Ataxia

Definition / Supporting Information

Ataxia refers to disorders marked by difficulties in smoothly performing co-ordinated voluntary movements, which may affect any body part, often affecting gait. In most children with acute-onset ataxia, a benign cause can be identified based on history, examination and investigations. There are both static and progressive causes of chronic ataxia. Clinical assessment focuses on excluding serious and treatable causes including central nervous system (CNS) infection, CNS inflammation and intracranial lesions.

Keywords / also known as: gait, balance difficulty, speech difficulty

Essential History

In paediatric patients, ataxia most frequently presents acutely.

- Time course (acute < 72hours)

Ask about:

- Recent or recurrent infection
  - Chicken pox, fever, rash, respiratory symptoms, vomiting and symptoms of ear infection (eg, discharging ear)
- Recent immunisation
- History of intoxication / toxin ingestion
  - Medication, alcohol, illicit drugs
- History of head and neck trauma
- Vertigo
- Associated symptoms:
  - Change in mental status including drowsiness, confusion, irritability and hallucinations
  - Double vision in older children
  - Obvious squint
  - Slurred speech
  - Difficulty in balance (eg, sitting or standing)
  - Difficulty in fine motor skills (eg, use of cutlery or handwriting)
  - Headache
- Signs (early, late, or intermittent) of elevated intracranial pressure (ICP):
  - Headache particularly worse:
    - During the night-time
    - Waking from sleeping

        - With coughing
        - When straining
        - When lying down
    - Recurrent vomiting especially early morning
    - Visual disturbances, including reduced acuity, problems with visual fields or diplopia
    - Cushing response (elevated systolic blood pressure (BP) accompanied by bradycardia) is a late, pre-terminal sign of raised ICP

Aetiology:

Acute post-infectious cerebellar ataxia (ACA), a toxin ingestion and Guillain-Barré syndrome are responsible for nearly 80% of cases.

- Acute-onset of symptoms suggests vascular (stroke), toxin-ingestion, or infectious (para- or post-infectious) cause
- Tumours and immune-mediated processes are typically subacute in progression
    - Acute haemorrhages into an existing tumour are the exception
    - Tumour symptoms are usually reported to have developed over the course of days
        - Subtle signs of weakness or lack of co-ordination may have been present for weeks or months
- In acute cerebellitis (ACA), ataxia is most prominent on awakening and improves over hours to days
- Migraine may manifest primarily as vertigo in children and in teenage girls
    - Family history of migraine, propensity to motion sickness, normal neurological examination between episodes
    - In children < 4 years old
        - Abrupt onset of unsteadiness lasting seconds to minutes
    - In adolescents
        - Isolated vertigo or headache and vertigo
- Labyrinthitis (primary or secondary to otitis media) presents as acute-onset of ataxia and nystagmus
    - Children appear more ill than in acute cerebellar ataxia
    - History of vertigo, which is usually, but not always, rotatory
        - Vertigo is constant, of sudden-onset and lasts for several hours to days before gradually improving
    - Prominent vomiting and purposeful stillness to avoid provoking symptoms
- Family history
    - Metabolic disease
    - Hereditary ataxia
    - Degenerative conditions

‘Red Flag’ Symptoms and Signs

A normal neurological exam is a pertinent negative finding. Involvement of brainstem and cerebellar structures in acute ataxia results in abnormalities in both history and physical examination. Move from the least to most invasive elements of examination, as child may be uncooperative.

Ask about:

- Vital signs

Look for:

- Behaviour
  - Is the child comfortable, agitated, reluctant to move, irritable?
- Decreased sensorium (eg, conscious state, orientation) or focal neurology
- Thorough neurological examination is mandatory
- Signs of raised intra cranial pressure
- Evidence of meningeal irritation (meningeal signs)
  - Neck stiffness, photophobia, discomfort with movement
- Eyes
  - Examine fundi if possible for papilloedema
  - Abnormal eye movements (eg, dancing eyes)
- Gait difficulties, standing posture
  - Romberg test to examine dorsal columns of the spinal cord
  - Result is positive (suggesting damage to the dorsal columns) when swaying or a fall occurs
- Asymmetric or unilateral weakness
- Difficulties with speech (eg, dysarthria, dysphasia), swallowing or drooling suggestive of bulbar dysfunction
- Features suggestive of acute cerebellar ataxia (ACA):
  - Cerebellar signs
  - Gait abnormalities
    - Staggering or wide-based
  - A negative Romberg test
  - Preservation of vibration and joint position sense
- Features suggestive of pseudoataxia include:
  - Ataxia, weakness and absent deep tendon reflexes
    - Consider Gullian-Barré syndrome
  - Ataxia, cranial nerve involvement and absent deep tendon reflex
    - Consider Miller Fisher syndrome
  - Mixed upper motor neurone and lower motor neurone lesions suggestive of acute disseminated encephalomyelitis or multiple sclerosis

