

Opis choroby *

Definicja

A rare systemic autoimmune disease characterized by exocrine gland dysfunction, resulting predominately in keratoconjunctivitis sicca and xerostomia, but also affecting exocrine glands of the skin, as well as respiratory, urogenital, and digestive tract. Extraglandular manifestations include arthritis, interstitial lung disease, renal disease, and peripheral neuropathy. The disease is accompanied by a substantially increased risk to develop B-cell non-Hodgkin lymphoma, especially MALT (mucosa-associated lymphoid tissue) lymphoma.

Dane

Klasyfikacja

Choroba

Synonimy

Primary Sjögren-Gougerot syndrome

Pierwotny zespół Sjögrena-Gougerota

Kod ORPHA

289390

Kod OMIM

270150

Kod ICD10

M35.0

Kod ICD11

4A43.20

*Źródło

orphanet