Abstract
Ataxia refers to motor incoordination that is usually most prominent during movement or when a child is attempting to maintain a sitting posture. The first part of the review focuses on the anatomic localization of ataxia — both within the nervous system and without — using a combination of historical features and physical findings. The remainder of the review discusses etiological considerations that vary depending on the age group under consideration. In infancy, certain specific diseases, such as opsoclonus myoclonus ataxia syndrome, must receive special mention because the underlying disease process may be amenable to surgical intervention. In the toddler- and school-age groups, certain conditions (such as stroke and acute cerebellitis) require immediate recognition and imaging, whereas others (such as post-infectious ataxia and concussion) require close follow-up. Finally, mention must be made of diseases outside of the central nervous system that can present with ataxia, such as Guillain-Barré syndrome.

The word ataxia is derived from the Greek word *ataktos*, which means “lack of order.” Ataxia is characterized by disturbances in the voluntary coordination of posture and movement. In children, it is most prominent during walking (the *sine qua non* being a staggering gait with impaired tandem), but it can also be present during sitting or standing, or when the child is performing movements of the arms, legs, or eyes.

This review focuses on the etiology and diagnostic considerations for acute ataxia, which for the purposes of this discussion refers to ataxia with a symptom evolution time of less than 72 hours.¹

Motor coordination requires sensory input from muscles and joints. This sensory information is transmitted through myelinated axons, via the posterior columns of the spinal cord, to higher centers in the cortex and basal ganglia.
These central structures generate their modulating output, which is conveyed through motor tracts that lay in the brain stem and spinal cord and then conveyed through peripheral nerves to the relevant muscle groups, thereby closing the action loop. The cerebellum exerts a modulating control at various levels of this loop. Furthermore, the vestibular system in the inner ear monitors angular and linear accelerations of the head. This information is also conveyed to the cerebellum, which utilizes this feedback to maintain posture. Ataxia can, therefore, arise from disturbances in various parts of the nervous system (eg, the cerebellum, brain stem, spinal cord, and peripheral nerves), as well as the inner ear (Figure 1). To equate ataxia with disease of the cerebellum alone is, therefore, inaccurate. Finally, certain non-neurological conditions, such as musculoskeletal pathology, can lead to clumsy gait. There are no clear data regarding incidence or gender prediction of acute ataxia in the pediatric age group.

CLINICAL EVALUATION

History

Important questions about patient history that may lead to an etiology are summarized in Table 1. As can be seen, there is significant overlap in symptomatology between different disease states, so it may be difficult to distinguish “central” from “peripheral” causes by history alone. Symptom onset may occur in just a few minutes or over the course of hours or days.

Physical Examination

Physical examination should focus on identifying life-threatening conditions that may require immediate intervention. Presence of confusion, hallucinations, mood disturbances, or somnolence must be noted. Such symptoms may indicate toxic ingestion, demyelinating diseases such as acute disseminated encephalomyelitis (ADEM), stroke, or meningo-encephalitis. The presence of papilledema, pupillary dilation, and lateral rectus palsy indicates elevated intracranial pressure. Motor examination should focus on eliciting weakness in an anatomically significant pattern, such as one-half of the body or both lower extremities. Often, a younger child with hemiparesis or paraparesis will present with ataxia as opposed to focal weakness of the affected body part (ie, “paretic ataxia”).

A detailed cerebellar exam should follow. Dysfunction of midline cerebellar structures causes truncal ataxia and head tremor. Disorders of the lateral aspects of the cerebellum cause limb ataxia, which can be evaluated by the finger-nose or finger-finger test, intention tremor, and dysdiadochokinesia. In the lower extremity, one can perform the heel-shin test to look for incoordination. Cerebellar incoordination tends to be most prominent when movements are performed slowly. Ataxia caused by pathology of the cerebellum is not exacerbated by eye closure; therefore, a Romberg test will be negative.

Presence of nystagmus should be noted with relevant details. Nystagmus may arise from pathology in the vestibular system (“peripheral” type) or the central nervous system (“central”). Certain specific types of nystagmus, such as opsoclonus, may rarely be noted.

