



## Complete Summary

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### GUIDELINE TITLE

ACR Appropriateness Criteria™ for ataxia.

### BIBLIOGRAPHIC SOURCE(S)

Johnson BA, Drayer BP, Anderson RE, Braffman B, Davis PC, Deck MD, Hasso AN, Masaryk T, Pomeranz SJ, Seidenwurm D, Tanenbaum L, Masdeu JC. Ataxia. American College of Radiology. ACR Appropriateness Criteria. Radiology 2000 Jun; 215(Suppl):573-8. [40 references]

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## SCOPE

### DISEASE/CONDITION(S)

Ataxia

### GUIDELINE CATEGORY

Diagnosis

### CLINICAL SPECIALTY

Family Practice  
Geriatrics  
Internal Medicine  
Medical Genetics  
Neurology  
Pediatrics  
Radiology

### INTENDED USERS

Health Plans  
Hospitals  
Managed Care Organizations  
Physicians  
Utilization Management

#### GUIDELINE OBJECTIVE(S)

To evaluate the appropriateness of initial radiologic examinations for ataxia

#### TARGET POPULATION

Patients with ataxia

#### INTERVENTIONS AND PRACTICES CONSIDERED

1. Magnetic resonance:
  - Plain (cranial to include upper cervical spine)
  - With contrast (cranial to include upper cervical spine)
  - Magnetic resonance spectroscopy
  - Magnetic resonance angiography
2. Computed tomography:
  - Plain
  - With contrast

#### MAJOR OUTCOMES CONSIDERED

Utility of radiologic examinations in differential diagnosis

## METHODOLOGY

#### METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

#### DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

The guideline developer performed literature searches of recent peer-reviewed medical journals, primarily using the National Library of Medicine's MEDLINE database. The developer identified and collected the major applicable articles.

#### NUMBER OF SOURCE DOCUMENTS

The total number of source documents identified as the result of the literature search is not known.

#### METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Expert Consensus (Delphi Method)  
Weighting According to a Rating Scheme (Scheme Not Given)

#### RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

#### METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review with Evidence Tables

#### DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

One or two topic leaders within a panel assume the responsibility of developing an evidence table for each clinical condition, based on analysis of the current literature. These tables serve as a basis for developing a narrative specific to each clinical condition.

#### METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus (Delphi)

#### DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Since data available from existing scientific studies are usually insufficient for meta-analysis, broad-based consensus techniques are needed to reach agreement in the formulation of the Appropriateness Criteria. Serial surveys are conducted by distributing questionnaires to consolidate expert opinions within each panel. These questionnaires are distributed to the participants along with the evidence table and narrative as developed by the topic leader(s). Questionnaires are completed by the participants in their own professional setting without influence of the other members. Voting is conducted using a scoring system from 1-9, indicating the least to the most appropriate imaging examination or therapeutic procedure. The survey results are collected, tabulated in anonymous fashion, and redistributed after each round. A maximum of three rounds is conducted and opinions are unified to the highest degree possible. Eighty (80) percent agreement is considered a consensus. If consensus cannot be reached by this method, the panel is convened and group consensus techniques are utilized. The strengths and weaknesses of each test or procedure are discussed and consensus reached whenever possible.

#### RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

#### COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

## METHOD OF GUIDELINE VALIDATION

Internal Peer Review

## DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Criteria developed by the Expert Panels are reviewed by the American College of Radiology (ACR) Committee on Appropriateness Criteria and the Chair of the ACR Board of Chancellors.

## RECOMMENDATIONS

### MAJOR RECOMMENDATIONS

ACR Appropriateness Criteria™

Clinical Condition: Ataxia

Variant 1: Child with ataxia.

Radiologic Exam Procedure	Appropriateness Rating	Comments
Magnetic resonance (cranial - to include upper cervical spine)	8	
Magnetic resonance with contrast (cranial - to include upper cervical spine)	4	
Magnetic resonance spectroscopy	4	
Computed tomography	4	If magnetic resonance imaging not available or contraindicated.
Computed tomography with contrast	4	If magnetic resonance imaging not available or contraindicated.
<u>Appropriateness Criteria Scale</u>  1 2 3 4 5 6 7 8 9  1=Least appropriate 9=Most appropriate		

Clinical Condition: Ataxia

Variant 2: Young adult with ataxia.

