Southern Caregiver Resource Center

Caring for those who care for others

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.

Persons who develop DLB have behavioral and memory symptoms of dementia like those of Alzheimer's disease and, to varying extents, the physical, motor system symptoms seen in Parkinson's Disease. However, the mental symptoms of a person with DLB might fluctuate frequently, motor symptoms are milder than for Parkinson's disease, and DLB patients usually have vivid visual hallucinations.

Facts

Dementia with Lewy Bodies is also called "Lewy Body Dementia" (LBD), "Diffuse Lewy Body Disease," "Lewy Body Disease," "Cortical Lewy Body Disease," "Lewy Body Variant of Alzheimer's Disease," or "Parkinson's Disease Dementia." It is the second most common dementia accounting for twenty percent of those with dementia (Alzheimer's Disease is first). Dementia is a gradual, progressive decline in mental ability (cognition) that affects memory, thinking processes, and behavior and physical activity. In addition to these mental symptoms, persons with DLB experience physical symptoms of parkinsonism including mild tremor, muscle stiffness, and movement problems. Strong visual hallucinations also occur.

DLB is named after smooth round protein lumps (alpha-synculein) called Lewy bodies, that are found in the nerve cells of the affected parts of the brain. These "abnormal protein structures" were first described in 1912 by Frederich Heinrich Lewy, M. D., a contemporary of Alois Alzheimer who first identified the more common form of dementia that 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 Alzheimer's, Parkinson's, Down Syndrome, and other disorders.

The cause of DLB is unknown, and no specific risk factors are identified. Cases have appeared among families, but there does not seem to be a strong tendency for inheriting the disease. Genetic research may reveal more information about causes and risk in the future. It usually occurs in older adults between 50- 85 years old, and slightly more men than women have the disease.

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Symptoms

Initial symptoms of DLB usually are similar to those of Alzheimer's or Vascular Dementia and are cognitive in nature such as acute confusion, loss of memory, and poor judgment. Other patients may first show the neuromuscular symptoms of parkinsonism--loss of spontaneous movement, rigidity (muscles feel stiff and resist movement), and shuffling gait--while still others may have visual hallucinations as the first symptom. Patients may also suffer from delusions or depression. Key symptoms are:

- Problems with recent memory such as forgetting very recent events.
- Brief episodes of unexplained confusion and other behavioral or cognitive problems. The individual may become disoriented to the time or location where he or she is, have trouble with speech, have difficulty finding words or following a conversation, experience visuospatial difficulty (for example, finding one's way), and have problems in thinking such as inattention, mental inflexibility, indecisiveness, lack of judgment, and loss of insight.
- 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 mute and unable to speak the next day. There are also fluctuations in attention, alertness, and wakefulness.
- Well-defined, vivid, recurrent visual hallucinations. These hallucinations are well formed and detailed. In DLB's early stage, the person may even acknowledge and describe the hallucinations. They are generally benign, and patients are not scared by them. 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 symptoms often seem to start spontaneously and may include flexed posture, shuffling gait, muscle jerks or twitches, reduced arm swing, loss of dexterity. limb stiffness, a tendency to fall, balance problems, bradykinesia (slowness of movement), tremor, shakiness, 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. About 50% of patients have this symptom.

Movement and motor problems occur in later stages for 70% of persons with DLB. But for 30% of DLB patients and more commonly those that are older, Parkinson's symptoms occur first,

before dementia symptoms. In these individuals, cognitive decline tends to start with depression or mild forgetfulness.

Testing and Diagnosis

Dementia with Lewy Bodies is difficult to diagnose. Not only does it resemble other dementias, it overlaps with Alzheimer's, Parkinson's and other disorders, which may result in it being difficult to rule out or exclude. Because no single test exists to diagnose DLB, a variety of medical, neurological, and neuropsychological tests are used to pinpoint it and its possible overlap with other illnesses. A definitive diagnosis can only be made by an autopsy at death. There are no medications currently approved to specifically treat DLB.

Although Lewy bodies are found in brains of patients with other diseases and because testing will involve several approaches, 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 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, especially in an area that also degenerates in Parkinson's disease, the substantia nigra located in the brainstem. 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 memory formation and learning, attention, and psychomotor (muscular movement) skills.
- Lesions called Lewy neurites that affect nerve cell function are found in DLB brains especially in the hippocampus, an area of the brain essential for forming new memories.

None of the symptoms of Dementia with Lewy Bodies is specific only to DLB. To address this problem, an international group of researchers and clinicians developed a set of diagnostic criteria in 1995 called the *Consensus Guidelines* that can reliably point to 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 will occur as DLB progresses. Attention, language, understanding and reasoning, ability to do arithmetic, logical thinking, and perceptions of space and time will be impaired.

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

- Fluctuating cognition and mental problems vary during the day especially attention and alertness.
- Visual hallucinations, detailed and well-formed visions occur and recur.
- Parkinsonism: motor related and movement problems appear.

