

# Disability Awareness Begins With You



## ATAXIA

Ataxia describes a lack of coordination while performing voluntary movements. It may appear as clumsiness, inaccuracy, or instability. Movements are not smooth, and may appear disjointed or jerky. Ataxia may affect any part of the body. When it affects the arms and hands, it may cause tremor due to over-correction of inaccurate movements, past-pointing when an attempted reach overshoots the target, and poor performance of regular, repeated movements, such as hand clapping. When ataxia affects mechanisms of walking, there will be instability with a tendency to fall. As a result, the child usually adopts a wide-based gait, with the feet spread further apart than the hips. This is done in an attempt to compensate for the instability. The gait may appear "drunken." Balance may also be affected; the affected child may fall spontaneously or be unable to compensate for variations in the ground or a mild push from the side. When ataxia affects speech, it leads to "scanning" speech. In this form of speech, the voice is relatively monotone, often with a breathy sound accompanied by unusual accelerations or pauses between syllables. When ataxia affects the eyes, rapid shifts of gaze to look at a particular object often miss. On careful examination, the eyes may be observed to overshoot or undershoot their mark, with "catch-up" movements.

Ataxia is usually caused by damage to the cerebellum or to its inputs or outputs. The cerebellum is a large structure at the lower back of the brain, just above and behind where the spinal cord enters the skull at the bottom.

The cerebellum exists in essentially all vertebrate animals.

Separate parts of the cerebellum are concerned with control

of arm movements, eye movements, trunk stability, balance, and gait. There are theories describing the function and purpose of the cerebellum, but these issues have not yet been resolved. In general, many researchers believe that the cerebellum receives sensory information from the entire body, as well as a copy of the motor commands being sent to move the body. It integrates this information using a densely interconnected network of nerves in order to determine the interactions between sensations coming from different parts of the body and the expected results of movement. The resulting information is then sent to motor areas of the brain. This is done to presumably adjust the motor commands in order to compensate for the location of the limbs in space and the interaction of the forces generated by different parts of the body. The cerebellum appears to have a set of signals that tell it when movement errors occur, and these errors are used to make rapid corrections and thereby learn to improve performance.

### Damage to the Cerebellum

When the cerebellum is damaged, the effects may often be understood by determining the regions that sustained the injury. A "focal" injury or an injury to a small local part of the cerebellum may affect only arm movements, balance, gait, or eye movements. This type of injury may be seen if there is a stroke, migraine, or tumor. A more "global" injury may result in loss of a particular cell type. The Purkinje cells are large and complex cells that perform much of the integration in the cerebellum. These cells seem to be particularly susceptible to injury. Certain poisons, medicines, and genetic diseases may lead to injury or loss of the Purkinje cells, causing symptoms that may

simultaneously affect many functions of the cerebellum. In some cases, a child's cerebellum may not have formed properly in the first place. Often the symptoms are relatively mild, as other brain areas may have taken over some lost functions (since parts of the cerebellum may have been absent during development of the rest of the brain).

In some cases, the metabolic disorders that cause ataxia may be treated with controlled diets and special medications. If the ataxia is due to a toxin, avoiding the toxin and particular anti-toxin measures may be appropriate. If due to a tumor, then surgery may be necessary. If ataxia is due to a stroke, it is important to prevent further strokes and minimize the current injury.

Most other causes of ataxia do not have specific treatments, and there is no replacement for cerebellar cells that die or which never developed in a proper manner. The most effective treatment often consists of physical therapy to train and strengthen muscles to compensate. Gait and balance training may also be helpful. The use of a cane, crutches, or walker is often beneficial. In some cases, adapted utensils and other tools may be helpful.

There are many diseases that lead to ataxia. Determining which disease is the cause for a particular child's ataxia may be very difficult, and requires a careful series of steps planned by an experienced physician. Often, the diagnosis cannot be made immediately, but must await careful examination of the progression or resolution of symptoms over time. In some cases, however, it is important to make the diagnosis rapidly so that treatment may be started before further damage occurs.

Speech therapy may sometimes be helpful for ataxic speech. Ataxia of eye movements rarely requires treatment.