Finally, a comprehensive sensory exam and an examination of deep tendon reflexes should be performed to rule out diseases of peripheral nerves or roots (ie, “sensory” ataxia). Plantar responses are not affected by diseases of the cerebellum; hence, a positive Babinski test is indicative of disease processes in the
### TABLE 1.
Questions about Patient History that May Lead to an Etiology of Ataxia

<table>
<thead>
<tr>
<th>History</th>
<th>Potential Etiology</th>
<th>Localization of Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>Basilar migraine</td>
<td>Brain stem</td>
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<tr>
<td>Frontal/temporal</td>
<td></td>
<td>Cerebellum</td>
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<td>Occipital</td>
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<tr>
<td>Acute onset in an unsupervised toddler or following a fall</td>
<td>Ingestion of toxins</td>
<td>Brain</td>
</tr>
<tr>
<td>Vomiting</td>
<td>Basilar migraine, stroke&lt;sup&gt;†&lt;/sup&gt;</td>
<td>Brain stem, cerebellum</td>
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<td></td>
<td>Space-occupying lesions&lt;sup&gt;†&lt;/sup&gt;</td>
<td>Posterior fossa</td>
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<td></td>
<td>Concussion</td>
<td>Labyrinth</td>
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<td></td>
<td>Benign paroxysmal vertigo</td>
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<tr>
<td>Recent history of fever, rash, gastrointestinal illness, infectious</td>
<td>ADEM</td>
<td>Brain stem</td>
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<tr>
<td>contacts</td>
<td>Acute post-infectious cerebellar ataxia</td>
<td>Cerebellum</td>
</tr>
<tr>
<td></td>
<td>Acute cerebellitis&lt;sup&gt;†&lt;/sup&gt;</td>
<td>Cerebellum</td>
</tr>
<tr>
<td></td>
<td>Kawasaki disease, polyarteritis nodosa, Henoch-Schöenlein purpura</td>
<td>Cerebellum</td>
</tr>
<tr>
<td>Recent immunizations</td>
<td>Guillain-Barré syndrome&lt;sup&gt;†&lt;/sup&gt;</td>
<td>Spinal roots and nerves</td>
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<td></td>
<td>Acute post-infectious cerebellar ataxia</td>
<td></td>
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<tr>
<td>Recent history of viral illness</td>
<td>Acute post-infectious cerebellar ataxia</td>
<td>Cerebellum</td>
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<td></td>
<td>Acute cerebellitis&lt;sup&gt;†&lt;/sup&gt;</td>
<td>Cerebellum</td>
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<tr>
<td></td>
<td>Miller-Fisher syndrome</td>
<td>Brain stem</td>
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<tr>
<td>Nystagmus</td>
<td>Benign paroxysmal vertigo</td>
<td>Peripheral vestibular system</td>
</tr>
<tr>
<td></td>
<td>Neuroblastoma&lt;sup&gt;†&lt;/sup&gt;</td>
<td>Sympathetic ganglia</td>
</tr>
<tr>
<td></td>
<td>Multiple sclerosis</td>
<td>Brain stem</td>
</tr>
<tr>
<td></td>
<td>Drug ingestion</td>
<td>Brain stem, cerebellum</td>
</tr>
<tr>
<td></td>
<td>Stroke&lt;sup&gt;†&lt;/sup&gt;</td>
<td></td>
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<tr>
<td>Head or neck trauma in an adolescent (eg, during skiing, diving,</td>
<td>Stroke</td>
<td>Brain stem</td>
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<td>martial arts)</td>
<td>Cerebellar concussion</td>
<td>Cerebellum</td>
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<tr>
<td>Seizures</td>
<td>Post-ictal ataxia</td>
<td>Brain</td>
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<td></td>
<td>Drug overdose</td>
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<td>Fluctuating drug levels</td>
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<tr>
<td>Encephalopathy (drowsiness, confusion, aggression, psychosis)</td>
<td>ADEM</td>
<td>Brain, spinal cord</td>
</tr>
<tr>
<td></td>
<td>Multiple sclerosis</td>
<td>Brain, spinal cord</td>
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<tr>
<td></td>
<td>Drug ingestion</td>
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<tr>
<td>Dizziness (true vertigo, spinning sensation)</td>
<td>Benign paroxysmal vertigo</td>
<td>Brain stem</td>
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<td></td>
<td>Basilar migraine</td>
<td>Brain stem</td>
</tr>
<tr>
<td></td>
<td>Stroke&lt;sup&gt;†&lt;/sup&gt;</td>
<td>Cerebellum or brain stem</td>
</tr>
<tr>
<td>Previous self-limiting episodes with family history</td>
<td>Genetic episodic ataxias</td>
<td>Not known</td>
</tr>
<tr>
<td>History of emotional trauma</td>
<td>Psychogenic</td>
<td>Not applicable</td>
</tr>
</tbody>
</table>

