



Disorders of Lymphocytes

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INTRODUCTION

The central cell of the immune system is the lymphocyte. Lymphocytes mediate the adaptive immune response, providing specificity to the immune system by responding to specific pathogens and conferring long-lasting immunity to reinfection. Lymphocytes are derived from pluripotent hematopoietic stem cells that reside in the bone marrow and give rise to all of the cellular elements of the blood. The two major functional classes of lymphocytes—B lymphocytes (B cells) and T lymphocytes (T cells)—are distinguished by their site of development, antigenic receptors, and function.

The major disorders of lymphocytes include neoplastic transformations of specific subsets of lymphocytes that result in an array of lymphomas or leukemias, congenital or acquired defects in lymphocyte development or function with resultant immunodeficiency syndromes, and physiologic responses to infection or

antigenic stimulation that may lead to lymphadenopathy, lymphocytosis, or lymphocytopenia.

LYMPHOCYTE DEVELOPMENT, FUNCTION, AND LOCALIZATION

B Cells

B cells are characterized by cell surface immunoglobulins (i.e., antibodies). Their major function is to mount a humoral immune response to antigens by producing antigen-specific antibodies.

B cells develop in the bone marrow in a series of highly coordinated steps that involve sequential rearrangement of the heavy- and light-chain immunoglobulin genes and expression of B-cell-specific cell surface proteins (Fig. 49-1). Rearrangement of the immunoglobulin genes results in generation of a large repertoire of B cells that are each characterized by an immunoglobulin molecule with unique antigenic specificity. Mature B cells

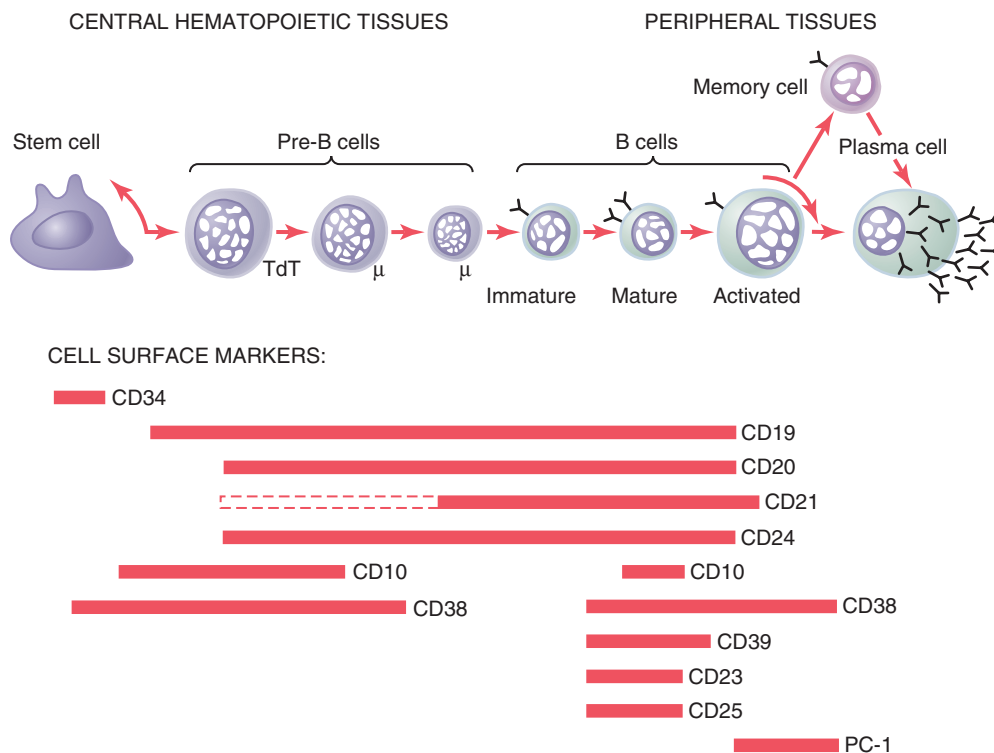


FIGURE 49-1 The maturation of B lymphocytes. *Top*, The changes in immunoglobulin production and maturation. *Bottom*, The appearance and disappearance of surface markers. TdT, Terminal deoxynucleotidyl transferase. (Modified from Ferrarini M, Grossi CE, Cooper MD: Cellular and molecular biology of lymphoid cells. In Handin RI, Lux SE, Stossel TP, editors: Blood principles and practice of hematology, Philadelphia, 1995, JB Lippincott, p 643.)