

**A GUIDE FOR PATIENTS**

# **LITERATURE**

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literature page



# ❖ Ataxia

## ➤ Disorders of the Cerebellum: Ataxia, dysmetria of Thought, and the Cerebellar Affective Syndrome

Many diseases involve the cerebellum and produce ataxia, characterized by incoordination of balance, gait, extremity, eye movements, and dysarthria. Cerebellar lesions do not always manifest with ataxic motor syndromes, however. The cerebellar cognitive affective syndrome (CCAS) includes impairments in executive, visual-spatial, and linguistic abilities, with affective disturbance ranging from emotional blunting and depression to disinhibition and psychotic features. The cognitive and psychiatric components of the CCAS, together with the ataxic motor disability of cerebellar disorders, are conceptualized within the dysmetria of thought hypothesis. This concept holds that a universal cerebellar transform facilitates the automatic modulation of behavior around a homeostatic baseline, and the behavior being modulated is determined by the specificity of anatomic subcircuits, or loops, within the cerebrocerebellar system. Damage to the cerebellar component of the distributed neural circuit subserving sensorimotor, cognitive, and emotional processing disrupts the universal cerebellar transform, leading to the universal cerebellar impairment affecting the lesioned domain.



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## ➤ Falls in degenerative cerebellar ataxias

We retrospectively and prospectively assessed the frequency and characteristics of falls in patients with degenerative cerebellar ataxias. The results show that falls occur very frequently in patients with degenerative cerebellar ataxias and that these falls are serious and often lead to injuries or a fear of falling. Clinicians should be aware of this problem in ataxia patients and should try to prevent falls. © 2005 Movement Disorder Society

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## ➤ Ataxia

This article introduces the background and common etiologies of ataxia and provides a general approach to assessing and managing patients with ataxia.

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## ➤ Spinocerebellar Ataxia

Ataxia is the absence of voluntary muscle coordination and loss of control of movement that affects gait stability, eye movement, and speech. Spinocerebellar ataxia (SCA) is an inherited (autosomal dominant), progressive, neurodegenerative, and heterogeneous disease mainly affecting the cerebellum.

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## ➤ **Brain and induced pluripotent stem cell-derived neural stem cells**

The ataxia telangiectasia mutated (ATM) kinase is a key transducer of the cellular response to DNA double-strand breaks and its deficiency causes ataxia-telangiectasia (A-T), a pleiotropic genetic disorder primarily characterized by cerebellar neuropathy, immunodeficiency, and cancer predisposition.

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## ➤ **Clinical analysis of adult-onset Spinocerebellar Ataxia in Thailand**

Non-ataxic symptoms of spinocerebellar ataxias (SCAs) vary widely and often overlap with various types of SCAs. Duration and severity of the disease and genetic background may play a role in such phenotypic diversity. We conducted the study in order to study the clinical characteristics of common SCAs in Thailand and the factors that may influence their phenotypes.

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## ➤ **Riluzole in Cerebellar Ataxia: A Randomized, Double-blind, Placebo-controlled pilot trial**

The pleiotropic effects of riluzole may antagonize common mechanisms underlying chronic cerebellar ataxia, a debilitating and untreatable consequence of various diseases.

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