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for the Expert Panel
on Neurologic
Imaging

Dementia and Movement Disorders

Dementia (Table 1)

Dementia is significant loss of cognitive function not due to impaired arousal affecting about 7% of those over 65, and 30% over 80. Delirium, focal brain lesions, and psychiatric problems must be excluded. Accurate diagnosis is important because therapy can delay progression.

Alzheimer disease (AD) causes 50%–80% of dementias. The National Institute of Neurologic and Communicative Disorders and Stroke (NINCDS) and the Alzheimer Disease and Related Disorders Association (ADRDA) established criteria for definite, probable, and possible AD.¹ Exclusion of other causes of dementia with imaging is required. MR imaging is preferable to CT due to greater sensitivity.² Mild Cognitive Impairment (MCI) is defined as cognitive decline greater than expected for an individual's age, but does not interfere notably with daily life.³ More than half of MCI patients progress to dementia within 5 years. The amnesic subtype may be prodromal AD.³ Direct diagnosis of AD by imaging is difficult. Positron emission tomography (PET) discriminates AD patients from normals.⁴ MR imaging hippocampal volumes are significantly smaller in mild AD than controls and other dementias, correlating with focal neuropathologic atrophy.⁵ SPECT imaging cannot be recommended for either initial or differential diagnosis of dementia.⁶ MR spectroscopy may permit identification of mild to moderate AD.⁷ In probable AD, PET or MR imaging increase diagnostic accuracy from 80%–85% to 90%–100%. In possible AD or atypical dementias, imaging studies permit more accurate diagnosis. A promising technique for AD is PET A β brain amyloid imaging.⁸

Frontotemporal dementia (FTD), rare after 75, includes sporadic and familial disorders⁹ causing behavioral or cognitive deficits with early progressive personality, behavior or language change. MR imaging may show atrophy of the anterior temporal and frontal lobes. PET shows metabolic disturbance in the frontal and temporal lobes.¹⁰ SPECT shows frontal hypoperfusion.¹¹

Dementia with Lewy bodies (DLB) diagnosis is useful due to the rapidly progressive course, risks of neuroleptics, and response to cholinesterase inhibitors. Features include prominent memory, attention, executive function, and visuospatial deficits, fluctuating cognition, visual hallucinations, and Par-

kinsonism. SPECT dopamine transporter striatal activity is normal in AD and low in DLB.¹² SPECT occipital hypoperfusion and occipital PET hypometabolism are seen in DLB. MR imaging shows preserved hippocampal and medial temporal volume and atrophy of the putamen.¹³

Vascular dementia (VaD), usually results from small-vessel disease. VaD may be prevented or arrested by preventing infarction. Cognition may improve, suggesting that some symptoms are caused by physiologic changes without infarct. Radiologic tests that distinguish VaD from other dementias are beneficial. Sudden onset of dysfunction, stepwise deteriorating course, focal neurologic signs, stroke risk factors, systemic vascular disease, and prior strokes suggest VaD. The NINDS-AIREN criteria for VaD include imaging findings of multiple large-vessel infarcts; single strategically placed infarct; multiple basal ganglia and white matter lacunes, or extensive periventricular white matter lesions or a combination. MR imaging is preferred for detecting vascular lesions.¹⁴ Differentiation of VaD from AD and VaD is difficult. When VaD is diagnosed, this pathologic diagnosis alone is confirmed in about 25% of cases; more commonly, a mixed disorder with neuropathologic changes of both AD and VaD is found. Vascular lesions on MR or CT favor VaD over AD.

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is a hereditary small-artery vasculopathy characterized by migraine with aura, strokes and progressive subcortical dementia. MR imaging shows hyperintense T2 or FLAIR lesions predominantly in the frontal, parietal, and anterior temporal cortex, and in the external capsule.¹⁵ Lesion load increases with age. Diagnosis is confirmed by skin biopsy pathogenic notch3 mutation.

