

THE LEWY BODY COMPOSITE RISK SCORE (LBCRS)

Purpose of Use

The Lewy body dementias, composed of two related disorders: Dementia with Lewy bodies (DLB) and Parkinson's disease dementia (PDD) are a challenge to diagnose, particularly outside of expert centers. One of the great challenges in differential diagnosis of neurodegenerative disorders is attributing clinical symptoms to specific pathologies to guide treatment choices and discuss prognosis and clinical course. While PDD provides a potentially easier route to diagnosis because the cognitive disorder begins in face of an established movement disorder and criteria have defined a mild cognitive impairment (MCI) state, DLB is a more difficult entity to diagnose with delays in diagnosis approaching 18 months leading to significant burden to patients and caregivers. Patients with DLB are often misdiagnosed. While consensus criteria for DLB have excellent specificity (79-100%), there is no standardized way to assess or operationalize many of the cognitive and behavioral symptoms which markedly decreases sensitivity in clinical practice (range 12-88%). We developed the Lewy Body Composite Risk Score (LBCRS) to improve the ability to detect DLB and PDD in clinic and research populations and increase the likelihood of determining whether Lewy bodies are contributing pathology to the cognitive diagnosis. The LBCRS was derived from clinical features in autopsy-verified cases of healthy controls, Alzheimer's disease (AD), DLB, and PD with and without dementia. The LBCRS was tested in a consecutive series of 256 patients compared with the Clinical Dementia Rating and gold standard measures of cognition, motor symptoms, function, and behavior. The LBCRS increases diagnostic probability that Lewy body pathology is contributing to the dementia syndrome and should improve clinical detection and enrollment for clinical trials.

Administration and Scoring Guidelines

The questions are completed by a clinician after interview with patient and caregiver and a complete physical and neurological exam. The operationalization of physical findings as being present for at least 6 months or symptoms occurring at least 3 times over the past 6 months permitted the scoring of the LBCRS by totaling the sum of signs and symptoms rated as present. In the context of a patient with cognitive impairment, the LBCRS can increase the probability that Lewy bodies are a significant contributor to the clinical diagnosis.

The LBCRS has 10 Yes/No questions; 4 questions cover motor symptoms while 6 questions cover non-motor symptoms. The clinician rates the presence or absence of physical signs and can elicit information from either the patient or caregiver regarding the presence or absence of symptoms.

Interpretation of the QDRS

A screening test in itself is insufficient to diagnose a dementing disorder. The LBCRS is, however, quite sensitive to suggesting that there is a high probability that Lewy bodies are a contributing pathology to the underlying cognitive decline either as a single pathology or as a mixed dementia. The LBCRS discriminates DLB, PDD and MCI due to Lewy body disease from other forms of cognitive impairment.

The LBCRS is scored on a continuous scale with a range of 0-10. Based on receiver operator characteristic curves from 265 individuals included in the development and validation samples, LBCRS scores differentiate with the following cut-points:

Non Lewy Body Case	0-2
Probable Lewy Body Case	3-10

Using the cutoff of 3 or greater, the LBCRS was able to discriminate:

	DLB vs. AD	DLB vs any dementia	MCI DLB vs MCI AD
Area Under Curve	0.94 (0.90-0.97)	0.94 (0.91-0.98)	0.96 (0.91-1.0)
Sensitivity	94.2	97.9	100
Specificity	78.2	86.1	72.9
Positive Likelihood Ratio	4.1	7.0	3.2
Negative Likelihood Ratio	0.08	0.02	0.0

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LEWY BODY COMPOSITE RISK SCORE

Please rate the following physical findings being present or absent for the past 6 months and symptoms as being present or absent for at least 3 times over the past 6 months. Does the patient...	Yes	No
Have slowness in initiating and maintaining movement or have frequent hesitations or pauses during movement?		
Have rigidity (with or without cogwheeling) on passive range of motion in any of the 4 extremities?		
Have a loss of postural stability (balance) with or without frequent falls?		
Have a tremor at rest in any of the 4 extremities or head?		
Have excessive daytime sleepiness and/or seem drowsy and lethargic when awake?		
Have episodes of illogical thinking or incoherent, random thoughts?		
Have frequent staring spells or periods of blank looks?		
Have visual hallucinations (see things not really there)?		
Appear to act out his/her dreams (kick, punch, thrash, shout or scream)?		
Have orthostatic hypotension or other signs of autonomic insufficiency?		
TOTAL SCORE		