

**Ataxia**  
**EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Strength of Evidence
1. Gilman S, Gelb DJ. Disorders of the Cerebellum. In: Griggs RC, Joynt RJ, eds. <i>Baker's Clinical Neurology</i> : Lippincott Williams & Wilkins; 2003.	15	N/A	Book chapter.	N/A	N/A
2. Bird TD. Hereditary Ataxia Overview. August 4, 2006; <a href="http://www.geneclinics.org/profiles/ataxias/details.html">http://www.geneclinics.org/profiles/ataxias/details.html</a> . Accessed 2006/8/21.	12	N/A	Review characteristics, diagnosis and management of hereditary ataxia.	Genetic forms of ataxia are diagnosed by family history, physical examination, and neuroimaging.	4
3. Neuromuscular Disease Center WU, St. Louis, MO. Ataxias: Classification. <a href="http://neuromuscular.wustl.edu/ataxia/aindex.html">http://neuromuscular.wustl.edu/ataxia/aindex.html</a> . Accessed 3/5/2009.	15	N/A	Extensive classification of disorders associated with ataxia.	N/A	N/A
4. Schapira AHV, Samuels MA, et al. <i>Spinocerebellar Degenerations: The Ataxias and Spastic Paraplegias</i> . Philadelphia, PA: Butterworth, Heinemann, Elsevier; 2007.	15	N/A	Book chapter.	N/A	N/A
5. American College of Radiology. ACR Appropriateness Criteria®: vertigo and hearing loss. Available at: <a href="http://www.acr.org/SecondaryMainMenuCategories/quality_safety/app_criteria/pdf/ExpertPanelonNeurologicImaging/VertigoandHearingLossDoc14.aspx">http://www.acr.org/SecondaryMainMenuCategories/quality_safety/app_criteria/pdf/ExpertPanelonNeurologicImaging/VertigoandHearingLossDoc14.aspx</a> . Accessed July 2009.	15	N/A	ACR Appropriateness Criteria® for “Vertigo and Hearing Loss.”	N/A	3
6. Abel TW, Baker SJ, Fraser MM, et al. Lhermitte-Duclos disease: a report of 31 cases with immunohistochemical analysis of the PTEN/AKT/mTOR pathway. <i>J Neuropathol Exp Neurol</i> 2005; 64(4):341-349.	13	31	To review histopathologic and molecular characteristics of Lhermitte-Duclos disease (LDD), and its association with Cowden Disease (CD).	Basic imaging findings and histopathology are illustrated. The pathogenesis of LDD is thought to relate to loss of inhibitory regulation on cell growth and migration. Search for manifestations of CD is needed.	3
7. Perez-Nunez A, Lagares A, Benitez J, et al. Lhermitte-Duclos disease and Cowden disease: clinical and genetic study in five patients with Lhermitte-Duclos disease and literature review. <i>Acta Neurochir (Wien)</i> 2004; 146(7):679-690.	14	5	Clinical, diagnostic and genetic study to determine association of LDD with CD. Both a case report and review of literature were performed.	4/5 patients treated for LDD were also diagnosed of CD. LDD is closely related to CD. Authors suggest LDD can also appear as an isolated condition.	4
8. Gaballo A, Palma M, Dicuonzo F, Carella A. Lhermitte-Duclos disease: MR diffusion and spectroscopy. <i>Radiol Med (Torino)</i> 2005; 110(4):378-384.	14	2	Describe MRI diffusion and spectroscopic imaging findings in LDD.	Diffusion coefficients were normal relative to surrounding cerebellar parenchyma. Proton spectroscopy demonstrated a lactate peak and reduction in the choline peak.	4

