



Fact Sheet:

Dementia with Lewy Bodies

Definition

Dementia with Lewy Bodies (DLB) is a progressive degenerative disease or syndrome of the brain. It shares symptoms and sometimes overlaps with several diseases especially Alzheimer's and Parkinson's.

People who develop DLB can have cognitive (thinking and memory) and behavioral symptoms similar to those of Alzheimer's disease and, to varying extents, to the motor and non-motor symptoms seen in Parkinson's Disease. However, there are some distinct contrasts to these conditions. The cognitive symptoms of a person with DLB might fluctuate from hour to hour or day to day, and attention and alertness may wax and wane. The motor symptoms may be similar to those features seen in Parkinson's, but their severity and treatment response may differ. Two of the most distinguishing symptoms of DLB from Alzheimer's disease include vivid visual hallucinations, particularly early in the course of the disorder, and a sleep disorder in which the person physically acts out his/her dreams, called REM sleep behavior disorder (RBM)

Caregiving and DLB

Treating and caring for a person with DLB requires frequent assessment and

reassessment. Some people with DLB can live at home with careful monitoring and supervision. Those with DLB who often experience repeat falls or fainting may benefit from a home safety assessment by an occupational or physical therapist. Ongoing attention to safety concerns from caregivers is also needed to minimize risk of falling. Particular care should be taken when a person with DLB is standing up from a chair or getting out of bed, as blood pressure can drop, causing the individual to become dizzy or lightheaded and lose his or her balance. Dementia can make it difficult for people to learn new actions that might help them overcome movement problems, such as using a cane, or walker. They may need more assistance on some days than other days. Some people with DLB can be reassured by a caregiver's help in turning attention away from the hallucinations, if present.

Caregivers must learn to navigate and adapt to cognitive, behavioral, and motor changes. Attending support groups and learning skills in how to communicate with someone with dementia, as well as learning skills in helping someone with a motor disorder, will reduce caregiver stress and frustration.

In California, caregivers can turn to a

California Caregiver Resource Center for assistance and to find a qualified diagnostic center for initial diagnosis and follow up. In California and other states, resources can be found through the Lewy Body Dementia Association's (LBDA) Research Centers of Excellence (RCOE), local and state offices on aging and health such as your Area Agency on Aging or the Alzheimer's Association in your area.

Facts

Dementia with Lewy Bodies is often called "Lewy Body Dementia" (LBD), which is the umbrella term that includes DLB and Parkinson's disease dementia. DLB is biologically related to Parkinson's disease, as both share overlapping disease processes in the brain and many clinical symptoms.

DLB is the second most common progressive dementia, accounting for twenty percent of those with dementia. (Alzheimer's disease is the most common). Dementia is a gradual, progressive decline in mental ability (cognition) that affects memory, thinking processes, and behavior, and performance of social, occupational, and daily activities.

The term DLB reflects the presence of Lewy bodies (the microscopic, smooth, round protein deposits made up of alpha-synuclein) that are found in the nerve cells of the affected parts of the brain. These "abnormal protein structures" were first described in 1912 by Frederick Heinrich Lewy, M.D., a contemporary of Alois Alzheimer, who identified clinical and pathological features of the more common form of dementia that now bears his name.

Lewy bodies are found throughout the outer layer of the brain (the cerebral cortex) and deep inside the midbrain and brainstem. They are often found in those diagnosed with Parkinson's approximately fifty percent of people with Alzheimer's and other disorders.

The cause of DLB is unknown. Risk factors include older age and the sleep disorder RBD, which may precede other LBD symptoms by years or even decades. Some cases of DLB have appeared among families, but there does not seem to be a strong tendency for inheriting the disease. Genetic research may reveal more information in the future about causes and risks. DLB usually occurs in older adults between 50- 85 years old, and slightly more men than women have the disease.

