

**American College of Radiology  
ACR Appropriateness Criteria®**

**Clinical Condition:** Ataxia

**Variant 1:** Slowly progressive ataxia, or ataxia of long duration (adult or child).

Radiologic Procedure	Rating	Comments	<a href="#">RRL*</a>
MRI head without and with contrast	8	See statement regarding contrast in text under "Anticipated Exceptions."	O
MRI head without contrast	7		O
MRI cervical thoracic and lumbar spine without and with contrast	7	Ataxia can be of spinal origin. Consider if brain imaging is negative or inconclusive. See statement regarding contrast in text under "Anticipated Exceptions."	O
MRI cervical thoracic and lumbar spine without contrast	6	Ataxia can be of spinal origin. Consider if brain imaging is negative or inconclusive.	O
CT head without and with contrast	5	The RRL for the adult procedure is ☼☼☼☼.	☼☼☼☼
CT head with contrast	5		☼☼☼
CT head without contrast	4		☼☼☼
FDG-PET/CT head	3		☼☼☼☼
MR spectroscopy head without contrast	2	Selectively used as an adjunct to conventional MRI for characterizing indeterminate lesion(s).	O
I-123 Ioflupane SPECT head (DaT scan)	2		☼☼☼
US transcranial with Doppler	1		O
<b>Rating Scale:</b> 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			<b>*Relative Radiation Level</b>

**Clinical Condition:** Ataxia

**Variant 2:** Acute ataxia (<3 hours) as a manifestation of suspected stroke (adult or child). (See the ACR Appropriateness Criteria® topic on “[Cerebrovascular Disease](#)”).

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with contrast	8	See statement regarding contrast in text under “Anticipated Exceptions.”	O
MRA head and neck without and with contrast	8	See statement regarding contrast in text under “Anticipated Exceptions.”	O
CTA head and neck with contrast	8	Combined vascular and cerebral evaluation should be considered. The RRL for the adult procedure is ☼☼☼.	☼☼☼☼
CT head without and with contrast	8	CT perfusion is less accurate in the posterior fossa. MRI and perfusion characterization is preferred if treatment will not be unreasonably delayed. Combined vascular and cerebral evaluation should be considered. The RRL for the adult procedure is ☼☼☼.	☼☼☼☼
CT head without contrast	8		☼☼☼
MRI head without contrast	7		O
CT head with contrast	7		☼☼☼
MRI cervical spine without and with contrast	5	Fat-saturated T1 axial images. See statement regarding contrast in text under “Anticipated Exceptions.”	O
MRI cervical spine without contrast	4		O
MRA head and neck without contrast	2		O
MR spectroscopy head without contrast	2		O
US transcranial with Doppler	2		O
<b>Rating Scale:</b> 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			<b>*Relative Radiation Level</b>

**Clinical Condition:** Ataxia

**Variant 3:** Acute or subacute ataxia as a manifestation of suspected infection (adult or child).

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with contrast	8	See statement regarding contrast in text under "Anticipated Exceptions."	O
MRI head without contrast	7		O
MRI cervical spine without and with contrast	6	Ataxia can be of spinal origin. Consider if brain imaging is negative or inconclusive. See statement regarding contrast in text under "Anticipated Exceptions."	O
MR spectroscopy head without contrast	6	May help distinguish abscess from other masses.	O
CT head with contrast	6		☼☼☼
CT temporal bone with contrast	6		☼☼☼
MRI cervical spine without contrast	5	Ataxia can be of spinal origin. Consider if brain imaging is negative or inconclusive.	O
MRA head without and with contrast	5	See statement regarding contrast in text under "Anticipated Exceptions."	O
CT head without and with contrast	5	The RRL for the adult procedure is ☼☼☼.	☼☼☼☼
CT temporal bone without contrast	5	Useful when skull base or middle ear disease is suspected.	☼☼☼
CTA head with contrast	4		☼☼☼
MRA head without contrast	4		O
CT head without contrast	4		☼☼☼
CT temporal bone without and with contrast	2		☼☼☼
US transcranial with Doppler	1		O
<b>Rating Scale:</b> 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			<b>*Relative Radiation Level</b>

**Clinical Condition:** Ataxia

**Variant 4:** Acute ataxia following head trauma, less than 24 hours (adult or child).

Radiologic Procedure	Rating	Comments	RRL*
CT head without contrast	9		☼ ☼ ☼
MRI head without contrast	8		○
CT temporal bone without contrast	7	Useful when skull base or middle ear disease is suspected.	☼ ☼ ☼
MRI head without and with contrast	7	See statement regarding contrast in text under "Anticipated Exceptions."	○
MRI neck without and with contrast	6	See statement regarding contrast in text under "Anticipated Exceptions."	○
CT head without and with contrast	6	The RRL for the adult procedure is ☼ ☼ ☼ .	☼ ☼ ☼ ☼
CTA head and neck with contrast	6	The RRL for the adult procedure is ☼ ☼ ☼ .	☼ ☼ ☼ ☼
MRA head and neck without and with contrast	6	See statement regarding contrast in text under "Anticipated Exceptions."	○
MRI neck without contrast	5		○
CT head with contrast	5		☼ ☼ ☼
CT temporal bone without and with contrast	2		☼ ☼ ☼
CT temporal bone with contrast	2		☼ ☼ ☼
MRA head and neck without contrast	2		○
US transcranial with Doppler	1		○
<b>Rating Scale:</b> 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			<b>*Relative Radiation Level</b>

