# Variant 1: Acute ataxia following recent head trauma. Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT head without IV contrast</td>
<td>Usually Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>CT temporal bone without IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>CTA head and neck with IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>CTV head with IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>MRA head and neck without IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>MRI head without IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>MRV head without IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>MRA head and neck without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>MRI head without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>CT head with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>MRV head with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>CT temporal bone with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>CT head without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>Radiography skull</td>
<td>Usually Not Appropriate</td>
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</tbody>
</table>

# Variant 2: Acute ataxia following recent spine trauma. Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT spine area of interest without IV contrast</td>
<td>Usually Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>CTA neck with IV contrast</td>
<td>Usually Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>MRI spine area of interest without IV contrast</td>
<td>Usually Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>MRA neck with IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>MRA neck without IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>MRI spine area of interest without and with IV contrast</td>
<td>May Be Appropriate (Disagreement)</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>Arteriography spine area of interest</td>
<td>Usually Not Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>CT myelography spine area of interest</td>
<td>Usually Not Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>CT spine area of interest with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>CT spine area of interest without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>Radiography spine area of interest</td>
<td>Usually Not Appropriate</td>
<td>Varies</td>
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</table>
**Variant 3:** Ataxia. No history of trauma. Suspected intracranial process. Stroke intervention not a consideration. Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>MRI head without and with IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRI head without IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>CT head with IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>CT head without IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>CT head without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>CTA head and neck with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>MRA head and neck without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRA head and neck without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>Arteriography cervicocerebral</td>
<td>Usually Not Appropriate</td>
<td>☢☢☢</td>
</tr>
<tr>
<td>CTV head with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☢☢☢</td>
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<tr>
<td>Ioflupane SPECT or SPECT/CT brain</td>
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<td>☢☢☢</td>
</tr>
<tr>
<td>MRV head with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRV head without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>DTPA cisternography</td>
<td>Usually Not Appropriate</td>
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</table>

**Variant 4:** Ataxia of any acuity. No history of trauma. Suspected spinal or spinal vascular process. Initial imaging.

<table>
<thead>
<tr>
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<tbody>
<tr>
<td>MRI spine area of interest without and with IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRI spine area of interest without IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRA spine area of interest with IV contrast</td>
<td>May Be Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>CTA spine area of interest with IV contrast</td>
<td>May Be Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>MRA spine area of interest without IV contrast</td>
<td>May Be Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>Arteriography spine area of interest</td>
<td>May Be Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>CT myelography spine area of interest</td>
<td>May Be Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>CT spine area of interest with IV contrast</td>
<td>May Be Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>CT spine area of interest without IV contrast</td>
<td>May Be Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>CT spine area of interest without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>Varies</td>
</tr>
<tr>
<td>Radiography spine area of interest</td>
<td>Usually Not Appropriate</td>
<td>Varies</td>
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</tbody>
</table>
ATAXIA

Expert Panel on Neurologic Imaging: Amy F. Juliano, MD; Bruno Policeni, MD; Vikas Agarwal, MD; Judah Burns, MD; Julie Bykowski, MD; H. Benjamin Harvey, MD, JD; Jenny K. Hoang, MBBS; Christopher H. Hunt, MD; Tabassum A. Kennedy, MD; Gul Moonis, MD; Jeffrey S. Pannell, MD; Matthew S. Parsons, MD; William J. Powers, MD; Joshua M. Rosenow, MD; Jason W. Schroeder, MD; Konstantin Slavin, MD; Matthew T. Whitehead, MD; Amanda S. Corey, MD.

Summary of Literature Review

Introduction/Background

Ataxia is a neurological sign and symptom that refers to loss of coordination of muscle movement and is due to dysfunction of one or more components of the nervous system [1]. Common manifestations include a wide-based, unsteady gait and poor coordination of the extremities. The causative lesion can be in the cerebellum (affecting neural information integration, coordination, and planning), the spinal cord and peripheral sensory nerves (affecting proprioception), or in the vestibular system (affecting balance and maintenance of equilibrium) [2]. Ataxia with hemiparesis can result from a cerebral infarct involving the thalamus, pons, corona radiata, or internal capsule [3]. A number of other pathological conditions may lead to symptoms that mimic ataxia, such as hydrocephalus, and should be excluded during its workup.

