

Opis choroby *

Definicja

Immunodeficiency due to a classical component pathway complement deficiency is a primary immunodeficiency due to a deficiency in either complement components C1q, C1r, C1s, C2 or C4 characterized by increased susceptibility to bacterial infections, particularly with encapsulated bacteria, and increased risk for autoimmune disease. Most commonly, these include systemic lupus erythematosus (SLE), SLE-like disease, Henoch-Schonlein purpura, polymyositis and arthralgia. Disease severity is variable and dependent on the complement affected.

Dane

Klasyfikacja

Choroba

Synonimy

Immunodeficiency due to C1, C4, or C2
component complement deficiency

Niedobór odporności z powodu deficytu
składników C1, C4, lub C2 układu dopełniacza

Immunodeficiency due to an early component of
complement deficiency

Kod ORPHA

169147

Kod OMIM

614380

Kod ICD10

D84.1

Kod ICD11

4A00.10

[*Źródło](#)

orphanet