

DISEASE:
Parkinson-dementia complex of Guam

NAME:	Parkinson-dementia complex of Guam
DESCRIPTION:	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 pelvicular 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.
ORPHACODE:	90020
SYNONYMS:	G-PDC Guam disease Guam parkinsonism-dementia complex Lytico-Bodig disease
XREF(S):	<u>Orphanet</u> <u>OMIM</u> <u>ICD-10</u>
ANALYTE(S):	<u>PARK7</u> <u>TRPM7</u>
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