WHAT IS ATAXIA?

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Ataxia is defined as a loss of voluntary motor control and is often used as an umbrella term to classify a group of diseases that effects movement coordination. Ataxia can affect all age groups, with the age of first ataxia symptoms varying widely from early childhood to late adulthood. While not all ataxia diseases are created equal, there are common symptoms of ataxia diseases that include slurred speech, stumbling, falling, and incoordination. Often there is a progressive nature to these diseases which overtime can affect a person's ability to walk, talk, use fine motor skills, and may ultimately be life-shortening.

What causes Ataxia?

Most disorders that result in ataxia are found to have degeneration, or atrophy, of the cells in the part of the brain called the cerebellum. The cerebellum is located at the back of the head. Its function is to coordinate voluntary muscle movements and to maintain posture, balance, and equilibrium.

The spine can also be affected. The terms cerebellar degeneration and spinocerebellar degeneration may be used to refer to this type of damage to the nervous system.

The various abnormal genes that cause ataxia do have something in common: they make abnormal proteins that affect nerve cells, primarily in the cerebellum and in the spinal cord. They may also affect other parts of the brain.

Types of Ataxia

Sporadic ataxias. Ataxias of this type usually begin in adulthood and have no known family history.

***Hereditary ataxias*.** These ataxias are caused by a defect in a gene that is present from the start of a person's life and can be either dominantly inherited or recessively inherited. Recessive disorders commonly cause symptoms to begin in childhood rather than in adulthood.

Genetic testing is now available. Friedreich's ataxia is recessively inherited and occurs in childhood but can have an adult onset in up to one third of patients. Dominant ataxia often begins in the twenties or thirties or sometimes even later in life.

Hereditary ataxias are degenerative disorders that may progress over a number of years. How severe the disability depends on the type of ataxia, the age of onset of symptoms, and other factors that are poorly understood.

What are the signs and symptoms of Ataxia

Symptoms and time of onset may vary according to the type of ataxia. Each individual may experience symptoms differently.

Typically, the most common are:

- Balance and coordination are affected first
- Incoordination of hands, arms, and legs
- Slurring of speech
- Wide-based gait
- Difficulty with writing and eating
- Slow eye movements

How is Ataxia Diagnosed

In addition to a thorough medical history, family history, and complete neurological and physical examination, the following diagnostic procedures may be performed:

- **Laboratory tests** (including blood and urine studies)
- **Magnetic resonance imaging (MRI).** A diagnostic procedure that uses a combination of large magnets, radiofrequencies, and a computer to produce detailed images of organs and structures within the body
- **Genetic testing.** Tests performed to determine if a person has certain gene changes (mutations) or chromosome changes which are known to increase risk for certain inherited conditions

Treatment for Ataxia

At this time there is no cure for the hereditary ataxias. There is also no medication currently available which treats the specific symptom of ataxia. As such, most of the treatment trials done routinely involve the use of medications designed to treat other conditions but felt to be useful to provide a limited improvement in symptoms and signs of ataxia.

If ataxia is due to a stroke, a low vitamin level, or exposure to a toxic drug or chemical, then treatment is aimed at treating those specific conditions.

The treatment for the incoordination or imbalance mostly involves the use of adaptive devices to allow the individual to maintain as much independence as possible. Such devices may include the use of a cane, crutches, walker, or wheelchair. Physical therapy, speech therapy, and medications to help symptoms, such as tremor, stiffness, depression, spasticity, and sleep disorders, may also be beneficial.

Research is being conducted on cerebellar and spinocerebellar degeneration, including work aimed at finding the cause(s) of ataxias and ways to treat, cure, and ultimately prevent them, according to the National Institute of Neurological Disorders and Stroke.