

Dementia with Lewy bodies For RIAPDA website for patients.  
Lewy body dementia

There is a lot of confusion about the disorder called either dementia with Lewy bodies or Lewy body dementia. This was first described in 1961 by Dr. Stanley Aronson, the founding dean of the medical school at Brown University, eminent neuropathologist and humanitarian, who also wrote a weekly column for the Pro Jo for over 20 years. When the first two cases were described, it was thought to be an extraordinarily rare condition, but with advancements in neuroscience, we have learned that it is not uncommon.

Firstly, what is it? And secondly, why did it become more common? The second question is easier to answer. For pathologists to look at tissue, it must be stained. Different stains color different organelles or chemicals within each cell. With the stains available in 1961 the Lewy body, which is one of the defining criteria for Parkinson's disease, could only be seen with one particular stain. The Lewy body, named after its discoverer, is a microscopic ball of protein that is found in certain brain cells in people with Parkinson's disease. These cells are in the "substantia nigra," which is in the upper front part of the brainstem, a structure that connects the bulk of the brain with the spinal cord. In DLB, these Lewy bodies were found in the upper part of the brain called the cortex, or outer lining, where they had only rarely been seen before. And in the two cases described in 1961, the abundance of the Lewy bodies, and their association with dementia, was unique. Over the last 25 years or so newer staining techniques have made it clear that Lewy bodies are much more commonly present than had been thought, and as more pathologists spent more time looking for them, with better stains, their true prevalence was finally noted.

What is DLB? It is a form of dementia, that is, a disorder in which memory and thinking abilities decline, more than can be accounted for by age alone. It was initially called Lewy body dementia, the name still embraced by the national support organization, but then altered to the "Lewy body variant of Alzheimer's disease," and then to DLB. The Alzheimer term is no longer used as it has become clear that this is not a variant of Alzheimer's disease. DLB is a disorder that causes memory and cognitive decline, usually associated with visual hallucinations and fluctuations during the day in level of memory and cognitive function, alertness and attention. Many patients have REM sleep disorder, a syndrome in which people act out dreams (REM, or Rapid Eye Movement, sleep), usually dreams of fighting, running, kicking, jumping, etc. Normal people are paralyzed when they dream. And almost all go on to develop all the motor features of Parkinson's disease. In many cases there is no dream enactment behavior (REM sleep behavior), or hallucinations, so that the overlap with Alzheimer's disease is so strong, that experienced clinicians cannot distinguish them. That is one problem that causes confusion, even though brain autopsies, however, show two distinctly different disorders.

The second confusing issue is that unfortunately, many people with PD develop dementia, and this looks exactly like the dementia in DLB, including the sleep disorder, hallucinations, clinical response to medications, and even the pathology seen at autopsy. So clinical researchers decided to set guidelines to distinguish DLB and PD with dementia and concluded with the "one year rule." By their consensus definition, if dementia preceded the development of PD motor problems (slowness, rigidity, tremor, posture and walking problems)

or if the dementia developed within one year of the motor features, the disorder should be called DLB (or LBD) but not Parkinson's disease.

Unfortunately, most doctors are unaware of these fine points and will simply tell families that the patient has DLB, not Parkinson's disease, even when the dementia began many years into the course. This is often confusing to patients and families, which is why I've written this essay.

Experts in the field debate whether DLB and PD with dementia are one disorder or two separate ones. My opinion is that they are different until they become the same, by which I mean that patients who have no signs of Parkinson's disease, no tremor, no slowness, no stopped posture, no stiffness, no masked facial expression, but have memory and thinking problems, act out their violent dreams, have visual hallucinations, have DLB, and will, with time, develop all the features of PD. At the time they meet criteria for a diagnosis of PD, I believe they should then be classified as have PD with dementia. I therefore believe that DLB is a variant of PD, but one in which the cognitive features occur first, whereas PD with dementia is a disorder in which the motor features develop first. When dementia and parkinsonism are both present, I believe the correct term is PD with dementia.

Does this argument matter to the patient? I think it does because the medical community currently studies drugs in one population or the other, so that some drugs are approved for PD with dementia but not DLB. Changing the definition will reduce the number and cost of studies to treat these disorders.