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# Neurology and Neurosurgery

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## Ataxia Center

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### What is Ataxia?

Ataxia is typically defined as the presence of abnormal, uncoordinated movements. This usage describes signs & symptoms without reference to specific diseases. An unsteady, staggering gait is described as an ataxic gait because walking is uncoordinated and appears to be 'not ordered'. Many motor activities may be described as ataxic if they appear to others, or are perceived by patients, as uncoordinated.

Ataxia can also refer to a group of neurological disorders in which motor behavior appears uncoordinated. Walking, speaking clearly, swallowing, writing, reading, and other activities that require fine motor control may be abnormal in patients with ataxia. Ataxia may result from abnormalities in different parts of the nervous system or different parts of the body, such as ataxic movements due to orthopedic injuries or pain from arthritis or muscle injury.

### What causes ataxia?

Ataxia may result from abnormalities in different parts of the nervous system, including the central nervous system (brain and spinal cord) and peripheral nervous system (roots and nerves that connect the central nervous system to muscles, skin, and the outside world). When patients experience abnormal walking or uncoordinated use of their hands or arms, dysfunction of the cerebellum is often responsible. The cerebellum is a rounded structure attached to the brainstem with a central portion (vermis) and two lateral lobes (cerebellar hemispheres). It sits beneath the

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back of the cerebral hemispheres (occipital cortices). The outer surface of the cerebellum is a continuous layer of nerve cells called the cerebellar cortex. The cortex is a three-layered sheet of neurons that are extensively interconnected and have a highly regular geometric organization. The cerebellar cortex receives information from most parts of the body and from many other regions of the brain. The cerebellum integrates this information and sends signals back to the rest of the brain that enable accurate and well-coordinated movements.

Although unsteady gait may result from problems in different parts of the nervous system or of the body, abnormal walking due to cerebellar dysfunction has distinct features that are usually recognizable. Persons with an ataxic gait due to cerebellar dysfunction keep their legs further apart than normal, referred to clinically as a 'broadened base'. They often stagger and resemble persons who have ingested excessive alcohol. The resemblance of ataxia to inebriation is not a coincidence as alcohol is known to affect the main nerve cells in the cerebellum. Although brief alcohol-induced staggering is usually reversible, repeated exposure to high doses of alcohol may cause degeneration of neurons in the cerebellum and result in persistent ataxia. A main group of cerebellar neurons is unusually susceptible to different forms of injury, including other toxins, prolonged seizures, and lack of oxygen. Cerebellar ataxia differs from gait problems due to abnormalities in other parts of the nervous system, such as the abnormal gait seen in Parkinson's disease, normal pressure hydrocephalus, or different forms of spasticity in the legs. Cerebellar ataxia is also distinguishable from abnormal walking due to pain and/or muscle or orthopedic abnormalities in the hips, legs, or feet.

## REQUEST AN APPOINTMENT

### Maryland Patients

To request an appointment or refer a patient, please contact the Johns Hopkins Ataxia Center at [410-616-2816](tel:410-616-2816).

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