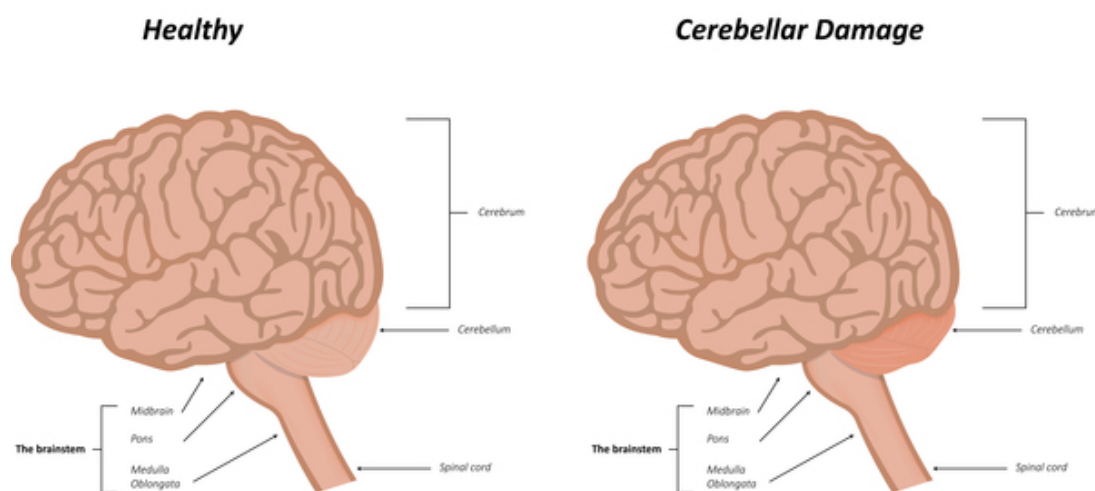


Ataxia in Childhood — Symptoms and Treatment

[See online here](#)

Ataxia results from the loss of voluntary muscle coordination, leading to unsteady gait, clumsiness, and imbalance. The symptoms of ataxia may be caused by neurologic disease, metabolic disease, or pseudo ataxia. Neurologic problems that may lead to ataxia include disorders of the cerebellum, basal ganglia, cerebral cortex, or deep sensation pathway. Pseudoataxia also sometimes occurs in response to weakness, acute illness, or anxiety.

Cerebellar Ataxia



Introduction to Ataxia

Ataxia may be classified as episodic, acute, intermittent, or chronic. Acute ataxia in children is caused by central nervous system (CNS) tumors, trauma, CNS infection, toxins, metabolic dysfunction, or stroke. Recurrent ataxia may be due to metabolic dysfunction, seizures, basilar artery migraine, or toxins. Chronic ataxia is usually the result of hereditary ataxia, CNS tumors, congenital anomalies, hydrocephalus, or metabolic disorders.

Acute Ataxia

Acute ataxia is usually due to trauma, infection, intoxication, or stroke. Head and neck trauma can both lead to acute ataxia.

