American College of Radiology
ACR Appropriateness Criteria®
Ataxia-Child

Variant 1: **Child. Acute ataxia, no history of recent trauma. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	☢☢☢
MRA head and neck without IV contrast	May Be Appropriate	O
MRA head without IV contrast	May Be Appropriate	O
MRA neck with IV contrast	May Be Appropriate	O
MRA neck without IV contrast	May Be Appropriate	O
MRI complete spine without and with IV contrast	May Be Appropriate	O
MRI complete spine without IV contrast	May Be Appropriate	O
CTA head and neck with IV contrast	May Be Appropriate	☢☢☢☢
CTA head with IV contrast	May Be Appropriate	☢☢☢☢
CTA neck with IV contrast	May Be Appropriate	☢☢☢
MR spectroscopy head without IV contrast	Usually Not Appropriate	O
MRI chest abdomen pelvis without and with IV contrast	Usually Not Appropriate	O
MRI chest abdomen pelvis without IV contrast	Usually Not Appropriate	O
MRV head and neck without IV contrast	Usually Not Appropriate	O
MRV head with IV contrast	Usually Not Appropriate	O
MRV head without and with IV contrast	Usually Not Appropriate	O
MRV head without IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CTV head with IV contrast	Usually Not Appropriate	☢☢☢☢
MIBG scan	Usually Not Appropriate	☢☢☢☢
CT chest abdomen pelvis with IV contrast	Usually Not Appropriate	☢☢☢☢
CT chest abdomen pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢☢
CT chest abdomen pelvis without IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without IV contrast	Usually Not Appropriate	☢☢☢☢

Variant 2: **Child. Acute ataxia, history of recent trauma. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	☢☢☢
MRA head and neck without IV contrast	May Be Appropriate	O
MRA head without IV contrast	May Be Appropriate	O
MRA neck with IV contrast	May Be Appropriate	O
MRA neck without IV contrast	May Be Appropriate	O
CTA head and neck with IV contrast	May Be Appropriate	☢☢☢☢
CTA head with IV contrast	May Be Appropriate	☢☢☢☢
CTA neck with IV contrast	May Be Appropriate	☢☢☢
MR spectroscopy head without IV contrast	Usually Not Appropriate	O
MRI complete spine without and with IV contrast	Usually Not Appropriate	O
MRI complete spine without IV contrast	Usually Not Appropriate	O
MRI head without and with IV contrast	Usually Not Appropriate	O
MRV head and neck without IV contrast	Usually Not Appropriate	O
MRV head with IV contrast	Usually Not Appropriate	O
MRV head without and with IV contrast	Usually Not Appropriate	O
MRV head without IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CTV head with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without IV contrast	Usually Not Appropriate	☢☢☢☢

<u>Variant 3:</u> **Child. Recurrent ataxia with interval normal neurology examination. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without IV contrast	Usually Appropriate	O
MR spectroscopy head without IV contrast	May Be Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
MRA head and neck without IV contrast	Usually Not Appropriate	O
MRA head without IV contrast	Usually Not Appropriate	O
MRA neck with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI complete spine without and with IV contrast	Usually Not Appropriate	O
MRI complete spine without IV contrast	Usually Not Appropriate	O
MRV head and neck without IV contrast	Usually Not Appropriate	O
MRV head with IV contrast	Usually Not Appropriate	O
MRV head without and with IV contrast	Usually Not Appropriate	O
MRV head without IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head without IV contrast	Usually Not Appropriate	☢☢☢
CTA head and neck with IV contrast	Usually Not Appropriate	☢☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢
CTA neck with IV contrast	Usually Not Appropriate	☢☢☢
CTV head with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without IV contrast	Usually Not Appropriate	☢☢☢☢

Variant 4: **Child. Chronic progressive ataxia. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
MRI complete spine without and with IV contrast	May Be Appropriate	O
MRI complete spine without IV contrast	May Be Appropriate	O
CT head without IV contrast	May Be Appropriate	☢☢☢
MR spectroscopy head without IV contrast	Usually Not Appropriate	O
MRA head and neck without IV contrast	Usually Not Appropriate	O
MRA head without IV contrast	Usually Not Appropriate	O
MRA neck with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRV head and neck without IV contrast	Usually Not Appropriate	O
MRV head with IV contrast	Usually Not Appropriate	O
MRV head without and with IV contrast	Usually Not Appropriate	O
MRV head without IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CTA head and neck with IV contrast	Usually Not Appropriate	☢☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢
CTA neck with IV contrast	Usually Not Appropriate	☢☢☢
CTV head with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without IV contrast	Usually Not Appropriate	☢☢☢☢

Variant 5: **Child. Chronic nonprogressive ataxia. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without IV contrast	Usually Appropriate	O
MRI complete spine without IV contrast	May Be Appropriate	O
CT head without IV contrast	May Be Appropriate	☢☢☢
MR spectroscopy head without IV contrast	Usually Not Appropriate	O
MRA head and neck without IV contrast	Usually Not Appropriate	O
MRA head without IV contrast	Usually Not Appropriate	O
MRA neck with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI complete spine without and with IV contrast	Usually Not Appropriate	O
MRI head without and with IV contrast	Usually Not Appropriate	O
MRV head and neck without IV contrast	Usually Not Appropriate	O
MRV head with IV contrast	Usually Not Appropriate	O
MRV head without and with IV contrast	Usually Not Appropriate	O
MRV head without IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CTA head and neck with IV contrast	Usually Not Appropriate	☢☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢☢
CTA neck with IV contrast	Usually Not Appropriate	☢☢☢
CTV head with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT complete spine without IV contrast	Usually Not Appropriate	☢☢☢☢

