

Delirium, Vascular Dementia, Frontotemporal Dementia and more

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Dementia is a syndrome of impairment of cognition and memory accompanied by interference with activities of daily living, without a disturbance in consciousness or level of alertness. Dementia should always be differentiated from normal memory loss that occurs with aging. This change associated with old age does not meet the criteria and severity for dementia that limits a person's physical activity.



Definition

Dementia is a **syndrome of impairment of cognition and memory** accompanied by interference with activities of daily living, without a disturbance in consciousness or level of alertness.

Impairment of cognition may involve:

- Language disturbances (aphasia)
- Loss of memory (amnesia)
- Inability to carry out motor activities (apraxia)
- Inability to recognize objects in the setting of intact sensory function (agnosia)
- Emotion and personality defects

Epidemiology of Dementia

Above 65 years of age, dementia will develop in approximately 17–20%. About 70% of those who develop dementia have Alzheimer disease dementia, 17% have vascular disease, and 13% have dementia with Lewy bodies, Parkinson-related dementia, alcoholic dementia, or frontal lobe dementia. It is estimated that **36 million people worldwide were diagnosed with dementia** in 2011.

The disease is more common among the elderly, and its prevalence increases with age to a rate of $\geq 20\%$ for patients over the age of 85 years. There is a 10–15% chance of developing Alzheimer disease from mild dementia. The average survival period of dementia patients is 4.5 years.

Due to alterations in the ability to perform daily functions, **the syndrome has a serious economic and social impact** as these patients require a helper and must take medications for the rest of their life.

Classification of Dementia

Dementia arises from neurodegeneration of the basal ganglia, thalamus, and cerebral cortex. Dementia has several possible causes, which categorizes it into different types. Although the causes of these types of dementias differ, their clinical presentations may overlap with some distinctive features.

The various types of dementia include:

- Alzheimer disease (AD)
- Vascular dementia
- Dementia with Lewy bodies (DLB)
- Parkinson disease dementia (PDD)
- Frontotemporal dementia (FTD)
- Other rare forms of dementia
- Creutzfeldt-Jakob disease
- Mixed dementia
- Wernicke-Korsakoff syndrome
- Huntington disease

Alzheimer disease, the most common cause of dementia, accounts for 70–80% of dementia cases.

Vascular Dementia

Definition

Vascular dementia is defined as a decline in mental function caused by brain damage after the insufficient or impaired blood supply to the brain.

Epidemiology

It is the **second most common cause of dementia after Alzheimer disease**, and it accounts for about 10% of cases.

Classification

Vascular dementia is classified into

1. Uncomplicated type
2. Vascular dementia with delirium
3. Vascular dementia with delusions
4. Vascular dementia with depressed mood

Etiology

It is **caused by a stroke that blocks an artery to a specific region of the brain** or by narrowing of blood vessels.

Pathophysiology

The **risk factors for stroke, such as alcohol use, smoking, and the presence of vascular disease**, also increase the risk of developing vascular dementia. Vascular blockage results from a stroke or a pathogenic narrowing of the vessel that compromises the distal flow of blood.

This leads to ischemia-induced demyelination or axonal loss in the areas that are responsible for memory and behavior control.

Clinical features

The diagnostic criteria entail:

- The patient must fulfill general criteria for dementia
- Multiple large infarcts in the cerebrum
- More than three microscopic infarcts identified in a systematic screening of the cortex and deep cerebral structures

The clinical presentation includes confusion, trouble paying attention, restlessness, reduced ability to organize thoughts, unsteady gait, depression, and neurological deficits, such as inability to understand speech or formulate speech and loss of vision associated with the stroke.

Investigations

There is **no specific diagnostic test** for vascular dementia, but tests are done to correlate clinical findings and to help confirm the diagnosis.

- Blood pressure, blood glucose level, and cholesterol level are measured to check for any risk factors
- Brain CT scan shows areas of shrinkage and hemorrhage
- MRI of the brain is more specific for identifying soft tissue damage and smaller infarcts

Neuropsychological tests assess one's ability to speak and work with numbers, memory, and problem-solving skills.

Treatment

Control of risk factors

- Lower blood pressure, cholesterol levels, and blood glucose level

Alzheimer disease medications

- Medications are used to alleviate symptoms and improve the quality of life in these patients

Prognosis

Studies show that **patients with vascular dementia survive for an average of 3 years**. However, some dementias may improve with the resolution of stroke symptoms. This feature is attributed to the development of new vessels that vascularize the ischemic part of the brain.

Dementia with Lewy Bodies (DLB)

Definition

Dementia with Lewy bodies is defined as a type of dementia that develops progressively with abnormal deposition of Lewy bodies responsible for impaired mental functions related to thinking, movement, behavior, and mood.

Epidemiology

It is the **third cause of dementia after Alzheimer disease and vascular dementia**, and it accounts for 10% of cases. It has a slight male predilection. The disease is more common among the elderly population with the age for peak incidence being 50–85 years.