Clinical evaluation by history and careful neurological examination will provide the localization information necessary to guide the choice of imaging. Truncal ataxia and titubation (rhythmic and spasmodic nodding or swaying of the head or body) are seen especially, although not exclusively, with disorders that involve the midline cerebellum. If the underlying pathology involves the cord, there may be weakness, hyperreflexia, spasticity, and sensory loss in addition to ataxia. If the pathology involves the vestibulo-cerebellar system, there may be nausea, vomiting, and vertigo. Ataxia that is due to peripheral neuropathy is associated with sensory loss, with hyporeflexia, and often weakness as well.

The purpose of this document is to identify the most common clinical scenarios and the most appropriate imaging for their assessment based on the current literature. This document does not address follow-up recommendations for patients with a known underlying etiology for ataxia. Given the frequent coexistence of ataxia and other neurologic, traumatic, and vascular processes, it is important to acknowledge the overlap of symptoms with other conditions referenced in independent ACR Appropriateness Criteria documents, and others beyond the scope of this document. To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4]. Please also reference the ACR Appropriateness Criteria® topic on “Suspected Spine Trauma” [6], the ACR Appropriateness Criteria® topic on “Myelopathy” [7], and the ACR Appropriateness Criteria® topic on “Hearing Loss and/or Vertigo” [8] in the appropriate clinical context.

Special Imaging Considerations

CT myelography has supplanted fluoroscopic myelography in most circumstances; however, there may be times when fluoroscopic myelography is also performed prior to CT imaging. For this document, the procedure term “CT myelography” is used to guide the referral to the radiologist. The ultimate judgment regarding the propriety of any specific procedure, lumbar versus cervical puncture route, amount of contrast, and the extent and modality of imaging coverage must be made by the radiologist, with appropriate documentation and coding [9].

*The views expressed in this manuscript are those of the author and do not reflect the official policy of the Department of Army/Navy/Air Force, Department of Defense, or United States Government.

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Discussion of Procedures by Variant

Variant 1: Acute ataxia following recent head trauma. Initial imaging.

A new neurologic deficit in the setting of head trauma is an indication for imaging to assess for intracranial hemorrhage or acute stroke, which requires expedited management. To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4]. As the two most common head trauma guidelines do not address ataxia, post-traumatic evaluation has been maintained separately from the ACR Appropriateness Criteria® topic on “Head Trauma” [5]; however, that guideline should also be referenced in the setting of altered Glasgow Coma Scale.

Gait disturbance is a frequent occurrence following head trauma, for example from a direct blunt forceful impact or a fall. It may persist in the setting of post-traumatic encephalopathy, or if there is sustained pathology or post-traumatic complication, such as an expanding cyst or hematoma anywhere from the frontal lobe along the frontopontocerebellar tract to the cerebellum [10]. If ataxia is accompanied by vertigo, injury to the vestibular system, including the inner ear or temporal bone, should be suspected.

Ataxia may also result from traumatic vertebral artery dissection, leading to vascular compromise to the cerebellum and brainstem. The most common presentation is gait ataxia [11], but may also be hemiataxia (involving the upper and lower limb) or monoataxia (involving only one limb) [12,13].

Ataxic hemiparesis is commonly related to an insult to the internal capsule [14]. Lesions in the pons, depending on location, may lead to ataxia, hemiparesis, and/or dysarthria-clumsy hand syndrome [15].

Radiography Skull
Radiographs are not adequate for assessing etiologies of ataxia.

CT Head
Head CT without intravenous (IV) contrast is the usual initial imaging procedure for a patient with a new neurologic deficit in the setting of acute head trauma, as reflected in the American College of Emergency Physicians clinical practice guidelines [16], and is often extrapolated, but not validated, for patients presenting >24 hours from recent head trauma. This variant is maintained separately from the ACR Appropriateness Criteria® topic on “Head Trauma” [5], as ataxia does not alter the Glasgow Coma Scale, and the New Orleans Head CT criteria [17] and the Canadian CT Head Rule [18] do not apply in the presence of a new neurologic deficit. Dual phase of both noncontrast and postcontrast imaging is usually not indicated as a first-line test.