# ATAXIA

Expert Panel on Neurologic Imaging: Daniel F. Broderick, MD<sup>1</sup>; Franz J. Wippold II, MD<sup>2</sup>; Rebecca S. Cornelius, MD<sup>3</sup>; Ashley H. Aiken, MD<sup>4</sup>; Edgardo J. Angtuaco, MD<sup>5</sup>; Douglas C. Brown, MD<sup>6</sup>; Kathryn Holloway, MD<sup>7</sup>; Charles T. McConnell Jr, MD<sup>8</sup>; Laszlo L. Mechtler, MD<sup>9</sup>; Joshua M. Rosenow, MD<sup>10</sup>; Christopher J. Roth, MD<sup>11</sup>; Konstantin Slavin, MD.<sup>12</sup>

## **Summary of Literature Review**

### **Introduction/Background**

Ataxia is a term that describes abnormality in the coordination of movement. Manifestations commonly include a wide-based unsteady gait and poor coordination of the extremities. Frequently associated findings include disorders of ocular motility, poor coordination of speech, dysmetria, dysdiadochokinesis, dysrhythmokinesis, hypotonia, and pendular reflexes [1]. Intention tremor may be present when initiating or performing an activity, especially when the ataxia is of cerebellar origin. Ocular nystagmus, skew deviation, disconjugate saccades, and altered ocular pursuit movements may also be seen as components of ataxia. A wide-based stance, with feet several inches apart, is the most commonly seen but nonspecific anatomically localizing sign of cerebellar disease. Truncal instability and rhythmic tremor of the body or head (titubation) occur especially, but not exclusively, in association with disorders that involve the cerebellar midline. Detailed physical findings associated with ataxia and the utility of these findings relative to anatomic localization are beyond the scope of this ACR Appropriateness Criteria<sup>®</sup> topic and have been reviewed elsewhere [1].

### **Overview of Imaging Modalities**

Ataxia may separately arise from disorders that involve the cerebellum, brainstem, vestibular nuclei, thalamic nuclei, cerebral cortex (especially the frontal lobes), and white matter tracts of the cerebral hemispheres, spinal cord, and peripheral sensory nerves. Because conditions involving many anatomic regions may produce ataxia, an appropriate imaging evaluation is often complex. Optimal detail of posterior fossa structures may be obscured by beam-hardening artifact in computed tomography imaging (CT), and therefore magnetic resonance imaging (MRI) is usually the preferred initial modality for evaluating patients of all ages [2]. Correlation with the patient's clinical history and physical findings is essential for appropriate study design and image interpretation. Some patients with distinct clinical ataxia may have initially normal imaging examinations, and repeat imaging may be necessary. This situation most commonly occurs early in the course of toxic, metabolic, degenerative, or other progressive disorders associated with ataxia.

### **Discussion of Imaging Modalities by Variant**

Medical disorders that cause ataxia are numerous, often individually quite uncommon, and have been the subject of several classification schemes [3-5]. The purpose of these guidelines, and of the designation of imaging variants, is to categorize the diverse disorders that may present with ataxia and to suggest imaging approaches that may be useful for patients with the most common clinical presentations and underlying disorders. For individual patients, imaging study design will be significantly compromised if the history that accompanies the imaging study is not sufficiently detailed regarding clinical and family history, physical findings, and the results of relevant laboratory studies.

Disorders associated with vertigo, the subjective illusion of movement, can be associated with clinical incoordination. See the ACR Appropriateness Criteria<sup>®</sup> on "[Hearing Loss and/or Vertigo](#)" [6].

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<sup>1</sup>Principal Author, Mayo Clinic Jacksonville, Jacksonville, Florida. <sup>2</sup>Panel Chair, Mallinckrodt Institute of Radiology, Saint Louis, Missouri. <sup>3</sup>Panel Vice-chair, University of Cincinnati, Cincinnati, Ohio. <sup>4</sup>Emory Healthcare, Atlanta, Georgia. <sup>5</sup>University of Arkansas for Medical Sciences, Little Rock, Arkansas. <sup>6</sup>Hampton Roads Radiology Associates, Norfolk, Virginia. <sup>7</sup>MCVH-Virginia Commonwealth University, Richmond, Virginia, American Association of Neurological Surgeons/Congress of Neurological Surgeons. <sup>8</sup>Good Samaritan Hospital, Cincinnati, Ohio. <sup>9</sup>Dent Neurologic Institute, Amherst, New York, American Academy of Neurology. <sup>10</sup>Northwestern Memorial Hospital, Chicago, Illinois, American Association of Neurological Surgeons/Congress of Neurological Surgeons. <sup>11</sup>Duke University Medical Center, Durham, North Carolina. <sup>12</sup>UIC Medical Center, Chicago, Illinois, American Association of Neurological Surgeons/Congress of Neurological Surgeons.