- Localising signs are NOT suggestive of ACA
- Nystagmus (eg, labyrinthitis, vestibular neuronitis)
- ENT assessment
- Exclude signs of infection:
  - Fever
  - Vomiting
  - Meningism

Differential Diagnosis / Conditions

Table 1: Causes and Classification of Ataxia in Children.

Table 1

Causes and Classification of Ataxia in Children.

Acute (<72 hours duration, previously well child)	Episodic (recurrent ataxia)	Chronic
▪ Post infectious* ▪ Toxins* ▪ Tumours* ▪ Trauma ▪ Metabolic ▪ Infections ▪ Vascular (Stroke) ▪ Immune inc ADEM ▪ Opsoclonus myoclonus Syndrome ▪ Labyrinthitis ▪ Epileptic ▪ Conversion disorder	▪ Toxin ingestion ▪ Basilar artery migraine ▪ Seizure disorder ▪ Metabolic ▪ Channelopathies, e.g. CACNA1A	▪ Brain tumours ▪ Hydrocephalus ▪ Metabolic ▪ Nutritional ▪ Congenital malformations ▪ Hereditary ataxias ▪ Functional

*Most common causes of Acute Ataxia

Investigations

To be undertaken by specialist practitioners (eg, Emergency Department / Paediatric and / or Paediatric Neurology Team(s)):

- Laboratory investigations to consider in a child with acute ataxia

Table 2

Investigation for Ataxia in Children

Screening	Indications / comments	Tests
Toxicology screening	Probably the most useful diagnostic test for children with acute ataxia. Consider even when the source of ingestion may not be immediately apparent.	Urine screen for drugs and alcohol Blood-specific drug levels (eg, anti-epileptic drugs)- informed by drug history of medications in the home (may require discussion with GP). Discussion with toxicologists will be helpful.
Hypoglycaemia	Consider this in all cases. Can be detected quickly with a bedside test.	Blood glucose
Metabolic work-up	Strongly consider if the history suggests: episodic acute ataxia AND other clues supporting an IMD (family history and impaired conscious level). Inherited Metabolic Disease (IMD) and genetic investigations are unlikely to be helpful (if first presentation, normal child development and no similar family history).	Urine- toxicology, amino acids and organic acids. Blood- blood gas, full blood count, liver function tests, plasma lactate, ammonia, acyl-carnitines and biotinidase
Cerebrospinal fluid (CSF) examination (do not delay treatment)	When a CNS infection is suspected, CSF should be obtained. Prior neuroimaging required if suspicion of raised ICP present. A significant pleocytosis (raised cell count) suggests meningitis or encephalitis. CSF protein is usually elevated in GBS, but may be normal in as many as 20% of children within a week of onset. Oligoclonal bands and elevated CSF/serum immunoglobulin index can be found in ACA, ADEM and multiple sclerosis.	Cell count Protein Glucose and Lactate (paired with serum) Culture and sensitivity Oligoclonal band (paired with serum) CSF immunoglobulin (paired with serum)

ACA, acute cerebellar ataxia; ADEM, acute disseminated encephalomyelitis; CNS, central nervous system; GBS, Guillain-Barré syndrome; ICP, intracranial pressure; IMD, inherited metabolic disease.

Neuroimaging is recommended for all patients with acute ataxia with or without other signs. However, urgent imaging within hours is required in certain circumstances.

Indications for urgent neuroimaging:

- Altered mental status
- Focal neurological signs
- Cranial neuropathies
- Obvious asymmetrical ataxia
- Signs of raised ICP
- History of trauma

Computed tomography (CT) identifies:

- Space-occupying lesions
- Trauma
- Haemorrhage

Magnetic resonance imaging (MRI) is better at detecting:

- Posterior fossa lesions
- Ischaemia and myocardial infarction (with diffusion-weighted MRI sequences)
- Cerebellar abscess
- Demyelinating diseases and brainstem encephalitis

Whilst urgent MRI may be preferable, it may be necessary to perform a CT in the first instance if there is likely to be a significant delay (several hours) in obtaining an MRI.

