Dementia (DSM-IV-TR #290.40–290.44, 294.10, 294.11, 294.8)

Dementia is a syndrome of multiple different etiologies characterized by a global decrement in cognitive functioning occurring in a clear sensorium. Though not confused, patients have difficulty with short-term memory and, to a relatively lesser degree, long-term memory. Intellectual abilities are likewise impaired. The ability to think abstractly fails, and patients become more and more concrete; relatively simple calculations are now beyond their grasp, and the ability to exercise "good judgment" in complex situations is lost.

In the past, dementia was also known as "chronic organic brain syndrome." Given, however, that not all dementias are necessarily chronic and irreversible, this term might best be dropped from our lexicon.

As most of the more common diseases that cause dementia occur in the elderly, the prevalence of dementia shows a striking association with age. Whereas anywhere from 5% to 10% of the entire population over the age of 65 is demented, at least 30% of the population over 80 is so affected.

ONSET

Both age and mode of onset are determined by the underlying disease. The onset may be acute (e.g., after head trauma) or insidious (e.g., with a slowly growing brain tumor). Furthermore, the onset may either be in early years (e.g., with adrenoleukodystrophy) or in later years (e.g., Alzheimer's disease).

CLINICAL FEATURES

The loss of short-term memory may manifest in a variety of ways. Patients may forget where they put their keys; they may forget to turn off the stove or lock the doors at night. Deficits in long-term memory, which are always less severe than shortterm memory losses, may become apparent when patients are unable to find their way home or forget where their grown children live. Short-term memory is formally tested by asking patients to recall three unrelated objects after 5 minutes; longterm memory is assessed by asking them to name the last four Presidents or to recall significant facts from their own distant personal history, such as where they went to school or when their parents died.

A deficiency in abstract thinking often becomes evident when the patient is faced with a new and complex task or situation. For example, the retired accountant may not be able to figure out and properly complete a new tax form, or a former champion chess player may be beaten again and again by relative novices. Abstract thinking may be formally tested by asking the patient to find similarities between apples and oranges or pencils and typewriters. Patients may also be asked to explain what old proverbs mean to them. The demented patient may be unable to see any similarity between a pencil and a typewriter and may give a "concrete" response to a proverb. For example, if asked what "don't cry over spilled milk" means, the demented patient may reply "well, it's already spilled."

The loss of calculating ability may become evident when the patient tries to make change or perhaps fails in balancing the checkbook. Formal testing is accomplished by asking the patient to perform simple addition and subtraction, and, if successful, to then attempt serial 7's by subtracting 7 from 100 and then 7 from the remainder, and so forth.

The loss of good judgment is often what first calls the patient to clinical attention. Subtleties are lost on the patient, and complex social situations may be approached with unaccustomed crudeness. A statesman might swear at a colleague, rather than use a "diplomatic" approach; an elderly retiree may begin propositioning teenagers. Manners and social graces are lost: food may be ravenously eaten; patients may indulge in crude jokes and overly boisterous laughter at parties. Grooming, dress, and hygiene often suffer. Patients may neglect to shave; makeup is applied haphazardly; crumbs of food are left on the shirt front.

Along with decay of good judgment, one often sees a "personality change." In some cases maladaptive personality traits become more prominent; a conscientious patient may become extremely rigid and cruelly critical; a circumspect patient may become suspicious and guarded. In other cases the personality may seem to change entirely: a prudish patient may become flirtatious and seductive; a happy-go-lucky patient may become irritable and demanding.

At times the deficits in abstracting ability or calculation may first become evident in what is known as an acute "catastrophic reaction." This may occur when patients, in facing a task that they had always been able to accomplish before, find that task completely beyond their ability to grasp. A typical example would be a checkout line in a store, when the patient, completely unable to manage the financial transaction, becomes extremely agitated. Such catastrophic reactions reflect the patient's enforced awareness of their deficits.

Often, hallucinations and delusions occur. The hallucinations may be either visual or, less commonly, auditory. Patients may see dead relatives or perhaps animals or complex scenes. Music and voices may be heard, or at times only creakings, footfalls, or sirens. A very common delusion is the belief that something has been stolen. Typically the patient puts something away and then, forgetting where it is and unable to find it, may accuse others of having stolen it. Another common delusion involves jealousy, and the patient may accuse the spouse of having an affair. Finally, one may encounter the "phantom boarder syndrome" wherein the patient believes that someone is hiding somewhere, perhaps in the attic or basement.

Other symptoms and signs, such as apraxia, agnosia, and aphasia, may or may not occur, depending on the underlying cause of the dementia.

COURSE

The course is determined by the underlying cause and may be either static or progressive. For example, the traumatic dementia following severe head injury tends to be "static," with symptoms generally remaining the same over the years. Conversely, Alzheimer's disease causes a relentlessly progressive dementia.

Although in the natural course of events almost all of the dementias are chronic, in some cases treatment may effect a remission. This may occur, for example, with treatment of hypothyroidism or hypovitaminosis B_{12} . In cases with structural damage, however, such as infarcts, a full remission is not possible.