The American College of Radiology seeks and encourages collaboration with other organizations on the development of the ACR Appropriateness Criteria through society representation on expert panels. Participation by representatives from collaborating societies on the expert panel does not necessarily imply individual or society endorsement of the final document.

Reprint requests to: Department of Quality & Safety, American College of Radiology, 1891 Preston White Drive, Reston, VA 20191-4397.

## **Variant 1: Gradually Progressive Ataxia or Ataxia of Long Duration (Adult or Child)**

### *Ataxia Associated with Intracranial Mass Lesion or Remote Mass Lesions*

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. In pediatric patients the most common intra-axial posterior fossa lesions are medulloblastomas, cystic astrocytomas, ependymomas, and brain stem gliomas. In adults, intra-axial hemangioblastomas, choroid plexus papillomas, extra-axial meningiomas, and intra-axial, extra-axial, or diffuse leptomeningeal metastatic processes become more prevalent. Isolated frontal lobe and thalamic mass lesions may also present with varying manifestations of gait and limb ataxia. Unless contraindicated, MRI of the entire brain, without and with contrast, is almost always superior to CT for the initial exclusion or characterization of a posterior fossa or other intracranial mass lesions.

Lhermitte-Duclos disease (dysplastic gangliocytoma) is a slowly growing benign cerebellar hamartoma or congenital malformation [7]. Symptoms correlate with local mass effect. MRI demonstrates a cerebellar hemisphere mass that involves the cortex and folia and is generally of increased T2 signal intensity. There are also characteristic internal curvilinear bands that are isointense with the cerebellar cortex [7-9]. The mass does not enhance with contrast, and it may demonstrate restricted diffusion [7-9]. Cowden syndrome, an autosomal dominant disorder characterized by cutaneous and noncutaneous hamartomas and by increased risk for breast, thyroid, gastrointestinal, and genitourinary neoplasias, is frequently associated [7,8].

Paraneoplastic cerebellar degeneration is clinically characterized by the subacute or acute onset of gait and limb ataxia, dysarthria, and ocular dysmetria [10]. Paraneoplastic syndromes may be caused by any primary tumor but are most commonly associated with breast, gynecologic, and lung tumors and with Hodgkin's disease [10]. Typically, tumor seeding of brain parenchyma is not demonstrated with tissue sampling or with imaging. Several antineuronal antibodies have been identified in serum and in tissue, depending on the originating tumor cell type. MRI is generally normal until late stages of the disorder when mild to moderate cerebellar cortical atrophy becomes evident [10]. Uncommonly, hyperintensity of the cerebellum or other involved brain parenchymal areas may be identified on T2-weighted MR sequences [11]. CT imaging and/or positron emission tomography (PET) imaging may be indicated when an underlying primary is not clinically evident [12].

### *Demyelinating Disorders*

Multiple sclerosis patients may present with or subsequently develop ataxia. In these patients, MRI (without and with contrast), diffusion imaging, spectroscopy, perfusion imaging, and magnetization transfer imaging can each support but not establish the diagnosis. The presence of multiple oval-shaped periventricular regions of hyperintensity on T2-weighted MRIs, generalized cerebral volume loss, callosal and optic nerve involvement, and ring enhancement of active lesions is typical. Detailed descriptions of specific MRI findings and the utility of advanced MRI techniques for patient evaluation and management are reviewed elsewhere [13,14].

### *Congenital Disorders*

Many ataxia-associated disorders are congenitally based [3]. For all of them MRI is preferred to CT. These disorders will most commonly manifest ataxia during early childhood. Some of them are sporadic, while others have a known or an apparent genetic basis [3]. Clinical abnormalities that occur in association with congenital ataxia may include mild to severe mental retardation, hearing loss, optic atrophy, cataracts, growth retardation, seizures, cleft palate, and either spasticity or diminished muscle tone. In these well-characterized congenital ataxia-associated disorders, imaging findings generally include nonspecific hypoplasia of the cerebellar vermis, hypoplasia of the entire cerebellum, or congenital cerebellar developmental dysplasia. Additionally associated imaging alterations can include brain stem hypoplasia, lissencephaly, aprosencephaly, microcephaly, or variable and less prominent cerebral developmental alteration.

Dandy-Walker syndrome is characterized by hypoplasia of the cerebellar vermis and/or demonstrates a cystic cerebrospinal fluid collection that is predominately posterior to the cerebellum but continuous with the fourth ventricle. Dandy-Walker patients present with ataxia, nystagmus, cranial nerve palsies, apneic episodes, hydrocephalus, and cognitive dysfunction [15,16]. The torcula is usually elevated and the posterior fossa usually enlarged. Hydrocephalus is frequently associated, and there may be anomalies of cerebral development that involve the cerebral cortex and corpus callosum [15]. Differentiation from other congenital or acquired posterior fossa cysts is essential.