CT Temporal Bone
In a trauma setting, this study is not the first imaging test performed; it is often used to further assess an abnormality found at the time of head CT. CT of the temporal bone may be obtained or reconstructed from head CT or CT angiography (CTA) with submillimeter slices and a small field of view, allowing detailed evaluation for fractures and their complications, including integrity of the bony labyrinth. Findings suggestive of a perilymph fistula include air in the inner ear structures and opacity in the round window niche. Late-stage labyrinthitis ossificans can be detected on CT by osseous density in the usually fluid-filled space of the otic-capsule [19]. IV contrast is not necessary for these indications.

CTA Head and Neck
CTA of the head and neck is appropriate if carotid or vertebral arterial injury, including dissection, is suspected based on examination or trauma mechanism [20,21].

CTV Head
CT venography (CTV) of the head can be performed if there is suspected venous injury, although this is not a first-line test in the setting of post-traumatic ataxia.

MRI Head
MRI is complementary to CT and in particular can more clearly depict processes in the posterior fossa and brainstem, which may contribute to ataxia. MRI is not a first-line test in the setting of post-traumatic ataxia but does have a role in the setting when the patient condition is not explained by the CT findings [22].

MRA Head and Neck
MR angiography (MRA) of the head and neck is performed if there is concern for arterial pathology, including dissection, occlusion, stenosis, aneurysm, vasculitis, and arteriovenous fistulas. In a trauma setting, CTA is often the preferred study; MRA may be performed if there is need for further assessment or problem-solving after an
MRA is useful in the setting of suspected arterial dissection in a relatively stable patient, and the addition of a fat-suppressed T1-weighted spin-echo sequence can be helpful for direct visualization of a mural hematoma, with sensitivity and specificity that is superior to those of CT [26,27].

**MRV Head**
MR venography (MRV) can be performed if there is suspected venous thrombosis or stenosis. In a trauma setting, CTV is often the preferred study, and an MRV is also performed if there is need for further assessment or problem-solving after an initial CTV. MRV may be performed with or without the use of IV contrast, utilizing time-of-flight technique. Please see the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4].

**Variant 2: Acute ataxia following recent spine trauma. Initial imaging.**
The body regions covered in this clinical scenario are the cervical, thoracic, and lumbar spine. These body regions might be evaluated separately or in combination as guided by physical examination findings, patient history, and other available information, including prior imaging.

The presence of an acute neurologic change in the setting of cervical spine trauma meets the NEXUS criteria, and appropriate cervical spine stabilization precautions should be maintained [28]. There are no detailed guidelines in the setting of thoracic or lumbar spinal trauma. Please see the ACR Appropriateness Criteria® topic on “Suspected Spine Trauma” [6] in the setting of additional traumatic symptoms and the ACR Appropriateness Criteria® topic on “Myelopathy” [7] if other neurologic symptoms are present.

Cord compression can occur in the setting of fractures, malalignment, traumatic exacerbation of spondylotic myelopathy, or ossification of the posterior longitudinal ligament. Spinal epidural hematoma may occur after minor trauma, though it more often results in local or radicular pain. Less commonly, ataxia may occur as a result of loss of sensory or motor function [29-31]. Spinal cord infarct is rare but does occur and is related to aortic dissection or complications of an aneurysm, thromboembolism, or systemic hypotension.

**Radiography Spine**
Radiography has largely been supplanted by CT for assessment of traumatic spine injury. Radiographs are insufficient to evaluate causes of spinal cord compression in the setting of a new post-traumatic neurologic deficit.

**CT Spine**
CT is considered the gold standard for identification of spine fractures, outperforming radiographs in identification of spine fractures [32-38]. CT is most useful in the acute setting to detect fracture, subluxation, and dislocation, which would necessitate immediate stabilization. CT may identify substantial cord deformity or a large epidural process; however, it is limited in detection of cord injury and compressive epidural or subdural processes causing acute ataxia [39]. IV contrast is not needed for CT assessment of the spine, especially in the trauma setting [40,41].

