Clinical Summary

Dementia with Lewy bodies

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ICD codes
ICD-9:
Dementia with Lewy bodies: 331.82

ICD-10:
Unspecified dementia: F03

OMIM
Dementia, Lewy Body; DLB: #127750

Synonyms
Cortical Lewy body disease; Diffuse Lewy body disease; DLB; DLBD; Lewy body dementia; Lewy body variant of Alzheimer disease; Senile dementia of Lewy body type; Synucleinopathy

Historical note and nomenclature

The first known description of abnormal deposits of protein, later termed Lewy bodies, dates back to 1912, when Fritz Heinrich Lewy (1885-1950) detailed these neuronal inclusion bodies that would later bear his name. His findings were first published in the *Handbook of Neurology*, compiled by Max Lewandosky in 1912 (Lewy 1912). The following year it was presented to the German Association of Psychiatrists and Neurologists in Breslau. In 1923, he published these findings in his own *Tonus and Bewegund* (Muscle Tone and Movement). In 1919, C. Tretiakoff was the first to ascribe the name "corps de Lewy" or "Lewy bodies" to the inclusions. Later, in 1961, Okazaki and colleagues described patients with dementia who had Lewy bodies in the neocortex (Okazaki et al 1961), but it was not until the mid-1980s that immunocytochemical methods to better identify cortical Lewy bodies were developed (ubiquitin and later alpha-synuclein immunocytochemical assays) (Holdorff 2002; Sweeney 1997).

The nomenclature for the combination of dementia with neocortical Lewy bodies evolved in 1989 with a proposal made by Perry and colleagues, who called it "senile dementia of Lewy body type" (Perry et al 1989). In 1990, Hansen and colleagues proposed "Lewy body variant

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of Alzheimer's disease" (Hansen et al 1990), and in 2000, Kosaka suggested it be named "diffuse Lewy body disease" (Kosaka 2000). In 1995, there was consensus among an international consortium of researchers that the phrase "dementia with Lewy bodies" be used for clinical diagnosis. Guidelines for clinical and pathologic diagnosis of dementia with Lewy bodies were established by this group. There have been 2 additional meetings (1999 and 2005) by this consortium of experts to refine these diagnostic guidelines (McKeith IG 1996; 1999; 2005).

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Clinical manifestations

Dementia with Lewy bodies is the second most common form of neurodegenerative dementia, accounting for up to 20% of cases in the elderly (Baskys 2004). It is characterized by fluctuating cognitive impairment, parkinsonism, and recurrent visual hallucinations. Consensus-based clinical criteria have been published and have been shown to have high specificity but still lack sensitivity (McKeith 2005) (see the criteria in the Diagnostic workup section). Patients can present with a wide range of signs and symptoms. Grouping the symptoms, as seen in Table 1, can facilitate the diagnosis.

Table 1. Clinical Features of Dementia with Lewy Bodies

Cognitive impairment

- forgetfulness (retrieval more impaired than recognition memory)
- apathy
- psychomotor slowing
- impairments of attention
- visual-spatial impairments
- subcortical dementia features
- increased risk of delirium

Neuropsychiatric features

- hallucinations
- delusions
- behavioral dyscontrol
- agitation
- aggression
- nocturnal wandering
- disinhibition
- depression
- emotional lability
- pseudobulbar affect
- anxiety
- obsessive/compulsive

Motor dysfunction

- extrapyramidal features, typically symmetric akinetic-rigid form of parkinsonism with mild to moderate rigidity and bradykinesia, but typically not a classic asymmetric resting tremor. Mild symmetric postural and kinetic tremor is common.

Sleep disorder

- REM sleep behavior disorder
- insomnia
- restless legs syndrome
- periodic limb movements of sleep
- excessive daytime sleepiness

Autonomic Dysfunction

- orthostatic hypotension
- poorly regulated sweating
- poorly regulated body temperature

Adapted from: (Boeve 2004).

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Clinical vignette

A 68-year-old man presented for further evaluation of recent confusion during a hospitalization for a urologic procedure 1 month earlier. Upon review, the patient's family stated that he had been somewhat more forgetful in the past year and had shown signs of visual-spatial difficulties, for example, having difficulty parallel parking his car. The changes, however, had been too subtle to prompt them to seek medical attention. He had also been slower in his walking speed; for instance, the family had recently taken a trip, during which he had not walked as fast as the others in the group, which was atypical for him.

