

Complete Information on Dentatorubral pallidoluysian atrophy with Treatment and Prevention

Dentatorubral pallidoluysian atrophy (DRPLA) is an autosomal predominant neurodegenerative disease. It is characterized by respective combinations of ataxia, choreoathetosis, myoclonus, epilepsy, and dementia as easily as a broad scope of ages at onslaught. In patients under the age of 20, dentatorubral pallidoluysian atrophy presents as seizures, ataxia, myoclonus, as well as progressive mental deterioration. In patients over the age of 20, dentatorubral pallidoluysian atrophy is suspected when a person develops ataxia, choreoathetosis, dementia, and psychiatric disturbances. A positive family history confirms the diagnosis. The size of the repeat transmitted to the next generation depends upon the size of the parent's repeat and the sex of the transmitting parent. Therefore, it is always important to evaluate both parents of an affected individual even if they appear to have no symptoms of DRPLA.

Dentatorubral pallidoluysian atrophy is sometimes initially thought to be Huntington disease. Patients with the liberal myoclonus epilepsy phenotype had larger expansions and an early age at onslaught. Furthermore, most of the patients with the progressive myoclonus epilepsy phenotype inherited their expanded alleles from their affected fathers. A possible diagnosis of dentatorubral pallidoluysian atrophy can be devastating for a family to experience - their once healthy child, or young adult, will begin to have seizures, involuntary movements, loss of control over voluntary movement, and delusions. Dentatorubral pallidoluysian atrophy as well as other genetic conditions, exhibits a phenomenon known as anticipation. Anticipation means that the disease increases in severity and presents at a younger age of onset with each successive generation.

Patients with Dentatorubral pallidoluysian atrophy have liberal disease, which means symptoms get worse over moment. Children with symptoms, however, normally gain from having a diagnosis established. The cardinal features of dentatorubral pallidoluysian atrophy are involuntary movements and dementia (inability to clearly think, confusion, poor judgement, failure to recognize people, places, and things, personality changes) regardless of the age of onset. A history of ataxia, epilepsy, and mental retardation in children, combined with a positive family history, are often the presenting signs of this condition. Adult onset dentatorubral pallidoluysian atrophy presents with ataxia, choreoathetosis, dementia, and psychiatric disturbances. There is currently no cure for dentatorubral pallidoluysian atrophy. Epilepsy is treated with anti-seizure medication.

About the Author

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