

434 Dementia with Lewy Bodies

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Lewy body disease (LBD), manifesting as Parkinson's disease and dementia (PDD) or dementia with Lewy bodies (DLB), is the second most common cause of neurodegenerative dementia, after Alzheimer's disease (AD) (**Chap. 431**). Approximately 10% of patients with PD develop PDD per year, with the majority of PD patients developing PDD over time. The incidence of DLB is approximately 7 per 100,000 person-years. The prevalence of both PDD and DLB increases with aging, and both affect men more often than women.

CLINICAL MANIFESTATIONS

Most researchers conceptualize PDD and DLB as points on a spectrum of LBD pathology. Cognitively, PDD and DLB usually manifest with severe executive, attentional, and visuospatial deficits but preserved episodic memory. Cognitive decline in LBD affects performance of daily living activities beyond other PD symptoms. Early psychosis including well-formed visual hallucinations, fluctuating cognition, rapid eye movement sleep behavior disorder (RBD), and parkinsonism are the main diagnostic features in DLB. The sense of a presence behind the person may precede well-formed hallucinations. Delusions are less frequent than hallucinations and are usually related to misidentification, infidelity, theft, or persecution. Fluctuating attention and concentration are other characteristic features. Minor day-to-day variation in cognitive functioning is common across dementias, but in DLB these fluctuations can be marked, with short periods of confusion or severe lethargy that may rapidly resolve. Patients with PDD and DLB are highly sensitive to infectious or metabolic disturbances. The first manifestation of DLB in some patients is delirium, often precipitated by an infection, new medicine, or other systemic disturbance.