Etiology and Pathophysiology

The **cause of the disorder is largely unknown** but several factors have been associated with increased incidence. The mutation in the **CYP2D6B** gene confers susceptibility to interruption of the metabolic pathway of environmental toxins in the brain. α -Synuclein proteins are the chief components of Lewy bodies. They are found abundantly in brain cells of people suffering from DLB, Parkinson disease dementia, and Alzheimer disease.

The presence of Lewy bodies alters the level of **neurotransmitters** and **neuromodulators** in the brain, principally dopamine. This **depletion compromises the flow of information from the striatum to the neocortex**. It begins in the brainstem and later spreads to the outer layers of the brain that are involved in perception and behavior.

Clinical features

Dementia with Lewy bodies commonly presents with features seen in other diseases. They **include motor features of Parkinson disease**, such as:

- Altered balance
- Hunched posture
- Visual hallucinations
- Rigidity
- Features of Alzheimer disease
- Anterograde memory loss
- Extrapyramidal symptoms
- Delusions
- Problems in understanding things and making judgments

Other features of the disease include:

- Fluctuating periods of alertness and loss of attention
- REM sleep disorder where the patient is violent and restless as he/she acts out his/her nightmares

Note: It is always necessary to differentiate the three conditions based on specific clinical findings in one condition but not in the other.

Important findings on physical examination include:

- Impaired cognitive function as seen in the mini-mental state examination (MMSE).
- Alternate periods of being alert and oriented vs confused and unresponsive.
- Motor signs that do not meet the criteria for Parkinson disease.

Investigations

The **diagnosis is made from clinical findings**. Investigations are done to support the diagnosis or to rule out other likely differential diagnoses.

1. Brain imaging by MRI/CT/PET scans may identify infarcts that suggest vascular dementia or brain atrophy that suggests Alzheimer disease.
2. CSF analysis: DLB can be diagnosed by identification of an increased level of tau proteins.
3. Low vitamin B12 levels showing deficiency could explain behavioral problems.

Differential diagnosis

Alzheimer's disease dementia	<ul style="list-style-type: none"> • Lacks hallucinations, motor features and fluctuations in alertness and lapse in concentration • Memory loss is more prominent
Parkinson's disease dementia	<ul style="list-style-type: none"> • Memory loss is a feature that is prominent and always seen • Motor symptoms are more pronounced
Vascular dementia	<ul style="list-style-type: none"> • Associated with a recent stroke

Treatment

There are **no medications to halt the progress or completely cure the disease**. Available medications focus on the improvement of the quality of life by symptomatic relief.

- Cholinesterase inhibitors (donepezil) increase the amount of available neurotransmitters and thus transmission of signals.

- Antipsychotics (clozapine and quetiapine) alleviate behavioral symptoms and may lead to altered consciousness, confusion, and hallucinations.
- Clonazepam is administered to treat REM sleep disorder.

Parkinson disease medications (carbidopa-levodopa) treat motor symptoms.

Parkinson's Disease Dementia (PDD)

Definition

Parkinson disease is the second most common neurodegenerative disorder and the most common movement disorder. It is characterized by progressive loss of muscle control, which leads to trembling of the limbs and head while at rest, stiffness, slowness, and impaired balance. The disease affects approximately 1 percent of persons older than 60 years, and up to 4 percent of those older than 80 years.

Pathophysiology

Parkinson disease dementia results from the presence of α -synuclein deposits like those seen in dementia with Lewy bodies. There is the loss of pigmented dopaminergic neurons in the substantia nigra pars compacta (SNpc) and the presence of Lewy bodies. This leads to destruction of brain cells and depletion of dopamine (the hormone that controls muscle coordination) in the basal ganglia.

The electrophysiologic changes, such as altered discharge rates, increased incidence of burst firing, interneural compatibility, oscillatory activity, and altered sensorimotor processing, in the basal ganglia, thalamus, and cerebral cortex, adversely affect voluntary movements of extremities and sensory functions. The disease thus represents a movement disorder; upon involvement of other brain areas that control memory and concentration, it results in dementia.

Clinical features

In addition to **diagnostic features of Parkinson disease (tremors, stiffness, slow movement, and loss of balance and coordination)**, Parkinson disease dementia has additional features of:

- Changes in memory, concentration, and judgment
- Visual hallucination
- Depression
- Delusions
- Irritability
- Anxiety
- REM sleep disorder

Investigations

Like other causes of dementia, available tests are used to support the diagnosis or rule out differential diagnoses. The clinical representation of Parkinson disease is the guideline for the diagnosis and **imaging of the brain should be done to rule out tumors or vascular diseases that may cause motor symptoms and dementia.**

Treatment

Symptomatic relief is achieved by:

- **Cholinesterase** inhibitors (**donepezil**) increase the available neurotransmitters and thus increase transmission of signals through neurons.
- **Antipsychotics** (**clozapine** and **quetiapine**) alleviate behavioral symptoms and may lead to altered consciousness, confusion, and hallucinations.
- **Clonazepam** is administered to treat REM sleep disorder.
- Antidepressant medications, such as SSRIs, may be effective in many cases.
- Parkinson disease medications (**carbidopa-levodopa**) treat motor symptoms (may aggravate hallucinations and confusion).

