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Dementia and Memory Disturbances

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MAJOR DEMENTIA SYNDROMES

Dementia is progressive loss of intellectual function coupled with loss of meaningful function in daily life. Memory loss is the central feature, and specific dementia syndromes characteristically cause particular forms of memory impairment. Dementia syndromes also produce specific abnormalities of cognition in language, spatial processing, praxis (i.e., learned motor behavior), and executive function (i.e., ability to plan and sequence events). *Cortical dementia* and *subcortical dementia,* although older terms, remain helpful for subdividing the dementias (Table 108-1).

Table 108-2 provides the differential diagnosis of neurodegenerative causes of dementia, and Table 108-3 outlines other causes of dementia. Neurodegeneration is the most common underlying cause of dementia and is seen in Alzheimer's disease (AD), frontotemporal dementia, and diffuse Lewy body disease.

Most causes of dementias are currently untreatable. Potentially correctable causes account for less than 5% of dementia cases. Structural processes or infections must be considered, along with metabolic and nutritional diseases. Every patient with dementia should have tests of serum electrolytes and vitamin B_{12} and assessments of liver, renal, and thyroid function. Serologic studies for syphilis and Lyme exposure should be done if risk factors are identified. Chronic infections (see Chapter 90) and normal-pressure hydrocephalus should be considered. Brain imaging should be performed.

Neuropsychological testing characterizes the pattern of cognitive and memory impairments and is helpful in the differential diagnosis. The Montreal Cognitive Assessment (MoCA) (Table 108-4) is a standard test that can be used as a bedside or office screening tool for identifying patients with dementia. This examination is superior to the Mini-Mental Status Examination (MMSE) in that it is sensitive to abnormalities in a wider array of cognitive domains, including visual-spatial or executive

TABLE 108-1 DISTINGUISHING CHARACTERISTICS OF CORTICAL AND SUBCORTICAL DEMENTIAS

CORTICAL DEMENTIA
Symptoms: major changes in memory, language deficits, perceptual deficits, praxis disturbances
Affected brain regions: temporal cortex (medial), parietal cortex, and frontal lobe cortex
Examples: Alzheimer's disease, diffuse Lewy body disease, vascular dementia, frontotemporal dementias
SUBCORTICAL DEMENTIA
Symptoms: behavioral changes, impaired affect and mood, motor slowing, executive dysfunction, less severe changes in memory
Affected brain regions: thalamus, striatum, midbrain, striatofrontal projections
Examples: Parkinson's disease, progressive supranuclear palsy, normal-pressure hydrocephalus, Huntington's disease, Creutzfeldt-Jakob disease, chronic meningitis

TABLE 108-2 ETIOLOGIC DIAGNOSIS OF NEURODEGENERATIVE DEMENTIA IN ADULTS

Alzheimer's disease*
Parkinson's disease*
Diffuse Lewy body disease*
Progressive supranuclear palsy
Corticobasal ganglionic degeneration
Striatonigral degeneration
Olivopontocerebellar degeneration
Huntington's disease
Frontotemporal dementias
Pick's disease
Frontotemporal dementia without characteristic neuropathology
Frontotemporal dementia with motor neuron disease
Hallervorden-Spatz disease

*Denotes conditions for which symptomatic treatment is available.

TABLE 108-3 OTHER CAUSES OF PROGRESSIVE DEMENTIA IN ADULTS

STRUCTURAL DISEASE OR TRAUMA	INFECTIOUS DISEASE
Normal-pressure hydrocephalus*	Human immunodeficiency virus type 1*
Neoplasms*	Tertiary syphilis*
Dementia pugilistica (multiple concussions in boxers)	Creutzfeldt-Jakob disease
VASCULAR DISEASE	Progressive multifocal leukoencephalopathy
Vascular dementia†	Whipple's disease*
Vasculitis*	Chronic meningitis*
HEREDOMETABOLIC DISEASE	Cryptococcal meningitis*
Wilson's disease*	Others
Neuronal ceroid lipofuscinosis (Kufs' disease)	**METABOLIC OR NUTRITIONAL DISEASE**
Other late-onset lysosomal storage diseases	Vitamin B_{12} deficiency*
DEMYELINATING OR DYSMYELINATING DISEASE	Thyroid hormone deficiency or excess*
Multiple sclerosis†	Thiamine deficiency* (Korsakoff's syndrome)
Metachromatic leukodystrophy	Alcoholism†
	PSYCHIATRIC DISEASE
	Pseudodementia from depression*

*Denotes conditions for which preventive or corrective treatment is available.
†Denotes conditions for which symptomatic treatment is available.