Acute cerebellar ataxia

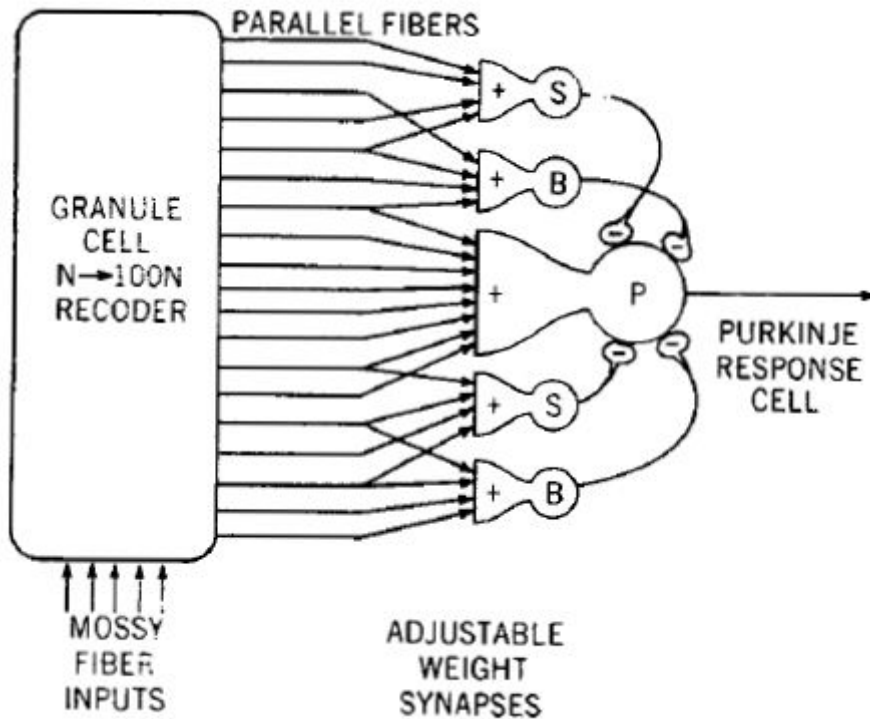


Image: "Model of Cerebellar Perception" by Albus, J.S., NIST. License: [Public Domain](#)

Acute cerebellar ataxia is the most common cause of acute ataxia in children. It is more common in children under the age of 5 years following an infection. It is usually benign and self-limited. Careful examination and work-up are needed to exclude more serious, life-threatening causes of ataxia.

Acute intoxication

Acute intoxication is a common cause of childhood ataxia due to the ingestion of medications affecting the CNS. Children present with altered mental status, ataxia, and behavior changes. **Anticonvulsants, alcohol, benzodiazepines, illicit drugs, and lead are common medications causing ataxia in children.**

Tumors

Tumors are a common cause of cerebellar ataxia. Cerebellar tumors or tumors of the posterior fossa (e.g., hemangioblastoma or neuroblastoma) can interfere with cerebrospinal fluid drainage, leading to early elevated intracranial tension. **Patients complain of headache, vomiting, blurring of vision, and focal neurological lesions.** Acute edema and hemorrhage lead to disturbed consciousness. Ataxia develops from cerebellar compression.

Paraneoplastic syndromes such as Opsoclonus-myoclonus syndrome (OMS)

Paraneoplastic syndromes are degenerative disorders that are triggered by the immune system in response to a tumor. Opsoclonus-myoclonus syndrome is a rare condition that can be associated with neuroblastoma. Its presentation includes opsoclonus, body

myoclonus, and truncal titubation, as well as ataxia. Encephalopathy and sleep problems may also arise.

Acute stroke

Acute stroke in children with sickle cell disease, systemic lupus erythematosus, nephrotic syndrome, or homocystinuria is a well-known cause of acute ataxia. **Hemorrhage due to vertebral artery dissection is the most common cause of stroke in children.**

The occurrence of cerebrovascular events leads to the deprivation of oxygen and nutrients to some parts of the brain, leading to brain damage that causes ataxia.

Intracranial hemorrhage

Intracranial hemorrhage in children can be the result of trauma or vascular malformation. It can lead to life-threatening elevation of intracranial tension and, subsequently, ataxia.

CNS infections



Image: "Varicella" by BenduKiwi. License: [Public Domain](#)

Both current and resolving CNS infections can lead to ataxia. Viral or bacterial agents with autoimmune-mediated cerebellitis can cause temporary ataxia in children; these usually resolve with no permanent sequelae. **Common viruses include Epstein-Barr, herpes simplex, varicella, parvovirus B19, mumps, and influenza.**

Cerebellar abscess, encephalitis, meningitis, and acute post-infectious demyelinating

encephalomyelitis are all causes of acute infectious ataxia. Patients present with fever, elevated intracranial tension, focal neurological symptoms, seizures, altered consciousness level, and ataxia. Acute postinfectious demyelinating encephalomyelitis sometimes presents with ataxia, as well as seizures and sensory and motor abnormalities.

Labyrinthitis

Labyrinthitis describes inflammation of the labyrinth due to viral or bacterial infection. It may complicate otitis media, leading to hearing loss, vertigo, and loss of balance.

Guillain-Barré syndrome (GBS)

Guillain-Barré syndrome usually causes motor paralysis, but ataxia may develop due to a loss of cerebellar sensory input.

Other Causes

Other causes of acute ataxia include inborn errors of metabolism, hypoglycemia, tick paralysis, and conversion disorder.

Hereditary Ataxia

There are many other types of autosomal dominant ataxia, most of which result from a gain of function mutation due to trinucleotide repeat expansion. They commonly present during adulthood, though are sometimes present during childhood. **A family history of autosomal dominant ataxia is the key determining factor.**

Autosomal recessive ataxia is common in children and may also manifest in adults. Sensory polyneuropathy, as well as other forms of organ dysfunction, is a common presentation. The most common hereditary ataxia is Friedreich ataxia.