ATAXIA-CHILD

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Summary of Literature Review

Introduction/Background

Ataxia is the inability to generate coordinated voluntary movement, which can manifest clinically with signs and symptoms such as clumsiness, nystagmus, dysmetria, abnormal or unsteady gait, dysdiadochokinesis, or dyssynergia [1]. Although dysfunction of the cerebellum is a dominant cause of ataxia in children, disruptions in several neuronal circuits impacting the basal ganglia, cerebral cortex, spinal cord, and peripheral nerves, as well as the sensory and vestibular system, can also result in ataxia [1-6]. Other causes such as muscle weakness and hypotonia may manifest with gait and postural abnormalities that may mimic ataxia [1]. Children with epilepsy may present with postictal ataxia. "Pseudoataxia" may occur with functional disorders [7,8].

Specific manifestations of ataxia may correspond to certain causes or may be discovered by particular triggers. For example, truncal ataxia is typical of cerebellar vermian pathology [1]. A lurching gait, when triggered by head rotation, is typical of vestibular dysfunction [9]. Sensory ataxia can be revealed by a positive Romberg test, which examines the dorsal columns. [9]. Additionally, ataxia with specific signs can suggest the underlying disorder. Pupillary abnormalities may suggest drug or toxin ingestion versus third cranial nerve compression. Torticollis or resistance to head and neck motion may indicate pathology at the craniocervical junction, cord compression, or posterior fossa tumor [9].

Based on a systematic review of European data, the estimated prevalence of childhood ataxia due to genetic and acquired causes is approximately 26 per 100,000 children, although the true worldwide prevalence may be higher [10]. Regional variations exist in the prevalence of childhood ataxia, with a higher prevalence of genetic causes of ataxia in countries with high consanguinity, and a higher prevalence of infectious causes such as malaria and varicella in other regions where these diseases are more common [10].

Evaluation of ataxia requires careful review of demographics, history (especially duration of symptoms and the presence of additional neurological deficits), clinical examination, laboratory testing, and neuroimaging to reach a cohesive diagnosis [2,7,11-13]. In young children and infants, a detailed neurological examination is often challenging, and initial imaging therefore plays a critical role in arriving at a diagnosis.

The time course of illness (eg, acute, recurrent, chronic with or without progression) may indicate or rule out potential etiology. Acute onset ataxia typically refers to ataxia that develops within hours or days and frequently presents within 72 hours, whereas chronic ataxia is defined as ataxia lasting longer than 2 months [1,14]. Acutely presenting ataxia in children may be due to infectious, inflammatory, toxic, ischemic, or traumatic etiology. Intermittent or episodic ataxia in children may be manifestations of migraine, benign positional vertigo, or intermittent metabolic disorders. Whereas nonprogressive childhood ataxia suggests a congenital brain malformation or early prenatal or perinatal brain injury, progressive childhood ataxia may be due to inherited causes or acquired posterior fossa lesions that result in gradual cerebellar dysfunction.

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The purpose of this document is to describe the most common clinical scenarios of ataxia in childhood and to provide the clinician with guidance based on the existing literature so that they can choose the most appropriate initial imaging. All scenarios described herein relate to the initial imaging encounter; the role of imaging in managing and monitoring patients with ataxia is beyond the scope of this document. For appropriate care, patients with ataxia and head trauma should also have imaging guided by the ACR Appropriateness Criteria® topic on "Head Trauma-Child" [15].

Special Imaging Considerations

CT imaging is helpful in the assessment of children with ataxia with clinical suggestion of etiologies such as acute head trauma, acute intracranial hemorrhage, intracranial mass, or stroke. CT imaging in children should be performed using dedicated pediatric protocols to keep radiation exposure "as low as reasonably achievable." The Image Gently® website (http://www.imagegently.org) provides additional information.

MRI is useful in the assessment of childhood ataxia because of its ability to distinguish tissue contrast based on intrinsic tissue magnetic relaxation properties and magnetic susceptibility. However, because of the diagnostic quality, MRI examinations require the patient to lay motionless, and it may be challenging to acquire MRI in young children and noncooperative patients. Rapid-acquisition MRI protocols may be useful in imaging the brain without sedation in these patients, but the sensitivity of rapid MRI compared with routine MRI brain protocols for identifying intracranial pathology is uncertain. The ACR–ASNR–SPR Practice Parameter for the Performance of Functional Magnetic Resonance Imaging (fMRI) of the Brain [16] and the ACR–ASNR–SPR Practice Parameter for the Performance of Magnetic Resonance Imaging (MRI) of the Pediatric Spine [17] provide additional information.

