

**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	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 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		☼ ☼ ☼
FDG-PET head	3		☼ ☼ ☼ ☼
MRI spectroscopy head	2	Selectively used as an adjunct to conventional MRI for characterizing indeterminate lesion(s).	O
<u>Rating Scale:</u> 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

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 or with contrast	8	MRI preferred if treatment is not unreasonably delayed. Combined vascular and cerebral evaluation should be considered. Fat saturated T1 axial images. Contrast administration may be appropriate if there is no evidence of infarction.	O
MRA head and neck with or without contrast	8	MRI preferred if treatment is not unreasonably delayed. Combined vascular and cerebral evaluation should be considered. If perfusion characterization is considered, MR with contrast is recommended.	O
CTA head and neck	8	MRI preferred if treatment will not be unreasonably delayed. Combined vascular and cerebral evaluation should be considered.	⊕ ⊕ ⊕
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.	⊕ ⊕ ⊕
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
MRI spectroscopy head	2		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

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
MRI spectroscopy head	6	May help distinguish abscess from other masses.	O
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
CTA head	4		☼ ☼ ☼
CT head without and with contrast	5		☼ ☼ ☼
CT temporal bone without contrast	5	Useful when skull base or middle ear disease is suspected.	☼ ☼ ☼
MRA head without contrast	4		O
CT head without contrast	4		☼ ☼ ☼
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

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		O
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."	O
MRI neck without and with contrast	6	See statement regarding contrast in text under "Anticipated Exceptions."	O
CT head without and with contrast	6		☼ ☼ ☼
CTA head and neck	6		☼ ☼ ☼
MRA head and neck	6		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

ATAXIA

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Summary of Literature Review

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. There can be associated abnormality in ocular motility and poor coordination of speech. Each of these alterations can relate to abnormal motor trajectory or placement during active movement (dysmetria), and/or to errors in the sequence and speed of motor activity. Abnormality in the rate, force, direction, and/or path of movement results in motion that is imprecise and dysfunctional. Purposeful, rapid alternating motion rates of muscle groups become slower and less fluid (dysdiadochokinesis), and the motion may have diminished rhythm (dysrhythmokinesis) [1]. Early in the course of an ataxia-causing disease process there may be decreased resistance to passive movement (hypotonia), and muscle stretch reflexes (DTRs) may demonstrate a “pendular” response. 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 can also be seen as components of ataxia. As an anatomic localizing sign, a wide-based stance, with feet several inches apart, is the most common, but not specific, clinical 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. Physical findings associated with ataxia and the utility of these findings relative to anatomic localization have been reviewed [1].

Overview of Imaging Modalities

Ataxia can 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 such a large number of anatomic regions can be responsible for ataxia, an appropriate imaging evaluation is often complex. Since optimal detail of posterior fossa structures can be obscured by beam-hardening artifact in computed tomography imaging (CT), magnetic resonance imaging (MRI) is usually the preferred initial modality for evaluating patients of all ages. Correlation with the patient's clinical history and physical findings is essential for appropriate study design and image interpretation. Among patients with distinct clinical ataxia, appropriate MRI or CT imaging of the cerebellum itself, or of the entire brain and spinal cord, may be entirely normal, and repeat imaging may be necessary after a variable period of time. This need is most likely to occur 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 [2-4]. 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 relative to 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 ACR Appropriateness Criteria[®] on “Vertigo and Hearing Loss” [5].

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

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hemangioblastomas, choroid plexus papillomas, extra-axial meningiomas, and a host of intra-axial, extra-axial, or diffuse leptomeningeal metastatic processes become more prevalent. Isolated frontal lobe and thalamic mass lesions can also present with varying manifestations of gait and limb ataxia. Unless there is a contraindication, 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 [6]. 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 [6-8]. The mass does not enhance with contrast, and it may demonstrate restricted diffusion [6-8]. Cowden syndrome, an autosomal dominant disorder characterized by cutaneous and noncutaneous hamartomas and by increased risk for breast, thyroid, gastrointestinal and GU neoplasias, is frequently associated [6,7].

Paraneoplastic cerebellar degeneration is clinically characterized by the subacute or acute onset of gait and limb ataxia, dysarthria, and ocular dysmetria [9]. It can occur in association with essentially any tumor type but most commonly occurs with breast, gynecologic, and lung tumors, and in association with Hodgkin's disease [9]. 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 [9]. Uncommonly there can be increased T2 signal intensity in the cerebellum or in other simultaneously involved brain parenchymal areas [10]. CT imaging and/or positron emission tomography (PET) imaging are indicated when an underlying primary is not evident [11].

