Ataxia in Childhood — Symptoms and Treatment

Ataxia results from disturbed voluntary muscle coordination, leading to an unsteady gait, clumsiness, and imbalance. It could be due to neurological or metabolic disease or pseudo ataxia due to weakness, acute illness or anxiety. Neurological causes of ataxia include disorders of the cerebellum, basal ganglia, cerebral cortex or the deep sensation pathway.

Cerebellar Ataxia

Introduction to Ataxia

Acute ataxia in children is caused by CNS tumors, trauma, CNS infection, toxins, metabolic causes or stroke. Recurrent ataxia can be due to metabolic causes, 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

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

Acute cerebellar ataxia is the most common cause of acute ataxia in children. It is more common in children less than 5 years following infections. It is usually benign and self-limited. Careful examination of work up 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 CNS. Children present with altered mental status, ataxia and behavior changes. Anticonvulsants, alcohol, benzodiazepines, illicit drugs and lead are common medication 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, are known to interfere with the CSF drainage leading to early elevated intracranial tension. Patients complain of a headache, vomiting, blurring of vision and focal neurological lesions. Acute edema and hemorrhage lead to a disturbed conscious level. Ataxia develops from cerebellar compression.

Paraneoplastic syndromes such as Opsoclonus-myoclonus syndrome (OMS)

These are degenerative disorders that are triggered by the immune system in response to a tumor. Opsoclonus-myoclonus syndrome (OMS) is a rare condition that can be associated with neuroblastoma present with opsoclonus, body myoclonus, and truncal
titubation, as well as ataxia. Encephalopathy and sleep problems can arise.

**Acute stroke**

Acute stroke in children with sickle cell disease, systemic lupus erythematosus, nephrotic syndrome and homocystinuria is a well-known cause of acute ataxia. **Hemorrhage due to vertebral artery dissection is the most common in children.** The occurrence of cerebrovascular events leads to deprivation of oxygen and nutrients to some parts of the brain that leads to brain damage causing ataxia.

**Intracranial hemorrhage**

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

**CNS infections**

Both current and resolving infections lead to ataxia. Viral or bacterial agents with autoimmune-mediated cerebellitis can cause temporary ataxia in children which usually resolves with no permanent sequelae. **Common viruses include; EBV, 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, disturbed conscious level and ataxia. Acute post-infectious demyelinating encephalomyelitis sometimes present with ataxia as well as seizures, sensory and motor abnormalities following infection due to demyelination.

Labyrinthitis
Labyrinthitis describes the inflammation of the labyrinth due to viral or bacterial infection complicating otitis media leads to hearing loss, vertigo and loss of balance.

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

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

Hereditary Ataxia
Autosomal dominant ataxia is numerous, most of which is due to a gain of function mutation due to trinucleotide repeat expansion mutation. They commonly present during adulthood, but less commonly can present during childhood. The family history of autosomal dominant ataxia is the key determining factor.

Autosomal recessive ataxia: are common in children and they can manifest early or later in adulthood. Sensory polyneuropathy, as well as other organ dysfunction, are common presentations. 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. Cases 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. An MRI shows late atrophy of the cerebellum and thin cervical spinal cord.

Ataxia Telangiectasia
Ataxia telangiectasia is also called Louis-Bar syndrome and 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 shows cerebellar atrophy.

Subacute and Chronic Ataxia
Most of the children presenting with subacute or chronic ataxia have long-standing disorders including; nutritional, inflammatory, autoimmune, infectious and endocrine
Clinical Evaluation of Ataxia

Any child presenting with ataxia should be evaluated for the following:
History of infection, trauma, medications, family history of ataxia or neurological disease, family history of a metabolic disease, headache, behavioral changes, prior motor development and the onset of ataxia.

**Note:** Vital signs correlating with increased intracranial tension include bradycardia, hypertension, and abnormal respiration, while fever correlates with infections. The examination should exclude life-threatening danger signs including; disturbed conscious level, meningism, and bulging fontanelles.
The examination should evaluate for:

- **Acute cerebellar ataxia:** abnormal gait, normal position and vibration sense and negative Rhomberg sign. Cerebellar lesions will affect the gait, speech and voluntary movements. Patients will present with wide based or staggering gait, speech dysarthria, truncal position abnormalities and over or under-shooting of voluntary movements with a finger to nose test or rapid alternating movements.
- **Pseudo-ataxia:** patients with weakness, areflexia, ADEM and multiple sclerosis.
- Infection: fever, neck rigidity, positive Kernig’s sign and Brudzinski’s sign.
- Labyrinthitis and ear vestibular neuritis 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.
- Motor examination will be remarkable for weakness, abnormal gait, and ataxia or pseudo-ataxia in Guillain-Barré syndrome, myasthenia gravis, and tick
paralysis.
- Sensory examination is noticeable for impaired proprioception in patients with sensorineural ataxia.

Investigations of Ataxia

A **urine toxicology screen should be the first step in child evaluation for 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 easily with a random blood glucose level.

Imaging of Ataxia

A CT scan is the preferred study in the ER 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 tried but with a lack of beneficial evidence.

References

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

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