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Dentatorubral-Pallidoluysian Atrophy

National Cancer Institute

Source

National Cancer Institute. *Dentatorubral-Pallidoluysian Atrophy*. NCI Thesaurus. Code C122653.

A rare, autosomal dominant inherited progressive neurodegenerative disorder. It is caused by a mutation in the ATN1 gene, resulting in a combined degeneration of the dentatorubral and pallidoluysian systems. It can appear at any age, but it usually affects individuals between 20 and 30 years and leads to death within 10-15 years. The clinical presentation depends on the age of the affected individual; juvenile patients develop severe progressive myoclonus epilepsy and cognitive decline, whereas adult patients develop ataxia, choreoathetosis and dementia.