Low marginal zone-like B lymphocytes and natural antibodies characterize skewed B-lymphocyte subpopulations in del22q11 DiGeorge patients.

Abstract

PURPOSE: Patients with DiGeorge syndrome suffer from T-lymphopenia. T-cells are important for the maturation and regulation of B-cell function. Our aim was to characterize the B-cell compartment in DiGeorge syndrome patients.

METHODS: B-cell subset phenotypization using flow cytometry. Serum BAFF (B-cell activating factor) and serum anti-alpha-galactosyl IgM measurement using ELISA. Serum IgG measurement using nephelometry.

RESULTS: We observed a significantly increased number of naïve B-cells and decreased number of switched memory B-cells in DiGeorge patients. Furthermore, we observed increased BAFF levels and a trend toward hypergammaglobulinemia.
later in life. Surprisingly, we detected a decrease in marginal zone-like (MZ-like) B-cells and natural antibodies in DiGeorge patients.

CONCLUSION: The maturation of B-cells is impaired in DiGeorge patients, with high naïve and low switched memory B-cell numbers being observed. There is a clear trend toward hypergammaglobulinemia later in life, coupled with increased serum BAFF levels. Surprisingly, the T-independent humoral response is also impaired, with low numbers of MZ-like B-cell and low levels of anti-alpha-galactosyl IgM natural antibodies being detected.


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