Demyelinating Disorders

Multiple sclerosis patients commonly present with or subsequently manifest persisting 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 periventricular regions of increased T2 signal intensity, generalized cerebral volume loss, callosal and optic nerve involvement, and ring enhancement of active lesions is typical. MRI findings and the utility of advanced MRI techniques for patient evaluation and management, are reviewed [12].

Congenital Disorders

A large number of ataxia-associated disorders occur on a congenital basis [3]. In each of these processes MRI is preferred to CT. All of these disorders will most commonly manifest ataxia during early childhood

development. 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 can 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. Additional associated imaging alterations can include brain stem hypoplasia, lissencephaly, aprosencephaly, microcephaly, or variable and less prominent cerebral developmental alteration.

Though the disorders that can present as congenital ataxia are individually uncommon, a small number are more frequently recognized [13,14]. Dandy Walker syndrome, with ataxia, nystagmus, cranial nerve palsies, apneic episodes, hydrocephalus, and cognitive dysfunction, is characterized by hypoplasia of the cerebellar vermis, with an associated central spinal fluid collection that is predominately posterior to the cerebellum but continuous with the fourth ventricle [13]. 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 [13]. 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) [15]. Four types have been identified, each with somewhat variable clinical and imaging features, and with genetic alterations that involve different loci [16].

Rhombencephalosynapsis is a rare cerebellar dysplastic process that can occur alone or in association with other developmental anomalies [14]. In rhombencephalosynapsis there is vermian agenesis with fusion of the cerebellar hemispheres, apposition or fusion of the dentate nuclei, and fusion of the superior cerebellar peduncles. There is usually enlargement of the lateral ventricles, and there may be fusion of the thalamic nuclei [14]. There is a wide spectrum of clinical symptomatology, and some patients are clinically normal [14].

Congenital ataxia can also occur in association with perinatal cerebral infarction and in association with congenital cytomegalovirus or other infectious processes of the central nervous system [13,17]. Though uncommon relative to other forms of cerebral palsy, the imaging correlates of ataxic cerebral palsy have not been well defined [18].

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 [19]. The designation of “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, and absence of an identified genetic marker [20]. In these patients MRI generally demonstrates cerebellar and pontine volume loss [21].

SCA2 is a form of AD-SCA that is caused by the presence of 32 or more CAG trinucleotide repeats on the ATXN2 gene. Symptoms include 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. 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.

SCA3 (Machado-Joseph disease or MJD) 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.

Dentorubral-pallidolucyan atrophy (DRPLA) 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. [22]. MRI demonstrates cerebellar and brainstem volume loss with cerebral cortical atrophy [23]. Less frequently there is increased T2 signal intensity in deep white matter of the cerebral hemispheres, in the thalamus, and in the brainstem [23]. Diagnosis is established through the identification of >48 CAG repeats in the DRPLA gene [22].

Autosomal recessive spinocerebellar ataxias (AR-SCA) can 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 [24,25]. Imaging findings include diminished cross-sectional area of the spinal cord and medulla, and inconsistently the presence of cerebellar volume loss [24,25]. Diagnosis is by molecular genetic testing with GAA expansions in the FRDA gene, and other FRDA gene mutations [25].

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 there is a population-based prevalence of 1/40,000-100,000. There is an increased risk of leukemia and lymphoma, in part relating 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 [26]. The vermis also becomes atrophic, more in its superior than in its inferior portions [26]. Laboratory testing supports the diagnosis of A-T by identifying elevated serum alpha-fetoprotein, the absence of ATM protein in blood mononuclear cells [27], 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 [2].

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 [28]. Fragile X syndrome results from silencing of the fragile-X mental retardation 1 (FMR1) gene by the presence of >200 CGG repeats at the 5’-UTR location on the x-chromosome [28]. Predominant clinical findings include a characteristic facies, developmental delay, and mental retardation [28]. FXTAS, however, involves adults, predominately males, who are *carriers* of the fragile-X syndrome [29]. All have “premutation” alleles that contain 55-200 CGG repeats within the FMR1 gene [30]. Significant and progressive clinical findings include tremor, ataxia, autonomic instability, Parkinsonism, and cognitive decline [29,31].

Age at onset and severity have a positive correlation with CGG repeat length. Characteristic FXTAS brain MRI findings include brain stem atrophy and cerebral cortical atrophy. There may be increased T2 signal intensity in the white matter of atrophic middle cerebellar peduncles, as well as in deep and in subependymal cerebral white matter [32].

Multiple system atrophy (MSA) is a sporadic neurodegenerative disorder that can initially be manifest by ataxia (MSAc) or by Parkinsonism (MSAp) [33,34]. 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-Dragger 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 [33]. MRI demonstrates atrophy of the pons, cerebellum, and putamen, and there may be increased T2 signal intensity in the pons and middle cerebellar peduncles. The putamen are generally low in signal intensity on T2-weighted images, though there is often a narrow band of increased T2 signal intensity at the lateral aspect of the putamen [33-35].