\* See Last Page for Key

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9. Bruylant K, Crols R, Humbel RL, Appel B, De Deyn PP. Probably anti-Tr associated paraneoplastic cerebellar degeneration as initial presentation of a squamous cell carcinoma of the lung. <i>Clin Neurol Neurosurg</i> 2006; 108(4):415-417.	14	1	Case report on paraneoplastic cerebellar degeneration associated with anti-Tr (anti-Purkinje cell) antibodies.	14 months after onset of symptoms, whole body PET-scan showed a pathological focus at the right hilus of the lungs. Anatomopathological analysis revealed a non-well differentiated squamous cell carcinoma. This is first report about the association between an anti-Tr associated paraneoplastic cerebellar degeneration and squamous cell carcinoma.	4
10. Scheid R, Voltz R, Briest S, Kluge R, von Cramon DY. Clinical insights into paraneoplastic cerebellar degeneration. <i>J Neurol Neurosurg Psychiatry</i> 2006; 77(4):529-530.	14	1	Case report on a patient with proven paraneoplastic cerebellar degeneration in whom cerebellar atrophy evolved very rapidly and was present in early imaging studies.	In these cases, MRI is recommended in addition to mammography and repeat FDG-PET may be necessary.	4
11. Rees JH, Hain SF, Johnson MR, et al. The role of [18F]fluoro-2-deoxyglucose-PET scanning in the diagnosis of paraneoplastic neurological disorders. <i>Brain</i> 2001; 124(Pt 11):2223-2231.	10	43	Retrospective review of case notes of unselected patients with suspected paraneoplastic neurological disorder referred for FDG-PET to determine value of this technique when conventional imaging is negative.	FDG-PET is useful in the detection of small tumors in patients with paraneoplastic neurological disorders.	3
12. Ge Y. Multiple sclerosis: the role of MR imaging. <i>AJNR Am J Neuroradiol</i> 2006; 27(6):1165-1176.	12	N/A	Review MRI use, techniques and findings in acute and chronic multiple sclerosis.	MRI is sensitive for detecting multiple sclerosis lesions and has proved to be useful for diagnosing multiple sclerosis and monitoring therapeutic trials.	4
13. Patel S, Barkovich AJ. Analysis and classification of cerebellar malformations. <i>AJNR Am J Neuroradiol</i> 2002; 23(7):1074-1087.	15	70	Retrospective review of MRI to provide a description and classification of cerebellar malformations.	Classification system helps in the segregation and understanding of the relationship among cerebellar malformations.	3
14. Boltshauser E. Cerebellum-small brain but large confusion: a review of selected cerebellar malformations and disruptions. <i>Am J Med Genet A</i> 2004; 126(4):376-385.	12	N/A	Review congenital disorders of the cerebellum.	No results stated.	4
15. Alorainy IA, Sabir S, Seidahmed MZ, Farooqu HA, Salih MA. Brain stem and cerebellar findings in Joubert syndrome. <i>J Comput Assist Tomogr</i> 2006; 30(1):116-121.	12	N/A	Illustrate the brainstem and cerebellar findings in Joubert syndrome.	Awareness of clinical and neuroimaging findings in Joubert syndrome and maintenance of a high index of suspicion are important in diagnosis.	4

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16. Valente EM, Marsh SE, Castori M, et al. Distinguishing the four genetic causes of Jouberts syndrome-related disorders. <i>Ann Neurol</i> 2005; 57(4):513-519.	15	N/A	Distinguish four genetic causes of Joubert syndrome. Their various brainstems, cerebellar and multi-organ involvement is described.	JBTS1 and -3 show features restricted to the central nervous system, with JBTS1 showing largely pure cerebellar and midbrain-hindbrain junction involvement, and JBTS3 displaying cerebellar, midbrain-hindbrain junction, and cerebral cortical features, most notably polymicrogyria. Conversely, JBTS2 is associated with multiorgan involvement of kidney, retina, and liver, in addition to the central nervous system features, and results in extreme phenotypic variability.	3
17. Mercuri E, He J, Curati WL, Dubowitz LM, Cowan FM, Bydder GM. Cerebellar infarction and atrophy in infants and children with a history of premature birth. <i>Pediatr Radiol</i> 1997; 27(2):139-143.	13	10	Describe cerebellar alteration associated with premature birth and cerebellar vascular insult. MRI findings were reviewed to determine nature and frequency of lesions of the cerebellum and results were correlated with clinical data.	Cerebellar infarction is not uncommon in premature infants with perinatal hemorrhage or hypoxic/ischemic injury. Focal infarction and cerebellar atrophy were the sequella demonstrated with MRI.	3
18. Himmelmann K, Hagberg G, Beckung E, Hagberg B, Uvebrant P. The changing panorama of cerebral palsy in Sweden. IX. Prevalence and origin in the birth-year period 1995-1998. <i>Acta Paediatr</i> 2005; 94(3):287-294.	15 (population-based study)	170	A study on the prevalence and origin of cerebral palsy in Sweden.	Ataxic cerebral palsy was demonstrated in 6% of the cohort. Prenatal insults were responsible for all cases of ataxic cerebral palsy when a cause could be identified.	3
19. Berciano J, Boesch S, Perez-Ramos JM, Wenning GK. Olivopontocerebellar atrophy: Toward a better nosological definition. <i>Mov Disord</i> 2006.	12	N/A	Review nosological definition of olivopontocerebellar atrophy (OPCA), multiple-system atrophy and idiopathic late onset cerebellar ataxia (ILOCA).	OPCA is a pathological label to describe cases that present with a clinical cerebellar-plus syndrome. It is the pathology of ILOCA, 25% of which evolve into multiple-system atrophy.	4
20. Kerber KA, Jen JC, Perlman S, Baloh RW. Late-onset pure cerebellar ataxia: differentiating those with and without identifiable mutations. <i>J Neurol Sci</i> 2005; 238(1-2):41-45.	13	38	Review clinical findings and quantitative oculomotor tests of patients with late onset pure cerebellar ataxia.	Genetic analysis uncovered a mutation in 11 patients. The SCA6 mutation was present in 8 patients. Patients without identified genetic mutations were characterized by: 1) a later age of onset, 2) truncal without extremity ataxia, 3) and down beat nystagmus. Although only a third of these idiopathic late onset ataxia patients had a positive family history, this homogeneous syndrome probably represents a yet to be identified genetic disorder.	3
21. Ormerod IE, Harding AE, Miller DH, et al. Magnetic resonance imaging in degenerative ataxic disorders. <i>J Neurol Neurosurg Psychiatry</i> 1994; 57(1):51-57.	13	53 patients with degenerative ataxias 96 controls	To describe MRI findings in a mixed group of patients with multiple degenerative ataxias.	Findings included cerebellar atrophy with or without brainstem atrophy, cerebral atrophy in late onset ataxias, and white matter lesions that were more prevalent than in controls.	2