Joubert syndrome, with congenital ataxia, hypotonia, and oculomotor ataxia, has unique imaging alterations that involve the midbrain and cerebellum ("molar tooth" contour of the brainstem or "bat wing" configuration of the fourth ventricle) [17]. Four types have been identified, each with somewhat variable clinical and imaging features and with genetic alterations that involve different loci [18].

Rhombencephalosynapsis is a rare cerebellar dysplastic process that can occur alone or in association with other developmental anomalies [16]. Rhombencephalosynapsis demonstrates vermian agenesis with fusion of the cerebellar hemispheres, apposition or fusion of the dentate nuclei, and fusion of the superior cerebellar peduncles. The lateral ventricles are usually enlarged, and the thalamic nuclei may be fused [16].

Congenital ataxia may also occur in association with perinatal cerebral infarction and in association with congenital cytomegalovirus or other infectious processes of the central nervous system [15,19].

Ataxic cerebral palsy is uncommon relative to other forms of cerebral palsy, and its imaging correlates have not been well defined [20].

#### *Hereditary and Idiopathic Degenerative Processes*

The hereditary ataxias are classified on the basis of their causative gene (when known) and their pattern of inheritance (autosomal dominant, autosomal recessive, x-linked, or mitochondrial). In each of these disorders MRI is the preferred imaging modality. Within this group of patients, a broad range of potential diagnostic considerations is often suggested by family history, by findings on physical examination, and by MRI evidence of atrophy involving the cerebellum and varying combinations of the pons, medulla, spinal cord, cerebral cortex, and optic nerves. Dentate calcification may be identified on CT imaging. Definitive diagnosis, however, relies on molecular genetic testing. While cerebellar ataxia is the dominant and occasionally the only clinical finding, spasticity, neuropathy, seizures, extrapyramidal symptomatology, mental retardation, cognitive decline, nystagmus, visual loss, spasmodic cough, and migraine-like episodes may also be associated. MRI is almost invariably the preferred imaging tool.

Among the identified autosomal dominant spinocerebellar ataxias (AD-SCAs), specific diagnostic nomenclature is replacing less specific terms such as spinocerebellar degeneration, Marie's ataxia, and olivopontocerebellar atrophy (OPCA). Among the AD-SCA disorders, over 30 separate and distinct genetic abnormalities have now been identified. The term OPCA continues to be used as a label only for those cases that have a clinical and pathology-related combination of "cerebellar-plus" symptomatology, have imaging correlates of cerebellar and brainstem atrophy, and have an unidentifiable genetic origin [21]. The designation "idiopathic late-onset cerebellar ataxia" is separately used to describe a different and significantly large group of adult patients with predominant cerebellar symptomatology, absence of a family history of ataxia, and absence of an identified genetic marker [22]. In these patients MRI generally demonstrates cerebellar and pontine volume loss [23].

Spinocerebellar ataxia type 2 (SCA2) is a form of AD-SCA that causes slowly progressive ataxia, dysarthria, nystagmus, and initially brisk but later absent tendon reflexes, with associated peripheral neuropathy. Dystonia, parkinsonism, and dementia may also be present. Symptoms are more rapidly progressive when they have an onset before age 20 years. Imaging findings include cerebellar and pontine volume loss and deep white matter alterations that may also involve the cerebral hemispheres. Transcranial brain parenchyma ultrasound demonstrates increased echogenicity of the substantia nigra in the majority of patients with SCA2 and without parkinsonism [24]. Increased echogenicity of the substantia nigra can also be seen in SCA3 and SCA 17; dentate nucleus hyperechogenicity and fourth ventricular enlargement are characteristic features of SCA3 on transcranial sonography [25]. Although SCA2 is a relatively common form of AD-SCA (13% of AD-SCA cases in one study), clinical and imaging features do not allow a definitive diagnosis. Molecular genetic analysis is necessary. Many but not all cases of Marie's ataxia and AD-OPCA are thought to represent SCA2.

Spinocerebellar ataxia type 3 (SCA3) (Machado-Joseph disease) accounts for 23% of patients with AD-SCA. Symptoms most commonly include an onset in the second to fourth decade of cerebellar ataxia, spasticity, peripheral neuropathy, bulbar dysfunction with facial and tongue atrophy, and occasional myoclonus or intellectual impairment. Subtypes have been described. MRI alterations include volume loss involving the cerebellum, the pons and medulla, and a linear region of high T2 signal intensity along the posterior and medial margins of the globus pallidus.

Quantitative MRI, including voxel-based morphometry, may be helpful in evaluating and characterizing SCA types 1, 3, and 6 [26,27], SCA2 [28], and 17 [29]. Qualitative and quantitative diffusion tensor imaging metrics of white matter integrity may also be useful in patients with sporadic and hereditary ataxia [29].