Creutzfeldt-Jakob disease (CJD) is a fatal prion disorder. Sporadic CJD (sCJD) occurs between ages 50 and 75 and variant CJD (vCJD) caused by contaminated beef occurs at average age 25–30 years. Definite diagnosis of CJD is based on histopathological findings, though biopsy is rarely performed. CJD produces rapidly progressive dementia with myoclonus, characteristic EEG, and 14–3–3 proteins in the CSF. The most common MR imaging abnormality is hyperintense signal intensity on T2WI in the basal ganglia, and less often in the cortex. MR imaging improves diagnosis of CJD.^{16,17} The most sensitive MR imaging sequences are DWI and PD.¹⁶ T2 pulvinar high signal intensity is highly suggestive of vCJD in clinical context.¹⁸ vCJD can be confirmed by tonsil biopsy.

Normal-pressure hydrocephalus (NPH) is characterized by dementia, gait disturbance, urinary incontinence and normal CSF pressure, communicating hydrocephalus on MR imaging or CT, and abnormal SPECT cisternography.^{19,20} MR imaging

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Table 1: Clinical condition: dementia and movement disorders

	MRI, Brain		CT, Head		PET, Brain	SPECT, Brain	MRS, Head	fMRI, Head
	Without Contrast	Without and With Contrast	Without Contrast	Without and With Contrast				
Probable AD	8	7	6	4	6 ^a	5 ^a	4	2 ^b
Possible AD	8	8	6	5	7 ^a	6 ^a	4	2
Suspected VaD or mixed VaD and AD ^c	8	8	6	5	6 ^a	5 ^a	2	2
FTD	8	8	6	4	7 ^a	6 ^a	4	2
DLB	8	8	6	5	7 ^a	7 ^a	3	2
Suspected prion disease (CJD, iatrogenic CJD or vCJD)	8 ^d	8 ^d	6	5	5 ^a	5 ^a	5	2
Suspected NPH ^e	8	8	6	5	N/A	N/A	3	N/A

Note:—MRI indicates MR imaging; MRS, MR spectroscopy; fMRI, functional MRI; NA, not applicable. Appropriateness criteria scale from 1 to 9; 1, least appropriate; 9, most appropriate.
^a For problem solving.
^b For research purposes.
^c MRA, head and/or neck; CT angiography, head and/or neck; and ultrasound, carotid, duplex = ratings of 6.
^d Includes DWI.
^e Cisternography = rating of 6.

Table 2: Clinical condition: dementia and movement disorders (degenerative diseases of the extrapyramidal system)

	MRI, Brain		CT, Head		PET, Brain	SPECT, Brain	MRS, Head	fMRI, Head
	Without Contrast	Without and With Contrast	Without Contrast	Without and With Contrast				
Suspected HD	8	7	5	3	5 ^a	5 ^a	3	2
Clinical features suggestive of NBIA	8	7	5	4	3 ^a	3 ^a	3	3
Parkinson's disease: typical clinical features and responds to levodopa	7 ^a	7 ^a	6	5	6 ^{a,b}	6 ^{a,c}	3	2
PD syndrome: atypical clinical features not responsive to levodopa	8	7	5	4	6 ^a	6 ^a	3	2

Note:—MRI indicates MR imaging; MRS, MR spectroscopy; fMRI, functional MRI. Appropriateness criteria scale from 1 to 9; 1, least appropriate; 9, most appropriate.
^a For problem solving.
^b Dopa PET.
^c Specific ligand.

findings include: moderate ventriculomegaly, and absent or mild cortical atrophy. CSF flow void at the cerebral aqueduct on MR imaging indicates hyperdynamic CSF.²¹

Movement Disorders (Table 2)

Huntington's disease (HD) presents with choreoathetosis, rigidity, dementia, and emotional disturbance in the 4th and 5th decades. It is autosomal dominant with complete penetrance. Genetic testing confirms HD, and identifies presymptomatic subjects. Neuroimaging and pathology show atrophy of the caudate and/or putamen.²² MR imaging shows signal intensity changes in the striatum.²³ Neuronal loss accompanied by loss of myelin and gliosis, and iron accumulation explain signal intensity abnormalities.