**CTA Neck**
CTA of the neck is appropriate if carotid or vertebral arterial injury, including dissection, is suspected based on examination or trauma mechanism [20,21] and may depict injury of the anterior spinal artery [42].

**MRI Spine**
MRI is complementary to CT, allowing for more detailed assessment of the soft tissues, including ligamentous integrity, intervertebral disc injury, and spinal cord injury [43,44]; however, in the trauma setting, CT is usually the first-line test because of stabilization concerns, and MRI is performed afterward as needed for further assessment or problem-solving. Recent suggestions include using diffusion tensor imaging (DTI) to detect intramedullary lesions that may not be apparent on conventional sequences in the early stage [45,46], or if conventional MRI is otherwise unrevealing. IV contrast is not necessary for MRI assessment of the spine in a trauma setting [40,47,48].

**MRA Neck**
MRA of the neck allows evaluation of the cervical arteries (internal carotid arteries and vertebral arteries) for potential vessel injury, including arterial dissection, and can be performed either without or with IV contrast. Addition of a fat-suppressed T1-weighted spin-echo sequence can offer direct visualization of a mural hematoma [26,27].

**CT Myelogram Spine**
There is no evidence to support the use of CT myelography as the initial imaging test for post-traumatic ataxia.
Arteriography Spine
Spinal angiography is the gold standard for assessing the vascular supply to and venous drainage from the cord, especially in cases of ambiguity after MRA or CTA have been performed [49]; however, it is not a first-line test in evaluation of post-traumatic ataxia.


To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4].

Information from the patient history, physical examination, and laboratory values can often help narrow the differential diagnoses and focus the workup of the patient with ataxia. The exclusion of a posterior fossa mass lesion is often an important consideration in evaluating ataxia. The suspected mass can be primary or metastatic, and it can be intra-axial or extra-axial in location. Isolated frontal lobe and thalamic mass lesions may also present with varying manifestations of gait and limb ataxia.

Paraneoplastic cerebellar degeneration is clinically characterized by subacute or acute onset of gait and limb ataxia, dysarthria, and ocular dysmetria [50]. Paraneoplastic syndromes may be caused by any primary tumor but are most commonly associated with breast, gynecologic, and lung tumors, and with Hodgkin disease [50].

Patients with acute cerebellitis present with truncal ataxia, dysmetria, and headache. In severe cases, there may be altered consciousness, additional neurological deficits, increased intracranial pressure, hydrocephalus, and even herniation [51]. Bacterial cerebellitis may occur in association with cerebritis and meningitis. Cerebellar atrophy may be a late-stage finding [52].

Superficial siderosis usually presents with slowly progressive ataxia and hearing loss, which is due to recurrent, often silent, subarachnoid hemorrhage that over time results in hemosiderin deposition on the subpial layers of the brain and spinal cord.

Vasculitides affecting the brain chronically, such as neuro-Behçet disease, can lead to ataxia [53]. Demyelinating diseases, such as acute disseminated encephalomyelitis and multiple sclerosis, can also result in ataxia [54]. Miller Fisher syndrome is considered a variant of Guillain-Barré syndrome, characterized by a triad of ataxia, areflexia, and ophthalmoplegia.

Ataxia may be seen with substance abuse, toxicity, or nutritional deficiencies, for example, as seen with chronic ethanol abuse [55], methanol toxicity [56], and heroin use [57]. Cerebellar atrophy and infarction can occur with opiate and solvent abuse [58]. Mercury poisoning/Minamata disease can cause cortical or cerebellar lesions and atrophy or peripheral neuropathy that may lead to symptoms simulating ataxia [59,60]. Metronidazole-induced cerebellar toxicity shows increased T2 signal and reduced diffusivity in the dentate nuclei on MRI [61]; the brainstem and corpus callosum can be affected as well [62]. Cerebellar atrophy and ataxia have been seen with vitamin E deficiency [63].