The hospitalization had been for an elective transurethral resection of the prostate. It was supposed to be a same-day procedure, but when the man awoke from anesthesia, he was agitated, disoriented, and hallucinating. He received Haldol and developed significant somnolence and rigidity that lasted 24 hours. The family stated that since his hospitalization, he had gradually improved, returning close to his baseline functioning.

On examination, the Mini-Mental State Examination (MMSE) was administered, yielding a score of 26 out of 30. He had difficulties with attention and constructional apraxia in his ability to draw intersecting pentagons and a clock face. He had intact delayed recall. On examination there were very subtle signs of parkinsonism, specifically, mild decreases in facial expression and volume of voice, traces of postural tremor but no resting tremor, and mild bilateral arm rigidity. His gait was abnormal, secondary to a stooped posture and decreased arm swing with walking. Stride length and gait speed were also decreased.

Upon further review of the history, the man's family described symptoms consistent with REM sleep behavior disorder, with yelling and intermittent acting out of his dreams, which had been present for the past several years, but not present prior to that point. These episodes had been becoming more frequent and severe. Since his hospital stay, he had had several occasions of visual hallucinations that had continued despite his otherwise overall improvement. He typically saw people, especially in the evenings or when he awoke from a daytime nap. He had also had some visual misperceptions since his hospital stay, where he would see things differently from others – for example, looking out the window and stating that it was raining when it was not.

Re-evaluation 6 months later showed continuing evidence of

parkinsonism and increasing cognitive impairment upon repeat testing (new MMSE score of 24). He was having increasing functional impairment in his everyday activities and still had intermittent visual hallucinations, especially in the evening hours. Diagnostic testing included MRI imaging of the brain, which showed mild generalized atrophy but no other structural cause of his symptoms. Laboratory tests included B12 and TSH, which were normal. Formal neuropsychological testing showed evidence of mild dementia with the subcortical pattern of deficits, including difficulty with executive cognitive function and attention and significant visual-spatial impairment. There were difficulties in memory, especially memory retrieval, but overall memory was relatively spared.

This patient's presentation and work-up is consistent with the diagnosis of dementia with Lewy bodies. The cognitive impairment and parkinsonism began approximately at the same time. He had evidence of dementia associated with parkinsonism and frequent visual hallucinations. By history, he had also had cognitive fluctuations. Other supportive features include REM sleep behavior disorder, susceptibility to an antipsychotic medication (Haldol), and susceptibility to delirium.

For management, the patient was placed on a cholinesterase inhibitor. This stabilized his cognitive function, and his hallucinations were less frequent. There appeared to be less frequent fluctuations in his symptoms. An antipsychotic medication was not needed. He was not having symptoms of depression. He was placed on a low dose of Klonopin at nighttime to suppress REM sleep behavior disorder symptoms and to improve his overall sleep.

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Etiology

The severity of cortical Lewy bodies correlates best with the degree of dementia in patients with dementia with Lewy bodies and is thought to be a marker for the underlying pathogenesis (see the Pathogenesis and pathophysiology section below). The disorders in the spectrum of Lewy body disorders that clinically includes dementia with Lewy bodies, Parkinson disease, and Parkinson disease with dementia are likely to share similar origins. There is also evidence that patients with dementia with Lewy bodies can demonstrate Alzheimer disease pathology or cerebrovascular disease pathology at autopsy. The cause of this overlap is poorly understood at this time (Yokota 2007).