Although MIBG (iodine-123 meta-iodobenzylguanidine) and CT imaging of the chest, abdomen, and pelvis is used in specific workup of children presenting with opsoclonus-myoclonus-ataxia syndrome, for diagnosing underlying neuroblastoma or ganglioneuroblastoma, they may not typically be the first line of imaging. It should be noted that opsoclonus-myoclonus-ataxia syndrome is one of the less common presentations of tumors such as neuroblastoma or ganglioneuroblastoma. A complete description of signs and symptoms of neuroblastoma or ganglioneuroblastoma and their workup are beyond the scope of this document.

Initial Imaging Definition

Initial imaging is defined as imaging at the beginning of the care episode for the medical condition defined by the variant. More than one procedure can be considered usually appropriate in the initial imaging evaluation when:

- There are procedures that are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

- There are complementary procedures (ie, more than one procedure is ordered as a set or simultaneously in which each procedure provides unique clinical information to effectively manage the patient's care).

Discussion of Procedures by Variant

Variant 1: Child. Acute ataxia, no history of recent trauma. Initial imaging.

Acute onset ataxia typically refers to ataxia that develops within hours or days and frequently presents within 72 hours [2]; subacute ataxia often refers to symptom onset of more than a week [22]. However, there is no clear clinical distinction between acute onset and subacute onset of ataxia in certain cases, and most studies of childhood acute ataxia combine these 2 groups together to include ataxia of several days' duration [2,13,18-21]. Most cases of acute childhood ataxia are transient and benign [20,22-25]. A recent single-center study suggests that postinfectious cerebellar ataxia is the most common cause of acute ataxia in children presenting to the emergency department, accounting for approximately 50% of these children [18]. In a multicenter study of 509 children (mean age 5.8 years) who presented to the emergency department with acute ataxia, the most common causes were infectious and postinfectious disorders (acute infectious cerebellitis, acute postinfectious cerebellar ataxia, or acute disseminated encephalomyelitis), which accounted for 33.6% of total ataxia cases; the next most common cause was brain tumors accounting for 11.2% of cases [19]. Other major causes of acute ataxia include intoxications, migraine-related ataxia, peripheral neuropathies, encephalitis, and vestibular dysfunction [18,19].

Opsoclonus-myoclonus-ataxia syndrome, a less common cause of immune-mediated acute childhood ataxia, only accounts for approximately 2% of acute childhood ataxias but is an important consideration because it is most commonly due to underlying neuroblastoma, ganglioneuroblastoma, or ganglioneuroma and less frequently due to infectious or postinfectious etiologies [18,19]. Opsoclonus-myoclonus-ataxia syndrome is one of the less common presentations of tumors such as neuroblastoma or ganglioneuroblastoma, and a complete description of signs, symptoms, and workup of neuroblastoma or ganglioneuroblastoma are beyond the scope of this document. Acute ataxia can also be a presenting feature (along with ophthalmoplegia and areflexia) of Miller-Fisher syndrome, a subtype of the Guillain-Barré syndrome characterized by the presence of the anti-GQ1b IgG antibody [1].

Cerebellar stroke is a rare cause of acute ataxia in children, with one series identifying 3 cases out of 364 children presenting to the emergency department with acute ataxia [26]. Despite its rarity, diagnosing acute cerebellar infarct is critical because of implications for appropriate diagnostic workup and management.

In a meta-analysis, Whelan et al [13] showed that clinically significant imaging findings were identified in 2.5% of patients with acute ataxia on CT and in 5% of patients on MRI. However, initial imaging was only performed in a small proportion of patients in the included studies; therefore, additional findings of interest may have been missed, limiting the generalizability of the results. Another retrospective study showed significant CT abnormalities in 7% and MRI abnormalities in 14% of children presenting with acute ataxia [18]. In a recent study of 141 children presenting to the emergency department with acute or subacute ataxia, 104 (73.8%) underwent neuroimaging, and abnormalities were found in 60.6% of those imaged (29.3% on head CT and 63.9% on brain MRI), but significant abnormalities—defined as requiring urgent surgical or medical management—were noted in only 13.5% of these children [20]. The vast majority (86%) of children with significant neuroimaging pathology had additional focal neurological findings, and only in 14% of the cases was ataxia the only presenting symptom [20].

Collective assessment of several studies suggests that, although acute imaging may reveal a clinically urgent condition [1,7,21,22,25-27], the yield of clinically significant findings may be highest in children >3 years of age with symptoms for >3 days duration and in the presence of extracerebellar symptoms, such as somnolence, encephalopathy, focal motor weakness, or cranial nerve involvement [13,18,20,26]. In young children presenting with acute cerebellar ataxia and a recent history of viral illness but without extracerebellar neurologic signs and symptoms and a negative urine drug screen, watchful waiting has been suggested [18], with imaging reserved for those with clinical deterioration. Therefore, the need for neuroimaging in the acute setting should be guided by the clinical presentation, history, and laboratory testing.