Congenital malformation of the midbrain and cerebellum can present with posterior fossa symptoms, including ataxia [64,65]. Chiari I, cerebellar hypoplasias or agenesis, rhombencephalosynapsis, and Joubert syndrome are examples [66-69]. Lhermitte-Duclos disease is variously considered a neoplasm or a hamartoma, and has an association with Cowden disease [70]. Cerebellar atrophy in childhood can be due to mitochondrial disorders, neuronal ceroid lipofuscinosis, ataxia telangiectasia, GM2 gangliosidosis, among others [71].

A number of genetic or inherited syndromes or diseases have ataxia as a component, including Christianson syndrome, Niemann-Pick disease type C, neuroferritinopathy, ataxia-telangiectasia [72,73], Huntington disease, Friedreich ataxia [74-79], fragile X-associated tremor/ataxia syndrome [80,81], and the spinocerebellar ataxias [82-86].

CT Head
CT is less sensitive and specific for comprehensive evaluation of these conditions compared to MRI in the nonemergent setting. CT head with IV contrast is preferred. Dual phase of both noncontrast and postcontrast imaging is usually not indicated as the first-line test in a nontraumatic setting, since the likelihood of there being acute blood as a confounder for enhancing pathology is low. To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4].
CTA Head and Neck
There is no evidence to support the use of CTA as the initial imaging test for ataxia with no history of trauma. To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4].

CTV Head
There is no evidence to support the use of CTV as the initial imaging test for ataxia with no history of trauma. To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4].

MRI Head
MRI of the entire brain without and with IV contrast is the preferred modality for initial assessment for a mass, any process that may result in edema and enhancement, and for neurodegenerative disorders. For the posterior fossa, in particular, MRI offers much better visualization compared to CT, and is especially preferred if the cerebellar finding is subtle (eg, mild parenchymal atrophy, architectural distortion, or mild parenchymal changes) that may not result in density differences significant enough for detection by CT. MRI with susceptibility weighted imaging or gradient echo T2-weighted sequences is useful for the detection of blood products in superficial siderosis, manifesting as a hypointense coating over the surface of the cerebellum, brainstem, and cord [87,88]. Diffusion weighted imaging can help to assess for an abscess [89] or a highly cellular tumor, and help identify and characterize certain infections, such as herpes encephalitis [90] and Creutzfeldt-Jakob disease [91]. Lhermitte-Duclos disease may demonstrate restricted diffusion [92]. MR spectroscopy has been used to assess abscesses [89], tumors, metabolic disorders, and neurodegenerative diseases and dementias. DTI, magnetization transfer imaging, MR perfusion imaging, and other techniques have also been investigated as assessment tools in the setting of multiple sclerosis and neurodegenerative disorders. To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4].

MRA Head and Neck
There is no evidence to support the use of MRA as the initial imaging test for ataxia with no history of trauma. To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4].

MRV Head
There is no evidence to support the use of MRV as the initial imaging test for ataxia with no history of trauma. To avoid delay of appropriate care, any patient with a suspected acute stroke should have imaging guided by the ACR Appropriateness Criteria® topic on “Cerebrovascular Disease” [4].

Arteriography Cervicocerebral
Digital subtraction angiography (DSA) is not a first-line test in the evaluation of ataxia.

Ioflupane SPECT or SPECT/CT Brain and DTPA Cisternography
Nuclear medicine scans are sometimes performed for assessment of some neurogenerative or movement disorders and dementias, such as Parkinson disease, Parkinson disease dementia, and dementia with Lewy bodies. See the ACR Appropriateness Criteria® topic on “Dementia and Movement Disorders” [93] for further information. These scans are not first-line tests in the evaluation of ataxia.

Variant 4: Ataxia of any acuity. No history of trauma. Suspected spinal or spinal vascular process. Initial imaging.
The body regions covered in this clinical scenario are the cervical, thoracic, and lumbar spine. These body regions might be evaluated separately or in combination as guided by physical examination findings, patient history, and other available information, including prior imaging.

