Opis choroby *

Definicja

A rare systemic autoimmune disease characterized by cutaneous lesions, hepatic dysfunction, hematological abnormalities, and/or cardiac arrhythmia, and caused by transplacental passage of maternal SS-A and SS-B autoantibodies. The most typical cutaneous manifestation is a macular annular erythema affecting the head, but also trunk and extremities. Other reversible features include anemia, neutropenia, thrombocytopenia, and elevation of liver parameters with hepatomegaly. The most severe presentation of the disease is irreversible congenital total atrioventricular block.

Dane

Klasyfikacja
Choroba

Kod ORPHA
398124

Kod OMIM
-

Kod ICD10
M32.8

Kod ICD11
KA07.0

---

*Źródło

orphanet