CT Head

In a large study assessing imaging in 104 children presenting with acute or subacute ataxia, CT abnormalities were identified in up to 29% of children [20]. Clinically significant findings have been identified on CT in 7% of children presenting with acute ataxia [18]. Although the study by Whelan et al [13] only showed significant intracranial abnormalities on CT in 2.5% of the children presenting with acute ataxia, their study was limited by the fact that only a small proportion of children had imaging. In the absence of preceding trauma, CT is useful in identifying nontraumatic acute intracranial hemorrhage but can also identify hydrocephalus, cerebellar edema, and calcifications [7,28]. However, CT is less sensitive than MRI in displaying intracranial pathology including posterior fossa abnormalities [20,26]. There is no relevant literature to support the use of contrast-enhanced head CT in the evaluation of acute childhood ataxia.

CT Complete Spine

There is no relevant literature to support the use of CT complete spine in the initial evaluation of a child with acute ataxia and no history of recent trauma.

CTA Head and Neck

CT angiography (CTA) of the head and neck may be helpful in children with ataxia due to a posterior circulation stroke, hemorrhage, or vascular malformation. However, this investigation may need to be decided based on the clinical presentation and concern for acute cerebellar infarct [7]. Stroke or vascular malformations are the etiology in only 1% to 3% of acute childhood ataxia [20,26]. Although there is no relevant literature distinguishing between CTA of head and neck versus CTA of either the head or neck alone in these rare vascular causes of acute childhood ataxia, CTA of the head and neck may be more beneficial than either CTA head or CTA neck alone because the site of vascular abnormality may be in either the head or the neck or both locations.

CTA Head
CTA of the head in conjunction with CTA of the neck may be helpful in children with ataxia due to a posterior circulation stroke, hemorrhage, or a vascular malformation. However, this investigation may need to be decided based on the clinical presentation and concern for acute cerebellar infarct [7]. Stroke or vascular malformations are the etiology in only 1% to 3% of acute childhood ataxia [20,26].

CTA Neck
CTA of the neck, in conjunction with CTA of the head, may be helpful in children with ataxia due to a posterior circulation stroke, hemorrhage, or a vascular malformation. However, this investigation may need to be decided based on the clinical presentation and concern for acute cerebellar infarct [7]. Stroke or vascular malformations are the etiology in only 1% to 3% of acute childhood ataxia [20,26].

CTV Head
There is no relevant literature to support the use of CT venography (CTV) head in the initial evaluation of a child with acute ataxia and no history of recent trauma.

MRI Head
In a study of 104 children imaged for acute or subacute ataxia, MRI abnormalities were identified in 64% of the cases [20]. However, clinically significant intracranial abnormalities on MRI have been reported in 14% of children presenting with acute or subacute ataxia [18,20]. MRI is more sensitive than CT in the evaluation of intracranial pathology in acute ataxia in children [20]. In a study by Luetje at al [20], abnormalities were identified on MRI but not on CT in 8 children. Intravenous (IV) contrast is helpful in infectious and postinfectious disorders, demyelinating conditions, and brain tumors, which are the common causes of acute cerebellar ataxia in children [19].

MRI Complete Spine
MRI of the spine may be helpful in children with ataxia due to causes such as acute disseminated encephalomyelitis, brain tumors, and demyelinating disease, all of which may show additional abnormalities of the spinal cord [29]. Generally, if there is a high clinical suspicion for a condition that may have spinal cord or paravertebral abnormalities, MRI of the spine can be included when imaging the neuroaxis. Although there is no relevant literature discussing the specific use of contrast in MRI of the spine in acute childhood ataxia, addition of postcontrast imaging may be useful in assessing neoplastic, demyelinating, and inflammatory lesions, which are common causes of acute cerebellar ataxia in children [20].

MR Spectroscopy Head
There is no relevant literature to support the use of MR spectroscopy head in the initial evaluation of a child with acute ataxia and no history of recent trauma.

MRA Head and Neck
MR angiography (MRA) of the head and neck may be a helpful tool in assessing ataxia due to a posterior circulation stroke, hemorrhage, or vascular malformation. However, a posterior circulation stroke is rare in children, and this investigation may need to be decided based on the clinical presentation and concern for acute cerebellar infarct [7]. Stroke or vascular malformations may present with ataxia in 1% to 3% of cases [20,26]. Although there is no relevant literature distinguishing between MRA of head and neck versus MRA of either the head or MRA neck alone in these rare vascular causes of acute childhood ataxia, MRA of the head and neck may be more beneficial than either MRA head or neck alone because the site of vascular abnormality may be in either the head or neck or both locations.

MRA Head
MRA of the head in conjunction with MRA of the neck may be a helpful tool in assessing ataxia due to posterior circulation stroke, hemorrhage, or vascular malformation. However, a posterior circulation stroke is rare in children, and this investigation may need to be decided based on the clinical presentation and concern for acute cerebellar infarct [7]. Stroke or vascular malformations may present with ataxia in 1% to 3% of cases [20,26].

MRA Neck
MRA of the neck in conjunction with MRA of the head may be a helpful tool in assessing ataxia due to posterior circulation stroke, hemorrhage, or vascular malformation. However, a posterior circulation stroke is rare in children, and this investigation may need to be decided based on the clinical presentation and concern for acute cerebellar infarct [7]. Stroke or vascular malformations may present with ataxia in 1% to 3% of cases [20,26].