Neurodegeneration with brain iron accumulation (NBIA) demonstrates neurodegeneration and excessive iron deposition in the basal ganglia.²⁴ There are two types of NBIA: early onset, rapidly progressive (classic) disease and late onset, slowly progressive (atypical) disease, both with relentless progression of gait impairment, rigidity, dystonic posturing, and mental deterioration.^{25,26} PANK two mutations accompany most cases of NBIA. MR imaging shows bilateral hyperintensity within a hypointense zone in the medial globus pallidus on T2-WI ("eye of tiger" sign).²⁵

Diagnosis of idiopathic *Parkinsonism* (PD) usually based on history and physical examination alone, is confirmed by effective dopaminergic therapy. Between 2%–3% of the population is expected to develop PD, with onset between 50 and 60 years of age. Decreased width of the pars compacta on MR imaging²⁷ may indicate neuronal loss, but substantia nigra

appears normal in most PD patients.²⁸ Increased lactate occipital lobe MR spectroscopy occurs in PD.²⁹ 18F-dopa PET can detect frontal changes in PD.³⁰

Multiple System Atrophy (MSA) can present with autonomic or motor deficits. Imaging and pathology show atrophy of the striatum due to neuronal loss, putamen more than caudate. MR imaging shows putaminal T2 hypointensity, equal to or greater than pallidal hypointensity correlating with severity of rigidity.³¹ PET can differentiate PD from MSA.³² MSA exhibits "hot cross bun" sign in the pons on MR imaging.³³ Putaminal and pontine abnormalities worsen as MSA progresses.³³

Progressive supranuclear palsy (PSP) is a gradually progressive disorder, onset over age 40, with vertical supranuclear gaze palsy, slowing of vertical saccades and postural instability with falls.³⁴ Putaminal hypointensity has been described on T2WI. Some patients show slight hyperintense signal intensity at T2WI in periaqueductal gray matter. Decreased midbrain size is always observed on MR imaging in PSP patients.³⁴

Degenerative Diseases of the Motor System (Table 3)

Amyotrophic lateral sclerosis (ALS) is the most frequent motor neuron disease, annual incidence 0.4 to 1.76 per 100,000. Symptoms progress relentlessly; half of patients die within 3 years, and 90% within 6 years due to degeneration of the corticospinal tract and lower motor neurons. MR imaging demonstrates atrophy and hyperintense foci of the corticospinal tract on T2WI³⁵ due to myelin loss and gliosis. Central cortical hypointense signal intensity on T2WI may occur due to iron deposition.³⁶ The cord may be atrophic and flattened due to loss of motor neurons in the anterior horns and corticospinal

Table 3: Clinical condition: dementia and movement disorders (degenerative diseases of the motor system)

	MRI, Brain		MRI, Spine		CT, Head		PET, Brain	SPECT, Brain
	Without Contrast	Without and With Contrast	Without Contrast	Without and With Contrast	Without Contrast	Without and With Contrast		
Motor neuron disease ^a	8	7	8 ^b	7 ^b	5	4	3 ^c	3 ^c

Note:—MRI indicates MR imaging. Appropriateness criteria scale from 1 to 9; 1, least appropriate; 9, most appropriate.

^a MR spectroscopy, head = rating of 3; functional MRI, head = rating of 2.

^b May need multilevel imaging.

^c For problem solving.

tracts. Proton MR spectroscopy reveals decreased *N*-acetylaspartate in the sensorimotor cortex and brain stem.³⁷ Diffusion tensor imaging shows involvement of the corticospinal tract in early ALS.³⁸

Review Information

This guideline was originally developed in 1996. The last review and update was completed in 2007.

Appendix

Expert Panel on Neurologic Imaging: Didier Dormont, MD, Co-Author, Hôpital de la Salpêtrière, Assistance-Publique-Hôpitaux de Paris, France; David J. Seidenwurm, MD, Co-Author and Panel Chair, Radiologic Associates of Sacramento, Sacramento, Calif; Patricia C. Davis, MD; James A. Brunberg, MD; Robert Louis De La Paz, MD; David B. Hackney, MD; John E. Jordan, MD; John P. Karis, MD; Suresh Kumar Mukherji, MD; Patrick A. Turski, MD; Franz J. Wippold II, MD; Robert D. Zimmerman, MD; Michael W. McDermott, MD, American Association of Neurologic Surgeons; Michael A. Sloan, MD, MS, American Academy of Neurology.

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