or CNS infection is suspected. Suspect basilar artery thrombosis in a comatose patient with "normal" results on head CT, in which the only finding may be a hyperdense basilar artery.[24] MRI or cerebral angiography is needed to make the diagnosis of basilar artery thrombosis.

SPECIAL CONSIDERATIONS IN COMA

If trauma is suspected, maintain stabilization of the cervical spine during assessment. If protection of the airway is in doubt or the coma state is likely prolonged, then protect the airway by intubating the patient. Rapid-sequence intubation techniques are discussed at length in chapter 29, Intubation and Mechanical Ventilation. **ingestions, infections, and child abuse in the appropriate clinical setting.**

Patients who have had generalized seizures and remain unresponsive may be in a continuing state of electrical seizures without corresponding motor movements. This is called **nonconvulsive status epilepticus** or **subtle status epilepticus** and can be described as electromechanical dissociation of the brain and body. **If the motor activity of the seizure stops and the patient does not awaken within 30 minutes, then consider nonconvulsive status epilepticus.** Obtain neurologic consultation and electroencephalography.[23]

TREATMENT

Treatment of coma involves identification of the cause of the brain failure and initiation of specific therapy directed at the underlying cause. Attend to airway, ventilation, and circulation. Evaluate and treat for readily reversible causes of coma, such as hypoglycemia and opioid toxicity.

Antidotes Rapid point-of-care glucose determination can identify the need for dextrose. Although thiamine should be administered before glucose infusion in patients with a suspected history of alcohol abuse or malnutrition, thiamine is not necessary for all patients. Routine use of flumazenil in coma of unknown cause is not recommended.[25] Naloxone, the opiate antagonist, is useful in coma because typical signs of opiate overdose may be absent.

Increased Intracranial Pressure If history, physical examination, or neuroimaging findings suggest increased ICP, specific steps can reduce or ameliorate any further rise in ICP. Any noxious stimulus, including "bucking" the ventilator, can increase ICP, so use paralytic and sedative agents. A general recommendation is to keep the head elevated about 30 degrees and at midline to aid in venous drainage. **Mannitol** (0.5 to 1.0 gram/kg IV) can decrease intravascular volume and brain water and may transiently reduce ICP. In cases of brain edema associated with tumor, dexamethasone, 10 milligrams IV, reduces edema over several hours. Hyperventilation with reduction of partial pressure of arterial carbon dioxide can reduce cerebral blood volume and transiently lower ICP. Current recommendations are to avoid excessive hyperventilation (partial pressure of arterial carbon dioxide ≤35 mm Hg) during the first 24 hours after brain injury. Brief hyperventilation may be necessary for refractory intracranial hypertension. Data to recommend specific therapy are lacking, and preferences among individuals and institutions vary greatly, so communicate early with consultants and admitting physicians.

DISPOSITION AND FOLLOW-UP

Patients with readily reversible causes of coma, such as insulin-induced hypoglycemia, may be discharged if home care and follow-up care are adequate and a clear cause for the episode is suspected. Admit patients with persistent altered consciousness. Most institutions depend on emergency physicians to stabilize the patient's condition and correctly assign a tentative diagnosis so that the patient may be admitted to the proper specialty service. If the appropriate service is not available, then consider transfer to another hospital after patient stabilization.

REFERENCES

The complete reference list is available online at www.TintinalliEM.com.

CHAPTER 169

Ataxia and Gait Disturbances

J. Stephen Huff

INTRODUCTION

Ataxia and gait disturbances may be symptoms of many disease processes and generally are not themselves diagnoses. **Ataxia** is uncoordinated movement. A **gait disorder** is an abnormal pattern or style of walking. The presenting problem may be articulated by the patient or family as weakness, dizziness, stroke, falling, or another nonspecific chief complaint. Such symptoms must always be viewed in the context of the patient's overall clinical picture. This chapter reviews the more common causes of acute ataxia and gait disorders (**Table 169-1**).

PATHOPHYSIOLOGY

Clinicians erroneously tend to think that ataxia and gait disorders result primarily from cerebellar lesions. However, such disorders result from many systemic or focal conditions that affect different elements of the central and peripheral nervous systems. Cerebellar lesions may indeed cause ataxia, but isolated lesions of the cerebellum are not the most common cause of these complaints. Ataxias are classified as either motor (cerebellar) or sensory.

