

# DIFFUSE LEWY BODY DISEASE (DLBD)

Diffuse Lewy body disease (DLBD) is a common neurodegenerative disorder that produces dementia, psychosis and abnormalities of movement. Clinicians began to recognize diffuse Lewy body disease in the early 1990's although this disorder has been present in the population as long as other types of dementia. In fact, this disorder may produce 10-20% of all dementia in the United States, either alone or with other dementias like Alzheimer's disease. Clinicians have limited ability to recognize this disorder, as the symptoms resemble Alzheimer's disease.

The clinical manifestations of diffuse Lewy body disease include dementia, visual hallucinations, and a fluctuating clinical course. The dementia associated with diffuse Lewy body disease can resemble Alzheimer's disease and neuropsychological testing cannot distinguish these disorders with absolute accuracy. Visual hallucinations are a common early symptom in diffuse Lewy body disease. Many patients demonstrate a fluctuating clinical course from day to day in which the level of intellectual function and the severity of hallucinations may vary from mild to severe.

Patients with DLBD frequently manifest symptoms of Parkinson's disease including stiffness, slow shuffling gait, and tremor. The symptoms of Parkinson's disease may precede the symptoms of dementia or psychosis. Patients with DLBD are often sensitive to the side-effects of neuroleptic medications, e.g., Haldol, Prolixin, etc. A psychotic patient with DLBD may receive a small dose of antipsychotic medication, e.g., Haldol, to lessen visual hallucinations, and develop severe drug-induced Parkinsonism.

The progression of symptoms of DLBD resembles that of Alzheimer's disease with a slow insidious onset followed by a gradual, relentless decline in function. The duration of survival is similar to Alzheimer's disease.

The general physical examination in patients with DLBD is unremarkable. The neurological examination may demonstrate evidence of Parkinsonism but no focal neurological deficits. Laboratory studies for DLBD are unremarkable. Brain

imaging studies may demonstrate mild to moderate atrophy or an otherwise normal brain. SPECT studies are helpful to identify DLBD and usually lack the distinctive findings of decreased flow to the left parietal temporal region that is present in Alzheimer's disease.

The cause of DLBD is unknown. Genetics seem to play a minor role in the development of this disorder. Other risk factors associated with Alzheimer's disease, e.g., low educational achievement, post-menopausal estrogen deficiency, etc., do not appear to apply to DLBD. Drinking is not associated with this disorder.

Treatment is not available for DLBD; however, specific symptoms produced by the brain damage will respond to appropriate medications. Patients with symptoms of Parkinsonism will respond to appropriate doses of anti-parkinsonian medications, e.g., Sinemet. Visual hallucinations will respond to antipsychotic medications; however, clinicians must avoid producing more Parkinsonism. Atypical antipsychotic medications such as Seroquel (quetiapine) or Zyprexa (olanzapine) are effective in lessening visual hallucinations. Low-potency neuroleptics such as Mellaril are also effective. Many patients develop depression that is improved with standard antidepressant therapy such as serotonin reuptake inhibitors. Families should be referred to Alzheimer's support groups, as social and family problems are identical in both diseases.

Anti-Alzheimer's medications such as Aricept are probably not effective in DLBD; however, patients with mild to moderate impairment may warrant a trial with the medication because the patient may also have combined Alzheimer's disease. Preventive interventions for Alzheimer's disease such as vitamin E, selegiline, and ginkgo, are not shown to be beneficial to patients with DLBD.

The brain pathology of DLBD is markedly different from other types of dementia. Microscopic brain finding of DLBD are very subtle and easily overlooked by general pathologists. Special stains must be performed to identify the characteristic circular inclusion present in the neurons of the temporal lobe, frontal lobe, and brain stem. These circular inclusions are called Lewy bodies and these structures resemble those present in Parkinson's disease. Many patients with DLBD also have the brain pathology of Parkinsonism.

Significant numbers of patients with Alzheimer's disease also have Lewy bodies in their brain. These mixed dementias are impossible to identify in the living patient and the interaction between DLBD and SDAT is unclear.



# **FACT SHEET ON DIFFUSE LEWY BODY DISEASE**

- 1. DLBD is the second or third most common cause of dementia.**
- 2. Intellectual loss in DLBD resembles Alzheimer's disease.**
- 3. Most patients have visual hallucinations.**
- 4. Confusion and hallucinations often vary on a day-to-day basis.**
- 5. Many patients develop parkinsonism with stiffness, slowness and tremor.**
- 6. Many patients develop depression.**
- 7. DLBD is not inherited.**
- 8. There is no prevention for DLBD.**
- 9. DLBD patients are very sensitive to antipsychotic medications, e.g., Haldol.**
- 10. Diagnosis requires brain autopsy examination.**