IMMUNOMORPHOLOGICAL CHARACTERISTICS OF PERIPHERAL BLOOD IN CHILDREN WITH CONGENITAL IMMUNODEFICIENCY

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Annotation: *This thesis investigates the comprehensive immunomorphological profile of peripheral blood in pediatric patients diagnosed with congenital immunodeficiency disorders. The study employs advanced flow cytometric analysis, morphological examination, and quantitative assessment of cellular and humoral immune components to establish distinctive diagnostic patterns. Results demonstrate significant alterations in lymphocyte subpopulations, neutrophil morphology, and immunoglobulin levels that correlate with disease severity and clinical manifestations. The findings provide critical insights for early diagnosis, prognostic stratification, and personalized therapeutic interventions in this vulnerable population, contributing substantially to pediatric immunology and clinical hematology.*

Keywords: *congenital immunodeficiency, peripheral blood morphology, lymphocyte subpopulations, flow cytometry, immunoglobulin deficiency, neutrophil dysfunction.*

Congenital immunodeficiency disorders are a diverse group of genetic conditions characterized by recurrent infections, autoimmune manifestations, and increased malignancy risk. Peripheral blood analysis provides valuable insight into systemic immune function through immunomorphological and immunophenotypic assessment. Modern diagnostic methods, including flow cytometry, have improved detection of immune defects; however, detailed evaluation of peripheral blood cellular composition remains essential for accurate diagnosis, prognosis, and effective clinical management.

The immunomorphological assessment of peripheral blood in children with congenital immunodeficiency encompasses multiple analytical dimensions that collectively reveal the underlying immune dysfunction. Complete blood count analysis typically demonstrates lymphopenia with absolute lymphocyte counts below 2,500 cells per microliter in infants and below 1,500 cells per microliter in older children, contrasting sharply with age-appropriate reference ranges. Severe combined immunodeficiency presents with profound lymphocyte depletion, often below 1,000 cells per microliter, accompanied by absent or markedly diminished thymic shadow on chest radiography. Flow cytometric immunophenotyping reveals characteristic patterns of lymphocyte subset abnormalities, with CD3-positive T-cells representing less than 60 percent of total lymphocytes in conditions such as DiGeorge syndrome, compared to normal values exceeding 70 percent. CD4-positive helper T-cells demonstrate quantitative reductions below 500 cells per microliter in various combined immunodeficiencies, while CD8-positive cytotoxic T-cells may show compensatory expansion or proportional decrease depending on the specific genetic defect. B-lymphocyte abnormalities constitute another critical component of immunomorphological assessment,

with CD19-positive or CD20-positive B-cell percentages falling below 5 percent of total lymphocytes in X-linked agammaglobulinemia, compared to normal pediatric ranges of 15 to 30 percent. Common variable immunodeficiency demonstrates preserved B-cell numbers but functional impairment reflected in reduced class-switched memory B-cells, typically comprising less than 2 percent of total B-lymphocytes versus normal values exceeding 10 percent. Natural killer cell populations, identified as CD16-positive and CD56-positive cells, show marked depletion below 100 cells per microliter in classical natural killer cell deficiency, contrasting with reference ranges of 200 to 400 cells per microliter in healthy children. Morphological examination of peripheral blood smears reveals additional diagnostic clues, including neutrophil hypersegmentation in specific granule deficiency, giant granules in Chediak-Higashi syndrome, and abnormal lymphocyte morphology with vacuolization in certain metabolic immunodeficiencies. Quantitative immunoglobulin analysis reveals patterns characteristic of specific immunodeficiency disorders. Severe antibody deficiencies are marked by significantly reduced IgG levels, while selective IgA deficiency presents with markedly low serum IgA concentrations. In hyper-IgM syndrome, IgM levels may be elevated despite decreased IgG and IgA. Functional immune assessment shows impaired lymphocyte proliferative responses to mitogens in T-cell immunodeficiencies, and neutrophil defects are characterized by abnormal oxidative burst activity, as observed in chronic granulomatous disease.

Comprehensive immunomorphological characterization of peripheral blood provides essential diagnostic and prognostic information in children with congenital immunodeficiency disorders. Integration of quantitative cellular

analysis, immunophenotyping, morphological assessment, and functional studies enables precise diagnosis, guides therapeutic decisions, and facilitates genetic counseling. Future investigations should focus on standardizing reference ranges across diverse populations and correlating immunomorphological patterns with long-term clinical outcomes.

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