Dentatorubral-pallidolusian atrophy is characterized by progressive ataxia, choreoathetosis, and dementia or character changes when the disorder occurs in adults. In children it is characterized by ataxia, myoclonus, epilepsy, and progressive intellectual deterioration. [30]. MRI demonstrates cerebellar and brainstem volume loss with cerebral cortical atrophy [31]. Less frequently signal intensity in deep white matter of the cerebral

hemispheres, in the thalamus, and in the brainstem may be increased on T2-weighted MRIs [31]. Diagnosis is established through the identification of genetic markers [30].

Autosomal recessive spinocerebellar ataxias (AR-SCAs) may be associated with multiple underlying genetic disorders. The most common is Friedreich's ataxia with a population frequency of 1-2/50,000. It commonly has its onset within the first or second decade. Symptoms are progressive and are characterized by ataxia, diminished muscle stretch reflexes, upgoing toes, sensory loss for vibration and position, pes cavus, and cardiomyopathy [32,33]. Imaging findings include diminished cross-sectional area of the spinal cord and medulla, and inconsistently the presence of cerebellar volume loss [32,33]. Diagnosis is by molecular genetic testing [33].

Ataxia-telangiectasia (A-T) is an AR-SCA with findings of progressive cerebellar ataxia, telangiectasias of the conjunctivae, oculomotor apraxia, choreoathetosis, and frequent infections. Symptoms begin between 1 and 4 years of age, and the population-based prevalence is 1/40,000-100,000. Patients are at risk of contracting leukemia and lymphoma, partly related to an increased sensitivity to ionizing radiation. MRI demonstrates initial selective atrophy of the lateral portions of the cerebellar hemispheres, with subsequent extension of volume loss to involve inferior and superior cerebellar cortical regions [34]. The vermis also becomes atrophic, more in its superior than in its inferior portions [34]. Laboratory testing supports the diagnosis of A-T by identifying elevated serum alpha-fetoprotein, the absence of ataxia-telangiectasia mutated protein in blood mononuclear cells [35], the presence of a 7:14 chromosome translocation in peripheral lymphocytes, and the presence of immunodeficiency. Several less common AR-SCAs will not be discussed here [5].

The fragile X tremor/ataxia syndrome (FXTAS) is an X-linked cause of progressive ataxia. Fragile X syndrome, a separate but related disorder, is the most common genetic cause of mental retardation [36]. Fragile X syndrome results from silencing of the fragile-X mental retardation 1 gene [36]. Predominant clinical findings include a characteristic facies, developmental delay, and mental retardation [36]. FXTAS, however, involves adults, predominately males, who are *carriers* of the fragile-X syndrome [37]. All have "premutation" alleles and demonstrate significant and progressive clinical findings of tremor, ataxia, autonomic instability, parkinsonism, and cognitive decline [38] [37,39]. Characteristic MRI findings of FXTAS include brain stem atrophy and cerebral cortical atrophy. There may be increased intensity in the white matter of atrophic middle cerebellar peduncles and in deep and in subependymal cerebral white matter on T2-weighted MRIs [40].

Multiple system atrophy (MSA) is a sporadic neurodegenerative disorder that may initially manifest by ataxia (MSAc) or by parkinsonism (MSAp) [41,42]. Onset is generally after age 50, and early additional findings can include prominent autonomic dysfunction and spasticity. Previously reported cases of sporadic OPCA and Shy-Drager syndrome most likely represent what is now known as MSA. MSAc is characterized by gait and limb ataxia, by dysarthria, and by oculomotor abnormalities that initially are similar to the findings observed in late-onset cerebellar ataxia. Dysautonomia and parkinsonism will eventually develop [41]. MRI demonstrates atrophy of the pons, cerebellum, and putamen. The pons and middle cerebellar peduncles may demonstrate hyperintensity on T2-weighted images. The putamen are generally low in signal intensity on T2-weighted MRIs, although a narrow band of increased signal intensity at the lateral aspect of the putamen may be seen [41-43].

Functional imaging of patients with idiopathic Parkinson's disease (PD) and atypical neurodegenerative parkinsonism disorders (such as MSA) includes I-123 Ioflupane SPECT head (DaT scan). Dopamine transporter (DaT) imaging may be useful in detecting dopaminergic dysfunction in suspected or premotor cases of striatal dopamine-deficient parkinsonism and in monitoring PD progression. Normal DaT imaging may be helpful in excluding vascular parkinsonism and other PD mimics [44-47].

Progressive ataxia is occasionally associated with mitochondrial disorders such as MERRF (myoclonic epilepsy with ragged red fibers), NARP (neuropathy, ataxia, and retinitis pigmentosa), Leigh syndrome, and Kearns-Sayre syndrome [48]. Additional clinical manifestations such as seizures, deafness, diabetes mellitus, cardiomyopathy, retinopathy, and short stature are often associated. In NARP, cerebral and cerebellar atrophy may be noted on MRI. In Leigh syndrome, bilateral symmetric low attenuation may be present in the basal ganglia on CT, and increased signal intensity may be seen in the brainstem and/or basal ganglia on T2-weighted MRIs [49].