Radiologic Exam Procedure	Appropriateness Rating	Comments
Magnetic resonance (cranial - to include upper cervical spine)	8	
Magnetic resonance with contrast (cranial - to include upper cervical spine)	4	Indicated if mass seen is suggestive of neoplasm or abscess or to improve diagnostic specificity.
Magnetic resonance spectroscopy	4	Application currently being evaluated.
Computed tomography	4	If magnetic resonance imaging not available or contraindicated.
Computed tomography with contrast	4	If magnetic resonance imaging not available or contraindicated.
<u>Appropriateness Criteria Scale</u>  1 2 3 4 5 6 7 8 9  1=Least appropriate 9=Most appropriate		

Clinical Condition: Ataxia

Variant 3: Elderly patient with ataxia.

Radiologic Exam Procedure	Appropriateness Rating	Comments
Magnetic resonance (cranial - to include upper cervical spine)	8	
Magnetic resonance with contrast (cranial - to include upper cervical spine)	6	
Computed tomography	5	If magnetic resonance imaging contraindicated or acute onset.
Computed tomography with contrast	4	
Magnetic resonance angiography	4	If vascular disease suspected.
Magnetic resonance spectroscopy	3	Application currently being evaluated.

## Appropriateness Criteria Scale

1 2 3 4 5 6 7 8 9

1=Least appropriate 9=Most appropriate

### Summary

Ataxia is the inability to coordinate muscles in the execution of voluntary movement. There are multiple causes and categories of ataxia which include hereditary and acquired disorders. Imaging studies contribute to the evaluation of patients who have known underlying disease and those without a previous diagnosis. Cerebellar symptoms from acute lesions may resolve due to compensation by other nervous system components making a clinical diagnosis difficult. In addition, lesions of the brain stem, spinal cord, and frontal lobe may produce a cerebellar type of ataxia with no visible abnormality in the cerebellum itself. This may occur due to the inherent links between the cerebral hemispheres, brain stem, spinal cord, and the cerebellum.

There are a number of systems for classifying the various forms of ataxia, but from an anatomic standpoint, it is useful to divide the hereditary ataxias into those that are predominantly spinal forms and those which arise from cerebellar diseases. Additional causes include metabolic disorders and ataxia-telangiectasia. There are also a number of acquired disorders in several disease categories that may present with ataxia. (See the Appendix of the guideline document for a list of the various forms of ataxia).

### Hereditary Ataxias

Inherited ataxias may be classified according to several systems. The atrophic ataxias are generally categorized as either predominantly spinal forms or predominantly cerebellar forms. Predominantly spinal forms include Friedreich's ataxia, which typically involves the spinal cord with relative sparing of the intracranial structures. With advanced Friedreich's ataxia, however, the vermis and medulla are frequently involved as well. Although imaging findings are often contributory, a strong family history and clinical symptoms often contribute more to the diagnosis. On the other hand, clinical criteria are often not sufficient to diagnose progressive ataxias. Magnetic resonance imaging is often useful to distinguish Friedreich's ataxia from non-Friedreich's diseases. The anatomopathologic classification by Greenfield considered Friedreich's ataxia, cerebello-olivary ataxia and olivopontocerebellar ataxias as distinct entities. He added a fourth category: Multisystem atrophy. These groups have distinct magnetic resonance (MR) imaging features. The more recently described infantile onset spinocerebellar ataxia has imaging features that overlap those of olivopontocerebellar atrophy, spinocerebellar, and cerebellar cortical atrophies.

Predominantly cerebellar forms of hereditary atrophic ataxias include olivopontocerebellar atrophy. Atrophy of the structures implicated in the name of this disorder are well demonstrated with multiplanar magnetic resonance imaging. Cerebello-olivary atrophy is an autosomal dominant inherited atrophic ataxia in which the pons demonstrates normal dimensions. Clinical and magnetic resonance

features of this disorder also permit a confident clinical diagnosis. Some of the primarily cerebellar forms of hereditary ataxia include congenital deafness of Hallgren, dentatorubral degeneration, familial corticocerebellar atrophy, and early onset cerebellar ataxia with retained tendon reflexes. Computed tomography studies demonstrate brain stem and cerebellar atrophy in a majority of patients with early onset cerebellar ataxia, while a minority develop cerebral atrophy. In a study by Pal and coworkers, there was no correlation between the duration of the disease and the degree of disability or computed tomography abnormalities. In comparing patients with spinal and cerebellar ataxia, de Michele and coworkers found that cervical cord atrophy was a consistent finding in Friedreich's ataxia, and in late stages there was often atrophy of the cerebellum and brain stem. The most frequent finding in early onset cerebellar ataxia was cerebellar atrophy, with variable involvement of the cervical cord and brain stem.