<sup>*Indicates conditions that are unlikely to present in an acute fashion.</sup>

<sup>†Indicates conditions that require immediate attention.</sup>

ADEM = acute disseminated encephalomyelitis.
upper motor neuron pathway, outside of the cerebellum.

ETIOLOGY ON THE BASIS OF AGE

Infancy

Opsoclonus Myoclonus Ataxia Syndrome

Opsoclonus myoclonus ataxia (OMA) syndrome can present as early as age 6 months. OMA syndrome is a paraneoplastic autoimmune phenomenon characterized by chaotic conjugate high-amplitude eye movements, body jerks, and truncal/limb ataxia, along with developmental regression and irritability. An underlying neuroblastoma or ganglioneuroblastoma is often recognized, although not universally present. Recognition is facilitated when the triad of symptoms manifest in close proximity to each other. However, ataxia alone may precede the eye findings, leading to diagnostic confusion and delays of months to years in initiating investigations. Nuclear scanning with metaiodobenzylguanidine scintigraphy (MIBG scan) has moderately high sensitivity, but children whose scans are negative should receive high-resolution computerized tomography (CT) or magnetic resonance imaging (MRI) of the chest and abdomen.

Children Age 1 to 4 Years (Preschool)

Acute Post-Infectious Cerebellar Ataxia

Acute post-infectious cerebellar ataxia (APCA), which accounts for up to 40% of cases of acute cerebellar ataxia in certain case series, typically occurs after a febrile illness or immunizations. A prior history of varicella was reported in up to 26% of patients by some authors. A large number of other viruses have been implicated, including coxsackie B, echoviruses, mumps, Epstein-Barr, and influenza A and B. The pathology is believed to be acute demyelination caused by cross-reacting antibodies to epitopes in the cerebellum. The disease onset can be up to 3 weeks after the systemic illness has subsided. Symptoms evolve over hours and are most prominent at initial presentation, with relatively rapid resolution over the next few days. The mental status usually remains clear throughout the course of the illness. The presence of extreme irritability should raise suspicion about the diagnosis.

Examination reveals a pure cerebellar syndrome with marked involvement of gait and significant truncal ataxia.

Accidental ingestion of drugs in children in the preschool years may account for up to 30% of cases of acute ataxia.

The average child recovers in less than 2 weeks after disease onset. It is a self-limited condition that requires no specific intervention or investigations. Cerebrospinal fluid (CSF) analysis can be performed with no risk of herniation, and it usually reveals a mild pleocytosis with negative viral and bacterial cultures. MRI tends to be normal or shows mild, non-specific changes.

Acute Cerebellitis

Acute cerebellitis may occur after a systemic illness or be the direct result of infection in the cerebellum. Common agents reported to cause acute cerebellitis are rotavirus, mycoplasma, and human herpesvirus 6. Clinical features include altered sensorium and manifestations of raised intracranial pressure, in addition to features of a pure cerebellar syndrome. CSF analysis is likely to reveal pleocytosis, and in rare cases, antibodies against an infectious agent have been demonstrated. It must be mentioned that performing a spinal tap in the face of significant cerebellar edema can be life-threatening. Imaging can reveal abnormalities suggestive of edema in the cerebellum. Fatalities have been reported from acute cerebellitis. In essence, acute cerebellitis differs from APCA by the presence, in some cases, of systemic symptoms such as fever and neck stiffness, symptoms and signs of raised intracranial pressure due to rapid compression of the fourth ventricle, and risk of death; therefore, there is a pressing need for early therapy. There is considerable overlap between APCA and acute cerebellitis, so it may be difficult to distinguish between the two conditions.

