

## Opis choroby \*

### Definicja

A rare tumor of pancreas caused by mutations in the *GCGR* gene characterized by pancreatic alpha cell hyperplasia, pancreatic neuroendocrine tumors and markedly increased serum glucagon levels in the absence of a glucagonoma syndrome. Clinical manifestations may include abdominal pain, pancreatitis, fatigue, diarrhea, and diabetes mellitus.

### Dane

#### Klasyfikacja

Choroba  
Mahvash disease  
Choroba Mahvasha

#### Kod ORPHA

438274

#### Kod OMIM

619290

#### Kod ICD10

E16.3

#### Kod ICD11

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#### \*Źródło

orphanet