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 [36]. 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 T2 signal intensity may be seen in the brainstem and/or basal ganglia on MRI [37].

A deficiency of coenzyme Q10 has been described in individuals with cerebellar ataxia, usually with childhood onset and often associated with seizures [38]. MRI may demonstrate cerebellar volume loss [38]. Symptoms may respond to treatment with coenzyme Q10.

Vanishing white matter disease is an inherited early childhood leucoencephalopathy whose most prominent initial symptom is ataxia [39]. Onset most commonly occurs at 2 to 6 years of age, though adolescent and adult onset have 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 [39].

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 [40]. MRI is the definitive diagnostic procedure [40]. It demonstrates low superficial T2 signal

intensity over cortex, brain stem, and/or spinal cord, and usually demonstrates cerebellar atrophy.

Spinal Cord and Peripheral Nerve-Related Ataxia

Ataxia that is potentially due to pathologic processes that originate within the spinal cord, or within the roots/nerves originating from the spinal cord, requires high resolution T1 and T2-weighted axial and sagittal imaging without and with contrast. Imaging needs to focus on the posterior columns and on the nerve roots. In pernicious anemia imaging findings depend on the duration and severity of the disorder. There may be early localized or relatively diffuse cord swelling with increased T2 signal intensity that is usually most evident in the posterior columns [41,42]. Late atrophy and persistent gliosis may develop, or all findings may resolve with treatment [41]. In patients with hypertrophic, inflammatory, or postinfectious polyneuropathies, nerve root enhancement and enlargement may be demonstrated with MRI [43].

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 [44].

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 [45,46].

Metronidazole (Flagyl)-induced cerebellar toxicity is associated with symptoms of ataxia, with MRI findings of increased T2 signal intensity and restricted diffusion in the dentate nuclei [47]. With symptom resolution MRI becomes normal [47].

In central pontine myelinolysis (osmotic demyelination syndrome), cerebellar or extrapyramidal symptoms have been observed. More typically, patients demonstrate coma, locked-in syndrome, or quadriparesis. This disorder is typically seen in chronic alcoholics or malnourished patients following the rapid correction of hyponatremia. Central pontine increased T2 signal intensity is the characteristic finding [48].

A leucoencephalopathy with initial symptoms of ataxia has also been reported to occur following the chronic inhalation of heroin vapors [44].

Vitamin E deficiency can occur in association with several acquired gastrointestinal disorders or with autosomal recessive defects in vitamin E transport [49,50]. Symptoms include ataxia with associated weakness, areflexia, and retinal degeneration. Imaging has demonstrated cerebellar atrophy and increased T2 signal intensity in the posterior columns of the spinal cord [49,50].

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 [48]. MRI demonstrates atrophy of the cerebellar vermis, especially superiorly, as well as volume loss involving pons, medulla, and cerebral hemispheres [48].

Wernicke encephalopathy is due to thiamine deficiency. It classically presents with ataxia, altered mental status, and abnormality of ocular motility. MRI demonstrates increased T2 signal intensity, reversible contrast enhancement, and reversible restricted diffusion in multiple areas including mammillary bodies, hypothalamus adjacent to the third ventricle, periaqueductal gray and white matter, pulvinar, and dorsomedial portions of the thalamic nuclei [48]. Small hemorrhagic foci can also be demonstrated in these regions.

Reversible posterior leukoencephalopathy is most commonly characterized by headache, altered consciousness, visual disturbance, and seizures [51]. Ataxia can be a component, especially when there is brainstem or cerebellar involvement [51,52]. Antecedent clinical conditions include hypertension, eclampsia, renal disease, and the use of cytotoxic or immunosuppressant drugs [51]. MRI findings include the presence of bilateral and generally symmetrically increased T2 signal intensity in the posterior parietal and occipital lobes. White matter is generally more involved than is cortex, and there is little or no enhancement following contrast administration [51,53]. Predominant brainstem involvement has also been described with apparent early restricted and subsequent increased diffusion in these same areas [52].