MRV Head and Neck
There is no relevant literature to support the use of MR venography (MRV) head and neck in the initial evaluation of a child with acute ataxia and no history of recent trauma.

MRV Head
There is no relevant literature to support the use of MRV head in the initial evaluation of a child with acute ataxia and no history of recent trauma.

MIBG Scan
In a child presenting with acute opsoclonus-myoclonus-ataxia syndrome, an MIBG scan may be helpful in the workup for identifying neuroblastoma, ganglioneuroblastoma, or ganglioneuroma as the causative etiology [2,13]. In the absence of any relevant literature, an MIBG scan may not be beneficial as a first-line imaging test in a child presenting with acute ataxia.

CT Chest, Abdomen, and Pelvis
In a child presenting with acute opsoclonus-myoclonus-ataxia syndrome with concern for neuroblastoma or ganglioneuroblastoma or ganglioneuroma, initial imaging with CT of the chest, abdomen, and pelvis with IV contrast may be helpful for diagnosis, although the clinical utility of CT as an initial imaging modality is less clear [2,13].

MRI Chest, Abdomen, and Pelvis
In a child presenting with acute opsoclonus-myoclonus-ataxia syndrome, with clinical suspicion for neuroblastoma, ganglioneuroblastoma, or ganglioneuroma, MRI of the chest, abdomen, and pelvis without and with IV contrast may be helpful to identify the underlying mass [2,13]; however, there is no literature to support the use of this modality as an initial procedure.

Variant 2: Child. Acute ataxia, history of recent trauma. Initial imaging.

Head trauma is a rare but important cause of ataxia in children, accounting for approximately 5% of acute childhood ataxias [18]. Ataxia after recent head and neck trauma may be due to intracranial hemorrhage, cerebral contusion, concussion, or traumatic vertebral dissection [18,30].

There is insufficient evidence in the literature regarding the optimal imaging evaluation of children presenting with acute ataxia after recent trauma. However, it is reasonable to image these children for traumatic injury that may require emergent intervention, especially to identify intracranial hemorrhage, significant mass effect, or brain herniation that may require emergent neurosurgical consultation [7,22,25,26]. Vascular imaging is typically included in the diagnostic workup if there is suspicion of traumatic vascular injury such as vertebral artery dissection that may result in a cerebellar stroke. Patients with head or spine trauma should also have imaging guided by the ACR Appropriateness Criteria® topics on "Head Trauma-Child" [15] and "Suspected Spine Trauma-Child" [31].

CT Head
In a child with recent trauma and neurologic symptoms including ataxia, CT of the head is a useful mode of emergent investigation to assess for acute intracranial processes such as bleeding, mass effect, and herniation [7]. The addition of IV contrast may obscure small hemorrhages in the setting of trauma; see the ACR Appropriateness Criteria® topic on "Head Trauma-Child" [15] for further guidance.

CT Complete Spine
There is no relevant literature to support the use of CT complete spine in the initial evaluation of a child with acute ataxia following recent head trauma. Children with spine trauma should have imaging guided by the ACR Appropriateness Criteria® topic on "Suspected Spine Trauma-Child" [31].

CTA Head and Neck
CTA head and neck may be useful in the diagnostic workup of a child presenting with acute ataxia with a recent history of trauma, because of a vertebral artery dissection that may result in a cerebellar stroke and present with ataxia. Although most vertebral artery dissections are extracranial in location, CTA of the head and neck may be more beneficial than either CTA head or CTA neck alone to assess entire extent of vascular injury.

CTA Head
CTA head in conjunction with CTA of the neck may be useful in the diagnostic workup if there is suspicion of traumatic vascular injury such as vertebral artery dissection that may result in a cerebellar stroke and ataxia.

Although most vertebral artery dissections are extracranial in location, CTA of the head and neck may be more beneficial than either CTA head or CTA neck alone to assess entire extent of vascular injury.

CTA Neck

CTA neck in conjunction with CTA of the head may be useful in the diagnostic workup if there is suspicion of cervical trauma resulting in cervical vascular injury such as vertebral artery dissection that may result in a cerebellar stroke and ataxia. Although most vertebral artery dissections are extracranial in location, CTA of the head and neck may be more beneficial than either CTA head or CTA neck alone to assess entire extent of vascular injury.

CTV Head

There is no relevant literature to support the use of CTV head in the initial evaluation of a child with acute ataxia following recent trauma.

MRI Head

MRI is more sensitive than CT in detecting intracranial injury in children with acute ataxia; see the ACR Appropriateness Criteria® topic on "Head Trauma-Child" [15] for further guidance. However, its utility in the setting of acute ataxia after trauma has not been evaluated. MRI may be useful in identifying small intracranial hemorrhages, diffuse axonal injury, cerebral contusion, and small cerebellar infarcts from traumatic dissection that may be missed by CT [4,26,32,33]; see the ACR Appropriateness Criteria® topic on "Head Trauma-Child" [15] for further guidance. There is no literature to support the use of contrast-enhanced MRI head for this scenario.

MR Spectroscopy Head

There is no relevant literature to support the use of MR spectroscopy of the head in the initial evaluation of a child with acute ataxia following recent trauma.