TABLE 169-1 Common Etiologies of Acute Ataxia and Gait Disturbances

- **Systemic conditions**
  - Intoxications with diminished alertness
    - Ethanol
    - Sedative-hypnotics
  - Intoxications with relatively preserved alertness (diminished alertness at higher levels)
    - Phenytoin
    - Carbamazepine
    - Valproic acid
    - Heavy metals—lead, organic mercurials
  - Other metabolic disorders
    - Hyponatremia
    - Inborn errors of metabolism
    - Wernicke's disease
- **Disorders predominantly of the nervous system**
  - Conditions affecting predominantly one region of the CNS
    - Cerebellum
      - Hemorrhage
      - Infarction
      - Degenerative changes
      - Abscess
    - Cortex
      - Frontal tumor, hemorrhage, or trauma
      - Hydrocephalus
    - Subcortical
      - Thalamic infarction or hemorrhage
      - Parkinson's disease
      - Normal pressure hydrocephalus
    - Spinal cord
      - Cervical spondylosis and other causes of spinal cord compression
      - Posterior column disorders
    - Conditions affecting predominantly the peripheral nervous system
      - Peripheral neuropathy
      - Vestibulopathy

MOTOR ATAXIAS

Motor ataxias (also referred to as **cerebellar ataxias**) are usually caused by disorders of the cerebellum. The sensory receptors and afferent pathways are intact, but integration of the proprioceptive information is faulty. Involvement of the lateral cerebellum (one of the cerebellar hemispheres) may lead to a motor ataxia of the ipsilateral limb. Lesions affecting primarily the midline portion of the cerebellum often cause problems with axial muscle coordination, which is reflected in difficulty maintaining a steady upright standing or sitting posture.

There are many reports of lesions in what would seem to be unlikely locations producing motor ataxia. Supratentorial infarctions, particularly small, deep infarctions, and lacunae of the posterior limb of the internal capsule have been reported to cause isolated hemiataxia. It is postulated that interruption of either ascending or descending cerebellar to cortical pathways are the cause of this motor-type ataxia.[1] Small infarctions or hemorrhages in thalamic nuclei may produce a clinical picture of motor- or cerebellar-like ataxia with hemisensory loss. These effects are seen contralateral to the lesion.[2] Lesions affecting the frontal lobe, such as tumor or cystic masses, may cause a motor ataxia of the contralateral extremities through poorly understood mechanisms.[3] Nontraumatic spinal cord compression may present with gait ataxia or abnormality.[4]

SENSORY ATAXIAS

Sensory ataxias are due to failure of transmission of proprioception or position sense information to the CNS. Failure may arise from disorders affecting the peripheral nerves, spinal cord, or cerebellar input tracts. Coordinated motor performance is faulty, even though motor systems and the cerebellum are intact. Sensory ataxias may be somewhat compensated by visual sensory information. Loss of visual information leads to the observation that sensory ataxias often worsen in poor lighting conditions and may be brought out during examination.

GAIT DISORDERS

No organized classification scheme exists for gait disorders, and different authors categorize abnormal gaits in descriptive terms. A **cerebellar or motor ataxic gait** is widely based with unsteady and irregular steps, and compensation to barriers in the environment may be lacking. The **gait of sensory ataxia** resulting from loss of proprioception is notable for abrupt movement of the legs and slapping impact of the feet with each step. A variety of other terms are used to describe abnormal gaits.

An **apraxic gait** is one in which the patient seemingly has lost the ability to initiate the process of walking, an "ignition failure." This may occur with right or nondominant hemispheric lesions. Frontal lobe dysfunction may result in a similar gait and may be seen in normal pressure hydrocephalus.[5]

The term **festinating gait** is used to describe narrowly based miniature shuffling steps and is common in Parkinson's disease. An abnormal gait with outward swinging or circumabduction of the leg suggests a mild **hemiparesis** reflecting the asymmetric weakness of the proximal lower extremity muscles. Bilateral weakness of the trunk and pelvic girdle muscles may result in a **waddling gait** from failure to maintain the normal position of the pelvis relative to the lower extremities.

A functional gait disorder is one in which the patient is unable to walk normally, although all motor pathways, sensory pathways, and cerebellar functions may be demonstrated to be functioning normally. The underlying problem is often a conversion disorder. Functional gaits may be bizarre, at times resembling a person balancing on a tightrope and seemingly threatening to fall but not falling. A dramatic functional gait with flailing movements without falling actually demonstrates that strength, balance, and coordination are intact.

A unifying concept defines gait disorders according to the level of processing of neurologic information (**Table 169-2**).[6,7] The classification scheme is not ideal but does allow a thoughtful approach to patient diagnosis.

