

## 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

#### Klasifikacja

Podtyp kliniczny

#### Synonimy

Familial amyloid nephropathy due to fibrinogen A alpha-chain variant  
Amylidoza fibrynogenu A łańcuchów alfa  
Dziedziczna amylidoza 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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