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22. Ikeuchi T, Koide R, Tanaka H, et al. Dentatorubral-pallidoluysian atrophy: clinical features are closely related to unstable expansions of trinucleotide (CAG) repeat. <i>Ann Neurol</i> 1995; 37(6):769-775.	13	65	To examine how the degree of expansion of the trinucleotide (CAG) repeat effects the clinical manifestations of dentatorubral-pallidoluysian atrophy.	Variability of clinical expression correlated with age at onset and with the length of CAG repeat expansion.	2
23. Koide R, Onodera O, Ikeuchi T, et al. Atrophy of the cerebellum and brainstem in dentatorubral pallidoluysian atrophy. Influence of CAG repeat size on MRI findings. <i>Neurology</i> 1997; 49(6):1605-1612.	13	26	Correlate CAG repeat length with imaging findings in subjects with dentatorubral pallidoluysian atrophy.	Atrophy of the brainstem and cerebellum, and increased white matter T2 signal intensity correlated with patient age and CAG repeat length.	3
24. Bhidayasiri R, Perlman SL, Pulst SM, Geschwind DH. Late-onset Friedreich ataxia: phenotypic analysis, magnetic resonance imaging findings, and review of the literature. <i>Arch Neurol</i> 2005; 62(12):1865-1869.	13	13	Describe the clinical and imaging findings of late onset Friedreich ataxia.	In contrast to imaging findings associated with the usual presentation of Friedreich ataxia, cerebellar and vermis atrophy is common in late onset Friedreich ataxia.	3
25. Filla A, De Michele G, Coppola G, et al. Accuracy of clinical diagnostic criteria for Friedreich's ataxia. <i>Mov Disord</i> 2000; 15(6):1255-1258.	9	142	To examine the accuracy of clinical diagnostic criteria for Friedreich's ataxia in patients with progressive unremitting ataxia of autosomal recessive inheritance or sporadic occurrence.	Authors suggest 3 levels of diagnostic certainty: <ul style="list-style-type: none"> <li>• Possible Friedreich's ataxia, defined as sporadic or recessive progressive ataxia with: a) lower limb areflexia and dysarthria, Babinski sign, or electrocardiographic repolarization abnormalities, or b) with lower limb retained reflexes and electrocardiographic repolarization abnormalities (95% sensitivity and 88% PPV);</li> <li>• Probable Friedreich's ataxia as defined by Harding's criteria (63% sensitivity and 96% PPV) or by Quebec Cooperative Study on Friedreich's Ataxia criteria (63% sensitivity and 98% PPV);</li> <li>• Definite diagnosis, molecularly confirmed.</li> </ul>	2
26. Tavani F, Zimmerman RA, Berry GT, Sullivan K, Gatti R, Bingham P. Ataxia-telangiectasia: the pattern of cerebellar atrophy on MRI. <i>Neuroradiology</i> 2003; 45(5):315-319.	13	19	Describe MRI finding and correlate findings with neurologic deficit in patients with patients with ataxia-telangiectasia.	Lateral cerebellar and vermis atrophy occurred by age 3-7 years. It progressed to severe volume loss by late teen age years.	3

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27. Butch AW, Chun HH, Nahas SA, Gatti RA. Immunoassay to measure ataxia-telangiectasia mutated protein in cellular lysates. <i>Clin Chem</i> 2004; 50(12):2302-2308.	10	21 ataxia telangiectasia patients 8 carriers and 22 controls	Report a rapid laboratory technique for the diagnosis of ataxia telangiectasia. Use of the immunoassay for ataxia-telangiectasia mutated protein is discussed.	Ataxia-telangiectasia mutated protein immunoassay can be used to confirm a diagnosis of ataxia telangiectasia in 2-days on small numbers of peripheral blood mononuclear cells and can potentially identify ataxia telangiectasia carriers and individuals at increased risk for cancer.	2
28. Verkerk AJ, Pieretti M, Sutcliffe JS, et al. Identification of a gene (FMR-1) containing a CGG repeat coincident with a breakpoint cluster region exhibiting length variation in fragile X syndrome. <i>Cell</i> 1991; 65(5):905-914.	15	N/A	Identify the gene and CGG repeat associated with Fragile X syndrome. The FMR-1 gene and abnormal CGG repeat associated with fragile X syndrome is described.	Localization of brain-expressed FMR-1 gene to EcoRI fragment suggests the involvement of this gene in the phenotypic expression of the fragile X syndrome.	4
29. Hagerman RJ, Leehy M, Heinrichs W, et al. Intention tremor, parkinsonism, and generalized brain atrophy in male carriers of fragile X. <i>Neurology</i> 2001; 57(1):127-130.	14	5	Report the fragile X permutation syndrome in elderly men who had a progressive action tremor associated with executive function deficits and generalized brain atrophy.	Authors propose that elevations of FMR-1 messenger RNA may be causative for a neurodegenerative syndrome in a subgroup of elderly men with the FMR-1 premutation.	4
30. Hagerman PJ, Hagerman RJ. The fragile-X premutation: a maturing perspective. <i>Am J Hum Genet</i> 2004; 74(5):805-816.	12	N/A	Review genetic, clinical and laboratory alterations associated with fragile X associated tremor/ataxia syndrome.	Carriers of 50-200 CGG repeats on the FMR-1 gene present with cognitive alteration, ataxia, tremor, parkinsonism and autonomic dysfunction.	4
31. Grigsby J, Brega AG, Jacquemont S, et al. Impairment in the cognitive functioning of men with fragile X-associated tremor/ataxia syndrome (FXTAS). <i>J Neurol Sci</i> 2006.	3b	25	To examine circumscribed aspects of cognitive functioning in men with fragile X associated tremor/ataxia syndrome.	Capacity for inhibition was severely affected in one-quarter of sample; information processing speed was impaired in most subjects. Although mean verbal and performance IQ scores were not significantly different from the general population, they were quite low given the sample's educational level. Cognitive and functional impairment was greater for men with more CGG repeats, although number of repeats was not associated with age of onset of either tremor or ataxia. Results provide evidence that fragile X associated tremor/ataxia syndrome involves marked impairment of executive cognitive abilities.	3