Nontraumatic pathology affecting the spinal cord can lead to symptoms of ataxia because of a disturbance in proprioception and motor function. Processes include inflammatory and demyelinating diseases [94], such as multiple sclerosis [95-104], neuromyelitis optica [105-112], and acute disseminated encephalomyelitis [34]. Other inflammatory causes, such as neurosarcoidosis [113]; neoplasms such as lymphoma [114] and metastases as well as paraneoplastic syndromes [115]; nutritional deficiencies, such as B12 deficiency [116] and copper deficiency; infections affecting the cord, such as neurosyphilis; and degenerative changes in the vertebral bodies and discs [117,118]; or ossification of the posterior longitudinal ligament [119] that lead to cord compression and edema.
Vascular lesions, such as spinal dural arteriovenous fistulae, can also cause cord edema and resultant ataxia [120,121]. Acute nontraumatic spinal cord syndrome is a medical emergency that may have neurologically devastating sequelae, in which ataxia results from suspected spinal cord involvement. Common causes include transverse myelitis, metastatic disease, as well as epidural hematoma and cord infarction. Depending on other symptoms associated with ataxia, the site of pathology may be narrowed down to a particular segment or level of the cord, and imaging can be tailored to that site to offer a smaller field of view and more detailed visualization of that particular portion of the cord for detection of even subtle pathology and small lesions. Please also reference the ACR Appropriateness Criteria™ topic on “Myelopathy” [7] in the appropriate clinical context.

**Radiography Spine**

Radiographs are not adequate for assessing etiologies of ataxia.

**CT Spine**

In the absence of trauma, ataxia from a spinal etiology would involve the cord and nerve roots, structures that are suboptimally assessed by CT compared to MRI. Degenerative changes and ossification of the posterior longitudinal ligament can be identified on CT, but any resultant sequelae on the cord and nerve roots cannot be adequately evaluated. IV contrast is not essential in CT assessment of spinal structures, in which the utility is primarily for bony evaluation. Obtaining both precontrast and postcontrast images is not necessary.

**CTA Spine**

CTA can help diagnose spinal vascular malformations, with definition of the supplied vessel and fistula; however, it is not comprehensive and is not a first-line test, and does not evaluate the spinal cord. Findings gathered from a CTA can help tailor any follow-up conventional angiography to be targeted to particular levels, shortening the time required for conventional angiography [122].

**MRI Spine**

MRI is the modality of choice for assessment of the spinal cord and nerve roots. It allows evaluation of the cord for edema, abnormal signal and/or enhancement (when IV contrast is utilized), mass and mass lesion, and compression. MRI can also assess the exiting nerve roots for compression, abnormal enhancement (when IV contrast is utilized), thickening, or mass. MRI allows evaluation of the thecal sac for mass and mass effect, enhancement (when IV contrast is utilized), hemorrhage, and abnormal flow voids that might suggest a potential vascular lesion. MRI allows assessment of vertebral bodies and discs for degenerative changes, mass effect, or diseases involving the vertebral bodies that may affect the cord, as well as sarcoidosis, tuberculosis, lymphoma, and metastasis, paraspinous soft tissue and musculature. For cord pathologies, such as transverse myelitis, infections, demyelinating diseases, neoplasm, vascular malformations, spondylosis, and neurodegenerative disorders, MRI without and with IV contrast is the initial imaging modality of choice. If there are clinical signs localizing the lesion level, dedicated imaging of the cervical versus thoracic spine may be performed rather than whole spine imaging, which allows for higher spatial resolution via smaller field-of-view and thinner image slices.

**MRA Spine**

MRA allows evaluation of vascular supply to the cord and for vascular abnormalities involving the thecal sac and cord. This is obtained when there is high suspicion for a vascular cause of ataxia based on clinical presentation or findings from initial imaging investigation. MRA of the spine may be performed using blood flow–dependent techniques with or without the use of IV contrast; however, a fast contrast-enhanced MRA has been shown to have the advantage of being able to depict the feeding artery to a potential spinal dural arteriovenous fistula or spinal arteriovenous malformation [123-126]. Unless a concurrent MRI is performed, the cord and other soft-tissue structures cannot be adequately assessed on the MRA sequences alone. MRA is not a first-line test for the evaluation of ataxia.

