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