key points

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Lewy body dementia (LBD) is a heterogeneous condition that is often difficult to treat. It comprises Parkinson's disease dementia (PDD), in which the cognitive and functional symptoms of dementia arise in the context of established Parkinson's disease, and dementia with Lewy bodies (DLB) where parkinsonism may develop concurrently with, or within a year of, dementia diagnosis. Both conditions are characterised by the same neuropathological mechanism; the cerebral aggregation of misfolded alpha-synuclein protein. Although the distinction between PDD and DLB is arbitrary, it is useful in understanding how patients experience their illness and determining the services in which they are likely to be treated.

LBD represents around 5% of dementia cases seen in clinical services; however, a large proportion of additional cases probably go undetected or are misdiagnosed. DLB is associated with higher rates of neuropsychiatric symptoms, inpatient admission and carer stress, as well as shorter survival, than other dementia subtypes. Accurate recognition can help guide clinicians towards specific treatment strategies and help avoid neuroleptic sensitivity.

A definite diagnosis of DLB can only be determined at post mortem, but the presence of two or more core clinical features of DLB confers a probable DLB diagnosis in patients with a dementia syndrome, with excellent sensitivity and good specificity. Core clinical features of DLB are: fluctuating cognition with pronounced variations in attention and alertness; recurrent visual hallucinations; REM sleep behaviour disorder (RBD); one or more features of spontaneous parkinsonism. Recurrent visual hallucinations occur in up to 80% of patients with DLB. RBD constitutes recurrent dream enactment and chasing or attacking themes are common. RBD symptoms can precede DLB diagnosis by ten years; 80% of individuals with RBD in the absence of dementia will develop a neurodegenerative disease within ten years.

Clinical assessment for any dementia subtype should follow the framework recommended by NICE, including the use of screening blood tests and assessment for reversible causes. A clinical history, including collateral history, is critical in the diagnosis of DLB. Assessment should include questions aimed at detecting each core symptom.

Following diagnosis acetylcholinesterase inhibitors rivastigmine and donepezil are the agents most frequently prescribed to patients with DLB and are recommended by NICE. Neither are licensed yet for DLB however, both drugs are associated with a modest reduction in carer burden and improvement in functioning.

Neuropsychiatric symptoms are among the most challenging to manage in LBD and often require referral to psychiatry of old age services. LBD is associated with a risk of severe neuroleptic sensitivity, which occurs in around a third of patients. Antipsychotics should be prescribed as a last resort and initiated by secondary care specialists.