As in other neurodegenerative diseases, it is suspected that environmental, genetic, and aging factors contribute to the risk of developing dementia with Lewy bodies (Tsuang 2004). Multiple cases within same families have been reported, but there does not seem to be a strong tendency for inheriting the disease (Brett 2002; Galvin 2002; Harding 2004). However, genetic studies are making some progress in revealing a matrix of different genes that may contribute to development of dementia with Lewy bodies and Lewy body disorders. This appears to be complex, but it may explain the relationship between dementia with Lewy bodies and the other primary Lewy body disorders, including Parkinson disease, and its association with Alzheimer disease. Also, there is genetic evidence that shows an overlap between dementia with Lewy bodies and Parkinson disease (Tsuang 2004). In addition, some genetic forms of Alzheimer disease have cortical Lewy bodies (Leverenz 2006). Some genes associated with Lewy bodies and dementia with Lewy bodies include the following:

- Alpha-synuclein (SNCA; 163890)
- Beta-synuclein (SNCB; 602569)
- APOE gene (107741)
- CYP2D6 gene (124030)
- Familial Parkinson disease-1 (PARK1; 168601)
- LRRK2 gene (609007)
- Parkinson disease-8 (PARK8; 607060)
- PRNP gene (M232R; 176640.0017)

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Pathogenesis and pathophysiology

In dementia with Lewy bodies, the protein alpha-synuclein aggregates into fibrils in Lewy bodies. The molecular characteristics of these aggregates are indistinguishable in dementia with Lewy bodies, Parkinson disease, and Parkinson disease with dementia, but their distribution differs. Alpha-synuclein aggregates by unknown mechanisms, but it is hypothesized that overproduction of alpha-synuclein or production of abnormal forms of the protein lead to abnormal aggregation (Eriksen 2003). There are thought to be multiple and complex mechanisms involved. Currently it is thought that Lewy bodies and alpha-synuclein cause pathologic conditions due to the following:

- Repeats or mutations in the alpha-synuclein gene cause familial dementia with Lewy bodies.
- Filamentous alpha-synuclein aggregates in Lewy bodies and Lewy neurites contain abnormally nitrated, phosphorylated, and ubiquitinated residues.
- Alpha-synuclein transgenic animal models develop a neurodegenerative disease with filamentous alpha-synuclein deposits.
- Cortical Lewy bodies correlate with the dementia in Parkinson disease with dementia and dementia with Lewy bodies.
- Double transgenic mice overexpressing human familial Alzheimer disease mutant alpha-synuclein show an augmentation in alpha-synuclein pathologies.
- Coexpression of heat shock proteins consisting of beta-synuclein with alpha-synuclein in mice ameliorates the disease phenotype.
- Antibodies specific to alpha-synuclein detect Lewy bodies and Lewy neurites, which are the hallmarks for amyloids of dementia with Lewy bodies (Spillantini 1997).
- It has been demonstrated that aggregated alpha-synuclein activates microglia and an inflammatory response as one mechanism of neurodegeneration (Zhang 2005).
- Ohtake and colleagues found that an alteration in beta-synuclein (SNCB) may impair its normal inhibitory action on the formation of alpha-synuclein fibrils (Ohtake et al 2004).

Pathologic findings. A Lewy body is composed of the protein alpha-synuclein along with ubiquitin, neurofilament protein, and alpha B crystallin. Khachaturian reported an autopsy series of elderly individuals with dementia and found that the second most common pathology after the senile plaques and neurofibrillary tangles of Alzheimer disease was Lewy bodies found in subcortical and cortical regions (Khachaturian 1995). Pathologically, Lewy bodies are present in a pattern more widespread than usually observed in Parkinson disease. In addition to Lewy bodies, patients with dementia with Lewy bodies have neuronal

loss, basal forebrain cholinergic deficits, Alzheimer disease, and vascular pathology. Some patients with such Lewy body dementia may also have a sufficient number of hippocampal and neocortical senile plaques to meet the diagnostic criteria for Alzheimer disease. Hansen and colleagues referred to such patients as having the "Lewy body variant of Alzheimer disease" (Hansen et al 1990). The term "diffuse Lewy body disease" is reserved for patients with brainstem and cortical Lewy bodies but an insufficient number of senile plaques to fulfill the diagnostic criteria for Alzheimer disease.

Wakabayashi and colleagues reported that pathologic examination of their 2 patients showed marked neuronal loss with Lewy bodies in the brainstem, pigmented nuclei, numerous cortical Lewy bodies, and ubiquitin-positive hippocampal neuritis (Wakabayashi et al 1998). Neuropathology of the proband reported by Ohara and colleagues demonstrated numerous Lewy bodies in the cerebral cortex and brainstem with no neurofibrillary tangles or neuritic plaques (Ohara et al 1999). Postmortem study revealed that cardiac sympathetic denervation occurs in Lewy body disease and indicates the presence of Lewy body pathology, which accounts for the decreased cardiac uptake of MIBG in Lewy body disease.

