

DISEASE:
Huntington disease-like 1

NAME:	Huntington disease-like 1
DESCRIPTION:	A rare, genetic, human prion disease characterized by adult-onset neurodegenerative manifestations associated with a movement disorder and psychiatric/behavioral disturbances. Patients typically present personality changes, aggressiveness, manias, anxiety and/or depression in conjunction with rapidly progressive cognitive decline (presenting with dysarthria, apraxia, aphasia, and eventually leading to dementia) as well as ataxia (manifesting with gait disturbances, unsteadiness, coordination problems), Parkinsonism, myoclonus, and/or chorea. Additional features may include generalized spasticity, seizures, urine incontinence and pyramidal abnormalities.
ORPHACODE:	157941
SYNONYMS:	Early-onset prion disease with prominent psychiatric features HDL1
XREF(S):	Orphanet ICD-10 OMIM
ANALYTE(S):	PRNP
CREATED:	13 May 2019 - 01:02
CHANGED:	22 Jun 2023 - 16:14

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