Frontotemporal Dementia (FTD)

Definition

Frontotemporal dementia is a **clinical syndrome associated with shrinkage of the frontal and temporal lobes of the brain**. It is also known as Pick's disease, frontal lobe dementia, or frontotemporal disorder. These disorders occur as a result of the damage of neurons in the frontal and temporal lobes of the brain with the death of the neurons; these lobes atrophy, which leads to behavioral change and diminished thought processes.

Epidemiology

Frontotemporal dementia is a rare cause of dementia that is **more common in the younger population**. The peak age of incidence of the disease is 45-65 years.

It has no sex predilection as seen with other types of dementia.

Etiology

A genetic mutation in **MAPY**, **GPN**, and **C9ORF72** genes.

Pathophysiology

The brain regions responsible for behavioral reactions, speech, and emotional reactions shrink due to progressive damage to the neurons in these areas. Death of these brain cells (neurons) is thought to arise from the presence of abnormal proteins in these cells. **Progression of the disease damages the connecting neurons in these regions and leads to changes in behavior, speech, and personality** with intact memory.

Clinical features

Presentation of the three main variants determines its classification:

1. Behavioral variant frontotemporal dementia (BvFTD) is the most common variant of FTD. It is **characterized by changes in social behavior and conduct**, with loss of social tact, neglect of personal hygiene, and poor impulse control. The individual lacks empathy and insight into personal behavior but has increased interest in sex and blunt emotions.
2. Semantic dementia (SD) is characterized by difficulty in understanding and making

a speech. **The individual suffers from impaired word comprehension with intact grammar and memory.**

3. Progressive nonfluent aphasia (PNFA) is **characterized by progressive difficulties in speech production.** The patient's speech is punctuated with omission of words that leads to grammatical errors. The speech also is sluggish.

Investigations

The diagnosis is made from clinical findings and investigations are done to support the diagnosis or to rule out other likely differential diagnoses.

- Brain MRI is superior for assessment for suspected brain atrophy.
- CT scans can help to rule out vascular dementia.
- PET scans may identify infarcts that suggest vascular dementia or brain atrophy that suggests Alzheimer disease.

Differential diagnosis

Atypical Alzheimer's disease	• Lacks memory loss
Schizophrenia	• Has prominent behavioral symptoms

Treatment

Care of these patients involves:

- Physiotherapy and occupational therapy to allow for some function of basic chores.
- Supportive care of the patient since he/she cannot express herself/himself.
- Speech therapy, such as encouraging the use of small sentences.
- Antipsychotic medications are considered in the control of extreme alterations in behavior.

Delirium

Just like dementia, **delirium is not a disease but a syndrome of disturbance in consciousness**, featuring abrupt change in cognition. The syndrome is characterized by clouding of consciousness, restlessness, confusion, psychomotor retardation or agitation, and affective lability.

Delirium is caused by

- General medical condition
- Drug/substance abuse
- Multifactorial delirium (multiple causes)
- Delirium not otherwise specified (cause not known)

It has been classified into

- Hyperactive delirium (restlessness, agitation, and rapid mood changes are the hallmarks)
- Hypoactive delirium (sluggishness or loss of motor function)
- Mixed delirium (includes both hyperactive and hypoactive symptoms and switches between the two)

Delirium and dementia are commonly confused due to the superficial similarities in the two diseases such as

- Both diseases are more common among the elderly population (> 65 years).
- Both diseases may occur concurrently and presence of dementia leads to brain damage that predisposes one to delirium.
- REM sleep disorder may be evident in both syndromes.
- **Both diseases lack tests that can confirm the diagnosis.**

The differences are clear and should be looked for during diagnosis. While **delirium is always of acute onset, dementia progresses gradually with worsening of the symptoms**. Patients with dementia are consciously alert and pay attention to the current happenings whereas patients with delirium have altered consciousness levels. Dementia symptoms are worse in daytime whereas delirium symptoms may fluctuate with some improvement during the day and worsening at night.

Difference between delirium and dementia

Delirium	Dementia
Clouding of consciousness	Loss of memory/intellectual ability
Acute onset	Insidious onset
Lasts 3 days to 2 weeks	Lasts months to years
Orientation impaired	Orientation often impaired
Immediate/recent memory impaired	Recent and remote memory impaired
Visual hallucinations common	Hallucinations less common
Symptoms fluctuate, often worse at night	Symptoms stable throughout the day
Usually reversible	15% reversible
Awareness reduced	Awareness clear
EEG changes (fast waves or generalized slowing)	No EEG changes

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