

Ataxia Information Guide



INDIAN ACADEMY OF
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Introduction

The word “ataxia” refers to clumsiness or a loss of balance and coordination that is not due to muscle weakness. Ataxia is not a specific disease, but rather a symptom of many diverse conditions that affect the nervous system.

Ataxias are caused by degeneration of some brain parts which mainly control the balance of the body. Ataxias can also involve the peripheral nervous system and can cause muscle weakness. However, there are some types of hereditary ataxias that can involve the heart, musculoskeletal system and can also cause diabetes and sexual dysfunction.

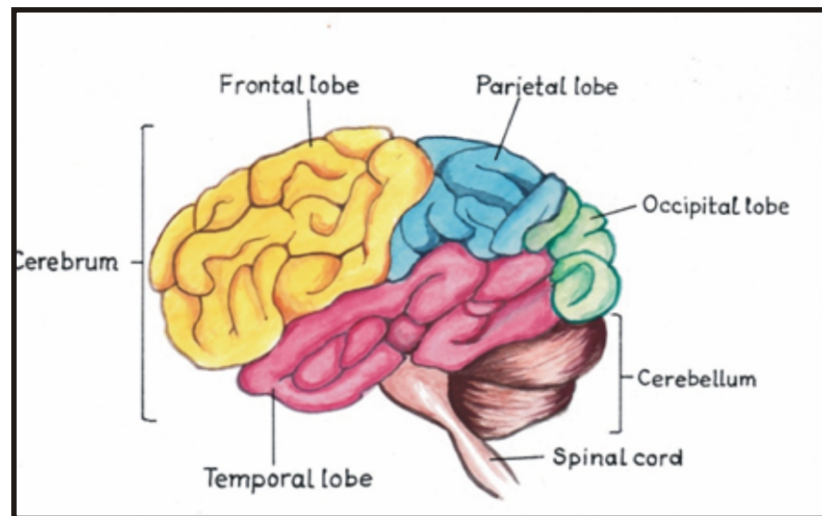


Figure: Sagittal section of the brain showing the cerebellum

Symptoms of Ataxia

People affected by ataxia may experience problems with using their fingers and hands, arms or legs, walking, speaking or moving their eyes. This loss of coordination may be caused by a number of different medical or neurological conditions.

The symptoms of ataxia may appear suddenly or gradually, and these include:

- Lack of coordination during various activities, could be from eating to buttoning of shirt.
- Difficulty in balancing and unstable gait. The persons affected with cerebellar ataxia may try to maintain balance by walking with legs wide apart. They initially experience difficulty in maintaining balance while turning. Those with sensory ataxia often look at their feet while walking.
- Swaying of the body from side to side.
- Some people may progress to experience the following symptoms:
 - Slurring of speech due to lack of coordination of facial muscles.
 - Involuntary eye movement that affects vision.
 - Difficulty swallowing.
 - Tremors while moving limbs.

Causes of ataxia

Damage to different parts of the nervous system can cause ataxia but many cases are caused by damage to a

part of the brain called the cerebellum, which serves as the balance and coordination centre. Damage to brain or its parts may be due to stroke, infection, intoxication with alcohol and drugs, tumours, multiple sclerosis, AIDS, lack of vitamins, or trauma. It may be hereditary also. Consult a neurologist to diagnose the exact cause of ataxia.

Different types of ataxia

Ataxia may be induced by drug/alcohol abuse, by deficiency of some nutritional element, trauma to certain brain parts, or due to viral infection. There may be some genetic (hereditary/sporadic) causes also.

The following is the list of some of the many medical and neurological conditions that can cause ataxia to appear more gradually:

- Problems with the liver, kidney, or thyroid gland
- Deficiencies of certain vitamins (especially Vitamin E or Vitamin B12)
- Exposure to certain drugs or toxins (heavy metals, alcohol, certain drugs)
- Sensitivity to gluten (wheat, rye and barley)
- Diabetes
- Multiple sclerosis
- Neurosyphilis
- Problems with the immune system
- Hidden cancers

Ataxias with genetic causes

The features of some hereditary ataxias are mentioned below:

Spinocerebellar Ataxias (SCAs)

Spinocerebellar ataxia is an **inherited ataxia** that mainly involves the cerebellum and the spinal cord. Other associated parts of the brain may also be affected. There are many different types of spinocerebellar ataxias, each having its specific features.