Additionally, these studies support current research showing an ascending pattern of Lewy body progression from brainstem to basal areas of the brain. Damage to these structures in dementia with Lewy bodies may affect a number of different neurochemical messenger systems, which possibly contributes to the clinical features of dementia with Lewy bodies (Whitwell et al 2007).

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Epidemiology

According to McKeith, the age of onset ranges from 53 to 83 years old, with death occurring between the ages of ages 68 and 92 (McKeith 2002). Median age at death for dementia with Lewy bodies is 78.0 years as compared to Alzheimer disease, which is 84.6 years (Williams 2006).

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Prevention

There are no known preventive measures.

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Differential diagnosis

- Alzheimer disease
- Parkinson disease
- Corticobasal ganglionic degeneration
- Frontotemporal dementia
- Hydrocephalus
- Prion-related diseases (eg, Creutzfeldt-Jakob disease)
- Progressive supranuclear palsy
- Frontotemporal dementia with parkinsonism linked to chromosome 17

(Also see Diagnostic workup section.)

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Diagnostic workup

Currently there is no single test to diagnose dementia with Lewy bodies; essentially it becomes a diagnosis of exclusion. Clinically, dementia with Lewy bodies is characterized by progressive cognitive impairment with fluctuating course, recurrent visual hallucinations, and parkinsonism. Although formal clinical criteria have been proposed, there is a pronounced clinical and neuropathologic overlap with Alzheimer disease as well as Parkinson disease with dementia. Four items of interest in the patient history suggesting dementia with Lewy bodies would be 1) acute-subacute onset, 2) early parkinsonism, 3) early hallucinations, and 4) early onset of urinary incontinence (Del Ser et al 2001). The relationship of dementia with Lewy bodies and Parkinson disease is an area of considerable controversy, particularly because dementia frequently occurs in Parkinson disease. Many investigators believe that a spectrum of Lewy body disorders exists. They use an arbitrary 1-year rule to distinguish dementia with Lewy bodies from Parkinson disease with dementia. If parkinsonism has been present for 12 months or longer before cognitive impairment is detected, the disorder is called Parkinson disease with dementia; otherwise, it is called dementia with Lewy bodies (Boeve 2004). Evaluation should begin with a complete medical history, including the patient's general health, past medical history, and family history. A possible diagnosis of dementia with Lewy bodies is more likely if the patient reports repeated falls, fainting, brief losses of consciousness, delusions, or is sensitive to antipsychotic drugs that are given to control hallucinations and other psychiatric symptoms. Blood tests may be done to rule out other causes of the dementia, such as vitamin B12 deficiency, thyroid deficiency, syphilis, or human immunodeficiency virus (HIV). Signs of inflammation may suggest infection, or certain biomarkers may suggest cancer, an autoimmune disorder, or possibly Creutzfeldt-Jakob disease (Knopman et al 2001). Cerebrospinal fluid should be sent for the 14-3-3 protein if a prion disease is suspected. A mental status evaluation should evaluate for deficits in orientation, attention, memory, and visual-spatial function, which are common. Neuropsychological testing will be especially helpful in trying to detect dementia at an early stage and typically can quantify a specific pattern of deficits that is supportive of a diagnosis of dementia with Lewy bodies. These include deficits of attention, executive function, memory (especially retrieval), and visual-spatial processing. Neuroimaging will help in ruling out structural causes of dementia. Specifically, CT scan or MRI scan should be used for ruling out abscess, normal pressure hydrocephalus, tumor, stroke, and subdural hematoma. MRI will often show cortical atrophy, but the extent of hippocampus atrophy is less than that seen in Alzheimer disease. PET scan and single photon emission computed tomography (SPECT) can detect temporal, parietal, and occipital hypoperfusion. SPECT brain imaging with the ligand (123) I-2beta-carbometoxy-3beta-(4-iodophenyl)-N-(3-fluoropropyl) nortropane (123)I-FP-CIT) can help in the diagnosis of dementia with Lewy bodies (McKeith et al 2007) The pattern is similar to that seen in Alzheimer disease and Parkinson disease with dementia, but on average there is greater occipital hypometabolism seen in dementia with Lewy bodies than in Alzheimer disease. EEG may show background slowing, but it is not diagnostic. Polysomnography will show an increase in electromyography tone associated with dream enactment behavior

during REM sleep to support a diagnosis of REM sleep behavior disorder or may identify evidence of periodic limb movements of sleep (Boeve et al 2004).

