

## Opis choroby \*

### Definicja

A form of hereditary cerebral hemorrhage with amyloidosis characterized by an age of onset of 45 years of age, progressive Alzheimer's disease-like dementia, and lobar intracerebral hemorrhage in some patients. This subtype is due to a mutation in the *APP* gene (21q21.2), encoding the beta-amyloid precursor protein. This mutation causes an increased accumulation of amyloid-beta protein in the walls of the arteries and capillaries of the meninges, cerebellar cortex and cerebral cortex, leading to the weakening and eventual rupture of these vessels.

### Dane

#### Klasyfikacja

##### Podtyp kliniczny

#### Synonimy

ABeta amyloidosis, Flemish type  
Amyloidoza zależna od ABetaA21G  
Beta amyloidoza, typ flamandzki  
Dziedziczny krwotok mózgowy z amyloidozą, typ flamandzki  
HCHWA, typ flamandzki  
ABetaA21G-related amyloidosis  
HCHWA, Flemish type  
Hereditary cerebral hemorrhage with amyloidosis, Flemish type

#### Kod ORPHA

324718

#### Kod OMIM

605714

#### Kod ICD10

I68.0\*

#### Kod ICD11

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#### \*Źródło

orphanet