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In Brief

Evaluation of Ataxia

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Ataxia, defined as impaired coordination of movement and balance, is associated with dysfunction of the cerebellum or the sensory or motor pathways connecting to it. The causes of ataxia range from a transient parainfectious process to a progressive degenerative neurologic condition. The challenge for the pediatrician is to distinguish among the possible etiologies.

The evaluation of ataxia begins with a thorough history and careful neurodevelopmental examination. The history must include the onset and duration of the symptom; family history of migraine or neurologic disease; recent infection, seizure, or head injury; and possible environmental exposure to heavy metals, gases, solvents, or medications. It is important to ask about other signs and symptoms, such as headache, vomiting, photophobia, vertigo, and altered mental status.

Distinguishing whether a young child has age-appropriate clumsiness or is truly ataxic requires familiarity with the developmental milestones related to coordinated movements. Ataxia also must be distinguished from other disorders of the nervous system or of the muscles. The child should be examined in an upright position with attention to mental status, balance, presence of nystagmus, deep tendon reflexes, and muscle tone and strength. Muscle

weakness from a myopathy or neuropathy may present with an altered gait and unstable balance, but it should be recognized on the basis of diminished muscle strength and deep tendon reflexes. Conversely, children who have spasticity may exhibit an unsteady, narrow-based gait, but they will demonstrate increased muscle tone and brisk reflexes. Movement disorders such as chorea and myoclonus, unlike ataxia, are evident even when the child is at rest.

The examiner must look for evidence of increased intracranial pressure or infection by checking for abnormal vital signs, meningismus, and papilledema. With a patient is old enough to cooperate, the neurologic examination should evaluate cranial nerve function, heel-to-knee movement, finger-to-nose movement, tandem gait, rapid alternating movement, and speech. It is particularly important to listen for dysarthria, specifically the scanning speech pattern associated with cerebellar dysfunction. Dysfunction of the cerebellar vermis presents with truncal ataxia—imbalance in the sitting position and head titubation. Dysfunction of the cerebellar hemispheres presents with a gait that veers toward the involved side and dysmetria of the ipsilateral extremity. Attenuation of sensory input because of peripheral nerve or posterior column damage leads to a careful high-stepping gait and a positive Romberg sign. Children who have sensory ataxia have problems with fine finger movements rather than dysmetria.

A typical presentation of acute ataxia is the sudden onset of a wide-based gait, with swerving and unsteadiness that is recognized easily by parent and physician. The most common

causes are drug ingestion and postinfectious cerebellitis. The differential diagnosis includes head trauma, vascular events, brain tumor, hydrocephalus, the Miller Fisher variant of Guillain-Barré syndrome (ataxia, ophthalmoplegia, and areflexia), labyrinthitis, seizure, and conversion reaction. Drugs associated with ataxia include phenytoin, carbamazepine, sedatives, hypnotics, and phencyclidine. Intoxication with alcohol, ethylene glycol, hydrocarbon fumes, lead, mercury, or thallium also may present with ataxia. A variety of pathogens can cause postinfectious cerebellitis, but it is observed most frequently after varicella.

The evaluation of the child who has acute ataxia is based on the history and findings on physical examination, but it also should include a complete blood count, measurement of electrolytes, toxicologic screening of blood and urine, brain imaging, and lumbar puncture. If magnetic resonance imaging or computed tomography demonstrates a mass lesion, hydrocephalus, or other intracranial abnormality, cerebrospinal fluid (CSF) evaluation may be deferred. The presence of cells in the CSF may indicate infection, and elevated CSF protein levels are seen with Guillain-Barré syndrome and multiple sclerosis. Ataxia, especially if accompanied by opsoclonus or myoclonus (“dancing eyes, dancing feet”), can be associated with neuroblastoma. If this diagnosis is suspected, examination of urine for vanillylmandelic acid/homovanillic acid (VMA/HVA) and imaging of the chest and abdomen are recommended.

If the initial evaluation fails to establish a diagnosis, what appears to be acute ataxia actually may be the initial presentation of a more chronic process.

Basilar migraine, seizure, and some rare metabolic disorders (Hartnup disease, maple syrup urine disease, pyruvate decarboxylase deficiency) may manifest with acute intermittent episodes of ataxia. Evaluation again is based on the history, but it may include brain imaging, electroencephalography, and electromyography. If metabolic disease is suspected, evaluation of amino acids, acid/base balance, lactate, pyruvate, ammonia, and ketones may be helpful.

The slow evolution of ataxia in a previously healthy child warrants rapid evaluation. The most common causes of progressive ataxia are brain tumors (medulloblastoma, cerebellar astrocytoma, brain stem glioma, ependymoma) and degenerative spinocerebellar diseases, some of which present in infancy and others of which are delayed in onset until adulthood. Friedreich ataxia, an autosomal recessive disorder linked to a defect on chromosome 9, presents in the first decade of life with ataxia,

pes cavus, scoliosis, and areflexia. Ataxia telangiectasia, also autosomal recessive, is marked by an abnormality on the long arm of chromosome 11 and is characterized by progressive truncal ataxia, oculocutaneous telangiectasias, and recurrent infections of the sinuses and lungs. If magnetic resonance imaging of a child who has progressive ataxia fails to disclose an intracranial mass, referral for a detailed metabolic and genetic evaluation is warranted.

In summary, ataxia may represent a benign, transient condition or a progressive, devastating neurologic disorder. As is usually true, a careful history and physical examination, followed by a focused radiologic and laboratory assessment, most often will lead to the proper diagnosis and intervention.

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Comment: As a disorder of coordination that depends on sensory, proprioceptive, motor, and integrative neural activities, ataxia can reflect dysfunction at virtually any level of the nervous system, not just the cerebellum. It also can be difficult to distinguish between ataxia and weakness when a child presents with, for example, an abnormal gait, and the differential diagnosis, although related, is not the same. Ataxia is particularly frightening because it represents a loss of physical control for the patient, and it poses some very nasty possibilities for the pediatrician. Clinical medicine is not easy, but it is less difficult if we go back to the basics, with a careful history and physical examination. It's amazing how often we can get to the answer without magnetic resonance imaging.

*Henry M. Adam, MD
Editor, In Brief*

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