Opis choroby *

Definicja

A rare, hereditary amyloidosis with primary renal involvement characterized by fibrinogen A-alpha-chain amyloid deposition predominantly in the kidney glomeruli and clinically presenting with hypertension, uremia, nephrotic syndrome slowly progressing to end-stage renal disease. Extra-renal involvement is possible, due to neurological, cardiac, visceral and vascular amyloid deposition.

Dane

Klasyfikacja
Podtyp kliniczny

Synonimy
Familial amyloid nephropathy due to fibrinogen A alpha-chain variant
Amyloidoza fibrynogenu A łańcuchów alfa
Dziedziczna amyloidoza nerek spowodowana wariantem fibrynogenu A łańcuchów alfa
Dziedziczna nefropatia amyloidowa spowodowana wariantem fibrynogenu A łańcuchów alfa
Rodzinna nefropatia amyloidowa spowodowana wariantem fibrynogenu A łańcuchów alfa
Fibrinogen A alpha-chain amyloidosis
Hereditary amyloid nephropathy due to fibrinogen A alpha-chain variant
Hereditary renal amyloidosis due to fibrinogen A alpha-chain variant

Kod ORPHA
93562

Kod OMIM
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Kod ICD10
E85.0

Kod ICD11
5D00.2Y

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*Źródło

orphanet