Friedreich's Ataxia

Friedreich's ataxia is an **inherited ataxia that affects sensory components** first. It usually appears around puberty. The initial symptoms often include imbalance, ataxia of the limbs, and speech disturbance. It affects the heart and may lead to diabetes as well. Spinal and foot deformities are often seen in individuals affected with Friedreich's ataxia.

Ataxia Telangiectasia

This is an **inherited type of ataxia** that affects the cerebellum. It occurs early in children. Beside the features of ataxia, the affected person also shows small, dilated blood vessels on the eye, nose or ears. The person may develop repeated infections due to reduced immunity. Death occurs due to respiratory failure, infections, or cancer.

Episodic Ataxias

These are hereditary ataxias that are often triggered by physical or mental stress or excitement. There are at

least six different types of episodic ataxias. Patients experience episodes of loss of coordination and balance. They may respond to drugs such as acetazolamide and anti-seizure medications.

Diagnosis of ataxia

A neurologist will perform an examination which can often determine whether the ataxia is caused by impairment in the cerebellum, its associated pathways, or other parts of the nervous system. Blood tests and magnetic resonance imaging (MRI) of the brain, can show whether the cerebellum or nearby parts of the brain or spinal cord have been affected by any degenerative disease that could cause ataxia.

Recovery of people affected with ataxia

Recovery depends on the cause of ataxia. If the cause can be completely treated, for example, in the case of low thyroid hormone levels/low vitamin levels, or some tumours, then the person can recover completely from the ataxia after treatment of the relevant cause.

However, if ataxia is genetic, then complete recovery of the person is not possible. In such cases, symptomatic treatment is given to slow down of progression of the disease. A variety of drugs may be used to treat gait and swallowing disorders. Physical therapy can strengthen muscles, while special devices or appliances can assist in walking and other activities of daily living.

Role of Physiotherapy

Physiotherapy aims to improve coordination skills, static and dynamic balance control of the affected persons. Exercises strengthen the individuals in repeating the daily life oriented coordination.

Diet for Ataxia affected individuals

Sound nutrition supports healthy body weight and normal bodily functions. It is important to achieve an appropriate body weight for improved movement ability and lower stress on joints. Diet for ataxia patients should contain high fibre content which may add regularity to bowel movements, which further leads to improved mood and spirit. Additionally, a multivitamin needs to be supplemented to provide MDRs (minimum daily requirements) of some specific micronutrients and to help with fatigue and general health.

Eat complex carbohydrates, e.g., unsweetened fruits, starchy vegetables, legumes, rice, and pasta. Avoid processed meats, e.g., cold cuts, hot dogs, bacon, and foods with additives and preservatives. Avoid sugar drinks and fruit juices. Drink plenty of water.

Age groups at risk of ataxia

There are no specified age groups which are at risk of ataxia. Some, inherited ataxias may possess a risk of onset before puberty (Friedreich's ataxia, SCA type 2), whereas late-onset of symptoms are also described in other types of ataxias like (SCA type 12, 3 etc.). However, ataxias associated with deficiency of some nutritional elements, trauma to certain brain parts, viral infection, etc. depend on the age of the individual.

Genetic factors

Ataxias which are caused by deficiency of certain vitamins or induced by drugs/alcohol etc. are not transmitted to next generation. These ataxias are curable. However, ataxias which have some genetic causes are transmittable to next generation, though the possibility of affected children would differ according

to the ataxia type.

Dominant ataxias: If any of the parents is affected, then the possibility of new born to get ataxia is 50%. Such transmission and types of ataxias are more common.

Recessive ataxias: Parents do not have symptoms but their children are affected. In such cases, the possibility of affected children is 25%. Family history of consanguinity (marriage within the same family or closely-related family members) is usually seen in this type of inheritance.

The natural course of hereditary ataxia

Ataxia is a progressive illness mainly involving the nervous system. The course of the disease may vary and depending upon the type of ataxia, quantum of genetic dysfunctions and other comorbidities in the affected individual. Development of disability in later stages is a feature of most of ataxias.

Disclaimer:

This brochure is for the general information of the public and the patients. People should not self-medicate themselves with the medicines and treatments mentioned here. Before taking any of the medications mentioned in the information brochure, please consult your neurologist.

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