

Selective IgM deficiency: An ignored and underestimated primary immunodeficiency**Sudhir Gupta**

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Although selective IgM deficiency (SIGMD) was described almost five decades ago it was largely ignored as a primary immunodeficiency (PID). Selective IgM deficiency is defined as serum IgM levels below two standard deviation of mean with normal serum IgG and IgA. It appears to be more common than originally realized. Selective IgM deficiency is observed in both children and adults. Patients with SIGMD may be asymptomatic; however, approximately 80% of patients with SIGMD present with infections with bacteria, viruses, fungi and protozoa. There is an increased frequency of allergic and autoimmune diseases in SIGMD. A number of B cell subset abnormalities have been reported, and impaired specific antibodies to *Streptococcus pneumoniae* responses are observed in more than 50% of cases. Innate immunity, T cells, T cell subsets and T cell functions are essentially normal. The pathogenesis of SIGMD remains unclear. Mice selectively deficient in secreted IgM are also unable to control infections from bacterial, viral and fungal pathogens and develop autoimmunity. Immunological and clinical similarities and differences between mouse models of deficiency of secreted IgM and humans with SIGMD will be discussed. Although etiopathogenesis of SIGMD is unclear, our recent whole exome sequence studies may shed light on possible gene mutations responsible for SIGMD. Patients with SIGMD presenting with recurrent infections and specific antibody deficiency responses appear to improve clinically on immunoglobulin therapy.

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