A deficiency of coenzyme Q10 has been described in individuals with cerebellar ataxia, usually with childhood onset and often associated with seizures [50]. MRI may demonstrate cerebellar volume loss [50]. Symptoms may respond to treatment with coenzyme Q10.

Vanishing white matter disease is an inherited early childhood leukoencephalopathy whose most prominent initial symptom is ataxia [51]. Onset most commonly occurs at 2 to 6 years of age, though adolescent and adult onset has been described. MRI demonstrates diffuse increased white matter T2 signal intensity and both a progressive and



extensive loss in volume of cerebral and cerebellar white matter. Subcortical white matter may initially be spared [51].

#### *Superficial Siderosis*

In this nonhereditary disorder hemosiderin accumulates in subpial layers of brain and spinal cord as the result of recurrent, often silent, subarachnoid hemorrhage. The most prominent symptoms are slowly progressive ataxia and hearing loss [52]. MRI is the definitive diagnostic procedure [52], demonstrating a hypointense coating over the cortex, brain stem, and/or spinal cord on T2-weighted MRIs. The cerebellum may be atrophied. The superior vermis and anterior cerebellar hemispheres may be preferentially affected. Both gradient-echo T2-weighted MRIs and images generated with magnets using high field strength improve sensitivity for hemosiderin deposition [53].

#### *Spinal Cord and Peripheral Nerve-Related Ataxia*

Evaluation of ataxia that is potentially due to pathologic processes originating within the spinal cord or within the roots/nerves originating from the spinal cord requires high-resolution T1 and T2-weighted axial and sagittal MRIs without and with intravenous contrast. Imaging should focus on the posterior columns and on the nerve roots. In pernicious anemia, imaging findings depend on the duration and severity of the disorder. The spinal cord may appear swollen and may demonstrate hyperintensity, especially in the posterior columns on T2-weighted MRIs [54,55]. Eventually, atrophy and persistent gliosis may develop, or alternatively all imaging findings may resolve with treatment [54]. In patients with hypertrophic, inflammatory, or postinfectious polyneuropathies, nerve root enhancement and enlargement may be demonstrated with MRI [56].

#### *Nutritional Deficiency, Toxins, and Drugs*

In each of these disorders MRI is the preferred imaging modality. Solvent abuse or toxic exposure to solvents can result in gait impairment and encephalopathy. MRI abnormalities are characterized by diffuse cortical atrophic changes and by hyperintensity on T2-weighted images in the white matter, basal ganglia, and thalami [57].

Methyl-mercury poisoning (Minamata disease) is a neurological illness caused by the ingestion of contaminated seafood. It is characterized by ataxia, visual loss, and sensory disturbance. MRI in affected patients demonstrates atrophy of the cerebellar vermis and hemispheres, as well as the calcarine cortex [58,59].

Metronidazole (Flagyl)-induced cerebellar toxicity is associated with symptoms of ataxia. MRI findings on T2-weighted images include bilateral symmetric hyperintensity, most commonly in the dentate nuclei; less common sites of abnormal signal intensity include the dorsal medulla and pons, the midbrain and the splenium of the corpus callosum. Restricted diffusion may be present in these locations as well [60]. With symptom resolution, MRI becomes normal [60,61].

In osmotic demyelination syndrome, formerly known as central and/or extrapontine myelinolysis, cerebellar or extrapyramidal symptoms have been observed. More typically, patients demonstrate coma, locked-in syndrome, or quadriparesis. This disorder is typically seen in the setting of hyponatremia and its rapid correction, often in chronic alcoholic and malnourished patients. Increased T2 signal intensity in the central pons with sparing of the tegmentum, ventrolateral pons, and corticospinal tracts is the characteristic finding [62,63].

A leukoencephalopathy with initial symptoms of ataxia has also been reported to occur following the chronic inhalation of heroin vapors [57]. A characteristic and highly specific pattern of bilateral symmetric increased T2 signal intensity in the cerebellar white matter with sparing of the dentate nuclei has been reported in heroin-induced leukoencephalopathy [64].

Vitamin E deficiency may occur in association with several acquired gastrointestinal disorders or with autosomal recessive defects in vitamin E transport [65,66]. Symptoms include ataxia with associated weakness, areflexia, and retinal degeneration. Imaging has demonstrated cerebellar atrophy and hyperintensity involving the posterior columns of the spinal cord on T2-weighted MRIs [65,66].

Chronic ethanol abuse is associated with ataxia and multiple other symptoms of neurologic dysfunction. These symptoms result from the neurotoxicity of ethanol and its metabolic products, from associated chronic liver disease, from secondary nutritional deficiencies, and from the effect of other toxins that are simultaneously ingested [62]. MRI demonstrates atrophy of the cerebellar vermis, especially superiorly, as well as volume loss involving pons, medulla, and cerebral hemispheres [62].