TABLE 169-2 Classification of Gait Disorders

Low-level gait disorders
Musculoskeletal problems
Arthritic gait or other joint or skeletal problems
Muscle weakness
Peripheral sensory problems
Sensory ataxic gait
Vestibular problems
Middle-level gait disorders
Hemiplegia
Paraplegia
Motor or cerebellar ataxia
Parkinson's disease
Dystonia, chorea, other movement disorders
High-level gait disorders
Senile gait (cautious gait)
Frontal ataxic gait
Apraxic gait (gait ignition failure)
Frontal disequilibrium

Low-level gait disturbance refers to disorders of proprioception or dysfunction of the musculoskeletal system. **Middle-level gait disturbance** causes distortion of appropriate interaction of postural and motor processes or synergies. This might include stroke with paralysis, cerebellar dysfunction, or diseases of the basal ganglia such as Parkinson's disease. **High-level gait disturbances** seemingly involve structures or processes that choose the appropriate responses for the support surface, body position in space, and intention of the patient. Cautious gait, apraxic gait, and the frontal gait disorder conceptually fall into this group with pathology that correlates with lesions in the frontal cortex or thalamus. This latter group is the least understood and the source of clinical confusion.

CLINICAL FEATURES

HISTORY

Collect historical information about the entire symptom constellation, and ask about headache, nausea, fever, weakness, or numbness. A history of fever, review of medication history, or family history of ataxia may lead to the diagnosis in individual cases. The nature of onset of symptoms and the time course of the process guide the pace of investigations. For example, abrupt onset of gait difficulty in a patient with severe headache, drowsiness, nausea, and vomiting should suggest an acute process within the CNS, possibly a hemorrhage into the cerebellum. The possible consequences of that diagnosis are severe and may require immediate attention. At the other extreme, a patient without significant medical history who is brought to the ED with a stumbling gait after an episode of binge drinking requires examination but may need nothing other than observation unless history or physical examination suggest trauma or some alternative cause for the symptoms.

PHYSICAL EXAMINATION

The following discussion of the neurologic examination assumes that the gait disorder is the dominating abnormality. Physical examination including testing of cranial nerves, mental status, sensation, and the motor system is necessary and may yield findings that lead to an unanticipated diagnosis.

General physical examination of a patient with ataxia or gait disturbance should include determination of orthostatic vital signs. Orthostatic hypotension may be present in hypovolemia, diabetic neuropathy, and other neurologic syndromes. Especially in the elderly, fluid replacement for simple hypovolemia may correct many symptoms of unsteadiness.

Gait testing is one of the most important parts of the directed neurologic examination. Observe the patient sitting upright in the stretcher, and then have the patient rise, stand, walk, and turn around. The patient should be asked to walk at a normal speed, then walk on the heels, and then the toes. Tandem gait is toe-to-toe walking and also tests many elements of the nervous system. **Do not assume a normal examination without observing ambulation.**

Cerebellar functions are tested by asking the patient to perform smooth voluntary movements and rapidly alternating movements. Dyssynergia (breakdown of movements into parts), dysmetria (inaccurate fine movements), or dysdiadochokinesia (clumsy rapid movements) may indicate a lateral cerebellar lesion. The rapid thigh-slapping test particularly examines rapidly alternating movements. This is correctly performed by asking the patient to pat the thigh with the palm then the back of the same hand in alternating fashion, making a sound with each rapid slap. The maneuver is performed with each hand in turn. The finger-to-nose test may be helpful in distinguishing between cerebellar and posterior column (proprioceptive) lesions. Performing this test with the eyes closed tests proprioception in the upper extremity. A test for cerebellar function that emphasizes the lower extremities is the heel-to-shin test. In cerebellar disease, the heel may initially overshoot the other shin or knee, and the action is done with a series of jerky movements. In posterior column disease, there may be difficulty locating the knee, and the movement down the shin typically weaves from side to side or falls off. Another test commonly used for cerebellar function is the Stewart-Holmes rebound sign (with sudden release of the flexed forearm, the individual fails to check the movement). Another example of rebound phenomena is when a tapped outstretched arm oscillates back and forth for several cycles.

The Romberg test is primarily a test of sensation, and if positive may distinguish sensory from motor ataxia. While standing with arms outstretched and eyes open, observe the patient for signs of unsteadiness. The feet should be narrowly spaced, and the posture should be easily maintained. The inability to maintain a steady standing posture (or, in extreme cases, a seated position) confirms that an ataxia is present but does not yet give any information about the type of ataxia. Then ask the patient to close the eyes, to eliminate visually orienting information. If the ataxia worsens with the loss of visual input, then the Romberg sign is present or positive, suggesting sensory ataxia with a problem of proprioceptive input (posterior column, vestibular dysfunction), or a peripheral neuropathy. Further neurologic examination is indicated to confirm the suspicion of sensory ataxia. In patients who show little or no change in unsteadiness with eye closure (Romberg test–negative), a motor ataxia is suggested, with possible localization of that problem to the cerebellum. Note that many normal individuals will have some small increase in unsteadiness with eye closure.

