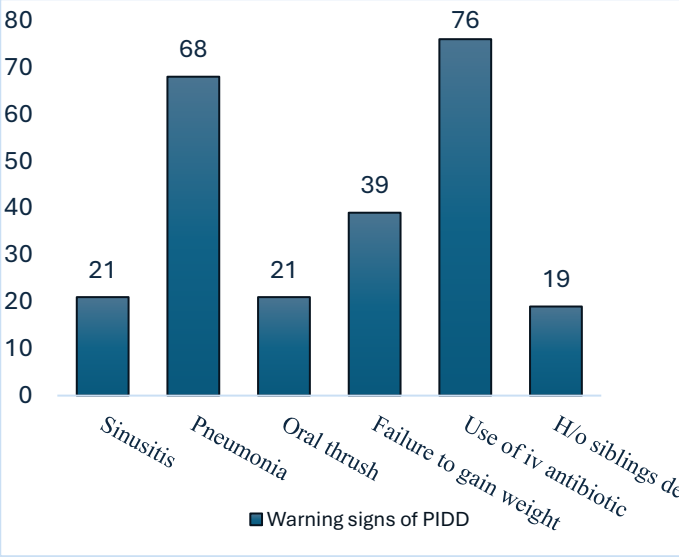

Figure-2: Distribution of warning signs of PIDD among study population

Distribution of different warning signs in the study population shows that most common warning sign was the recurrent use of intravenous antibiotics to clear infection (76%) followed by recurrent pneumonia (68%). 19% of patient's had a family history of sibling death due to similar infections (Figure-2).

Discussion:

In the present study, 23% of the clinically suspected cases were confirmed to have primary immunodeficiency diseases (PIDDs) based on laboratory evidence. This detection rate is within the range reported from similar hospital-based studies in developing countries, where diagnostic facilities and clinical awareness are often limited. Early recognition of PIDDs remains a significant challenge, particularly in resource-constrained settings where recurrent infections are frequently attributed to environmental or nutritional factors rather than underlying immune defects[11]. This study demonstrates that predominantly antibody deficiencies constitute the largest group of PIDDs among paediatric patients, consistent with international trends[5,6]. The observed frequency (64.3%) mirrors reports from other developing regions, where delayed diagnosis often leads to recurrent infections before confirmation. In our study, CVID and agammaglobulinemia were the leading antibody deficiency disorders, similar to findings from India[12] and Iran[5]. Combined immunodeficiencies accounted for 35.7% of cases, consistent with studies from the Middle East[7] but slightly higher than Western data[1], possibly reflecting diagnostic referral bias toward more severe cases. The median age of onset (5 months) and diagnosis (21 months) indicate a diagnostic delay, which is a persistent issue in resource-limited settings due to low clinical suspicion and limited access to advanced immunologic testing[3]. Recurrent pneumonia and repeated antibiotic use were the most common clinical manifestations, consistent with other regional studies[8,12]. Family history of infection-related mortality further underscores the genetic basis of these disorders and the need for family screening. Improved awareness, establishment of diagnostic infrastructure, and inclusion of flow cytometry-based lymphocyte subset analysis can significantly aid early identification of PIDDs.

Conclusion:

Primary immunodeficiency disorder is not uncommon in our country. Predominantly antibody deficiency was the most common PIDD. Routine immunological screening and integration of molecular testing can improve diagnostic accuracy and patient outcomes.

Limitations:

The study was limited by its single-center design and small sample size. Genetic testing was unavailable, limiting precise molecular diagnosis. Future studies should incorporate multicenter collaboration and longitudinal follow-up.

Recommendations:

- Conduct multicenter studies with larger cohorts.
- Include CD45RA/CD45RO markers in T-cell subset analysis for combined immunodeficiency.
- Use CD27 and IgD markers to identify switched memory B-cells in CVID cases.
- Establish genetic testing facilities for definitive PIDD diagnosis.

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