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32. Brunberg JA, Jacquemont S, Hagerman RJ, et al. Fragile X premutation carriers: characteristic MR imaging findings of adult male patients with progressive cerebellar and cognitive dysfunction. <i>AJNR Am J Neuroradiol</i> 2002; 23(10):1757-1766.	13	17	To characterize MRI findings of the brain of adult male fragile X premutation carriers with a recently identified disorder characterized by ataxia, tremor, rigidity, and cognitive dysfunction.	MRI findings in symptomatic male fragile X premutation carriers are characteristic of this disorder. Recognition of these alterations may support a specific diagnosis and may have implications for the potential occurrence of fragile X syndrome in the children of reproductive age female relatives.	3
33. Geser F, Wenning GK. The diagnosis of multiple system atrophy. <i>J Neurol</i> 2006; 253 Suppl 3:iii2-iii15.	12	N/A	Describe the clinical and laboratory diagnosis of multiple system atrophy.	MRI shows brainstem, cerebellar and putamen atrophy. There is increased T2 signal intensity in the middle cerebellar peduncles, and low T2 signal intensity in the putamen with a lateral plane of increased T2 signal intensity.	4
34. Quinn NP. How to diagnose multiple system atrophy. <i>Mov Disord</i> 2005; 20 Suppl 12:S5-S10.	12	N/A	Review the clinical multiple system atrophy diagnostic criteria.	MRI shows brainstem, cerebellar and putamen atrophy. There is increased T2 signal intensity in the middle cerebellar peduncles and low T2 signal intensity in the putamen with a lateral plane of increased T2 signal intensity.	4
35. Arai K. MRI of progressive supranuclear palsy, corticobasal degeneration and multiple system atrophy. <i>J Neurol</i> 2006; 253 Suppl 3:iii25-iii29.	12	N/A	Review the MRI findings in multiple system atrophy, progressive supranuclear palsy and corticobasilar degeneration.	Clinical and imaging findings in multiple system atrophy relate to degeneration occurring in extrapyramidal and cerebellar systems. Pathways and anatomic alterations are reviewed.	4
36. DiMauro S, Bonilla E. Mitochondrial Encephalomyopathies. In: Rosenberg R, Prusiner S, DiMauro S, al. e, eds. <i>The Molecular and Genetic Basis of Neurological Disease</i> . Boston: Butterworth-Heinemann; 1997:201-236.	15	N/A	Book chapter.	N/A	N/A
37. Rossi A, Biancheri R, Bruno C, et al. Leigh Syndrome with COX deficiency and SURF1 gene mutations: MR imaging findings. <i>AJNR Am J Neuroradiol</i> 2003; 24(6):1188-1191.	14	3	Report the MRI findings associated with Leigh Syndrome.	Regions of increased T2 signal intensity in the substantia nigra, subthalamic nuclei, central tegmental tract, medulla, pons, dentate nuclei and inferior cerebellar peduncles. The putamen was variably involved. Imaging finds are characteristic but not specific for a given genetic defect.	4
38. Lamperti C, Naini A, Hirano M, et al. Cerebellar ataxia and coenzyme Q10 deficiency. <i>Neurology</i> 2003; 60(7):1206-1208.	13	135	Measured coenzyme Q10 (CoQ10) concentration in muscle biopsies from patients with genetically undefined cerebellar ataxia. 13 patients with childhood-onset ataxia and cerebellar atrophy had decreased levels of CoQ10.	Associated symptoms included seizures, developmental delay, mental retardation, and pyramidal signs. These findings confirm the existence of an ataxic presentation of CoQ10 deficiency, which may be responsive to CoQ10 supplementation.	3