MRA Head and Neck

MRA head and neck may be useful in the diagnostic workup if there is suspicion of traumatic vascular injury such as a vertebral artery dissection that may result in a cerebellar stroke and ataxia. Although most vertebral artery dissections are extracranial in location, MRA of the head and neck may be more beneficial than either MRA head or MRA neck alone to assess entire extent of vascular injury.

MRA Head

MRA head in conjunction with MRA of the neck may be useful in the diagnostic workup if there is suspicion of traumatic vascular injury such as a vertebral artery dissection that may result in a cerebellar stroke and ataxia. Although most vertebral artery dissections are extracranial in location, MRA of the head and neck may be more beneficial than either MRA head or MRA neck alone to assess entire extent of vascular injury.

MRA Neck

MRA neck may be useful in the diagnostic workup if there is suspicion of traumatic cervical vascular injury such as a vertebral artery dissection that may result in a cerebellar stroke and ataxia. Although most vertebral artery dissections are extracranial in location, MRA of the head and neck may be more beneficial than either MRA head or MRA neck alone to assess entire extent of vascular injury.

MRI Complete Spine

There is no relevant literature to support the use of MRI complete spine in the initial evaluation of a child with acute ataxia following recent head trauma. Initial evaluation of a child with acute ataxia following recent spine trauma should follow the ACR Appropriateness Criteria® topic on "Suspected Spine Trauma-Child" [31].

MRV Head and Neck

There is no relevant literature to support the use of MRV head and neck in the initial evaluation of a child with acute ataxia following recent trauma.

MRV Head

There is no relevant literature to support the use of MRV head in the initial evaluation of a child with acute ataxia following recent trauma.

Variant 3: Child. Recurrent ataxia with interval normal neurology examination. Initial imaging.

To meet the definition of recurrent ataxia, symptoms of each ataxic episode must resolve—or nearly completely resolve—before the onset of the next attack [34]. This mode of presentation is relatively infrequent in children. In one Canadian study of 185 pediatric patients with chronic ataxia, only 11.4% were categorized as episodic or intermittent ataxia [34]. Recurrent ataxia in a child may be a manifestation of basilar migraines, benign paroxysmal vertigo, genetic disorders such as autosomal dominant episodic ataxia, and metabolic disorders [1,22,25,27,34,35]. Benign paroxysmal vertigo is typically diagnosed by history, physical examination, and when vestibular testing and neuroimaging are normal. Basilar migraines are typically diagnosed based on clinical symptoms and with normal neuroimaging. Autosomal dominant episodic ataxias are a clinically heterogeneous group (at least 6 types described), with imaging findings such as vermian atrophy described in type 2 episodic ataxia. Inborn errors of metabolism, such as intermittent maple syrup urine disease, pyruvate dehydrogenase deficiency, and Hartnup disease can also present with intermittent ataxia during times of stress or illness [1,36,37]; imaging in these cases may be helpful in identifying and characterizing the underlying neurometabolic disease. Children with rotational occlusion of the vertebral artery, also known as bow hunter syndrome, may present with episodic neurologic symptoms including ataxia [38]; however, this condition is rare enough that it is not a diagnostic target for initial evaluation for a child presenting with recurrent ataxia and interval normal neurology examination.

CT Head
There is no relevant literature to support the use of head CT in the initial evaluation of a child with recurrent ataxia.

CT Complete Spine
There is no relevant literature to support the use of CT complete spine in the initial evaluation of a child with recurrent ataxia.

CTA Head and Neck
There is no relevant literature to support the use of CTA head and neck in the initial evaluation of a child with recurrent ataxia.

CTA Head
There is no relevant literature to support the use of CTA head in the initial evaluation of a child with recurrent ataxia.

CTA Neck
There is no relevant literature to support the use of CTA neck in the initial evaluation of a child with recurrent ataxia.

CTV Head
There is no relevant literature to support the use of CTV head in the initial evaluation of a child with recurrent ataxia.

MRI Head
Although there is inadequate literature regarding the utility of MRI in intermittent or episodic ataxia in childhood, head MRI can be useful in diagnosing metabolic disorders and genetic abnormalities [1,25,34]. Neuroimaging in patients with basilar migraine and childhood benign paroxysmal ataxia is typically normal, and establishing normal neuroimaging supports the diagnosis of these conditions in the appropriate clinical setting [1,25,34]. In these aforementioned causes, there is not enough evidence in the literature to support the use of contrast-enhanced MRI in the initial evaluation of a child with episodic or intermittent ataxia [1,25,34].

MRI Complete Spine
There is no relevant literature to support the use of MRI complete spine in the initial evaluation of a child with recurrent ataxia.

MR Spectroscopy Head
Some episodic ataxias may be due to underlying metabolic disorders, and in specific conditions in which an underlying metabolic disorder is suspected, MR spectroscopy may be useful in investigation.

MRA Head and Neck
There is no relevant literature to support the use of MRA head and neck in the initial evaluation of a child with recurrent ataxia.

MRA Head
There is no relevant literature to support the use of MRA head in the initial evaluation of a child with recurrent ataxia.

MRA Neck
There is no relevant literature to support the use of MRA neck in the initial evaluation of a child with episodic or intermittent ataxia.