A DLB diagnosis is even more likely if the patient also experiences repeated falls, fainting, brief loss of consciousness, delusions, or is sensitive to neuroleptic drugs that are given to control hallucinations and other psychiatric symptoms.

Finally, the timing of symptoms is a reliable clue*: if both mental and motor symptoms appear within one year of each other, DLB is more likely the cause.* Signs of stroke or vascular dementia usually negate the likelihood of DLB.

Testing is usually done to rule out other possible causes of dementia. 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. Scans using SPECT and PET technology have shown promise in detecting differences between DLB and Alzheimer's disease.

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:

- Memory and other cognitive problems occur in both DLB and Alzheimer's. However, in DLB, they fluctuate frequently.
- DLB patients experience more depression than do Alzheimer's patients.
- Hallucinations are experienced by Alzheimer's patients in late stages and by Parkinson's patients who take medications to improve movement and tremor. In DLB, hallucinations occur in early stages, 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 Parkinson's in some DLB patients.
- Life expectancy is slightly shorter for DLB than for Alzheimer's patients.
- At autopsy, the brains of DLB patients have senile plaques, a hallmark of Alzheimer's. Another Alzheimer's feature, neurofibrillary tangles, are absent or found in fewer numbers and

are concentrated in the neocortex. Other Alzheimer's features-- regional neuronal loss, spongiform change and synapse loss, neurochemical abnormalities, and neurotransmitter deficits-- are also seen. However, DLB-afflicted brains are less damaged than are Alzheimer's brains.

- In DLB, movement problems are spontaneous; the symptoms begin suddenly.
- Tremor is less pronounced in DLB than in Parkinson's. Also, DLB patients respond less dramatically to drugs such as Levodopa that are used to treat Parkinson's. Nerve cell loss in the subtantia nigra is not as severe in DLB.
- Both DLB and Parkinson's patients may sometimes experience fainting and wide alterations in blood pressure.
- Some Parkinson's patients develop dementia in later stages. Dementia is usually the presenting symptom in DLB.
- Parkinson's patients lose the neurotransmitter dopamine; Alzheimer's patients lose the neurotransmitter acetylcholine. DLB patients lose both.
- In DLB, Alzheimer-like and Parkinson-like symptoms appear within one year of each other.

Despite these differences, a diagnosis of Dementia with Lewy Bodies does not preclude a positive diagnosis of Alzheimer's, Parkinson's or other diseases common in older age.

Duration and Treatment

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

Caution must be used in treating a person with DLB. Medications must be monitored closely for proper balance because some patients are adversely affected by some drugs. Neuroleptic (tranquilizing) anti-psychotic drugs such as haloperidol (Haldol) or thioridazine (Mellaril) as well as benzodiazepines (Valium, Ativan) and anti-histamines can cause extreme adverse reactions in DLB patients. Side effects include motor related symptoms, catatonia (non-responsiveness), loss of cognitive function, and/or development of muscle rigidity. These medications are sometimes used in Alzheimer's patients to help with hallucinations and behavioral symptoms but should not be used in patients with DLB. Levodopa may be given to treat the parkinsonism, however, it may increase the hallucinations of DLB patients and aggravate other symptoms such as cognitive functioning. It is less effective in treating tremor in DLB patients than in Parkinson's patients. Aricept or other cholinesterase inhibitors are given to treat the hallucinations. Some anti-depressants have shown positive results while others are counter- indicated.

When considering surgery, families should meet with the anesthesiologist to discuss possible side effects of

anesthesia as DLB patients are prone to delusions and a decline in motor functioning after anesthesia.

Caregiving and DLB

DLB patients can live at home with frequent reassessment and careful monitoring and supervision. Caregivers must watch the patient closely because of the tendency for them to fall or lose consciousness. Particular care should be taken when a patient is standing up from a chair or getting out of bed as blood pressure can drop causing the patient to lose his or her balance. Dementia prevents patients from learning new actions that might help them overcome movement problems such as learning to use a walker. They may need more assistance some days than others and can be reassured by a caregiver's help in turning attention away from hallucinations.

Caregivers must learn skills in dealing with cognitive, behavioral, and motor disabilities. 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.

Caregivers can turn to a California Caregiver Resource Center for assistance and to a qualified diagnostic center for initial diagnosis and follow-up. In other states, resources can be found through local and state offices on aging and health such as your Area Agency on Aging or the Alzheimer's Association in your area.

Credits and References

Lewy Body Dementia Association. www.lbda.org

Riding the Roller Coaster with Lewy Body Dementia by Helen Whitworth.

LewyNet, The University of Nottingham, Division of Pathology, University Park, Nottingham, England NG7 2RD. Telephone +44 115 9515151.

Web site: http://www.nottingham.ac.uk/pathology/lewy/lewyinfo.html

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Resources

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

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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
http://www.lbda.org/

Reviewed by William Jagust, MD and prepared by Family Caregiver Alliance. February 2001. Updated June, 2010. Funded by the Alameda County Area Agency on Aging and the California Department of Mental Health.

Rev. 4/2017