

LEWY BODY DEMENTIA DIAGNOSIS

Neurological exam

A thorough neurological examination should be conducted by an experienced clinician at the time of the initial assessment. Areas of importance include:

- Cognitive function, including language and speech
- Eye movements (can be abnormal in some types of atypical parkinsonism)
- Gait, balance, fine/coarse motor movements, reflexes
- Presence of involuntary movements such as tremor
- Alteration of smell

Blood pressure should be taken at every visit to assess whether blood pressure drops significantly when a person moves from sitting or lying to standing (a measure of an impaired autonomic nervous system). As hallucinations are common, evaluation of vision and hearing can establish baseline acuity. Assessment of a person's functional status, including the ability to perform activities of daily living (bathing, dressing) and instrumental activities of daily living (managing money, shopping), can provide insight into the ability of patients to care for themselves.

Clinical follow-up should be done in 6 month intervals or whenever changes are reported by the patient or family.

Brief cognitive assessments

Select a brief standardized instrument sensitive for both amnesic and non-amnesic cognitive decline. The Montreal Cognitive Assessment (MoCA) has become the standard, whereas the Folstein Mini-Mental State Exam (MMSE) is also available. Each of these tests take 5-10 minutes to administer.

Psychiatric symptoms are also common and may include anxiety, depression, and psychotic features such as hallucinations and delusions. REM sleep behavior disorder (dream enactment) frequently precedes the observed onset of LBD and the sleep partner should be asked about a history of acting out dreams.

Any ambiguous abnormalities will require referral for a more in-depth evaluation by a neuropsychologist.

Blood tests and imaging

Reversible causes of dementia (e.g. post-traumatic hydrocephalus, drug and alcohol toxicity, electrolytes, B12 deficiency, HIV, thyroid disorders), though rare in this setting, should be screened for and treated if confirmed.

Imaging by computed tomography (CT) or magnetic resonance (MRI) should be done to rule out stroke, brain tumors, intracranial bleeding, hydrocephalus or other structural causes of dementia. Imaging in DLB is usually normal. Dopamine imaging of the brain can be done when indicated to differentiate LBD (DLB and PDD) from Alzheimer's disease by using single photon emission computerized tomography (SPECT). The scan in LBD usually shows reduced uptake of an injected radioactive compound scan compared with a normal result in Alzheimer's disease.

Research is under way to identify neuro-imaging tests, and brain, spinal fluid and serum proteins that can be used to help identify various types of dementia.

DLB Diagnostic criteria

Consensus criteria for possible and probable DLB (Table 1) were developed and updated most recently in 2005 by the Consortium on DLB International Workshop¹ for the clinical diagnosis of DLB.

Table 1: Features of dementia with Lewy bodies

The diagnostic criteria for **probable DLB** require:

- The presence of dementia
- At least two of three core features:
 - fluctuating attention and concentration,
 - recurrent well-formed visual hallucinations, and
 - spontaneous parkinsonian motor signs.

Suggestive clinical features include:

- Rapid eye movement (REM) sleep behavior disorder
- Severe neuroleptic sensitivity
- Low dopamine transporter uptake in basal ganglia demonstrated by SPECT or PET imaging

*In the absence of two core features, the diagnosis of **probable DLB** can also be made if dementia plus at least **one suggestive feature** is present **with one core feature**. **Possible DLB** can be diagnosed with the presence of **dementia plus one core or suggestive feature**.*

Supportive clinical features include repeated falls, syncope, a transient loss of consciousness, severe autonomic dysfunction, depression, systematized delusions, or hallucinations in other sensory and perceptual modalities. While these features may support the clinical diagnosis, they lack diagnostic specificity and can be seen in other neurodegenerative disorders.

A diagnosis of DLB is less likely

- In the presence of cerebrovascular disease evident as focal neurologic signs or on brain imaging
- In the presence of any other physical illness or brain disorder sufficient to account in part or in total for the clinical picture
- If parkinsonism only appears for the first time at a stage of severe dementia.

PDD Diagnostic criteria

A consensus statement² by a task force from the Movement Disorder Society for the diagnosis of PDD was published in 2007, providing criteria for probable and possible PDD (Table 2).

A diagnosis of probable PDD requires the core features and a typical presentation of clinical features which is defined as having deficits in at least two out of four cognitive domains. There may or may not be behavioral symptoms, although their presence would support a diagnosis of probable PDD. There must not be any features present from groups III and IV, as the abnormalities and conditions described in these categories can cause too much uncertainty in a potential diagnosis.

A diagnosis of possible PDD also requires the core features, but can have a more non-characteristic pattern of symptoms in at least one of the cognitive domains. There may or may not be any behavioral symptoms. One or more features of group III may be present, and none in Group IV.

Table 2. Features of dementia associated with Parkinson's disease

Group I - The core feature requires a prior diagnosis of Parkinson's disease, and dementia causing a decline in function severe enough to impair the patient in daily activities and in at least one cognitive domain.

Group II - The clinical features include both the cognitive and behavioral domains described below:

Cognitive domains:

- Attention – The patient shows a level of impairment in attention, which may fluctuate over time
- Executive function – Impairment in complex thought processes such as in initiating an action, planning, or organization
- Visuo-spatial ability – Marked deficits in the processing of visuo-spatial material
- Memory – There is noticeable impairment in both the recall of existing memories and in the learning of new material
- Language – Basic language features are largely intact, although there may be difficulties in finding words and understanding complex sentences.

Behavioral domains:

- Apathy – Decreased spontaneity, motivation, effortful behavior
- Changes in personality and mood – Can include depression and anxiety
- Hallucinations - Usually complex and visual
- Delusions - Usually paranoid delusions, such as infidelity or perceived unknown guests in the home
- Excessive daytime sleepiness

Group III - The third category includes two features that will not rule out a diagnosis of PDD, but may make the diagnosis more uncertain:

- Existence of an abnormality such as vascular disease which causes cognitive impairment although not determined to cause dementia
- If the duration of time between the onset of motor and cognitive symptoms is not known

Group IV - The last domain contains two features which suggest that other existing conditions impair the patient's cognitive functioning to such an extent that reliable diagnosis of PDD becomes impossible.

1. Cognitive or behavioral symptoms which occur only in the context of existing conditions, such as systemic diseases, drug intoxication, or major depression
2. Symptoms compatible with vascular dementia, confirmed by an established relationship between brain imaging results and impairment in neurological testing

References

- 1 McKeith IG, Dickson DW, Lowe J, *et al.* Diagnosis and management of dementia with Lewy bodies: third report of the DLB Consortium. *Neurology* 2005;65:1863-72.
- 2 Emre M, Aarsland D, Brown R, *et al.* Clinical diagnostic criteria for dementia associated with Parkinson's disease. *Mov Disord* 2007; 22:

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