MRV Head and Neck
There is no relevant literature to support the use of MRV head and neck in the initial evaluation of a child with recurrent ataxia.

MRV Head
There is no relevant literature to support the use of MRV head in the initial evaluation of a child with recurrent ataxia.

Variant 4: Child. Chronic progressive ataxia. Initial imaging.

Chronic progressive ataxia is a frequent presentation of ataxia in children. Signs and symptoms are typically of >2 months duration, but occasionally only a few weeks of symptoms may be present [5,14,34]. Cerebellar tumors, brain stem gliomas, and inflammatory disorders are collectively a common cause of chronic progressive ataxia in childhood [1,4,5,25]. Less commonly, inherited ataxias cause chronic progressive ataxias in children [1,4,5,25]. Inherited ataxias are a heterogenous group of clinically and genetically distinguished neurodegenerative disorders, which include autosomal dominant cerebellar ataxias, such as spinocerebellar ataxias, and autosomal recessive cerebellar ataxias, such as Friedreich ataxia.

Imaging plays an important role in children with chronic progressive ataxias. Brain imaging is crucial in the assessment of children with suspected brain tumors. In children with inherited chronic progressive ataxias, certain imaging findings can be helpful in the diagnostic workup [1,4,11,28,39-47]. Several hereditary cerebellar ataxias disorders have a progressive clinical course and varying progression of cerebellar hemispheric and vermian volume loss. In addition to cerebellar and vermian atrophy, associated signal abnormality and atrophy of the spinal cord and additional areas of the brain may be seen in certain conditions. Initial imaging provides a baseline for assessment of these abnormalities. Because of the phenotypic heterogeneity and the progressive nature of hereditary cerebellar ataxias, imaging in early childhood may be normal or subtly abnormal with imaging abnormalities becoming more apparent on follow up [14,46,48].

CT Head
Few studies describe the utility of CT in identifying intracranial calcifications in specific conditions such as Cockayne syndrome, which can present with chronic progressive ataxia in children [14,28]. Although CT may identify major structural abnormalities or intracranial mass lesions, MRI is more sensitive for this purpose.

CT Complete Spine
There is no relevant literature to support the use of CT complete spine in the initial evaluation of a child with chronic progressive ataxia.

CTA Head and Neck
There is no relevant literature to support the use of CTA head and neck in the initial evaluation of a child with chronic progressive ataxia.

CTA Head
There is no relevant literature to support the use of CTA head in the initial evaluation of a child with chronic progressive ataxia.

CTA Neck
There is no relevant literature to support the use of CTA neck in the initial evaluation of a child with chronic progressive ataxia.

CTV Head
There is no relevant literature to support the use of CTV head in the initial evaluation of a child with chronic progressive ataxia.

MRI Head
MRI of the head is useful in the clinical workup of children with chronic progressive ataxia and can identify brain tumors and hereditary neurodegenerative disorders [1,4,11,28,39-47]. In a study of 82 patients with spinocerebellar ataxia type 1, 3, or 6, and 32 normal controls, Schulz et al [46] identified significant atrophy of the brainstem,

cerebellar vermis, and cerebellar hemispheres in affected children. Contrast-enhanced MRI of the head is useful in characterization of brain tumors that are a common cause of chronic progressive ataxia in children [1,4,5,25].

MRI Complete Spine
MRI of the spine may be helpful in children with chronic progressive ataxia due to certain causes, such as central nervous system tumors and spinocerebellar ataxias. Postcontrast imaging may be required depending on case specifics, such as in the setting of central nervous system tumors to identify spinal metastatic disease.

MR Spectroscopy Head
There is no relevant literature to support the use of MR spectroscopy head in the initial evaluation of a child with chronic progressive ataxia. Altered metabolite ratios or presence of specific metabolites on MR spectroscopy may be useful in distinguishing posterior fossa tumor types and molecular subtypes.

MRA Head and Neck
There is no relevant literature to support the use of MRA head and neck in the initial evaluation of a child with chronic progressive ataxia.

MRA Head
There is no relevant literature to support the use of MRA head in the initial evaluation of a child with chronic progressive ataxia.

MRA Neck
There is no relevant literature to support the use of MRA neck in the initial evaluation of a child with chronic progressive ataxia.

MRV Head and Neck
There is no relevant literature to support the use of MRV head and neck in the initial evaluation of a child with chronic progressive ataxia.

MRV Head
There is no relevant literature to support the use of MRV head in the initial evaluation of a child with chronic progressive ataxia.

Variant 5: Child. Chronic nonprogressive ataxia. Initial imaging.