Regardless of whether the course is static or progressive, patients with dementia are prone to the development of delirium during intercurrent illnesses. Urinary tract infections, attacks of bronchitis, and mild degrees of dehydration, all of which might have passed without complication before the dementia, now often cause a superimposed delirium. Thus the course of dementia may be "punctuated" by recurrent deliria.

COMPLICATIONS

Patients may wander away from home, perhaps only to become lost, but perhaps also out into a busy street. Some may insist on driving or using hazardous machinery long after they have lost the ability to do so. Those who insist on handling their finances and business affairs may become bankrupt.

In more advanced cases, patients are prone to decubiti, falls and fractures, dehydration, and the like.

ETIOLOGY

The box on p. 285 lists most of the important causes of dementia. Of all these, the most common cause of dementia is Alzheimer's disease; other common causes include certain of the vascular dementias (e.g., multi-infarct dementia, lacunar dementia, Binswanger's disease), alcoholic dementia, diffuse Lewy body disease, advanced Parkinson's disease, various tumors, normal pressure hydrocephalus, subdural hematoma, and, especially among younger adults, AIDS. Although only 10% or less of all cases of dementia are due to potentially fully reversible causes, one should nevertheless diligently search for these, with special attention to Wilson's disease, Hashimoto's encephalopathy, systemic lupus erythematosus, subdural hematoma, certain tumors (e.g., meningiomas or low-grade gliomas), normal pressure hydrocephalus, hyper- or hypothyroidism, and either vitamin B_{12} or folate deficiency.

When, after a thorough history and physical examination, the cause is not clear, instituting a "screen" of laboratory tests is

Causes of Dementia

SUBSTANCE RELATED	**MASS LESIONS AND HYDROCEPHALUS**
Inhalants	Brain tumor
Alcoholic dementia	Brain abscess
Methanol intoxication	Hydrocephalus
Marchiafava-Bignami disease	Normal-pressure hydrocephalus
HEREDODEGENERATIVE DISORDERS	**ENDOCRINOLOGIC DISORDERS**
Alzheimer's disease	Hyperthyroidism (apathetic type)
Pick's disease	Hypothyroidism
Parkinson's disease	**METAL POISONING**
Diffuse Lewy body disease	Lead encephalopathy
Huntington's disease	Manganese dementia
Wilson's disease	Thallium poisoning
Progressive supranuclear palsy	Arsenic poisoning
Multiple system atrophy	Mercury poisoning
Myotonic dystrophy	Dialysis dementia
Amytrophic lateral sclerosis	**VITAMIN DEFICIENCIES**
Cerebrotendinous xanthomatosis	Pellagra
Adrenoleukodystrophy	Vitamin B_{12} deficiency
Metachromatic leukodystrophy	Folic acid deficiency
Hallervorden-Spatz disease	**INFECTIOUS AND RELATED DISORDERS**
VASCULAR DISORDERS	Neurosyphilis
Multiinfarct dementia	Tuberculosis
Lacunar dementia	Lyme dementia
Binswanger's disease	Whipple's disease
Polyarteritis nodosa	Progressive rubella panencephalitis
Cranial arteritis	Subacute sclerosing panencephalitis
Granulomatous angiitis	Encephalitis lethargica
Hypertensive encephalopathy	AIDS dementia
CADASIL	Progressive multifocal leuko-encephalopathy
Cerebral amyloid angiopathy	Cytomegalovirus encephalopathy

Wegener's granulomatosis	Mycoses
IMMUNE-MEDIATED DISORDERS	Toxoplasmosis
Limbic Encephalitis	**METABOLIC DISORDERS**
Multiple sclerosis	Hypoglycemia
Hashimoto's encephalopathy	Hypocalcemic encephalopathy
Systemic lupus erythematosus	Acquired hepatocerebral degeneration
Sarcoidosis	
BRAIN INJURY	
Postanoxic dementia	
Delayed postanoxic encephalopathy	
Traumatic dementia	
Subdural hematoma	
Dementia pugilistica	
Delayed radiation encephalopathy	

prudent. Among adults the vast majority of cases are identified by one of the following: MRI scan with enhancement, ANA, ESR, thyroid profile with TSH, B12 and folate levels (or, for enhanced sensitivity, homocysteine and methylmalonic acid levels), calcium, HIV and FTA-ABS (neither an RPR nor a VDRL are adequate for testing for neurosyphilis as both may be normal in such cases). If Wilson's disease is suspected, copper and ceruloplasmin levels are obtained, and if Hashimoto's encephalopathy seems likely, antithyroid antibodies. If this first "screen" is negative, consideration may be given to a second level screen with an EEG and lumbar puncture. If a lumbar puncture is performed, the following are routinely ordered: total protein, glucose, cell count and differential, VDRL, HIV antibodies, myelin basic protein, oligoclonal bands, IgG level, India ink preparation, gram stain and routine culture and sensitivity. A simultaneous blood glucose and serum protein electrophoresis should also be obtained. Other CSF studies (e.g., PCR assay for tuberculosis, fungal cultures) are ordered as indicated.