**CT Myelogram Spine**

CT myelogram can help delineate the cord contour, configuration of the thecal sac, and outline exiting peripheral nerve roots. It may be useful in select circumstances when MRI is nondiagnostic or when physical findings do not correlate with MRI findings [9,127]. The caliber and position of the cord and nerve roots and mass effect upon these structures, for example from degenerative disease, can be appreciated. However, assessment of cord parenchyma is suboptimal when compared to MRI, and this is not a first-line test for the evaluation of ataxia.
Arteriography Spine

DSA is the gold standard for assessing the contour, caliber, anatomy, and drainage patterns of vessels supplying and draining the cord, and can help characterize a vascular abnormality or malformation that is suspected on initial cross-sectioning imaging studies; however, DSA is not a first-line test for the evaluation of ataxia.

Summary of Recommendations

- **Variant 1:** A CT head without IV contrast is usually appropriate for the initial imaging of acute ataxia following recent head trauma.

- **Variant 2:** A CT spine area of interest without IV contrast, CTA neck with IV contrast, or MRI spine area of interest without IV contrast are usually appropriate for the initial imaging of acute ataxia following recent spine trauma, depending on whether the clinical suspicion is for bony fracture, vascular pathology or dissection, or soft-tissue injury respectively. The panel did not agree on recommending MRI spine area of interest without and with IV contrast in this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from this procedure. The use of MRI spine area of interest without and with IV contrast in this patient population is controversial but may be appropriate.

- **Variant 3:** A MRI head without and with IV contrast or MRI head without IV contrast is usually appropriate for the initial imaging of ataxia in patients with no history of trauma, suspected intracranial process, and when stroke intervention is not considered, noting that the use of IV contrast is preferred.

- **Variant 4:** A MRI spine area of interest without and with IV contrast or MRI spine area of interest without IV contrast is usually appropriate for the initial imaging of ataxia of any acuity and no history of trauma and suspected spinal or spinal vascular process, noting that the use of IV contrast is preferred.

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at https://acsearch.acr.org/list. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents go to www.acr.org/ac.

Appropriateness Category Names and Definitions

<table>
<thead>
<tr>
<th>Appropriateness Category Name</th>
<th>Appropriateness Rating</th>
<th>Appropriateness Category Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usually Appropriate</td>
<td>7, 8, or 9</td>
<td>The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.</td>
</tr>
<tr>
<td>May Be Appropriate</td>
<td>4, 5, or 6</td>
<td>The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.</td>
</tr>
<tr>
<td>May Be Appropriate (Disagreement)</td>
<td>5</td>
<td>The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel’s recommendation. “May be appropriate” is the rating category and a rating of 5 is assigned.</td>
</tr>
<tr>
<td>Usually Not Appropriate</td>
<td>1, 2, or 3</td>
<td>The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.</td>
</tr>
</tbody>
</table>
Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® Radiation Dose Assessment Introduction document [128].

<table>
<thead>
<tr>
<th>Relative Radiation Level*</th>
<th>Adult Effective Dose Estimate Range</th>
<th>Pediatric Effective Dose Estimate Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>☐</td>
<td>0 mSv</td>
<td>0 mSv</td>
</tr>
<tr>
<td>☢</td>
<td>&lt;0.1 mSv</td>
<td>&lt;0.03 mSv</td>
</tr>
<tr>
<td>☢☢</td>
<td>0.1-1 mSv</td>
<td>0.03-0.3 mSv</td>
</tr>
<tr>
<td>☢☢☢</td>
<td>1-10 mSv</td>
<td>0.3-3 mSv</td>
</tr>
<tr>
<td>☢☢☢☢</td>
<td>10-30 mSv</td>
<td>3-10 mSv</td>
</tr>
<tr>
<td>☢☢☢☢☢</td>
<td>30-100 mSv</td>
<td>10-30 mSv</td>
</tr>
</tbody>
</table>

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as “Varies”.

References