Ataxia telangiectasia is a hereditary multisystem disease consisting of progressive cerebellar ataxia with onset in infancy or childhood. There are progressive oculomucocutaneous telangiectasias and a proneness to sinus and lung infections, as well as neoplasms. It is inherited in a Mendelian recessive fashion. Imaging studies may demonstrate vermian atrophy, a prominent fourth ventricle and cisterna magna. Its multiplanar capabilities make magnetic resonance preferable to computed tomography for evaluation of this entity. In addition, diffuse hyperintense signal abnormalities may be seen in the white matter in early stages of ataxia telangiectasia. If these imaging findings are discovered prior to the development of more typical clinical features of this entity, an erroneous diagnosis of leukodystrophy may be made.

Several metabolic disorders can cause progressive cerebellar manifestations. Evaluation of these disorders relies on clinical and laboratory findings. Although the magnetic resonance findings are often nonspecific, they may contribute to limiting the differential diagnosis in combination with clinical and laboratory data. In addition, magnetic resonance spectroscopy may provide additional diagnostic clues to a specific metabolic abnormality.

Although migration abnormalities are often associated with seizures, developmental and motor delay, ataxia may also accompany the symptom complex in patients with extensive migration abnormalities. Magnetic resonance imaging allows the identification of subtle neuronal migration defects, and may also help diagnose patients with ataxia who have concomitant supratentorial defects.

#### Acquired Ataxias - Inflammatory

A number of infectious and postinfectious etiologies for the development of ataxia have been reported. To detect abnormalities in the cerebellum, magnetic resonance imaging provides a well-known advantage over computed tomography due to superior contrast resolution and absence of artifacts due to adjacent bone in the posterior fossa. Postinfectious white matter lesions in the cerebellar hemispheres have been reported in patients with acute cerebellar ataxia with subsequent resolution of the hyperintensities on T2-weighted images and the gradual development of cerebellar atrophy. The Fisher syndrome, which is considered a variant of Guillain-Barre' syndrome, involves the peripheral and central nervous systems. It is characterized by ophthalmoplegia, areflexia, and

cerebellar ataxia and is also associated with transient high-signal-intensity lesions in the cerebellum on long TR images. Long TR magnetic resonance images or fluid attenuated inversion recovery sequences demonstrate the lesions to best advantage in the acute phase. All imaging sequences demonstrate the atrophic changes which often occur during the convalescent phase.

## Neoplasms

Cerebellar tumors are an important consideration, especially in the child presenting with new onset ataxia and other posterior fossa symptoms. Between the second year of life and early in the second decade of life, posterior fossa tumors are more common than supratentorial tumors, which are more common during the first year of life and after the 12-14th year. Cerebellar and brain stem astrocytomas, medulloblastomas and ependymomas are the most common posterior fossa lesions seen in children. The previous fifteen years of clinical experience and the imaging literature have demonstrated that magnetic resonance is more sensitive than computed tomography for the detection and characterization of posterior fossa neoplasms.

## Trauma

Rarely does posttraumatic sequelae include ataxia. Although this is typically due to damage to the posterior fossa structures, frontal lobe injury (which is much more common) may also result in subsequent ataxia. Gait difficulties resulting from frontal lobe disease are less common but well recognized. One possible explanation for the development of ataxia due to a frontal lobe lesion implicates involvement of the frontopontocerebellar tract, which originates mostly in Brodmann's areas 8, 9, 10, 45, 46 and carries information regarding intentional movement to the contralateral cerebellum via the pontocerebellar peduncle. Interruption of this tract deprives the cerebellum of this information, resulting in impairment of coordination and locomotion.

## Metabolic/Toxic

Cases of substance and solvent abuse resulting in gait impairment and associated magnetic resonance abnormalities have been described. In addition to atrophic changes, hyperintensity on T2-weighted images has been reported in the white matter, basal ganglia and thalami. Methyl-mercury poisoning (Minamata disease) is a neurological illness caused by ingestion of contaminated seafood. Magnetic resonance imaging in affected patients may reveal atrophy of the cerebellar vermis and hemispheres, as well as the calcarine cortex. In rare cases of central pontine myelinolysis, cerebellar or extrapyramidal symptoms have been observed. More typically, patients demonstrate coma, locked-in syndrome or quadriplegia. In this metabolic disorder, typically seen in chronic alcoholics or malnourished patients following the rapid correction of hyponatremia, hyperintensity seen in the pons on long TR images is a characteristic pattern which is more difficult to detect using other imaging modalities.

## Vascular Etiologies

Strategically located ischemic insults may also result in ataxia. Lesions in the medullary tegmentum, the ventroposterior thalamus, and in the cerebellum may

result in ataxia. These lesions manifest with T1 and T2 prolongation on magnetic resonance, and are more difficult to detect on computed tomography, especially when small lesions are evaluated early in the course of disease. In three patients described with ataxic hemiparesis by Helweg-Larsen and coworkers, lesions were contralateral to the side of involvement involving the posterior limb of the internal capsule, the midpons and the red nucleus. None of the lesions were identified on computed tomography. Superficial siderosis of the central nervous system is a rare fatal disorder characterized by ataxia and sensorineural hearing loss caused by recurrent bleeding into the cerebral spinal fluid. The diagnosis can be reliably confirmed using magnetic resonance imaging.