Treatment Approach

Acute post-infectious cerebellar ataxia (ACA) a toxic ingestion and Guillain-Barré syndrome are responsible for nearly 80% of cases. Those with acute post-infectious cerebellar ataxia resulting from varicella or viral illness can be discharged following senior review, and reviewed by a general paediatrician in 1–2 weeks, with appropriate safety netting around signs of concerns. However, those children who have unexplained ataxia require admission for investigation and discussion with a local paediatric neurology team.

To be undertaken by specialist practitioners (eg, Paediatric / Paediatric Neurology Team(s)):

- Localising signs
- Raised ICP
- History not typical of acute post infectious cerebellar ataxia
- Child requiring care beyond the comfort level of the hospital

Treatment is directed at the underlying diagnosis.

Specific Treatment

- Acute stroke
  - Decompression of posterior fossa hemorrhage (neurosurgical team)
  - Vertebral artery dissection
    - Early anticoagulation with heparin seems to reduce the chance of stroke progression
- Acute disseminated encephalomyelitis
  - Consider immune modulating therapy such as corticosteroids, intravenous immunoglobulin (IVIG), or plasma exchange
  - Immunological therapy may hasten recovery, but may not affect ultimate outcome
- Infections such as cerebellar abscess or brainstem encephalitis
  - Initial treatment with broad antimicrobial therapy as well as antiviral therapy (aciclovir for herpes simplex virus (HSV) infection) until causative agent is identified
  - Neuromodulation with IVIG and / or corticosteroids may be indicated for brainstem encephalitis
- Guillain-Barré syndrome and Miller Fisher syndrome
  - Treatment with IVIG (preferred), or plasma exchange indicated in children who have become non-ambulant, or have features of respiratory or bulbar compromise
  - IVIG should not be used in patients who are immunoglobulin A (IgA) deficient
- Acute post-infectious cerebellar ataxia
  - Full recovery is typical, but can take as long as 3 to 6 months
  - No evidence that immune therapy such as corticosteroids alters outcome

When to Refer

Refer urgently to specialist practitioners (eg, Emergency Department / Paediatric / Paediatric Neurology Team(s)) if:

- Altered mental status
- Focal neurological signs
- Cranial neuropathies
- Obvious asymmetrical ataxia and absent reflex
- Signs of respiratory distress or failure
- Signs of raised ICP
- History of trauma

When to Admit

- Ischaemic stroke
  - Admission to specialist paediatric centre
- Guillain-Barré syndrome
  - For monitoring of vital signs and respiratory function

Escalate care to specialist practitioners (eg, Paediatric / Paediatric Neurosurgery / Paediatric Neurology Team(s)) if:

- Neurological consultation
  - Posterior fossa haemorrhage
    - Urgent consultation
  - Stroke
  - Brain tumours
    - Neurosurgery and oncology
  - Acute disseminated encephalomyelitis (ADEM)
  - Brain stem encephalitis
  - Cerebellar abscess
  - Guillain-Barré syndrome
  - Opsoclonus-myoclonus syndrome

‘Safety Netting’ Advice

Patients assessed to have acute post infectious cerebellar ataxia should be reviewed by their local paediatric team in 1–2 weeks

Patient / Carer Information

Please note: whilst these resources have been developed to a high standard they may not be specific to children.

- Ataxia (Webpage), the NHS website
- Ataxia (Website), Ataxia UK

Resources

Medical Decision Support

Hughes RA, Swan AV, van Doorn PA. Intravenous immunoglobulin for Guillain-Barré syndrome. Cochrane Database Syst Rev 2010;16(6):CD002063.

Management of the ataxias towards best clinical practice, 3rd ed. Ataxia UK.

Suggested Resources

Please note: these resources include links to external websites. These resources may not have national accreditation and therefore PCO UK cannot guarantee the accuracy of the content.

Prasad M, Whitehouse W, Ong MT, et al. Fifteen-minute consultation: the Child with acute ataxia. Arch Dis Child Educ Pract ED. 2013. Dec;98 (6):217-23.

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Gieron-Korthals MA, Westberry KR, Emmanuel PJ, et al. Acute childhood ataxia: 10-year experience. J Child Neurol 1994;9(4):381–384.

Jones HR. Guillain-Barré syndrome: perspectives with infants and children. Semin Pediatr Neurol 2000;7(2):91–102.

Rust RS. Multiple sclerosis, acute disseminated encephalomyelitis, and related conditions. Semin Pediatr Neurol 2000;7(2):66–90.

Ryan MM, Engel EC. Acute ataxia in childhood. J Child Neurol 2003;18(5):309–316.

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