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Autosomal recessive cerebellar ataxia with late-onset spasticity

INSERM

Source

INSERM. (1999). *Orphanet: an online rare disease and orphan drug data base. Autosomal recessive cerebellar ataxia with late-onset spasticity. ORPHA:352641*

Autosomal recessive cerebellar ataxia with late-onset spasticity is a rare, genetic neurodegenerative disease characterized by childhood or adolescent-onset of cerebellar ataxia with dysarthria which slowly progresses and associates pyramidal signs, including lower limb spasticity, brisk reflexes, and Babinski and Hoffman signs. Patients typically present cerebellar ataxia with development of increasing asymmetric spasticity in upper and lower limbs, and variable axonal sensory or sensorimotor neuropathy. Additional heterogeneous features, including pes cavus, scoliosis, and abnormalities of the brain (e.g. cerebral atrophy), may also be associated.