In 2003, the International Consortium on Dementia with Lewy Bodies revised the guidelines for the clinical and pathologic diagnosis of dementia with Lewy bodies.

Table 2. Guidelines for the Clinical and Pathologic Diagnosis of Dementia with Lewy Bodies

Central features (essential for the diagnosis of possible or probable dementia with Lewy bodies).

- Dementia defined as progressive cognitive decline of significant magnitude to interfere with normal social or occupational function.
- Prominent or persistent memory impairment may not necessarily occur in the early stages, but it is evident with progression.
- Deficits on tests of attention, executive function, or visual-spatial ability may be especially prominent.

Core Features (Two core features are sufficient for the diagnosis of probable dementia with Lewy bodies, 1 for possible dementia with Lewy bodies.)

- Fluctuating cognition with pronounced variation in attention and alertness.
- Recurrent visual hallucinations that are typically well formed and detailed.
- Spontaneous features of parkinsonism.

Suggestive features (If 1 or more of these is present in the presence of one or more core features, diagnosis of probable dementia with Lewy bodies can be made. Diagnosis of probable dementia with Lewy bodies should not be made on the basis of suggested features alone.)

- REM sleep behavior disorder
- Severe neuroleptic sensitivity
- Low dopamine transporter uptake in the basal ganglia demonstrated by SPECT or PET imaging

Supportive features (commonly present but not proven to have diagnostic specificity)

- Repeated falls and syncope
- Transient unexplained loss of conscious
- Severe autonomic dysfunction, orthostatic hypotension, and urinary incontinence
- Hallucinations other than visual
- Systemized delusions
- Depression
- Relative preservation of medial temporal lobe structures on CT/MRI scan
- Generalized low uptake on SPECT/PET perfusion scan with reduced occipital activity
- Low uptake of MIBG scintigraphy
- Distinct slow-wave activity of EEG with temporal lobe

transient sharp waves

Diagnosis of dementia with Lewy bodies is less likely:

- In the presence of significant cerebrovascular disease, evident on neurologic examination or brain imaging, that is sufficient to explain cognitive and motor features.
- 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.

Temporal sequence of symptoms

Dementia with Lewy bodies should be diagnosed when dementia occurs before or along with parkinsonism. The timing of symptoms is a reliable clue: if both mental and motor symptoms appear within 1 year of each other, dementia with Lewy bodies is more likely the cause.

The sensitivity of an initial clinical diagnosis of dementia with Lewy bodies was reported to be 75% and the specificity was reported to be at 42%. The sensitivity of the (123)I-FP-CIT scan for diagnosing dementia with Lewy bodies was 88%, and the specificity was 100%.

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Prognosis and complications

Dementia with Lewy bodies is a neurodegenerative disorder that results in progressive intellectual and functional deterioration. There are no known therapies to alter the course of dementia with Lewy bodies. Average survival after the time of diagnosis is about 8 years. Importantly, patients with this form of dementia often respond well to medications that may improve the cognitive and neuropsychiatric symptoms and, to a lesser extent, motor symptoms. It will inevitably progress over time, which will debilitate the patient even more.

Lewy body dementia progresses into severe dementia and ultimately eliminates a person's ability to speak or move. The disease, which typically lasts 8 years, leads to fainting and fluctuations in blood pressure. The most common cause of death is pneumonia or other primary infection. This is usually brought on by dysphagia, inhaling food or drink into the airway, from a catheter inserted into the body, and/or from becoming bedridden. Infection also may be introduced by a urinary catheter (urinary tract infection). As the disease advances, people may lose all ability to care for themselves. They may have difficulty eating, become incontinent, or be unable to take a walk and find their way back home. Other problems will come about by falls and related injuries. Patients may become disoriented or have tremor or gait problems. These lead to head injuries and fractures. The surgical risks increase in the elderly, as does the risk of pulmonary embolus with prolonged immobilization.