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39. van der Knaap MS, Pronk JC, Scheper GC. Vanishing white matter disease. <i>Lancet Neurol</i> 2006; 5(5):413-423.	12	N/A	Review the topic of vanishing white matter disease.	Childhood progressive ataxia and encephalopathy with diffuse increased T2 signal intensity and volume loss of white matter is diagnostic.	4
40. Kumar N, Cohen-Gadol AA, Wright RA, Miller GM, Piegras DG, Ahlskog JE. Superficial siderosis. <i>Neurology</i> 2006; 66(8):1144-1152.	13	30	Review the clinical and imaging features of superficial siderosis.	Hearing loss and slowly progressive ataxia were the most common presenting symptoms. Cerebellar atrophy and low T2 superficial signal intensity were typical.	3
41. Bassi SS, Bulundwe KK, Greeff GP, Labuscagne JH, Gledhill RF. MRI of the spinal cord in myelopathy complicating vitamin B12 deficiency: two additional cases and a review of the literature. <i>Neuroradiology</i> 1999; 41(4):271-274.	14	2	Report two cases of MRI of the spinal cord in myelopathy complicating vitamin B12 deficiency and review the literature.	MRI of early B12 related myelopathy can show cord swelling and increased T2 signal intensity with or without enhancement following contrast administration. There may be late atrophy or findings may resolve with treatment. The cord may also appear normal, even though symptomatic.	4
42. Facchini SA, Jami MM, Neuberg RW, Sorrel AD. A treatable cause of ataxia in children. <i>Pediatr Neurol</i> 2001; 24(2):135-138.	14	1	Describe and illustrate the imaging findings associated with B12 deficiency. MRI is illustrated, findings are discussed, and the literature is reviewed.	Spinal MRI demonstrated extensive demyelination of the posterior columns along the entire length of the cord, and areas of contrast enhancement.	4
43. Inoue N, Ichimura H, Goto S, Hashimoto Y, Ushio Y. MR imaging findings of spinal posterior column involvement in a case of Miller Fisher syndrome. <i>AJNR Am J Neuroradiol</i> 2004; 25(4):645-648.	14	1	Describe the MRI findings in Miller-Fisher syndrome.	The significant MRI was initial abnormal enhancement of the lower spinal nerves, with cerebellum normal. Five months later there was increased T2 signal intensity in the posterior column of the spinal cord at C1-T12.	4
44. Borne J, Riascos R, Cuellar H, Vargas D, Rojas R. Neuroimaging in drug and substance abuse part II: opioids and solvents. <i>Top Magn Reson Imaging</i> 2005; 16(3):239-245.	12	N/A	To review the imaging finding associated with opiate and solvent abuse.	Imaging alterations are mediated by vascular, infectious, cytotoxic and demyelinating mechanisms. Cerebellar atrophy and infarction are frequent.	4
45. Korogi Y, Takahashi M, Okajima T, Eto K. MR findings of Minamata disease--organic mercury poisoning. <i>J Magn Reson Imaging</i> 1998; 8(2):308-316.	12	N/A	Describe the MRI findings of Minamata disease.	There is prominent atrophy of the visual cortex, cerebellar vermis and cerebral cortex, though most prominently in pre and post central cortex. There is increased T2 signal intensity in occipital cortex.	4
46. Abbaslou P, Zaman T. A Child with elemental mercury poisoning and unusual brain MRI findings. <i>Clin Toxicol (Phila)</i> 2006; 44(1):85-88.	14	1	Describe unusual brain MRI findings in a child with mercury vapor poisoning.	Multiple regions of high T2 signal intensity were demonstrated in cerebral white matter, left globus pallidus and putamen.	4

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47. Heaney CJ, Campeau NG, Lindell EP. MR imaging and diffusion-weighted imaging changes in metronidazole (Flagyl)-induced cerebellar toxicity. <i>AJNR Am J Neuroradiol</i> 2003; 24(8):1615-1617.	14	1	Describe MR imaging changes associated with metranidazole toxicity.	MRI included increased T2 signal intensity in the dentate nuclei with associated restricted diffusion. Follow-up imaging 8 weeks after cessation of metronidazole therapy showed resolution of imaging findings, including diffusion changes.	4
48. Spampinato MV, Castillo M, Rojas R, Palacios E, Frascheri L, Descartes F. Magnetic resonance imaging findings in substance abuse: alcohol and alcoholism and syndromes associated with alcohol abuse. <i>Top Magn Reson Imaging</i> 2005; 16(3):223-230.	12	N/A	Review imaging alterations associated with chronic ethanol abuse. Mechanisms of brain toxicity and imaging findings associated with Wernicke encephalopathy, fetal alcohol syndrome, Marchiafava-Bignami disease, chronic hepatic encephalopathy, osmotic demyelization syndrome, and methanol toxicity are reviewed and illustrated.	No results stated.	4
49. Vorgerd M, Tegenthoff M, Kuhne D, Malin JP. Spinal MRI in progressive myeloneuropathy associated with vitamin E deficiency. <i>Neuroradiology</i> 1996; 38 Suppl 1:S111-113.	14	1	Describe spinal MRI in progressive myeloneuropathy associated with vitamin E deficiency.	MRI of the cervical spine demonstrated high-signal lesions on T2-weighted images in the posterior columns, correlating with the clinical findings.	4
50. Battisti C, Toffola ED, Verri AP, et al. Clinical and stabilometric monitoring in a case of cerebellar atrophy with vitamin E deficiency. <i>Brain Dev</i> 1998; 20(4):253-257.	14	1	Describe MRI alterations associated with vitamin E deficiency, ataxia and cerebellar atrophy.	MRI demonstrated diffuse cerebellar atrophy.	4
51. Stott VL, Hurrell MA, Anderson TJ. Reversible posterior leukoencephalopathy syndrome: a misnomer reviewed. <i>Intern Med J</i> 2005; 35(2):83-90.	14	6	Illustrate and review the syndrome of posterior leukoencephalopathy.	Increased T2 and fluid attenuated inversion recovery signal in posterior regions of the cerebral hemispheres is compatible with the multiple causes, each of which is reviewed.	4
52. Kitaguchi H, Tomimoto H, Miki Y, et al. A brainstem variant of reversible posterior leukoencephalopathy syndrome. <i>Neuroradiology</i> 2005; 47(9):652-656.	14	2	Present two patients with reversible brainstem encephalopathy: one with hypertension and the other without hypertension.	Pontine swelling and increased signal intensity is demonstrated on FLAIR and T2-weighted imaging. Reversible brainstem encephalopathy with characteristic MRI features was found in both hypertensive and non-hypertensive patients.	4
53. Ugurel MS, Hayakawa M. Implications of post-gadolinium MRI results in 13 cases with posterior reversible encephalopathy syndrome. <i>Eur J Radiol</i> 2005; 53(3):441-449.	13	13	Retrospective review of MRI findings and clinical data to evaluate contrast enhanced MRI in reversible posterior leukoencephalopathy.	Distinct contrast enhancement is not evident in the majority of cases with reversible posterior leukoencephalopathy.	3

