



Consensus clinical management guideline for pantothenate kinase- associated neurodegeneration (PKAN)

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Methods:

Guideline development procedure:

This guideline largely reflects the consensus opinion of clinical experts in PKAN. In addition, peer-reviewed publications were systematically searched for, evaluated, and their conclusions incorporated into this document when applicable.

A draft guideline was developed and submitted for review in four phases (approved by the OHSU IRB as protocol e8165). The first was to an international group of clinical experts, each with more than 10 years of experience with PKAN. Based on their recommendations, revisions were made and a second phase of review was sought by leaders of international patient advocacy groups. After further revisions were incorporated, a draft was shared with people with PKAN and their family members from nine affected families who commented further. Finally, an open call to the NBIA community was made via the NBIAcure.org website for final reviews and comments.

Comprehensive, systematic review of the literature

PubMed (<http://www.ncbi.nlm.nih.gov/pubmed>) was searched January–February 2016 using the following search terms: ‘PKAN’, ‘pantothenate kinase-associated neurodegeneration’, ‘HSS’, ‘Hallervorden-Spatz syndrome’, ‘NBIA’, and ‘neurodegeneration with brain iron accumulation.’ Limited consideration was made of publications utilizing aggregated data from multiple NBIA disorders or in which a diagnosis of PKAN could not be confirmed based on the information provided.

The guideline is based on expert consensus, due to limited available evidence.

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Reference to original guideline:

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