

Opis choroby *

Definicja

A clonal B-cell disorder characterized by the aggregation and deposition of insoluble amyloid fibrils derived from misfolding of monoclonal immunoglobulin light chains. It usually presents as systemic AL amyloidosis with involvement of one or more parenchymal organ(s) and, less frequently, as localized amyloidosis with usually nodular deposits restricted to a single organ and/or system.

Dane

Klasyfikacja

Choroba

Synonimy

Light-chain amyloidosis

Amyloidoza immunoglobulinowa

Amyloidoza łańcuchów lekkich

Amyloidoza pierwotna

Primary amyloidosis

Kod ORPHA

85443

Kod OMIM

254500

Kod ICD10

E85.9

Kod ICD11

5D00.0

*Źródło

orphanet