Symptoms

Initial symptoms of DLB are often similar to those of Alzheimer's or vascular dementia and are cognitive in nature, such as progressive decline in memory, judgment, planning and organizing, or acute confusion. Some people may first show the motor symptoms of parkinsonism – slowness, stiffness in muscles, tremors, decreased dexterity, or show, shuffling gait. Others may have visual hallucinations as the first symptom. Those with DLB may also suffer from delusions, depression, or autonomic (i.e., blood pressure, bladder, bowels) changes.

Key symptoms are:

- Problems in thinking such as inattention, lack of judgment, loss of insight, and decline in problem solving and organizational skills. People with DLB experience visuospatial difficulty (for

example, finding one's way around places), as well as trouble with finding the words they want to say, or following a conversation. Memory may be relatively intact in the early stage, but problems with attention and alertness may mimic memory problems.

- Fluctuation in the occurrence of cognitive symptoms from moment to moment, hour to hour, day to day, or week to week. For example, the person may converse normally one day and be more confused the next day. There are also fluctuations in attention, alertness and wakefulness.
- Recurrent, visual hallucinations. These hallucinations are well formed, vivid, and detailed. In DLB's early stage, the person may even acknowledge and describe the hallucinations. They can be benign and not frightening, but in some, can be troublesome. Hallucinations may also be auditory (hearing sounds), olfactory (smelling or tasting something), or tactile (feeling or touching something that is not there).
- Movement problems of parkinsonism, sometimes referred to as "extrapyramidal" signs. These motor symptoms often seem to start spontaneously and may include bradykinesia (slowness of movement), tremor, shakiness, loss of dexterity, rigidity (limb stiffness), flexed posture, shuffling gait, muscle jerks or twitches, reduced arm swing, a

tendency to fall, balance problems and lack of facial expression.

- Rapid Eye Movement Sleep Behavior Disorder. This is characterized by vivid dreaming, talking in one's sleep, and excessive movement while asleep, including occasionally hitting a bed partner. The result may be excessive daytime drowsiness, and this symptom may appear years before DLB is diagnosed. Over 50% of patients have this symptom.

Testing and Diagnosis

DLB can be difficult to diagnose. Not only does it resemble other dementias, it overlaps with Parkinson's and psychiatric disorders. Because no single test, such as a blood test or brain scan exists to diagnose DLB, a variety of medical, neurological, and neuropsychological tests are used to identify its symptoms and the possible overlap with other illnesses. A definitive diagnosis can only be made by an autopsy at death but probably and possible diagnoses can be made in life.

Although Lewy bodies are found in brains of patients with other diseases, it is useful to understand what happens to the brain of a person who has DLB. Three significant changes or pathological features are seen in brains afflicted by DLB:

- The brain's cerebral cortex (the outer layers of the brain) degenerates or shrinks. This brain tissue shrinkage (or atrophy) can affect reasoning and complex thinking, understanding personality, movement, speech

and language, sensory input, and visual perceptions of space. Degeneration also occurs in the limbic cortex at the center of the brain, which plays a major role in emotions and behavior. Lewy bodies form throughout these degenerating cortical areas.

- Nerve cells die in the midbrain region. This occurs especially in the substantia nigra in the brainstem, an area that also degenerates in Parkinson's disease. These cells are involved in making the neurotransmitter (brain messenger) dopamine. Lewy bodies are found in the nerve cells that remain. The midbrain is involved in motor control, sleep/wake, arousal, sensory and autonomic functions.
- Another cellular structure, Lewy neurites, that affect nerve cell function are found in the brains of people who have DLB, especially in the hippocampus, an area of the brain essential for forming new memories.

None of the symptoms found in DLB are specific only to this disorder. To address this problem, an international group of researchers and clinicians developed a set of diagnostic criteria called the *Consensus Guidelines*, most recently updated in 2017, that can reliably point to features of DLB:

Must be present:

- Progressive cognitive decline (decrease in thinking ability) that interferes with normal social or occupational activities. Memory problems do not necessarily occur in the early period but can

occur as DLB progresses. Attention, problem-solving, reasoning, and visual perceptions of space are likely to be impaired early.