### Degenerative/Demyelinating Diseases

Some patients with multiple sclerosis manifest with cerebellar or gait ataxia. Evaluation of these patients with magnetic resonance provides an accurate means for assessing lesions. However, it does not confirm demyelination or neuronal loss. Proton magnetic resonance spectroscopy has the potential to detect axonal loss, which manifests as a decrease in N-acetyl aspartate.

Nonhereditary cerebellar ataxia is an idiopathic degenerative condition resulting in progressive cerebellar ataxia, typically with onset after 20 years of age. In these patients, a greater degree of pontine atrophy is seen in patients with rigidity, hyperreflexia, and/or hypokinesia, as compared with those who manifest with pure cerebellar signs. Approximately 1/3 of patients have hyperintense foci in the pons and medulla on long TR images. Patients with a smaller ventral pons demonstrate a more rapid progression and poorer prognosis as well. Magnetic resonance has proven to be more useful than computed tomography in demonstrating atrophy of cerebellar structures, brain stem, and spinal cord, and facilitates distinguishing the diagnosis from other diseases, such as multiple sclerosis.

The noninvasive evaluation of patients with degenerative spinal ataxia requires high resolution T2-weighted images in order to detect abnormalities in the posterior columns. Vivo studies and postmortem histopathologic correlative studies reveal volume loss and high signal intensity in the posterior columns associated with subtle deformity of the dorsal surface of the cord, which corresponds to tract degeneration on histopathologic examination.

### CLINICAL ALGORITHM(S)

Algorithms were not developed from criteria guidelines.

## EVIDENCE SUPPORTING THE RECOMMENDATIONS

### TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The recommendations are based on analysis of the current literature and expert panel consensus.

## BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

### POTENTIAL BENEFITS

Appropriate selection of radiologic exams for the diagnosis of ataxia.

### POTENTIAL HARMS

Not stated

## QUALIFYING STATEMENTS

### QUALIFYING STATEMENTS

An American College of Radiology (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 exams 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 U.S. Food and Drug Administration (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.

## IMPLEMENTATION OF THE GUIDELINE

### DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

## INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

### IOM CARE NEED

Living with Illness

### IOM DOMAIN

Effectiveness

## IDENTIFYING INFORMATION AND AVAILABILITY

### BIBLIOGRAPHIC SOURCE(S)

Johnson BA, Drayer BP, Anderson RE, Braffman B, Davis PC, Deck MD, Hasso AN, Masaryk T, Pomeranz SJ, Seidenwurm D, Tanenbaum L, Masdeu JC. Ataxia. American College of Radiology. ACR Appropriateness Criteria. Radiology 2000 Jun; 215(Suppl):573-8. [40 references]

### ADAPTATION

Not applicable: The guideline was not adapted from another source.

### DATE RELEASED

1999

### GUIDELINE DEVELOPER(S)

American College of Radiology - Medical Specialty Society

### SOURCE(S) OF FUNDING

The American College of Radiology (ACR) provided the funding and the resources for these ACR Appropriateness Criteria™.

### GUIDELINE COMMITTEE

ACR Appropriateness Criteria™ Committee, Expert Panel on Neurologic Imaging

### COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Names of Panel Members: Thomas Masaryk, MD; Burton P. Drayer, MD; Robert E. Anderson, MD; Bruce Braffman, MD; Patricia C. Davis, MD; Michael D. F. Deck, MD; Anton N. Hasso, MD; Blake A. Johnson, MD; Stephen J. Pomeranz, MD; David Seidenwurm, MD; Lawrence Tanenbaum, MD; Joseph C. Masdeu, MD, PhD.

### FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

### GUIDELINE STATUS

This is the current release of the guideline.

The ACR Appropriateness Criteria™ are reviewed after five years, if not sooner, depending upon introduction of new and highly significant scientific evidence. The next review date for this topic is 2004.

## GUIDELINE AVAILABILITY

Electronic copies: Available from the [American College of Radiology \(ACR\) Web site](#).

Print copies: Available from ACR, 1891 Preston White Drive, Reston, VA 20191.  
Telephone: (703) 648-8900.

## AVAILABILITY OF COMPANION DOCUMENTS

None available

## PATIENT RESOURCES

None available

## NGC STATUS

This summary was completed by ECRI on July 31, 2001. The information was verified by the guideline developer as of August 24, 2001.

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