



Function and dysfunction of B-cells in primary immunodeficiencies

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The generation of antibody responses and the formation of B-cell memory require strictly controlled B cell differentiation that can be dissected into several critical steps. Many of these steps have been found disrupted in patients with a primary immunodeficiency (PID). Immunological studies are crucial in the process of identification of genetic defects in these patients. A molecular diagnosis is important for instalment of adequate therapy and therefore contributes to good patient care. However, immunological studies in PID patients also generate new insights in B cell differentiation and antibody responses. In addition, some genetic defects not only affect the humoral immune response, but also the cellular immune response. Still, in many patients with impaired antibody formation, it remains unclear how B cells and sometimes T-cells are affected, indicating that further research on PID will unravel new aspects of the immune system.