

Professional Brief: New DLB Diagnostic Criteria

In June, 2017 the international Dementia with Lewy Bodies (DLB) Consortium published updated diagnostic criteria for dementia with Lewy bodies in the journal *Neurology*.

Compared to the previous criteria, the new criteria now distinguish between ***clinical features*** and ***diagnostic biomarkers*** and provide guidance on how to establish and interpret them to make a DLB diagnosis.

Based on the diagnostic specificity, clinical features are divided into *core* and *supportive* categories and biomarkers are divided up into *indicative* and *supportive categories*. These revised categorizations provide greater weight to those features and biomarkers that are highly associated with the presence of Lewy body pathology.

KEY CHANGES

- Biomarkers are divided into the categories 'indicative' and 'supportive.'
- Compared to previous criteria, greater weight is now given to the presence of rapid eye movement (REM) sleep behavior disorder and iodine-123 - metaiodobenzylguanidine (MIBG) myocardial scintigraphy.
- Hypersomnia and hyposmia are new supportive clinical features.

As DLB is a heterogeneous disorder, this new diagnostic construct allows healthcare providers the ability to diagnose the disease based on the patient's individualized clinical presentation and biomarker profile.

Reference: (OPEN ACCESS)

McKeith IG, Boeve BF, Dickson DW, et al. Diagnosis and management of dementia with Lewy bodies: Fourth consensus report of the DLB Consortium. *Neurology*. 2017 Jul 4;89(1):88-100.

Patient Resource:

Patient Checklist for Diagnostic Symptoms: <http://www.lbda.org/content/lbd-diagnostic-symptoms-checklist>

Revised Criteria for the Clinical Diagnosis of Probable and Possible DLB

Essential for a diagnosis of DLB is dementia, defined as a progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational functions, or with usual daily activities.

- Prominent or persistent memory impairment may not necessarily occur in the early stages but is usually evident with progression.
- Deficits on tests of attention, executive function and visuo-perceptual ability may be especially prominent and occur early.

<p style="text-align: center;">Core Clinical Features</p> <p>(NOTE: The first 3 typically occur early and may persist throughout the course)</p> <ul style="list-style-type: none"> • Fluctuating cognition with pronounced variations in attention and alertness • Recurrent visual hallucinations that are typically well formed and detailed • REM sleep behavior disorder (RBD) which may precede cognitive decline • One or more spontaneous cardinal feature of parkinsonism – these are bradykinesia (defined as slowness of movement and decrement in amplitude or speed), rest tremor, or rigidity 	<p style="text-align: center;">Indicative Biomarkers</p> <ul style="list-style-type: none"> • Reduced dopamine transporter (DaT) uptake in basal ganglia demonstrated by SPECT or PET • Abnormal (low uptake) ¹²³Iodine-MIBG myocardial scintigraphy • Polysomnographic confirmation of REM sleep without atonia <p style="text-align: center;">See open access article in Neurology (referenced on Page 1) for examples of abnormal scan results.</p>
<p style="text-align: center;">Supportive Clinical Features</p> <p>Severe sensitivity to antipsychotic agents; postural instability; repeated falls; syncope or other transient episodes of unresponsiveness; severe autonomic dysfunction e.g. constipation, orthostatic hypotension, urinary incontinence; hypersomnia; hyposmia; hallucinations in other modalities; systematized delusions; apathy, anxiety and depression</p>	<p style="text-align: center;">Supportive Biomarkers</p> <ul style="list-style-type: none"> • Relative preservation of medial temporal lobe structures on CT/MRI scan • Generalized low uptake on SPECT/PET perfusion/metabolism scan with reduced occipital activity +/- the cingulate island sign on FDG-PET imaging • Prominent posterior slow wave activity on EEG with periodic fluctuations in the pre-alpha/theta range

PROBABLE or POSSIBLE DLB

Probable DLB can be diagnosed if:

- a) two or more core clinical features of DLB are present, with or without the presence of indicative biomarkers,
- or**
- b) only one core clinical feature is present, but with one or more indicative biomarkers.

Probable DLB should not be diagnosed on the basis of biomarkers alone.

Possible DLB can be diagnosed if:

- a) only one core clinical feature of DLB is present, with no indicative biomarker evidence,
- or**
- b) one or more indicative biomarkers is present but there are no core clinical features.

DLB is LESS LIKELY:

- a) in the presence of any other physical illness or brain disorder including cerebrovascular disease, sufficient to account in part or in total for the clinical picture, although these do not exclude a DLB diagnosis and may serve to indicate mixed or multiple pathologies contributing to the clinical presentation,
- or**
- a) if parkinsonian features are the only core clinical feature and appear for the first time at a stage of severe dementia.

Differentiating between DLB and Parkinson's Disease Dementia

DLB should be diagnosed when dementia occurs before, or concurrently with parkinsonism. The term Parkinson's disease dementia (PDD) should be used to describe dementia that occurs in the context of well-established Parkinson's disease.

In a practice setting the term that is most appropriate to the clinical situation should be used and generic terms such as LB disease are often helpful.

In research studies in which distinction needs to be made between DLB and PDD the existing one-year rule between the onset of dementia and parkinsonism continues to be recommended.

Assessing Cognition in the Primary Care Setting

DLB is frequently misdiagnosed as Alzheimer's disease. While dementia screens like the Mini Mental State Assessment (MMSE) and Montreal Cognitive Assessment (MoCA) are useful tools to detect global impairment, neither are able to differentiate DLB from Alzheimer's disease (AD).

TIP: *If DLB is suspected, a comprehensive neuropsychological assessment is recommended to determine the full range of cognitive domains affected.*

People with DLB also experience spontaneous variations in cognition, attention and arousal. These includes behavioral fluctuations, episodes of incoherent speech, altered consciousness and varying degrees of attention. While the patient may not be able to report these fluctuations, questions to an informant about daytime drowsiness, lethargy, staring into space or episodes of disorganized speech are useful in detecting fluctuating cognition.

TIP: *When detected early in the course of dementia, these fluctuations can help differentiate between DLB and AD.*

Visual Hallucinations

Visual hallucinations occur in up to 80% of people with DLB and may present early on when the person is experiencing only mild cognitive symptoms. These hallucinations are typically vivid and feature people, children or animals.

TIP: *If visual hallucinations occur with mild dementia, it is suggestive of DLB over AD.*

Parkinsonism

Most people with DLB will experience parkinsonism over the course of the disorder, though it may be very subtle or not clinically apparent in the early stage. As such, it is not required for diagnosis. In addition to bradykinesia, only one cardinal feature of Parkinson's disease (rest tremor, rigidity or postural instability) is needed to document the presence of parkinsonism in DLB.

TIP: A dopamine transporter uptake scan may be helpful if the presence of parkinsonism is in doubt.

Rapid Eye Movement Sleep Behavior Disorder (RBD)

RBD, a sleep disorder in which people physically act out their dreams, can present years or even decades before the onset of other DLB symptoms. While the patient often does not know they are moving, a bed partner can be a helpful informant to determine if they are thrashing about violently and injuring themselves or someone else in the bed. A polysomnogram is the gold standard in diagnosis RBD.

TIP: RBD is highly associated with Lewy body disorders, but not Alzheimer's disease.

Supportive Clinical Features

The table on Page 2 lists clinical features that support a DLB diagnosis.

TIP: Screen for all of these as part of the diagnostic process. Comprehensive treatment of DLB symptoms improves quality of life for the patient and their primary caregiver.