NEUROCOGNITIVE DISORDERS

Major or Mild Neurocognitive Disorder with Lewy Bodies

DLB

DSM-5-TR 331.83 / 331.84 | ICD-10-CM G31.83, G31.84 WITH F02.8X

Alpha-synuclein dementia with fluctuating cognition, visual hallucinations, REM sleep behavior disorder, parkinsonism, and neuroleptic sensitivity.

SHARE OF DEMENTIAS	TYPICAL ONSET	SEX RATIO	COURSE
~5% clinical, 15-20% autopsy	Age 50-85; peak mid-70s	~1.5:1 male:female	5-8 yr median survival

CLINICAL PICTURE

- Cognitive fluctuation dominates: episodes of staring, incoherent speech, and daytime drowsiness that families describe as good days and bad days.
- Recurrent well-formed visual hallucinations, typically of people or animals, are often non-threatening and accompanied by preserved insight early on.
- Deficits are attentional, executive, and visuospatial rather than amnestic, so clock drawing and pentagon copying fail well before memory does.
- REM sleep behavior disorder with dream enactment can precede cognitive decline by 10 or more years and is frequently the earliest red flag.
- Parkinsonism is symmetric and bradykinetic with less resting tremor; gait instability and repeated falls are common at first presentation.
- Severe antipsychotic sensitivity, dysautonomia with orthostasis and syncope, constipation, and hyposmia complete the systemic picture.

DIAGNOSTIC CRITERIA AT A GLANCE

- Major or mild neurocognitive disorder is established first, then Lewy body etiology is assigned as probable or possible from supporting features.
- Core features are fluctuating cognition with variable attention, recurrent formed visual hallucinations, and spontaneous parkinsonism after cognitive decline.
- Probable diagnosis requires two core features, or one core feature plus a suggestive marker such as REM sleep behavior disorder or low dopamine transporter uptake.
- Onset is insidious with gradual progression, which separates the syndrome from delirium and from abrupt vascular events.
- The one-year rule separates this disorder from Parkinson disease dementia: cognitive decline must begin before or within a year of motor signs.

NEUROBIOLOGY & PHYSIOLOGY

- Misfolded **alpha-synuclein** aggregates as Lewy bodies in brainstem, limbic, and cortical neurons, and most cases also carry Alzheimer copathology.
- Cholinergic loss from nucleus basalis degeneration exceeds that seen in Alzheimer disease and underlies the fluctuations and visual hallucinations.
- Nigrostriatal dopamine loss produces the parkinsonism, and **DaTscan** shows reduced striatal dopamine transporter uptake with roughly 78% sensitivity.
- Occipital hypometabolism on **FDG-PET** with a relatively preserved posterior cingulate island sign is a characteristic imaging signature.
- Synuclein pathology in the subcoeruleus abolishes REM atonia, and polysomnography documents REM sleep without atonia underlying dream enactment.
- **GBA** and **SNCA** variants raise risk, and cardiac **MIBG** scintigraphy shows reduced uptake reflecting sympathetic denervation.

PSYCHOLOGICAL MECHANISMS

- Insight is often preserved early, so patients experience genuine distress and shame about hallucinations they can correctly identify as unreal.
- Fluctuating capacity confuses families and clinicians, and lucid intervals are frequently misread as volitional or attention-seeking behavior.
- Visuoperceptual failure generates illusions and misidentification, so patterned wallpaper, mirrors, and low lighting reliably worsen symptoms.
- Depression and apathy affect roughly 40% and are often prodromal, preceding recognized cognitive decline by several years.
- Caregiver burden exceeds that in Alzheimer disease because of falls, dream enactment injury, hallucinations, and unpredictable day-to-day variability.

DIFFERENTIAL & COMORBIDITY

- Severe antipsychotic sensitivity occurs in up to 50%: rigidity, immobility, sedation, worsened confusion, and neuroleptic malignant syndrome that can be fatal.
- Avoid **haloperidol** and every first-generation agent; if psychosis is dangerous, use low-dose **quetiapine** or **clozapine** with documented consent.
- Differentiate from Alzheimer disease by early visuospatial and attentional loss, and from Parkinson disease dementia by the one-year rule.
- Delirium mimics the fluctuation, so screen for infection, dehydration, and anticholinergic burden before attributing change to disease progression.
- Comorbid depression, orthostatic hypotension with falls, and constipation are near-universal and drive most avoidable hospitalizations.

Treatment

EVIDENCE-BASED MANAGEMENT

PHARMACOLOGIC

- **Rivastigmine 6-12 mg/day** oral or the **9.5 mg/24 h patch** is first-line and yields larger gains in cognition and hallucinations than in Alzheimer disease.
- **Donepezil 5-10 mg/day** is a reasonable alternative; monitor for bradycardia, syncope, gastrointestinal upset, and worsening dream enactment.
- Antipsychotics carry a boxed mortality warning plus severe sensitivity here, so if unavoidable use **quetiapine 12.5-100 mg** nightly with informed consent.
- **Melatonin 3-12 mg** at bedtime is first-line for dream enactment, while **clonazepam 0.25-1 mg** works but adds fall, sedation, and confusion risk.
- Treat parkinsonism cautiously with low-dose **carbidopa-levodopa**, since dopaminergic agents worsen hallucinations and orthostatic blood pressure drops.

PSYCHOTHERAPY

- Psychoeducation covering fluctuation, hallucinations, and the danger of antipsychotics is the highest-yield intervention for patient and caregiver alike.
- Hallucination coping strategies include reality testing, distraction, brighter lighting, and removing mirrors and visually ambiguous patterns.
- **Cognitive stimulation therapy** delivered in roughly 14 group sessions has modest evidence in mild to moderate dementia and is recommended by NICE.
- Adapted **CBT** or behavioral activation helps depression and anxiety in mild disease when sessions are shortened and heavily externally cued.
- Structured caregiver interventions such as **START** reduce caregiver depression and delay institutional placement.

ADJUNCT & SUPPORTIVE

- Make the bedroom safe for dream enactment: lower or pad the bed, clear nightstands, consider separate beds, and secure windows and firearms.
- Manage falls and orthostasis by checking lying and standing pressures, deprescribing antihypertensives, and adding compression, salt, and physical therapy.
- Aggressively remove anticholinergics, benzodiazepines, and sedating antihistamines, which markedly amplify confusion and fluctuation in this population.
- Track cognition with the **MoCA** and behavior with the Neuropsychiatric Inventory, since informant report captures fluctuation better than office testing.
- Flag antipsychotic sensitivity in the chart and on a medical alert bracelet, and involve palliative care early given the aggressive trajectory.

CLINICAL PEARLS

- Neuroleptics can kill in DLB: no haloperidol, ever. Quetiapine or clozapine only, lowest dose.
- Dream enactment plus formed visual hallucinations means DLB until proven otherwise.
- Cholinesterase inhibitors work better here than in Alzheimer disease.

References

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NOTE

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