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

Strategically located ischemic or hemorrhagic insults can result in distinct and often prominent ataxia. Ataxia-associated infarction can be isolated to vascular distributions within portions of the cerebellum, medulla, pons, mesencephalon, red nucleus, thalamic nuclei, posterior limb of the internal capsule, or to frontal or parietal cerebral cortex [54-60]. Medially positioned infarctions involving the pons and medulla are uncommonly associated with ataxia [54,55]. Several named syndromes are associated with ataxia and focal regions of brainstem infarction [60,61]. 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. While brainstem and cerebellar infarction are predominately arterial in origin, venous infarction should also be considered [62]. The radiologic evaluation of ataxia generally requires MRI, with water diffusion characterization, and with time-of-flight MR angiography. Use is also made of neck vessel MRI, using T1-weighted images, without and with fat saturation, to exclude the possibility of dissection [63-65]. MR venography should be accomplished if there is

consideration of central or dural venous thrombosis. Catheter-based diagnostic angiography and/or CT angiography is occasionally necessary [65].

When posterior fossa or supratentorial brain parenchyma is involved, vascular malformations, angiopathy, or aneurysm rupture can each lead to the acute development of ataxia. When there is consideration of acute or subacute hemorrhage, CT imaging of the brain and CT angiography may replace or supplement MRI.

Recurrent or paroxysmal ataxia has been associated with several disorders, including epilepsy and migraine, as well as with a transient limb and trunk ataxia that can occur with high systemic fever in otherwise healthy children. These disorders can be idiopathic or can be associated with abnormalities in membrane calcium or potassium channel function, or with altered synaptic glutamate transport [66-68]. MRI may be normal, may demonstrate cerebellar volume loss, or may demonstrate extensive areas of cortical increased T2 signal intensity that may correlate with the possible simultaneous occurrence of hemiplegic migraine or recent seizure activity. MRI is the imaging modality of choice.

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

A number of infectious and postinfectious processes can be responsible for the development of ataxia. To detect infectious-process-related alterations in the cerebellum, MRI, without and with contrast administration, provides a distinct advantage over CT. This advantage is due to superior contrast resolution of MRI and to the absence of CT-related artifacts that can occur in association with bone at the margins of the posterior fossa.

Bacterial cerebellitis can occur in association with meningitis or with cerebritis involving the cerebral hemispheres. It can also occur independently. Penetrating trauma or transdural extension of an epidural process, most commonly from the temporal bone, also needs to be considered. Diffusion imaging and MR spectroscopy can narrow the differential diagnosis [69,70]. Multiple viral processes, including herpes and arbovirus, can also be associated with brainstem or cerebellar involvement [71].

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 each present with behavioral, emotional, and intellectual deterioration, followed by development of ataxia and dysarthria. Progression is to stupor and coma, with associated myoclonus being prominent in sCJD. The clinical course is more prolonged in vCJD. MRI demonstrates increased T2 signal intensity, and increased signal intensity on inversion recovery and diffusion-weighted sequences, in the heads of the caudate nuclei, the putamen, and regions of frontal, parietal, and occipital cortex. These alterations in signal intensity can initially be asymmetric [72]. There is eventual diffuse volume loss. While all forms of CJD can have increased T2 signal intensity and restricted diffusion in the thalamic nuclei

and in the pulvinar bilaterally, this focal alteration is especially prominent in vCJD [72].

Acute cerebellitis, also called acute cerebellar ataxia, is a parainfectious disorder that predominately, but not exclusively, occurs in childhood. Symptoms include headache, ataxia, photophobia, and varying findings associated with potential increased intracranial pressure or brainstem involvement. MRI demonstrates increased T2 signal intensity in the cerebellar hemispheres with associated mass effect [73]. Bilateral abnormality is more common than unilateral abnormality. There may be obstruction of CSF flow with enlargement of the lateral ventricles and upward herniation of posterior fossa structures [73]. There may also be cerebellar meningeal enhancement following contrast administration. Surgical decompression of the posterior fossa may be necessary [74]. Follow-up imaging may demonstrate cerebellar atrophy [75].

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, increased T2 signal intensity and restricted diffusion within portions of the pons, medulla and cerebellum [76,77].

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, and it is associated with transient high T2 signal intensity within the cerebellum and/or brainstem [78]. Enhancement of cranial nerves and spinal nerve roots can be demonstrated, and there may be increased T2 signal intensity within the posterior portions of the spinal cord [43,79]. Fluid-attenuated inversion recovery (FLAIR) images 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 due to damage to cerebellar, vestibular, or brain stem structures. Persisting ataxia can also relate to frontal lobe injury [80]. One possible explanation for the association of ataxia with a frontal lobe lesion of any origin is interruption of the frontopontocerebellar tract (Arnold’s bundle). This tract originates in Brodmann’s area 10 and carries information regarding intentional movement to the contralateral cerebellum via the middle cerebellar peduncle [80]. 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.

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/1.73m²), 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/1.73m². For more information, please see the [ACR Manual on Contrast Media](#) [81].

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
○	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 NS (not specified).		

Supporting Document(s)

- [ACR Appropriateness Criteria® Overview](#)
- [Procedure Contrast 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.