



DIFFUSE LEWY BODY DISEASE

Diffuse Lewy Body Disease (DLBD) is a degenerative disorder of the brain that leads to loss of memory and difficulty concentrating (dementia). After Alzheimer's disease, it is the second most common cause of degenerative dementia. In the early stages of the disease there is often simply a problem with attention, e.g. sustaining a line of thought. Memory loss does not necessarily occur in the early stages, but usually appears as the disease progresses. Patients with DLBD are often anxious and may be depressed. These may even be the first symptoms of the disease.

Other features of Diffuse Lewy Body Disease are:

- 177 **Fluctuations in thinking.** Especially early in the disease, patients may be very confused at some times and be able to think clearly at others.
- 177 **Hallucinations.** These are most often well formed and detailed visual hallucinations; the patient will see animals or people that are not really there.
- 177 **Symptoms of Parkinson's Disease,** e.g. tremor, stiffness, slowness of movement. DLBD may first present itself with these symptoms. It is therefore not unusual that the initial diagnosis is Parkinson's Disease and later, when dementia develops, is changed to DLBD.

Most people have never heard of Diffuse Lewy Body Disease. It was only in the 1980s that DLBD was first described. Pathologists looking at post mortem brains of some people who had dementia before they died noticed that some of the brains didn't have the classical features of Alzheimer's disease (plaques and tangles) that they expected to see. These patients were then found to have had different features of dementia according to their clinic records, so researchers realized that these patients had something different, while they were alive. What the pathologists found were Lewy bodies. This new disease was called " Diffuse Lewy Body Disease".

What is a "Lewy body"?

Lewy bodies are named after Dr. Friederich Heinrich Lewy who first described these structures that he discovered in brains of patients who had died with Parkinson's disease. In Parkinson's disease patients, these structures are found in brain cells in the brain stem (at the base of the brain), which are involved in the control of movement. In DLBD, Lewy bodies are also found in the outer layer of the brain called the cortex, which is responsible for mental function. These "cortical Lewy bodies" are the essential pathological feature of Diffuse Lewy Body Disease.

What causes Diffuse Lewy Body Disease?

We don't know the cause of DLBD. Genetic factors may play a role in some cases, but there is no clear pattern of inheritance. There is evidence that suggests that DLBD is caused by the same mechanism that causes Parkinson's disease. Unfortunately, we still don't know the cause of most cases of Parkinson's disease, nor do we know why some people develop Parkinson's disease and others Diffuse Lewy Body Disease.

How is the diagnosis of Diffuse Lewy Body Disease made?

The only way to be certain about the diagnosis is to examine the brain after death. In life the diagnosis of DLBD is made based on a patient's symptoms. There are no tests such as a blood test or brain scan that can provide a certain diagnosis of DLBD. A doctor will often order some blood tests, or a brain scan, to exclude other illnesses that may cause similar symptoms. The following symptoms are used to make a clinical diagnosis:

- 177 **Dementia** – must develop during the course of the disease – specific difficulties with *inattention, visuospatial skills* and *lack of initiative*.
- 177 Two of the following must be present:
 - **Fluctuations in memory**
 - **Visual hallucinations**
 - **Parkinsonism**

Features that further support the diagnosis are:

- 177 **Difficulty with balance** - may lead to repeated falls
- 177 **Transient loss of consciousness**
- 177 **Inability to tolerate neuroleptic medications**

Many different names are given for different subtypes of Diffuse Lewy Body Disease. They include: dementia with Lewy bodies, Lewy body variant of AD, and Lewy body disease. These subtypes are all variants of the same disease. Physicians may use these names interchangeably. The precise subtype does not matter in clinical practice, as treatment is the same.

How is Diffuse Lewy Body Disease different from AD?

Although dementia is present in both AD and DLBD, the features of dementia differ between the two. It's unusual to have visual hallucinations in Alzheimer's disease and fluctuations in intellectual functioning are more prominent with Diffuse Lewy Body Disease. Visuospatial skills and attention are more affected in DLBD, whereas language function is more affected in AD. Patients have problems with memory in both DLBD and Alzheimer's disease, but they are different. In AD, the patients have difficulty consolidating memory (forming new memories) whereas in DLBD patients have difficulty retrieving memory (accessing previously memorized information). These differences stem from pathological differences between the two disorders. In AD there is wide spread cell loss in the cortex producing degeneration throughout the brain. If you look at a MRI or CT scan of a patient with Alzheimer's disease the brain appears to have shrunk in size compared to a normal brain. In contrast a MRI scan of a patient with DLBD looks normal. This is because a MRI shows structural abnormalities and in Diffuse Lewy Body Disease the problem is chemical. It is possible to show the difference between brains with DLBD and normal brains using a PET scanner. The remarkable feature of PET scanning is its ability to show levels of specific chemicals in the brain.

How is Diffuse Lewy Body Disease different from Parkinson's disease?

In Parkinson's disease, dementia occurs in about 30% of patients and usually late in the disease. Memory problems in Parkinson's disease are often quite mild. Since some patients with Parkinson's disease have dementia and some patients with DLBD have Parkinsonism, there is considerable overlap between the symptoms of the two diseases. Do all Parkinson's disease patients who have dementia have DLBD? No (not according to our current criteria). A patient with Parkinson's disease can for instance also develop AD, in which the pattern of dementia is different as described earlier.

Is there a Cure for Diffuse Lewy Body Disease?

Unfortunately, there is no cure for DLBD. There are, however, some drugs that can help some symptoms of the disease. Patients with parkinsonian symptoms may be helped by some of the drugs used to treat Parkinson's disease. There are also new medications that seem particularly helpful for the thinking problems of Diffuse Lewy body disease.

Source: Pacific Parkinson's Research Centre, University of British Columbia, Vancouver, B.C.

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