Opis choroby *

Definicja

A rare, systemic amyloidosis characterized by slowly progressive renal dysfunction, increased serum creatinine, mostly normal urine analysis with no significant proteinuria and associated heart disease. Cardiac involvement presents as hypertrophic obstructive cardiomyopathy, left ventricular outflow tract obstruction, coronary artery disease and conduction system abnormalities. Histology reveals renal tubular atrophy, interstitial fibrosis, glomerular sclerosis, and medullar amyloid deposits.

Dane

Klasyfikacja
Choroba

Synonimy
Apolipoprotein A-IV amyloidosis
Amyloidoza apolipoproteinowa A-IV

Kod ORPHA
439232

Kod OMIM
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Kod ICD10
E85.8

Kod ICD11
5D00.2Y

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*Źródło

orphanet