Wernicke encephalopathy is due to thiamine deficiency. It classically presents with ataxia, altered mental status, and abnormality of ocular motility. MRI demonstrates hyperintensity on T2-weighted sequences, reversible enhancement following intravenous gadolinium injection, and reversible restricted diffusion in multiple structures, including mammillary bodies, hypothalamus adjacent to the third ventricle, periaqueductal gray and

white matter, pulvinar, and dorsomedial portions of the thalamic nuclei [62,63]. Corresponding small hemorrhagic foci may also occur in these regions.

Reversible posterior leukoencephalopathy is most commonly characterized by headache, altered consciousness, visual disturbance, and seizures [67]. Ataxia can be a component, especially when there is brainstem or cerebellar involvement [67,68]. Antecedent clinical conditions include hypertension, eclampsia, renal disease, and the use of cytotoxic or immunosuppressant drugs [67]. MRI findings include the presence of bilateral and generally symmetric patchy and confluent hyperintensity in the posterior parietal and occipital lobes on T2-weighted images. Signal abnormalities may also involve the basal ganglia, cerebellum and brainstem, and the frontal lobe. White matter is generally more involved than is cortex, although the deep gray matter may also be affected. Little or no enhancement following contrast administration typically occurs [67,69]. Intracranial hemorrhage adjacent to the foci of abnormal signal intensity has been reported. Predominant brainstem involvement has also been described, with restriction of water diffusion demonstrated on diffusion-weighted MRIs; water diffusion may subsequently increase with resolution of the clinical syndrome [68,70,71].

### **Variant 2: Acute Ataxia as a Possible Manifestation of Stroke (Adult or Child)**

Ischemic or hemorrhagic events may cause ataxia when isolated to vascular distributions supplying portions of the cerebellum, medulla, pons, mesencephalon, red nucleus, thalamic nuclei, or posterior limb of the internal capsule, or to the frontal or parietal cerebral cortex [72-79]. Medially positioned infarctions involving the pons and medulla are uncommonly associated with ataxia [72,73]. Several named syndromes are associated with ataxia and focal regions of brainstem infarction [78,80]. Infarctions in the distribution of the posterior inferior cerebellar artery (lateral medullary syndrome or Wallenberg syndrome) are the most common pattern of brainstem infarction that is associated with a specific syndrome of ataxia. Symptoms include ipsilateral hemiataxia, vertigo, dysarthria, ptosis, and miosis. Although brainstem and cerebellar infarction are predominately arterial in origin, venous infarction should also be considered [81]. The imaging evaluation of ataxia generally requires MRI, with water diffusion characterization and with time-of-flight MR angiography. Evaluation of the neck vessels with MRI, using T1-weighted images, without and with fat saturation, aids in excluding arterial dissection [82-84]. MR venography should be considered if central or dural venous thrombosis is suspected. Catheter-based diagnostic angiography and/or CT angiography may be necessary for confirmation [84].

Vascular malformations, angiopathy, or aneurysm rupture may cause acute ataxia when posterior fossa or supratentorial brain parenchyma is involved. CT imaging of the brain and CT angiography may replace or supplement MRI when evaluating for acute or subacute hemorrhage.

Recurrent or paroxysmal ataxia has been associated with several disorders, including epilepsy and migraine. Transient limb and trunk ataxia may occur with high systemic fever in otherwise healthy children. These disorders may be idiopathic or may be due to abnormalities in membrane calcium or potassium channel function or altered synaptic glutamate transport [85-87]. MRI is the imaging modality of choice. MRI may be normal, may demonstrate cerebellar volume loss, or may demonstrate extensive areas of cortical hyperintensity on T2-weighted images. These imaging findings may correlate with the possible simultaneous occurrence of hemiplegic migraine or recent seizure activity.

### **Variant 3: Acute or Subacute Ataxia as a Manifestation of Suspected Infection (Adult or Child)**

Several infectious and postinfectious processes may cause ataxia. Detection of infectious-process-related alterations in the cerebellum is best with unenhanced and enhanced MRI rather than CT because of the superior contrast resolution of MRI in the posterior fossa.

Bacterial cerebellitis may occur in association with meningitis or with cerebritis involving the cerebral hemispheres. Etiologies include penetrating trauma or transdural extension of an epidural infection, most commonly arising from the temporal bone. Diffusion-weighted images and MR spectroscopy may narrow the differential diagnosis [88,89]. Multiple viral processes, including herpes and arbovirus, may also involve brainstem and cerebellum [90].

Variant Creutzfeldt-Jakob disease (vCJD) (also known as bovine spongiform encephalopathy [BSE]) familial Creutzfeldt-Jakob disease (fCJD), and sporadic Creutzfeldt-Jakob disease (sCJD) are the most common prion-associated spongiform encephalopathies. vCJD and sCJD and each may present with behavioral, emotional, and intellectual deterioration. Eventually, ataxia and dysarthria may develop. Rapid progression to stupor, coma, and myoclonus occurs in sCJD, whereas the clinical course may be more prolonged in vCJD. MRI demonstrates hyperintensity on T2-weighted images, inversion sequences, and diffusion-weighted sequences within the heads of the caudate nuclei, the putamen, and regions of frontal, parietal, and occipital cortex. These alterations in signal

intensity may initially be asymmetric [91]. Eventually, diffuse brain atrophy develops. Although all forms of CJD demonstrate hyperintensity within the thalamic nuclei and pulvinar bilaterally on T2-weighted and diffusion-weighted images, these focal alterations are especially prominent in vCJD [91].