Toxic Ingestion

Accidental ingestion of drugs in children in the preschool years may account for up to 30% of cases of acute ataxia. The Purkinje cells of the cerebellum are especially susceptible to toxic injury. Accidental ingestion of anticonvulsants, lead, eucalyptus oil, insecticides such as parquat and phosphine, dextromethorphan, and shellfish poisoning may cause prominent cerebellar symptoms. Clinical features include depressed mentation or agitation, seizures, and cerebellar signs. The latter may be masked by the overall acuity of the situation. Parents should be asked to bring in all prescription medication for household members, and a urine/serum drug screen is essential in the initial battery of tests.

Benign Paroxysmal Vertigo

Benign paroxysmal vertigo (BPV) must be differentiated from benign paroxysmal positional vertigo, which is the most common cause of vertigo in adults. BPV is characterized by brief spells of vertigo and ataxia. An otherwise healthy child suddenly looks frightened, pale, and wants to hold on to a parent for support during episodes of BPV. The duration of symptoms is a few minutes at most, after which the child experiences no confusion or sleepiness. The typical age of onset is age 1 to 4 years, and resolution occurs around the age of 7 to 10 years. Examination is invariably normal between spells, as are imaging studies. A family history of migraine is found in
Acute Disseminated Encephalomyelitis

ADEM is an immune-mediated phenomenon that presents after a viral illness or immunization and is characterized by encephalopathy (confusion, irritability, somnolence, personality changes) and acute onset of multifocal neurological deficits — most commonly ataxia. Seizures, cranial nerve palsies, hemiparesis, fever, and meningismus are frequently present. Imaging reveals bright lesions in the subcortical white matter, cerebellum, and basal ganglia. CSF is abnormal about half the time, with elevated white cells and protein. ADEM is usually a monophasic illness.

Inner Ear Disease

One of the rare complications of acute otitis media is labyrinthitis, which is an extension of the middle ear bacterial infection to the inner ear or labyrinth, causing sensorineural hearing loss, tinnitus, nystagmus, and vertigo. The vertigo leads to ataxia. Fever, fullness and pain in the ear, and mastoiditis may serve as clues to the diagnosis. The presence of nystagmus in a child with acute otitis media is often a reliable clue to the presence of impending labyrinthitis, so it should be treated expeditiously. The presence of contrast enhancement of the labyrinth on MRI is a sensitive indicator of disease process in this area. CT is less reliable.

Children Age 5 to 16 years (School Age)

Concussion

Cerebellar concussion is a clinical syndrome wherein a head injury is followed by transient deficits in functioning of the cerebellum, with intact consciousness throughout. Patients typically present with a wide-based gait, truncal instability, and dysarthria. Most cases follow severe injuries, however, a few children have been noted to have cerebellar concussion following relatively minor head trauma. The pathophysiology is believed to be damage to the connections between the cerebellum and the cortex, especially the superior cerebellar peduncle, following severe head injury. Transient vasospasm is the purported mechanism behind ataxia following mild injuries. No specific intervention is required and resolution is usually complete.

Stroke

Strokes in the region of the cerebellum (posterior circulation) are easily missed, as strokes are uncommon in childhood and posterior circulation strokes present with subtle symptoms that are often non-specific, with no clear-cut motor or sensory disturbances. Posterior circulation strokes present with dizziness about three-quarters of the time, nausea/vomiting or gait disturbances about one-half of the time, and headache about one-third of the time. Onset of symptoms is abrupt. Examination may reveal ataxia with tendency to fall to the affected side, as well as vertical nystagmus, in addition to other classic cerebellar signs. Presence of altered level of consciousness, hemiplegia, and cranial nerve weakness, when present, may quickly guide the clinician to an appropriate diagnosis. MRI is the diagnostic test of choice because CT scan can miss a stroke in the cerebellum and brain stem. The majority of cerebellar strokes are caused by arteriopathy that may follow infection (such as varicella), vasculitis (such as Kawasaki disease), or dissection. Edema following a stroke is most prominent in the first 24 hours, and if it is in the region of the posterior circulation, then it is particularly ominous. Rapid swelling may compress the brain stem. The vast majority of such patients will die without surgical intervention. Therefore, all cerebellar strokes must be closely observed in an intensive care setting.