Chronic nonprogressive ataxias are comprised of a heterogenous group of cerebellar ataxias with early onset of cerebellar symptoms and no clinical change in severity during follow-up [1,4,25]. Commonly, symptom onset in these cases is within the first year of life, and this group of ataxias are referred to as chronic nonprogressive congenital ataxia [49]. Chronic nonprogressive ataxia in children may be due to syndromic, nonsyndromic, genetic, and acquired etiologies. In some syndromic causes such as Joubert syndrome and related disorders, ataxia is characteristic [50]. There are genetic causes of nonprogressive congenital ataxia with autosomal dominant, autosomal recessive, and x-linked inheritance patterns but with poor correlation between genotype and phenotype defined by imaging and clinical status [49]. Mutations in the Reelin signaling pathway are one of the autosomal recessive nonprogressive congenital ataxias with marked cerebellar and vermian hypoplasia and gyral simplification [4]. Developmental causes of nonprogressive congenital ataxia include posterior fossa malformations, such as rhombencephalosynapsis, Dandy-Walker syndrome, and Chiari II malformations [4]. Acquired causes of nonprogressive congenital ataxia include prenatally acquired cerebellar injury such as congenital cytomegalovirus infection, in utero cerebellar stroke, and perinatal disrupted development of the cerebellum as can be seen in premature infants and those with perinatal ischemic injury [51].

In the setting of chronic nonprogressive ataxia, imaging can be helpful in identifying the presence of congenital cerebellar, vermian, brainstem, and supratentorial brain malformations and acquired cerebellar disruptions due to prenatal or perinatal insult, although imaging can be normal in genetic etiologies [1,4,25,40].

CT Head
Although CT may provide an assessment of major structural abnormalities in the posterior fossa, CT is less sensitive than MRI for assessment of intracranial structures [14,28]. The use of IV contrast is generally not warranted in assessment of brain structural abnormalities that present with chronic nonprogressive ataxia in children.

CT Complete Spine
There is no relevant literature to support the use of CT complete spine in the initial evaluation of a child with chronic nonprogressive ataxia.

CTA Head and Neck
There is no relevant literature to support the use of CTA head and neck in the initial evaluation of a child with chronic nonprogressive ataxia.

CTA Head
There is no relevant literature to support the use of CTA head in the initial evaluation of a child with chronic nonprogressive ataxia.

CTA Neck
There is no relevant literature to support the use of CTA neck in the initial evaluation of a child with chronic nonprogressive ataxia.

CTV Head
There is no relevant literature to support the use of CTV head in the initial evaluation of a child with chronic nonprogressive ataxia.

MRI Head
MRI is the most widely utilized imaging method to evaluate chronic nonprogressive ataxia [4,32]. In a child with chronic nonprogressive ataxia, MRI is useful in identifying cerebellar and brainstem malformations to aid diagnosis. Imaging can be particularly useful in the diagnosis of certain syndromic causes of chronic nonprogressive ataxia such as rhombencephalosynapsis, Dandy-Walker or Joubert syndrome, and related disorders that have characteristic imaging findings [50]. Diffusion tensor imaging can facilitate assessment of white matter tract structure in congenital brainstem and cerebellar anomalies [4,6,43,52]. For example, absence of decussation of the superior cerebellar peduncles in Joubert syndrome and abnormal dorsal pontine transverse white matter bundles in pontine tegmental cap dysplasia are seen on diffusion tensor imaging [4]. There is no literature to support the use of IV contrast in this setting.

MRI Complete Spine
Because some congenital brainstem and cerebellar anomalies that present with chronic nonprogressive ataxia are associated with spinal anomalies, MRI of the complete spine may be helpful in individual cases [44]. There is no literature to support the use of contrast-enhanced imaging of the spine in this scenario.

MR Spectroscopy Head
There is no relevant literature to support the use of MR spectroscopy head in the initial evaluation of a child with chronic nonprogressive ataxia.

MRA Head and Neck
There is no relevant literature to support the use of MRA head and neck in the initial evaluation of a child with chronic nonprogressive ataxia.

MRA Head
There is no relevant literature to support the use of MRA head in the initial evaluation of a child with chronic nonprogressive ataxia.

MRA Neck
There is no relevant literature to support the use of MRA neck in the initial evaluation of a child with chronic nonprogressive ataxia.

MRV Head and Neck
There is no relevant literature to support the use of MRV head and neck in the initial evaluation of a child with chronic nonprogressive ataxia.

MRV Head
There is no relevant literature to support the use of MRV head in the initial evaluation of a child with chronic nonprogressive ataxia.

Summary of Recommendations

- **Variant 1**: MRI head without and with IV contrast or MRI head without IV contrast or CT head without IV contrast is usually appropriate for the initial imaging of a child with acute ataxia and no history of recent trauma. These procedures are equivalent alternatives (ie, only one initial procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- **Variant 2**: MRI head without IV contrast or CT head without IV contrast is usually appropriate for the initial imaging of a child with acute ataxia and history of recent trauma. These procedures are equivalent alternatives (ie, only one initial procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- **Variant 3**: MRI head without IV contrast is usually appropriate for the initial imaging of a child with recurrent ataxia with interval normal neurology examination.
- **Variant 4**: MRI head without and with IV contrast or MRI head without IV contrast is usually appropriate for the initial imaging of a child with chronic progressive ataxia. These procedures are equivalent alternatives (ie, only one initial procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- **Variant 5:** MRI head without IV contrast is usually appropriate for the initial imaging of a child with chronic nonprogressive ataxia.

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

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	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.
May Be Appropriate (Disagreement)	5	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.
Usually Not Appropriate	1, 2, or 3	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.

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 [53].

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."

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