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54. Kataoka S, Hori A, Shirakawa T, Hirose G. Paramedian pontine infarction. Neurological/topographical correlation. <i>Stroke</i> 1997; 28(4):809-815.	13	49	Analyzed clinical signs and their association with MRI findings to describe clinical correlates of paramedian pontine infarction.	A faciobrachial dominant hemiparesis with dysarthria, somatosensory alteration and horizontal gaze abnormality are most common. Ataxia is not a frequent component.	2
55. Katoh M, Kawamoto T. Bilateral medial medullary infarction. <i>J Clin Neurosci</i> 2000; 7(6):543-545.	13	13 cases	Present a case of bilateral medial medullary infarction demonstrated by MRI and review 12 previously reported cases.	MRI showed upper medial medullary infarction bilaterally that extended to the pontomedullary junction. Authors propose that the prognosis of type 2 bilateral medial medullary infarction is better than that of type 1.	3
56. Kim JS, Kim J. Pure midbrain infarction: clinical, radiologic, and pathophysiologic findings. <i>Neurology</i> 2005; 64(7):1227-1232.	13	40	Report the MR findings and pathogenesis of pure midbrain infarction.	Clinical-radiologic correlation study yields 4 distinct subgroups: anteromedial, anterolateral, combined, and lateral. Large vessel disease and small vessel disease are usual pathogenic mechanisms, whereas cardiogenic embolism is rare.	3
57. Kim JS, Lee JH, Im JH, Lee MC. Syndromes of pontine base infarction. A clinical-radiological correlation study. <i>Stroke</i> 1995; 26(6):950-955.	13	37	Report the clinical-imaging correlation of lacunar infarction involving the base of the pons.	Study suggested large lesions involving the paramedian caudal or middle pons correlate with severe hemiparesis, whereas lesions of similar size located in the paramedian rostral pons tended to produce dysarthria-clumsy hand syndrome. In pontine lacunar infarction various manifestations of ataxia are frequent, but they are less common than sensory-motor alteration.	3
58. Luijckx GJ, Boiten J, Lodder J, Heuts-van Raak L, Wilmink J. Isolated hemiataxia after supratentorial brain infarction. <i>J Neurol Neurosurg Psychiatry</i> 1994; 57(6):742-744.	14	3	Describe hemiataxia in association with infarction involving the posterior limb of the internal capsule.	Hemiataxia can occur in association with isolated supratentorial infarction involving the posterior limb of the internal capsule.	4
59. Melo TP, Bogousslavsky J. Thalamic ataxia syndrome. <i>Neurology</i> 1995; 45(3 Pt 1):598-599.	15 (letter to editor)	17	Describe ataxia with thalamic infarction.	Infarction of the thalamus is frequently associated with ataxia.	4
60. Mossuto-Agatiello L. Caudal paramedian midbrain syndrome. <i>Neurology</i> 2006; 66(11):1668-1671.	14	5	Examine patients with MRI evidence of unilateral paramedian caudal midbrain infarction to define clinical and radiologic picture.	3 cases had ocular movement abnormalities. A constant MRI finding was bilateral inferior olivary degeneration, but only one patient displayed a delayed palatal tremor. A single strategically placed unilateral lesion can cause bilateral dysfunction. A bilateral cerebellar syndrome can occur with unilateral lesions in the lower midbrain with a wide range of other clinical features.	4

