

Opis choroby *

Definicja

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 pneumoniae*), 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.

Dane

Klasyfikacja Choroba	Synonimy Complete factor I deficiency		
Kod ORPHA 200418	Kod OMIM 610984	Kod ICD10 D84.1	
Kod ICD11 4A00.1Y			

*Źródło

orphanet