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Management

Pharmacologic. There have been few placebo-controlled clinical trials of medications to treat patients with dementia with Lewy bodies. One area of focus has been on enhancing cholinergic neurotransmission in patients with dementia with Lewy bodies. There is post-mortem

evidence of a significant decrease in neocortical cholinergic enzymatic activity, which is greater than that seen in Alzheimer disease patients. This supported a clinical trial of the cholinesterase inhibitor rivastigmine for the treatment of patients with a clinical diagnosis of dementia with Lewy bodies. This study randomized 120 patients to placebo or rivastigmine for 20 weeks and examined their performance on neuropsychological tests and a computerized cognitive assessment system and measured their neuropsychiatric symptoms (McKeith et al 2000). Twice as many patients on rivastigmine (63%) than on placebo (30%) showed at least a 30% improvement from baseline on the computerized cognitive assessment program and neuropsychological tests, especially tasks with a substantial attentional component. There was no significant difference in MMSE scores at the end of the trial, but there was a trend toward improvement in the MMSE score in the rivastigmine group (1.5 point improvement vs. 0.1 point decline, p=0.072). There was a significant improvement in neuropsychiatric symptoms as measured by the total Neuropsychiatric Inventory score (NPI-10) at study end in those receiving rivastigmine (3.8 point improvement, p=0.048), with significant improvements in apathy, anxiety, delusions, and hallucinations. Side effects that were significantly more common in the treatment group included nausea, vomiting, anorexia, and somnolence, as has been seen in other trials of cholinesterase inhibitors. There have been open-label trials of other cholinesterase inhibitors (eg, donepezil, galantamine) for the treatment of patients with dementia with Lewy bodies that have suggested similar benefits, but there have been no other placebo-controlled trials of cholinesterase inhibitors for the treatment of patients with dementia with Lewy bodies.

Psychiatric symptoms, including hallucinations, delusions, anxiety, and depression, are a source of significant morbidity for patients with dementia with Lewy bodies, but there has been little research into the most effective medications to treat these symptoms. In patients with severe psychotic symptoms, including delusions and hallucinations, an antipsychotic medication trial should be considered. Neuroleptic antipsychotics should be avoided because of the risk of worsening parkinsonism and the risk of neuroleptic hypersensitivity reactions. On the other hand, the atypical antipsychotic class (eg, clozapine, quetiapine, aripiprazole, risperidone, olanzapine, ziprasidone) is associated with less risk of worsening parkinsonism. There have been placebo-controlled trials of this class of medications for the treatment of psychosis in patients with Parkinson disease, but not specifically in patients with dementia with Lewy bodies (Friedman and Fernandez 2002; Miyasaki et al 2006). The literature supports the use of clozapine or quetiapine for psychosis in patients with Parkinson disease, but it does not support the use of olanzapine in this situation (Miyasaki et al 2006). It is assumed that similar recommendations would apply to patients with dementia with Lewy bodies, but rigorous evidence in these patients is lacking. The potential benefits of atypical antipsychotic medications need to be weighed against potential side effects of these drugs, including agranulocytosis seen rarely with clozapine and the potential for somnolence, worsening of gait or parkinsonism, cognitive decline, and the small increase in risk of cerebrovascular events or death that has been reported with drugs in the class when used to treat patients with dementia (Schneider et al 2005; 2006). There have been no placebo-controlled trials of medications to treat symptoms of

depression and anxiety specifically in patients with dementia with Lewy bodies, despite the common occurrence of these symptoms. Clinical experience and open-label studies suggest that selective serotonin reuptake inhibitor (SSRI) antidepressants and serotonin-norepinephrine reuptake inhibitor (SNRI) antidepressants are safe and effective for the treatment of symptoms of depression and anxiety in patients with dementia with Lewy bodies.

