Immunodeficiency with factor I anomaly

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Immunodeficiency with factor I anomaly. ORPHA:200418

Immunodeficiency with factor I anomaly is a rare, genetic, primary immunodeficiency disease characterized by increased susceptibility to recurrent, usually severe, infections (particularly by Neisseria meningitidis, Haemophilus influenzae and Streptococcus pneumonia), typically manifesting as otitis, sinusitis, bronchitis, pneumonia, and/or meningitis. Autoimmune disease (e.g. systemic lupus erythematosus, glomerulonephritis) and atypical hemolytic uremic syndrome may be associated. Laboratory serum analysis reveals, in addition to diminished or undetectable complement factor I, variably decreased complement C3, complement factor B and complement factor H.