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Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Strength of Evidence
61. Cormier PJ, Long ER, Russell EJ. MR imaging of posterior fossa infarctions: vascular territories and clinical correlates. <i>Radiographics</i> 1992; 12(6):1079-1096.	13	18 cases	Retrospective study to describe MR findings associated with posterior fossa infarction.	The vascular supply and MRI findings of posterior fossa infarctions are grouped by anatomy.	3
62. Krespi Y, Gurol ME, Coban O, Tuncay R, Bahar S. Venous infarction of brainstem and cerebellum. <i>J Neuroimaging</i> 2001; 11(4):425-431.	14	2	Describe clinical and imaging findings of brainstem and cerebellar infarction.	Venous infarctions are often hemorrhagic and they are not in an arterial territory of distribution. Clinical and neuroimaging features can lead to earlier diagnosis and, potentially, more effective management.	4
63. Caplan LR, Biouesse V. Cervicocranial arterial dissections. <i>J Neuroophthalmol</i> 2004; 24(4):299-305.	12	N/A	Review the topic of cervicocranial arterial dissections. Diagnosis and management is reviewed.	Common manifestations are pain and neuro-ophthalmic symptoms and signs.	4
64. Shah GV, Quint DJ, Trobe JD. Magnetic resonance imaging of suspected cervicocranial arterial dissections. <i>J Neuroophthalmol</i> 2004; 24(4):315-318.	14	1	MRI of suspected cervicocranial arterial dissection is reviewed.	A protocol for combined MRA and MRI, using fat suppressed sequences is proposed.	4
65. Tay KY, JM UK-I, Trivedi RA, et al. Imaging the vertebral artery. <i>Eur Radiol</i> 2005; 15(7):1329-1343.	12	N/A	Review the current state of vertebral artery imaging. Techniques for imaging of vertebral artery dissection, using CTA, catheter based angiography and MRI are discussed.	CTA may have a sensitivity of 100% and specificity of 98%.	3
66. Browne DL, Gancher ST, Nutt JG, et al. Episodic ataxia/myokymia syndrome is associated with point mutations in the human potassium channel gene, KCNA1. <i>Nat Genet</i> 1994; 8(2):136-140.	12	N/A	Identify the genetic alteration associated with episodic ataxia.	Episodic ataxia and myokymia is associated with mutation of the voltage gated K+ channel gene KCNA1.	4
67. Jen J, Kim GW, Baloh RW. Clinical spectrum of episodic ataxia type 2. <i>Neurology</i> 2004; 62(1):17-22.	13	27	Describe the findings and mutations associated with episodic ataxia type 2.	Alterations in neuronal CA channels, heavily expressed in the cerebellum, are associated with episodic ataxia, occasional progressive ataxia, episodic hemiplegia and migraine.	3
68. Jen JC, Wan J, Palos TP, Howard BD, Baloh RW. Mutation in the glutamate transporter EAAT1 causes episodic ataxia, hemiplegia, and seizures. <i>Neurology</i> 2005; 65(4):529-534.	14	1	Describe the excitatory amino acid transporter (EAAT)1 mutation and associated episodic ataxia, hemiplegia and seizures.	Mutation in the glutamate EAAT can contribute to neuronal hyper-excitability, and thereby cause seizures, hemiplegia and episodic ataxia.	4
69. Garg M, Gupta RK, Husain M, et al. Brain abscesses: etiologic categorization with in vivo proton MR spectroscopy. <i>Radiology</i> 2004; 230(2):519-527.	13	75	To use proton MR spectroscopy to categorize the etiologic agent causing brain abscess. MRI and in vivo single-voxel proton MR spectroscopic data obtained from patients with brain abscesses were retrospectively analyzed.	Proton MR spectroscopy demonstrates the absence of acetate and succinate in abscess due to obligate aerobes or facultative anaerobes. Acetate and succinate are present with obligate anaerobes alone or in combination with facultative anaerobes.	2