DIFFERENTIAL DIAGNOSIS

Among children mental retardation may be distinguished from dementia by the course. In mental retardation cognitive development increases only up to a point (at the most to about a sixth-grade level), then "flattens out," without any decrement. By contrast, in dementia, cognitive ability undergoes a definite falling off. Occasionally, as in Down's syndrome, the same disease may cause both mental retardation and a dementia.

"Benign senescent forgetfulness" is the term applied to the mild degree of memory loss that is a normal part of aging. Unfortunately, given a person with a recent slight loss of memory, one cannot definitively tell, prospectively, whether the memory loss will remain mild or progress and be joined by other cognitive losses. Thus only long-term follow-up can reliably distinguish between benign senescent forgetfulness and a very gradually progressive dementia.

Delirium is distinguished from dementia primarily by the presence of confusion. At times, however, certain diseases can cause both delirium and dementia. A common example is multi-infarct dementia, wherein each fresh infarct creates a delirium, which in turn gradually clears along with the peri-lesional edema, leaving the patient, however, one "step" further down into dementia. Another important example is diffuse Lewy body disease, wherein delirium typically occurs early on in the course of the dementia.

Depression, especially in the elderly, may cause at times profound cognitive deficits. This "dementia syndrome of depression" (or, as it has also been called, "pseudodementia") is suggested by a history of depressive episodes, the presence of typical "vegetative" signs (e.g., insomnia, weight loss) and by the patient's tendency to give up trying on being asked difficult mental status questions. In doubtful cases, a "diagnosis by treatment response" to an antidepressant may be justified; however, one must keep in mind that the cognitive deficits of any given elderly patient may be due not only to depression but also to some other process, such as Alzheimer's disease.

Amnesia, as may occur in Korsakoff's syndrome, may superficially resemble dementia. However, in these cases only memory is lost; abstracting and calculating abilities are retained.

TREATMENT

The first task is to arrive at an accurate diagnosis of the causative disease and then, if any specific treatment is available, to institute it. Concurrent with this, symptomatic measures, if required, may be instituted.

The demands placed on patients should be reduced commensurate with their reduced cognitive abilities. Weapons should be removed and, eventually, patients will have to surrender the car keys, credit cards and the checkbook. Guardianship may be required.

Familiar routine and surroundings should be maintained for as long as possible. A move from a lifetime home to a retirement apartment may be catastrophic for patients who are unable to remember and familiarize themselves with new surroundings. Night-lights are a necessity, and for patients who wander, locking doors and even windows may be necessary.

If hospitalization or institutionalization is required, certain measures can reduce the risk of a catastrophic reaction. The patient's room should have a window, a large calendar (marked off day by day), and a large clock, preferably a digital one. Familiar photographs and personal items (even furniture, if possible) should be brought, and arrangements should be made for delivery of the patient's local newspaper. Visits from family and friends should generally be encouraged.

At times, institutionalization may be avoided by the use of home sitters, visiting nurses, “Meals on Wheels,” and adult day-care centers.

Symptomatic pharmacologic treatment of dementia may be required for agitation or aggression, delusions or hallucinations, depression and insomnia.

Agitation or aggression may be treated with antipsychotics, antiepileptic drugs, or trazodone. Both low-dose risperidone and haloperidol (i.e., 0.5 to 1.5 mg) are effective, but risperidone is better tolerated. Carbamazepine in low doses (yielding blood levels at the low end of the “therapeutic range”) is helpful, and there is some indication that divalproex may also be effective. Trazodone, in doses of 50 to 250 mg, was found equivalent to haloperidol in one study. Choosing among these agents is difficult, as there are few comparison studies. Certainly, if there were concurrent delusions or hallucinations, an antipsychotic would be a reasonable first choice; however, these agents must be used with extreme care in cases of diffuse Lewy body disease, given the risk of severe antipsychotic-induced parkinsonism. Initial doses should be low, and titration should be gradual, and this is especially the case with carbamazepine and trazodone, given the risk of hypotension and falls.

Delusions or hallucinations generally respond to risperidone; however, in the case of diffuse Lewy body disease, there is now evidence that rivastigmine, a cholinesterase inhibitor, may also be effective.

Depression may be treated with an SSRI, such as citalopram; if a tricyclic is deemed preferable, consideration may be given to nortriptyline.

Insomnia may be treated with low dose lorazepam or zolpidem.

Certain dementing disorders have specific treatments (e.g., cholinesterase inhibitors for Alzheimer’s disease) and these are specified in the appropriate chapter.

All medication regimens should be kept as simple as possible, with the lowest number of pills and the least number of dosings per day possible.

Proper glasses, hearing aids and dentures, if needed, are essential. Quad canes and walkers should be encouraged, and wheelchairs avoided if at all possible. Rigorous internal medical follow-up is required in all cases, and it must be borne in mind that seemingly trivial disorders, such as urinary tract infections, may cause significant agitation in elderly demented patients.

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