

## Opis choroby \*

### Definicja

A rare neurodegenerative disease characterized by extrapyramidal symptoms (rigidity, tremor, bradykinesia) and dementia, typically beginning in the fifth or sixth decade of life and progressing to a vegetative state with pelvicurral flexion contractures within few years. Oculomotor signs, olfactory dysfunction, and autonomic disturbances may also be observed. Neuropathological hallmarks are frontotemporally accentuated cerebral atrophy, as well as neurofibrillary tangles and neuronal loss in a characteristic distribution in cortical and subcortical regions. The disease is endemic to the Pacific island of Guam.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

G-PDC

Choroba Guama

Choroba Lytico i Bodiga

Kompleks parkinsonizm-demencja-ALS

PDALS

Stwardnienie zanikowe boczne -parkinsonizm-demencja Guama

Guam disease

Guam parkinsonism-dementia complex

Lytico-Bodig disease

#### Kod ORPHA

90020

#### Kod OMIM

105500

#### Kod ICD10

G12.2

#### Kod ICD11

6D85.Y

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#### \*Źródło

orphanet