Multiple Sclerosis

Multiple sclerosis (MS) is an autoimmune demyelinating disease that affects different parts of the nervous system over time. About 50% of the children younger than the age of 5 years who have MS and 5% to 15% of adolescents who have MS present with acute ataxia, making it a relatively frequent disease manifestation. Children with MS have more frequent disease relapses than their adult counterparts, and they experience significant cognitive decline and severe residual disability in their early adult years. Early imaging in the form of MRI with administration of contrast material and referral to a neurologist for initiation of therapy is essential.

Prescription Drug Use, Substance Abuse, and Poisoning

Adolescents who present with acute ataxia should be questioned regarding drugs of abuse as well as therapeutic agents they may be using for medical purposes. Benzodiazepines, phenytoin, and carbamazepine are the anticonvulsants most likely to cause ataxia following acute intoxication. Free as well as total levels of drugs must be measured in these situations. Antineoplastics, such as fluorouracil (5-FU), cytarabine (ara-C), and methotrexate, may induce acute cerebellar damage. Drugs of abuse that may cause ataxia are toluene, cocaine, heroin, and phencyclidine. Toluene intoxication can occur due to glue-sniffing or acute exposure in areas that are poorly ventilated. Cocaine use predisposes to cerebellar infarctions, as does envenomation due to a scorpion sting.

Basilar Migraine

Children with basilar migraine present with an aura consisting of ataxia, diplopia, tinnitus, tingling in their extremities, or alteration in the level of their consciousness that lasts from 5 minutes to 1 hour. A severe, pounding headache follows within 1 hour of these
symptoms and is typically accompanied by nausea/vomiting and photophobia/phonophobia. The headache may be unilateral or bilateral and is relieved by sleep. The average duration of the headache is 30 minutes to 3 days. Examination is unremarkable, with no clear cerebellar signs other than ataxia. Initial episodes warrant an MRI.

**Guillain-Barré Syndrome**

Children may present with ataxia due to disease processes in the peripheral nerves that do not allow appropriate sensory signals to reach the higher coordinating centers. A classic example of this is Guillain-Barré syndrome. A preceding viral infection or gastroenteritis is noted in at least half of all cases. Children may experience severe pain in the lower extremities or back prior to the onset of weakness. Weakness and ataxia reach a nadir within 4 weeks. Rare instances may progress to involvement of the respiratory muscles and quadripareisis. Diagnostic testing may include imaging of the spinal cord to rule out myelitis and spinal fluid analysis, although in most instances the diagnosis remains a clinical one that can be made with assurance at the bedside due to the triad of ataxia, areflexia, and motor weakness. Nerve conduction studies may be performed in atypical cases to confirm the diagnosis. Coordination with a neurologist and intensivist is vital in children being prepared for treatment of Guillain-Barré syndrome.

Figure 2. Algorithm for investigating acute ataxia. ADEM = acute disseminated encephalomyelitis; CT = computed tomography; ECG = electrocardiogram; ED = emergency department; GBS = Guillain-Barré syndrome; MIBG = metaiodobenzylguanidine scintigraphy; MS = multiple sclerosis; MRI = magnetic resonance imaging.
**Episodic Ataxia**

Hereditary episodic ataxia (EA) refers to a group of dominantly inherited conditions characterized by periods of cerebellar dysfunction that lead to acute ataxia. Seven different subtypes are recognized, of which EA2 is the most common. Attacks are precipitated by fatigue or strong emotion (such as anger or sadness) and may last for hours. In addition to ataxia, patients may experience vertigo, nausea, vomiting, and (rarely) seizures. Patients may have migranous headaches and downbeat nystagmus between spells. The mutation responsible for EA2 is located on the CACNA1A gene.

**CONCLUSION**

An algorithm for work-up of acute ataxia is presented in Figure 2. Ataxia can cause significant anxiety for the child and family due to the dramatic nature of the symptoms. The fact that most children with acute onset ataxia will have rapid resolution of their symptoms must be emphasized to parents and caregivers. Collaboration with a child neurologist, radiologist, and intensive care physician may be of benefit in cases where serious intracranial pathology is a consideration.

**REFERENCES**