**Ataxia**  
**EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Strength of Evidence
70. Jaggi RS, Husain M, Chawla S, Gupta A, Gupta RK. Diagnosis of bacterial cerebellitis: diffusion imaging and proton magnetic resonance spectroscopy. <i>Pediatr Neurol</i> 2005; 32(1):72-74.	14	1	Describe diffusion imaging and proton spectroscopy in brain abscess and briefly review the literature.	Diffusion is restricted in abscess, and spectroscopy in the presence of obligate anaerobes demonstrates elevated lactate, acetate and succinate.	4
71. Kato Z, Kozawa R, Teramoto T, Hashimoto K, Shinoda S, Kondo N. Acute cerebellitis in primary human herpesvirus-6 infection. <i>Eur J Pediatr</i> 2003; 162(11):801-803.	14	1	Describe cerebellitis due to herpesvirus-6.	Herpes virus can cause an isolated cerebellitis. Imaging findings consisted of increased T2 signal intensity and restricted diffusion in the cerebellum.	4
72. Mendonca RA, Martins G, Lugokenski R, Rossi MD. Subacute spongiform encephalopathies. <i>Top Magn Reson Imaging</i> 2005; 16(2):213-219.	12	N/A	Describe the clinical course, pathology and imaging of the subacute spongiform encephalopathies.	Diffusion imaging is the most sensitive initial sequence, while MRI has an overall sensitivity of 91%, specificity of 95% and accuracy of 94%.	4
73. De Bruecker Y, Claus F, Demaerel P, et al. MRI findings in acute cerebellitis. <i>Eur Radiol</i> 2004; 14(8):1478-1483.	14	4	Describe the clinical, CT and MRI finding in four cases and review existing literature.	Most common imaging finding was bilateral diffuse hemispheric abnormalities. The development of cerebellar atrophy following an initial normal MRI is a new finding. In atypical clinical presentation, MRI can lead to the diagnosis. MRI findings have, however, no prognostic value.	4
74. de Ribaupierre S, Meagher-Villemure K, Villemure JG, et al. The role of posterior fossa decompression in acute cerebellitis. <i>Childs Nerv Syst</i> 2005; 21(11):970-974.	5	2	Discuss the role of posterior fossa decompression in the management of acute cerebellitis.	External ventricular drainage and posterior fossa decompression may limit secondary brainstem or cerebellar lesions.	4
75. Adachi M, Kawanami T, Ohshima H, Hosoya T. Cerebellar atrophy attributed to cerebellitis in two patients. <i>Magn Reson Med Sci</i> 2005; 4(2):103-107.	14	2	Describe the late findings of cerebellitis in two rare patients considered to be in late-stage cerebellitis.	Findings included isolated cerebellar atrophy and slightly increased cortical signal intensity on FLAIR images.	4
76. Mondejar RR, Santos JM, Villalba EF. MRI findings in a remitting-relapsing case of Bickerstaff encephalitis. <i>Neuroradiology</i> 2002; 44(5):411-414.	14	1	Describe the imaging findings in one case of remitting-relapsing Bickerstaff encephalitis and their significance when a clinical differentiation between Bickerstaff encephalitis and Miller-Fisher syndrome is attempted.	Signs and symptoms may occasionally overlap. However, because Miller-Fisher syndrome is related to the peripheral nervous system and Bickerstaff encephalitis is a central disease, the recognition of brain stem hypointense lesions on T1-weighted images, which are hyperintense on T2-weighted sequences, could be a reliable tool when the clinical diagnosis is unclear.	4

**Ataxia  
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Strength of Evidence
77. Weidauer S, Ziemann U, Thomalske C, Gaa J, Lanfermann H, Zanella FE. Vasogenic edema in Bickerstaff's brainstem encephalitis: a serial MRI study. <i>Neurology</i> 2003; 61(6):836-838.	14	1	Describe the MRI findings of a patient with Bickerstaff's brainstem encephalitis, disclosing caudal migration of an initial upper midbrain lesion and review literatures.	High apparent diffusion coefficient values imply a vasogenic rather than cytotoxic edema as the cause of the hyperintense signal changes on T2-weighted images.	4
78. Suzuki K, Meguro K, Nakayama J, Aoki T, Tsurushima H. MRI of an infant with Fisher syndrome. <i>Childs Nerv Syst</i> 1997; 13(2):95-96.	14	1	Describe the MRI findings of an infant with Miller-Fisher syndrome.	The significant MR finding was increased T2 signal intensity lesion in the left cerebellar hemisphere and left middle cerebellar peduncle.	4
79. Garcia-Rivera CA, Rozen TD, Zhou D, et al. Miller Fisher syndrome: MRI findings. <i>Neurology</i> 2001; 57(10):1755.	14	1	Describe the imaging findings in Miller-Fisher syndrome.	The significant MR finding was abnormal enhancement of the lower cranial nerves.	4
80. Terry JB, Rosenberg RN. Frontal lobe ataxia. <i>Surg Neurol</i> 1995; 44(6):583-588.	14	1	Case report on a patient with large bilateral, medial-orbital, frontal lobe lesion who manifested gait impairment and dysarthria. Lesion is defined by MRI and PET.	Disruption of the frontopontocerebellar pathway, originating from Brodman's area 10 in the frontal cortex, is the likely mechanism for frontal ataxia.	4
81. American College of Radiology. <i>Manual on Contrast Media</i> . Available at: <a href="http://www.acr.org/SecondaryMainMenuCategories/quality_safety/contrast_manual.aspx">http://www.acr.org/SecondaryMainMenuCategories/quality_safety/contrast_manual.aspx</a> .	15	N/A	Guidance document on contrast media to assist radiologists in recognizing and managing risks associated with the use of contrast media.	N/A	3

## Evidence Table Key

### Study Type Key

*Numbers 1-7 are for studies of therapies while numbers 8-15 are used to describe studies of diagnostics.*

1. Randomized Controlled Trial — Treatment
2. Controlled Trial
3. Observation Study
  - a. Cohort
  - b. Cross-sectional
  - c. Case-control
4. Clinical Series
5. Case reviews
6. Anecdotes
7. Reviews
8. Randomized Controlled Trial — Diagnostic
9. Comparative Assessment
10. Clinical Assessment
11. Quantitative Review
12. Qualitative Review
13. Descriptive Study
14. Case Report
15. Other (Described in text)

### Strength of Evidence Key

- Category 1 - The conclusions of the study are valid and strongly supported by study design, analysis and results.
- Category 2 - The conclusions of the study are likely valid, but study design does not permit certainty.
- Category 3 - The conclusions of the study may be valid but the evidence supporting the conclusions is inconclusive or equivocal.
- Category 4 - The conclusions of the study may not be valid because the evidence may not be reliable given the study design or analysis.