Two of the following are present (one also indicates possibility of DLB):

- Fluctuating cognition: Mental problems vary during the day, especially attention and alertness.
- Visual hallucinations: Detailed and well-formed visions, which occur and recur.
- Parkinsonism: Movement problems.
- RBD: Physically acting out dreams while asleep.

A DLB diagnosis is even more likely if the individual also experiences any of the following: repeated falls, fainting, brief loss of consciousness, delusions, apathy, anxiety, problems with temperature and blood pressure regulation, urinary incontinence, chronic constipation, loss of smell, or sensitivity to neuroleptic medications that are given to control hallucinations and other psychiatric symptoms.

Finally, the timing of symptoms is a reliable clue: *if cognitive symptoms appear before or within a year of motor symptoms, DLB is more likely the cause than Parkinson's disease.* Signs of stroke or vascular dementia usually negate the likelihood of DLB.

Testing is usually done to rule out other possible causes of dementia, motor, or behavioral symptoms. Brain imaging (CT scan or MR imaging) can detect brain shrinkage and help rule out stroke,

fluid on the brain (normal pressure hydrocephalus), or subdural hematoma. Blood and other tests might show vitamin B12 deficiency, thyroid problems, syphilis, HIV, or vascular disease. Depression is also a common cause of dementia-like symptoms. Additional tests can include an electroencephalogram (EEG) or a spinal tap (lumbar puncture).

New research confirms several tests are highly associated with the presence of Lewy bodies in the brain and are now increasingly being used to make a DLB diagnosis. Scans using SPECT and PET technology can detect differences between DLB and Alzheimer's disease. A sleep study can confirm RBD, and special cardiac imaging can detect changes in cardiac nerves associated with DLB.

Alzheimer's and Parkinson's: Differences and Overlap with DLB

DLB's similarity to Alzheimer's and Parkinson's diseases and the fact that Lewy bodies are often found in the brains of patients with these diseases, means that clinicians must pay close attention to the factors that distinguish DLB. These factors include:

- Memory and other cognitive problems occur in both DLB and Alzheimer's. However, in DLB, they fluctuate frequently.
- In DLB, Alzheimer- like and Parkinson- like symptoms may appear within one year of each other.
- Hallucinations are experienced in the late stages of Alzheimer's and by people with Parkinson's

who take medications to improve movement and tremor. In DLB, hallucinations occur in early stages, even without Parkinson's medications, and they are frequent, vivid, and detailed.

- Neuroleptic drugs (sometimes called psychotropic drugs) prescribed to lessen the so-called psychiatric symptoms of dementia, such as hallucinations, agitation, or restlessness will induce or worsen Parkinson's-like symptoms in some DLB patients.
- Tremor is likely less pronounced in DLB than in Parkinson's. Also, individuals with DLB respond less dramatically to drugs such as levodopa that are used to treat Parkinson's.
- Some people with Parkinson's develop dementia in later stages. However, dementia is usually the presenting symptom in DLB.
- Someone with Parkinson's loses the neurotransmitter, dopamine; in Alzheimer's, people lose the neurotransmitter, acetylcholine. A person with DLB or Parkinson's disease dementia loses both.
- Life expectancy is slightly shorter for DLB than Alzheimer's and Parkinson's.
- At autopsy, the brains of those with DLB look essentially the same as a person with Parkinson's disease with dementia, as Lewy bodies are spread throughout the brain. People with DLB frequently have some co-existing changes of Alzheimer's disease in the brain as well.

Duration and Treatment

With an average lifespan after onset of 5 to 7 years, the progression of dementia with Lewy bodies is relentless; however, the rate of decline varies with each person. DLB does not follow a set pattern of stages as is seen in some other dementias. There is neither cure nor specific treatment to arrest the course of the disease. Death usually occurs from pneumonia or other illness.

