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Autoimmunity and Immunodeficiency.

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Abstract

The references provided include data from evidence A and B studies based on the relevant populations. Because many primary immunodeficiencies associated with autoimmune diseases are rare, illustrative cases (evidence D) also are referenced. On the basis of level A evidence, immunoglobulin A deficiency is the most common primary immunodeficiency and is associated with defective mucosal immunity and autoimmune disease. On the basis of strong evidence (level A), Wiskott Aldrich syndrome presents early in life and is associated with autoimmune arthritis and anemia. On the basis of strong evidence in the literature, a number of primary immunodeficiencies are associated with defects in T regulatory cell number and development, cytokine aberrancies, and, as a consequence, production of autoantibodies. On the basis of strong evidence (level A) and case reports (level D), complement deficiency can be associated with autoimmune disease, most notably systemic lupus erythematosus.

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