A trial of levodopa should be considered in patients with dementia with Lewy bodies, if parkinsonism is causing significant functional disability. However, the clinical response to levodopa is often less dramatic in patients with dementia with Lewy bodies than in patients with idiopathic Parkinson disease (Molloy et al 2005). The dose of levodopa should be kept low, because dopaminergic stimulation may worsen neuropsychiatric symptoms, especially psychosis. Similarly, dopamine agonists should be avoided because of a higher risk of exacerbating neuropsychiatric symptoms, and anticholinergic medications should be avoided because of a high risk of causing confusion and memory loss in patients with dementia with Lewy bodies. Low-dose clonazepam (0.25mg to 0.5mg at bedtime) can be used to help improve symptoms related to REM sleep behavior disorder but may cause sedation and imbalance in some patients (Boeve et al 2004). For autonomic system dysfunction such as orthostatic hypotension, fludrocortisone and midodrine can be used (Boeve 2004).

Behavioral strategies. Not all cases of dementia with Lewy bodies will require pharmacologic intervention. Modifying daily tasks to encourage personal interaction may help reduce agitation and aggression (for example, sponge bathing a person in bed rather than having them take a bath or shower). Other management options would include physical therapy, massage therapy, and aromatherapy. Difficulties with sleep disturbances may be improved by reducing daytime naps and caffeine intake. Keeping patients stimulated throughout the day until bedtime may be helpful and necessary. Maintaining a predictable routine and avoiding overstimulating environments and situations can help decrease confusion and agitation. Exercise should be encouraged, as it gives opportunity for the patient and caregiver to spend time together. Exercise improves strength and cardiovascular health, helps with depression, and helps in daily motor functioning. It also helps sustain strength, flexibility, and balance, which can help lesson the injuries associated with possible falls (Lingler and Kaufer 2002). There is evidence that supports the use of psychological treatments for disruptive behaviors that occur in individuals with dementia. These can include behavioral problem-solving therapies that identify and modify antecedents and consequences of problem behaviors and individualized interventions based upon progressively lowered stress threshold models that include problem solving and environmental modification (Logsdon et al 2007).

Caregivers. Currently there is no cure for patients with dementia with Lewy bodies, so once dementia is apparent, it can be assumed that progressively increasing levels of care will be required. For family members, caregiving can become challenging and overwhelming. Specific education about dementia with Lewy bodies and its associated symptoms is important for caregivers to better understand support and management needs and treatment options and to anticipate future problems. Educational materials and support groups are available through national caregiver support organizations for both Alzheimer

disease and Parkinson disease. In addition, in many regions of the country there are psychologists or social workers who can provide individual counseling and education concerning behavioral management strategies that are specific to dementia caregivers. There is evidence-based support for several forms of psychological treatments to lower distress in family caregivers of older adults with dementia, including skill training programs focused on behavioral management, cognitive behavioral therapy for the caregiver, and combinations of individual caregiver counseling and support group attendance (Gallagher-Thompson and Coon 2007). These approaches may all be used to help lower distress in caregivers of patients with dementia with Lewy bodies, especially those that focus on behavioral symptoms that are very common in patients with dementia with Lewy bodies and distressing to caregivers.

As with all patients with dementia, caregivers should ensure that the environment is safe for someone with dementia. Medicines, toxic household chemicals, alcohol, weapons, and power tools should be locked up. Wandering can become a problem and will require interventions such as a card with the patient's home phone number or phone number of a loved one. The patient should wear a bracelet with his or her name and phone number and identified with "Memory impaired." The Alzheimer's Association offers a 24-hour hot line (800-272-3900) that offers a Safe Return program.

Legal and financial issues should be discussed with the patient, their family, social workers, and an attorney. These include assigning a durable power of attorney to help with medical and financial decision in the future if the patient is unable to make decisions. Courts can appoint a guardian if the patient has progressed to moderate-to-severe dementia. Patients and families should discuss and record advanced directives for end-of-life care.

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Pregnancy

Not applicable.

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Anesthesia

Hemodynamic monitoring is important.

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Dementia in Parkinson disease
Dementia in progressive supranuclear palsy
Diffuse Lewy body disease with gaze palsy
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Progressive supranuclear palsy
Fronto-temporal dementia with parkinsonism linked to chromosome 17

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Demographics

For more specific demographic information, see the Epidemiology, Etiology, and Pathogenesis and pathophysiology sections of this clinical summary.

Age
45-64 years
65+ years

Population
None selectively affected.

Occupation
None selectively affected.

Sex
male>female, >1:1

Family history
Family history may be obtained.

Heredity
Heredity may be a factor and show autosomal dominant pattern.

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