While there are no medications currently approved to specifically treat DLB, there are many medications developed for other conditions that can help reduce some of its symptoms. A comprehensive treatment plan can improve the quality of life for a person with DLB and their family caregiver.

However, caution must be used in treating a person with DLB. Medications must be monitored closely for proper balance because some people are negatively affected by some drugs. Neuroleptic (tranquilizing) antipsychotic drugs such as haloperidol (Haldol) or thioridazine (Mellaril), some newer atypical antipsychotics (especially risperidone, olanzapine) as well as benzodiazepines (Valium, Ativan), and certain antihistamines like diphenhydramine (Benadryl) can cause extreme adverse reactions in those with DLB. Side effects include worsened motor-related symptoms, catatonia (non-responsiveness), loss of cognitive function, and/or development of muscle rigidity. These medications are sometimes used in Alzheimer's to help with hallucinations, agitation, and behavioral symptoms, but should not be used in individuals with DLB. Levodopa may be given to treat the parkinsonism, however, it may increase the

hallucinations in someone with DLB and aggravate other symptoms, such as confusion or low blood pressure. It is generally less effective in treating motor symptoms in those with DLB than in those with Parkinson's, especially since higher doses are rarely given. Dementia symptoms may be treated with medications originally developed for Alzheimer's disease, called cholinesterase inhibitors. Research shows these medications can be even more effective in people with DLB. As such, some of these treatments have been approved by regulatory agencies for Lewy body disorders; donepezil was approved in Japan for DLB and rivastigmine was approved by the US Food and Drug Administration (FDA) for treatment of dementia in Parkinson's disease. Treating hallucinations can be challenging since typical and atypical antipsychotics can worsen motor function; some newer antipsychotics with greater serotonin (another brain chemical) mechanisms (such as quetiapine and clozapine) are preferred by DLB experts to treat hallucinations in this disorder. Some antidepressants have also shown positive results, while others are counter-indicated. More research is urgently needed in this area to determine which medications are both safe and effective to treat hallucinations and delusions in DLB.

When considering surgery, families should meet with the anesthesiologist to discuss possible side effects of anesthesia, as persons with DLB are prone to delirium or confusion with anesthesia. Sometimes this 'stepping down' of cognitive ability noticed after anesthesia can linger or remain. In addition, families of those with DLB who are having surgery or who are hospitalized should discuss current

medications and any potentially anticipated medications with the healthcare professional team in advance; neuroleptics (antipsychotics) and some anti-nausea medications can worsen motor parkinsonism and should be avoided.

Resources

Southern Caregiver Resource Center

3675 Ruffin Road, Suite 230
San Diego, CA 92123
(858) 268-4432 | (800) 827-1008 (in CA)
Fax: (858) 268-7816
Email: scrc@caregivercenter.org
Website: www.caregivercenter.org

The Southern Caregiver Resource Center offers services to family caregivers of adults with chronic conditions in San Diego and Imperial counties. Services include specialized information, family consultation/ case management, respite care, short-term counseling, support groups, legal/ financial consultation, education and training, and employer resources.

Family Caregiver Alliance National Center on Caregiving

(415) 434-3388 | (800) 445-8106
E-mail: info@caregiver.org
Website: www.caregiver.org

Family Caregiver Alliance (FCA) seeks to improve the quality of life for caregivers through education, services, research and advocacy. FCA's National Center on Caregiving offers information on current social, public policy and caregiving issues; provides assistance in the development of public and private programs for caregivers; publishes timely reports, newsletters and fact

sheets; and assists caregivers nationwide in locating resources in their communities.

Lewy Body Dementia Association
www.lbda.org

This fact sheet was updated by Angela Taylor, Director of Programs at the Lewy Body Dementia Association and reviewed by Dr. Jennifer Goldman, Chair, Lewy Body Dementia Association Scientific Advisory Committee, ©2001, 2010, 2018 Family Caregiver Alliance. All rights reserved.

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