Acute cerebellitis, also called acute cerebellar ataxia, is a parainfectious disorder that predominately, but not exclusively, occurs in childhood. Symptoms include headache, ataxia, and photophobia. Intracranial pressure may be increased and the brainstem may be involved. MRI demonstrates hyperintensity in the cerebellar hemispheres with associated mass effect on T2-weighted images [92]. Bilateral involvement occurs more commonly than unilateral involvement. Obstruction of CSF flow may cause ventriculomegaly and upward herniation of posterior fossa structures [92]. The cerebellar meninges may enhance following intravenous contrast administration. Surgical decompression of the posterior fossa may be necessary [93]. Follow-up imaging may demonstrate cerebellar atrophy [94].

Bickerstaff encephalitis is a brainstem and cerebellar inflammatory disorder that most commonly follows a viral illness. It is characterized by ataxia and ophthalmoplegia, with MRI demonstrating mass effect, hyperintensity on T2-weighted MRIs, and restricted diffusion within portions of the pons, medulla, and cerebellum [95,96].

Fisher syndrome, a variant of the Guillain-Barré syndrome, involves the peripheral and central nervous system. It is clinically characterized by ophthalmoplegia and cerebellar ataxia. Transient hyperintensity within the cerebellum and/or brainstem may be seen on T2-weighted images [97]. Enhancement of cranial nerves and spinal nerve roots may be demonstrated, and the posterior portions of the spinal cord may be hyperintense on T2-weighted MRIs [56,98]. Fluid-attenuated inversion recovery (FLAIR) MRIs often demonstrate this alteration to greatest advantage in the acute phase. Cerebellar atrophy is generally demonstrated during the convalescent phase.

#### **Variant 4: Acute Ataxia Following Head Trauma (Adult or Child)**

Gait instability is a frequent component of concussion syndrome, and it may persist in association with a post-traumatic encephalopathy. Symptoms may be caused by damage to cerebellar, vestibular, or brain stem structures. Persisting ataxia may also relate to frontal lobe injury [99]. Interruption of the frontopontocerebellar tract (Arnold's bundle) may explain the association of ataxia with a frontal lobe lesion. This tract originates in Brodmann's area 10 and carries information regarding intentional movement to the contralateral cerebellum via the middle cerebellar peduncle [99]. Interruption of this tract along its course, or at its origin, deprives the cerebellum of frontal cortical input, resulting in impaired coordination and locomotion. In the presence of acute trauma, or in subjects with progressive post-traumatic ataxia, an expanding cyst or extra-axial hematoma should be considered separately. MRI is the preferred method for evaluation.

#### **Summary**

- MRI is usually the preferred initial modality for evaluation of ataxia in patients of all ages.
- Correlation with the clinical history and physical findings is essential for appropriate exam selection and interpretation.
- Ataxia may be of spinal origin. MRI of the spine may be considered if brain imaging is negative or inconclusive.
- Modalities such as PET, SPECT, MR spectroscopy and transcranial ultrasound are usually reserved for more challenging clinical problems.

#### **Anticipated Exceptions**

Nephrogenic systemic fibrosis (NSF) is a disorder with a scleroderma-like presentation and a spectrum of manifestations that can range from limited clinical sequelae to fatality. It appears to be related to both underlying severe renal dysfunction and the administration of gadolinium-based contrast agents. It has occurred primarily in patients on dialysis, rarely in patients with very limited glomerular filtration rate (GFR) (ie, <30 mL/min/1.73m<sup>2</sup>), and almost never in other patients. There is growing literature regarding NSF. Although some controversy and lack of clarity remain, there is a consensus that it is advisable to avoid all gadolinium-based contrast agents in dialysis-dependent patients unless the possible benefits clearly outweigh the risk, and to limit the type and amount in patients with estimated GFR rates <30 mL/min/1.73m<sup>2</sup>. For more information, please see the [ACR Manual on Contrast Media](#) [100].

#### **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, both because of 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 to 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.

Relative Radiation Level Designations		
Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
O	0 mSv	0 mSv
⊕	<0.1 mSv	<0.03 mSv
⊕ ⊕	0.1-1 mSv	0.03-0.3 mSv
⊕ ⊕ ⊕	1-10 mSv	0.3-3 mSv
⊕ ⊕ ⊕ ⊕	10-30 mSv	3-10 mSv
⊕ ⊕ ⊕ ⊕ ⊕	30-100 mSv	10-30 mSv
*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”.		

### Supporting Documents

- [ACR Appropriateness Criteria® Overview](#)
- [Procedure Information](#)
- [Evidence Table](#)

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The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.