Opis choroby *

Definicja

A rare, hereditary amyloidosis with primary renal involvement characterized by amyloid deposition in the kidney glomeruli and medulla, gastrointestinal tract, liver, spleen and slow disease progression. Symptoms and signs include nausea, vomiting, dyspepsia, gastritis, gastrointestinal hemorrhage, abdominal pain, hepatic rupture, sicca syndrome, purpura and petechiae, lymphadenopathy and renal dysfunction.

Dane

Klasyfikacja	Synonimy
Podtyp kliniczny	Familial amyloid nephropathy due to lysozyme variant Amyloidoza lizozymowa Dziedziczna amyloidoza nerek spowodowana wariantem lizozymu Dziedziczna nefropatia amyloidowa spowodowana wariantem lizozymu Rodzinna amyloidoza nerek spowodowana wariantem lizozymu Rodzinna nefropatia amyloidowa spowodowana wariantem lizozymu Familial renal amyloidosis due to lysozyme variant Hereditary amyloid nephropathy due to lysozyme variant Hereditary renal amyloidosis due to lysozyme variant Lysozyme amyloidosis

Kod ORPHA	**Kod OMIM**	**Kod ICD10**
93561	-	E85.0

Kod ICD11
5D00.2Y

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*Źródło

orphanet