Friedreich's Ataxia

Friedreich's ataxia is **due to the expansion of the GAA repeat in the FXN gene responsible for iron clusters, with resultant iron overload in the mitochondria.** Patients present with ataxia, weakness, absent reflexes, and dorsiflexion of the toes. Other associated presentations may include scoliosis, visual and auditory dysfunctions, cardiomyopathy with conduction abnormalities, and diabetes mellitus. Magnetic resonance imaging (MRI) scan shows late atrophy of the cerebellum and a thin cervical spinal cord.

Ataxia Telangiectasia

Ataxia telangiectasia, also known as Louis-Bar syndrome, is considered the second-most common cause of autosomal recessive ataxia. It is **caused by a mutation in the ATM gene responsible for managing DNA repair.** The mutation leads to cerebellar ataxia, telangiectasias, oculomotor apraxia, delayed puberty, dysarthria, and diabetes. The immune system is also impaired, increasing the risk of infections and cancer. An MRI scan shows cerebellar atrophy.

Subacute and Chronic Ataxia

Most children presenting with subacute or chronic ataxia have a long-standing disorder such as nutritional issues or inflammatory, autoimmune, infectious, and endocrine diseases. Vitamin B12 and folate deficiencies are common causes of sensorineural ataxia in adults and children.

Clinical Evaluation of Ataxia

Any child presenting with ataxia should be evaluated for the following:

- History of infection, trauma, or medications
- Family history of ataxia or neurological disease, metabolic disease, headache, behavioral changes, prior motor development, and onset of ataxia

Vital signs correlating with increased intracranial tension include bradycardia, hypertension, and abnormal respiration, while fever correlates with infections. An examination should exclude life-threatening danger signs including disturbed consciousness, meningism, and bulging fontanelles.

The examination should evaluate for:



[Image](#): "illustration of the altered walking gait of a woman with cerebellar disease" by André Thomas. License: [Public Domain](#)

- **Acute cerebellar ataxia: abnormal gait, normal position and vibration sense, and negative Romberg sign.** Cerebellar lesions affect gait, speech, and voluntary movements. Patients will present with wide-stance or staggering gait, speech dysarthria, truncal position abnormalities, and over- or under-shooting of voluntary movements during a finger-to-nose test or rapid alternating movements.
- **Pseudo-ataxia:** patients present with **weakness, areflexia, acute disseminated encephalomyelitis, and multiple sclerosis.**
- **Infection:** fever, neck rigidity, positive Kernig's sign and Brudzinski's sign.
- **Labyrinthitis and ear vestibular neuritis:** confirmed by performing the Dix-Hallpike maneuver.

- **Cranial nerve abnormalities:** suggest **posterior fossa tumor compressing the cranial nerves**, as they originate from the brain stem or Miller Fisher syndrome.
- **Weakness, abnormal gait, and ataxia or pseudo-ataxia:** as in Guillain-Barré syndrome, myasthenia gravis, and tick paralysis.
- **Impaired proprioception:** seen during sensory examination in patients with sensorineural ataxia.

Investigations of Ataxia

A urine toxicology screen should be the first step in the pediatric evaluation of ataxia. Accidental ingestion of drugs or medications may be responsible for acute ataxia in many children.

Basal metabolic profile and blood gases are important to exclude life-threatening metabolic abnormalities. Amino acid determinations of blood and urine, pyruvate level, ammonia levels, and urine organic acid are also useful for determining inborn errors of metabolism. Diabetes can be screened using a random blood glucose level.

Imaging of Ataxia

A computed tomography scan is the preferred study in the emergency room setting. It is useful for detecting posterior fossa lesions including hemorrhage, tumors, and traumatic lesions. An **MRI allows better visualization of the brain and brain stem but is less useful in emergency cases.** Imaging is indicated in cases with acute loss of consciousness, focal neurological signs, or evidence of increased intracranial pressure.

Management of Ataxia

Management of children with acute ataxia is supportive. Most of the cases resolve within a few weeks. Prednisolone and IV immunoglobulin have been tried for persistent cases. Intermittent and chronic cases are difficult to manage. Transcranial magnetic stimulation and deep brain stimulation have been attempted but with a lack of beneficial evidence.

References

Childhood Cerebellar Ataxia; J Child Neurol. 2012 Sep; 27(9): 1138-1145

[Approach to the child with acute ataxia](#) via uptodate.com

[Diagnostic approach to the ataxic child](#) via aacpdm.org

[Ataxia with Identified Genetic and Biochemical Defects](#) via emedicine.medscape.com

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