Historically, tabes dorsalis (neurosyphilis) was a common cause of sensory ataxia. In tabes dorsalis, the posterior columns and posterior spinal roots degenerate, primarily in the lumbosacral region. The loss of proprioceptive information from the lower extremities renders the patient dependent on visual cues for correct gait. The classic description is that of a patient who walks slowly with wide gait while staring at the ground. In darkness or with interruption of vision, the patient is unable to walk. The gait in this condition is peculiar, with the foot first raised and then slapped to the ground with each step. These abnormalities reflect the loss of proprioceptive information from the posterior roots and posterior columns. Consider vitamin B_{12} deficiency in patients with evidence of posterior column disease. If the deficiency is left untreated, an initial unsteady gait may progress to weakness, spasticity, and ataxia. The finding of a megaloblastic anemia may be a clue, but the neuropathy may precede the anemia.

Sensory examination in a patient with unsteady gait or movements should include position or vibration testing (posterior columns), as well as testing sensation to pinprick. Testing of the deep tendon reflexes will serve largely to discover asymmetry or spasticity that might suggest an alternative diagnosis. Acute cerebellar injury may result in muscle hypotonia for a few days or weeks.[8]

Nystagmus is seen in many different disorders due to lesions in a variety of different locations of the CNS, but the presence of nystagmus does suggest that the pathologic process is intracranial (CNS or vestibular) and not in the spinal cord or peripheral nervous system (see chapter 170, Vertigo).

DIAGNOSIS

Assuming a primary complaint of ataxia, the first task is to determine whether the ataxia is sensory or motor and whether the primary process is systemic or within the nervous system. If the ataxia is thought to result from problems within the nervous system, the next question is one of localization to the peripheral nervous system versus the CNS and perhaps to a more specific anatomic location. Finally, the tempo of the illness, comorbid diseases, and other clinical findings guide investigations and may allow a disease-specific diagnosis.

A patient with acute gait failure over hours to days needs thorough evaluation in the ED, often requiring CT scan and MRI, or lumbar puncture if cerebrospinal fluid infection is suspected. Acute ataxia or gait disturbance may also be evaluated by consultation if available, and possible admission, in contrast to a patient with gradual loss of abilities over weeks or months where outpatient referral and evaluation may be more appropriate.

SPECIAL POPULATIONS

THE GERIATRIC PATIENT

The gait changes with advancing age. A typical constellation includes gait slowing, shortening of the stride, and widening of the base. This results in the appearance of a guarded gait—that is, the gait of someone about to slip and fall. Many patients are aware of the loss of speed and adaptive balance and acknowledge the need to be careful. The nature of the senile gait is not fully understood but may represent a mild degree of neuronal loss, failing proprioception, slowing of corrective responses, or weakness of the lower extremities. Senile gait disorder is thought to exist in up to one fourth of the elderly population. Some authorities divide this disorder into components of gait ataxia with mild truncal instability and widened gait, and gait slowing with diminished spontaneous arm swing and bradykinesia.[9] However, elements of the senile gait are also found in neurodegenerative diseases, so consider the possible presence of a neurodegenerative disorder such as Parkinson's disease or normal pressure hydrocephalus in elderly patients with gait impairment.[9] Patients unable to walk or care for themselves, or with increasing falls at home need admission for supportive care.

THE ALCOHOLIC PATIENT

A history of alcoholism or malabsorption problem in the patient with ataxia or gait disorder raises the possibility of a potentially remedial nutritional problem. If acute motor ataxia is present with confusion or eye movement abnormalities, consider **Wernicke's disease** and administer IV thiamine.[10] The entity of alcoholic cerebellar degeneration (sometimes referred to as rostral vermis syndrome, because a portion of the cerebellar vermis is preferentially affected) may represent the same nutritional deficiency and not the direct toxic effects of alcohol.

CHILDREN

In evaluating children with acute ataxia or gait disorder, examination must exclude weakness and musculoskeletal disorders. The child may be awake, alert, and playful but is visibly unsteady or wobbly sitting on a stretcher. The differential diagnosis is extensive (**Table 169-3**). Acute or deteriorating presentation generally mandates an aggressive search for the underlying cause and will likely need inpatient management.[11]

Intoxications are a cause of ataxia in children, and the ingestion may be surreptitious. History should include queries about any medications in the household. Acute ataxia may follow immunizations, viral illnesses, or varicella and also has been rarely reported in the preeruptive phase of varicella.[12] Most children are in the 2- to 4-year-old range. Acute cerebellar ataxia of childhood is thought to be a postinfectious demyelinating disorder. The onset of gait ataxia is abrupt, and only occasionally is fever present at the time ataxia begins.